Clinical Cardiology Update 2010

Course Book

Contents

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FOREWORD

It gives us great pride in organizing the Clinical Cardiology Update 2010 at the Madras Medical Mission. The Madras Medical Mission has been in the forefront of teaching, training and therapeutics in cardiology over the last 25 years. This Cardiology Update has been an effort by the cardiology group at the Madras Medical Mission to provide candidates in training in cardiology an opportunity to interact with best of teachers and updating their skills in clinical cardiology.

I thank the entire faculty and wish all delegates a useful and interactive update. This would be an annual programme at the Madras Medical Mission.

Dr. Mullasari Ajit S.
Course Director

Preface

Cardiology is the fastest developing superspeciality in medicine due to introduction of high gadget investigative tools along with advanced interventional and surgical treatment. However clinical cardiology is losing its charm and importance in the examination of a cardiac patient.

To improve clinical skills in cardiology, we at the Madras Medical Mission decided to hold a Clinical Cardiology Update for the benefit of cardiology postgraduates across the country. We are also coming out with a course book, which will primarily focus on improving clinical cardiology skills and strengthening your preparation for the clinical cardiology examinations.

I am thankful to my Director of Cardiology at the Madras Medial Mission, Dr.Ajit Mullasari for giving me the responsibility of organizing this conference. I am also thankful to Dr. R. Suresh Kumar, Head of Pediatric Cardiology Department for his guidance and support.

I will be failing in my responsibilities if I do not thank my colleagues, Dr.Latchumanadhas and Dr. Ulhas Pandurangi and all other faculty members, residents and secretaries for their immense support.

I appreciate the enthusiasm and cooperation of all the postgraduates who attended and made the 2009 Update a grand success. We are confident that at the end of the update you will be more confident than before to face the challenge of clinical cardiology examination. We also welcome suggestions from you to improve the course book and the update sessions for the future.

Dr. J. Ezhilan
Organising Secretary

Acknowledgements:

We thank all Registrars and Ms. Sujatha K.
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GENERAL EXAMINATION OF CARDIOVASCULAR SYSTEM

Dr. J. Ezhilan

Physical signs are objective indications of disease whose significance is enhanced when they confirm a disease already suggested by the patients history. At times the physical sign may be the only evidence for the disease. The physical examination should be performed methodically and thoroughly also taking into consideration the patients comfort and modesty. The examination must extend from head to foot in an objective search for abnormalities. Skill in physical diagnosis is acquired with experience and knowledge, for the eye sees only what the brain knows. A good history taking, detailed symptom analysis and thorough general examination is half way through to the diagnosis of a cardiovascular disorder. However a meticulous general examination in a cardiovascular case is inadequately utilized. A common pitfall in cardiovascular medicine is the failure by the cardiologist to recognize that a patient's heart disease is part of a systemic illness and failure of the noncardiologist to recognize the presence of a cardiac disorder accompanying the systemic illness. Hence it is imperative to do detailed general examination of a cardiovascular patient

Cardiovascular Disorders With Findings In General Examination

Cardiovascular disorders with findings in general examination can be broadly classified as follows

1) Chromosomal abnormalities
2) Heritable conditions
3) Connective tissue disorders
4) Inborn errors of metabolism
5) Sporadic disorders
6) Teratogenic disorders
7) Endocrine disorders

1) Chromosomal Abnormalities

Chromosomal abnormalities and mutation of single genes accounts for less than 10% of cardiac malformations. The common chromosomal abnormalities are

a) Trisomy (Additional chromosome)
b) Monosomy (Missing chromosome)
c) Triploidy (1 additional set of chromosome)
d) Tetraploidy (2 additional set of chromosomes).

Of the above abnormalities Trisomy is the commonest abnormality. Trisomy occurs for all chromosomes but only trisomy 13, 18, 21 and sex chromosome are compatible with survival to term.

GE	CVS
Down's Syndrome (Triosomy 21)	
Flat facial profile	Single atrium
Oblique palpaberal fissure	Endocardial cushion defect
Epicanthus fold	
Abundant neck skin	

Protruding tongue
Low set ears
Poorly formed nasal bridge
Hypoplastic mandible

Edward's Syndrome (Triosomy 18)

Micrognathia
Rocker bottom feet

VSD
PDA
Polyvalvular dysplasia

Patau's Syndrome (Triosomy 13)

Microcephaly
Microophthalmia
Cleft lip
Mid facial defects

VSD
PDA
DORV

Cri Du Chat Syndromme (5P-)

Microcephaly
Antimongoloid slant
Cats cry

VSD

Turners Syndrome

Short stature
Shield chest
Web neck
Cubitus valgus
Lymphoedema of hand & feet
Micrognathia
Epicanthal fold
Low set or deformed ears
Ptosis
Fish like mouth

Coarctation of aorta
Bicuspid aortic valve

2) Heritable Conditions

Mutations of single gene leads to the following conditions

Ellis Van Crevald

Chondrodystrophic dwarfisim
Nail dysplasia
Polydactyly

Single atrium
ASD (OP)

Tar Syndrome

Thrombocytopenia
Abscent radius

ASD
TOF

Holt Oram Syndrome (12q21-Q3)

Skeletal upper limb defects
Hypoplasia of clavicle

ASD

Kartageners Syndrome

Situs inversus

Dextrocardia

Sinusitis
Bronchiectasis

Laurence Moon Biedel Bardet

Moon face	Variable defect
Obesity	
Polydactyly	
Mental retardation	
Retinitis pigmentosa	
Renal abnormalities	
Hypogonadotropic hypogonadisim	
Diabetes mellitus	

Ullrich Noonan Syndrome (12q24) Male Turners

Short stature	Pulmonary stenosis
Shield chest	HOCM
Web neck	ASD
Cubitus valgus	
Hypogonadisim	
Scoliosis	
Renal abnormalities	

Tuberous Sclerosis / Bournevilles Disease / Epiloa

Type-I – 4Q / Type II 16P	
Mental retardation	Rhabdomyoma
Seizure disorder	Cardiomyopathies
Adenoma sebaceum	
Shagreen patches	
Leaf shaped hypopigmented macules	
Subungual fibroma	
Angioleiomyoma	
Brain tumors	

Lepoard Syndrome (Multiple Lentigenes)

L- Lentigines	Pulmonary stenosis
E- ECG abnormalities (Cond defects)	Subaortic stenosis
O- Ocular hypertelorism	PPH
P- Pulmonary stenosis	
A- Abnormal genitials (Cryptorchadism/Hypospadiasis)	
R- Retardation of growth	
D- deafness (Sensorineural)	

Rubenstein-Taybi Syndrome (16p13.3)

Slanted palpaberal fissure	PDA
Hypoplastic maxilla	
Broad thumbs and toes	

Familial Deafness

Sensorineural deafness	Arrhythmias
	SCD

Osler Rendu Weber Syndrome (9q33)

Telengiactisis of face & mucosa	Pul A-V Fistula
Haemoptysis	

Aperts Syndrome (10q26)

Craniosynostosis	VSD
Midfacial hypoplasia	
Syndactyly	
Brachycephaly/Acrocephaly	
Proptosis	
Beaked nose	
Antimongoloid slant	
Cleft palate	

Crouzon (10q26,4p16.3)

Craniosynostosis	PDA
Ptosis	Coarctation of aorta
Shallow orbits	
Maxillary hypoplasia	

Incontinentia Pigmenti

Mental retardation	PDA
Seizures	
Strabismus	
Cataract	
Patchy alopecia	
Hypodentia	
Hyper/Hypopigmentation	
Spastic paraplegia	

Alagille (20p12) Arteriohepatic Dysplasia

Prominent forehead	Peripheral Pulmonic stenosis
Deep set eyes	Pulmonary stenosis
Mongoloid slant	
Prominent nasal bridge	
Small pointed chin	
Biliary hypoplasia	
Vertebral anomalies	

Di George (22q11) CATCH-22

Hypoplastic mandible	TOF
Defective ears	Truncus arteriosus
Short philtrum	Interrrupted Ao arch(TypeB)
Thymic hypoplasia	
Parathyroid hypoplasia	

Sphrintzen (22q11) Velocardiofacial Syndrome

Long vertical face	VSD
Prominent face	TOF
Cleft palate	Right Aortic arch

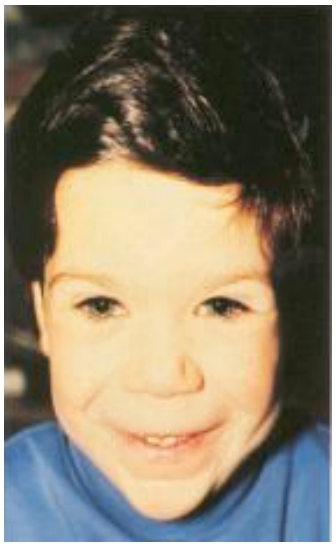
Retruded mandible
Slender hands
Learning disability

Williams Syndrome (7q11.23)

Broad forhead
Hypertelorism
Medial eyebrow flare
Internal squint
Stellate pattern of iris
Epicanthal fold
Low set ears
Upturned nose/Long philtrum
Baggy cheeks
Large upper lip with pouting
Hypoplastic mandible
Malformed teeth/Malocclusion
Hypercalcemia

Supravalvular aortic stenosis
Pulmonary artery stenosis

Williams Syndrome
Elfil facies



Long QT Syndrome (11p15.5,7q35,3p21,21q22)

Jervell & Lange-Nielsen / Romano Ward
Congenital deafness
Sudden Cardiac Death

Long QT
Ventricular arrhythmias

Progeria (Werners Syndrome)

Premature ageing
Alopecia
Atrophy of subcutaneous fat
Skeletal hypoplasia

Accelerated atherosclerosis

Conotrunkal Facies

Ocular hypertelorism

TOF

Narrow eye fissures	DORV
Bloated eye lids	Truncus arteriosus
Small mouth	TGV
Deformed ear lobes	

3) Connective Tissue Disorders

Cutix Laxa

Gen disruption of elastic fibers	Peripheral Pulmonic stensosis
Diminished skin resilience	
Hernias	

Ehlers Danlos(2q31)

Hyperelastic & Friable	Arterial dilation & rupture
Hyperextensible joints	Mitral regurgitation
Keratoconus	

Marfans (15q21.1)

Gracile habitus	Arterial dilation
Arachnodactyly	Mitral regurgitation
Pectus excavatum/carinatum	Aortic regurgitation
Kyphoscoliosis	
Higharched palate	
Hyperextensibility	
Lens dislocation	
Irridodonesis	
Myopia	
Retinal detachment	

Osteogenesis Imperfecta (4.17)

Fragile bones	Aortic regurgitation
Blue sclera	

Pseudoxanthoma Elasticum

Degeneration of elastic fibres in skin	Coronary artery disease
Bull dog appearance of mouth	peripheral artery disease
Fissured thickened yellow skin (Plucked chicken appearance)	
Retinal angiod streaks	

4) Inborn Errors of Metabolism

Pompe Disease

Acid Maltase deficiency	Glycogen storage disease of heart
Muscular weakness	

Homocystinuria

Cystathionine syntethase def	Aortic & PA dilation
Lens subluxation (Posterior)	Intravascular thrombosis
Osteoporosis	

Mucopolysaccharidosis

a) Hunter

Def of L-iduranosulfate	Multivalvular disease
Coarse facies	Great artery disease
Growth & mental retardation	Coronary artery disease
Clear cornea	Cardiomyopathy

b) Hurler

Def of alpha- L Iduronidase	Multivalvular disease
Corneal clouding	Great artery disease
Coarse facies	Coronary artery disease
Growth & mental retardation	Cardiomyopathy

c) Morquio

Def of N-acetyl hexosamine sulfate sulfatase	
Cloudy cornea	AR
Normal intelligence	
Bony changes involving vertebrae & epiphysis	

d) Scheie

Def of alpha L-Iduronidase	AR
Cloudy cornea	
Normal intelligence	
Peculiar facies	

e) Maroteaux-Lamb

Def of arylsulfatase B	AR
Cloudy cornea	
Osseous changes	
Normal intelligence	

5) Sporadic Disorders

VATER Association

V - Vertebral anomalies	VSD
A - Anal atresia	
T- Tracheo Eesophageal fistula	

R-Radial & Renal anomalies

CHARGE Association

Colobomas	TOF
Choanal atresia	Other defects
Mental & growth deficiency	
Genital anomalies	
Ear anomalies	

Cornelia De Lange

Micromelia	VSD
Synophrys	
Mental & growth deficiency	

6) Teratogenic Disorders

Rubella

Microcephaly	ASD
Microopthalmia	PDA
Hypoplastic iris	Pulmonary valvular stenosis
Cataract	Pulmonary arterial stenosis
Deafness	
Thrombocytopenia	

Alcohol

Microcephaly	VSD
Growth & mental retardation	
Short palpebral fissures	
Smooth philtrum	
Thin upper lip	

Dilantin

Growth & Mental deficiency	Pulmonary stenosis
Hypertelorism	Aortic stenosis
Prominent epicanthal fold	Coarctation of aorta
Hirsute forehead	PDA
Bowed upper lip	
Midfacial hypoplasia	
Depressed nasal bridge	
Microcephaly	
Cleft lip	

Thalidomide

Phocomelia	Variable

Lithium

None	Ebsteins anomaly
	Tricuspid atresia

General Examination Findings in Cardiovascular Cases

The general examination of a patient with cardiovascular disorder should be done in the following order not to miss any finding

1. Stature & Build
2. Eyes
3. Nose
4. Ears
5. Lips
6. Tooth
7. Neck
8. Skin
9. Extremeties
10. Chest & Abdomen

STATURE	EYES	NOSE	EARS	LIPS
Tall	Hypertelorism	Broad nose	Ear lobe crease	Thick lips
Short	Exopthalmos	Broad flat nose	Cauliflower ear	Absent philtrum
Obese	Nystagmus	Beaked nose	Low set ears	Cyanosis
	Ptosis		Deafness	Quinkes sign
	Xanthelesma		Preauricular sinus	Rhagades
	Conjuctival pallor			Petichae
	Conjuctival suffusion			Telengiactasia
	Subconjuctival haemorrage			
	Conjuctivitis			
	Jaundice			
	Blue sclera			
	Arcus juvenialis			
	Arcus senilis			
	Corneal clouding			
	Brushfield spots			
	Coloboma			
	Argyll Robertson pupil			
	Cataract			
	Subluxation of lens			
	Fundoscopy			

ORAL CAVITY	NECK	SKIN	LIMBS	CHEST & ABDOMEN
TEETH	Short neck	Bronze pigmentation	Cyanosis	Barrel chest
Delayed dentition	Webbed neck	Café au lait spots	Clubbing	Pectus excavatum
Premature dentition	Low hair line	Multiple lentigines	Pedal edema	Pectus carinatum
Pegshaped teeth		Adenoma sebaceum	Terry nails	Khyposcolisois
Malformed teeth		Erythema marginatum	Lindsay nails	Straight back syndrome
Widely spaced teeth		Janeway lesions	Subungal haemorraes	Harrisons sulci
GUMS		Hyperextensible rubber like skin	Koilonychia	Hypomastia
Gingival hypertrophy		Plucked chicken like appearance	Raynauds phenomenon	Gyanecomastia
TONGUE		Xanthomas	Arachonodactyly	Left Thorocatomy scar
Macroglossia		Subcutaneous nodules	Polydactyly	Right thoracotomy scar
Glosoptosis		Oslers nodes	Syndactyly	Shield chest
PALATE			Clindactyly	Abscent Pectoralis
Cleft Palate			Brachydactyly	Abdominal striae
High arched palate			Clenched hand	Abdominal obesity
			Fingerized thumb	Ascites
			Pes cavus	
			Phocomelia	
			Jaccouds arthritis	

Stature & Build

Before commenting about the build and stature of the patient the following measurements have to be taken

1. Height of the patient
2. Weight of the patient
3. Upper segment The upper segment of the body is the distance from the top of the head to pubic ramus
4. Lower segment The lower segment of the body is measured from the pubic ramus to floor
5. Arm Span
6. Arm span/Height ratio
7. Body mass index is derived by dividing the weight of the individual in Kgs by height in meters2
8. Hip circumference
9. Waist circumfrence
10. Hip Circumfrence/Waist circumfrence Ratio

Short Stature

When the height of the individual is < 2 standard deviations of the mean height for age and race he is considered to be short. Conditions where short stature is encountered with cardiovascular manifestations are

1. Cretinism
2. Pitutary dwarf
3. Cushings syndrome
4. Turners Syndrome
5. Noonans syndrome
6. Morquios Syndrome
7. Multiple lentigines syndrome
8. Ellis Van Crevald syndrome
9. Achondroplasia
10. Osteogenesis imperfecta

Tall Stature

When the height of the individual is more than 2 standard deviation of the meam height for his age he is considered to be tall. Conditions with tall stature and cardiovascular manifestations are

Upper segment=Lower segment

1)Constitutional

2)Pitutary

Upper segment<Lower segment

1)Marfans Syndrome

2)Homocystinuria

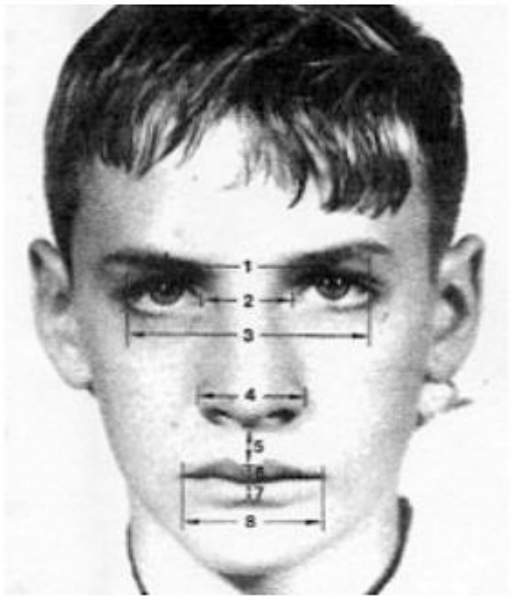
3)Klinfelter Syndrome

Obese

Obesity is generalised if BMI is >25 and it is abdominal if waist hip ratio is abnormal. It is seen in

1. Generalised obesity

2. Cushing Syndrome
3. Cretinism
4. Tickwickiam syndrome
5. Laurence Moon Biedel Syndrome
6. Prader Willi syndrome
7. Metabolic syndrome



1. Interpupillary distance
2. Innercanthal distance
3. Outercanthal distance
4. Inneralar distance
5. Philtral length
6. Upper lip thickness
7. Lower lip thickness
8. Lip length

CRANIOFACIAL MEASUREMENTS

Eyes

Hypertelorism

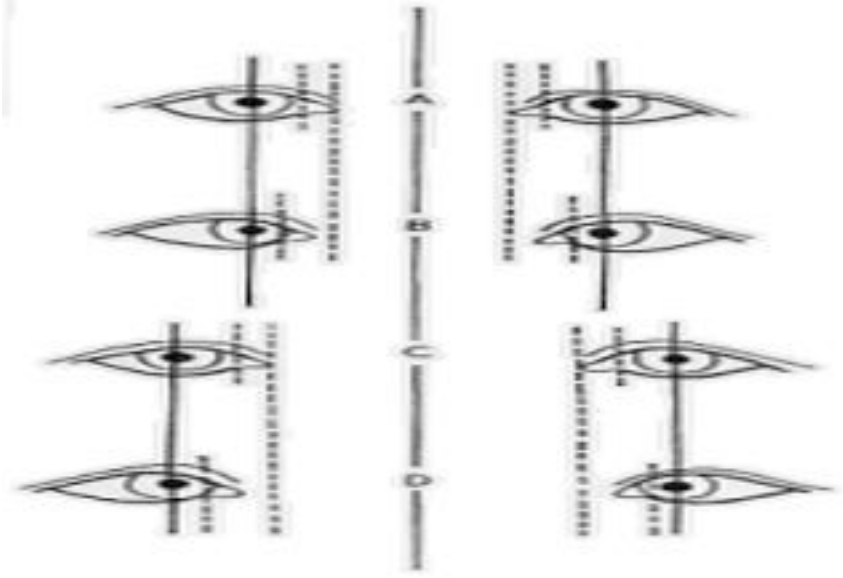
Hypertelorism is defined as increased space between the orbital cavities. The normal interorbital distance at birth is < 20 mm and in adults > 25 mm is considered abnormal. This can only be measured by radiology. At bedside Hypertelorism can be measured by measuring

a)Inner intercanthal distance
b)Outer intercanthal distance

If the inner intercanthal distance is .50% of outer intercanthal distance Hypertelorism is present.

Hypertelorism is seen in

1)Ullrich Noonans Syndrome	—	PS
2)Turners Syndrome	—	Coarctation of Aorta
3)LEOPARD Syndrome	—	PS/HCM
4)Hurlers Syndrome	—	Valvular regurgitation
5)Williams Syndrome	—	PS/Supravalvular AS
6)Klippel-Feil Syndrome	—	VSD



A. Normal Interocular distance
B. Primary telecanthus
C. Hypertelorism
D. Hypetelorism with secondary telecanthus

Mongoloid Slant

The eyes are normally horizontally placed but when it slants upwards and outwards it is called Mongoloid slant

1) Down's Syndrome
2) Alagille Syndrome

Antimongoloid Slant

When the eyes instead of being horizontal are slanted downwards and outwards it is called Antimongoloid slant

1) Cri du chat syndrome
2) Aperts syndrome

Epicanthus

Prominent medial folds of the eye are called epicanthus. This is normal in certain races. It is seen in:

1) Down's Syndrome
2) Turner's syndrome
3) William's syndrome
4) Endocardialcushion defect
5) Phenytoin teratogenic defect

Lid Retraction / Proptosis

Lid retraction causes a staring appearance and can occur in any form of thyrotoxicosis and is the result of sympathetic overactivity

Proptosis is visualization of the sclera between the lower border of the iris and lower eyelid with the

eye in primary position. It can be measured using an exopthalmometer. Proptosis if severe can cause corneal exposure and damage. This is seen only in Graves disease it is due to hypertrophy of extraocular muscles.

Ptosis

Ptosis is abnormal drooping of the eyelids and can be unilateral or bilateral. It can be due to congenital or acquired caused. Ptosis can be Mechanical ptosis, Aponeurotic ptosis, Myogenic Ptosis and Neurogenic ptosis

MYOGENIC PTOSIS

1)Kearns Sayre Syndrome - Complete Heart block
2)Myotonic dystrophy
3)Klippel-Feil Syndrome — VSD
4)Multiple Lentigenes Syndrome – PS / Sub aortic stenosis / PPH

Xanthalesma

Xanthalesma is yellowish deposition of cholosterol near the inner canthus in dyslipedemias.

Conjuctival Pallor

Conjuctival pallor is seen in anaemia

Conjuctival Suffusion

Conjuctival sufision is seen in Polycythemia

Subconjuctival Haemmorage

Subconjuctival hemorrhage is seen in infective endocarditis

Jaundice

Jaundice is yellowish discoloration of tissues resulting from deposition of bilurubin. Slight increase in bilurubin is best detected by examining the sclera which has a high affinity for bilurubin due to their elastin content. Scleral icterus indicates a serum bilurubin of 3.0 mg/dl.

Cardiovascular causes for jaundice are

1)Congestive cardiac failure
2)Pulmonary infarction
3)Cardiac cirrhosis
4)Prosthetic valves
5)Group A Beta Haemolytic infection

Blue Sc lera

Blue sclera is seen in Osteogenesis imperfecta which causes a generalized decrease in bone mass (Osteopenia) and makes the bones brittle. It is associated with blue sclera, dental abnormalities (dentino-genesis imperfecta) and a positive family history. The sclera in osteogenesis imperfecta can be normal, slightly blue or bright blue. The colour is probably caused by a thinness of the collagen layers of the sclera that allows the choroids layers to be seen. Blue sclera as an inherited trait can be seen in some families without bone fragility.

1)Familial
2)Osteogenesis imperfecta
3)Ehlers Danlos Syndrome Type VI

Arcus Senilis

Arcus senilis is a white or gray opaque ring in the corneal margin. Most commonly found in the elderly and hence the name. However if it is seen in individuals less than 50 due to dyslipedemiait is called as arcus juvenalis. It results from cholesterol deposition and hyalinosis in the deep layers of the corneal stroma. It is a deep often incomplete yellowish white ring with a sharp outer margin and a poorly defined inner margin.

It is seen in

1)Normal individuals
2)Dyslipedemia
3)Carotid artery disease (Unilateral arcus)

Corneal Clouding

Clouding of the cornea with cardiovascular manifestations occurs in

1. Mucopolysacchariodosis
a. Hurlers
b. Sheie
c. Morquio
d. Maroteaux-Lamb
e. Not seen in Hunters and Sanflippino
2. Scleroderma
3. Congenital rubella

Brushfield Spots

Thomas Brushfield a physician first described Brushfield spots. Brushfield spots are small white or grayish brown spots on the periphery of the iris in the eye due to aggregation of a normal iris element. They are focal areas of stromal hyperplasia surrounded by relative hypoplasia and are more common in patients with lightly pigmented iris. It is more common in Down's syndrome children of Caucassians race than Asian heritage.

1) Normal children
2) Down's Syndrome (35-78%)

Coloboma

Coloboma or cats eye is a fissure in the iris changing the papillary shape from circular to oblong, which is normal for a cats eye. It is seen in

1)Trisomy 22 (Cat eye syndrome) TAPVC / TOF / VSD
2)Tetrasomy 22
3)CHARGE Syndrome Conotruncal anomalies

Argyll Robertson Pupil

Argyll Robertson Pupil is small irregular unequal pupils with absent light reflex but spare accommodation and convergence. It is seen in

1)Neurosyphillis AR

Nystagmus

Nystagmus is rhythmic oscillation of the eye occurring physiologically from vestibular and optokinetic stimulation, or pathologically in a wide variety of diseases. There are four types of Nystagmus 1)Jerk Nystagmus 2)Gaze evoked nystagmus 3)Vestibular nystagmus and 4)Downbeat nystamus.

Freidrichs ataxia HCM

Cataract

Cataract is clouding of the lens sufficient to reduce vision. Cataract develops commonly after aging, ocular trauma, uvetis Glucocorticoid therapy, radiation therapy and diabetes mellitus. With cardiovascular manifestations it is seen in

1)Myotonic dystrophy	Cardiomyopathy
2)Neurofibromatosis Type II	Phaechromocytoma
3)Refsums disease	DCM/HCM
4)Rubella	Congenital Heart disease
5)Werners Syndrome	Premature atherosclerosis

Subluxation Of Lens

Subluxation of lens is dislocation of the liament from its normal position, which is held by suspensory ligaments. It can be displaced either forward or backward. Dislocation of the lens may be readily apparent but usually diagnosis needs pupillary dilation and slit lamp examination. The displacement is usually not progressive but may contribute to the formation of cataract. It is seen in

1)Marfans Syndrome (Anterior)	Aortic dilation/AR/MR
2)Homocystinuria (Posterior)	CAD/CVD/PVD/DVT

Fundus Findings

Fundus examination is mandatory in cardiovascular diseases. Some diseases with fundus findings are

FINDINGS	DISEASE
1)Hypertensive Retinopathy	Systemic Hypertension
2)Papilledema	Malignant Hypertension
3)Arterisclerotic retinopathy	Atherosclerosis
4)Wreath like AV anastomosis	Takayasus arteritis
5)Corkscrew retinal arteries	Coarctation of aorta
6)Roths spots	Infective endocarditis
7)Angiod streaks	Pseudoxanthoma elasticum

Nose

Broad Nose

Broad nose is seen in Acromegaly

Broad Flat Nose

Broad flat nose is part of facial dysmophosis and is seen in

1) Down's Syndrome
2) William's Syndrome
3) Hurlers
4) Cornelia-de-lange

Beaked Nose

Thin beaked nose is seen in

1)Werners syndrome
2) Rubenstein Taybi syndrome

Ears

Ear Lobe Crease (Frank Sign)

A horizontal crease in the ear lobule may be seen in some individuals unilaterally or bilaterally. It was though that people with ear lobe crease had

1)Premature Coronary artery disease
2)Diabetes mellitus
However it is now seen that the association with CAD is dwindling.

Floppy Ear (Cauliflower Ear)

1) Polychondritis
   Saddle nose — Pericarditis
   Arthritis — AR/TR

Microtia

Microtia which is deformed atretic ear is seen in:
1. Oculo-vertebral dysplesia
2. HMC syndrome

Preauricular Sinus/ Preauricular Appendage

Pre auricular sinus and preauricular appendage are both sen in Oculoauriculo vertebral dysplasia.

Oral Cavity

Lips

Thick lips

Thick lips which have diameter more than the normal are seen in:
1) Hurlers
2) Acromegaly
3) Myxedema
4) Cretinism

Thin Lips

Thin upper lip where the diameter of the upper lip is less than normal is seen in
1) Prader Willi syndrome
2) Fetal alcohol syndrome

Absent Philtrum

Absent philtrum is the absence of the vertical furrow seen above the centre of the lips extending to the nose. It is seen in
1)Prader Willi Syndrome
2) Fetal alcohol syndrome

Rhagades

Rhagades are ulcerative fissures at the angle of the mouth observed in congenital syphilis and has to be differentiated from angulostomatitis

Capillary Pulsation

Capillary pulsations can be seen by pressing a slide on the mucous membrane of the lips in severe Aortic regurgitation

Teeth

Premature Dentition

Ellisvan crevald syndrome

Delayed Dention

1) Cretinism
2) Down's syndrome

Malformed Teeth

Non hereditary form of Supravalvular AS with PS

Widely Spaced Teeth

1) Acromegaly
2) Morquios syndrome
3) William's syndrome

Peg Shaped Teeth

1) Hurler's syndrome
2) William's syndrome
3) Congenital syphilis

GUMS

Gingival Hypertrophy

Gingival hyperplasia of hypertrophy is seen in

1) Idiopathic familial gingival fibromatosis
2) Cyclosporin
3) Phenytoin
3) Ca Channel blockers
4) Ellisvan Crevald syndrome

Tongue

Macroglossia

Macroglossia or enlarged tongue is seen in

1) Downs syndrome
2) Acromegaly
3) Cretinism
4) Amylydosis
5) Hurlers syndrome

Glosoptosis

Glosoptosis or retracted tongue occurs in
Pierre Robin syndrome VSD

Palate

Cleft Palate

Cleft palate is cleft in the floor of the mouth and is seen in

1) tertiary syphilis AR
2) Velocardiofacial syndrome

High Arched Palate

High arched palate is the roof of the mouth being higher than normal. In congenital cause for high arched palate it is due to persistence of a certain stage of fetal development. It results in a narrow maxilla in the upper jaw leading to crowding of the teeth. Severe high arched palate with marked

dental crowding is also called as Cathedral palate. There are 177 medical conditions with high arched palate. Some of the conditions with cardiovascular involvement are

1) Normal variant
2) Marfans Syndrome
3) Turners syndrome
4) Noonans syndrome
5) Pierre Robin Syndrome
6) Pseudoxanthoma elasticum
7) Freidrichs ataxia.

Neck

Short Neck

Short neck is due to failure of normal segmentation of cervical somites during 3-8th week of gestation. It is associated with decreased flexion and extension of neck inability to rotate the neck and low hair line.

The ratio of height to the distance beween external occipital protuberance and C7 spinous process (Birds index)< 12.8 is normal, while >13.6 indicates short neck. It is seen in

1)Klippel-Feil syndrome
2)Morquios syndrome

Webbed Neck

Webbing of the neck is seen in

1) Noonans syndrome
2) Turners syndrome
3) Trisomy Syndrome.

Low Hair Line

When the posterior hair line extends below the level of C5 spinous process or the ratio of the distance from the external occipital protuberance to the hair line and the distance from the hair line to C7 spinous process is > 1/6 in males and >1/4 in females is defined as low hair line and is seen in

1) Noonans Syndrome
2) Turners Syndrome
3) Klippel-Feil syndrome
4) Cornelia de Lange syndrome

Skin

Bronze Pigmentation

Bronze pigmentation of the skin is seen in Haemochromatosis a common disorder of iron storage in which inappropriate increase in intestinal iron absorption results in deposition of excess iron in parenchymal cells leading to tissue damage and organ dysfunction. Cirrhosis of liver diabetes mellitus, arthritis, cardiomyopathy and hypogonadotropic hypogonadism are common manifestations. Excessive skin pigmentation is seen in over 90% of symptomatic patients. The pigmentation has a metallic slate grey hue called bronzing and results from increased melanin and iron in the dermis. The pigment is usually diffuse and generalized but may be prominent on the face neck extensor aspect of forearms, dorsum of the hands, lower legs, genital regions and in scars.

Café-Au-Lait Spots

Café-au-lait spots are flat hyperpigmanted skin lesions that have rough or smooth borders. The café-au-lait spots in Neurofibromatosis have smooth borders and also called as Coast of California. The café-au-lait spots in Micune Albright syndrome have rough borders and are also called as Coast of Maine. Café-au-lait spots are seen in

1) Neurofibromatosis — Phaechromocytoma
2) Tuberos sclerosis — Rhabdomyoms
3) Leopard syndrome
4) Watson syndrome — Pulmonary stenosis
5) MiCune Albright syndrome
6) Multiple endocrine neoplasia

Multiple Lentigines

Lentigines are hyperpigmented spots due to proliferation of melanocytes. In majority they are due to sun exposure which explains their distribution. It is seen in

1) Leopard syndrome — HCM
2) LAMB syndrome — Atrial myxoma
3) NAME syndrome — Atrial myxoma
4) Peutz Jeghar syndrome

Addisons disease

Adenoma Sebaceum

Adenoma sebaceum are angiofibromas of the face are orange red to yellow naevi of a few mm to cm on the face symmetrically distributed over malar and nasal skin. It is seen in Tuber Sclerosis or Bournewilles disease. Shagreen patches – Yellowish thickening of skin over the lumbosacral region of the back and Ash leaf shaped hypopigmented macules are the other two cutaneous lesions seen in Tuberos Sclerosis along with adenoma sebaceum.

Hyperelasticity Of Skin

Hyperelasticity of skin and hypermobile joints is seen in Ehlers-Danlos syndrome a inherited disorder of connective tissue. There are 11 types of Ehler Danlos syndrome defined based on the extent of skin, joints and other tissue involved. The genetic mutation leads to defects in synthesis of collagen or conversion of procollagen to collagen. It is because of this that the skin varies from very thin and velvety to dramatically hyperextensible (Rubber man syndrome) or scarred.

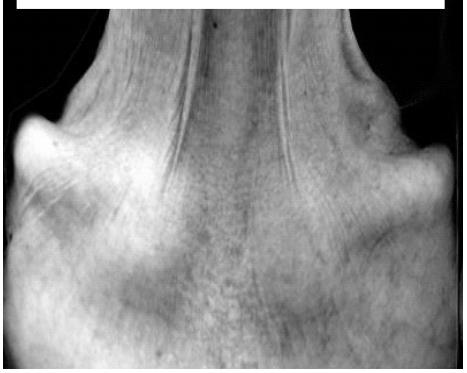
Type I patients develop cigarette paper scars

Type IV the skin may be so thin that subcutaneous blood vessels are visible.
Easy bruisability occurs in several types of Ehlers Danlos syndrome. Laxity and hypermobility of joints vary from mild to unreducable dislocations of hips and other large joints. The cardiovascular manifestations in Ehlers Danlos syndrome are arterial dilation and rupture and mitral regurgitation.

Plucked Chicken Appearance

In Pseudoxanthoma elasticum there is an abnormal deposition of calcium on the elastic fibres of the skin eye and blood vessels. In the skin the flexural areas such as neck, axilla, antecubital fossa and inguinal areas are the primary sites of involvement. Yellow plaques coalesce to form reticulated plaques that have an appearance similar to that of plucked chicken skin. In severely affected skin, hanging redundant folds develop. Biopsy specimen of involved skin shows swollen and irregular clumped elastin fibres with deposits of calcium.

PLUCKED CHICKEN
APPEARANCE
PSUEDO ZANTHOMA
ELASTICUM



Erythema Marginatum

Erythema marginatum are transient, macular, non pruritic rashes with serpinginous erythematous borders and normal skin surrounding it. It is about 1 inch in diameter, blanches completely on pressure, localized in distribution to trunk and proximal limbs. It is difficult to recognize in dark skinned people and can be brought out by 1)Warmth 2)Hot towels and 3)Warm bath. It is directly proportional to the severity of carditis.

Also seen in 1) Sepsis
2) Drug reactions
3) Glomerulonephritis
4) Idiopathic.

Vasculitis is the underlying pathological proicess in erythema marginatum

Subcutaneous Nodules

They are round, firm, painless, freely mobile transient nodules occurring in crops over bony prominences and extensor tendons (1)Elbow 2)Knee 3)Ankle 4)Suboccipital region 5)Medial border of scpula 6)over spinous process. The skin above the nodule is pinchable and lasts for a week or two then disappearing without a trace. It is a rare major manifestation of acute rheumatic fever and is seen in association with severe carditis. The histology of subcutaneous nodule is similar to Aschoff body consisting of central fibrinoid necrosis surrounded by epithelial and mononuclear cells.

Oslers Nodes

Oslers nodes are small tender subcutaneous nodules on the pads of fingers or toes and palms or soles seen in 7-10% of infective endocarditis patients. It is due to deposition of immune complexes in mucocutaneous vessels and infected microemboli. It is a minir criteria in the Dukes criteria to diagnose infective endocarditis.

Cyanosis

Cyanosis is bluish discoloration of skin and mucous membrane resulting from an increased quantity of reduced hemoglobin or of abnormal hemoglobin pigments in the blood perfusing these areas. Cyanosis can be central or peripheral. During clinical examination in a cyanotic patient the following physical examination observations can take us to the diagnosis, 1)Onset of cyanosis 2) Type of cyanosis 3)Association with failure 4)Association with squatting or its equivalent 5)Association with a wave 6)Association with Collapsing pulse 7)Association with continuous murmur 8)Differential cyanosis 9)Reversed differential cyanosis 10)Association of left axis deviation ECG 11)Association with LV/RV apical impulse.

Clubbing

Clubbing of fingers also known as Hippocratic fingers is due to hypervascularity of the nailbed which increases the hydrostatic pressure in the capillariespromoting interstitial edema and hypertrophy of soft tissues. The degree of clubbing can be graded into 4 grades. Clubbing could be unidigital, unilateral or bilateral and symmetrical. Causes for clubbing could be familial. Cardiovascular,pulmonary and Gastrointestinal. Cardiovascular causes are

1) Cyanotic congenital heart disease
2) Infective endocarditis

Pedal Edema

Pedal edema is defined as accumulation of fluid in the legs due to increase in the interstitial component of extracellular fluid volume. One has to see whether the pedal edema is unilateral or bilateral and whether it is pitting or nonpitting. The cardiovascular causes for pedal edema are

1) Congestive cardiac failure
2) Constrictive pericarditis
3) Post phlebitis syndrome
4) Varicose veins
5) Chronic venous insufficiency
6) Calcium channel blocker induced

Terry Nails/Lindsay Nails

Alternating dark and light strips parallel to the tips of the nails occurs in

1) Protein losing enteropathy
2) Cirrhosis
3) Constrictive pericarditis

Koilonychia

Koilonychia or spoon shaped nails occurs when the normal transverse curvature of the nail plate becomes flat or concave due to iron deficiency anaemia

Subungual Haemorrages

Splinter haemor\ages or subungual haemorrages are dark red linear or occasionally flame shaped streaks in proximal nail bed seen in infective endocarditis.

1) Infective endocarditis
2) Trauma

Raynauds Phenomenon

Raynauds phenomenon is characterized by episodic digital ischemia \, manifested clinically by sequential development of triphasic colour changes.

1. Blanching (White) – vasospasm of digital vessels causing pallor
2. Cyanosis (Blue) – vasospasm – ischemia – dilatation of papillaries and venules with stagnation of blood causing the blue colour.
3. Rubor (Red) – reverting the digital vasospasm causes increased flow into the vessels and causes the redness.

It is seen in:

1. Primary
2. Secondary

a. Collagen vascular disease
b. TAO

c. Atherosclerosis
d. Thoracic outlet syndrome
e. Trauma
f. Drugs
i. Ergot derivatives
ii. Methysergive
iii. Beta blockers
iv. Bleomycin
v. Cisplatin

Arachnodactyly

Arachonodactyly or spider finges are unduely long and thin fingers and toes. Patients with arachnodactyly will have positive thumb sign and wrist sign. Their metacarpal index will also be > 8.

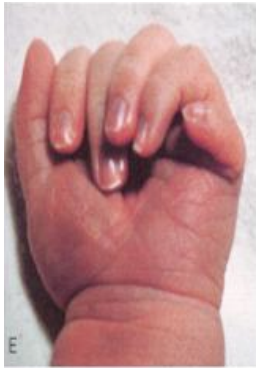
Thumb Sign (Steinburg Sign) – Thumb sign is the protrusion of the thumb behind the medial border of the hand in a closed fist.

Wrist Sign (Walker Murdock Sign) – Ability to oppose the thumb abd little finger of one hand over the wrist of the other hand.
It is seen in
1) Marfans syndrome
2) Homocystinuria
3) Sickle cell anaemia

Polydactyly

Polydactyly is the presence of extra or supranemeray fingers or toes. It is seen in
1) Trisomy 13
2) Ellis van Crevald syndrome
3) Rubenstein taybi syndrome
4) Laurence Moon Biedel syndrome

Polydactyly



Syndactyly

Syndactyly I is the fusion of adjacent fingers or toes and may be dermal or osseous.
1) Aperts syndrome
2) Cornelie de lange syndrome
3) Holt Oram syndrome
4) Trisomy 13/18/21
5) Laurence Moon Beidle syndrome

Brachydactyly

Brachydactyly is the presence of shortened fingers resulting in short hands. It is seen in

1) Downs syndrome
2) Turners syndrome
3) Achondoplasia

Fingerized Thumb

Thumb with an extra phalanx and the finger lying in the same plane as the rest of the fingers making it difficult to oppose with other fingers is called fingerized thumb. It is seen in Holt oram syndrome

Phocomelia

Limbs without hands, like flipper is associated with

1) Holt Oram syndrome
2) Corneliadelange syndrome
3) Thalidomide

Simian Crease

Simian crease or single palmar crease is normal finding in monkeys. It is seen as a normal finding in 4% of normal Caucassian population unilaterally and bilaterally in 1%. It is seen in

1) Down's syndrome
2) Edward's syndrome
3) Cri-du-chat syndrome
4) Cornelia de lange syndrome.

Jaccouds Arthritis

Jaccouds arthritis is named after Sigismond Jaccoud a French physician. It is deforming non-erosive arthropathy after repeated attacks of arthritis in Rheumatic fever. This is the only deforming arthritis complication of rheumatic fever. The deformity is flexion and ulnar deviation of metacarpophalangeal joints of 4^{th} and 5^{th} finger and flexion and fibular deviation of metatarsal phalangeal joints. It is seen in

1) Scleroderma
2) SLE
3) Rheumatoid arthritis
4) Ehlers Danlos syndrome
5) Aggamaglobinemia

Hammer Toe/Pes Cavus

In Frederick ataxia patient has ataxia, kyphoscoliosis, speech defects, hammertoe and pes cavus. The Cardiological manifestations are cardiomyopathy and conduction defects.

CHEST AND ABDOMEN

Barrel Chest

In a normal adult the shape of the chest is truncated cone with AP diameter being less than the lateral diameter in the ratio of 5:7. When the chests anteroposterior diameter is more than the lateral diameter it is known as barrel chest. It is seen in

1) Chronic bronchitis
2) Emphysema

Pectus Excavatum

Pectus excavatum or Funnel chest is a congenital deformity of the lower portion of the sternum which is depressed posteriorily and the anterior ribs are markedly bowed, which results in a depressed panel in the anterior chest. Respiratory symptoms are inconspicuous and Pulmonary function tests are normal. It is seen in Marfans syndrome. It is quantified as mild moderatr 2-5 ml and severe >5 ml.

Pectus Carinatum

Pectus Carinatum or pigeons chest is the reverse of pectus excavatum with the sternal portion protruding anteriorly. This also does not cause any respiratory symptoms or abnormalities in the PFT. It is seen in

1) ASD
2) VSD
3) Marfans
4) Prolonged childhood asthma

Kyphoscoliosis

Kyphoscoliosis is a combination of excessive AP and lateral curvature of the thoracic spine. Kyphoscoliosis occurs in 3% of the population however symptoms related to it are seen in only 3% of the above population. The pathophysiological consequences of kyphoscoliosis are restrictive lung disease and ventilation perfusion imbalances that result in chronic alveolar hypoventilation, hypoxic vasoconstriction and eventually pulmonary arterial hypertension and cor pulmonale. If the scoliosis is < 60 degrees ventiloatory impairment is rare while if it is >90 degree marked ventilatory abnormalities develop. It is seen in

1) Isolated
2) MVP
3) Marfans
4) Neurofibromatosis
5) Metabolic disorders
6) Myopathies.

Harrisons Sulci

Harrisons sulci extend transversly as grooves from the sides of the xiphisternum on either side giving the thorax an appearance of transverse constriction. These grooves correspond to the costal attachments of the diaphragm and are due to the pulling of the softened ribs. It is seen in

1) L-R shunts
2) Rickets
3) Nasopharangeal obstruction
4) Bronchial asthma

Straight Back Syndrome

The normal thoracic vertebrae has mild khyposis, loss of this normal thoracic kyphosis is called Straight back syndrome. This reduction in thoracic space results in expiratory splitting of S2, parasternal systolic impulse, ejection systolic murmur and prominence of pulmonary artery in Chest X-Ray mimicking atrial septal defect. It is seen in

1) MVP
2) ASD

Shield Chest

A broad chest has widely spaced nipples and prominent angle between the manubrium and body of the sternum. It is seen in

1) Turners syndrome
2) Noonans syndrome

Hypomastia

Hypomastia or decreased breast tissue is seen in

1) Marfans syndrome
2) MVP

Gynecomastia

Gynecomastia refers to enlargement of the male breast due to excess estrogen action and is result of increased estrogen/androgen ratio. True gynecomastia is associated with breast tissue > 4 cm in diameter and is tender. It is seen in

Physiological
1) New born
2) Pregnancy
3) Aging

Pathological
4) Klinfelters syndrome
5) Peutz Jeghars syndrome
6) Sertoli cell tumor
7) Carey complex
8) Drugs 1)Oral contraceptive
   2) Estrogens
   3) Digitalis
   4) Spironolactons
   5) Ketoconazole

Left Thoracotomy Scar

If a patient has Left thoracotomy scar the possible surgeries that he has undergone are

1) Closed Mitral commissurotomy
2) PDA ligation
3) Coarctation of aorta repair
4) BT Shunt
5) PA banding.

Right Thoracotomy Scar

If a patient has Right thoracotomy scar the possible surgeries that he has undergone are

1) Potts shunt
2) Waterstons shunt
3) Mitral valve replacement
4) ASD closure

Abdominal Striae

Abdominal striae are purpish striae seen in the flangs of the abdomen in patients with Cushing syndrome. They also have central obesity, moon facies and hypertension.

Ascites

Accumulation of fluid in the peritoneal cavity is called ascites. Cardiovascular causes for ascites are :

1. Congestive cardiac failure
2. Tricuspid valve disease
3. Constrictive pericarditis
4. Hepatic vein thrombosis

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CYANOSIS

Dr J.Ezhilan

Si-a-no sis – Greek word meaning dark blue colour

Kyanos – French word meaning blue substance

Cyanosis was first described by C.Lundsgaard a Norwegian Pulmonologist.

Cyanosis is both a sign and symptom.

Definition:

Cyanosis is a bluish discoloration of the skin and mucous membrane resulting from an increased quantity of reduced hemoglobin or of abnormal hemoglobin pigments in the blood perfusing these areas.

Actually the term reduced hemoglobin should not be used for during these reactions iron stays in the ferrous state and only if it becomes Ferric form one can call it reduced haemoglobin. (as occurs in Methemoglobinemia)

Cyanosis could be central cyanosis or peripheral cyanosis

Central Cyanosis

Central Cyanosis is due to arterial blood unsaturation or an abnormal Hb derivative with the mucous membrane and skin both being affected. Central cyanosis is seen in

1) R-L Shunt
2) Impaired pulmonary function
3) Abnormal hemoglobin

Peripheral Cyanosis

Peripheral cyanosis is secondary to cutaneous vasoconstriction due to a) low cardiac output b)exposure to cold air or water causing slowing of blood flow and an abnormally greater extraction of oxygen from normally saturated blood. It is seen in

1) Decreased cardiac output
2) Cold exposure
3) Redistribution of blood flow
4) Arterial obstruction
5) Venous obstruction

Central Cyanosis	Peripheral Cyanosis
1) O_2 saturation is < 85%	O_2 saturation is normal
2) Mechanism- Arterial blood unsaturation	Increased peripheral utilization
3) Skin & mucous membrane involved	Skin only is involved
4) Skin over cyanosis is warm	Skin over cyanosis is cold & clammy
5) Clubbing	No clubbing
6) Polycythemia	No Polycythemia
7) O_2 therapy has no effect	O_2 therapy improves
8) Warming – No effect	Warming – Improves
9) Exercise – worsens	Exercise – improves
10) AV O_2 difference is normal - 5 vol%	AV O_2 diff is greater >12 vol%
11) P O_2 is decreased	PO_2 is normal

Only 2 congenital heart disease where oxygenation increases O_2 saturation are

1) TAPVC

2) Pulmonary AV fistula

Cyanosis causes

Central cyanosis

1. DECREASED ARTERIAL O_2 SATURATION
   a. DECREASED ATMOSPHERIC PRESSURE
      i. High altitude
   b. IMPAIRED PULMONARY FUNCTION
      i. Alveolar hypoventilation
      ii. Ventilation Perfusion mismatch
      iii. Impaired oxygen diffusion
   c. ANATOMIC SHUNTS
      i. Congenital heart disease
      ii. Pulmonary A-V Fistula
      iii. Multiple small intrapulmonary shunts
   d. HAEMOGLOBIN WITH LOW AFFINITY FOR OXYGEN
2. HAEMOGLOBIN ABNORMALITIES
   a. Methemoglobinemia
   b. Sulfhaemoglobinemia
   c. Carboxyhaemoglobinemia

Peripheral Cyanosis

1) Reduced Cardiac output
2) Cold exposure
3) Reduction of blood flow
4) Arterial obstruction
5) Venous obstruction

Differential Diagnosis For Cyanosis

GENETIC CAUSES	Hyperpigmentation of eyelids, axilla & nailbeds
METABOLIC CAUSES	Haemochromatosis Ochronosis
NUTRITIONAL CAUSES	Chronic Nutritional insufficiency
CHEMICAL CAUSES	Heavy Metal Intoxication 1) Silver (Argyria) 2) Gold (Chrysiasis) 3) Bismuth 4) Lead Topical mercury – Face creams Fixed drug eruptions – Phenothiazines
NEOPLASTIC CAUSES	Metastatic melanoma

Before going into the pathophysiology of cyanosis one has to get into the physiology of haemoglobin and oxygen transport system for better understanding. Haemoglobin is a red oxygen carrying pigment in the red blood cell. It is globular molecule with a molecular weight of 64,450. It is made of 4 subunits each subunit consisting of

HAEME + GLOBIN

IRON + PORPHYRINS — 4 POLYPEPTIDE CHAINS

2 Pairs of alpha chains (141)

2 pairs of Beta chains (146)

Within the RBC's of embryo, fetus, child and adults six different haemoglobins may be detected

Embryonic Haemoglobins

1) Gower 1 ($\zeta 2, \varepsilon 2$) — Appear during 4th week of gestation
2) Gower 2 ($\alpha 2, \varepsilon 2$) — Dominant Hb from 4th-8th week
3) Portland Hb ($\beta 2, \gamma 2$) — Disappears by 3rd month

Fetal Haemoglobins

1) Fetal Haemoglobin ($\alpha 2, \gamma 2$)
   Dominant Hb from 8th week of gestation
   6th month – 90% of total haemoglobin
   At birth – 70% of total Hb
   6th month -<1% of total Hb

Adult Haemoglobin

1) Haemoglobin A1 ($\alpha 2, \beta 2$)
   A1A- Appears during 16th-20th week
   A1B-6th month gestation – 5-10%
   A1C-30% at term / 6-12 mths Normal adult patter
2) Haemoglobin A2 ($\alpha 2, \delta 2$)
   A1 to A2 ratio (30:1)
   At birth < 1%
   12 months – 2 to 3.4%

ADULT HAEMOGLOBIN
Hb A1 – 97%
Hb A2 – 2-3.4%
Hb F – 1%

Hb at birth – 22 gms
Hb at 3 months – 11 gms
Hb at 2 years – 13 gms

Reactions Of Haemoglobin

Haemoglobin + oxygen - Oxyhaemoglobin
Haemoglobin + Carbondioxide - Carboxy haemoglobin
Haemoglobin + Hydrogen sulphide - Sulphoxyhaemoglobin
Haemoglobin + Iron(Ferric form) - Methaemoglobinemia

(<.01 second)

$Hb + O_2 \rightarrow$ Oxyhaemoglobin

Affected by

1) pH
2) Temperature
3) 2,3 Diphospho glycerate

Normal Skin Colour

Light impinging on human skin is partly reflected and partly transmitted inward to successive skin layers epidermis, dermis and subcutaneous tissues. Each layer contains pigments, which act as colour screens or biological optical filters. Light striking each layer is absorbed, scattered or transmitted to the layer below. The colour perceived at the skin surface is determined by the wavelength ultimately remitted e.g.

If all the wavelengths were absorbed skin would appear black.

If all the wavelengths were remitted skin would appear white.

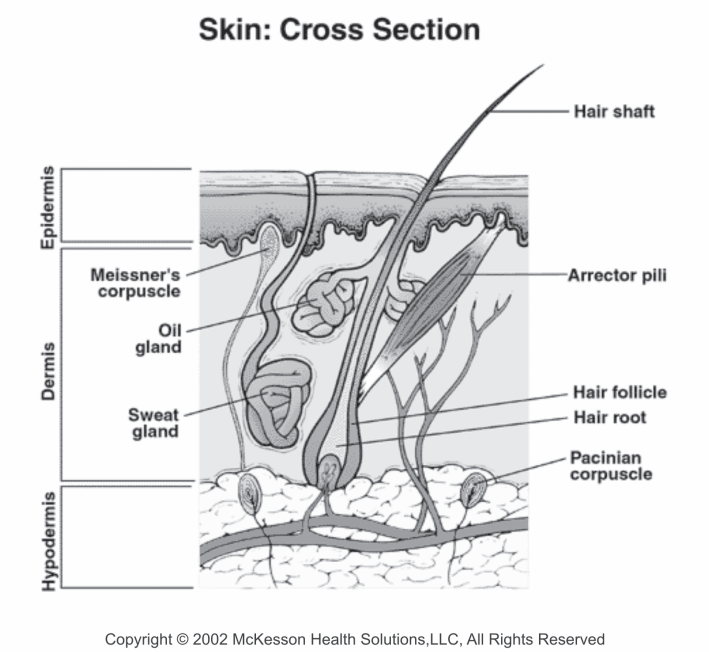
Light reflected at the outer surface of the skin does not contribute significantly to skin colour. Normal skin colour is determined by the absorption protein density and distribution of four major pigments in the skin

1)	Melanin	I Epidermis
2)	Carotene	Cornified layer of epidermis
		Sebaceous glands
		Subcutaneous fat
3)	Oxygenated Hb	Blood perfusing the skin
4)	Deoxygenated Hb	Blood perfusing the skin

The skin contains 1) Subpapillary plexus and 2) Deep plexus

The Subpapillary plexus is horizontal and largely responsible for skin colour.

The deep plexus normally contains loop, which come from deep plexus and are vertical. At certain sites they are also horizontal and hence cyanosis is clearly made out at these sites.



Human body contains 15 gms of Hb/100 ml of blood. 1 gm of Hb can combine with 1.34 ml of O_2. 15 gm of Hb would combine with 20 ml of O_2. Since only 94.97% of Hb is saturated 15 gm of Hb would combine with 19 ml O_2 (19 vol%). During passage of blood through capillaries to veins O_2 is given off to tissues 22% (5-6 vol%)

Thus O_2 in venous blood is 14-15 vol%. Oxygen unsaturation in venous blood is 5-6 vol%. It is this 5-6 vol% unsaturation that imparts a blue colour to the veins. If the O_2 unsaturation in the capillaries exceeds 6.5% cyanosis is seen.

Sites Of Cyanosis

1)	Lips	5)	Cheeks
2)	Ear lobes	6)	Hand and feet
3)	Nose tip	7)	Finger tips
4)	Nail bed	8)	Trunk

Cyanosis is better observed in hands and feet because 1) skin is thin 2) Skin is unpigmented and 3) capillaries are numerous. Cyanosis is rarely seen in trunks because there is not enough blood vessels in the skin.

Degree Of Cyanosis

Degree of cyanosis depends on

1) Skin pigmentation

2) Amount of haemoglobin

3) Amount of deoxygenated haemoglobin

4) Colour of blood plasma

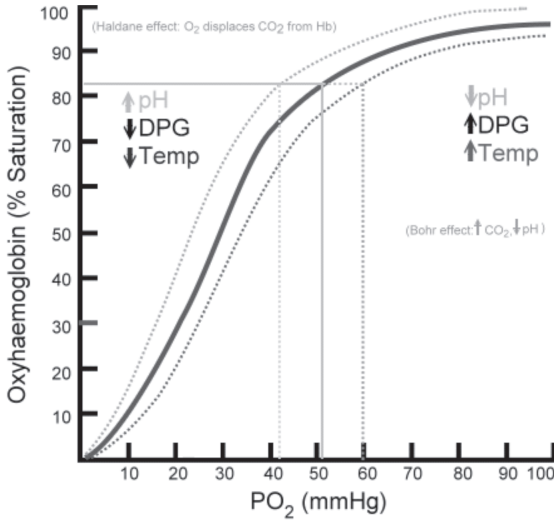
5) Skin thickness

6) Capillary density

7) Capillary orientation

8) Vessel caliber

Oxy-Haemoglobin Dissociation Curve



1)	Increase in plasma pH	Shifts the curve to the left and a relative decrease in O_2 unloading
2)	Decrease in temperature	
3)	Decrease in 2-3 DPG	

1)	Decrease in plasma pH	Shifts the curve to the right and a relative increase in O_2 unloading
2)	Increase in temperature	
3)	Increase in 2-3 DPG	

Cyanotic Point

1) O_2 saturation	<	85%
2) Deoxyhaemoglobin	>	5 gm/100 ml
3) Methaemoglobin	>	2 gm/100 ml
4) Sulphaemoglobin	>	0.5 gm/100 ml
5) O_2 unsaturation	>	6.5 vol%
6) R-L shunt	>	25% of LV output.

Cyanosis Overlooked

1) Anaemia (Hb < 33% of normal)
2) Secondary Polycythemia (Not yet developed)
3) Arterial Blood O_2 saturation (85-93%)
4) Increased skin pigmentation
5) R-L shunt < 40%
6) Blood shunted away from skin.

Types Of Cyanosis

1) Central cyanosis
2) Peripheral cyanosis
3) Mixed cyanosis
4) Differential cyanosis
5) Reversed differential cyanosis
6) Deffered cyanosis
7) Transient cyanosis
8) Persistent cyanosis
9) Cyanosis tardive
10) Acrocyanosis
11) Orthodeoxia
12) Tuft erythema
13) Malar flush
14) Heliotrope cyanosis
15) Intermittent cyanosis
16) Autotoxic Enterogenous cyanosis

Mixed Cyanosis

When both the causes for central and peripheral cyanosis are there mixed cyanosis can occur eg

1)	Pulmonary edema (Central cyanosis)	with	Cardiogenic shock (Peripheral cyanosis)
2)	Chronic Bronchitis (Central cyanosis)	with	Cor Pulmonale (Peripheral cyanosis))
3)	PPH (Central cyanosis)	with	Raynauds phenomenon (Peripheral cyanosis)

Differential Cyanosis

Presence of differential cyanosis in a patient indicates

1) Presence of a congenital heart disease

2) Patency of ductus arteriosus.

If not obvious clinically differential cyanosis can be provoked by

1) exercise

2) Warming the hand and feet (Which increases skin blood flow)

Confirmation – Simultaneous O_2 saturation in femoral and brachial artery

Upper Part Pink Lower Part Blue

1) PDA with reversal of shunt

2) Coarctation of aorta with PDA and reversal of shunt

3) Interruption of Aortic arch with PDA and reversal of shunt

4) Tubular hypoplasia of Aortic arch with PDA and reversal of shunt

Right Arm Pink Left Arm Blue Lower Part Blue

1) If unoxygenated blood enters the left subclavian artery

2) If ductus arises from Left Subclavian artery

Right Arm Blue Left Arm Pink Lower Part Blue

1) Right sided ductus

2) Aberrant Rt subclavian distal to the ductus.

Head Right Shoulder Right Arm Cyanosed

1) Persistent fetal circulation with R-L ducal shunting

Reversed differential cyanosis

Transposition of great Vessels with Co of aorta with PDA with reversal of shunt
Oxygenator blood → Pulmonary veins → LA → LV → PT → Ao → Supplies lower half of body

Taussing Bing anomaly

DORV with Subpulmonic VSD with PDA with reversal of shunt
Oxygenated blood → PV → LA → LV → Through VSD → Into PA → Through PDA → Aorta → Lower limbs.. RV blood enters aortic root and supplies upper half.

Deffered Cyanosis

In deferred cyanosis the child is not cyanotic at birth but after 3-6 months the child becomes cyanotic and thereafter the cyanosis is persistent. Causes are

1) Persistence of ductal patency

2) Persistence of fetal hemoglobin

3) Decreased energy expenditure at birth

4) Progressive infundibular stenosis

Deferred cyanosis is seen in

1) Tetralogy of Fallot

2) L-TGV VSD with PS

3) DORV VSD with PS

4) SV VSD with PS

5) TGV VSD with PS

Transient Cyanosis

In Transient cyanosis the child is born at birth with cyanosis and after a month when the PVR falls the cyanosis regresses. The cyanosis is due to R-L shunt caused by the increased PVR, which ceases when the PVR comes down. It occurs in

1) ASD

2) Ebsteins anomaly

3) DORV with subaortic VSD without PS.

4) Truncus arteriosus

ASD – At birth the high PVR and decreased compliance of RV causes R-L shunt and cyanosis, which gradually ceases as the PVR falls down.

EBSTEINS ANOMALY- Transient neonatal cyanosis due to R-L shunt, which disappears as neonatal PVR, normalizes and recurs later as the filling pressures in the functionally abnormal ventricle rises. Recurs a decade or more later.

DORV WITH SUBAORTIC VSD WITHOUT PULMONARY STENOSIS- From the LVsaturated blood reaches aorta through the subaortic VSD. From the RV blood is flown into the pulmonary artery. Because of the increased PVR flow is less after PVR falls, pulmonary flow increases and cyanosis disappears.

TRUNCUS ARTERIOSUS- Because of high neonatal pulmonary vascular resistance less flow occurs across from the truncus into the pulmonary vessel. Later when PVR falls increased flow into the pulmonary circulation causes cyanosis to cease.

Intermittent Cyanosis

Intermittent cyanosis occurs in these conditions where strain, exercise or arrhythmia brings out a transient R-L shunt and cyanosis. Causes are

1) Ebsteins anomaly

2) TAPVC unobstructed

3) Complete A-V canal defect

4) Eissenmengers with bidirectional shunt

Persistent Cyanosis

Persistent cyanosis appears at birth and after that it remains persistent and is either static or progressive. It occurs in

1) Transposition of great vessels

2) Single ventricle

3) Hypoplastic Left Heart syndrome

4) Tricuspid atresia

5) Pulmonary atresia

6) Ebsteins anomaly

Ebsteins anomaly is one condition which can have all the three types of cyanosis a)Transient b) Intermittent and c)Persistent.

Cyanosis Tardive

Cyanosis tardive or very late onset cyanosis is cyanosis which occurs after first decade due to reversal of L-R shunt as seen in

1) ASD 2) VSD 3) PDA 4) A-P Window

5) Triology of Fallot (PS with R-L shunt at PFO)

Ruddy Cyanosis

Ruddy cyanosis is not a true cyanosis is seen in patients with Polycythemia Vera due to expanded blood volume with resulting dilation of cutaneous vessels and sluggish flow

Orthodeoxia

It is a rare syndrome characterized by orthostatic accentuation of a right to left shunt across an ASD or PFO in the presence of normal pulmonary arterial pressures. Seen in

1) Normal elderly individuals 2) ASD

Tuft Erythema

Tuft erythema is a reddish discolouration of finger tips due to mild cyanosis and is seen in ASD

Malar Flush

Hyper pigmentation of the cheeks around the nose due to long standing peripheral cyanosis in chronic mitral stenosis. It is well made out in whites and difficult to appreciate in dark races

Autotoxic Entergeneous Cyanosis

Autotoxic Enterogeneous cyanosis is a disorder attributed to the formation of intracellular methemglobin or sulfhaemoglobin by toxic substances, possibly of bacterial origin absorbed from GI tract.

Acrocyanosis

Arterial vasoconstriction and secondary dilation of capillaries and venules resulting in peripheral cyanosis of hands and less frequently feet is acrocyanosis. Cyanosis is intensified by exposure to cold and women are more frequently affected. Age of onset < 30 years and patients are usually asymptomatic. The patients have normal pulses, peripheral cyanosis and moist palms. It is distinguished from Raynauds phenomenon by being persistent. Management is to dress warmly and avoid cold exposures.

Approach To Cyanotic New Born

In a new born with cyanosis it has to be due to the following four systems

1)	CARDIAC	-	1)	R-L Shunt
2)	PULMONARY	-	1)	Upper airway obstruction
			2)	Hyaline membrane disease
			3)	Pneumonitis
			4)	Atelectesis
3)	CNS	-	1)	CNS depressants-Sedatives/Hypnotics
			2)	Birth trauma-ICH
4)	HAEMATOLOGICAL	-	1)	Methaemoglobinemia
			2)	Sulphaemoglobinemia
			3)	Carboxyhaemoglobinemia

Differentiating Causes For Cyanosis In New Born

Cardiac

- Vigorous or labored respiration
- Increased depth of respiration
- Tachypnoea < 60/mt
- Auscultatory findings

Pulmonary

- Vigorous or labored respiration
- Tachypnoea > 80-110/mt
- Flaring of alae nasi
- Intercostals retraction
- Expiratory grunting
- Increased PCO_2
- Response to oxygen therapy

Central Nervous System

- Irregular shallow breathing
- Lethargy
- Hypotonicity
- Reduced spontaneous movements
- Convulsions

Haematological

- Cyanosis from birth or after 3-6 months
- No abnormal breathing pattern
- Autosomal recessive pattern of inheritance
- Mild polycythemia but no clubbing
- No evidence of lung or heart disease
- Venous blood becomes chocolate brown on exposure to air
- Normal Pa O_2 but low O_2 saturation
- Spectroscopic examination

Normal Respiratory Rate At Birth And Infancy

At birth respiratory rate is 60-70/mt decreases by day 1 to 30-55/mt

Minimal intercostals retraction and expiratory grunting present at birth disappears within 1 hour. Both the above should not be taken for respiratory embrassment.

Approach To Cyanotic Heart Disease

The following issues should be looked into in a patient with cyanotic heart disease to get clues that will lead us to the diagnosis

1) History of exposure to drugs?
2) Type of cyanosis?
3) Cyanosis with failure
4) Cyanosis with squatting
5) Differential cyanosis

6) Reversed differential cyanosis
7) Cyanosis with a wave in JVP
8) Cyanosis with Collapsing pulse
9) Cyanosis with continuous murmur
10) Cyanosis with LAD in ECG
11) Cyanosis with LV/RV/Combined apical impulse.

History of Exposure to Drugs

Exposure to the following drugs can cause cyanosis

1) Nitrites
2) Nitrates
   a) Sodium nitrate
   b) Amyl nitrate
   c) Nitroglycerine
   d) Sodium Nitroprusside
   e) Silver Nitrate
3) Chlorate
4) Phenazopyridine
5) Sulfonamides
6) Aniline dyes
7) Acetanilid
8) Phenacitin
9) Lignocaine
10) Chlorate
11) Phenazopyridine
12) Sulfonamides

History Of Type Of Cyanosis

Certain diseases have particular type of cyanosis and thus knowing the type of cyanosis one can hypothesis possible conditions

Transient Cyanosis

1) Atrial septal defect
2) Ebsteins anomaly
3) DORV with subaortic VSD without pulmonary stenosis
4) Truncus arteriosus

Intermittent Cyanosis

1) Ebsteins anomaly
2) TAPVC unobstructed
3) Complete A-V canal defect
4) Eissenmengers with bidirectional shunt

Persistent And Progressive Cyanosis

1) Transposition of great vessels

2) Single ventricle

3) Hypoplastic left heart syndrome

4) Tricuspid atresia

5) Pulmonary atresia

6) Ebsteins anomaly

Deferred Cyanosis

1) Tetralogy of Fallot

2) Transposition of great vessels with VSD with PS

3) Corrected Transposition of great vessels with VSD with PS

4) Double outlet right ventricle with VSD with PS

5) Single ventricle with VSD with PS

Cyanosis With Failure

Congestive cardiac failure in a patient with cyanosis denotes cyanotic heart disease with increased pulmonary blood flow physiology.

1) Transposition of great vessels

2) Taussing Bing anomaly

3) Truncus arteriosus

4) Total anomalous Pulmonary venous connection

5) Single ventricle with low PVR and no pulmonary stenosis

6) Common atrium

7) Tetralogy of Fallot with pulmonary atresia with inc collateral flow

8) Tricuspid atresia with nonrestrictive VSD

9) Complete Interruption of aortic arch with VSD and PDA

Cyanosis With Squatting

History of squatting or squatting equivalents like 1) Knee chest position 2) Lying down 3) Sitting with legs drawn underneath 4) Legs crossed while standing and 5) Carrying a child with legs flexed upon its abdomen all indicate that the patient is having cyanotic heart disease with decreased pulmonary blood flow physiology. Of conditions with decreased pulmonary flow all do not have history of squatting or squatting equivalent

1) Tetralogy of Fallot

2) Tricuspid atresia

3) Pulmonary atresia

4) Double outlet right ventricle

5) Single ventricle

6) Transposition of great vessel

7) Eissenmengers syndrome

In Transposition of great vessels a condition with increased pulmonary flow squatting occurs in 15% of patients because of dynamic subpulmonary stenosis due to left ventricular outflow obstruction.

Differential Cyanosis

Differential Cyanosis where the upper limbs are pink and the lower limbs are blue is seen in

1) PDA with reversal of shunt

2) Coarctation of aorta with PDA and reversal of shunt

3) Interruption of aortic arch with PDA and reversal of shunt

4) Tubular hypoplasia of aortic arch with PDA and reversal of shunt

5) Persistent fetal circulation with PDA and reversal of shunt.

Reversed Differential Cyanosis

In reversed differential cyanosis the upper limbs are blue and the lower limbs are pink

1) TGV with Coarctation of aorta with PDA with reversal of shunt

2) Taussing Bing anomaly.

Cyanosis With A Wave In Jvp

Cyanotic heart diseases with prominent a waves seen in JVP are

1) Tricuspid atresia

2) Pulmonary atresia with intact IVS

3) Transposition of great vessels with intact IVS

4) Eissenmengers ASD

5) Tetralogy of Fallot

Cyanosis With Collapsing Pulse

Cyanotic heart disease with collapsing pulse are

1) Tetralogy of Fallot with AR

2) Truncus arteriosus

3) TOF/PA with increased aorto-pulmonary collaterals

4) Following shunt surgeries

5) Cyanotic CHD with PDA

Cyanosis With Continuous Murmur

Cyanotic heart diseases with continuous murmur on auscultation are

1) Pulmonary atresia with VSD

2) Truncus arteriosus

3) TAPVC with obstruction

4) TOF with peripheral artery stenosis

5) Post shunt surgery done

6) Associated PDA

7) Hypoplastic left heart syndrome

Cyanosis with LVAI/RVAI/CAI

Decreased Pulmonary Blood Flow

RVAPICAL IMPULSE	1)	Tetralogy of Fallot
	2)	Pulmonary atresia with intact septum
	3)	Eissenmengers
	4)	Triology of Fallot
LV APICAL IMPULSE	1)	Tricuspid atresia
	2)	Pulmonary atresia with hypoplastic RV
COMBINED APICAL IMPULSE	1)	TGV with Pulmonary stenosis
	2)	Truncus arteriosus with hypoplastic PA

Increased Pulmonary Blood Flow

RV APICAL AIMPULSE	1)	Hypoplastic left heart syndrome
	2)	TAPVC
	3)	Transposition of great vessels
LVAI/COMBINED AI	1)	TGV with VSD
	2)	Single ventricle
	3)	Tricuspid atresia with TGV

Cyanosis With Left Axis Deviation In Ecg

Cyanotic heart disease with LAD in ECG are

1) Tricuspid atresia
2) Complete AV canal defect
3) Single ventricle –LV type
4) Transposition of great vessels with VSD with PS
5) Large Pulmonary AV fistula
6) DORV with subpulmonary VSD
7) Ebsteins anomaly

Cyanotic Heart Disease Symptoms

Hyperviscosity Symptoms

1) Headache
2) Faintness/Dizziness
3) Visual disturbances
4) Fatigue/Lassitude
5) Myalgia
6) Paresthesia
7) Depressed mentation

Haemorragic Diathesis

1) Haemoptysis
2) Easy bruising
3) Epistaxis
4) Gingival bleeding
5) Heavy menstruation

Urate Metabolism

1) Arthralgias
2) Acute gouty arthritis

Cyanotic Heart Disease Complications

1) Polycythemia
2) Clubbing
3) Cerebro vascular accident (<2yrs –Haemoconcentration is more)
4) Cerebral abscess (>18 months –Tooth eruption & mastication occurs)
5) Paradoxical emboli
6) Retinopathy
7) Hemoptysis
8) Impaired growth

Investigations Needed In A Patient With Cyanosis

1) Blood hemoglobin
2) PCV
3) Venous blood exposure to air
4) Arterial blood gas analysis
5) Saturation response to 100% oxygen
6) Spectroscopy
7) ECG
8) X-Ray Chest
9) Echocardiogram
10) MCV
11) MCHC
12) Total serum iron
13) Total iron binding capacity
14) Serum ferritin

Management of Cyanotic CHD

A) Management Of Anoxic Spells

1) Knee chest position/Squatting equivalent position
2) Oxygen - 4 lt / mt
3) Morphine - 0.1 to .2 mg / kg SC injection
4) Propranolol - 0.1 mg/Kg IV followed by .5-1 mg/kg PO 6th hourly
5) Sodabicarbonate - 5 milliequivalent/Kg
6) Correction of anemia
7) Correction of hypoglycemia
8) Emergency Shunt surgery
9) Vasopressors and Trihydroxy Methane (THM) which were used in the past are not used nowadays.

B) Management Of Hyperviscosity

Phlebotomy

The symptom of hyper viscosity is due to increased haemoconcentration of blood impairing its flow. The treatment is Phlebotomy.

- In decompensated erythrocytosis patient becomes symptomatic when the PCV>65%.
- When there is compensated erythrocytosis no symptoms occur even if PCV >70%.
- Associated iron deficiency anemia signs of hyper viscosity occur at a much lower level.
- Without iron deficiency symptoms occur at PCV of 68%
- With iron deficiency symptoms occur at PCV of 52-58%
- Rule out dehydration as cause for rise in PCV
- Remove 500 ml in 30-45 minutes
- Replace 500 ml of isotonic saline.
- Record BP in supine, sitting and standing position 15 mts before and after procedure.
- After 24 hours if no relief relieve another 500 ml.
- Maximum one can extract thrice.

Identifying Anaemia In Cyanotic Patients With Polycythemia

18% of patient with cyanotic congenital heart disease and polycythemia have iron deficiency anemia in spite of having a high HB and PCV level. Since polycythemia symptoms occur at a much lower level it is important to identify this group and correct the anemia. Iron deficiency anemia can be detected by doing

Total Serum Iron	- <	16% (Normal 20-50%)
Serum Ferritin	- <	15 ng/ml (N- Males 20-323/Females 10-383)
Treatment	–	60 mg of elemental iron 1 tid for 1-5 weeks
		Stop if Hb raises by > 2 gm%

Cyanosis In Special Situations

Cyanosis In TGA

In newborn with TGA severe cyanosis occurs when

1) Intact IVS

2) Restrictive ASD

3) Closure of existing PDA

This should be recognized early and treated because associated metabolic acidosis can cause myocardial failure.

Cyanosis In ASD

Uncomplicated ASD

1) Common atrium
2) TAPVC
3) LSVC draining into LA
4) Eustachian valve directing IVC blood into fossa ovalis
5) Unroofed coronary sinus
6) Anomalous drainage of IVC
7) Platypnoea Orthodeoxia syndrome

Complicated ASD

1) RV failure

2) Eissenmengers

Cyanosis With PA O_2 Saturation > AO O_2 Saturation

1) TGV

2) TAPVC

3) Taussing Bing anomaly

4) Sub Pulmonic VSD

Cyanosis With Atrial Isomerism

Right atrial isomerism - Cyanosis

Left atrial isomerism - CCF

Cyanosis On Exercise

Cyanosis can sometimes occur with strenuous exercise when the flow through the lungs is so rapid that there is no time of oxygen to diffuse.

Cyanosis & Clubbing

Clubbing in cyanotic congenital heart disease is not present at birth and takes 3 months to develop.

Cyanosis And Lesion Severity

Severe cyanosis – Severe lesion

Mild cyanosis – does not exclude severe lesion

Earlier the onset of cyanosis – severe the lesion.

Greater the R-L shunt – severe the cyanosis this applies to all cyanotic CHD except TGV where greater the shunt lesser the cyanosis.

Demarcation Line In Differential Cyanosis

Line of demarcation in differential cyanosis is brim of pelvis because abdominal wall receives blood supply from

1) Subscapular

2) Internal mammary

3) Superficial Epigastric

all are branches arising above the ductus.

Asymptomatic Cyanotic CHD

1) Persistence LSVC draining into LA

2) Pulmonary A-V Fistula

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PULSE

Dr. Jaishankar

Definition

Pulse is the palpability over peripheral arteries, a pulse wave, which is a transmitted wave from the root of aorta along the vessel wall traveling 10 times faster than blood.

Blood travels at speed of - 0.5 mt/sec.

Pulse travels at speed of - 5 mt/sec.

Pulse Wave

Pulse wave is a pressure wave that travels about 5 mt/sec and is propagated by the incompressible blood both forwards and laterally. The lateral movement distends the arterial wall and is felt as pulse.

Pulse Wave Components

- PERCUSSION WAVE
- TIDAL WAVE
- DICROTIC WAVE
- ANACROTIC NOTCH
- INCISURA

Percussion wave is impulse generated by LV ejection

Tidal wave is Percussion wave reflected from upper part of the body

Dicrotic wave is Percussion reflected from lower part of the body often recorded but not palpable

Anacrotic notch occurs towards the end of rapid ejection phase just before maximum pressure is reached

Incisura occurs in Isovolumic relaxation phase prior to aortic valve closure.

Dicrotic Notch

Usually occurs about 300 ms after the onset of pulse wave when corrected for heart rate

Dicrotic Wave

Wave seen after diacrotic notch, it is thought to be related to the reflection of wave from periphery.

Height of dicrotic wave decreases with

1) Age

2) Hypertension

3) Arteriosclerosis

Central Pulse

The central pulse begins with AV opening and onset of LV ejection

The rapid rising portion of the arterial pressure curve is termed anacrotic limb (Greek – upbeat)

An anacrotic notch is frequently recorded on the ascending limb towards the end of rapid ejection phase.

Peak Aortic flow velocity occurs slightly earlier than the peak pressure.

The Pulse shows 2 systolic waves Percussion wave and tidal wave & 1 diastolic wave Dicrotic wave

The descending limb of the carotid arterial pulse is less steep than the ascending limb

The descending limb is interrupted by a incisura a sharp downward deflection related to isovolumic relaxation phase during which a transient reversal of flow from the central arteries towards the ventricle occurs, just prior to AV closure.

The subsequent small positive wave is attributed to

1) Elastic recoil of aorta and AV

2) Reflected waves from most distal arteries.

Alterations In Central Pulse Peripherally

Upstroke becomes steeper

Systolic peak becomes higher (Pulse pressure increases)

Anacrotic notch disappears

Systolic upstroke time becomes shorter (120)

Systolic ejection time becomes more (320)

The dicrotic notch occurs much later

Systolic pressure increases

Diastolic pressure & mean pressure decreases.

Dicrotic wave follows the notch

Changes In Pulse With Aging

1) Increase in the height of tidal wave

2) Increase in the height of the incisura

3) Systolic upstroke time is longer

4) Amplitude & duration of diacrotic wave decreases

Normally PW is taller than TW and TW is not palpable. In old age, diabetes & arteriosclerosis TW is taller than PW and this is clinically appreciated as the pulse reaching a peak in late systole.

Peripheral Accessible Arteries

1) Head & Neck
    a. Superficial Temporal
    b. Carotids
2) Upper Limb
    a. Subclavian
    b. Axillary
    c. Brachial
    d. Radial
3) Abdomen
    a. Abdominal aorta

4) Lower Limb
    a. Femoral
    b. Popliteal
    c. Posterior Tibial
    d. Dorsalis Pedal

Superficial Temporal

It is the terminal branch of External Carotid artery and is palpable over the zygomatic process just anterior to the tragus of the ear.

Carotids

Carotid Sinus (Pressor Receptor)

Is dilatation at the origin of internal carotid artery.

Increase in BP stimulates the carotid sinus & causes slowing of heart rate & vasodilation

Carotid Body (Chemo Receptor)

Tucked deep to the bifurcation of ECA & ICA.

Decrease in oxygen tension stimulates carotid body & causes reflex increase in respiration

Common Carotids

The CCA terminates at C4 level (upper border of thyroid cartilage & then divides into ECA & ICA.

The sternocleidomastoid muscle should be relaxed & the head rotated slightly towards the examiner. The carotid pulse should be palpated in the lower half of the patients neck to avoid inadvertant carotid sinus compression.

Right thumb is applied to patients' LCCA, left thumb is separately applies to patients' RCCA. This technique permits comfortable application of the thumb without awkward bending of the wrist, a maneuver that decreases sensitivity in the finger tips.

External Carotid

The ECA is palpated medial to the sternocleidomastoid above upper border of the thyroid cartilage

Internal Carotid

The Internal Carotids are palpated placing a hand in the mouth and palpating the tonsillar fauces.

Subclavian

The subclavian artery is felt in the anterior & inferior angle of the posterior triangle. It can be effectively compressed against the first rib with the shoulder depressed, pressure is exerted down back and medially in the angle between sternocleidomastoid and clavicle.

Brachial

Midway in the arm medial to the humerus pressure should be exerted slightly posterolaterally. Palpation of the right brachial pulse is accomplished with the thumb of the examiners right hand as the patients arm lies supinated at his or her side. As the thumb explores the antecubital fossa for the brachial artery pulse the patient's elbow can rest in the palm of the examiners right hand while the free left hand passively raises & lowers the patients forearm to achieve maximum relaxation of muscles around the antecubital fossa.

Axillary

Axillary compression against the humerous.

Radial

For radial pulse palpation the patient's hand should be supinated & comfortably supported. The examiners thumb or tip of a single finger preferably the index is applied to the pulse.

In infants palpation of radial pulse has inherent limitations

1) Radial artery is very small

2) Padding of subcutaneous fat is more.

Femoral

Mid Inguinal Ligament

Mid point of Pubic symphisis – Anterior Illiac spine

Mid Point Of Inguinal Ligament

Mid point of Pubic tubercle – Anterior Illiac spine

Femoral

At the mid inguinal point – Anterior superior Illiac spine to Pubic symphasis.

2cm below the junction of medial 1/3rd and lateral 2/3rd of inguinal ligament.

Popliteal

Patient prone, knee flexed, muscles relaxed by resting the leg on the examiners arm. Pulse is sought by firm pressure downwards against the popliteal fossa of the leg.

Posterior Tibial

Fingers breadth below and behind the medial malleoli.

Dorsalis Pedis

It is felt between the tendons of extensor hallucis longus and extensor digitorum at the dorsum of the foot over the base of the first metatarsal bone.

Pulse examination includes

1. Observation
2. Palpation
3. Auscultation

Pulse - Observation

Corrigans Pulse

AR

Dancing Brachialis (Serpentine Brachialis)

1) AR

2) Atherosclerosis

3) Arteriosclerosis

Pulse - Palpation

1) Rate

2) Rhythm

3) Volume

4) Character

5) Palpability of all peripheral accessible vessels

6) Thickening of vessels

7) Radiofemoral delay

8) Pulse deficit

9) Grading of Pulses

10) Peripheral signs of AR

Pulse - Auscultation

1) Supraclavicular region

Innocent Systolic murmur

2) Interscapular region

Coarctation of Aorta

3) Abdominal aorta & bifurcation

Iliofemoral obstruction

4) Flanks

Renal artery stenosis.

Grading of Pulses

GRADES 0 – 4+

1) GRADE 0 – Absent pulse
2) GRADE 1+ - feeble
3) GRADE 2+ - palpable but diminished
4) GRADE 3+ - normal
5) GRADE 4+ - high volume / bounding pulse

Abnormal Pulses

1) Pulsus Parvus
2) Pulsus Tardus
3) Hypokinetic Pulse
4) Hyperkinetic Pulse (Bounding)
5) Brisk or Jerky Pulse
6) Water Hammer Pulse
7) Collapsing Pulse
8) Corrigans Pulse
9) Anacrotic Pulse
10) Bisferiens Pulse
11) Dicrotic Pulse
12) Pulsus Paradoxsus
13) Pulsus Alternans
14) Pulsus Bigeminny

Pulsus Parvus

1) A slow rising pulse is Pulsus Parvus
2) It is a low volume pulse
3) Best appreciated in carotids
4) Seen in severe AS
5) Also seen in severe heart failure.

Pulsus Tardus(Anacrotic Pulse)

1) Pulse with late peaking
2) Peak is delayed nearer to second sound
3) Best appreciated by simultaneous auscultation of the heart and palpation of carotid pulse
4) Seen in all forms of fixed obstruction to the LV outflow.

Hypokinetic Pulse

Small or diminished pulse is called Hypokinetic pulse and the pulse pressure should be decreased.

1) Low CO
2) LV Dysfunction
3) CCF
4) Hypotension
5) LVOT Obstruction

In Hypokinetic pulse:-

Normal upstroke indicates decreased stroke volume

Slow upstroke indicates LVOT obstruction

Hyperkinetic Pulse

Hyperkinetic pulse has a larger than normal amplitude and results from

1) Increased LV ejection velocity
2) Increased Stroke volume
3) Increased arterial pressure.

It is seen in:-

1) Anxiety
2) Anaemia
3) Thyrotoxicosis
4) Exercise
5) Hot & humid environment
6) Alcohol intake
7) Cigarette smoking
8) SHT with Atherosclerosis
9) Isolated Systolic HT

Jerky Pulse

Jerky pulse is a pulse with a brisk or sharp upstroke that literally taps against the palpating fingers. The pulse volume is not increased. Rapid upstroke / Normal downstroke / Normal volume. Typically, the pulse of HCM is decribed as jerky pulse.

Collapsing (Or) Water Hammer Pulse

Thomas Watson (1844) coined the term after victorian toy.

Water Hammer was a popular toy in Victorian England and consisted of a sealed glass tube containing water in a vaccum. When the glass tube is quickly inverted, the water or mercurry falls abruptly from one end to the other and the finger tip holding the inverted end feels a sudden impact or jolt.

The collapsing pulse is due to:

i) Diastolic run off into the LV

ii) Reflex vasodilatation mediated by carotid baroreceptors secondary to large stroke volume

iii) Rapid run off to the periphery due to decreased systemic vascular resistance.

A Water Hammer pulse is best appreciated at the radial pulse with the palmar side of the examiner's hand and with the patient's arm suddenly elevated above the shoulder. This may be related to the artery becoming more in the line with the central aorta, allowing direct systolic ejection and diastolic backward flow.

Collapsing pulse can be seen in:-

- Conditions with aortic run off:
  AR, PDA, AP window, rupture of sinus of Valsalva into the right chambers and arteriovenous fistula.
- Cyanotic diseases like Truncus arteriosus with truncal run off in to PA or truncal insufficiency, Pulmonary atresia with Aortopulmonary collaterals, TOF with AP collaterals/associated PDA/ associated AR / after BT shunt.
- Hyperkinetic states
  Pregnancy, Anemia, thyrotoxicosis, Beriberi, Fever, Paget's disease of
  Bone
- Normal Volume Collapsing Pulse
  1) MR 3) VSD

Corrigans Pulse

The visible AR pulsation seen over carotid artery was called Corrigans Pulse by Dominic Corrigan.

Anacrotic Pulse

Ana: Up

Crotos: Beat

(Greek word)

Anacrotic notch is normally recorded on the ascending limb towards the end of rapid ejection phase of the arterial pulse curve. The rapid rising portion of the arterial pulse curve is termed anacrotic limb and indicates rapid ejection phase.

Anacrotic pulse is Pulsus parvus et tardus with accentuation of the anacrotic notch and a small volume pulse. Thus it is characterized by

1) Slow upstroke
2) Delayed peak
3) Small volume

It is well felt in the carotids

The anacrotic notch should occur in the early or mid portion of the ascending limb to be significant.

Earlier the anacrotic notch severe the stenosis

Two waves may be felt, anacrotic and percussion wave

In As there occurs flattening of dicrotic notch especially with valvular AS

In HT & MS a notch can occur but occurs high up in the ascending limb

Characteristics Of Anacrotic Pulse

1) Pulsus parvus
2) Pulsus tardus
3) Small volume
4) Prominent anacrotic notch
5) Anacrotic notch appears earlier
6) Dicrotic notch disappears

Bisferiens Pulse

Bis: Two

Feriere: To beat

Bisferiens in Latin means twice beating pulse.

In Bisferiens pulse there is a double systolic peak.

It is better felt in the Carotids. To appreciate the character of the pulse, either use graduated pressure technique or totally obliterate the pulse and then gradually release the pulse till two peaks are palpable.

The 2 peaks may be equal or either the first or second can be larger.

The 2 peaks can produce double Korotkoff sounds

Bisferiens nature disappears with failure.

Causes

- AR
- AR with AS
- HOCM
- Normal persons
- Hyperkinetic circulation

Mechanism of bisferiens pulse:

Normally percussion wave is felt but not the tidal wave. In all the conditions where percussion wave is prominent, tidal wave also becomes prominent. This mechanism is applicable in severe AR and other hyperkinetic conditions. In combined AS and AR, the stenotic component permits a jet, lateral to the jet, there is a fall in pressure(Bernoulli Phenomenon), this results in a dip or inward movement in the pulse with secondary outward movement in a pulse or tidal wave. Normally Both waves are prominent in patients with severe AR.

In HOCM, the initial part of left ventricular ejection is rapid, resulting in rapid upstroke. As obstruction to the outflow starts later in the systole, due to SAM, a sudden interruption to left ventricular ejection occurs resulting in a dip in the pressure pulse followed by the slow rising pulse wave, which

is characteristic of HOCM (spike and dome pattern). The percussion wave is more prominent than tidal wave in HOCM.

Dicrotic Pulse

Di: Twice

Crotos: beat

Greek word

Dicrotic pulse has an accentuated dicrotic wave and hence is a twice beating pulse, one in systole and one in diastole. The dicrotic notch is > 50% of pulse pressure.

For dicrotic pulse to occur

1) Hypotension

2) Reduced Peripheral Vascular Resistance is needed

Rarely present when BP > 130 mm/hg and in patients beyond 50 years of age.

When the reflection wave travels rapidly and meets the original wave well in advance, it is lost in it.

In rigid and nondistensible arterial system, as in SHT, dicrotic pulse in never present.

It is differentiated from the bisferiens pulse by the simultaneous auscultation of the heart sounds.

It is more noticeable in the beat following a premature ventricular contraction.

It is better appreciated during inspiration or inhalation of amyl nitrite.

Seen in

1) Healthy young adults

2) Fever

3) Hypovolumic Shock

4) CCF

5) Cardiac Tamponade

6) Sepsis

7) Aortic valve replacement.

Twice Beating Pulse

Anacrotic, Bisferiens, Dicrotic all 3 are twice beating pulse.

Differentiation

1)The double peaking occurs

A) On the upstroke in Anacrotic Both in Systole

B) On the peak in Bisferians

C) On the downstroke in Dicrotic One in Systole

One in Diastole

Anacrotic pulse is slow rising, late peaking and small volume.

Bisferiens pulse is rapid rising

Dicrotic pulse is normal rising.

In Anacrotic pulse first wave is of low amplitude (anacrotic notch) & second wave is large (Percussion wave)

In Bisferiens first wave is large (Percussion wave) & second wave is small (Tidal wave)

Pulsus Paradoxus

History

Pulsus Paradoxus was first seen by Adolph Kussmaul in a case of Constrictive Pericarditis

The terminology was first coined by him in 1873.

Paradoxus

Paradox about the pulse is absence of pulse during inspiration but presence of heart sounds & was coined by Adolph Kussmaul in 1873.

MISNOMER

Misnomer about the term paradoxus is that normally there is a fall in BP during inspiration (4-6mm/hg) which in PP is exaggerated (>10mm/hg)

Definition

Pulsus Paradoxus is an exaggerated fall in systolic BP with inspiration usually more than 10 mm/hg.

Pulsus Paradoxus is suspected if the pulse varies with inspiration in all accessible arteries.

Mechanism

Inspiratory increase in right heart filling and output was necessary for the development of PP. Inspiration causes increase in RV dimensions, & pulmonic and Tricuspid velocities & a decrease in LV dimensions and aortic and mitral velocities and LV diastolic compliance which results in a fall in systolic BP.

LV filling is reduced during inspiration because exaggerated RV filling causes

1) Leftward shift of IVS reducing LV volume & diastolic compliance

2) Elevated intrapericardial pressure which is transmitted to the LA but not the extraparenchymal pulmonary veins and hence a decreased pulmonary vein – LA pressure gradient

Inspiratory pooling of blood in the pulmonary bed produces decline in LA and LV filling.

Underfilled LV may be operating in the steep ascending limb of Starling curve so that any inspiratory reduction of LV filling results in marked depression of the LV stroke volume and the systolic pressure.

Measuring Pulsus Paradoxus

To detect pulsus paradoxus inflate the cuff rapidly above the systolic pressure and then slowly deflate it.

The difference of the systolic pressure at which sounds are first heard only during expiration and later during both expiration and inspiration is a measure of the magnitude of PP.

In normal persons deep inspiration but does not cause the radial pulse to disappear. Hence disappearance of the radial pulse on deep inspiration suggests significant PP

Pulsus Paradoxus - Causes

Physiological -

1) Obesity 2) Pregnancy

Respiratory Causes

3) Bronchial Asthma 4) Emphysema

5) COPD 6) Large Bilateral Pleural effusion

Cardiovascular Causes

7) Cardiac Tamponade 8) Constrictive Pericarditis (1/3rd)

9) Hypovolemic shock 10) Pulmonary embolism

11) RV Infarct 12) Cardiomyopathy

13) SVC Obstruction 14) Post Thoracotomy

Determinants Of PP

1) Venous return 2) LV afterload

3) Diastolic ventricular interdependence.

4) Lung volume.

5) Circulatory reflexes.

Echocardiographic Correlates Of PP

1) Exaggerated respiratory changes in diastolic ventricular dimensions

2) Leftward displacement of IVS during inspiration

3) Abnormal respiratory changes in aortic (Decreased opening) & Mitral (Decreased EF slope) movements.

4) Exaggerated respiratory changes in venous & transvalvular flow.

5) Decreased transmitral diastolic E/A ratio during inspiration.

Cardiac Cause

Inspiratory increase in venous pressure (Kussmauls sign)

Respiratory Cause

Expiratory increase in venous pressure.

Cardiac Tamponade Without PP

1) LVH 2) RVH 3) PHT 4) ASD 5) AR 6) Regional Tamponade

Mechanism for absence of PP is lack of competitive ventricular filling during inspiration.

Pulsus Paradoxus In COPD

The decrease in lung compliance magnifies the normal inspiratory decrease in LV volume and systolic arterial pressure and expiration may be accompanied by an excessive rise in systolic pressure above normal.

In COPD PP occurs when FEV1 < 1 lt.

Reversed PP

In Reversed Pulsus Paradoxus there is an increase in systemic pressure with inspiration seen in

1) HOCM : Mechanism unknown.

2) Isorhythmic AV dissociation : Atrial activity preceds QRS duration during inspiration and marches into QRS during expiration. The atrial activity during inspiration increases the stroke volume and its lack during expiration decreases the stroke volume and systolic pressure.

3) Intermittent positive pressure breathing : Intrathoracic pressure is higher during inspiration and lower during expiration.

Pseudo PP

Some patients with AV dissociation can have phasic swings in systolic blood pressure that mimics pulsus paradoxus.

Pulsus Alternans

Pulsus Alternans is a pulse pattern in which the beats occur at regular intervals but in which there is a regular attenuation of the systolic height of the pressure pulse.

It was first described by Traube in 1872.

Pulsus Alternans is a peripheral manifestation of LV failure

In Pulsus Alternans there is

1) Alteration in the height of the pressure pulse
2) alteration in the rate of rise.

It is the latter that is appreciated during palpation.

PA is better felt in distal vessels than proximal, reason being rate of rise & peak pressure developed are accentuated during peripheral transmission of the arterial pulse pressure.

Light pressure is applied to palpate Pulsus alternans.

Mild degree of PA is detected by sphygmomanometer. Inflate the BP cuff rapidly above systolic pressure and then deflate slowly until Korotkoffs sounds are audible. At this point beats are heard at one half of the heart rate. When the cuff is deflated further the rate doubles.

Pulsus Alternans - Mechanism

Pulsus alternans is due to alteration of the contractile state of at least part of the myocardium, caused by failure of electromechanical coupling in some cells during weaker contraction. A subsequent contraction then represents contraction of all cells some of which were potentiated.

How To Look For PA

1) Regular in rhythm

2) Should be felt in peripheral arteries

3) Light pressure should be applied.

4) Breath should be held in mid expiration

5) Can be brought out or exaggerated by decreasing venous return by

a) Sitting b) Standing c) Head up tilting or

Premature ventricular contractions, rapid atrial pacing, inferior vena caval occlusion, myocardial ischemia and intracoronary injection of contrast during coronary arteriography are known to induce alternans.

By infusion of nitroglycerine, Valsalva maneuver and in the presence of aortic regurgitation or systemic hypertension, pulsus alternans can be exaggerated.

6) It is usually associated with S3.

Pulsus Alternans – Causes

LVF

Myocariditis

Acute pulmonary embolism

Severe AS with failure

Severe PS with failure

Severe AR with failure specially after aortic valve replacement.

Briefly during or after supraventricular tachycardia

Severe systemic hypertension.

Transient right ventricular outflow occlusion during balloon dilatation of pulmonary stenosis.

Pulsus Alternans After VPC

When precipitated after a VPC the extent of difference of systolic pressure in alternate beats declines for several cycles.

This is due to increased duration of LV filling after the premature beat resulting in greater end-diastolic volume and hence increased contractile force due to Frank Starling mechanism.

Differentiating PA From Bigeminy

Pulsus Bigeminy caused by VPCs alternating with normal beats also stimulates Pulsus alternans

1) Pulsus Alternans is associated with LVS3
2) In PA the interval between the weak & strong beats are equal. In Pulsus Bigeminy the weaker beats arise prematurely and the stronger beats occur after a pause resulting in ventricular cycles that are alternatively short and long.

Radiofemoral Delay

Time taken by aortic pulse wave to reach

1) Carotids - 30 ms.
2) Brachials - 60 ms.
3) Femoral - 75 ms.
4) Radial - 80 ms.

When we say radiofemoral delay it is not the delayed arrival of the femoral pulse wave but instead a slow rate of rise to a delayed peak.

Causes

Coarctation of Aorta.

Occlusive disease of the bifurcation of the aorta, common iliac or external iliac arteries.

Coarctation of aorta with absent radio femoral delay:-

Coarctation of aorta + Bicuspid aortic valve with AS

Coarctation of aorta + Bicuspid aortic valve with AR

Coarctation of aorta with MR

Coarctation of aorta with Supravalvular AS

Pseudo Coarctation.

Right radio femoral delay:-

Supravalvular AS

Peripheral Signs Of AR

Head & Neck

1)	De Mussets sign	Head bobbing
2)	Light House Sign	Alternate flushing & blanching of face
3)	Landolfis sign	Alteration in pupillary size
3)	Quinckies sign	Capillary pulsation over lips
4)	Mullers sign	Uvula pulsation
5)	Carotid shudder	Thrill over carotid during upstroke
6)	Corrigans Pulse	Visible carotid pulse of AR
7)	Julians sign	Pulsation of retinal vessels.
8)	Minervini's sign	Strong lingual pulsations. Tongue depressor moves up and down when tongue is depressed.
9)	Logue's sign	Pulsation of sternoclavicular junction when AR is associated with aortic dissection.

Limbs

10)	Bisferiens Pulse	Double peaked Pulse
11)	Locomotor Brachi	Dancing Brachialis
12)	Hills sign	LL BP > 20 mmHg than UL
	Mild 20-40 mmhg	
	Moderate 40-60 mmHg, severe >60mmhg	
13)	Pistol shot Femoralis -	Systolic sounds over FA
14)	Traubes sign	Systolic & Diastolic sounds over femoral artery
15)	Duroziez's murmur.	Distal occlusion of femoral artery causes diastolic murmur and Proximal occlusion systolic murmur
16)	Palfrey's sign	Pistol shot sound over radial artery

Abdomen

17) Rosenbachs sign - Liver Pulsation

18) Gerhardts sign - Splenic Pulsation

19) Dennison's sign - Presence of pulsations in cervix

Differentiating Frequent VPC From AF By Pulse

VPC – 2 beats in quick succession followed by a large pause. (Normal beat followed by premature beat)

APC – 2 beats in quick succession followed by a short pause.

AF - Irregular in rate rhythm & force

Long pause that is not preceded by 2 beats in quick succession.

Pulse Deficit

Difference between apex beat and radial pulse > 10 beats/mt occurs in AF & VPC if they are too weak to open the aortic valve. Bruit over an artery

Less severe obstruction- short systolic bruit

More obstruction:-

- Continuous bruit
- High pitched bruit
- No bruit

Unequal Upper & Lower Limb Pulse

Coarctation of Aorta

Aortoarteritis

Dissection of Aorta

Atherosclerosis

Trauma

Unequal Carotids

Aortoarteritis

Dissecting aneurysm of Aorta

Atherosclerosis

Thromboembolic occlusion

Supravalvular AS

Unequal Radials

Aortoarteritis

Dissecting aneurysm of Aorta

Thromboembolic obstruction

Previous catheterization

Cervical rib

Scalenus Anticus syndrome

Anomalous Rt Subclavian artery

Aberrant course of Radial artery

Arteritis.

Absent Femorals

Dissecting aneurysm

Coarctation of aorta

Pseudoxanthoma elasticum

Hypoplastic External Iliac artery.

Hypertrophic cardiomyopathy:

1) Pulsus Bisferiens is common, whether or not there is associated obstruction. Bisferiens pulse in HCM has initial brisk upstroke, and the initial systolic wave is wider and of greater amplitude. The initial wave is followed by relatively wide second systolic wave, usually of lower amplitude than first, but may be of same amplitude too. In HCM characteristic pulse is probably due to ascending aortic flow which is biphasic, peak systolic flow followed by some flow in later systole, the second systolic wave corresponds to this late systolic flow, but it is also probably consequence of wave reflection from the first systolic impulse.
2) The trough between the two systolic waves corresponds in time with the falling limb of the aortic flow wave rather than its peak as in aortic stenosis and incompetence.
3) The timing of second wave is relatively constant in relation to first wave. When ejection is abbreviated following an extra systole, the second wave may appear in diastole and generate diastolic pulse. This type of phenomenon is never seen in Bisferiens pulse of aortic stenosis and incompetence.

Aortic incompetence:

1) High volume pulse with a rapid upstroke and rapid decline in diastole.
2) Incisura is poorly defined.
3) The systole is often prolonged (more than 300msec) and pressure wave is only slightly altered between the central and the peripheral arteries.

Aortic Stenosis:

1) Pulsus parvus et tardus (slow rising and small pulse). Pulse pressure is low.
2) Pulse is described as anacrotic pulse, with attention directed to the initial peak on the upstroke, the subsequent slow rise, and later systolic peak. The anacrotic pulse also has been used to describe the pulse in HT and arteriosclerosis by some authors, but in these situations, the initial peak is high on the first part of the wave, the initial upstroke is normal, and pulse pressure is high.
3) The severe the stenosis lower the anacrotic notch
4) Characteristically well felt in carotids.
5) Severity is related to the rate of rise of the pulse during systole and to the amplitude of the pulse.
6) Amplification of pulse is decreased between central and peripheral pulse.
7) In severe AS, incisura becomes indistinct.

Aortic stenosis and incompetence:

1) Bisferiens pulse. The mechanism appears to be venturi effect
2) The first systolic peak is always lower than the second systolic peak, which differentiates it from the bisferiens pulse of HCM.
3) Carotids are best sites for detecting bisferiens pulse.

N.B. If the arterial pulse is regular in a patient with established atrial fibrillation on digitalis therapy, digitoxicity with AV nodal rhythm should be considered.

High volume pulse in MS occurs with aortic regurgitation, anemia or systemic hypertension.

Causes of Rapid Regular pulse

1) Sinus tachycardia
2) Supraventricular tachycardia
3) Paroxysmal atrial tachycardia
4) Junctional tachycardia
5) Atrial tachycardia with fixed block
6) Atrial flutter with fixed block
7) Ventricular tachycardia
8) Atrial fibrillation
9) Atrial flutter with varying block
10) Atrial tachycardia with varying block
11) Multifocal ventricular tachycardia
12) AF with WPW syndrome
13) Frequent multifocal atrial and ventricular ectopy

Causes of Bradycardia

1. Sinus bradycardia
2. Complete heart block
3. High grade heart block
4. Bigeminal rhythm with impalpable premature beat
5. Pulsus alternans with impalpable weak beat

THE ARTERIAL BLOOD PRESSURE

Dr. Mullasari Ajit S., Dr. Rajendra Deshmukh

The arterial BP, a measure of lateral force per unit area of vascular wall, is quantitated as millimeters of mercury (mmHg) or dynes per square centimeter (d/cm^2).

History:

In the nineteenth century, the invention by Jules Herisson of the sphygmograph to measure blood pressure non-invasively and its further refinements by Karl Vierordt, Etienne Jules Marey, F. A. Mahomed and others has evolved into a clinically useful bedside device.

In 1905, Dr. Nikolai Korotkoff (1874-1920), a Russian surgeon, discovered a simple and precise technique to measure arterial pressure.

Korotkoff described four distinct phases of sounds: first sound, then compression murmurs, second sound, and disappearance of sounds. Korotkoff was also able to demonstrate the same auscultatory finding in healthy persons. He failed to notice only the muffled second sound, which was demonstrated a little later.

Factors responsible for the peak systolic BP:

1. Volume and velocity of LV ejection
2. The peripheral arteriolar resistance
3. The distensibility of the arterial wall
4. The viscosity of the blood
5. The end-diastolic volume in the arterial system.

Factors responsible for the diastolic BP:

1. Blood viscosity
2. Arterial distensibility
3. Peripheral resistance to flow
4. Length of the cardiac cycle

Measurement of blood pressure:

1. Direct methods
2. Indirect methods

Direct method: Use the electromanometer, a transducer that converts mechanical energy into an electric signal. The artery is cannulated with a saline-filled catheter or needle that mechanically couples the circulation to the arterial manometer. Pressures are recorded using atmospheric pressure as the *zero* reference level and intravascular pressures are further referenced to the level of the heart by addition or subtraction of a gravitation factor.

Indirect methods: For the noninvasive evaluation of arterial BP, a pneumatic cuff with a mercury or aneroid manometer is the most frequently used technique.

Important Aspects of Blood Pressure Measurement:

Patient should

- Sit or lie quietly for at least 5 minutes before the measurement is taken.
- Seated comfortably, back supported, bared upper arm, legs uncrossed
- Arm should be at heart level - the mid-point of the sternum
- Cuff length/width should be 80%/40% of arm circumference
- Cuff should be deflated at <3 mm Hg per second
- Column or dial should be read to nearest 2 mm Hg
- First audible Korotkoff sound is systolic pressure; last sound is diastolic pressure
- No talking between subject and observer

Measurements made while the patient is on an examining table do not fulfill these criteria and should preferably be made while the patient is seated in a chair.

Factors causing significant deviations in measured blood pressure: Room temperature, exercise, alcohol or nicotine consumption, positioning of the arm, muscle tension, bladder distension, talking, and background noise.

Choice of Blood Pressure Measurement Devices

The "gold standard" device for office blood pressure measurement is the mercury sphygmomanometer.

Cuff size:

An undersized cuff - falsely high readings, and An oversized cuff - falsely low readings

The recommended cuff sizes are:

- Arm circumference 22 to 26 cm, 'small adult' cuff, 12 x 22 cm
- Arm circumference 27 to 34 cm, 'adult' cuff: 16 x 30 cm
- Arm circumference 35 to 44 cm, 'large adult' cuff: 16 x 36 cm
- Arm circumference 45 to 52 cm, 'adult thigh' cuff; 16 x 42

Effects of Body Position:

Blood pressure measurement is most commonly made in either the sitting or the supine position.

Sitting position- diastolic pressure is higher than when measured supine (by 5 mm Hg), although there is less agreement about systolic pressure.

Supine position: The systolic pressure is 8 mm Hg higher in the supine than the upright position.

If the back is not supported (as when the patient is seated on an examination table as opposed to a chair), the diastolic pressure may be increased by 6 mm Hg.

Crossing the legs: May raise systolic pressure by 2 to 8 mm Hg

In the supine position, the right atrium is approximately halfway between the bed and the level of the sternum; thus, if the arm is resting on the bed, it will be below heart level. For this reason, when measurements are taken in the supine position the arm should be supported with a pillow. In the sitting position, the right atrium level is the midpoint of the sternum or the fourth intercostal space.

Effects of Arm Position:

If the upper arm is below the level of the right atrium- High BP

If the upper arm is above the level of the right atrium- low BP

Difference: 2 mm Hg for every inch above or below the heart level

These differences can be attributed to the effects of hydrostatic pressure

Differences Between the 2 Arms:

Blood pressure should be measured in both arms either in rapid succession or simultaneously; normally the measurements should differ by <10 mm Hg, independent of handedness. As many as 20 percent of normal subjects, however, have a >10 mm Hg arm blood pressure difference.

A blood pressure difference of >10 mm Hg can be associated with-

- Subclavian artery disease (atherosclerotic or inflammatory),
- Supravalvular aortic stenosis,
- Aortic coarctation
- Aortic dissection.

Differences Between arms and legs: Systolic leg pressures may be as much as 20 mm Hg higher than arm pressures.

Greater leg-arm pressure differences are seen in patients with-

- Severe AR (Hill sign) and
- Patients with extensive and calcified lower peripheral arterial disease (PAD)
- Aortic dissection
- Coarctation of aorta

When there is a consistent interarm difference, the arm with the higher pressure should be used.

Cuff Placement and Stethoscope:

- First palpate the brachial artery
- The lower end of the cuff should be 2 to 3 cm above the antecubital fossa to allow room for placement of the stethoscope
- Pull the cuff snugly around the bare upper arm
- phase 1 (systolic) and phase 5 (diastolic) Korotkoff sounds are best heard using the bell of the stethoscope over the palpated brachial artery in the antecubital fossa.

Traditionally, the sounds have been classified as 5 phases:

- Phase I- appearance of clear tapping sounds corresponding to the appearance of a palpable pulse
- Phase II,-sounds become softer and longer
- Phase III- sounds become crisper and louder
- Phase IV- sounds become muffled and softer
- Phase V- sounds disappear completely. The fifth phase is thus recorded as the last audible sound.

The sounds are thought to originate from a combination of turbulent blood flow and oscillations of the arterial wall. There is agreement that the onset of phase I corresponds to systolic pressure but tends to **underestimate** the **systolic pressure** recorded by direct intra-arterial measurement. The disappearance of sounds (phase V) corresponds to diastolic pressure but tends to occur before diastolic pressure determined by direct intra-arterial measurement. No clinical significance has been attached to phases II and III.

The Korotkoff sound method tends to give values for systolic pressure that are lower than the true intra-arterial pressure, and diastolic values that are higher.

There has been disagreement in the past as to whether phase IV or V of the Korotkoff sounds should be used for recording diastolic pressure, but phase IV tends to be even higher than phase V when compared against the true intra-arterial diastolic pressure and is more difficult to identify than phase V. There is now general consensus that the fifth phase should be used, except in situations in which the disappearance of sounds cannot reliably be determined because sounds are audible even after complete deflation of the cuff.

Use of phase IV as a diastolic pressure:

- Pregnancy
- Arteriovenous fistulas (eg, for hemodialysis)
- Aortic insufficiency

Most of the large-scale clinical trials that have evaluated the benefits of treating hypertension have used the fifth phase

Inflation/Deflation:

The cuff should initially be inflated to at least 30 mm Hg above the point at which the radial pulse disappears.(THIS AVOIDS THE AUSCUTATORY GAP) The rate of deflation has a significant effect on blood pressure determination. Deflation rates >2 mm per second can lead to a significant underestimation of systolic and overestimation of diastolic blood pressure. It is recommended that a deflation rate of 2 to 3 mm Hg per second.

Number of Measurements:

When a series of readings is taken, the first is typically the highest. A minimum of 2 readings should be taken at intervals of at least 1 minute, and the average of those readings should be used to represent the patient's blood pressure. If there is >5 mm Hg difference between the first and second readings, additional (1 or 2) readings should be obtained, and then the average of these multiple readings is used.

Automated Methods

When they are used in the office, the readings are typically lower than readings taken by a physician or nurse.

Advantages :

- Elimination of observer error
- Minimizing the white coat effect
- Increasing the number of readings

Disadvantages:

- The error inherent in the oscillometric method
- The fact that epidemiologic data are mostly based on auscultated blood pressure measures.

Required Competencies:

Vision- The observer must be able to see the dial of the manometer or the meniscus of the mercury column at eye level without straining or stretching, and must be able to read well enough to see the sphygmomanometer or digital display no further than 3 feet away.

Hearing- The observer must be able to hear the appearance and disappearance of Korotkoff sounds.

Eye/hand/ear coordination- This is required for the use of mercury and aneroid sphygmomanometers but not for the newer electronic technologies.

Blood Pressure Recording in Special Situations:

Elderly Patients:

Blood pressure should also be taken in the standing position routinely because the elderly may have postural hypotension.

Pulseless Syndromes:

Rarely, patients present with occlusive arterial disease in the major arteries to all 4 limbs (eg, Takayasu arteritis, giant cell arteritis, or atherosclerosis) so that a reliable blood pressure cannot be obtained from any limb. In this situation, if a carotid artery is normal, it is possible to obtain retinal artery systolic pressure and use the nomogram in reverse to estimate the brachial pressure (oculoplethysmography), but this procedure and the measurement of retinal artery pressures are not generally available.

Arrhythmias:

When the cardiac rhythm is very irregular, the cardiac output and blood pressure varies greatly from beat to beat. The blood pressure should be measured several times and the average value used. If severe regular bradycardia is present (eg, 40 to 50 bpm), deflation should be slower than usual to prevent underestimation of systolic and overestimation of diastolic blood pressure

Obese Patients:

A longer and wider cuff is needed for adequate compression of the brachial artery in the obese patient with a very large upper arm. The error of overestimating the pressure when measuring with a cuff that is too small for an obese arm can be considerable and can lead to misclassification of an individual as hypertensive and to unnecessary concern and therapy.

Children:

Blood pressure is most conveniently measured in children by auscultation with a standard mercury sphygmomanometer. Select a cuff that has a bladder width that is at least 40% of the arm circumference midway between the olecranon and the acromion. This will usually be a cuff bladder that will cover 80% to 100% of the circumference of the arm.

- Newborn–premature infants: a cuff size of 4-8 cm
- Infants: 6-12 cm; and
- Older children: 9-18 cm.

Systolic blood pressure is determined by the onset of the auscultated pulsation or first Korotkoff sound. The phase of the Korotkoff sounds that defines diastolic blood pressure has been somewhat controversial.

Pregnant Women:

Hypertension is the most common medical disorder of pregnancy and occurs in 10% to 12% of all pregnancies. The International Society for the Study of Hypertension in Pregnancy currently

recommends using K5 for the measurement of diastolic blood pressure in pregnancy. When sounds are audible with the cuff deflated, K4 should be used.

Diabetes Mellitus: Present recommendations suggest the blood pressure in diabetics be controlled to 130/80, and if renal disease or microalbuminuria is also present the pressure should be 120/70 or lower.

Classification/Subtypes of Hypertension:

The classification is based on the average of >2 seated blood pressure measurements, properly measured with well-maintained equipment, at each of >2 visits to the office or clinic.

JNC 7 :

Isolated Systolic Hypertension: When the average systolic blood pressure is >140 and diastolic blood pressure is <90, the patient is classified as having isolated systolic hypertension.

Isolated Diastolic Hypertension: More commonly seen in some younger adults, isolated diastolic hypertension is defined as a systolic pressure <140

and a diastolic >90.

White-Coat Hypertension or Isolated Office Hypertension: In 15% to 20% of people with stage 1 hypertension, blood pressure may only be elevated persistently in the presence of a health care worker, particularly a physician. When measured elsewhere, including while at work, the blood pressure is not elevated. When this phenomenon is detected in patients not taking medications, it is referred to as white-coat hypertension (WCH).

The commonly used definition is a persistently elevated average office blood pressure of >140/90 and an average awake ambulatory reading of <135/85 mm Hg. In some patients, WCH may progress to definite sustained hypertension, and all need to be followed-up indefinitely with office and out-of-office measurements of blood pressure.

White coat hypertension has been defined and classified into three groups:

1. White coat hypertension: Abnormal office systolic blood pressure >150 mmHg and daytime average systolic blood pressure <140 mmHg. (Patients not on antihypertensives).
2. White coat syndrome—normotensive: Patients' blood pressures controlled on antihypertensives. Their daytime average systolic blood pressure <140 mmHg and office blood pressure reading of >150 mmHg.
3. White coat syndrome—hypertensive: Patients may be on or off antihypertensive medications with daytime average systolic blood pressure of >140 mmHg and office systolic blood pressure measurement of >150 mmHg, which is at least 15 mmHg higher than the average daytime systolic blood pressure.

Masked Hypertension or Isolated Ambulatory Hypertension:

Somewhat less frequent than WCH but more problematic to detect is the converse condition of normal blood pressure in the office and elevated blood pressures elsewhere, eg, at work or at home. Lifestyle can contribute to this, eg, alcohol, tobacco, caffeine consumption, and physical activity away

from the clinic/office.

Pseudohypertension:

When the peripheral muscular arteries become very rigid from advanced (often calcified) arteriosclerosis, the cuff has to be at a higher pressure to compress them. The brachial or radial artery may be palpated distal to the fully inflated cuff in these instances (positive Osler sign). When suspected, an intra-arterial

radial artery blood pressure can be obtained for verification. It was present in 7.2% of 3387 persons older than 59 years screened for the Systolic Hypertension in the Elderly Program (SHEP) study.

Orthostatic or Postural Hypotension:

Orthostatic hypotension is defined as a reduction of systolic blood pressure of at least 20 mm Hg or 10 mm Hg in diastolic blood pressure within 3 minutes of quiet standing. An alternative method is to detect a similar fall during head-up tilt at 60 degrees.

Pheochromocytoma:

The great majority (e"90%) of patients have essential or primary hypertension. A history of the "5 P's" (ie, paroxysmal hypertension with postural hypotension, head pain, palpitations, pallor, and perspiration) in a thin, anxious, hypermetabolic person with pheochromocytoma (rememberthere are "no fat pheo's").

Coanda effect:

In supravalvular aortic stenosis the direction of the jet of flow tends to be directly directed into the innominate artery. This often results in the direct impact pressure of the central jet being transmitted to the right arm, thus making the right arm pressure higher than the left due to a Coanda effect.

The Ankle-Brachial Index

The ankle-brachial index (ABI) is the ratio of the systolic blood pressure at the ankle divided by the higher of the two arm systolic blood pressures.

It reflects the degree of lower-extremity arterial occlusive disease, which is manifest by reduced blood pressure distal to stenotic lesions. Either the posterior tibial or dorsalis pedis artery pressures can be used. An arm systolic pressure of 120 mmHg and an ankle systolic pressure of 60 mmHg yields an ABI of 0.5 (60/120).

The ABI is inversely related to disease severity. A resting ABI <0.9 is considered abnormal.. An ABI <0.3 is consistent with critical ischemia, rest pain, and tissue loss. For screening purposes, ABI is usually measured only at rest. However, a resting ABI >0.9 does not exclude significant PAD.

Pulsus Paradoxus:

A normal person can exhibit a 10 mmHg drop in systolic pressure during normal inspiration. A greater decline can be identified in patients with acute cardiac tamponade, constrictive pericarditis, severe obstructive lung disease, and restrictive cardiomyopathy.

Pulsus paradoxus is best detected by inflating the BP cuff above systolic pressure and then slowly releasing it. As the cuff pressure is gradually reduced, the BP sounds become audible during expiration. The difference in pressure between the first audible sound heard on expiration and the pressure level at which the sounds are heard during all phases of respiration gives a measurement of magnitude of pulsus paradoxus.

Increased Pulse Pressure

Physiological: Fever, anemia, hot weather, exercise, pregnancy, hyperthyroidism, or arteriovenous fistulas

Pathological : AR, patent ductus arteriosus, and truncus arteriosus

Reduced Pulse Pressure

Increased peripheral resistance (increased circulating catecholamines in heart failure), decreased stroke volume (severe as in arteriosclerosis [AS]), and/or markedly decreased intravascular volume (diabetic ketoacidosis)

JUGULAR VENOUS PULSE

Dr. K. Latchumanadhas, Dr. Kalaichelvan

Introduction:

- The venous system contains about 70 – 80% of the circulating blood volume which is non pulsatile.
- However, changes in flow and pressure caused by the right ventricular and right atrial filling produce pulsation in the central vein that are transmitted to the peripheral venous system (Jugular venous system).
- The jugular vein pulse waves and pressure provide events from the right side of the heart – right atrial pressure during systole and right ventricular filling pressure during diastole.
- Hence, an accurate assessment of the venous pulse, the jugular venous pulse (JVP) reflects the dynamics of the right sided of the heart.

History:

- Lancis (1728) first described the venous pulse of external jugular vein.
- The classic graphic recording of the JVP were done by chaurea and marey (1863).
- Potain (1869) who accurately described the wave pattern in the internal jugular vein.
- Subsequently, James Mackenzie (1902) provided nomenclature of JVP and applied this principle of examination at the beside.
- In 1950, Paul wood rekindled this interest of assessment of JVP.

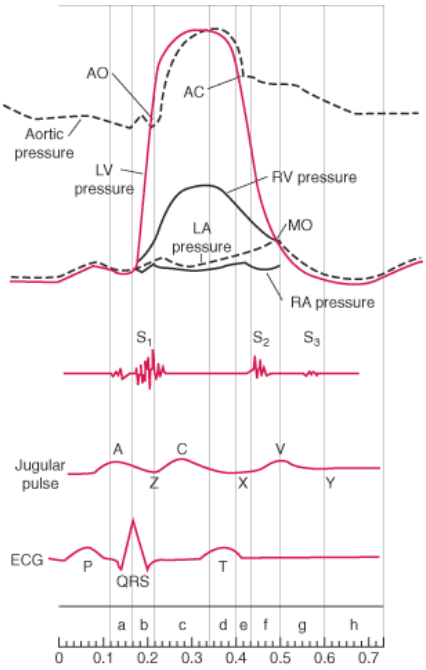
Jugular venous system:

- The jugular venous system is in direct continuity with right atrium and right ventricular in diastole. During systole, the tricuspid valve is closed and therefore the ventricle is excluded from this system.
- RA pressure during systole and RV filling pressure during diastole are producing pulsation and pressure waves in jugular veins.
- Right atrial pressure is transmitted through the superior vena cava to the internal jugular vein. The internal jugular vein runs underneath the sternocleidomastoid muscle and jugular pulsations are transmitted to overlying skin.
- It extends in direction from angle of the jaw to the hollowness between the heads of the sternocleidomastoid muscle attachments to the upper sternum and medial portion of the clavicle.
- Since RA pressure waves (a and v waves) have slow rises and are often of low amplitude, they are usually not well appreciated in jugular veins.
- On the other hand, the descents in the RA pressure pulse are better transmitted and appreciated in the jugular veins.
- The descents in the internal jugular vein reflect a fall in pressure in right atrium during cardiac systole and diastole.
- The descents are generally rapid movements, moving away from the eye and thus easily seen.

Figure 1: Cardiac Cycle

Right IJV is preferred for examination of JVP:

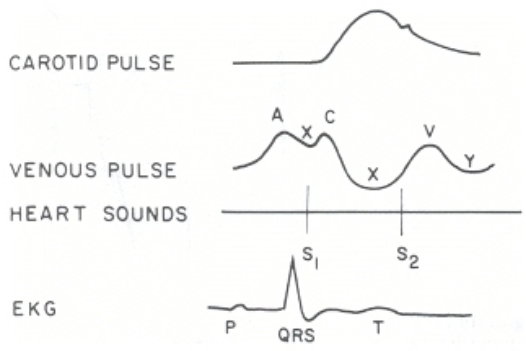
- Right IJV is usually assessed both for wave from and estimation of central venous pressure.
- Right IJV is in direct continuity with SVC and right atrium. It has straight line course through SVC to right atrium.
- Left IJV drains in to left innominate vein, which is not in straight line from RA.
- Right IJV and innominate vein is not compressed by adjacent structures in the neck and thorax.
- There are no or less numbers of valves in IJV than EJV.
- Less impact of vasoconstriction on IJV due to sympathetic activity than in external jugular vein.



Difference between IJV and carotid pulses:

Internal Jugular Vein	Carotid pulse
Superficial and lateral in the neck	Deeper and medical in neck
Better seen than felt	Better felt than seen
Has two peaks and two troughs	Has single upstroke only
Descents more obvious than crests	Upstroke brisker and visible
Gradual upstroke and dramatic collapse	Dramatic upstroke and gradual collapse
JVP falls during inspiration	Does not change with respiration
Abdominal compression elevates jugular pressure	Abdominal compression has no effect on carotid pulse
Mean JVP falls during standing	Does not changing with standing
Digital compression abolish jugular pulse	No effect on carotid pulse

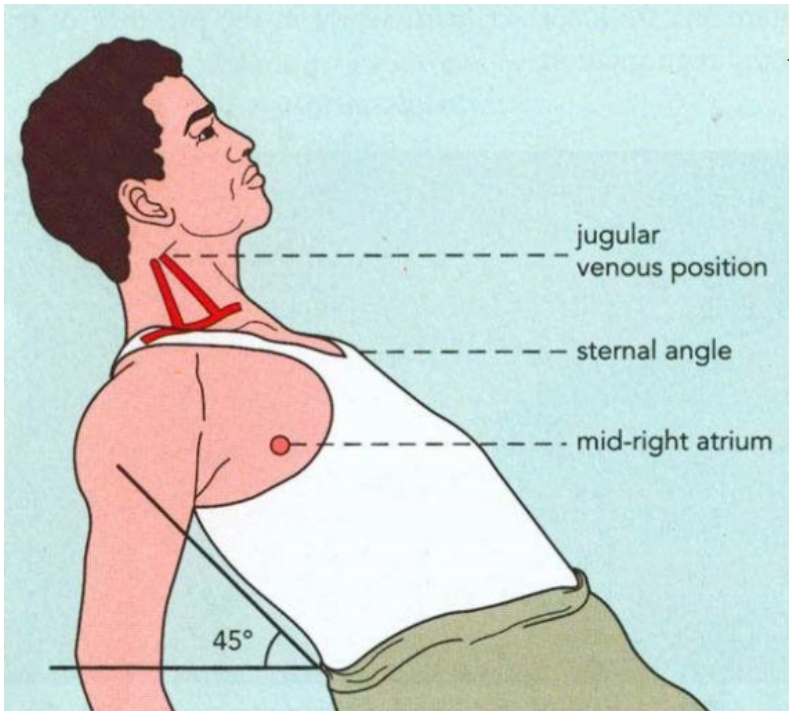
Figure 2: Timing of JVP in comparison with carotid pulse, heart sounds and ECG



Examinations of JVP (Lewis method):

- Patient should lie comfortably and trunk is inclined by 45° position.
- Elevates chin and slightly rotate the head to the left.
- Inclination angle should be subtended between trunk and bed, while neck and trunk should be in same line.
- When neck muscles are relaxed, shining the light tangentially over the skin and see top of the venous column and venous pulsations.
- In patients with low jugular pressure, a lesser (<30°) inclination is desirable.
- In patients with high jugular pressure, a greater (60 – 90°) inclination is required to obtain visible pulsation.
- Simultaneous palpation of left carotid artery and / or apical impulse aids in timing of the venous pulsations in cardiac cycle.

Figure 3: Positioning of the patient in JVP Exam

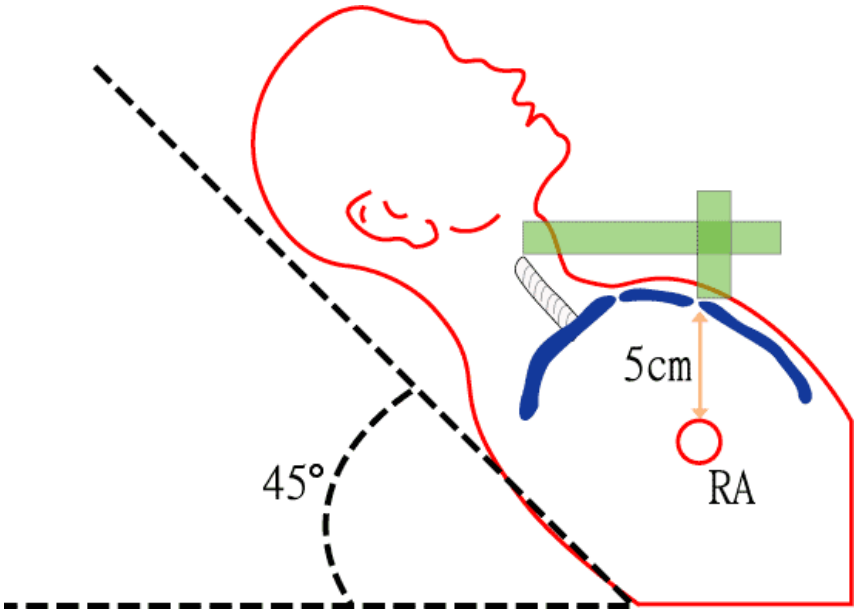


Measurement of JVP:

- Sternal angle or angle of Lowis is a surface anatomical mark, is used as a reference point for JVP measurement (Paul wood).
- Sternal angle is found approximately 5 cm above the center of the right atrium.
- Distance between sternal angle and center of right atrium remain relatively constant regardless of position of the thorax.
- Use sternal angle as zero reference point.
- For measuring of mean jugular venous pressure, first identify 'z' point in the jugular venous column.
- Two-scale method is commonly used.
- A horizontal scale at the top of oscillating venous column (z point) in IJV cuts the vertical scale kept at the sternal angle gives jugular venous pressure in cm of water.
- Normally jugular venous pressure does not exceeds 4 cm above the sternal angle.

- Since RA is approximately 5 cm below the sternal angle, the jugular venous pressure (RA mean pressure) is corresponds to 9 cm.
- By way to conversion, normal mean jugular venous pressure does not exceed 7 mm Hg (9 cm column of water / 1.3 = 6.9).
- Elevated JVP means that the jugular venous pressure of > 4 cm above sternal angle.

Figure 4 - Measurement of JVP:



Normal JVP waves:

- JVP reflects the phasic pressure changes in RA during systole and RV during diastolic part of cardiac cycle.
- Two visible positive waves (a and v) and two negative troughs (x and y) were seen in JVP.
- Two additional positive waves can be recorded. 'c ' wave interrupts x - descent and 'h' wave precedes the next a wave.

Presystolic ' a' wave in JVP:

- First positive presystolic 'a' wave is due to right atrial contraction results in retrograde blood flow into SVC and jugular veins.
- Effective RA contraction is needed for visible a wave.
- This is dominant wave in JVP and large than v wave.
- It precede upstroke of the carotid pulse and S1 but follow the P wave in ECG .
- It usually correspond to fourth heart sound in cardiac cycle.

'x' - descent in JVP:

- Systolic x - descent (systolic collapse) is due to atrial relaxation during ventricular systole.
- x-descent is most prominent motion of normal JVP which begin during systole and ends just before S2.
    - It is large than y - descent
    - x - descent more prominent during inspiration.

'C' Wave in JVP:

- Second positive wave recorded in JVP which interrupts the x-descent.
- c wave is produced by
  Carotid artery impact on JVP
  Upward bulging of closed TV in to RA during isovolumetric contraction.

'X' descent in JVP:

- x′ descent is systolic trough after c wave
- x′ descent is due to fall of RA pressure during early systole causing downward pulling of tricuspid valve by contracting right ventricle .
- Descent of RA floor by contracting RV also producing x′ descent.

'V' wave in JVP:

- Third positive wave in JVP which begins in late systole and ends in early diastole.
- Rise in RA pressure due to continued RA filling during ventricular systole when tricuspid valve closed.
- It is roughly synchronous with carotid upstroke and corresponds to S2.

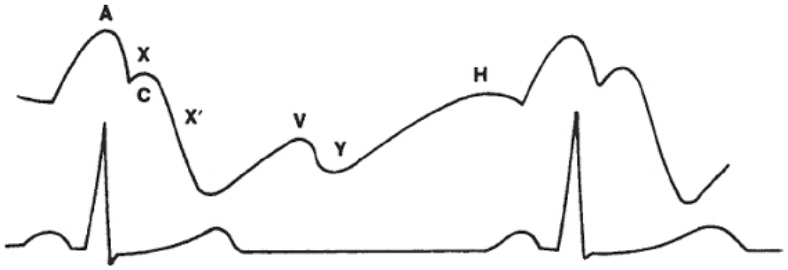
'Y' – descent in JVP:

- Diastolic collapse wave (down slope of v wave)
- It begins and ends during diastole well after S2.
- Decline of RA pressure due to RA emptying during early diastole when tricuspid valve opens causing y descent.
- Initial y descent is corresponds to rapid RV filling and later part of y descent is produced by continued diastolic inflow into RV.

'h' wave in JVP:

- Small brief positive wave following y - descent just prior to a wave during period of diastasis.
- 'h' wave is first described by Hieschfelder in 1907
- It usually seen when diastole is long (as in slow heart rates)
- With increasing heart rate, y - descent is immediately followed by next a wave.

Figure 5 – Normal JVP waves:



Prominent 'a' wave in JVP:

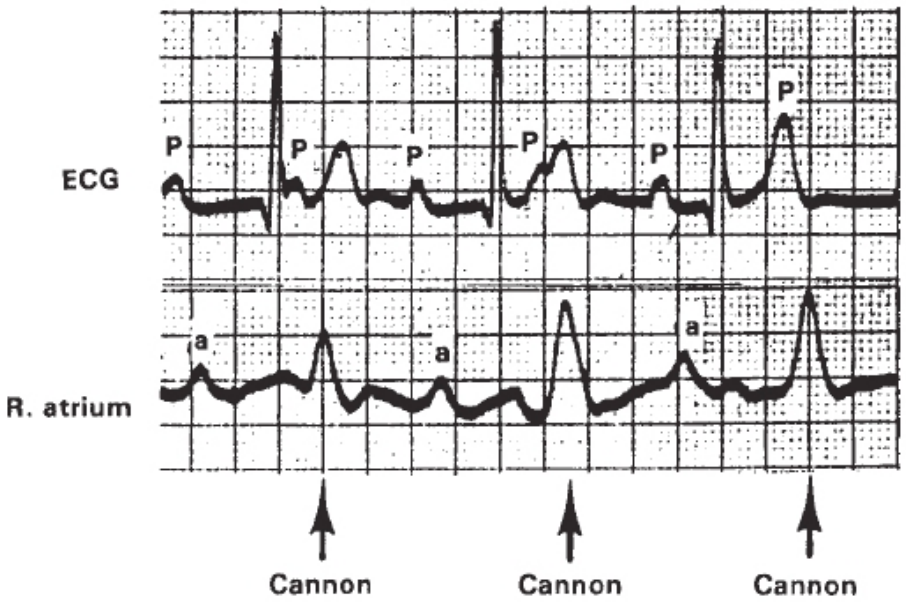
- Forceful atrial contraction when there is resistance to RA emptying or increased resistance to ventricular filling lead to prominent 'a' wave in JVP.
- RV inflow obstruction:
  Tricuspid stenosis or atresia

  RA myxoma
- Decreased ventricular compliance
  Associated with increased resistance to ventricular filling
  Force RA contraction is need to over come the elevated filling pressure in right ventricle lead to prominent 'a' wave in JVP.
- Condition associated with decreased ventricular compliance:
  Pulmonary stenosis
  PHT of any cause
  RV infarction
  RV cardiomyopathy (including HOCM)
  Acute pulmonary embolism

Cannon waves:

- Whenever RA contracts against closed tricuspid valve during RV systole, the prominent systolic cannon wave seen in JVP.
- Cannon wave is also called as venous Corrigan by Paul wood.
- Regular cannon waves:
  Junctional rhythm
  VT with 1:1 retrograde VA conduction
  Isorhythmic AV dissociation.
- Irregular cannon waves:
  Complete heart block
  Classic AV dissociation
  Ventricular pacing or ventricular ectopics.

Figure 6 – Cannon wave in CHB:



Absent 'a' wave in JVP:

- 'a' wave become absent when there is no effective atrial contraction as in AF.
- In Atrial fibrillation, both atrial contraction and atrial relaxation become ineffective .
- In sinus tachycardia, when a wave may fuse with preceding v wave, a wave become absent.

Prominent 'x' - descent in JVP:

- Presence of atrial relaxation with intact tricuspid valve and good RV contraction is need for prominent x descent.

- Causes of prominent x - descent:
  PHT with compensated RV function
  Constrictive pericarditis
  Early pericardial effusion
  Atrial septal defect

Absent 'x' - descent in JVP:

- Absent x - descent is early sign of moderate to severe TR due to obliteration of x - descent with early build up of large systolic wave during ventricular systole (cv wave).

- Absent x - descent is also seen in decompensated RV dysfunction with loss of effective RV contraction as in PHT with RV failure and Severe RV infarction.

- Chronic atrial fibrillation : loss of atrial relaxation lead to absent of x descent.

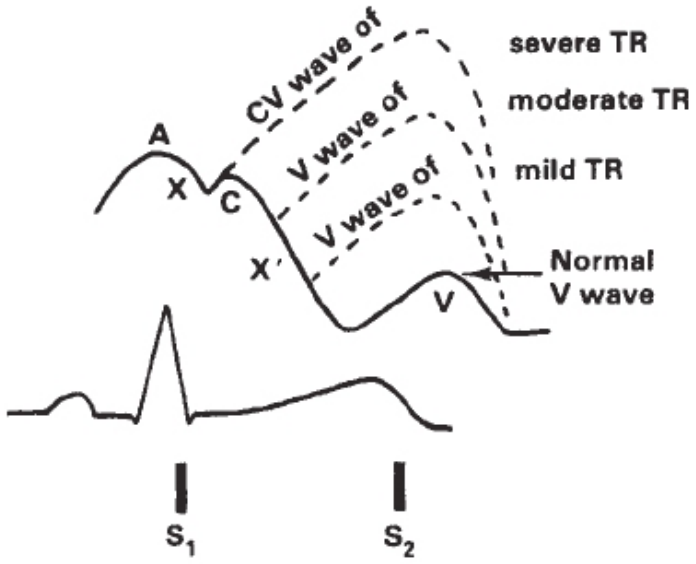
Prominent 'v' wave in JVP:

- Increased RA volume during ventricular systole produce prominent v wave in JVP.

- Severe tricuspid regurgitation can cause giant v wave – Lanci sign (ventricularisation of atrial pressure).

- Giant 'v' waves sometimes causes:
  Systolic movement of ear lobe
  Head bobbing with each systole
  Systolic pulsation of liver
  Pulsatile exophthalmos
  Pistol shots heard over IJV.

- Prominent 'v' wave in JVP:
  Tricuspid regurgitation
  ASD with mitral regurgitation
  Severe congestive heart failure
  VSD of LV to RA shunt (gerbode's detect)
  Corpulmonale
  Atrial fibrillation with CCF.



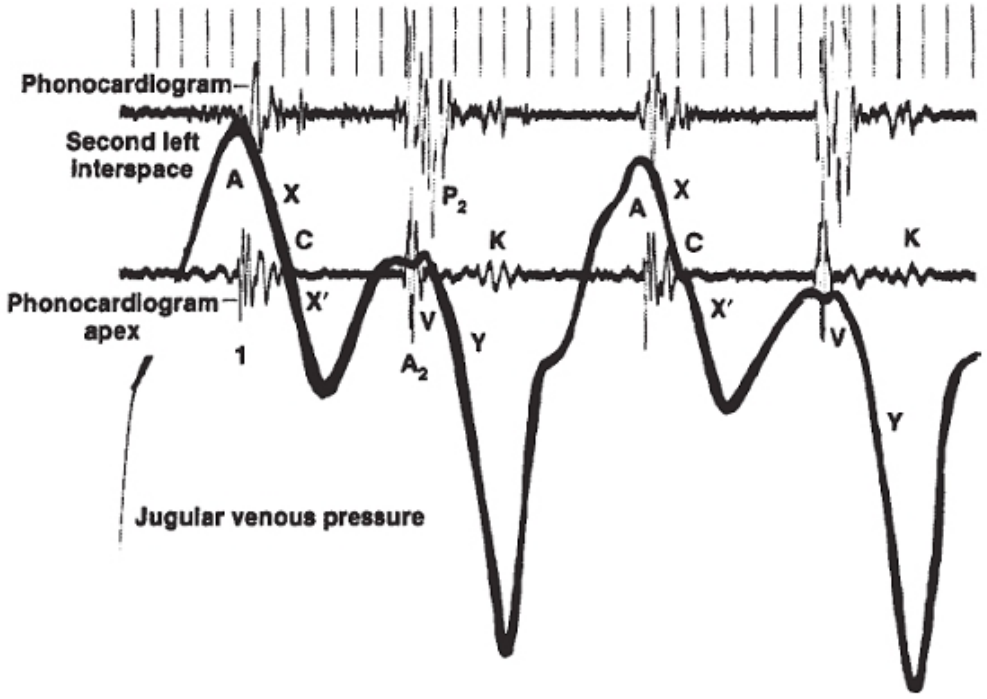
- Right atrial pulsus alternans:

  Prominent a and v waves may alternate with each other, seen in RVF with sinus rhythm.

Rapid 'y' - descent in JVP:

- Severe TR
- Constrictive pericarditis (friedreich's sign) (early rapid ventricular filling)
- PHT with RV dysfunction
- Severe RV failure
- ASD with mitral regurgitation
- Severe RV infarction

Figure 8 – Prominent y descent:



Slow y - descent in JVP:

- When RA emptying and RV filling are impaired, y descent is slow and gradual.
- Seen in Tricuspid stenosis or atresia
  RA myxoma
  Pericardial tamponade (y - descent may even be absent).

Double descents in JVP:

- Prominent x and y - descents in JVP
- Constrictive pericarditis.
- RV infarction with mild RV dysfunction
- PHT with early RV decompensation
- ASD
- Isolated prominent x descent in JVP :
  Cardiac tamponade
- Isolated prominent y descent in JVP :
  - Severe TR with RV dysfunction
  - Chronic AF with CCF
  - EMF with TR.

Estimation of venous pressure:

- Assessment of jugular venous pressure reflect mean right atrial pressure.
- Various methods are used to assess the jugular venous at the bed side.
  Measuring internal jugular venous pressure
  Hepato jugular reflux
  Examining the vein on the dorsum of the hand
  Examining the veins of the under surface of tongue (May sign).

Elevated JVP:

- Increased RV filling pressure and reduced ventricular compliance
  Severe pulmonary stenosis
  Severe pulmonary hypertension
  Right ventricular failure
  Severe RV infarction .
- Increased RV inflow impedance:
  Tricuspid stenosis / atresia
  RV myxoma
  Constrictive pericarditis
- Circulatory overlode
  Renal failure
  Cirrhosis of liver
  Excessive fluid administration
  SVC obstructions (non pulsatile JVP elevation)
  COPD

KUSSMAUL'S SIGN:

- Mean jugular venous pressure increases during inspiration as a result of impaired right heart filling due to reduced RV compliance, is called Kussmaul's sign:
- Constrictive pericaridtis
  Severe right heart failure
  RV infarction
  Restrictive cardiomyopathy

- Early RV decompensation
  JVP may be elevated
  a wave is prominent and larger than v wave.
  x and y - decent seen and equal.
- Decompensated RVF:

  JVP —> always elevated
  a and v wave prominent
  v wave larger than a wave
  x – descent is diminished or absent
  Rapid y descent due to TR.

JVP in EMF:

- JVP is usually elevated
- a wave prominent
- v wave is prominent due to TR
- x - descent is usually obliterated due to TR
- Rapid y - descent is due to TR
- Kussmauls sign is negative

JVP in ASD:

- JVP is normal and equal a and v waves seen
- x - decent is more prominent than y - descent
- Elevated JVP may seen in ASD with severe PAH and ASD with right ventricular failure.
- Prominent a wave seen in ASD if associated with severe PS or MS.
- Prominent v and rapid y - descent seen in ASD with TR due to right ventricular failure.

JVP in VSD:

- JVP is usually normal in isolated VSD
- Prominent a wave seen in associated pulmonary stenosis
- Prominent v wave seen in VSD of LV to RA shunt (Gerbodes shunt)
- Elevated JVP seen in non-restrictive VSD with congestive heart failure.
- VSD with Eisenmenger complex
  JVP is usually normal. CHF and TR is rare .
  Normal a and v waves

JVP in Ebstein anomaly:

- JVP is usually normal
- Unimpressive JVP is attributed to damping effect of large capacitance RA and thin, toneless atrialized RV (hypokinetic TR)
- Attenuated x - descent and systolic v wave are not reflected in jugular pulse despite appreciable TR.
- Prominent a and v wave with elevated JVP seen if right ventricular failure developed.

Cyanosis with prominent 'a' wave:

- It usually indicates intact IVS
- Severe PS with intact IVS and right to left shunt
- Tricuspid atresia
- TAPVC with restrictive ASD

- Eisenmenger syndrome with PDA
- PPH with right to left shunt via PFO

Cyanosis with elevated JVP:

- Elevated JVP in cyanotic CHD usually indicates intact IVS or increased PBF or both.
- Common in TGA or TAPVC with increased PBF
- Severe PS with RVF and right to left shunt
- Tricuspid atresia with restrictive ASD
- PPH with RVF and right to left shunt
- Eisenmenger syndrome with ASD and PDA.
- Adult TOF.

Cyanosis with normal JVP:

- It usually indicates cyanotic CHD with VSD and reduced PBF
- Tetralogy of fallot
- DORV with VSD + PS
- TGA with VSD +PS
- Single ventricular with PS
- VSD with Eisenmenger complex
- Pulmonary AV fistula.

JVP in Arrhythmias:

- In sinus bradycardia: slow normal regular sequence of a and v waves is maintained. 'h' wave can be recorded in slow rhythm.
- In Atrial fibrillation: JVP simulated TR as 'v' wave is prominent due to the absence of a wave and diminution of x - descent .
- Irregular heart rhythm:
  - Absence of a wave – Atrial fibrillation
  - Irregular cannon waves – Ventricular premature beat
  - In atrial premature beats, normal sequence of a wave, carotid pulse and v wave is maintained.
- In VT and Junctional rhythm: cannon waves are characteritic.
- VT with AV disassociation: irregular cannon wave occurs.

JVP in Conduction defects:

- PR interval can be estimated from interval between a wave and carotid pulse (C) so, increased a – C duration indicates prolonged PR interval.
- 1° AV block – prolonged a – C interval.
- Mohitz type I AV block: Gradual lengthening of sequential a-C intervals, ending with an a wave that are not followed by carotid pulse (non –conducted beat) can be made out.
- Mobitz type II AV block: a – C interval does not vary but is suddenly interrupted by isolated a waves that are not followed by a carotid pulse (non conducted beat).
- In 2:1 AV block, two 'a' waves for every one carotid pulse are present.
- Complete heart block: Intermittent cannon waves in a patient with bradycardia.

EXAMINATION OF THE PRECORDIUM

Dr S.Shanmugasundaram

Examination of the precordium is an important aspect of clinical evaluation of the cardiovascular system, but unfortunately often overlooked by the hasty physician. Low frequency vibrations of the heart like third and fourth heart sounds are better felt and seen than heard. Cardiomegaly can be quickly identified by simply palpating the apex. Specific chamber enlargements are easily made out even before radiological and ECG changes could manifest. Thus it is very important that one spends enough time in inspecting and palpating the chest before beginning the auscultation.

Methodology:

Each experienced clinican can devise his own method of examining the chest. I will be describing the methodology with which I am comfortable. This sequence has helped me in identifying most if not all of the abnormalities and in forming a mental picture of the hemodynamics. My preference is to combine inspection and palpation together to save time and achieve brevity of expression without repetition. One should remember that outward impulses are better felt and inward or retractile impulses are better seen than felt. While trying to palpate, it is a good practice to start placing both hands on either side of the chest, in order not to miss cardiac malpositions. Once the location of the heart in chest cavity is identified place the entire hand over the precordium to identify abnormal impulses and thrills. Finally selected areas of the hand might be used for bringing out a particular event better - heel of the palm to be used for assessing parasternal impulse ; finger tips for locating apex beat and assessing its quality and for localized impulses like pulmonary artery pulsation, RVOT or aortic pulsation and the region of heads of metacarpals for feeling the thrills. Practise with left hand as the tactile sensitivity of a nondominant infrequently used hand is better. Sometimes keeping the left hand over the chest and applying pressure over that with right hand may be useful in making out thrills. Timing of the events should be done by simultaneous palpation of carotid artery with the left thumb or by simultaneous auscultation. Palpation should be begun by firmly applying the hand over the area of interest and gradually reducing the applied pressure. Sharp high pitched sounds like opening snap and loud S1 are better felt with firm pressure. Low frequency vibrations like S3 or S4 are better felt by lighter pressure. Apical impulse may be better felt in end expiration and if behind the rib it may be better brought out by a slight inspiratory effort.

Sequence:

Precordial examination should begin with ***observing the entire chest*** for symmetry, deformity, dilated vessels and scars. ***Tracheal position*** is identified next to identify the presence of mediastinal shift. Locating the ***apex beat*** subsequently helps in assessing heart size. Nature of the apical impulse and ***parasternal impulse*** give information about the left and right ventricular hemodynamics. A dilated Pulmonary artery may be felt in the **left II interspace** and RVOT impulse in the ***left III interspace***. An abnormal aortic impulse may be seen / felt in ***right II interspace***. A pulsation over the ***right lower sternal border*** may imply a dilated overloaded right atrium. ***Supraclavicular, Suprasternal and Subxiphoid*** areas should also be examined. Inspecting and palpating the ***axillary and interscapular***

regions provide information in certain situations. While looking for the abnormal impulses in these areas, the presence of palpable sounds and thrills should be simultaneously ascertained.

Posture:

Ideally the patient should be examined in supine, semirecumbent (30' elevation with a backrest) and sitting postures. If the patient coughs or develops dyspnea on lying flat, examination in the supine posture may be avoided. Assuming left lateral posture may bring out the apex better but turning the patient to more than 45' left lateral decubitus shifts the apex down and out and makes it more heaving. However the area is not bigger than 3 cm^2 in normals even with extreme left lateral posture. If the apex beat is not felt in supine or sitting posture, turning the patient to <30' left lateral posture is allowed with a presumption that this degree of turning does not alter the position or the character. Anyhow extreme left lateral posture can be adopted to make out palpable S4 and thrills. Inspection is carried out from the right side and from the foot end of the bed tangentially with a suitably directed light source.

Place:

Examination is better carried out in a warm well lit and well ventilated room where privacy can be achieved and extraneous noise and distraction can be avoided. A very cold airconditioned room may make the patient shiver which hinders a proper examination. A cold palpating hand is also detrimental to a meticulous examination.

Steps:

The chest and abdomen should be bared. After inspecting the chest for ***symmetry***, one should look for ***deformities*** like pectus excavatum, straight back with narrow anteroposterior diameter and kyphoscoliosis. These deformities might result in shifting of apex beat resulting in apparent cardiomegaly. Pectus carinatum (pigeon chest) is anterior protrusion of sternum and ribs that may occur in Marfan's syndrome. Davies chest refers to anterior bowing of entire chest that occurs in children having large VSD with marked biventricular enlargement. Harrison's sulcus is the depression on the lower edge of the thorax at the level of insertion of diaphragm, giving the chest the shape of a pear. It occurs in rickets and in cardiopulmonary diseases that increase the work of breathing in early childhood – asthma or the noncompliant lung of heart failure being the commonest causes. **Precordial bulge** indicates dilated heart particularly the RV and it manifests in children and young adults because the ribs are pliable enough to be pushed out by the hyperactive heart. A localized **prominence of left II rib** is frequently noticed in ASD. ***Dilated veins*** over the chest wall may occur in SVC obstruction. A ***dilated*** tortuous lateral thoracic ***artery*** in the axillary region may suggest the diagnosis of coarctation. ***Surgical scars*** may give an insight into the nature of surgery done before – a midline sternotomy scar indicates open heart surgery – ASD, VSD closure, TOF repair, CABG, Valve Replacements; left anterolateral thoracotomy scar indicates previous closed mitral valvotomy or subtotal pericardiectomy (nowadays pericardiectomy is mostly done through midline sternotomy); Posterolateral thoracotomy scar indicates previous BT shunt, PDA ligation or coarctation repair.

Trachea should be palpated first for assessing mediastinal position. Trachea is in the centre with slight deviation to right. The direction of the anterior ridge of trachea or the space between the trachea and sternomastoid on either side should suggest the upper mediastenal position . This will help in ascertaining the cause of apical impulse shift – intrinsic or secondary to lung disease - collapse

or fibrosis (shifted to ipsilateral side) or pneumothorax or pleural effusion (shifted to contralateral side)

Apical impulse – the outermost and the lowermost point on the precordium – is formed by the anterior wall of left ventricle close to apical inter ventricular septum. The true anatomic LV apex is slightly lower and lateral to the clinically palpable apical impulse. Counterclockwise rotation of the apex with relatively fixed basal part gives rise to anterior movement of LV apex imparting the palpating finger an outward thrust. The maximum anterior movement occurs during the isovolumic phase of contraction and early ejection. Continued ejection and emptying of LV makes the apical impulse to recede from the palpating finger in mid late systole. In diastole clockwise rotation of apex allows it move posteriorly and in normal individuals no impulse is felt over the apex in diastole. The term Point of Maximal Impulse should be avoided as an alternative term for apex beat because a maximal impulse may be elsewhere in precordium eg: an ectopic dyskinetic impulse in anterior wall infarction may be more prominent than the true apical impulse.

While trying to palpate the apex, the following features should be elicited - **L**ocation, **A**mplitude, **S**ize, **D**uration & **C**haracter or **C**ontour (L A S Du C). A normal apical impulse is located in the 5^{th} or 4^{th} left interspace, 1-2 cm medial to midclavicular line (10 cm from midsternal line or 7- 8 cm from left sternal edge). It occupies not more than one interspace, the area of the impulse being 2 to 2.5 cm^2 . It is a brief gentle impulse lasting for < 1/3 of systole. There is a zone of retraction medial to apex, as the right ventricle recedes in systole. In a tall asthenic individual, apical impulse may be felt in 6^{th} interspace and > 2 cm internal to midclavicular line. On the contrary, in individuals with short stocky build the apex may be in 4^{th} left interspace at or lateral to midclavicular line.

A ***tapping apical impulse*** is a hypokinetic (underfilled LV) apex beat with a palpable S1, the latter giving rise to the tapping quality (tap = to strike gently with a light blow). It typically occurs in mitral stenosis. A ***Hyperdynamic apical impulse*** occurs in volume overload states of LV like mitral regurgitation, aortic regurgitation, VSD, PDA, RSOV and Hyperkinetic circulatory states like anemia, hyperthyroidism etc . It occupies two or three interspaces exceeding 3 cm in diameter. The apex is usually displaced down and to the left. It is increased in amplitude with a brisker upstroke when compared to a normal apical impulse but ill sustained – the duration not exceeding initial half of systole. The medial retraction is exaggerated giving rise to a rocking nature of precordial pulsation. Milder degrees of volume overload may slightly increase the amplitude and rapidity of upstroke giving a hyperdynamic feeling to the palpating finger but may be confined to one interspace without getting significantly displaced. An apical impulse of this nature occurs in moderate MR / AR or moderate left to right shunts, febrile states, after exertion and during excitement. Hyperdynamic apical impulse denotes increase in LV end diastolic volume with preserved contractile function; in other words the end systolic volume is normal. Chest deformities like thin flat chest, pectus excavatum or rib resection (eg: closed mitral valvotomy done in earlier days) may give rise to a hyperdynamic quality though LV volume is not increased. A ***heaving apical impulse*** indicates pressure overload states like Aortic Stenosis, HOCM, Systemic Hypertension. It is increased in amplitude and duration (sustained) generally lasting for > 2/3 of systole. It is usually not displaced, occupying not more than one interspace. Apical heave indicates LVH accompanied by increased force of contraction but without increase in LV volumes. Prolonged contraction and ejection allows the apex to be in contact with the chest wall for longer time, the amplitude being more because of increased force of contraction or increased LV systolic

pressure. Hypertrophic cardiomyopathy may give rise to sustained impulse even in the absence of obstruction. A long standing severe volume overload state particularly chronic severe AR may give rise to a heaving apical impulse particularly with the onset of LV systolic dysfunction. A heaving apical impulse may also occur in LV dysfunction (ischemic or idiopathic dilated cardiomyopathy) or Apical LV aneurysm. In spite of poor systolic performance the apical impulse is heaving in LV dysfunction because of increased end systolic volume and prolonged systolic contraction / ejection, allowing the apex to remain in contact with the palpating finger for longer time. In conditions that produce marked RVH and dilatation, RV may form the apex and it may be hyperdynamic or sustained with a lateral retraction caused by the posteriorly rotated LV. In this situation the parasternal impulse is continuous with the apex. In Biventricular overload, there may be an area of quiescence or retraction in the mid precordium corresponding to the region of anterior septum. A ***retractile apical impulse*** occurs in constrictive pericarditis – in systole it is negative or inward and in diastole it is outward. Apical impulse may not be palpable in obesity, muscular chest wall, barrel chest (elderly), emphysema, large pericardial or left pleural effusion.

Parasternal Impulse:

Parasternal impulse is generated by the right ventricle. In an anatomical sense, the RV is a crescent attached to LV as an appendage and it contracts with a bellow like action. Thus when LV undergoes counterclockwise rotation in long axis the RV recedes from the chest wall. Moreover RV is thin walled and generates smaller systolic pressure < 25 mm Hg. That is how the normal RV systole does not produce any palpable activity. In thin chest wall individuals and in children, one may be able to feel a very brief outward impulse followed by mostly a retractile pulsation – better seen than felt. When there is RV volume overload state (ASD, Non pulmonary hypertensive TR) the retractile impulse is exaggerated to give rise to a wavy parasternal impulse – seen better than felt. This is equivalent to Grade I Parasternal impulse where the RV impulse is obliterated by a finger. When there is RV pressure overload without RV dilatation, as in TOF, severe pulmonary stenosis and DCRV, one may get a stronger or forcible outward impulse but not a sustained one. This jerky impulse is called as parasternal or RV systolic shock. This impulse is better felt in sitting posture in end expiratory apnea and is accompanied by a much more forcible subxiphoid RV thrust. This is equivalent to Grade II parasternal impulse, defined as an outward impulse in the parasternal region, which could be obliterated by firm pressure with the heel of the palm. In Valvular Pulmonary Stenosis, the forcible RV impulse is felt over left III, IV & V interspaces. In TOF, where the obstruction is subvalvular, the RV impulse is confined to left IV & V interspaces. In DCRV, a parasternal impulse may not be felt but the subxiphoid impulse may be quite forcible. In RV systolic overload states with RV dilatation, as in Pulmonary Hypertension with tricuspid regurgitation, Pulmonary stenosis with RV failure, the parasternal impulse is a sustained forcible outward impulse. This is equivalent to Grade III parasternal impulse, a sustained RV impulse which cannot be obliterated by the heel of the hand, whatever force you apply.

While trying to feel the RV impulse from subxiphoid region, it is preferable to examine the patient in 30' semirecumbent posture during inspiratory apnea with the examiner's palpating middle three fingers insinuated in the subxiphoid area. RV impulse may impart the feeling to the tips of the insinuated fingers in contrast to abdominal aortic pulsation, which is felt over the pads of fingers not the tips. In COPD and barrel chest RV impulse is palpable only in the subxiphoid region.

If a systolic impulse is felt over left III interspace, a dilated RVOT may be responsible as it does occur in Ebstein's anomaly and RV EMF. In both the conditions a quiet lower left parasternal area (a dilated noncontractile atrialized RV in Ebstein's anomaly and obliterated RV apex in EMF are responsible for quiet lower parasternal region) is in sharp contrast to vigorous pulsation in left III space. Ascending aorta in CTGA may also produce a localized impulse in left III interspace.

In severe mitral regurgitation, a dilated left atrium receiving a large influx of regurgitant blood may produce an impulse in the left parasternal region. The systolic expansion of LA displaces RV anteriorly and produces a palpable event. This differs from the parasternal impulse of RV systolic overload in the following respects – LA pulsation is late systolic with a slow rise, late systolic poorly sustained peak and rapid fall and is felt little away from the left sternal edge in a localized area. RV impulse of systolic overload is felt close to the sternum, more widespread with a slow rise, slow fall and a sustained peak. Rarely a dyskinetic septum of anteroseptal MI may produce an impulse in the parasternal region – a bulging anterior septum may push the RV anterior wall and impart a palpable impulse. Severe TR may produce pulsation along right lower sternal edge.

Systolic Impulse In Left II Interspace is due to dilated pulmonary artery which occurs in **I**ncreased Flow (left to right shunts) **I**ncreased Pressure (Pulmonary Hypertension) and **I**diopathic Dilatation. The impulse is hyperdynamic and ill sustained in situations producing increased flow. It is sustained in PAH. Dilated ascending aorta as in aortic regurgitation (mostly in conditions due to dilated aorta like annulo aortic ectasia), Aortic Dissection and aortic aneurysm may produce visible and palpable **impulse in right second interspace** and at times lower down. Post stenotic dilatation of ascending aorta in Valvular Aortic Stenosis may produce a visible but nonpalpable impulse localized to right second interspace. An **ectopic impulse** (usually above and medial to apex) may occur in post infarction dyskinesia and LV aneurysm. **Suprasternal** pulsation may be due to hyperkinetic circulatory states, Aortic Run Off situations like AR, PDA and arch aneurysms. Dissection of aorta and right aortic arch may produce pulsation of right sternoclavicular joint. A giant LA may produce a systolic impulse in ectopic sites like right anterior or lateral chest or left axillary region. A **supraclavicular** pulsation commonly occurs on the right side due to kinked tortuous carotid artery. Collaterals of Coarctation of Aorta may at times be felt over supraclavicular area. When the patient bends forward in standing posture with arms dangling down, collateral arteries of coarctation of aorta may be seen and felt over the back (inter and infrascapular regions) – Suzman's sign. Broadbent's sign is **retraction of intercostal space** coinciding with the systolic phase of cardiac cycle – a sign of adherent (constrictive) pericarditis – detected near the posterior axillary line.

Diastolic Impulses are usually the palpable low frequency events like presystolic LV distension (palpatory counterpart of S4) and early diastolic impulse due to exaggerated rapid filling of LV (palpatory counterpart of S3). S3 is better felt with gentle palpation with the entire hand while S4 is better felt by firm pressure with one or two fingers (tip). Early diastolic impulse is felt in volume overload states like MR, VSD PDA etc and in LV dysfunction. Pericardial knock is at times palpable along the sternal edge. Palpable presystolic impulse over the apex indicates augmented atrial contraction in the presence of decreased LV compliance as in AS, HCM, CAD and SHT. A bifid apical impulse of HOCM is usually due to palpable S4 and heaving systolic LV impulse. Rarely it may be trifid – S4, Early systolic and late systolic LV impulse with midsystolic retraction. In MVP also there may be midsystolic retraction but it is usually a recordable than palpable event. Palpable S4 is always pathological whatever be the

age of the patient. Remember a fourth heart sound may be normally heard in elderly. Palpable S4 in Aortic Stenosis indicates severe obstruction – the gradient being more than 70 mm Hg. Dilated cardiomyopathy may produce palpable S3 and / or S4. It is better to form a mental picture of the palpatory diastolic events along with trans mitral Doppler echo findings. Palpable S3 occurs in conditions with Doppler echo evidence of grade III diastolic dysfunction (restrictive physiology) and palpable S4 in conditions with grade I diastolic dysfunction (impaired relaxation). S4 and S3 of RV origin may be felt along the lower sternal edge or in subxiphoid region - better felt in inspiration.

Sounds and Thrills:

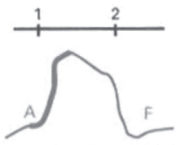
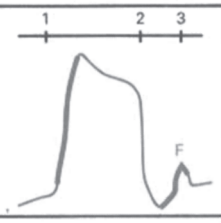
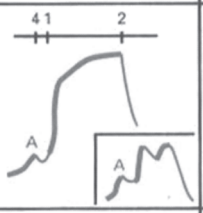
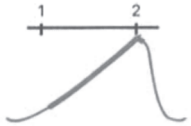
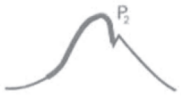
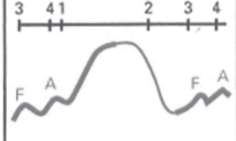
A loud S1 may be felt over apex in Mitral stenosis; Opening snap of MS in midprecordium ; Sail sound of Ebstein's along lower left sternal edge; a loud P2 of PAH in left II interspace; Aortic ejection sound over the apex, Pulmonary ejection sound at the upper left sternal edge and loud A2 along left sternal edge in SHT, aortic aneurysms, TGA and CTGA. Systolic thrill occurs over the apex in MR; along lower left sternal edge in VSD, DCRV, HOCM, Discrete membranous subaortic stenosis or IPS; in left second interpace in supracristal VSD and valvular PS and in right second interspace with radiation to right or both sides of neck and suprasternal notch in Aortic Stenosis. Diastolic thrill at apex usually indicates MS. Diastolic thrill along left III and IV interspaces may occur in certain conditions producing acute severe AR. But whenever an early diastolic murmur is accompanied by thrill it usually denotes PR rather than AR. The thrill of PR is better felt over left II interspace along the left sternal edge and little away from it also. Continuous thrill over left II space occurs in PDA and along lower left sternal or right sternal border in RSOV (a peculiar superficial purring sensation in this condition is virtually diagnostic).

Palpation should be completed by looking for hepatic pulsations – systolic in TR and presystolic in Tricuspid stenosis.

Percussion:

The diagnostic yield by percussion is little. It may be more useful to percuss the chest for identifying the visceral situs – liver dullness on the right side and gastric tympany on the left side. An astute physician may employ percussion to diagnose cardiomegaly when apex is poorly felt, RA enlargement, PA dilatation and pericardial effusion (dullness at apex extends beyond the palpable apex beat).

Diagnostic yield by careful inspection and palpation is so much that nearly complete diagnosis is achieved before auscultation. For example if there is wavy pulsation along the left sternal edge, RV volume overload is present- it may be caused by ASD or TR. If there is no prominent v wave in neck, ASD is diagnosed. A quiescent RV inflow with tumultuous RVOT indicates either Ebstein's anomaly or RV EMF- neck veins might clinch the diagnosis – steep x and y (w pattern) suggest RV EMF and elevated JVP without a prominent v wave in neck is diagnostic of Ebstein's. There are innumerable such examples and for a good clinician precordial examination is challenging, interesting and informative.

Type of movement and associated clinical condition		Location and accompanying features
NORMAL ADULT APEX IMPULSE		Cardiac apex; moderate systolic thrust; A and F waves usually imperceptible
HYPERKINETIC APEX IMPULSE "Normal Child "Hyperdynamic states "Ventricular septal defect "Patent ductus arteriosus "Mitral regurgitation "Aortic regurgitation		Exaggerated thrust at cardiac apex; F wave may be palpable, coincident with third heart sound
HYPERKINETIC RIGHT VENTRICULAR IMPULSE "Atrial septal defect "Pulmonary regurgitation	Same as above	Maximal at left sternal edge in third and fourth intercostal spaces
SUSTAINED APEX IMPULSE "Left ventricular hypertrophy, ""as in: "Aortic stenosis "Hypertension "Insert: a variation that ""may occur in hypertrophic ""cardiomyopathy		Maximal at cardiac apex; A wave may be visible and palpable coincident with fourth heart sound
SUSTAINED RIGHT VENTRICULAR IMPULSE "Right ventricular ""hypertrophy, as in: "Pulmonary hypertension "Pulmonary stenosis	Same impulse as in Sustained above	Maximal at left sternal edge in third and fourth intercostal spaces
ECTOPIC LEFT VENTRICULAR IMPULSE "Ventricular aneurysm	Same impulse as in Sustained above	Maximal over mid-precordium rather than at apex
LEFT ATRIAL EXPANSION "Severe mitral regurgitation		Left sternal edge or entire precordium; hyperkinetic apex impulse due to left ventricular volume overload
PULMONARY ARTERY PULSATION "Pulmonary hypertension		Second left intercostal space; palpable P_2
INWARD MOVEMENT DURING SYSTOLE "Constrictive pericarditis "Tricuspid regurgitation; ""primary		Cardiac apex or entire precordium; reversal of direction during systole as compared with preceding examples
DIASTOLIC MOVEMENTS "Cardiomyopathy		Cardiac apex; systolic movement may be inconspicuous; diastolic movements F and A correspond to 3rd and 4th heart sounds which may merge in tachycardia to form a summation gallop

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AUSCULTATION OF THE HEART

Dr. J. Balachandar

Mitral stenosis

The First Heart Sound (S1)

In Mitral Stenosis (MS) a tricuspid component may precede the delayed M1. Intra-cardiac phonocardiograph has shown that a right sided S1 component precedes a left-sided S1 component in about a fourth of patients with MS. In MS, S1 is loud because the high pressure in the left atrium that makes the gradient at the end of diastole requires the ventricle to reach a higher pressure before it can close the valve. The LV, therefore, has more time to accelerate before it closes the valve.

The relatively stiff mitral valve, as in MS, makes the M1 short and snapping (i.e. it causes a "closing snap")[1]. The resistance to movement of the fibrotic and tethered edges causes the still flexible bodies or bellies of the anterior leaflet to billow upward like a snapping sail. The posterior leaflet usually has relatively little "belly" to billow unless it has redundant tissue, as in mitral valve prolapse syndrome. Therefore, the anterior leaflet is mainly responsible for the M1 snapping effect. When porcine mitral valves become fibrosed, the high frequencies become dominant. [2]

When the belly of the anterior leaflet is very stiff and immobile due to either calcium or severe fibrosis, there is no snapping M1.

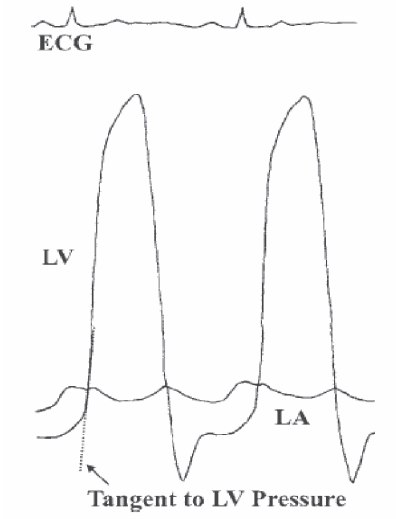


Fig. 1. Diagrammatic illustration of the left ventricular (LV) and left atrial (LA) pressure curves in a patient with mitral stenosis showing the diastolic pressure gradient between LA and the LV reflecting the mitral stenosis. When the rising LV pressure with onset of systole exceeds that of the LA, the mitral valve will close. Note that the tangent to LV pressure drawn at the point of the crossover of the two pressure curves during this phase of LV systolic pressure rise is steep, showing that the ventricle has achieved a faster rate of contraction and higher dP/dt.

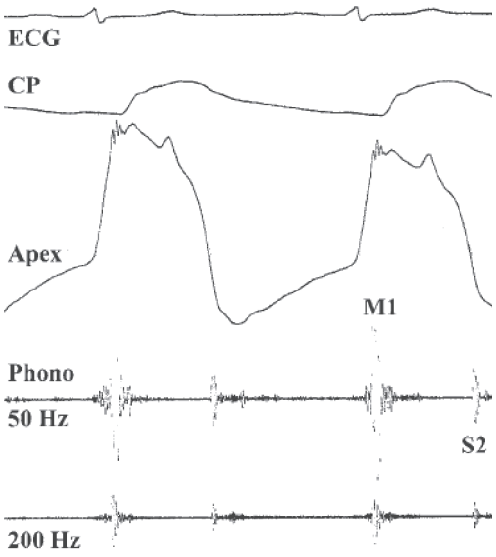


Fig. 2. Phonocardiographic (Phono) recordingfrom a patient with mitral stenosis showing theloud intensity first heart sound caused by the loud mitral component M1.

Mitral stenosis – M1 Loudness and AF :

Three types of M1 Loudness variations.

Type 1 : If the MS is mild and although there is softening with short diastoles as with a normal valve, there is less tendency for the M1 to become louder after long diastoles. [3]

Type 2 : If the valves are severely stenosed and calcified (i.e. no opening snap is present), the M1 depends entirely on end-diastolic volume and on the preceding and pre-preceding RR intervals (starling and post extrasystolic potentiation

effect). Thus, the S1 becomes louder in proportion to the length of the previous diastole.[3]

Type 3 : If the valves are moderately stenosed, the S1 loudness varies inversely with the duration of the previous diastole (i.e., the shorter the previous RR interval, the louder the M1 because of dependence on the end-diastolic left atrial-to-LV gradient). [3]

It used to be thought that the loud S1 (M1) characteristic of severe MS was caused by closure of the valve that was kept wide open by high left atrial pressure.

In MS, S1 (M1) is loud because the valve closure occurs at a time when the dp/dt in the ventricle is high as a result of a higher pressure cross over point. In some patients with very severe MS, usually associated with heavily calcified valves, the M1 may not be loud. This probably stems from the fact that the LV in such patients are grossly underfilled from the mitral obstruction and therefore are unable to achieve good contractility and dp/dt.

The second heart sound (S2) :

In MS the absolute loudness of the P2 is a good sign of pulmonary hypertension only if it is loud.

If the S1 is louder than the S2 at the base it suggests that an extra-loud S1 is present, as in MS.

Normal or narrow splitting is expected in most patients with MS and pulmonary hypertension. The level of pulmonary hypertension does not correlate with the width of splitting in MS.[4]

The Opening Snap (OS) :

For opening snap (OS) to occur, there is usually some MS due to fibrous thickening and often calcification of the margins of the mitral leaflets, especially the large anterior leaflet. The commissures are also fused by fibrosis or calcium. The mitral valve belly may act like a sail that billows downward into a dome in diastole as the left ventricle (LV) attempts to "suck" left atrial blood into the LV cavity. This sudden diastolic doming causes the anterior leaflet to bulge downward with a snap. It is the anterior leaflet that is responsible for the OS of MS. Not only is the anterior leaflet three times broader than the posterior leaflet, but the surface of the chordae tendinae of the anterior leaflet are also attached to the peripheral zone of the valve, leaving the bellies intact, whereas the surface chordae of the posterior leaflet are attached to its entire ventricular surface.[5] Therefore, even though MS tends to convert the two semi-independent leaflets into a continuous funnel-like sleeve with a fish-mouth opening. The belly of the anterior leaflets contributes most to the loud S1 and OS.

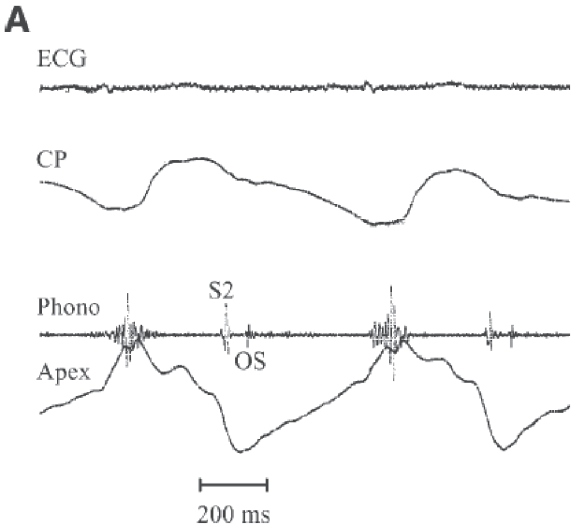


Fig.3. (A) Phonocardiogram (Phono) recording taken at the apex area from a patient withrheumatic mitral stenosis who had a previous mitral valve commissurotomy for relief of the obstruction. The S1 is relatively loud. Note a sharp sound following the S2, which is the opening snap (OS). The OS occurs almost simultaneously with the most nadir point of the apex tracing, which is termed the O point.

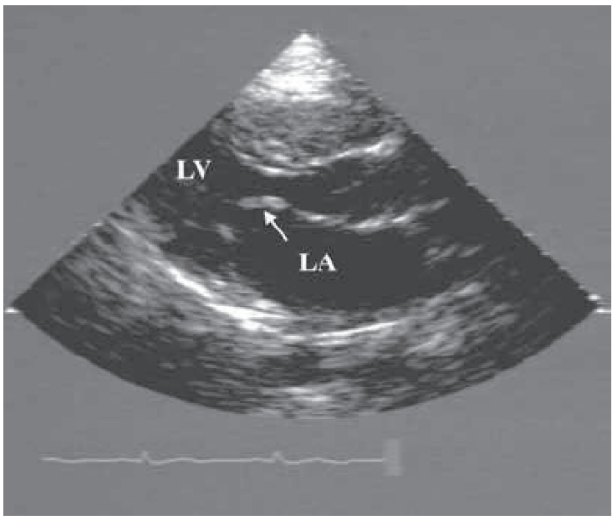


Fig.4.Stop frame of a two-dimensional echocardiogram from a patient with mitral stenosis inthe parasternal long axis at onset of diastole showing the typical bowing of the anterior mitral leaflet (arrow).

Note that the leaflet tip is pointing posteriorly because of tethering caused by thestenosis making a funnel-like opening. Part of the column of blood trying to enter the left ventricle (LV) from the left atrium (LA) during diastole is oriented toward the belly of the leaflet. When the leaflet excursion reaches its anatomical limits caused by the tethering, this column of blood is suddenly decelerated. This leads to the production of the opening snap (OS).

Other causes of OS are :

1. Dominant mitral regurgitation MR : if the regurgitation is due to a thickened, rolled, immobile posterior (mural) leaflet and if the anterior (septal) leaflet still has a mobile belly.[6,7]
2. Ventricular Septal Defect (VSD)
3. Persistent Ductus Arteriosus (PDA)
4. Tricuspid Atresia
5. Thyrotoxicosis
6. After a BT shunt for TOF
7. Congenital MS
8. Mitral valve prolapse[8,9]
9. Massive ascites[10] (Pseudo-Knock sound)
10. Tumor plop of LA Myxoma (same timing as O.S?)

Relation between the S2-OS and the severity of Mitral stenosis :

The factors that control the duration of isovolumic relaxation or the S2-OS interval are

1. The pressure in the left atrium at the time the mitral valve opens
2. The heart rate. The more rapid the heart rate, the shorter the S2-OS interval because isovolumic relaxation is faster under the influence of increased sympathetic tone or catecholamines that cause the faster rate, and left atrial pressure is higher due to the reduced atrial emptying time with short diastoles.
3. The stiffness of the mitral valve (due to mostly calcium)[6]
4. The rate and strength of relaxation of the myocardium (i.e., the state of myocardial function or inotropic state [contractility] of the myocardium.
5. The pressure at which the aortic valve closes (near systolic pressure).

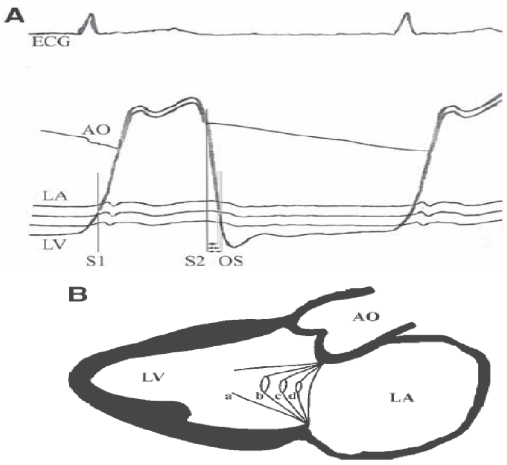


*Fig. 5.**(A)** Diagram showing simultaneous left ventricular (LV) and left atrial (LA) pressuresin mild, moderate, and severe degrees of mitral stenosis. The more severe the stenosis, the higher will be the left atrial pressure. The opening snap (OS) occurs at the end of the isovolumic relaxation phase of the left ventricle when the left ventricular pressure falls just below the left atrial pressure. The OS will therefore tend to occur earlier with higher LA pressure and later with lower LA pressure. Thus, the S2-OS interval is short with severe mitral stenosis and long with mild mitral stenosis. **(B)** Visual representation of the excursion of the mitral leaflets in mitral stenosis of different degrees of severity: a, normal; b, mild; c, moderate; d, severe. With milder stenosis the column of blood has to travel further before decelerationagainst the valve, thereby making a late OS.*

The greater the stenosis, the greater the obstruction to flow, and thus the slower and more incomplete empyting of the left atrium. Therefore, the greater the MS, the higher the V wave.

The V wave is built up during ventricular systole when the mitral valve is closed. If the V wave begins to build up from a high pressure, the wave rises even higher. If the left atrium did not empty well in diastole due to MS, the V wave starts to build up from an already high pressure.

A high left atrial pressure makes the S2-OS interval shorter because the LV pressure does not have to fall as far to open the mitral valve after closure of the aortic valve or A2.

The high V wave caused by concomitant MR does not necessarily cause an early OS, probably because the OS in MS occurs not at the peak but early on the J descent.[11]

Even though the descent of the mitral leaflets begins at the cross over of left ventricular and left atrial pressure, maximum excursion of the leaflets may take another 25-70 msec, and the OS occurs at the moment of maximum excursion.

The beat that follows a long diastole in AF produces a long S2-OS because a long diastole gives more time to empty the left atrium, allowing its pressure to drop. The next systole begins and ends with a lower left atrial pressure, thus making a long S2-OS.

If the mitral valve is very stiff, the S2-OS interval will be lengthened, because the stiffer (less mobile) the valve, the lower the LV pressure has to fall below atrial pressure before it can "suck" the valve open.[6]

Calcification has been shown to slow the velocity of mitral valve opening. The S2-OS interval is the sum of the true isovolumic relaxation period (A2 to the beginning of the opening of the mitral valve) plus mitral valve excursion. Therefore, even though a high left atrial pressure shortens the true isovolumic relaxation time, valve calcification and stiffness lengthen the mitral valve excursion period. The result is a longer than expected S2-OS for the degree of MS.[12] Echophonocar diogrpahic correlations have shown that the true isovolumic relaxation period (A2 to the onset of mitral valve opening) correlates better with MS severity than does the S2-OS interval.

Causes of a late OS

1. Mild degree of MS
2. Heavily calcified mitral valve
3. Bradycardia
4. Poor myocardial function
5. Aortic regurgitation (AR)
6. A low left atrial pressure due to a large left atrium and severe failure with low flow.
7. High aortic pressure : At an aortic systolic pressure of more than 130mm.Hg, the S2-OS interval is unreliable in indicating the degree of MS.[13]

A narrow S2-OS is more reliable in predicting the degree of MS. Unequivocally tight MS is suggested by a resting S2-OS of 59ms or less, even if only after exercise.[14,15]

Soft or absent OS in MS

1. Obese chest
2. Emphysematous chest
3. Calcification of the mitral valve :An OS can be heard even with heavy calcification but it will be soft, late, and low pitched (i.e. it loses high frequencies).[16] This is opposed to the degeneration of a bioprosthetic valve, which, loses low frequencies when it becomes fibrotic.
4. Fibrosis alone is rarely responsible for a soft OS.
5. Congenital MS has no OS due mainly to the rubbery consistency of the valve.
6. Extremely low flow due to
    a. Exceptional severity of the stenosis.
    b. Severe pulmonary hypertension
    c. Concomitant aortic or tricuspid valve disease
    d. Myocardial dysfunction.
7. A large right ventricle (RV) (usually due to pulmonary hypertension or tricuspid regurgitation) that pushes the LV away form the chest wall.
8. Moderate to severe AR that can "cushion" the mitral valve anterior leaflet as it comes downward.

When a patient with both AR and MS sits up, you may hear a soft OS for the first time, probably because the decreased venous return of the sitting position reduces the amount of AR.

Standing can cause a soft OS to disappear, and raising the legs can make it louder.

A mitral commissurotomy will eliminate the OS in about half the cases.

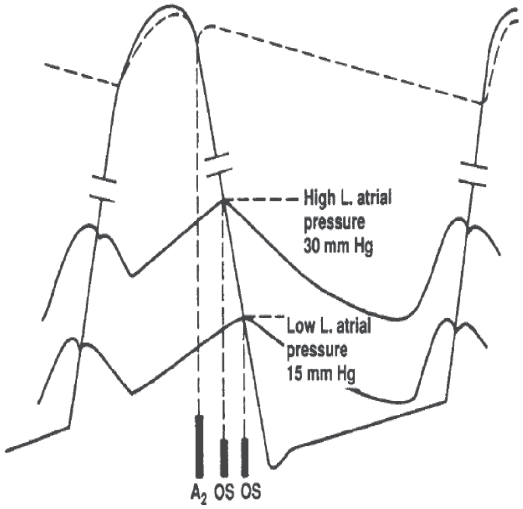


Fig. 6. Note that the distance between the A2 and the OS is shorter with the higher left atrial pressure.

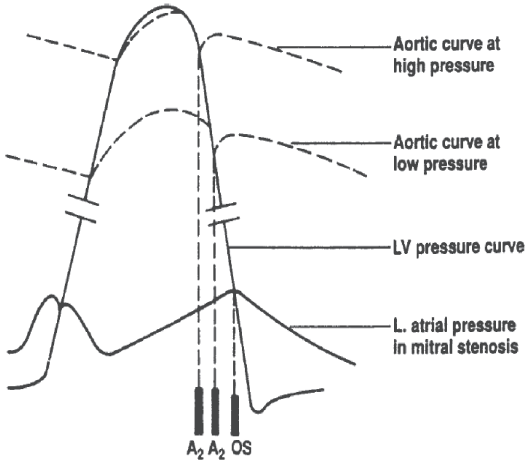


Fig. 7. Note that the higher the aortic pressure the longer the A2-OS interval.

Differentiating A2 P2 from A2 OS :

1. If the second component of a split S2 is louder at the apex as elsewhere, it is probably an OS.
2. If the second component of an S2 split becomes softer on inspiration at the lower left sternal border (in the absence of LBBB), it is probably a mitral OS.
3. A widely split S2 on inspiration, that appears to become, wider on expiration is an OS, in the absence of LBBB.
4. A triple second sound, in which the three sounds are close enough together to sound like a snare-drum, implies than an OS is present as the final component.
5. If a split second sound becomes wider on standing, second component is an OS.
6. If the S1 is soft the second component of the S2 is not likely to be an OS.

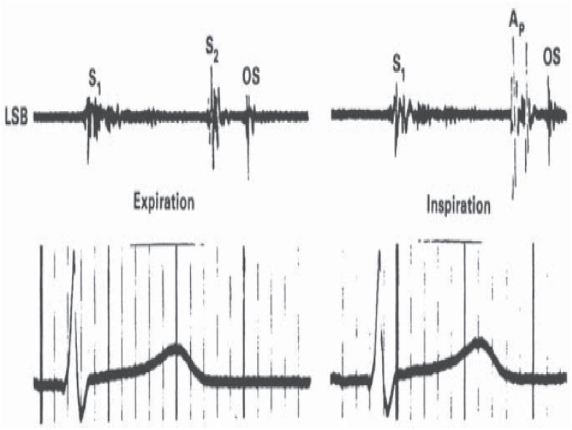


Fig. 8. On inspiration, the S2 split opened up into its A2 and P2 components. Together with the OS, a triple second sound is heard that produces a snare-drum effect.

Differential Diagnosis of an Opening Snap :

1. The early S3 of constrictive pericarditis – "Pericardial Knock"
2. The Tumor-plop of a left atrial myxoma.
3. A vegetation on the mitral valve that moves rapidly form the left atrium into the LV and strikes the base of the ventricular septum.[17]

The S3 and Mitral Stenosis :

If the Diastolic murmur of MS begins with a loud sound, that sound is probably on S3. One study showed that when the S3 was recorded in MS, its presence and intensity were independent of the severity of MS. The S3 varied only with the intensity of LV expansion and recoil in early diastole and could be present even in severe MS with reduced flow. The S3 probably reflects good LV function. In the French literature the loud S3-like beginning of the diastolic murmur has been called the "initial jerk" of the MS murmur.

A right ventricular S3 may be heard in MS if the right atrial pressure is high and the RV is dilated due to pulmonary hypertension and congestive

failure. If the enlarged RV usurps the apex area, the S3 may be heard well into the middle of the left thorax and may be mistaken for an LVS3.

The diastolic murmur of mitral stenosis :

The diastolic murmur of MS begins just after the opening snap (OS). This means that there must be a pause due to isovolumic relaxation between the A2 and the diastolic murmur. Because of the pause that usually occurs after the S2, the MS murmur is called an early delayed diastolic murmur. There often seems to be an slight additional pause between the OS and the diastolic murmur of MS when listening with the sthethoscope. Although inflow into the left ventricle (LV) begins as soon as the mitral valve opens, the gradient and flow increase for a short period because the LV is still rapidly expanding. This increase in gradient often shows on a phonocardiogram as a short, early cresundo-decrescendo. We here the OS and then the peak of the crescendo as if there were a pause between the OS and the beginning of the murmur. The typical shape of the diastolic murmur of MS on auscultation is initial crescendo, decrescendo rumble and late crescendo upto to the M1.

The crescendo murmur to the M1 in MS ("Presystolic murmur") :

The time between the onset of ventricular contraction and closure of the mitral valve or M1 is called the Pre-isovolumic contraction period.

This period is prolonged in MS because both the high left atrial pressure and the stiffness of the mitral valve have to be overcome before the mitral valve can be closed. Most of the murmur, however, is actually an pre-systolic murmur, because it occurs during the pre-isovolumic contraction period of LV contraction.

As the mitral orifice is reduced by LV contraction, the velocity of flow increases as long as the pressure is higher in the left atrium than in the LV.[18] Several studies have shown that even the slightest closing motion of a stenotic mitral valve during diastole can produce a diastolic murmur.[19,20]

In AF the late crescendo occurs at the end of short diastoles because only during short diastoles is the left atrial pressure high enough to maintain high-velocity flow during preisovolumic contraction. It requires a gradient of more than 10mmHg at the onset of LV contraction to create a crescendo murmur to the M1.

The presence of a crescendo murmur to the M1 indicates that the valve must be sufficiently flexible to change the size of the orifice; that is, it must not be rigidly calcified (although it may be too fibrosed or calcified for a valvotomy).

Important MR complicating MS can eliminate this pre-M1 accentuation even in sinus rhythm.[21] The loss of presystolic accentuation here may be due to a poorly contracting left atrium secondary to both the dilatation and the greater rheumatic damage of the atrium associated with the combined lesion.[22]

Pitch and Quality :

The diastolic murmur of MS is low in pitch because a murmur that is produced by flow than by gradient produces low frequencies. High frequencies are produced if the velocity of flow across the orifice is increased due to a good circulation time, a strong LV expansion, a strong atrial contraction, MR or a tadpole-shaped orifice in which. Commissural fusion and volvular thickening are more marked anterolaterally than posteromedially.[23] The crescendo to the M1 is sually rich in frequency. If a wide, rough S1 is confused with a presystolic crescendo, form pressure with the diaphragm brings out the high-pitched Crescendo components leading to the M1. A rough S1, on the other hand, separates into split sound components.

Factors increasing MS murmur loudness :

1. Expiration
2. Healthy LV
3. Concomitant MR

A Grade 4/6 mitral diastolic murmur means that there is atleast moderate stenosis in the absence of MR. If, however, it radiates to the base,

it almost always signites severe MS. (It also denies systemic levels of pulmonary hypertension).

4. When the patient is turned to left lateral position and auscultation done during end-expiration with very light pressure exerted by the bell of the stethoscope.
5. The flow across the mitral valve can be increased by
    a. Coughing a few times
    b. Post-valsalva release phase.
    c. Listening after heart rate has slowed down or after digitalis
    d. Squatting or during a handgrip maneuver. Cardiac output is increased for a few beats after squatting. During handgrip the mitral diastolic gradient has been shown to increase as a result of both the increase in cardiac output and the increase in heart rate.
    e. Exercise : 3 minutes of moderate exercise
    f. Amylnitrite : Increases venous return and cardiac output.

Factors softening the MS murmur :

1. Mild Ms
2. Obesity
3. Emphysema
4. Low flow
5. A large RV pushing the LV posteriorly
6. Coincidental ASD
7. Severe pulmonary hypertension
8. Tricuspid or aortic stenosis
9. Very much dilated left atrium : due to severe rheumatic damage can lower the left atrial pressure even with severe MS and decrease the flow.
10. Cardiomyopathy
11. AF : Loss of atrial contraction

In atrial fibrillation, the murmur of MS may disappear at the end of a long diastole because

a. There may be such mild MS that the gradient disappears by the end of diastole.
b. There may be such low flow that although the gradient is still high at the end of a long diastole, the flow is too low to permit a murmur to be heard at the end of a long diastole.

Silent MS :

1. An almost completely immobile mitral valve, usually with adhesions, thickening and shortening of the chordae, which causes a second area of stenosis below the valve.[23]
2. Posteromedially deviated mitral valve orifice.
3. A large left atrial thrombus deviating the stream away from the apex.
4. A large ASD (lutembacher syndrome)

Etiology and Differential Diagnosis :

Etiology :

1. Rheumatic Heart Disease
2. A large left atrial myxoma
3. Congential MS : "Parachute" mitral valve
4. Calcified bacterial vegetation
5. Mitral ring constriction due to localized constrictive pericarditis
6. Carcinoid Syndrome with PFO / ASD

Differential Diagnosis :

1. Flow murmur – Severe MR, VSD
2. Hypertrophic cardiomyopathy
3. Normally functioning porcine valve
4. S3 which resembles MS
5. The Austin Flint Murmur
6. Coarctation of Aorta with secondary Subendocardial fibroelastosis.

Mitral Regurgitation (MR)

First heart sound M1 :

The torrential flow across a mitral valve causes a gradient only in early and mid diastole, whereas a stenotic valve, even with less-than-normal flow, causes a gradient across the mitral valve throughout diastole. About half the patients with pure MR have a soft M1. If the LV is not damaged, the M1 may be loud. About 70% of patients with papillary muscle dysfunction murmurs have a loud S1.

The Second Heart Sound S2

It has been assumed in the post that a widely split S2 occurring in severe mitral regurgitation (MR) is due to an early A2 secondary to shortening of the ejection time because of presence of two outlets for systole. However, even though there may be two outlets, there is

also more volume to be ejected. The S2 is not widely split in mild to moderate MR. LV ejection times are normal in MR unless the output is decreased at rest. MR can delay the onset of LV contraction.

MR can produce a widely split S2 and this indicates that the MR is at least moderately severe.

The Third Heart Sound S3 :

A loud S3 can occur in moderate to severe MR.

Differentiating an S3 from an OS

1. The OS is not usually more than 100ms from the S2. The S3 is rarely less than 120ms from the A2.
2. The OS is usually a short, sharp click, best heard with the diaphragm near the left sternal border. The S3 is a thud or boom, best heard by applying light or moderate pressure with the bell near the apex.
3. The OS is associated with a sharp, loud S1. The S3 may or may not have loud S1.
4. An OS separates further from the A2 when the patient stands. An S3 does not change its distance from the A2 on standing.

The systolic regurgitant murmur of MR :

"Holos" is a Greek Word meaning "Wholly", "Complete" "entire" and "all". Pan is a Greek Work meaning "each" "every" and "all". "Holosystolic" has only one meaning, which well fits the timing of the murmur (from S1 to S2).

The effect of long diastoles on MR murmur.

1. The loudness usually remains about the same because the LV has two outlets during systole. The quantity of retrograde flow in MR actually does not increase during the isovolumic contraction phase in the beat after the long pause but this does not affect the loudness of the murmur during the major part of systole, when the amount regurgitated is not increased.[24]

The MR murmur can become softer after a long diastole in.

1. The mitral valve prolapse syndrome
2. In some papillary dysfunction murmurs, when myocardial ischemia rather than fibrosis causes the MR. The long diastole may decrease myocardial ischemia by allowing more time for coronary filling and by decreasing the after load due to the increased time allowed for aortic pressure to fall.[25]

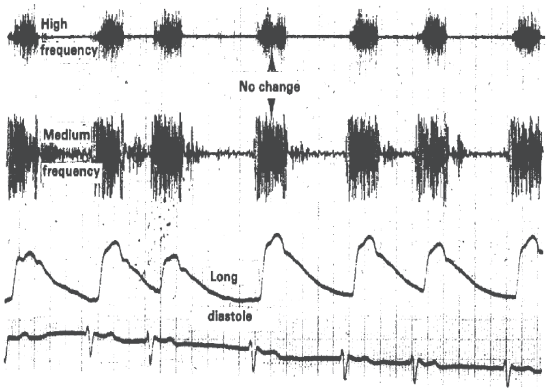


Fig 9:A high- and medium-frequency phonocardiogram taken at the apex together with an external carotid tracing from a 45-year-old woman with moderately severe chronic rheumatic MR, with few symptoms on digitalis alone. Because of atrial fibrillation,short and long diastoles are present, demonstrating that the murmur does not grow louder after long diastoles than after short or average diastoles.

In WPW type B preexcitation, the MR murmur is made louder after a long diastole.[26]

Causes of MR murmurs in the adult :

1. Rheumatic heart disease
2. Mitral valve prolapse
3. Papillary muscle dysfunction
4. Ruptured chordae
5. Left atrial myxoma
6. Calcified mitral annulus
7. Endocardial cushion defects with a cleft anterior leaflet
8. ASD with MR.

Causes of MR murmur at the apex in an infant :

1. Endocardial cushion defect
2. ALCAPA
3. Endocardial Fibroelastosis
4. Acute Myocarditis

5. Myxomatous degeneration of the mitral valve with or without Marfan's Syndrome.
6. Ebstein's Anomaly of the left atrioventricular (AV) valve (actually tricuspid valve) in corrected transposition of the great vessels.

Mitral annular dilatation is itself a rare cause of MR.

Causes of papillary muscle dysfunction murmurs:

1. Myocardial infarction, recent or old, with or without papillary muscle fibrosis.
   HOCM can cause MR
   MR murmurs of different shapes
1. Disproportionate lengths of papillary muscle-crescendo murmur to the S2
2. Papillary muscle shortend by fibrosis or is attached to an aneurysm or dilated akinetic area, then regurgitation is pansystolic.
3. The shape of the murmur may be decrescendo if dilatation was the major cause of the regurgitation. If the murmur is crescendo to the S2, it is more likely to be due to papillary muscle dysfunction. Rheumatic MR is rarely crescendo to the S2 except during the healing phase of acute rheumatic fever, usually in a patient under age 20, who has had an attack of rheumatic fever within a few months, during which there was also a pansystolic murmur.

Papillary muscle dysfunction due to HOCM almost always produces a decrescendo murmur.

If the murmur becomes louder as the patient compensates for failure, fixed rheumatic MR is suggested. Papillary muscle dysfunction murmurs tend to become softer as the heart becomes smaller with improvement of heart failure.

If an S4 is present, is strongly suggests papillary muscle dysfunction. Ruptured chordae superimposed on rheumatic heart disease produce a sudden onset of severe MR and failure (a loud S3, a murmur of grade 3/6 or more, and an increase in symptoms). A decrescendo, mixed frequency murmur associated with symptoms of high left atrial pressure (orthopnea or paroxysmal nocturnal dyspnea) suggests ruptured chordae of recent onset. The poor left atrial compliance may raise the V wave pressure to a very high peak in. systole (pressure as high as 70mm Hg has been recorded). This rise in left atrial pressure plus a precipitous fall in LV pressure toward the end of systole decrease the gradient and murmur toward the end of systole.

In sudden severe MR, the LV diastolic pressure may rise rapidly enough to exceed the left atrial pressure by mid or late diastole, and the mitral valves may close. This makes the M1 soft or inaudible.

If posterior chordae rupture, producing a flail posterior cusp, the stream of regurgitation may strike the atrial septum in such a way that murmurs with the shape and radiation (into the carotids) typical of AS are produced. Radiation of a posterior chordal rupture murmur may sometimes be heard better in the lower back than in the neck.[27]

The MR murmur caused by a rupture of the anterior chordae may radiate along the spine, and if loud, even to the top of the head.

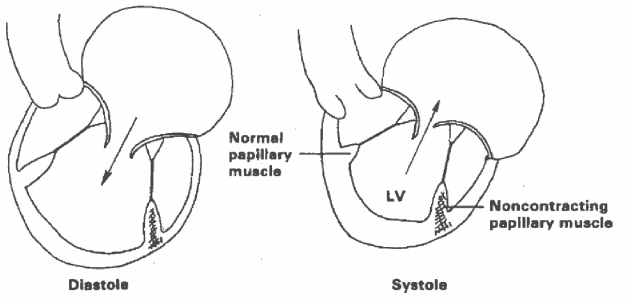


Fig 10. *A noncontracting papillary muscle may make its chordae-plus-papillary muscle relatively longer as the ventricle becomes smaller. This is most likely to produce a murmur that becomes progressively louder as systole proceeds*

Loudness, sites and radiation :

MR murmurs are loudest slightly lateral (1-2cm) to the site of the maximum apex impulse.

The MR murmur radiates best to the axilla and the left posterior interscapular area of the chest.

If the murmur is due to an endocardial cushion defect with a cleft anterior mtiral leaflet, the murmur may radiate better to the right than to the left of the apical impulse.

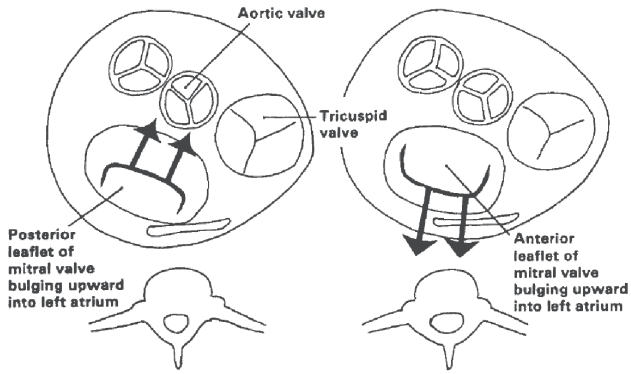


Fig 11.*These views of the valve rings from above show how posterior ruptured chordae (on left) can direct the regurgitant stream against the aorta and cause the murmur to be transmitted like an aortic ejection murmur. The diagram at right shows how ruptured anterior chordae can direct the regurgitant stream posteriorly against the spine.*

Causes of silent, severe MR :

1. Concomitant Ms
2. Obesity
3. Emphysema
4. Prosthetic Mitral Valve MR due to suture breakdown

Shape, Pitch and Duration :

Pansystolic murmurs with a slight crescendo-decrescendo (spindle shape on a phonocardiogram) are usually the loudest.

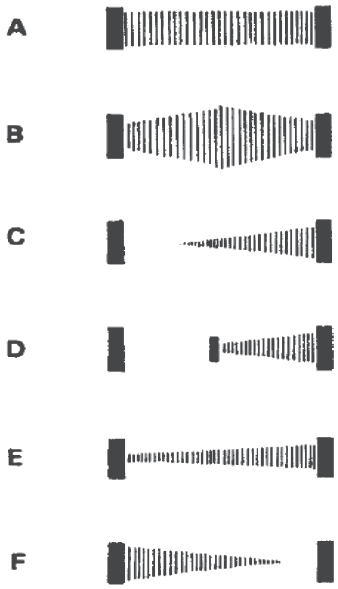


Fig 12:SHAPES OF MR MURMUR

Note that when the MR murmurs begin late, they always go to the second sound, and when they begin early, they always start with

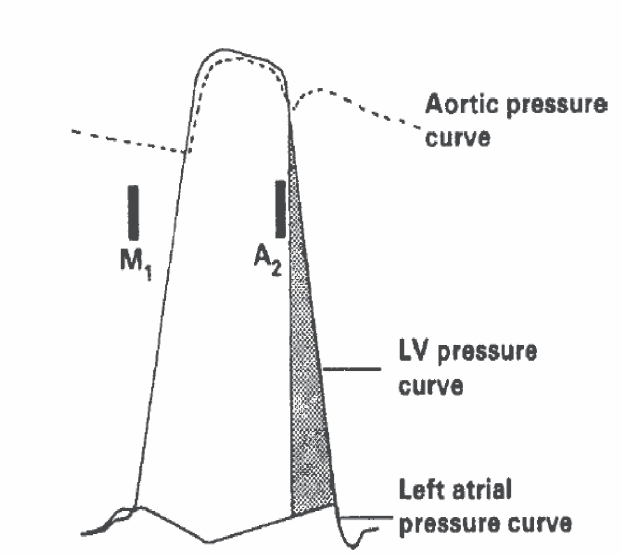


Fig 13 MR MURMUR EXTENDS BEYOND S2

Note that the LV pressure is above left atrial pressure, even after the A2.

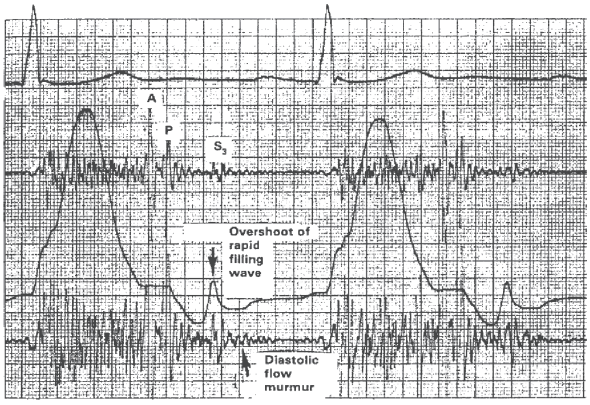


Fig 14. *This phonocardiogram and apical pulse tracing is from a 15-year-old girl with severe rheumatic MR. The pulse tracing was taken over the LV impulse in the supine position and is therefore an apex precordiogram instead of an apex cardiogram, which is taken in the left lateral decubitus position. The phonocardiograms are from the third left parasternal interspace. The upper one is taken at medium frequency; the lower one brings out low and medium frequencies. Note the following signs of severe MR: (1) the widely split S2 of 50 ms; (2) the diastolic flow murmur after the S3; (3) the exaggerated early rapid filling peak of the apical impulse (this would be palpable in the left lateral decubitus position).*

Quantitating Degrees of MR :

The MR is greater.

1. Large LV by palpation
2. Greater and later LPSH (left atrial pulsation)
3. Palpable S3
4. Louder and longer the apical systolic murmur (ruptured chordae may be decrescendo but are atleast Grade 4/6 in loudness).
5. More low and medium frequencies are present
6. Loud S3
7. Wide split S2

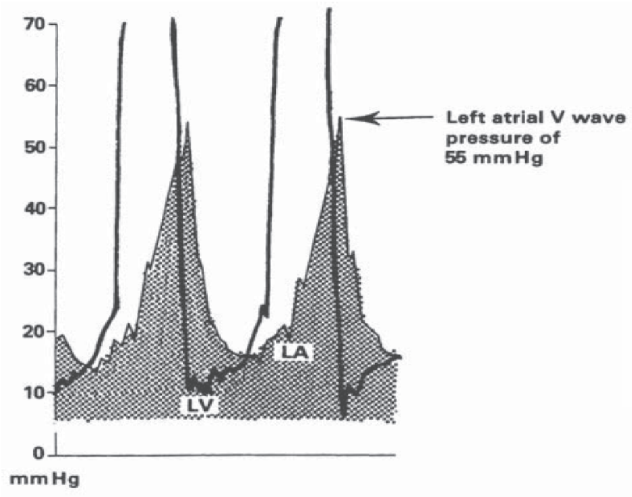


Fig 15: *This is a left atrial (wedge) and LV pressure tracing from a 23-year-old woman with ruptured mitral chordae. The shaded area is under the left atrial (wedge) pressure curve. The slight delay in the peak wedge pressure is due to the fact that wedgepressures (taken by a catheter wedged into the distal pulmonaryarterial branches) always show a delay in comparisonwith direct left atrial pressure tracings. The rapid increase inV-wave pressure during systole rapidly decreases the gradient across the mitral valve and will tend to cause both adecrescendo gradient and murmur. The decompressing effecton the LV of the massive loss of blood into the left atriumcauses a late systolic fall in LV pressure. This end-systolicdecrease in LV pressure further decreases the gradient acrossthe mitral valve toward the end of systole.*

Effects Of Drugs And Maneuvers

Increasing peripheral resistance:

Left sided regurgitant murmurs become louder.

Differentiating long systolic murmurs due to Aortic Stenosis (AS) or MR

With Handgrip, squatting or Phenylephrine, AS murmurs are unchanged, but MR murmurs become louder.

Standing, even though increases peripheral vascular resistance, does not increase MR

Decreasing Peripheral Resistance

Amyl Nitrite makes the MR murmur softer by decreasing peripheral vascular resistance.

Mitral Valve Prolapse Auscultatory Findings: One or more crisp systolic sounds or clicks and a late systolic or pansystolic MR murmur. This delayed systolic murmur is crescendo to S2.

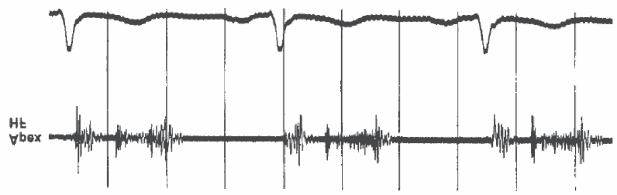


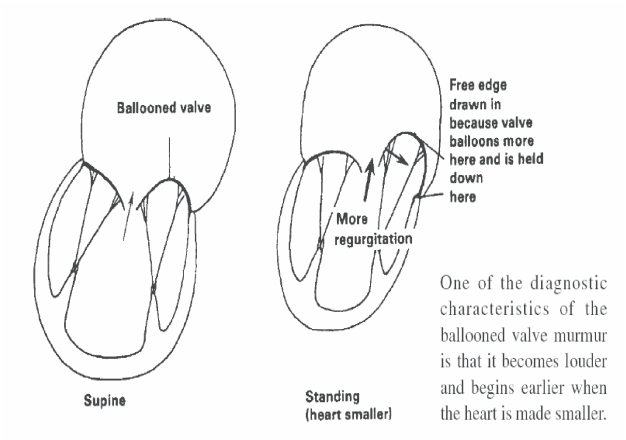
Fig 16: The midsystolic sound was a click heard loudest at the apex in this 45- year-old woman. The murmur following it is crescendo to the S2. This is the classic ballooned valve click-murmur complex by auscultation.

A pansystolic murmur can occur with even only late systolic prolapse on echocardiogram. This is because hearts with MVP usually have varying degrees of annular dilatation and those with pansystolic murmurs have the greatest dilatation.

Changing the Click and murmur with maneuvers or drugs.

Standing, Inspiration or a Valsalva Strain: The click and the murmur occur earlier and often become louder. The click may also become louder in the left lateral decubitus position. If the blood pressure (systolic pressure) decreases, the click and murmur become softer.

With Amyl Nitrite: The click occurs earlier and usually becomes softer. The smaller volume causes the click to come earlier and the low systolic blood pressure makes it softer. Usually the murmur becomes softer immediately and occurs earlier, but after about 30 sec it may become louder due to the overshoot of blood pressure. If the control murmur is only late systolic amyl nitrite may cause it to become pansystolic.



One of the diagnostic characteristics of the ballooned valve murmur is that it becomes louder and begins earlier when the heart is made smaller.

Differentiation of the late systolic murmur of papillary muscle dysfunction from the late systolic murmur of a prolapsed mitral valve (without click) by auscultation.

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Second Heart Sound in Congenital Heart Disease

R J Manjuran

Second heart sound evaluation is the most important assessment in physical diagnosis of congenital heart disease.

Normal Second Heart Sound (S_2)

Mechanism – The S_2 is due to vibration of cardiac structures and great vessels due to sudden cessation of blood flow at the end of systole. This corresponds to the coaptation of semilunar valves.

Components – The S_2 has two components – Aortic component (A_2) and Pulmonary component (P_2). Most investigators have documented that A_2 and P_2 correspond to Aortic and Pulmonary valve closure respectively.

Respiratory variation – In normal individuals both components of S_2 occur together during expiration while during inspiration A_2 and P_2 are separated (normal inspiratory splitting). The mechanisms of inspiratory splitting are:

1. Increased hang out interval of P_2
2. Increased venous return to right side of heart causes longer right ventricular ejection time.
3. Decrease in venous return to left side of heart results in shorter left ventricular ejection time.

Increased hang out interval is the most important cause of delayed P_2 during inspiration.

Clinical evaluation of S_2

1. Intensity
   - Relative to each component
   - Absolute increase
2. Respiratory variation

Abnormalities of S_2 in Congenital Heart Disease

Intersity of S_2 –

Palpable S_2 - Palpable S_2 in pulmonary area indicates palpable pulmonary component and is due to pulmonary arterial hypertension (PAH) eg. Eisenmenger syndrome.

Caution - In very thin chest walled individuals sometimes P_2 is palpable without PAH.

Loud P_2 - Indicates PAH – eg. Eisenmenger Syndrome. Normally P_2 is not audible at apex. However, audible P_2 at apex indicates PAH or ASD. In ASD, apex may be formed by right ventricle and in that situation P_2 is audible at apex.

Soft P_2 - P_2 can be soft because the sound is feeble as in valvular pulmonary stenosis or the pulmonary artery is posterior and hence P_2 is not well conducted to the chest as in Transposition of great arteries. In mild form of Tetralogy of Fallot sometimes a feeble P_2 is audible.

Loud A_2 - Loud A2 in congenital heart disease is due to

(1) augmented aortic closure as in coarctation of aorta, the aorta is anteriorly placed as in Transposition of Great. Arteries, Tetralogy of Fallot (TOF) especially, severe TOF
(2) abrupt closing motion of a pliable domed stenosed aortic Valve.

Soft A_2 - Can occur in congenital Aortic stenosis due to distortion of Aortic leaflet. A2 is soft in congenital aortic regurgitation.

Splitting of s2

Normal splitting - normal splitting of s_2 is heard in mild acyanotic congenital heart lesions like small ventricular septal defect (vsd) mild aortic stenosis., or mild pulmonary stenosis.

Wide variable split – in this, the s_2 is split during inspiration and expiration, but the split widens during inspiration. This type of split of s_2 can be present in moderate to severe pulmnonary stenosis, ebstein's anomaly, severe mitral regurgitation, moderate to large vsd and small atrial septal defect (asd).

Wide fixed split - The S_2 split is fixed during expiration and inspiration. This is a feature of moderate to large Atrial septal defect. Diagnosis of fixed splitting of S_2 must be made during normal breathing both in lying and upright positions.

Close splitting – Normal splitting of S_2 is appreciated when the interval between A_2 and P_2 is more than 20 msec. However, in PAH even when the split is equal to or less than 20 msec the split is appreciated. This is what is meant by close splitting. Patients of shunt lesion with PAH can have close splitting of S_2.

Single S_2 – The S_2 split is not appreciated during expiration or inspiration. This is because both the P2 and S_2 occur together as in Eisenmenger VSD or the A_2 is heard loud and the P_2 very feeble and hence a single S_2 is appreciated as in TOF or the pulmonary artery is posterior and aorta is anterior resulting in easily audible loud A_2 and poorly transmitted P_2 as in Transposition of great arteries or when there is only a single great vessel as in Truncus Arteriosus.

Conditions which can cause paradoxical split may present as single S_2 if the movement of P_2 is such that the $A_2 – P_2$ interval during inspiration and $P_2 – A_2$ interval during expiration is equal to or less than 20 msec.

Paradoxical split – The S_2 split is appreciated during expiration and during inspiration the split shortens or becomes a single S_2. This can occur in congenital severe AS, severe AR or large patent ductus arteriosus. The paradoxical splitting can have the following patterns.

Type I - Single S_2 during inspiration and split S_2 during expiration. This is the classic paradoxical split.

Type II - There will be normal split A2 – P_2 during inspiration while during expiration also the split is appreciated (P_2-A_2). This can masquerade as fixed split of S_2

Type III In this, the $P_2 – A_2$ patterns occur during expiration and $A_2 – P_2$ patterns during inspiration. However the separation of sounds both during inspiration and expiration is equal to or less than 20 msec and these results in a single S_2.

In type II paradoxical split the P_2 can be identified by auscultating from pulmonary area to apex and the sound, which softens and becomes inaudible is P_2. Valsalva maneuver also helps to identify paradoxical split. In strain phase paradoxically split S_2 widens and during release phase S_2 narrows while the opposite occurs with normal S_2.

Summary – The evaluation of S_2 is the most important single clinical assessment in the diagnosis of congenital heart disease.

DYNAMIC AUSCULTATION

Dr. G. Gnanavelu

Definition:

'Technique of altering circulatory dynamics by means of respiration and variety of physiological and pharmacological maneuvers and determining their effects on heart sounds and murmurs'

History:

ANTON MARIA VALSALVA (1707) 'De Aure Humane'(Anatomy and diseases of ear) described a simple way of raising the intrathoracic pressure WEBER (1860) described the cardiovascular effects of valsalva maneuver and observed that all the sounds associated with the movement of the heart disappear in response to the strain. POTAIN (1866) first noted normal respiratory variation in splitting of second sound. HAMILTON et al (1944) first reported an abnormal cardiovascular response with valsalva maneuver in patients with congestive heart failure AUBREY LEATHAM (1954) emphasized respiratory variation of second sound HENCKE (1960) first noticed augmentation of murmur of valvular aortic stenosis following longer cardiac cycle lengths. RIVERO CARVELLO observed increase in intensity of mid diastolic murmur of Tricuspid stenosis and holosystolic murmur of Tricuspid regurgitation during Inspiration Auscultatory changes may be induced by maneuvers or may spontaneously occur.

Spontaneous events :

1. Normal respiration
2. Premature beats
3. Myocardial ischemia

Maneuvers commonly used:

1. Postural changes
2. Valsalva maneuver
3. Muller maneuver
4. Isometric exercise
5. Vasoactive agents – Amyl nitrite, Methoxamine, Phenylephrine

TABLE 1 : IMPORTANT HEMODYNAMIC CHANGES IN MANEUVERS:

	Venous return	Systemic resistance	Stroke volume	Cardiac output	Arterial pressure	Heart rate	Heart size
Standing	⇓		•		•	•	•
Squatting	⇑	⇑	•		•	•	•
Valsalva Phase II	⇓	•	•	•	⇓	•	•
Isometric exercise				•	⇑	⇑	
Amylnitrite	•	•		•	⇓	•	•
Phenylephrine		•		•	⇑	•	•

Primary change : ⇓ decrease ⇑ increase Reflex changes: • Decrease • Increase

Respiration:

* Generally, it is best to assess respiratory variation in heart sounds and murmurs during normal respiration.

* Respiration exerts more pronounced & consistent alterations on murmur originating from right than from left side of heart.

* Right ventricular filling pressure is very high in right ventricular failure and pulmonary hypertension, there will be no increase in venous return with inspiration. Therefore there will be no inspiratory augmentation of right heart murmurs and gallops.

* Absence of respiratory influence is of no particular diagnostic value and cannot be used as evidence against any lesion.

* Effects of inspiration may be accentuated by use of Muller maneuver – forced inspiration against a closed glottis.

Muller Maneuver: the patient attempts to inhale with his mouth closed and his nostrils plugged, which leads to collapse of the airway. This leads to marked negative intrathoraic pressure resulting in increased systemic venous return.
Muller maneuver-technique : Patient is asked to pinch his nostrils and close his mouth and asked to inspire at the end of forced expiration.

Effects on splitting of S2:

* Observations are best made with patient in semiupright posture at 30 – 45 deg. angle or in the sitting position, since significant separation of A2-P2 may be heard during expiration in many normal subjects in supine position.

* When splitting of S2 is not readily apparent, separation of A2-P2 becomes obvious by inspiration immediately following valsalva maneuver.

* Muller maneuver may also be useful. Delay in P2 becomes even more obvious than with normal respiration.

Effects on murmurs:

* High frequency diastolic murmur of aortic regurgitation is most easily heard at the end of deep expiration. This is due to removal of insulation effect – volume of air acting as insulation between heart and stethoscope on inspiration.

* Right sided events are amplified due to increase in systemic venous return.

* Mid diastolic murmur of TS and holosystolic murmur of TR increase during inspiration. In sinus rhythm the presystolic murmur of TS is also selectively enhanced. When severe TS accompanies TR, the systolic murmur fails to increase with inspiration since stenosis prevents drop in right atrial pressure during inspiration and augmentation of right ventricular inflow.

* Early diastolic tricuspid flow murmur in ASD also increases with inspiration.

* Holosystolic murmur heard at lower left sternal border that is augmented by quiet inspiration is almost always due to TR. Holosystolic murmur due to other causes like MR, VSD, HOCM are dampened due to insulating effect of inspired air, if they change at all.

* Low frequency diastolic murmur of congenital pulmonary regurgitation increases on inspiration. High frequency diastolic murmur of pulmonary regurgitation due to pulmonary hypertension is less likely to be influenced by respiration due to hypertrophied right ventricle which has low compliance.

* Ejection systolic murmur of RVOT obstruction fails to vary predictably with respiration. Hence respiratory variation cannot be used to distinguish this murmur from innocent murmurs.

Effects on added sounds:

* Pulmonary ejection sound of pulmonary stenosis decreases in intensity on inspiration and moves closer to S1.
* RVS3 and RVS4 get augmented by inspiration as opposed to LVS3 and LVS4 which fade. But respiratory variation often lacks and therefore LV vs RV diastolic sounds are more reliably differentiated based on location.

Post premature ventricular contraction:

When PVC is followed by significant pause both increase in ventricular filling and augmentation of cardiac contractility occur.

1. Systolic murmur of Aortic stenosis/Pulmonary stenosis/HOCM gets augmented
2. Systolic murmur of Rheumatic Mitral regurgitation and Ventricular septal defect is not altered
3. Diastolic murmur of Aortic regurgitation becomes louder due to rapid ventricular filling and elevated arterial pressure
4. Increase in left ventricular size delays systolic click and systolic murmur of Mitral valve prolapse
5. Systolic murmur of Papillary muscle dysfunction actually decreases in intensity following a PVC due to post extrasystolic potentiation of contractile strength of ischemic papillary muscle and restraining effect of increased end diastolic volume on mitral valve apparatus.

Similar changes follow prolonged diastolic pauses in atrial fibrillation and sinus arrhythmia.

Effects of transient myocardial ischemia:

Hemodynamic changes during spontaneous angina or induced by exercise test:

LVEDP increases
Diastolic compliance of LV decreases
Wall motion abnormality
Papillary muscle dysfunction
Reduced stroke volume
Increase in arterial pressure

Auscultatory changes:

* Paradoxic splitting of S2 becomes obvious due to delayed left ventricular ejection
* S4 may develop during angina
* Late or midsystolic murmur of Papillary muscle dysfunction may appear or may get augmented .

Postural changes:

Recumbency & passive leg raising:

Hemodynamic Changes: venous return increases when assuming sudden supine position from sitting position and further enhanced by passive leg raising.

* Midsystolic murmur of pulmonary stenosis increases in intensity and duration during first few beats.
* Mid systolic murmur of aortic stenosis increases in intensity and duration after 3 – 6 seconds.
* Obstructive murmur of HOCM diminishes in intensity and duration due to increase in end diastolic volume.

Standing:

Rapid standing or sitting up from lying position or rapid standing from squatting posture results in opposite effect i.e venous return decreases due to venous pooling in legs and splanchnic vessels leading to momentary decrease in heart size, stroke volume, mean arterial presure followed by reflex increase in heart rate & systemic resistance.

Technique: Auscultation is carried out immediately before and after the change since effects may be quite transient persisting for only 10- 15 heart beats.
If patient is unable to sit upright or stand, rapid application of tourniquets at upper thigh level may result in trapping of blood to reduce venous return and reproduce similar response.

* Obstructive murmur of HOCM increases due to decrease in left ventricular volume
* Systolic murmur of Valvular pulmonary stenosis, aortic stenosis, mitral regurgitation and tricuspid regurgitation decreases.
* This posture is helpful to distinguish S4-S1 and S1-ES. S4 becomes softer and moves closer to S1 and after 30 seconds S4 may disappear altogether. Ejection sound remains audible..
* Systolic click with late systolic murmur of MVP : click occurs earlier and duration of murmur increases.
* Continuous murmur of PDA decreases in duration or disappears in sitting or standing position and this response suggests that the ductus is small. Hence it is important to examine children in supine position to avoid missing small PDA.

Prompt squatting:

Hemodynamic changes: There is momentary increase in venous return and stroke volume associated with increase in mean arterial pressure which is due to combined effect of increase in cardiac output and increased peripheral vascular resistance due to acute compression of leg arteries.

Technique: Examiner is seated on a chair/stool with stethoscope in place while patient is still standing so that he can be prepared to assess changes occuring within first few beats after squatting position is assumed.
In patients unable to squat, the same circulatory changes can be produced by bending patient's knees on his abdomen while he is in the supine position.

* Early diastolic murmur of Aortic regurgitation increases due to increased diastolic pressure.
* Holosystolic murmur of MR and VSD increase due to increased afterload.
* Midsystolic murmur of Aortic stenosis and Pulmonary stenosis increase due to increase in venous return and stroke volume.
* Systolic murmur of HOCM decreases due to greater LV filling and larger end diastolic volume leading to reduced obstruction.

Prone position & knee chest position :

Pericardial friction rub gets accentuated in these positions.

Valsalva maneuver:

Requires forced expiration against a closed glottis.

Technique:

Manometer method:

This method standardises the increase in intrathoracic pressure. Patient blows into the mercury manometer and maintains 40 mm Hg for 15 seconds.

Valsalva equivalent:

Patient is asked to push back against examiner's hand which is pressed downward on mid abdomen. This brings out sustained valsalva effort for 10 – 15 seconds needed to produce hemodynamic changes.

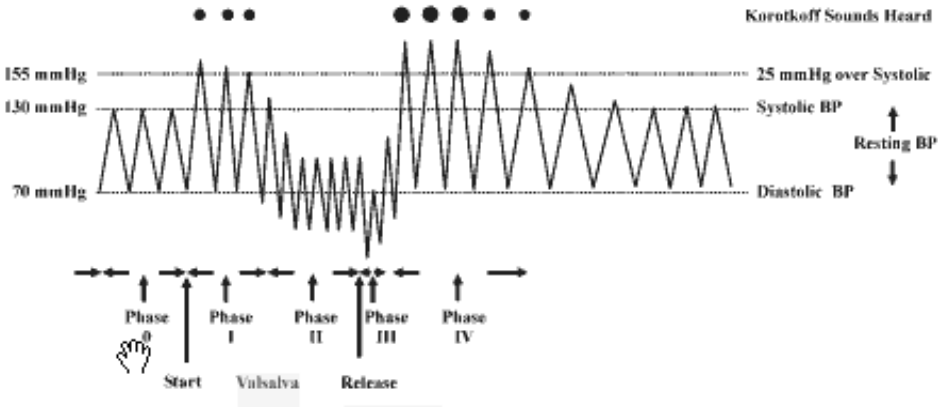
The maneuver is demonstrated to patient and the patient is allowed to practice the maneuver before trying to make assessments about the behaviour of the cardiac events.

Table 2 : PHASES OF VALSALVA MANEUVER & HEMODYNAMIC CHANGES

VALSALVA	HEART RATE	BLOOD PRESSURE	MECHANISM
Phase I 'Onset of strain'	No change	Increases	Increased intrathoracic pressure compresses vessels within thorax producing an abrupt increase in arterial pressure
Phase II 'Maintenance of strain'	Increase	Systolic pressure & Pulse pressure decrease	Increased intrathoracic pressure hinders venous return to right atrium leading to decreased cardiac output and systolic arterial pressure. Carotid baroreceptors trigger reflexes in the brainstem and inhibit the vagus nerve and stimulate the vasomotor center. Reflex tachycardia and peripheral vasoconstriction result.
Phase III 'Release of strain'		Further transient decrease in arterial pressure	Venous return and the capacity of the pulmonary vascular bed increase abruptly. Arterial pressure further diminishes because of transient pooling in the expanded pulmonary vascular bed.
Phase IV 'Relaxation'	Decrease	Overshoot of systolic pressure and pulse pressure increases	Accumulated venous return reaches the left ventricle and the increased stroke volume is pumped into the constricted systemic vascular bed, causing an overshoot of arterial pressure. This overshoot is detected by the carotid sinuses, resulting in excitatory effect on the vagus nerve center which manifests as a transient reflex bradycardia

Mechanical and reflex changes in circulation during valsalva maneuver depend on two factors:

1. level of cardiac function and effective central blood volume
2. speed and magnitude of baroreceptor responses to change in arterial pressure



The 4 Phases of Valsalva

Fig. 1 Four phases of Valsalva – Normal response : When the blood pressure cuff is inflated 25 mm Hg and maintained above the resting systolic pressure of the patient, the hearing of Korotkoff sounds are depicted as dots at the top.

Phase two can be used to distinguish fixed left ventricular outflow obstruction (valvular aortic stenosis) from dynamic obstruction. During phase two the murmurs of hypertrophic obstructive cardiomyopathy and mitral valve prolapse may increase as a result of the decreased stroke volume. Most other murmurs (including valvular aortic stenosis) decrease in intensity.

It is important to continue listening after Valsalva release. Phase four auscultation is useful in distinguishing left-sided from right-sided murmurs. Right-sided murmurs (such as pulmonic stenosis) that decrease in intensity during phase two will return to baseline intensity almost immediately after Valsalva release. Left-sided murmurs (such as aortic stenosis) require five to ten cardiac cycles to return to baseline.

Valsalva ratio:

Longest RR interval (following maneuver)

Shortest RR interval (late phase 2 or early phase 3)

Provides index of parasympathetic function. Test is performed three times and maximum value is recorded.
Normally ratio should exceed 1.4 in healthy children, but varies with age with higher ratios expected in younger patients.

Cardioaccelerator index: is the difference between resting heart rate & peak heart rate during phase 2.
An increase of > 20 bpm is expected.

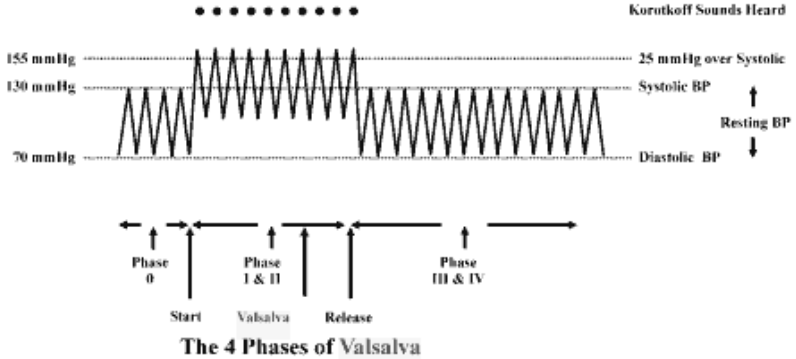
Change in heart rate vs change in Blood Pressure is used as an index of baroreceptor function.

Abnormal responses with normal autonomic function can occur in the presence of

Decreased intravascular volume
Congestive heart failure
Aortic stenosis/Mitral stenosis
Constrictive pericarditis
Atrial septal defect.

Square wave response: persistent elevation of systolic and diastolic pressures with no change in heart rate during forced expiration. There is no overshoot of blood pressure and no bradycardia when straining is stopped. Absence of bradycardia at the end of procedure is the easiest sign to detect.

This abnormal response occurs in congestive heart failure due to increased pulmonary blood volume with continued left heart filling for sustained period of time even during active straining. Left ventricular volume and stroke



The 4 Phases of Valsalva

Fig. 2 Four phases of Valsalva – Square wave response : When the blood pressure cuff is inflated 25 mm Hg and maintained above the resting systolic pressure of the patient, the hearing of Korotkoff sounds are depicted as dots at the top.

Isometric exercise:

Technique:

Isometric hand grip should be carried out using calibrated handgrip device or tennis ball or rolled up blood pressure cuff. First the maximum effect is measured. Then patient exerts 70 – 100% of this maximum for about 30 seconds.

* Useful to carryout isometric exercise bilaterally simultaneously.
* Should be avoided in patients with ventricular arrhythmias and myocardial ischemia, both may be intensified by this activity.
* Handgrip should be sustained for 20-30 seconds. But valsalva maneuver during handgrip should be avoided.
* Recent myocardial infarction, uncontrolled hypertension, cerebrovascular disease, suspected aortic dissection are contraindications to any form of isometric exercise.

Hemodynamic changes:

Transient but significant increase occurs in

Arterial pressure
Heart rate
Cardiac output
LV filling pressure
Heart size

Auscultatory changes:

1. In normal subjects sounds are little changed by isometric exercise.
2. LV S3 and LV S4 get accentuated
3. Appearance of S4 during isometric exercise is a more reliable indication of Left ventricular dysfunction than its presence during rest.
4. Systolic murmur of aortic stenosis gets diminised due to reduction of pressure gradient across aortic valve
5. Systolic murmur of HOCM diminishes and click and murmur of Mitral valve prolapse get delayed due to increase in left ventricular volume
6. Systolic murmur of Rheumatic mitral regurgitation and ventricular septal defect increases
7. Systolic murmur of Papillary muscle dysfunction gets augmented or is first recognised during handgrip.
8. Diastolic murmur of Aortic regurgitation increases
9. Diastolic murmur of Mitral stenosis gets louder

Pharmacological agents:

Commonly two agents are used in dynamic auscultation:

1. Amyl nitrite – vasodilator
2. Phenylephrine – vasoconstrictor

Amyl nitrite:

Technique:

Inhalation carried out by placing an ampoule in gauze near supine patient's nose and then crushing the ampoule. Patient is asked to take three or four deep breaths over 10 to 15 seconds after which amylnitrite is removed.

Hemodynamic changes:

Two phases: During first 30 seconds vasodilatation occurs leading to reduction in systemic arterial pressure. During next 30- 60 seconds reflex tachycardia occurs leading to increase in cardiac output and velocity of blood flow.

Most of the auscultatory changes occur in the first 30 seconds following inhalation.

1. S1 is augmented; A2 diminishes
2. MVOS; TVOS get louder; A2 OS interval shortens
3. RV S3 and LV S3 get augmented due to greater rapidity of filling
4. S3 of Mitral regurgitation is reduced since Mitral regurgitation decreases
5. Systolic murmur of Valvular Aortic stenosis/Pulmonary stenosis/HOCM/functional Tricuspid regurgitation gets accentuated
6. Systolic murmur of TOF diminishes since right to left shunt increases leading to decreased flow from right ventricle to pulmonary artery
7. Diastolic murmur of mitral stenosis/tricuspid stenosis/pulmonary regurgitation gets augmented due to increase in cardiac output.
8. Systolic murmur of Mitral regurgitation and ventricular septal defect and diastolic murmur of Aortic regurgitation diminish due to fall in systemic arterial pressure
9. Austin Flint murmur of aortic regurgitation, Continuous murmur of PDA and Systemic AV fistula diminish
10. In mitral valve prolapse – click occurs earlier due to reduced cardiac size and the intensity of murmur may have variable response

Table 3 : AMYL NITRITE IS USEFUL TO DISTINGUISH

	Augmented	**Diminished**
Systolic murmur	Aortic stenosis	Mitral regurgitation
Systolic murmur	Tricuspid regurgitation	Mitral regurgitation
Systolic murmur	Pulmonary stenosis	TOF
Systolic murmur	Pulmonary stenosis	Ventricular septal defect
Diastolic murmur	Mitral stenosis	Austin Flint of AR
Early diastolic murmur	Pulmonary regurgitation	Aortic regurgitation

Methoxamine & phenylephrine:

Technique:

Methoxamine is given intravenously 3 – 5 mg; elevates arterial pressure by 20-40 mmHg for 10 to 20 minutes

Phenylephrine is given intravenously 0.3 to 0.5 mg. 10 mg vial is diluted in 250 ml of isotonic saline and is given intravenously. It has shorter duration of action and elevates systolic pressure by approximately 30 mm Hg for only 3 to 5 minutes. BP is monitored and infusion is stopped when 20 mmHg rise in BP occurs or when intensity of murmur clearly changes.

Hemodynamic changes:

Two phases: During first 3- 5 mins. systemic arterial pressure increases followed by reflex bradycardia and decreased contractility and cardiac output.

They should not be used in the presence of congestive heart failure and essential hypertension.

Auscultatory changes are assessed during first phase:

1. S1 and A2 are diminished in intensity
2. A2-MV OS interval gets prolonged
3. S3 & S4 show variable response
4. Systolic murmur of HOCM gets softer due to increased LV size
5. Click and murmur of MVP get delayed due to increased LV size
6. Systolic murmur of aortic stenosis becomes softer due to reduced cardiac output
7. Diastolic murmur of Mitral stenosis diminishes
8. Diastolic murmur of Aortic regurgitation increases.
9. Systolic murmur of Rheumatic Mitral regurgitation, Ventricular septal defect, TOF, continuous murmur of PDA and systemic AV fistula get louder due to increase in Arterial pressure

Other maneuvers:

Carotid sinus massage:

During rapid heart rate, gallop sound in mid diastole is difficult to identify as S3 or S4. Adequate slowing of heart rate occurs during carotid sinus massage and closer proximity of S1 or S2 of the gallop is identified and differentiated as S3 or S4.

Carotid sinus stimulation may cause asystole or ventricular escape rhythm especially in elderly patients or in patients with underlying heart disease. Hence this maneuver is used only if sufficient slowing does not occur after reasonable period of rest.

Carotid sinus massage is contraindicated in the presence of carotid bruit. Therefore always auscultate over carotids before attempting carotid sinus massage.

External compression of chest wall:

Pulmonic mid systolic murmur in straight back syndrome gets accentuated by external compression of chest wall with stethoscope. Heart and great vessels closely approximate anterior chest wall so that firm pressure over pulmonary area magnifies the murmur.

Table 4 : DIFFERENCES BETWEEN MURMUR OF HOCM & MVP

	HOCM	MVP
Strain phase of Valsalva	Murmur increases in intensity	Murmur longer but not louder
AmyInitrite	Murmur louder	No change in intensity of murmur
Post PVC	Increase in intensity and in duration of murmur	Murmur remains unchanged or decreases
Standing	Murmur increases in intensity	Click occurs earlier, intensity of murmur is variable

Table 4 : USEFUL MANEUVERS IN DIFFERENTIATING SIMILAR AUSCULTATORY FINDINGS:

Systolic murmur	Valvular Aortic stenosis vs HOCM	sudden squatting, Valsalva maneuver
Systolic murmur	Valvular aortic stenosis vs mid-late MR	sudden standing, Amyl nitrite
Systolic murmur	Valvular Aortic stenosis vs Mitral regurgitation	Amyl nitrite, Phenylephrine, Variation in cycle length
Diastolic murmur	Mitral stenosis vs Austin Flint	Amyl nitrite
Diastolic murmur	Mitral stenosis vs Tricuspid stenosis	Respiration
Systolic murmur	Mitral regurgitation vs Tricuspid regurgitation	Respiration
Systolic murmur	Supraclavicular bruit vs Aortic stenosis	Extension of shoulder, compression of subclavian artery
Ejection click	Aortic stenosis vs Pulmonary stenosis	Respiration
Systolic murmur	Small VSD vs Pulmonary stenosis	Amyl nitrite, Phenylephrine
Systolic murmur	Large VSD with fixed vs hyperkinetic PAH	Amyl nitrite
Systolic murmur	Pulmonary stenosis vs TOF	Amyl nitrite
Continuous murmur	PDA vs cervical venous hum	compression of neck veins
S4 S1 vs Split S1		Respiration, Sudden standing, Lying with passive leg raising
S2OS vs split S2		Respiration, Phenylephrine, Sudden standing

Practical dynamic auscultation:

Dynamic auscultation is useful in differentiating murmurs with similar patterns during standard examination. Remember the dynamic alterations that occur during a maneuver and apply this knowledge to the physiology of the murmur in question, rather than memorizing the particular effect of a maneuver on a given murmur. Basically, the maneuvers of dynamic auscultation cause the following:

1. Increased ventricular volume/ejection due to increased venous return (right sided murmurs such as TR, PS are louder due to increased volume/flow)

   * Inspiration

2. Decreased ventricular volume/ejection due to decreased venous return (HOCM murmur is louder, MVP click moves closer to S1 and mitral regurgitation duration prolongs but the intensity of murmur may vary

   * Squatting to standing

   * Valsalva

3. Increased systemic arterial pressure (regurgitant murmurs such as MR and VSD are louder due to "back pressure," AS is softer due to decreased gradient across valve)

   * Isometric handgrip

4. Both increased systemic arterial pressure and increased venous return (HOCM murmur is softer due to increased ventricular volume, MVP click moves away from S1 and duration of mitral regurgitation is reduced but the intensity of murmur may vary)

   * Passive leg raise

   * Standing to squatting

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X-RAY OF HEART

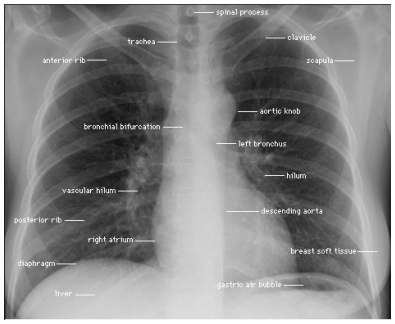
Dr. R. Ravikumar

How to go about reading a Chest X ray?

1. Check for patient details.
2. Check whether technical parameters are correct such as rotation, inspiratory/expiratory film, penetration etc….
3. Position, size and contour of the heart
4. Tracheal position
5. Mediastinal contours
6. Lung parenchyma and its vascular markings
7. Diaphragm and costophrenic angle
8. Soft tissues
9. Bones

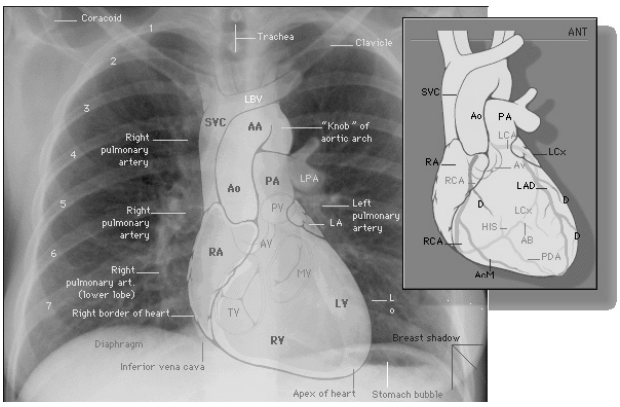
DO NOT FORGET TO REVIEW : Costophrenic angles, apices, retrocardiac portion, diaphragm and below it, breast shadows (in female).

Normal structures seen on chest X ray:

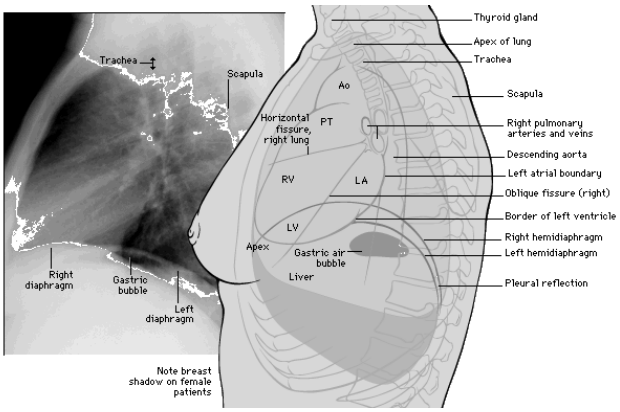


Right heart border is formed by Right brachiocephalic vein, SVC and right atrium.
Left heart border is formed by left subclavian artery, aortic knob, undivided segment of pulmonary artery, left atrium and left ventricle.

Aortic knob: In normal people the aortic knob measures less than 35mm and this is measured from the lateral border of trachea to lateral border of aortic knob.



PA View

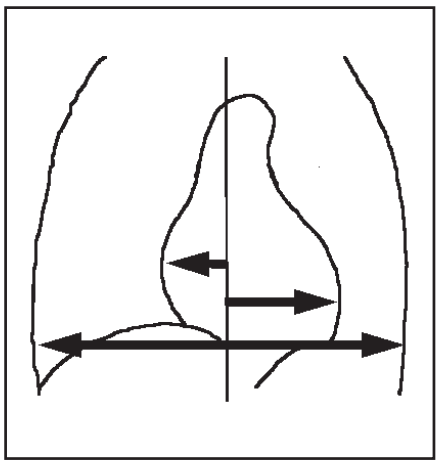


Lateral view:

Cardiothoracic (C-T) ratio:

It is the maximum transverse diameter of the heart divided by the greatest diameter of the horacic cage.
Normal C-T ratio- less than 50%

If CT ratio is greater than 50% it is suggestive of cardiomegaly.



Cardio-thoracic ratio

Psedoenlargement of CT ratio: Obesity, ascites, pectus excavatum, straight back syndrome.

Evaluation of Pulmonary Vasculature:

Right descending pulmonary artery measures <17.0mm.

Distribution of flow: Normally Increased vascular markings are noted within base in comparison to apex. In pulmonary venous hypertension blood flow to the apex becomes equal to or greater than the blood flow to base **(cephalization)**.

Central to peripheral marking: Normally pulmonary vessels taper gradually from central to peripheral. When peripheral vessels appear too small for the size of the central vessels (not gradual tapering) it is called pruning and results due to pulmonary arterial hypertension.

Identifying pulmonary vascular disease from vascular markings:

Normal pulmonary vasculature:

Right descending pulmonary artery measures <17.0mm
Lower lobe vessels are larger than the upper lobe vessels.
Gradual tapering of blood vessels from central to peripheral should be seen.

Pulmonary venous hypertension:

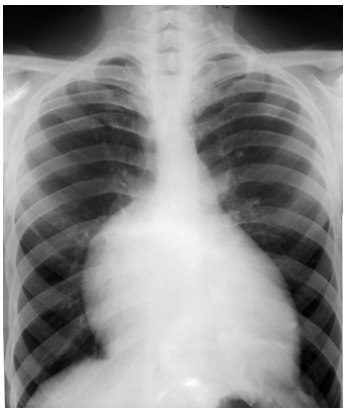
Right descending pulmonary artery e• 17.0mm
Cephalization of vascular markings.
Gradual tapering of blood vessels noted from central to peripheral.

Causes: Mitral stenosis, Left atrial myxoma, congestive heart failure, Hypoplastic left heart syndrome, Cor triatriatum, TAPVR from below the diaphragm, AS, coarctation of aorta.

Pulmonary arterial hypertension:

Right descending pulmonary artery >17.0mm
Lower lobe vessels are large than upper lobe vessels.
Rapid decreasing in size of peripheral vessels in comparison to central vessels (pruning)

Causes: Primary or Idiopathic Secondary to Lung disease- COPD, Fibrosis Pulmonary artery disease like aortitis, multiple emboli Chronic hypoxia, high altitude.

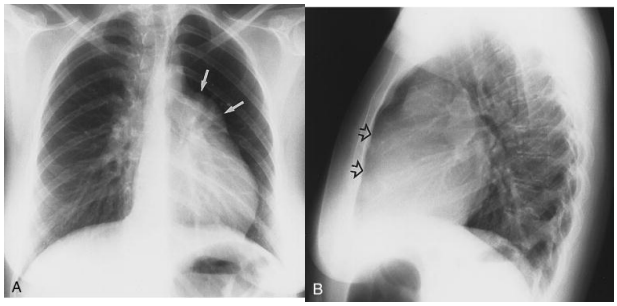


Identifying chamber enlargement:

Enlargement of right side more difficult to recognize.

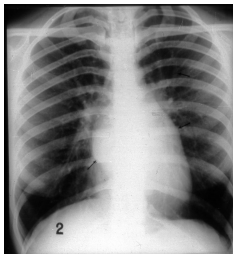
RA forms the right lateral cardiac border. Prominent RA extending more than 5 cms from mid line or exceed one third of the total diameter of the heart.

RV is better seen on lateral chest X ray. In RV enlargement the right ventricle fills in the space behind the sternum.



Left atrial enlargment:

Double density of the right cardiac shadow
Bulging of left atrial appendage along the middle of the left cardiac border
Posterior bulge of upper cardiac border on lateral view
Elevation of left main bronchus

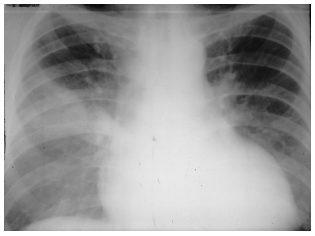


Arrow points to Double density shadow due to LA enlargment

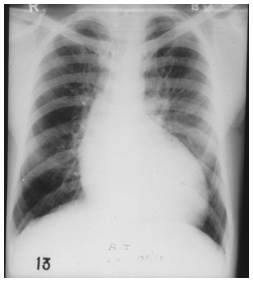
LV enlargement:

LV dilatation causes the cardiac apex to be displaced downwards towards the diaphragm and to the left.

LV hypertrophy causes the cardiac apex to become rounded.



LV dilatation



LV hypertrophy

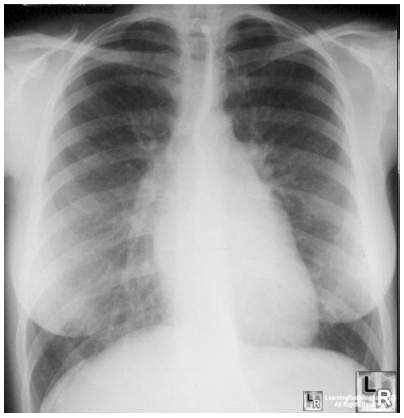
X ray findings in cardiac failure:

Enlarged Cardiac size
Peribronchial cuffing with air bronchogram
Loss of definition of subsegmental and segmental vessels
Perihilar or diffuse consolidation
Bat's wing appearence
Septal lines (Kerley B lines): short horizontal white linear densities very close to the peripheral margins of the lung parallel to diaphragm
Small pleural effusion
Thickening of interlobar fissure
Widening of Vascular Pedicle Width **(Normal 48±5 mm)**

Chest X ray findings can lag behind hemodynamic alterations but the following pattern can predict Pulmonary Artery Wedge Pressure (PAWP)

Grade 0: Normal PAWP <12 mm Hg
Grade 1: Pulmonary venous HT seen as pulmonary vascular redistribution (PAWP 12-19 mm Hg)
Grade 2: Interstitial edema ,Kerley B lines (PAWP 20-25 mm Hg)
Grade 3: Generalized or perihilar alveolar edema (PAWP >25 mm Hg)

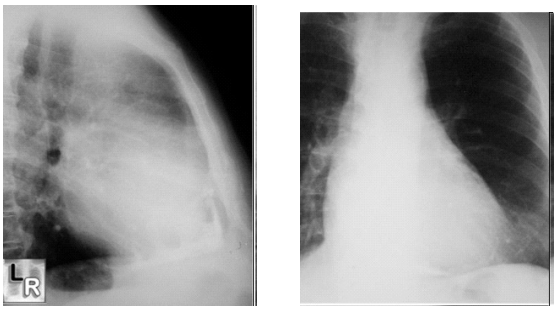
Mitral stenosis:



Normal or slightly enlarged C-T ratio
Straightening of left heart border
Small aortic knob from decreased cardiac output
"Double density" sign due to LA enlargement
Calcification of mitral valve (NOT annulus) seen best on lateral film
Calcification of left atrial wall indicating chronic MS
Calcification of pulmonary arteries due to PAH
Cephalization of pulmonary vascular markings
Elevation of left main bronchus

Constrictive Pericarditis:

Calcification of the pericardium is detected in upto 50%.
PN: A calcified pericardium is not necessarily a constrictive one.
Lateral chest film useful for its detection in the atrioventricular groove along the anterior and diaphragmatic surfaces of right ventricle.
Pleural effusion is seen in about 60%



Pericardial calcification

DD for pericardial calcification: coronary artery calcification, valvular calcification, calcified myocardial infarct, ventricular aneurysm, left atrial calcification, calcification outside the heart.

Differential diagnosis in cardiac X ray findings:

Cyanosis with decreased vascularity:

Tetrology of Fallot
Truncus arteriosus type 4
Tricuspid atresia
Transposition of great arteries
Ebsteins anomaly

Cyanosis with Increased vascularity:

Truncus type 1,2 3
TAPVR
Tricuspid atresia
Transposition
Single ventricle

Causes of left atrial enlargement:

Congestive heart failure
Mitral stenosis
Mitral regurgitation
Prolapsed mitral valve
Papillary muscle dysfunction
Left atrial myxoma

Causes of increase in main pulmonary artery:

L-R shunt
Pulmonary arterial hypertension
Pulmonary stenosis
Idiopathic dilatation of pulmonary artery

Prominent thoracic aorta:

Hypertension
Atherosclerosis
AS, AR
Aneurysm
Dissecting aneurysm
Aortitis
Posttraumatic rupture of aorta.

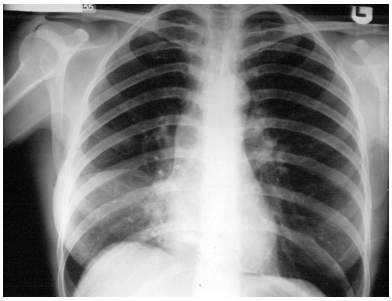
Causes of increased cardiac silhoutte:

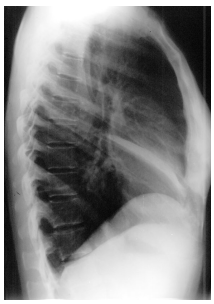
Cardiomyopathy
Severe Congestive cardiac failure
Pericardial effusion
Multi valve disease

SILHOUTTE sign:

An intra-thoracic radio-opacity, if in anatomic contact with a border of heart or aorta, will obscure that border. An intra-thoracic lesion not anatomically contiguous with a border or a normal structure will not obliterate that border.

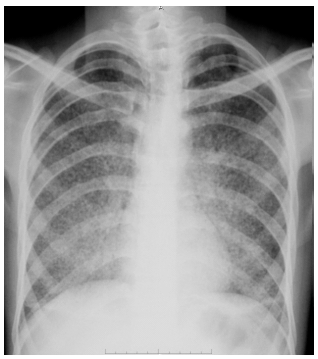
Middle lobe collapse causing obscuration of right heart border



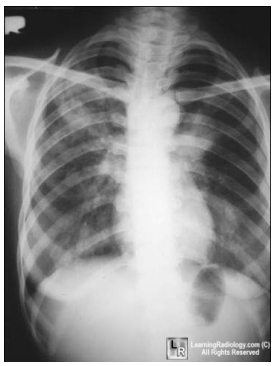


Masking of right heart border- think of right middle lobe pathology
Masking of posterior diaphragm- think of right lower lobe pathology
Masking of left ventricle – think of left upper lobe pathology
Masking of descending aorta- think of left lower lobe pathology
Masking of SVC and IVC- think of right lower and middle lobe pathology.

Pulmonary Tuberculosis:



Rt upper lobe cavitaion with bilateral hilar adenopathy



Miliary mottling

- Almost always affects apical or posterior segments of upper lobe or superior segment of lower lobe- Bilateral upper lobe disease is very common
- Pneumonic consolidation
- Cavitation
- Secondary bronchiectasis
- Bronchostenosis causing distal atelectasis " middle lobe syndrome"
- Tuberculoma usually single can be multiple. When single, presents as Solitary Pulmonary nodule (SPN) and is usually associated with small discrete shadows in the immediate vicinity of the lesion " satellite lesion"

- Pleural effusion. Formation of pleural effusion in post primary TB almost always means direct spread and should be regarded as empyema.
- Miliary tuberculosis

Acute Respiratory Distress syndrome (ARDS):

- There is delay in onset of any X –ray findings for 12 hours post onset of symptoms
- 12-24 hrs Patch alveolar infiltrates appear in both lungs and frequently peripheral in location
- 24-48 hrs Patch infiltrates coalesce producing massive air space consolidation of both lungs and frequently with air bronchogram
- 5-7 days Clearing is frequently secondary effect of CPP ventilation rather than true healing. If CPP ventilation has been administered watch for PULMONARY INTERSTITIAL EMPHYSEMA
- Pneumonia may superimpose. Always look for new focal infiltrates and pleural effusion
- Long term effects is seen as coarse reticular interstitial disease which may progress to fibrosis

DD for ARDS:

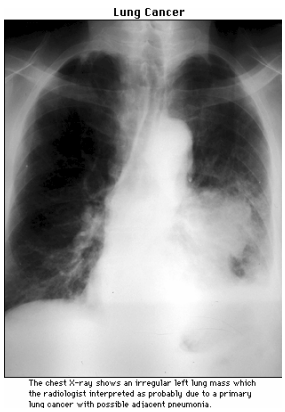
- Diffuse bacterial pneumonia
- Cardiogenic pulmonary edema
- Renal based pulmonary edema

PN: Kerley B lines and peribronchial cuffing not seen in ARDS and is seen in cardiac and renal pulmonary edema. Air bronchogram is seen in 70% of ARDS.

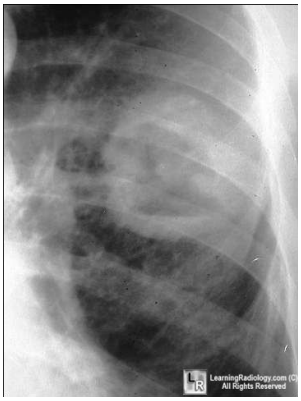
Solitary Pulmonary Nodule (SPN):

Well circumscribed, approximately round lesion that is less than 4-6 cms in diameter. By definition it is completely surrounded by aerated lung.

Cavitating lesion, lesion with multilobulated or speculated contour and lesion with shaggy or extremely irregular borders tend to be MALIGNANT.



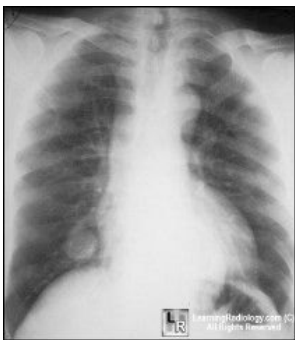
The chest X-ray shows an irregular left lung mass which the radiologist interpreted as probably due to a primary lung cancer with possible adjacent pneumonia.



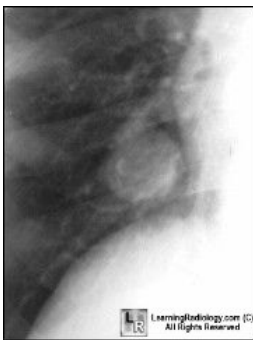
Malignant nodules grow at a steady, predictable exponential rate.

Growth of a nodule is conventionally defined as the doubling time. Doubling time is defined as the time required for its volume to double which corresponds to an increase in diameter by a factor of 1.26.

In general doubling time of > 16months or < 1 month are associated with benign process. If a nodule has not increased in size over a 2 year period the probability that the lesion is benign is >99%.



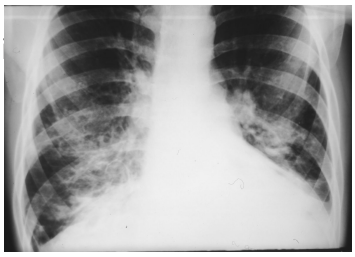
SPN



Hamartoma

Six common pattern of calcification is seen in SPN. If calcification within the lesion is diffuse, central, popcorn and laminar it fits into a benign lesion.
If the calcification within the lesion is stippled and eccenteric it fits in to being
a malignant lesion.

Bronchiectasis:



- Tramline or honeycombing represents dilated, thickened bronch
- Loss of lung volume due to destruction of lung tissue
- Multiple small nodular densities from plugged alveoli
- Lack of bronchial tapering
- Nonuniform bronchial dilatation
- Bronchial wall thickening

Interstitial lung disease (ILD):

Reticular pattern: Lung parenchyma is replaced by many thin walled cysts (lesions less than 10.0mm). These cysts are barely perceptible, round or oval thus giving the lung the radiologic appearance of a fine network.

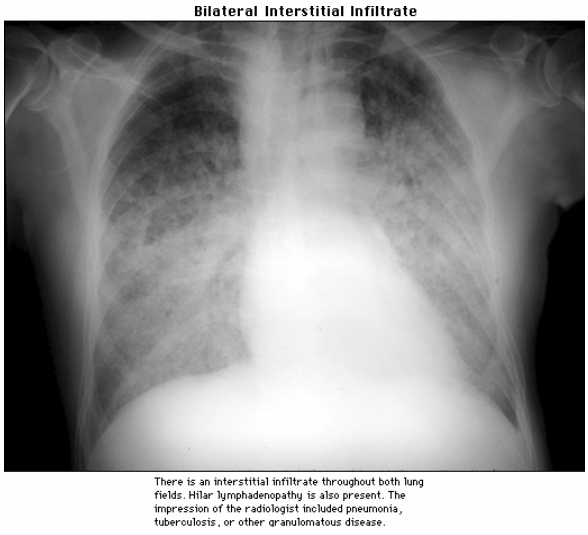
Miliary, Nodular: Numerous discrete, tiny less than 5.0mm uniform densities evenly distributed throughout the lungs.

Retculo-nodular: Mixture of both patterns

Kerley B lines: Usually less than 2cms in length and about 1 mm in thickness. Attributed to increased tissue and or fluid accumulation in interlobular septa

Kerley A lines: usually 4 cms in length, relatively staright, linear densities. Tend to be oriented perpendicular to the nearest pleural surface. Attributed to increased tissue and or fluid accumulation in communicating lymphatics between vein and bronchi.

Exaggerated bronchovascular markings.



Points to cover in a X ray chest report:

The domes of the diaphragms are evenly shaped and positioned in proper height.
The sinuses are not obliterated.
The pleura shows no thickening.
Both lung fields have the same transparency and no geographic or rounded densities.
There is a harmonic bronchovascular branching right into the periphery of the lungs.
The upper mediastinal shadow is not enlarged.
The tracheal band is not narrowed.
The hila are not enlarged.
There is no pathologic transformation of the cardiac silhouette.
The visualized parts of the skeleton are normal
The soft tissue of the chest wall is not conspicuous.

ACUTE RHEUMATIC FEVER IN INDIA – A MYTH OR A REALITY AT PRESENT

Dr. S. Thanikachalam

Rheumatic fever is an immunologically mediated connective tissue disease as a consequence of throat infections with group A Streptococci. Histopathological hallmark is an inflammatory process involving collagen fibrils and the ground substance of the connective tissue. Similarly involving joints, (transient) heart and to lesser extent central nervous system. The most dreaded consequence is rheumatic heart disease often affecting more than one valve maiming the prime youth of the affected person and often ending with significant morbidity and mortality.

The average age of presentation had been reported by Dr.Padmavathi to be between 10 to 14 years even though early developments of rheumatic heart disease less than 5 years of age have been reported. The prevalence of acute rheumatic fever was 0.05 to 1.7/ 1000 from 1940 to 1983 and 0.18 to 3/1000 from 1984-1995 and a much lower in the last decade.

Even though there has been steady decline in the incidence of acute rheumatic fever and rheumatic heart disease in the Western countries, there is selective trend of obvious decline in incidence of acute rheumatic fever only, in our country. This trend may be attributed to the improved nutrition of children as a result of improved socio-economic status of the community or may be due to diminishing streptococci virulence or because of presence of fewer Rheumatogenic serotypes in the last decade.

PATHOLOGY AND PATHOPHYSIOLOGY:

Rheumatic fever is a systemic disease affecting the peri-arteriolar connective tissue and can occur after an untreated Group A Beta hemolytic streptococcal pharyngeal infection. It is believed to be caused by antibody cross-reactivity. This cross-reactivity is a Type II hypersensitivity reaction and is termed molecular mimicry. Usually, self reactive B cells remain anergic in the periphery without T cell co-stimulation. During a Streptococcal infection, mature antigen presenting cells such as B cells present the bacterial antigen to CD4-T cells which differentiate into helper T2 cells. Helper T2 cells subsequently activate the B cells to become plasma cells and induce the production of antibodies against the cell wall of Streptococcus. However the antibodies may also react against the myocardium and joints, producing the symptoms of rheumatic fever.

Group A streptococcus pyogenes has a cell wall composed of branched polymers which sometimes contain "M proteins" that are highly antigenic. The antibodies which the immune system generates against the "M proteins" may cross react with cardiac myofiber protein myosin, heart muscle glycogen and smooth muscle cells of arteries, inducing cytokine release and tissue destruction. However, the only proven cross reaction is with perivascular connective tissue. This inflammation occurs through direct attachment of complement and Fc receptor-mediated recruitment of neutrophils and macrophages. Characteristic Aschoff bodies, composed of swollen eosinophilic collagen surrounded by lymphocytes and macrophages can be seen on light microscopy. The larger macrophages may become Aschoff giant cells. Acute rheumatic valvular lesions may also involve a cell-mediated immunity reaction as these lesions predominantly contain T-helper cells and macrophages.

In acute RF, these lesions can be found in any layer of the heart and is hence called pancarditis. The inflammation may cause a serofibrinous pericardial exudates described as "bread-and-butter" pericarditis, which usually resolves without sequelae. Involvement of the endocardium typically results

in fibrinoid necrosis and verrucae formation along the lines of closure of the left-sided heart valves. Warty projections arise from the deposition, while subendothelial lesions may induce irregular thickenings called MacCallum plaques.

Chronic rheumatic heart disease is characterized by repeated inflammation with fibrinous resolution. The cardinal anatomic changes of the valve include leaflet thickening, commissural fusion and shortening and thickening of the tendinous cords.

The classical lesion is an exudative and proliferative inflammatory reaction involving the collage and connective tissue. There is generalized vasculitis. The pathological hallmark is the presence of Aschoff cells. This is one of the modified forms of Histiocytic cells. There is pancarditis myocarditis, valvulitis and pericarditis and pathologically it is characterized by cellular infiltration, edema and hyaline degeneration. There is eventual thickening, fibrosis and calcification.

Even though the association of infection with Group A Beta hemolytic infection of the Upper respiratory tract and the subsequent development of acute rheumatic fever is well known, the exact mechanism is unknown largely due to lack of an animal model and only evidence of an Group A streptococcal infection is required for the confirmation after critical diagnosis of acute rheumatic fever. Only about 11% have throat cultures positive. This may be due to elimination of the organisms by host defense mechanisms during the latent period between the throat infection and subsequent development of rheumatic fever.

Clinical features:

Revised Jones criteria as a guide for diagnosis for Rheumatic heart disease have been approved by WHO study group.

Major Criteria:

1. Carditis
2. Polyarthritis, migratory
3. Erthema marginatum
4. Chorea
5. Subcutaneous nodules

Minor criteria

1. Fever
2. Arthralgia
3. Elevated acute phase reactants(ESR, CRP)
4. Prolonged PR interval in ECG

Plus evidence of preceding group-A streptococcal infection (culture, rapid antigen, and antibody rise/elevation)

Two major and one minor criteria or 2 minor criteria and plus evidence of preceding streptococcal infection indicate a high probability of rheumatic fever. In the three special categories listed below, the diagnosis of rheumatic fever is acceptable without 2 major or one major and 2 minor criterion. However, only for a and b can the requirement for evidence of a preceding streptococcal infection be ignored.

a. Chorea, if other causes have been excluded
b. Insidious or late-onset carditis with no other explanation

c. Rheumatic recurrence in patients with documented rheumatic heart disease or prior rheumatic fever, the presence of one major criterion, suggests a presumptive diagnosis of recurrence. Evidence of previous streptococcal infection is needed.

MAJOR CRITERIA:

CAROTIS:

The carditis of acute rheumatic fever is pan-carditis affecting pericardium, epicardium, myocardium and endocardium. Carditis occurs in 40-60% cases of rheumatic fever.

Valvulitis:

a. Valvular insufficiency is the most common defect commonly affecting Mitral Valve.
b. Carrey-coombs murmur of acute rheumatic fever is a sign of active mitral valvulitis. It is a soft high pitched early diastolic murmur, varying from day to day.
c. Isolated aortic valvular involvement is rare and other valvular involvements are unusual.

Pericarditis:

Manifested as chest pain, pericardial rub typical ECG changes or pericardial fluid on echocardiography

Myocarditis:

Myocarditis manifests itself as disproportionate tachycardia, soft heart sounds, cardiomegaly or congestive cardiac failure.

POLYARTHRITIS:

It occurs in 75% of cases and the usual presentation with red, swollen, warm and tender joints especially elbows, knees, ankles and wrist joint and is migratory in nature. Fingers, toes and spine are rarely affected. Resolution of joint symptoms occurs in 6 weeks and seldom produces chronic joint disease. In the last decade the classical presentation of swollen, red warm and tender presentation was absent in many children. The presentation was in the form of migrating arthralgia

CHOREA:

Even though it has been reported to occur as high as in 20% of patients, it is an uncommon manifestation in the present era.

ERYTHEMA MARGINATUM:

Though unique it is an infrequent clinical finding seen in less than 10% of patients. It presents as an evanescent, erthematous, non-tender and non-pruritic macular rash with pale center and serpenginous border over the trunk. Erythema marginatum is usually associated with chronic carditis. It is very rare in Indian patients.

SUBCUTANEOUS NODULES:

They present as non-tender, firm pea-sized nodules seen on extensor surfaces of the joint and it is seen in less than 3% of patients and chroinic carditis is usually present.

MINOR CRITERIA:

1. Fever: it ranges from 101-102 degree F.
2. Arthralgia: Diagnosed only in the absence of arthritis
3. Evidence of group A streptococcal infection: Positive throat culture, history of scarlet fever, or Elevated streptococcal antibodies ASO, Anti-DNAse B or anti-hyaluronidase

LABORATORY DIAGNOSIS:

Throat culture:

At the time of acute rheumatic fever, only 11% of the patients have positive throat culture for group A Streptococci.

Streptococcal antibody test:

ASO titre rise in 80% of patients. ASO titres vary with age, geographical region, and other fevers influencing streptococcal infection. ASO titre rises to maximum in 2-3 weeks after streptococcal infection and rapidly falls in next few months upto six months. Arthritis occurs at or close to the peak of the antibody response. Rising titre is more diagnostic. Anti DNAase –B and AH are indicators of recent streptococcal infection. Streptozyme test can be used as an adjuvant.

Acute phase reactants:

ESR, CRP are raised in almost all patients with carditis, arthritis and sometimes in Chorea.

ECG:

Prolonged PR interval is seen in ECG due to carditis. Slow tachycardia, AV block, QRS-T changes suggest myocarditis.

Chest radiography: Useful to assess cardiac size and pericarditis, pulmonary edema and increased pulmonary vascularity are other findings.

Echocardiography:

It may show endocardial, Myocardial and /or pericardial involvement and useful in cases with valve affection. In global hypokinesia of left and right ventricle, multiple non-infectious verrucae over the atrial surface, atrio-ventricular valves at the point of contact of leaflets, thickening of the valve leaflets and trivial mitral and aortic regurgitation are often identified during acute rheumatic fever. Further transient pericardial effusion and thickening of the epicardium can be observed transiently.

Treatment:

Treatment of group A streptococcal infection:

All patients with rheumatic fever must be treated irrespective of isolation of organism. Oral penicillin for 10 days or single intramuscular dose of 12, 00,000 units can be given. Patients allergic to penicillin, erythromycin can be given for 10 days

Treatment with Clinical Manifestations:

Arthritis:

Aspirin 100mg/kg/day to attain a serum level of 20% are required. However salicylates have to be withheld until diagnosis is established.

Carditis:

In absence of congestive cardiac failure salicylates are beneficial otherwise prednisolone 1-2 mg/kg/day is given. While tapering the steroids after 4 weeks salicylates has to be added and continued for 3-4 weeks to prevent rheumatic rebound. Bed rest is advised during carditis.

Guidelines for bed test:

Carditis status Management

No carditis	Bed rest for 2 weeks and gradual ambulation over 2 weeks even if no salicylates
Carditis with no cardiac enlargement	Bed rest for 4 weeks and gradual ambulation over 4 weeks
Carditis with cardiac enlargement	Bed rest for 6 weeks and gradual ambulation over 6 weeks
Carditis with heart failure	Strict bed rest as long as heart failure is present and gradual ambulation over 3 months

Syndenhams Chorea:

Diazepam is sufficient, in severe case haloperidol can be used.
Skin manifestations: No treatment is required

PROPHYLAXIS OF RHEUMATIC FEVER:

PRIMARY PROHYLAXIS

INTRAMUSCULAR	Benzathine Pencillin G	12,00,000 U(6,00,000U if weight<27 kg) once daily
ORAL	Penicillin V	500mg bid for 10
	Erthromycin	250mg qid daily for 10 days
	Others(Clindamycin nafcillin, ampicillin, amoxicillin,cephalexin)	Doses varies

Secondary prophylaxis:

INTRAMUSCULAR	Benzathine Penicillin G	12,00,000 U every 3-4 weeks
ORAL	Penicillin V Sulfadiazine	250mg bid daily 1god(0.5g od in children)
	Erythromycin stearate	250mg bid daily

RECOMMENDED READINGS:

1. Padmawati S. Rheumatic fever and rheumatic disease in development countries. *Bull WHO* 1978;56:543

2. Kutumbiah P. Rheumatic Fever and rheumatic heart disease in India. Review of 2 5 years of study and progress. *Indian J Pediat*
1958;25:240•5

3. Padmavati S. Present Status of Rheumatic Fever and Rheumatic Heart Disease in India. *Indian Heart J.* 1995, 47:395-398.

4. Padmavati S. Rheumatic fever and Rheumatic heart disease in India at the turn of the country. *Indian Heart Journal* 2 001;53:35-

5. Krishnaswami S, Joseph G, Richard J: Demand on tertiary care for cardiovascular disease in India: analysis of data for 1960-89. *Bull WHO* 1991;69:325-30.

6. Krishnaswami S, Joseph G, Chandy ST, Rajendran G, Zacharious TU - Rheumatic fever chemoprophylaxis in Adults. *JAPI* 1998;46:593-594.

7. WHO Technical Report Series. Rheumatic Fever and Rheumatic Heart Disease 198. Geneva, No. 764,35-6.

ASSESSMENT OF MULTIVALVULAR LESIONS

Dr. N. Sudhayakumar

Accurate assessment of isolated valve lesions is fairly simple by a meticulous clinical evaluation supported by judicious application of investigation modalities. However the issue may become difficult quite often in the setting of multivalvular lesions, as the hemodynamics of one will significantly influence those of the other. Though in general the proximal lesion influences the hemodynamics of the distal lesion, the reverse is also important. Basically the clinical and investigation findings are determined by the hemodynamics – volume of flow , rate of flow and pressure gradients. eg. in mitral stenosis the findings are influenced by the left atrial volume, left atrial pressure and transmitral gradient. The following discussion will concentrate on the influence of aortc valve lesion on mitral valve lesion and vice versa.

Aortic stenosis vs mitral stenosis

Basic hemodynamic abnormality in mitral stenosis is the diastolic pressure gradient from left atrium to left ventricle i.e. the difference between the left atrial and left ventricular pressure in diastole. In the presence of significant aortic stenosis the left ventricular relaxation is impaired and the left ventricular diastolic pressure is likely to be elevated thereby attenuating the transmitral gradient. In addition, due to impairment of relaxation the isovolumetric relaxation time is prolonged. These changes can result in the following modifications of clinical data.

a) isovolumetric relaxation is the period from the end of systole (A2) to the opening of mitral valve (OS) . In severe aortic stenosis the isovolumetric relaxation time is prolonged and hence the A2 – OS interval can become longer and the transmitral flow time (length of mitral middiastolic murmur) can be shorter, thereby under assessing the severity of mitral stenosis.

b) In severe aortic stenosis, the left ventricular diastolic pressure gets elevated resulting in reduction in the transmitral diastolic gradient thereby reducing the intensity of the mitral diastolic murmur.

c) As the elevated left ventricular diastolic pressure interferes with the left atrial emptying, the left atrial pressure can go up further resulting in aggravation of symptoms of pulmonary congestion.

Thus in presence of significant aortic stenosis, the symptoms of mitral stenosis can get exaggerated with attenuation of the auscultatory findings.

The radiologic findings of mitral stensis are not modified by aortic stenosis ; but the echocardiographic data can be altered. One of the simple methods of calculating the mitral valve area is by applying the pressure half time principle(MVA = 220 / PHT). Rate of drop of transmitral gradient is influenced by the left ventricular compliance. When the LV compliance is reduced, the pressure half time is prolonged thereby under assessing the valve area. However the calculation of valve area by applying the principle of continuity equation is unaltered and hence it has to be applied (A1 x V1 = A2 x V2) ; pulmonary flow velocity integral , pulmonary annular area and mitral flow velocity integral should be used.

Hemodynamics of mitral stenosis do not significantly alter those of aortic stenosis unless the mitral stenosis is severe and has produced pulmonary arterial hypertension / cardiac failure, when the low cardiac output attenuates the transaortic gradient and the auscultatory findings of aortic stenosis. The low cardiac output is likely to exaggerate the symptoms of aortic stenosis. One of the important

finding of severe aortic stenosis ie. S4 will be absent in presence of mitral stenosis. Mitral stenosis increases the risk for development of atrial fibrillation which can be detrimental in aortic stenosis.

Aortic stenosis vs mitral regurgitation

As the hemodynamic abnormalities in both these lesions are in systole, one lesion can influence the other. In the presence of aortic stenosis (AS), the LV systolic pressure is elevated and hence the LV – LA systolic gradient for mitral regurgitation (MR) is exaggerated leading to aggravation of MR. The regurgitant fraction increases and the forward stroke volume comes down. Hence in presence of AS the findings of MR are accentuated and those of AS are attenuated as follows:

- Pulse – volume becomes lower; but character is unaltered.
- Apex – more sustained
- S2 – MR results in early A2 ; AS results in delayed A2
- MR murmur is accentuated ; AS murmur is attenuated
- Echo – exaggeration of findings of MR ; reduction in LV- aorta gradient.

Calculation of aortic valve area — continuity equation principle is the better method. Velocity time integral at LV outflow tract (below aortic valve), area of LV outflow tract and velocity time integral at the aortic orifice are applied in the equation A1 x V1 = A2 x V2 to get the aortic valve area.

Aortic regurgitation vs mitral stenosis.

Before analyzing the hemodynamic data, the usual bedside problem of diagnosing mitral stenosis (MS) in presence of significant aortic regurgitation (AR) shall be discussed. Though this is not a major issue with the availability of echocardiographic facilities, it continues to be a problem in the examination. I shall mention the following points to diagnose MS in presence of AR ie Austin Flint murmur Vs mitral stenosis.

- Opening snap ; loud S1
- Apical diastolic thrill
- Long loud MDM with presystolic accentuation (though described with Austin Flint murmur, it is a rare finding without MS)
- Findings of pulmonary arterial hypertension
- Features of chronic severe pulmonary venous hypertension
- Presence of atrial fibrillation

The hemodynamics and hence the clinical findings of AR are not significantly altered by presence of MS unless the patient has severe pulmonary hypertension or heart failure when the findings of AR can be attenuated. However the auscultatory findings of MS can be attenuated by severe AR as the elevated LV diastolic pressure can reduce the transmitral gradient. Hence assessment of severity of mitral stenosis by pressure half time can be modified by the presence of AR.

Aortic regurgitation vs mitral regurgitation

In AR the LV systolic pressure is high and hence the MR gets exaggerated. Because of the regurgitation of blood into left atrium during systole, the forward stroke volume into the aorta is reduced thereby producing systolic decapitation of the blood pressure. The following changes can occur:

- Pulse , BP – systolic decapitation of BP ; however the diastolic pressure and pulse character of AR are not modified by MR.
- S2 - AR - delayed A2 ; MR — early A2
- Murmur — MR murmur is accentuated ; AR murmur is unaltered
- Echocardiographic findings of MR are exaggerated ; AR findings are not modified by MR

Mitral stenosis + mitral regurgitation

Combined mitral lesion is the commonest lesion in rheumatic heart disease. Usually one lesion dominates; severe stenosis and severe regurgitation can't coexist. Mitral stenosis doesn't significantly influence the hemodynamics and clinical findings of MR. However MR can enhance the findings of mitral stenosis. The increase in left atrial volume due to MR elevates the left atrial pressure and hence can exaggerate the clinical and echocardiographic findings of MS.

Sometimes the diagnosis of associated mitral stenosis in severe MR itself is difficult as we can get a mid-diastolic flow murmur in severe MR. In addition the LVS3 of MR can be confused with an opening snap though the pitch of the sounds are different. A loud S1, diastolic thrill and presystolic murmur favour associated stenosis whereas in rheumatic severe isolated MR, S1 is usually soft and the flow rumble is a decrescendo one confined to middiastole.

Aortic stenosis + aortic regurgitation

A common dilemma in examination is the diagnosis of aortic stenosis in a patient with severe AR as the latter can produce a systolic murmur in the aortic area which may be conducted to the carotids. A significant aortic gradient also will be there in severe AR. Systolic decapitation of BP, slow upstroke of the carotid pulse, systolic thrill and long systolic murmur with late systolic peaking favour associated aortic stenosis.

Influence of tricuspid valve disease

Tricuspid valve disease – both stenosis and regurgitation – reduces the right ventricular output to the pulmonary circulation resulting in reduction in the left heart volume. This leads to

a. reduction in left atrial volume and pressure thereby resulting in partial relief of pulmonary congestion symptoms; the murmur of mitral stenosis becomes shorter and A2 – OS interval lengthens. The transmitral gradient also decreases. Thus the severity of mitral stenosis is likely to be under assessed.
b. The reduction in the left ventricular preload due to the tricuspid lesion produces a fall in the LV stroke volume and cardiac output which can exaggerate the symptoms of aortic stenosis. The pulse volume becomes still lower. Thus the severity of aortic stenosis will be overestimated. However the murmur of aortic stenosis will be relatively short.

The above discussion may give an impression that it is extremely difficult to assess the severity of individual lesions in multivalvular disease; this is not true. With a clear understanding of the hemodynamics, the clinical findings have to be interpreted cautiously and a meticulous echocardiographic evaluation with the limitations in mind along with ECG and skiagram of the chest can help us in the accurate assessment of each lesion and seldom is there a need for an invasive procedure.

CLINICAL APPROACH TO CYANOTIC CONGENITAL HEART DISEASE

G. Madhusudan, R. Suresh Kumar

Cyanosis is a bluish discolouration of the fingers, lips and mucous membranes. **Central cyanosis** is associated with reduced Hemoglobin concentration of >3g% in arterial blood gas or >5g% in capillary blood gas. **Peripheral cyanosis** is seen in conditions of sluggish peripheral blood flow and has normal arterial oxygen saturation.

Cyanosis in newborns

In newborns, central cyanosis has many causes, which may be broadly divided into – cardiac, pulmonary, neurologic or neuromuscular. This differentiation has important implications in the management of the patient. Clinical findings point to the correct system that causes cyanosis. A patient with a respiratory cause of cyanosis has associated respiratory distress and lung signs. A patient with cyanosis due to neuromuscular disease usually has weak respiratory effort. Patients with CNS impairment have associated changes in sensorium, neurological signs and weak respiratory effort. Babies with a cardiac cause usually have a 'peaceful' cyanosis and tachypnea. Crying may improve the cyanosis caused by lung disease or CNS depression; however, crying usually worsens cyanosis in patients with cyanotic heart defects.

Causes and clinical features of central cyanosis in newborns:

Evaluation of the cyanotic newborn:

The newborn with apparent cyanosis needs confirmation of the arterial desaturation by either bedside measurement of the pulse-oximeter Oxygen saturation (SpO2), or by performing an arterial blood gas. Clinical evaluation helps in differentiating the basic group of diagnosis – cyanotic congenital heart disease with reduced pulmonary blood flow or with increased pulmonary blood flow. Chest X-ray and ECG help in narrowing the diagnostic possibilities.

Hyperoxia test:

This test helps to differentiate cyanosis caused by cardiac disease from that caused by pulmonary disease. When central cyanosis has been confirmed by arterial partial pressure of oxygen (Po2), one tests the response of arterial Po2 to 100% oxygen inhalation (hyperoxia test). Oxygen should be administered through a plastic hood (such as an oxyhood) for at least 10 minutes in order to fill the alveolar space completely with oxygen. With pulmonary disease, arterial Po2 usually rises to more than 100 mm Hg. When there is a significant intracardiac right-to-left shunt, the arterial Po2 does not exceed 100 mm Hg, and the rise is usually not more than 10 to 30 mm Hg. However, some infants with cyanotic defects with a large pulmonary blood flow, such as total anomalous pulmonary venous connection (TAPVC), may have a rise in arterial Po2 to 100 mm Hg or higher. Conversely, infants with a massive intrapulmonary shunt from lung disease (but with a normal heart) may not have a rise in arterial Po2 to 100 mm Hg. Therefore, the response of Po2 to 100% oxygen inhalation should be interpreted in light of clinical picture, especially the degree of pulmonary pathology seen on chest x-ray.

Clinical evaluation:

Cyanotic congenital heart disease can be divided according to whether pulmonary blood flow is decreased (Tetralogy of Fallot (TOF), pulmonary atresia with intact ventricular septum, tricuspid atresia) or increased (transposition of the great vessels, single ventricle, truncus arteriosus, total anomalous pulmonary venous return).

A quiet precordium, single S2 and a prominent ejection systolic murmur characterise cyanotic CHD with reduced pulmonary blood flow. Active precordium, epigastric pulsation and features of congestive heart failure, usually with mild cyanosis, characterise cyanotic CHD with increased pulmonary blood flow. Profound cyanosis, no murmur and peaceful tachypnoea point to TGA.

Cyanotic lesions with decreased pulmonary blood flow:

These lesions must include both an obstruction to pulmonary blood flow and a large VSD. Common lesions in this group include TOF, double outlet right ventricle with PS, single ventricle with PS, corrected transposition of great vessels / VSD / PS, AV canal defect / PS or Critical PS / pulmonary atresia with intact IVS. Tricuspid atresia with reduced pulmonary blood flow would also belong to this category. Clinically all such children have a quiet precordium, single S2 and pulmonary ejection murmur. ECG shows RVH and RAD. LV dominance suggests tricuspid atresia. Left axis deviation suggests tricuspid atresia, AV canal defect with PS or single ventricle with PS.

Cyanotic CHD with normal / increased pulmonary blood flow:

This group comprises transposition of great arteries (TGA), tricuspid atresia with increased pulmonary blood flow, truncus arteriosus, AV canal defect, single ventricle, TAPVC, TGA with intact IVS presence on day 1 with profound cyanosis. Characteristically the baby has SpO2 <60%, tachypnea and no murmur. ECG shows RVH and RAD. Chest x-ray shows typical egg-on-side silhoutte with narrow pedicle and normal lung vascularity. Indeed, finding normal lung vascularity in a baby with profound hypoxemia is very suggestive of TGA.

All the other conditions tend to present at 3 – 6 weeks of age with congestive heart failure. Cardiomegaly, single S2 (except in TAPVC where S2 is widely split) and systolic murmur at the left sternal border are common features. Cyanosis is mild. Bounding pulses point to truncus arteriosus. LVH in ECG suggests tricuspid atresia whereas left axis deviation points to tricuspid atresia, AV canal defect or some form of single ventricle. Biventricular hypertrophy is seen in complete AV canal defect and truncus arteriosus. Cardiomegaly with increased pulmonary blood flow is seen in all the cases. High take off of central pulmonary arteries is a feature of truncus arteriosus. "Figure of 8" appearance and sometimes, pulmonary oedema characterise supracardiac TAPVC.

Cyanotic CHD in older children and adults:

The two common causes are Eisenmenger syndrome or Tetralogy of Fallot. Complex CHD has very low chance of survival. History of recurrent respiratory infection in early childhood and present good effort tolerance point to Eisenmenger syndrome. History of squatting and cyanotic spells suggest TOF. Squatting helps the TOF child by increasing systemic vascular resistance (SVR), by kinking femoral arteries, locking up desaturated systemic venous blood by kinking femoral veins and by displacing better saturated mesenteric venous blood into the heart. Cyanotic spell is a rapid worsening of cyanosis accompanied by hyperventilation. If unchecked, it progresses to unconsciousness, seizures and death. Abrupt decrease in SVR and infundibular spasm are the incriminated triggering mechanisms, which increase right to left shunt. Progressive decrease in SVR, acidosis and hyperventilation set up a vicious cycle.

Eisenmenger syndrome is characterised by features of pulmonary hypertension and right to left shunt. The patients have good effort tolerance till late in the natural history. Presentation in late adulthood, cardiomegaly, raised JVP, atrial fibrillation and widely split S2 point to ASD. Chest x-ray shows cardiomegaly and prominent central pulmonary arteries. Presentation in childhood is a feature of VSD or PDA. Single S2 suggests VSD. Normally split S2 with loud P2 is a feature of PDA. PDA also has differential cyanosis. Both VSD and PDA tend to have normal heart size on x-ray with dilated pulmonary arteries.

Conclusion:

Clinical evaluation assisted by x-ray and ECG goes a long way in the diagnosis of cyanotic CHD. Further refinement in diagnosis is achieved by echocardiography and other imaging techniques.

PRESSURE MEASUREMENT

Dr. Lakshmi Sriram

Pressure is force per unit area. Standard unit for pressure is Pascal (Pa). A Pascal is force of one Newton acting uniformly on an area of $1m^2$.

In cardio-vascular physiology, units include cm of water and mm of mercury.

Mm of mercury (mmHg) is defined as the pressure exerted by a column of liquid 1mm in height with a density of 13.5951 gm/cm^3 under standard acceleration due to gravity. In practice, this definition is realized by a column of mercury 1mm in height at 0^0 c and at standard atmospheric pressure.

Pressure conversion:

1mmHg = 133.3 Pa.
1mmHg = 1.36 cm H_20

In catheterization practice, pressures are often measured with reference to a point at mid chest level in the supine position and thus, are relative, not absolute values. Other choices include relating zero to the sternal angle or 10 cm above table top, but these levels have no reference to the patient's size. Some identify the center of the heart, using a lateral X-ray done with the patient standing, and relate zero to this point. This is time consuming and has no relation to the position of the heart when the patient is supine.

Pressure recording system consists of many parts namely the catheter, transducer, amplifier and recorder. Any such system must meet the following criteria: (a) amplitude linearity, (b) adequate frequency response and (c) phase linearity.

Amplitude linearity:

It refers to the ability of the transducer – processor – recorder system to produce an output signal that is directly proportional in magnitude to the amplitude of the input signal.

Calibration:

Must be carried out over a range of input amplitudes. Calibration of BP recording system are done against a column of mercury. Incremental increases in mmHg (input) are applied to the pressure transducer and the resulting output signals are measured and a plot of the input-output values results in a linear calibration relationship.

Frequency response and phase linearity:

It refers to the ability of recording system to provide a signal that is identical (except in amplitude) to the event presented to it. The dynamic frequency response of a recording system must be great enough to detect the highest harmonic of the cardio-vascular variable being recorded.

With regard to frequency characteristics, linearity means that input plotted against output gives a straight line. In real systems, however, as the frequency of a given signal is increased, a point is reached when the amplitude of the output signal is no longer equal to the amplitude of input signal. This phenomenon defines the frequency response of the system. A system with a frequency response range that is flat to 20Hz is necessary for accurate pressure reproduction.

Damping:

Any means of dissipating the energy of the natural frequency of the system such as friction is called damping. "Optimal damping" dissipates the energy gradually, maintaining the frequency response curve flat. It helps to prevent overshoot artifacts.

In practice micro mano-meter tipped catheters (Millar) and standard fluid filled catheters are used to measure pressure. Micro mano-meter catheters have a semiconductor strain gauge built into the catheter tip with frequency characteristics that will enable precise measurements of pressure waveforms.

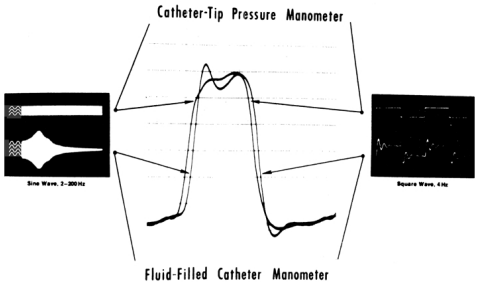


Figure 7.4 Left ventricular pressure **(center)** measured with a fluid-filled standard catheter and micromanometer (catheter-tip pressure manometer) in a patient undergoing cardiac catheterization. **Left and right.** In vitro frequency response for micromanometer (*upper*) and fluid-filled (*lower*) systems. The left panel recordings were obtained by continuously increasing the input frequency of a sine wave pressure waveform from 2 to 200 Hz. The fluid-filled system resonated (natural frequency) at 37 Hz but was flat (±5%) only to 12 Hz. Therefore its useful range is only to approximately 12 Hz. The right panel shows the response of each system to a square wave pressure input signal. (From Nichols WW, Pepine CJ, Millar HD, et al. Percutaneous left ventricular catheterization with an ultraminiature catheter-tip pressure transducer. *Cardiovasc Res* 1978; 12:566, with permission.)

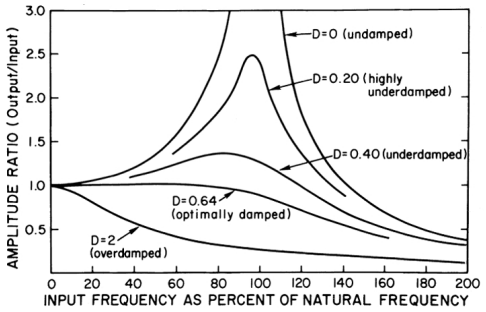


Figure 7.3 Frequency response curves of a pressure measurement system, illustrating the importance of optimal damping. The amplitude of an input signal tends to be augmented as the frequency of that signal approaches the natural frequency of the sensing membrane. Optimal damping dissipates the energy of the oscillating sensing membrane gradually and thereby maintains a nearly flat natural frequency curve (constant output/input ratio) as it approaches the region of the pressure measurement system's natural frequency (see text). D, damping coefficient.

Normal Pressure Waveforms

RA pressure waveforms:

Pre - a wave pressure: This is the pressure in diastole during the slow filling phase when RV and RA pressures become equal. The level at which this equalization occurs is determined by the compliance of the Right ventricle and the pericardium.

a wave: Irrespective of what the pre – a wave pressure is, the 'a wave' height is determined by the strength of the atrial contraction (atrial systole).

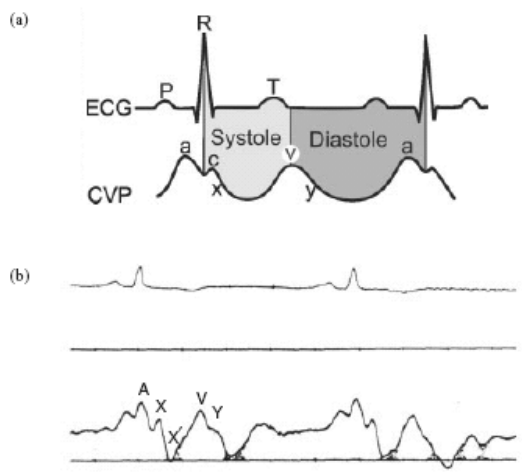
x descent: Atrial relaxation causes pressure fall termed x descent. It precedes the x' descent in timing.

x' descent: This is the major fall in atrial pressure in ventricular systole and is caused by right ventricular contraction pulling on the closed tricuspid valve and ring. A good and dominant x' descent indicates good RV systolic function.

'v' wave: The 'v' wave rise in the RA pressure in the absence of significant tricuspid regurgitation is result of venous filling of the right atrium in the later part of systole whether the actual pressure is normal or high. The normal right atrium, being a capacitance chamber, has good distensibility. Hence the 'V' wave pressure build up is often quite low, not exceeding 5mmHg.

'y' descent: Is the fall in the RA pressure in diastole immediately following the opening of the tricuspid valve (early rapid filling phase of diastole). The steepness of the 'y' descent will depend on the 'v' wave pressure head that is present at the time of the tricuspid valve opening (assuming there is no tricuspid stenosis).

The normal 'v' wave pressure being low, the 'y' descent is usually not very prominent.



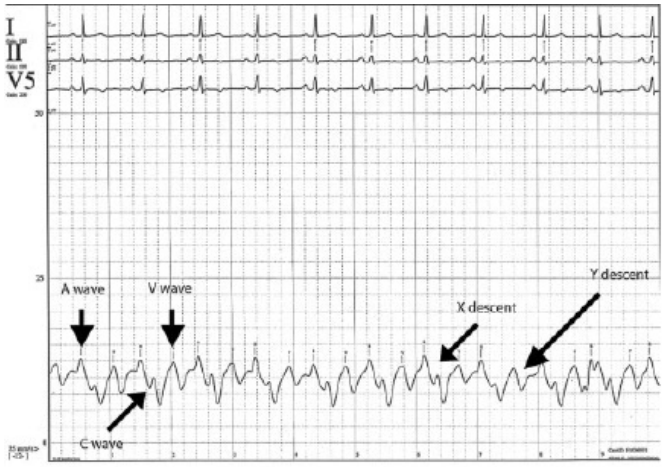


Figure 3.5 Right atrial pressure waveform.

Data usually reported include mean RA pressure, peak 'a' and 'v' wave pressures. The 'a' wave is usually the highest wave recorded in RA.

LA pressure waveform:

A normal 'LA' pressure wave form is similar to that of the RA. The 'v' wave however is the highest wave in the normal LA.

Important points:

The atrial waveform is composed of the positive atrial (A) and ventricular (V) waves and the X and Y descents.
The 'a' wave is prominent with abnormalities of LV, RV compliance.
The 'v' wave is prominent in Atrio-Ventricular valve regurgitation.
The X and Y descents are diminished in pericardial tamponade and accentuated in constriction and restrictive diseases.

RV pressure waveform:

Consists of a small rapid-filling wave, a slow filling wave, the 'a' wave co-incident with RA systole and then RV systolic pressure. The shape is somewhat "triangular".

Pressures usually reported include end-diastolic pressure which is measured just after the 'a' wave and the peak systolic pressure. In atrial fibrillation, when no 'a' wave is present, end diastolic pressure is measured 0.04 sec, after the peak of the 'R' wave on ECG.

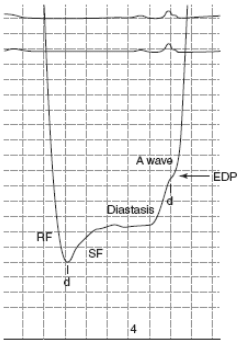


Figure 3.7 Ventricular diastole—the ventricular diastolic pressure waveform can be divided into three phases: the early or rapid filling (RF) phase occurs with the opening of the tricuspid valve. The slow filling (SF) phase follows and extends until the onset of right atrial systole (A wave). The nadir in pressure following the A wave is the right ventricular end-diastolic pressure (EDP).

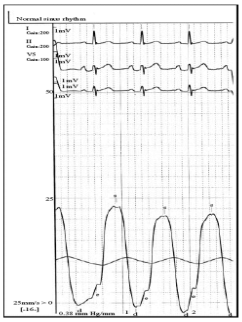


Figure 3.6 Ventricular pressure tracing.

LV pressure waveform:

Is less "triangular" with more rapid upstroke and descent than RV pressure pulse.

The early filling wave is followed by late filling, the 'a' wave of 'LA' contraction and later LV systole.

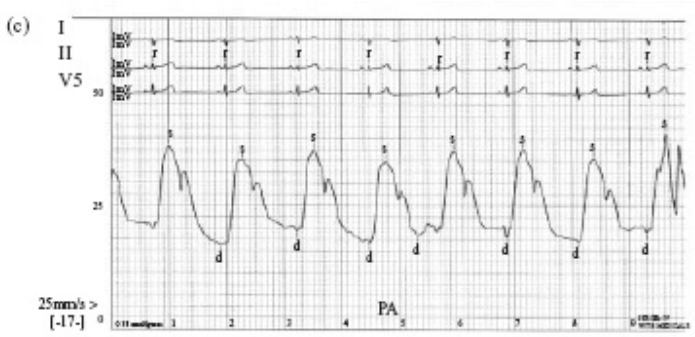
As 'LV' pressure rises, the mitral valve closes and isovolumic contraction continues until Aortic valve opens. Following LV ejection, as LV pressure falls below the aortic pressure, the aortic valve closes. The period between aortic valve closure and mitral valve opening is termed isovolumic relaxation. Values reported include end diastolic (measured at the end of the 'a' wave) and peak systolic LV pressure.

There is little meaning in measuring the initial diastolic pressure when fluid filled catheters are used, as this point is highly variable.

PA pressure waveforms:

Consists of a systolic pulse coincident with RV emptying. As RV ejection ends, the PA pressure begins to fall and the 'incisura' occurs as RV pressure beings to fall below that in the pulmonary artery and the pulmonary valve closes. As the PA pressure falls, the nadir of this phase is measured as PA end diastolic pressure.

The pressures usually reported are peak systolic, mean and PA diastolic pressures.



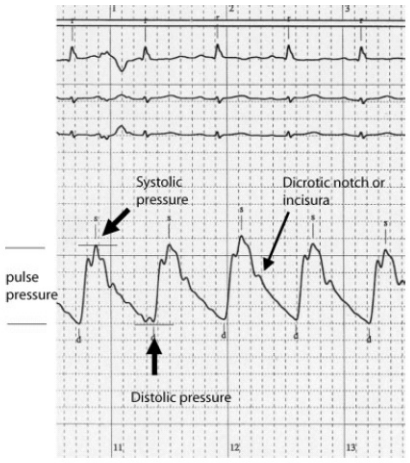
Aortic pressure waveforms:

Consists of a rapidly rising systolic pulse, an 'incisura' and a subsequent decay of pressure similar to PA.

Values reported include peak systolic, end diastolic and mean.

Measured central aortic pressure wave form is a conjugate of both forward (antegrade pressure wave) and reflected or retrograde pressure wave.

The components vary with disease, pharamacologic manipulations and LV function and these changes will be reflected in the waveforms.



1 Schematic of aortic pressure.

Factors that influence the magnitude of reflected waves:

Factors the augment pressure wave reflections:

Vasoconstriction
Heart failure

Hypertension
Aortic or ilio femoral obstruction
Valsalva – after release.

Factors that diminish pressure wave reflections:

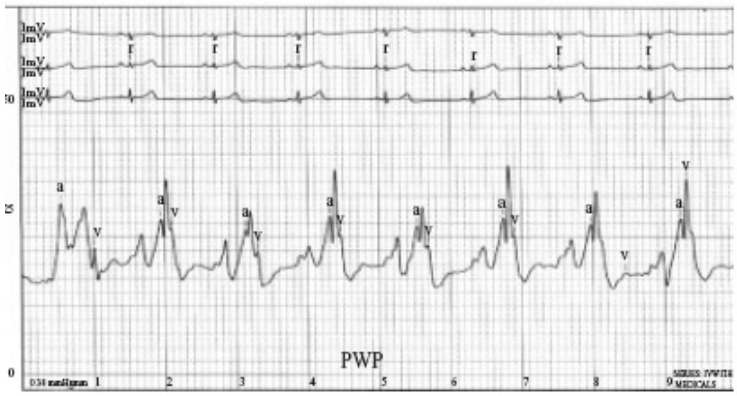
Vaso-dilatation
Hypovolemia
Hypotension
Valsalva – strain phase

Pulmonary capillary wedge pressure:

Obtained when an end-hole catheter is positioned in a pulmonary capillary with its open end hole facing pulmonary capillary bed with no connecting vessels conducting flow into or away from catheter tip and capillary bed.

It has a configuration similar to LA with added effects of delay and damping of pressure waves due to transmission through the pulmonary capillary and venous beds.

Values reported include mean PCWP, peak 'a' and 'v' wave pressures.



e tracings encountered during the insertion of a PA catheter.

Respiratory variation:

Breathing influences hemodynamic measurements. During spontaneous respiration, intra-thoracic pressure may reduce from 3 to 4 mmHg at end-expiration to 7 to 8mmHg during end-inspiration. This decreases the pressure in the LA, RA and the Aorta resulting in underestimation. Since end-expiration more closely resembles atmospheric pressure, intra-cardiac pressures should be recorded at end-expiration.

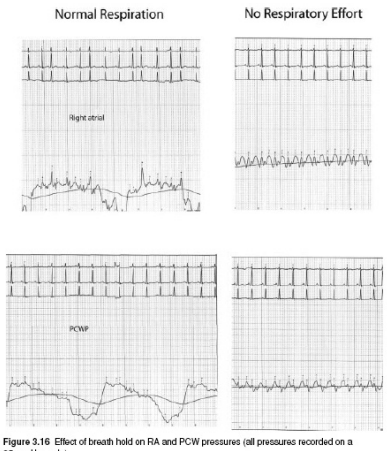
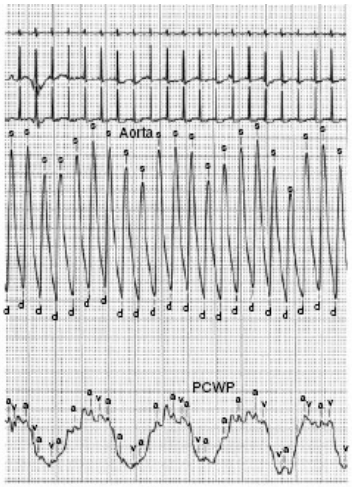


Figure 3.16 Effect of breath hold on RA and PCW pressures (all pressures recorded on a 25mm Hg scale).



Cardiac Output

Quantity of blood delivered to the systemic circulation per unit time is 'cardiac output' expressed in liters per minute. It is regulated by preload, heart rate any myocardial contractility. Normalization of cardiac output for body size is expressed as cardiac index = (liters / min / m2BSA).
(BSA is calculated by Dubois formula. BSA (m^2) = 0.007184 x weight (kgs) 0.725 x height (cm)).

Other variables like age, posture, body temperature and humidity must be considered in interpreting cardiac output measured in clinical setting.

Techniques for determination of cardiac output (CO) in cath lab:

1. Fick oxygen technique.
2. Indicator dilution technique.

Fick's O_2 technique
Fick's principle states that the total uptake or release of any substance by an organ is the product of blood flow to the organ and the arterio-venous concentration difference of the substance.

$$\textbf{CO} = \frac{O_2 \text{ Consumption}}{(\text{arterial} - \text{venous})O_2 \times \text{Hb concentration} \times 1.36 \times 10}$$

$$\textbf{CO} = \frac{130 \times \text{BSA}}{(\text{Sa } O_2 - \text{Sv } O_2) \times \text{Hb} \times 1.36 \times 10}$$

O_2 consumption is measured by polarographic and paramagnetic method. To avoid technical difficulties and expense, some labs assume O_2 consumption based on BSA as 125ml/m^2 BSA. AV O_2 difference is measured by reflectance oximetry method.

Error associated with CO determination by Fick's method is about 10%. Fick's method is most accurate in low output states, and in conditions with irregular heart rate (Atrial fibrillation, ventricular bigeminy)

Thermodilution method:

It involves injection of 10ml of cold saline into the right atrium and temperature changes are measured in the pulmonary artery. A transient decrease in PA temperature will be caused by the injectate and a thermodilution curve is generated by plotting the temperature of the pulmonary artery versus time. The curve has a smooth upslope and a more gentle decline. The area under the curve is inversely related to the cardiac output and is used by the computer to calculate to cardiac output.

$$\textbf{CO} = CC \times (T_b - T_i).$$

Where cc is the computation constant, T_b the blood temperature and T_i the injectate temperature.

Severe TR is a contra-indication to use of this method and the method is most accurate when used in high output states and less accurate in low output states. Under ideal conditions, error range associated with this method is 5 – 20%.

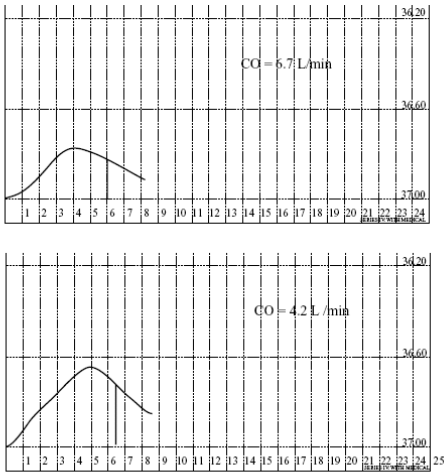


Figure 6.1 Thermodilution curves. Pulmonary artery temperature versus time is plotted in patients with thermodilution cardiac outputs (CO) of 6.7 L/min and 4.2 L/min.

Vascular Resistance

Resistance to blood flow is a function of viscosity, vessel radius and vessel length. The relationship is Poiseuille's law. When used in blood vessels it only provides an approximation of resistance since there are four assumptions namely viscosity is unchanging, tube is rigid and cylindrical, length exceeds diameter of the tube and flow is steady and non-pulsatile, which are violated when the equation is applied to blood flow in body.

In clinical practice, however, systemic and pulmonary vascular resistance are calculated based on Ohm's law relating blood flow, resistance and pressure.

$$\text{Systemic vascular resistance} = \frac{\text{Ao (mean)} - \text{RA (mean)}}{Q_s}$$

$$\text{Pulmonary vascular resistance} = \frac{\text{PA(mean)} - \text{LA(mean)}}{Q_p}$$

Where Ao (mean) is mean systemic arterial pressure
RA (mean) is mean RA pressure
PA (mean) is mean PA pressure
LA (mean) is mean LA pressure
Q_s systemic blood flow
Q_p pulmonary blood flow

Flows are in liters/min.
Pressures are in mmHg
Resistance is in "hybrid resistance units" (HRU) or "wood units" expressed in mmHg per liter per minute.
Wood units can be converted to metric units by conversion factor 80 expressed as dynes.sec.cm $^{-5}$

Vascular resistance is normalized for BSA giving a Resistance Index.

$$\text{SVRI} = \frac{\text{Ao(mean)} - \text{RA(mean)} \times 80}{\text{CI}}$$

Where CI is Cardiac Index.

Thus SVRI = SVR x BSA.

Normal Values For Vascular Resistance

Systemic vascular resistance	-	1170 $\pm$ 270 dynes – sec – cm $^{-5}$
Systemic vascular resistance index	-	2130 $\pm$ 450 dynes – sec – cm^{-5} m^2
Pulmonary vascular resistance	-	67 $\pm$ 30 dynes – sec – cm^{-5}
Pulmonary vascular resistance index	-	123 $\pm$ 54 dynes – sec – cm^{-5} m^2

Pressure tracings in diseased heart – MS, MR, AS, AR, TS, TR, PS, PR, HOCM, EMF, CP).

Dr. Sriram Rajagopal

Pressure tracing interpretation – some general principles:

1. Requires a good understanding of the pathophysiology of the underlying primary lesion as well as of compensatory mechanisms.

2. Technical aspects of pressure recording need close attention – e.g. scale on which pressures recorded, proper zero reference, presence of artifacts etc.

3. Information from the pressure tracing has to be interpreted in the context of other information such as clinical setting, flow data, time intervals, resistance etc.

4. The heart is an electromechanical device – remember always to correlate pressure information with electrical events as shown in the ECG trace.

5. Requires an ability to interpret changes in the tracing due to modifying factors such as heart rate, contractility, changes in preload or after load, effects of drugs or maneuvers etc.

Valve stenosis and regurgitation - General principles:

Stenotic lesions - Valve stenosis requires generation of a higher pressure proximal to the valve in order to maintain forward flow. This results in the generation of a pressure gradient across the valve. The magnitude of the pressure gradient across the valve is directly related to the square of the flow rate and inversely related to the square of the valve orifice area. Doubling of the flow rate at any given valve orifice area will result in quadrupling of the gradient.

The doubling of the flow rate can result from either an increase in volume of flow with the time duration for flow remaining constant or from the volume of flow remaining the same while the time duration for flow is decreased due to an increase in heart rate. Commonly both mechanisms, i.e. increase in flow and decrease in heart rate occur together, e.g. during exercise, resulting in a pressure gradient at rest which is hemodynamically tolerated but which stresses the system on exercise.

The obvious corollary of the above is that measurement of gradient alone is not an adequate estimate of severity of valve stenosis.
For purposes of calculations, mean gradients are usually used (i.e. the area between the relevant pressure traces in planimetered and divided by the length corresponding to the filling period. The number obtained is then expressed as pressure with reference to the scale being used to record the pressure). It is essential to average measurements of at least five beats in sinus rhythm and ten beats if the patient is in atrial fibrillation.

Regurgitant lesions – While stenotic lesions are gradual in onset and progression, severe regurgitation can occur as an acute event (e.g. in infective endocarditis or after balloon valvotomy). The key difference between these in terms of the hemodynamics and pressure tracings is that the

proximal chamber would not have time to adapt in the case of acute regurgitant lesions. Further, the volume load imposed by chronic regurgitation causes secondary changes in both the upstream and downstream chambers and this affects the pressure tracings in these chambers.

Specific lesions: Atrioventricular valves:

Mitral stenosis:

The presence of a gradient between the left atrium and the left ventricle throughout diastole is characteristic.

Pulmonary capillary wedge pressure is sometimes used as a surrogate for left atrial pressure, but the presence of phase lag and variable amounts of damping render this a less than ideal choice.

For a given degree of stenosis, doubling of flow rate will quadruple the gradient. The influence of heart rate is particularly prominent here, as the diastolic period is disproportionately shortened as heart rate increases. Hence, with a given orifice area, assuming cardiac output remains constant, increase in heart rate from say 60/min to 120/min (by use of atropine or supine leg exercise) will cause a fourfold rise in the gradient. This effect is also seen during spontaneous variation in cycle length in patients with atrial fibrillation.

Attention should also be paid to the relatively slow "y" descent in the left atrial pressure trace and to the influence of the stenosis on the early rapid filling phase of the left ventricular trace.

Pressure half time (which is used in estimating valve area) used to originally be calculated from hemodynamic pressure tracings, but is now usually estimated during echo studies.

In patients in sinus rhythm, the forceful atrial contraction during the "a" wave is not transmitted well to the left ventricle as a result of the stenosis and the gradient increases in late diastole.

Co-existing disease (e.g. hypertension or coronary artery disease or other valve lesions) may increase the LVEDP and this may in turn affect the interpretation of the tracing.

The development of pre-capillary pulmonary hypertension may add another element which is sometimes referred to as the "second stenosis".

Pulmonary venous pressures are raised. Pulmonary arterial hypertension often develops with time and can result in elevated right ventricular pressures and tricuspid regurgitation.

One should also be familiar with the change in hemodynamics following intervention (balloon mitral valvotomy).

Mitral regurgitation:

Acute severe mitral regurgitation (e.g after balloon valvotomy or following chordal rupture in MVPS or during infective endocarditis) usually causes very tall "v" waves in the left atrial pressure trace (due to direct transmission of the ventricular pressure in systole to a small non-compliant left atrium). There will also be severe pulmonary venous and arterial hypertension and the tall "v" waves will be seen in the PCWP.

In contrast, chronic mitral regurgitation (even when severe) may not be associated with such an impressive "v" wave as the left atrium has time to dilate and accommodate the regurgitant volume.

However, the chronic volume load on the left ventricle may result in elevated left ventricular end diastolic pressures and reduced dp/dt as the ventricle starts to fail. Pulmonary venous and pulmonary arterial hypertension develops over time with associated right heart failure.

Aortic systolic and pulse pressure may be slightly reduced.

Maneuvers which increase or decrease afterload will increase or decrease the degree of regurgitation and associated pressure changes.

Tricuspid valve disease:

Tricuspid stenosis: Similar principles as discussed under mitral stenosis apply here.

A dual catheter technique (two venous catheters) one each in the right atrium and ventricle is used. To cross check, tracings are often recorded and then the catheter connections to the respective transducers are "reversed" and tracings recorded again.

Since absolute levels of pressure are usually lower, meticulous attention to technique (zero referencing, scale factors etc) is essential.

Respiratory variations in pressures are often noted and must be documented.

Adequate filling pressures must be ensured, as a small gradient may be missed if the patient is dehydrated or over diuresed and right atrial pressure is therefore too low.

Also, many of these patients may have very low cardiac output, and hence the pressures must be correlated with the flow rates.

Tricuspid regurgitation:

As in mitral regurgitation, the hemodynamic hallmark of severe tricuspid regurgitation is a tall "v" wave in the right atrial pressure trace. Systemic venous hypertension is also seen.

Acute severe tricuspid regurgitation can lead to volume overload which may manifest as "restrictive physiology" in the presence of normal pericardial restraint (as the heart has not had time to adapt to the volume load and the normal pericardium acts as if it was adding an element of restriction).

Semilunar valve lesions:

Aortic stenosis:

The systolic gradient between the left ventricle and aorta (at the valvar level) is the most commonly used example to understand the hemodynamics of valve stenoses.

Elevated left ventricular systolic pressure and a characteristic small amplitude, slow rising aortic pressure wave contour (often associated with a notch on the upstroke, the anacrotic notch) with delayed rise to a peak (as compared to the left ventricular peak pressure) are hallmarks of aortic stenosis.
The left ventricular pressure trace also often shows an elevated LVEDP with a prominent late diastolic rise due to the prominent atrial contribution ("a" wave in the LVEDP).

Simultaneous measurement of ascending aortic and left ventricular pressures is preferable to using a "pullback" tracing from the left ventricle. This is because simultaneous pressures give details of pressures of both chambers in the same cardiac cycle (with identical rates, ejection periods and filling pressures

etc) whereas the presence of ectopics or a mild change in heart rate can affect pullback tracings and make superimposition of aortic and left ventricular tracings difficult to interpret.

During a planned pullback from the left ventricle to the aorta, the aortic pressure should be carefully recorded to note any significant rise (>10mm Hg) in aortic systolic pressure when the catheter is withdrawn from across the vavle (Carabello's sign – indicative of critical AS).

Meticulous attention to technique is essential for accurate assessment of aortic stenosis – including attention to catheter position, scale factors, simultaneous measurement of cardiac output, technical aspects of pressure recording etc.

The gradient used for calculations is usually the mean gradient. "Peak to peak" (highest pressure in the LV trace to highest pressure in the aortic trace) has no physiologic basis. Peak instantaneous (highest instantaneous difference between LV and aortic pressures) is more easily obtained generally with echo- Doppler studies.

Every effort should be made to compare LV pressures (taken from well within the left ventricular cavity, preferably near the apex) with ascending aortic pressures. Using the femoral arterial sheath side arm pressure as a surrogate for aortic pressure results in several artifacts due phase lag, peripheral augmentation of arterial pressure wave form etc and results in fallacies in evaluating the degree of stenosis.

The gradient can be interpreted only with reference to flow across the valve and, as mentioned earlier, flow rates, heart rate etc have an important bearing on the interpretation of pressure data.

"Low-flow low-gradient" aortic stenosis presents a challenge and the use of measures of contractile reserve to differentiate true severe aortic stenosis with a low gradient due to poor left ventricular contractility from a low gradient associated with mild stenosis and depressed left ventricular function is an area of active research.

Aortic regurgitation:

The hemodynamics of acute and chronic aortic regurgitation are significantly different.

In chronic aortic regurgitation, typical findings include a large aortic pulse pressure, low aortic diastolic pressure and elevation of LVEDP. In later stages, left ventricular dysfunction and pulmonary hypertension may develop.

In acute aortic regurgitation, the sudden volume load imposed on the left ventricle results in marked elevation of the LVEDP (rising to the level of the aortic diastolic pressure). The aortic pulse pressure is not wide and the aortic diastolic pressure is not low. The elevated LVEDP exceeds left atrial pressure and results in premature closure of the mitral valve. The aortic diastolic and left ventricular pressure may be identical.

Pulmonary stenosis and regurgitation:

Similar considerations as in aortic stenosis and regurgitation generally apply. The right ventricular systolic pressure may be supra-systemic in severe pulmonary stenosis when the ventricular septum is intact. The elevated right ventricular filling pressure and RVEDP result in a prominent "a" wave in the right atrial pressure tracing.

Hypertrophic obstructive cardiomyopathy (HOCM) :

The classic finding is that of a dynamic pressure gradient between the left ventricular inflow or apex and the sub-aortic region of the left ventricle. The gradient is usually in the LVOT, but occasionally a mid-cavitary gradient may be observed.

It should be differentiated from fixed sub-aortic obstruction (e.g. from a sub-aortic membrane). The presence of a "spike and dome" aortic pressure contour is characteristic of HOCM.

The gradient must be recorded with either micromanometer tipped catheters or with an end-hole catheter. The use of catheters with multiple side-holes results will usually result in unsatisfactory tracings and the gradient may be missed or underestimated.

Catheter entrapment in the hypertrophied and hypercontractile ventricle must be avoided.

Brockenbrough's sign (decrease or failure of the aortic pulse pressure to increase in the beat following a ventricular ectopic) may be elicited.

Response of the gradient to maneuvers and drugs which affect loading conditions or contractility must be documented. Specifically reduction in filling and increase in contractility will increase the gradient while increasing the afterload may diminish it.

Since maximum ejection occurs in early systole there will be a "spike" followed by a "dome" (due to transient reduction in ejection followed by resumption in late systole).

Constrictive pericarditis:

The pressure tracings reflect the pattern of ventricular filling. Most of ventricular filling occurs in early diastole, followed by a plateau as the constricting pericardium does not permit further filling. (Dip and plateau or "square root sign").

Ventricular filling pressures are elevated, with resultant elevation in atrial, pulmonary venous and pulmonary arterial diastolic pressures. The mean RA pressure, RV EDP, PA diastolic, PCWP mean, LA mean and LV EDP are all within 5mmHg of each other.

Pulmonary arterial hypertension is usually not marked (PA systolic < 50mmHg), in contrast to restrictive cardiomyopathy.

The RV EDP is usually greater than 1/3 of RV systolic pressure.

Ventricular interdependence, i.e., reciprocal change in right and left ventricular systolic pressures as a result of respiratory influences, can be demonstrated.

CARDIAC OUTPUT

Dr. Thomas George

Introduction

Cardiac output is the volume of blood ejected by the heart per minute. At rest the cardiac output for a healthy adult is 5 L/min and during exercise it rises upto 35 L/mt. Conversely during heart failure cardiac output falls. **Cardiac index** is the cardiac output expressed in body surface area and is between 2.7 – 4.3 l/min/m^2 of BSA in healthy adults. **Stroke volume**(SV) is the amount of blood ejected by the heart per contraction. The normal stroke volume for both ventricles are same and is 35 – 55ml/m^2 of BSA. The stroke volume is the difference between the end diastolic volume (EDV) (maximum filling of ventricle in diastole) and the end systolic volume (ESV) (volume of blood in the heart at the end of ejection).

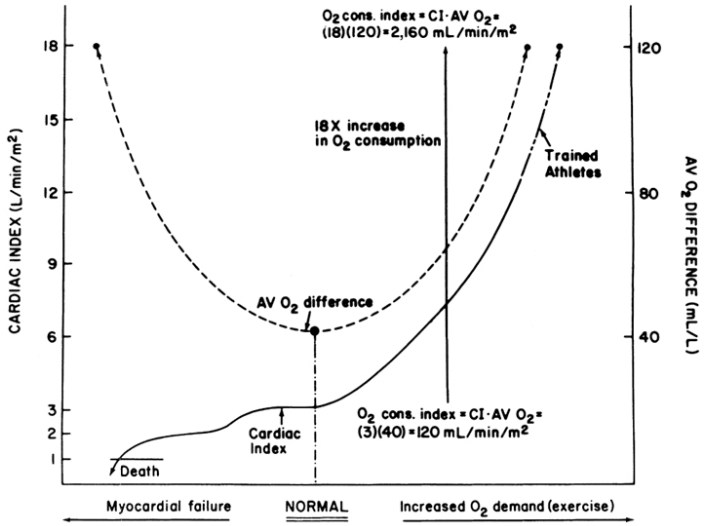


Figure 8.1 Relationship between arteriovenous oxygen (AV O_2) difference (*broken line*) and cardiac index (*solid curve*) in normal subjects at rest **(center)** and during exercise **(right)**, and in the patient with progressively worsening myocardial failure **(left)**. (See text for discussion.)

Factors influencing cardiac output:

1. Body surface area (BSA) – cardiac output is a function of the body's oxygen consumption or basal metabolic rate. This correlates best with the BSA.
2. Age – normal cardiac output varies with age, steadily falling from 4.5 l/min/m^2 at 7 years to 2.5 l/min/m^2 at 70 years. This is expected as the BMR falls with age.
3. Posture – the cardiac output falls by 10% when patient rises from lying to sitting position and falls by 20% when patient stands up from a recumbent position.
4. Body temperature
5. External environmental temperature
6. Anxiety and humidity also influence the cardiac output.

Measurement of cardiac output at rest and in some cases during exercise is needed for the assessment of cardiovascular function, calculating vascular resistances, regurgitant fractions and valve orifice areas.

Cardiac output can be measured by (i) Invasive and (ii) Non-invasive methods

Invasive methods:

1. Ficks technique
2. Indicator dilution method
3. Angiographic technique

Non invasive methods:

1.Doppler principle
2.Aortovelography
3.Trans-thoracic impedance

There is no technique which provide flawless results for cardiac output estimation. It is for the clinicians to decide what level of accuracy is required in a given situation.

Ficks principle

"*The total uptake or release of a substance by an organ is the product of blood flow to the organ and of the arteriovenous concentration of the substance*" [1].

Ficks principle states that flow through a vascular bed is equal to the amount of substance (A) produced or used by that bed divided by the outflow concentration (C-out) of that substance minus the input concentration (c-in) of the substance.

$$\text{Flow} = \frac{A}{C_{out} - C_{in}}$$

As pulmonary blood flow is similar to systemic blood flow, it can be used to measure cardiac output, as at a given time the cardiac output from both the ventricles are similar. Oxygen (O_2) is utilized by the systemic vascular bed and resupplied by the lungs by an equal volume. From the Ficks equation the substance measured (A) is taken as oxygen. Both the outflow (C_{out}) and inflow (C_{in}) concentration of O_2 across the pulmonary vascular bed is measured. By measuring the amount of oxygen extracted from inspired air by lungs and the AVO_2 difference across the lungs the pulmonary blood flow can be calculated.

Further extrapolating the Ficks principle, the cardiac output is equal to the minute consumption of oxygen (A) divided by the difference in O_2 content of pulmonary outflow from the pulmonary inflow.

$$\text{Cardiac output in L/mnt} = \frac{O_2 \text{ consumption ml/min}}{AVO_2 \text{ difference across lungs ml/L}}$$

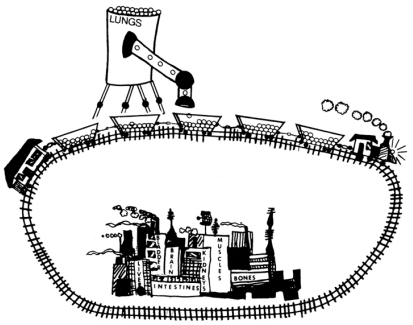


Figure 8.3 Illustration of Fick's principle. A train, representing the circulation, passes by a hopper (the lungs) that delivers marbles (oxygen) to the train's boxcars at a rate of 20 marbles per minute. Because the boxcars each contain 16 marbles before and 20 marbles after passing under the hopper, each boxcar is picking up 4 marbles and must be taking only 0.20 minute to pass under the hopper, since it would pick up 20 marbles in each full minute under the hopper. If each boxcar takes only 0.20 minute to pass by the hopper, the train is moving at a speed sufficient to deliver 5 boxcars per minute to any point down the line. This could have been calculated as

Train's speed (boxcars/minute) = marble delivery rate (marbles/minute)/"AV" marble difference (marble/minute)

= (20 marbles/minute)/(4 marbles/boxcar)

= 5 boxcars/minute

If one boxcar is 1 L of blood and each marble is 10 mL of oxygen, then we have an arteriovenous oxygen difference of 40 mL/L, an O_2 consumption of 200 mL/minute, and a cardiac output of 5 L/minute. (Illustration kindly provided by Jennifer Grossman, age 11.)

Measurement of oxygen consumption:

The normal oxygen consumption is 110 – 150 ml/min/m^2 of BSA. Normally the oxygen consumption is higher for men than women

and increases with hyperthyroidism, exercise and hyperthermia. There are two methods of calculating oxygen consumption [2].

1. Polarographic method
2. Paramagnetic method

In the former a metabolic rate meter is used. It basically contains a polarographic oxygen sensor cell, a hood which covers the patents head and a blower. The blower maintains an unidirectional flow of air from the room through the hood and via a connecting hose to the polarographic oxygen sensory cell. The patient's oxygen consumption is given by the formula.

$VO_2 = (F_{RO2}.V_R) - (F_{MO2}.V_M)$

Where F_{RO2} & F_{MO2} are fractional contents of oxygen in room air and in air flowing past the polarographic cell. V_R is the rate at which room air enters the blood in ml/min. This is determined by the blowers discharge rate (V_M in ml/mt)as well as the patents ventillatory rate (Vi – insp & V_E – expiratory)

$V_M = V_R\text{-}Vi + V_E$

(Or) $V_R = V_M + Vi - V_E$

Substituting, $VO_2 = V_M(F_RO_2\text{-}F_MO_2)+F_RO_2(Vi\text{-}V_E)$

The fractional content of O_2 in room air is 0.209

Substituting $VO_2 = V_M\ (0.209 - F_MO_2) + 0.209(Vi\text{-}V_E)$

In practice F_MO_2 is set at 0.199 and in a steady state $Vi = V_E$

So, $VO_2 = 0.01\ V_M$

The paramagnetic method utilizes a sensor medics device. A constant flow rate V_M leaves the hood and enters the metabolic monitor unit. The paramagnetic sensor measures oxygen and an infra red sensor measures CO_2.

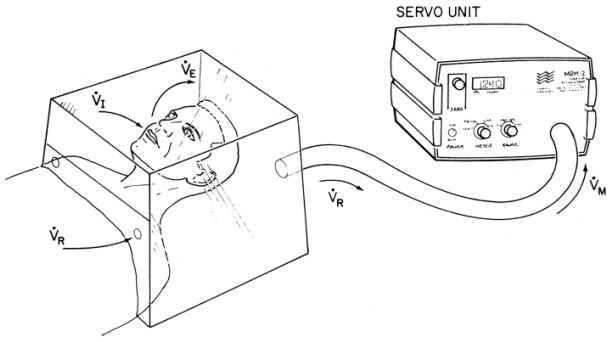


Figure 8.4 Measurement of O_2 consumption by a polarographic cell technique using the Waters Instruments metabolic rate meter (MRM). A transparent hood fits snugly over the patient's head, resting on a pillow. Air enters the hood through holes in a plastic sheet at a flow rate of V_R. The patient's inspiratory (V_I) and expiratory (V_E) flow rates subtract and add to V_R to yield V_M, the flow rate leaving the hood and entering the servo unit. A blower motor in the servo unit adjusts V_M to keep the O_2 sensed by a polarographic cell constant. (See text for details.)

Measurement of AV O2 difference

For calculating the AV O_2 difference, the O_2 content of pulmonary arterial (mixed venous) and systemic arterial blood is to be measured. First the oxygen saturation of the blood and its hemoglobin (Hb) concentration is taken. The oxygen saturation of pulmonary outflow is to be measured at pulmonary veins which will be equal to the systemic arterial saturation and can be taken from the systemic arteries. The oxygen saturation at the pulmonary inflow is measured by sampling blood from the pulmonary artery.

O_2 content = Hb (in gm/dl) x 1.36 (ml O_2 per gram Hb) x 10 x saturation

1.36 is the maximum oxygen carrying capacity of 1 gram of Hb. The normal A-V O_2 difference is 3.0 – 4.5 ml O_2/dl of blood.

Eg: - a patient with a Hb of 14.0 gm/dl has a systemic arterial O_2 saturation of 95% (corresponds to pulmonary venous saturation) and a pulmonary arterial saturation of 65%. His O_2 consumption is 250 ml/min. 10 is taken as conversion factor (dl to L)

CO = 250/14 x 1.36 x 10 x 0.95 – 14 x 1.36 x 10 x 0.65

Step 1. Theoretic oxygen-carrying capacity:
Hemoglobin (gm/dL) × 1.36 (mL of O_2/gm of Hb) × 10 = _____ mL O_2/L blood

Step 2. Saturation of arterial (BA, FA, Ao) blood = _____ %

Step 3. Oxygen content of arterial blood:
Theoretic capacity (step 1) × % saturation (step 2) = _____ mL/L

Step 4. Saturation of mixed venous (PA) blood = _____ %

Step 5. Oxygen content of mixed venous blood:
Theoretic capacity (step 1) × % saturation (step 4) = _____ mL/L

Step 6. AV O_2 difference:
Arterial O_2 content (step 3) − venous O_2 content (step 5) = _____ mL/L

Figure 8.5 Calculation of oxygen content and AV oxygen difference when using the reflectance oximetry method.

Drawbacks of Ficks method(3)

The most common source of error is the incomplete collection of expired air. This underestimates O_2 consumption leading to a low value for cardiac output.

Incorrect timing of collection of expired air – again leads to an inaccurate estimation of oxygen consumption.

If Douglas bag method is used then analysis is to be done immediately or air may diffuse in and out of the bag.

When the polarographic system is used the gas sensors have to be calibrated regularly or the values for fractional content of oxygen and carbon dioxide will be inaccurate.

O_2 saturation values from spectrometry may be modified by presence of certain substances such as indocyanine green.

Mixed venous blood sample should be from pulmonary artery and not be from pulmonary capillary wedge position. Also there should not be any left to right shunt which may alter the value. If sample is taken from RA ,IVC or coronary sinus it will lead to false values during calculation.

Narrow AV O2 differences are more prone to error than wide AV O2 differences. Hence the Fick method is more accurate in low cardiac output states where AV O2 difference is narrow.

Indicator dilution technique

This technique was first used by Stewart for measuring cardiac output [4]. There are 2 types of indicator dilution methods – the continuous infusion method and the single injection method

This technique works on the same principle as Ficks method – where oxygen is the indicator, site of injection is the lungs and the injection procedure is a continues infusion. This works on the principle that the volume of fluid within a container can be determined if one adds a known quantity of indicator to the fluid and then measures the concentration of the indicator after it has completely mixed with the fluid [6]. Once the indicator is injected into the circulation it is allowed to mix completely in the blood. A time concentration curve is then plotted and the computer calculates the area. The area under the photometric curve after a known amount of dye is injected determines the cardiac output by Stewart's historic method. Extrapolation of the dye decay curve yields measurements of blood volume. This is a limitation of this method in comparison with non-first pass indicators.

Dye dilution technique:

A known amount of dye is injected into the pulmonary artery, and its concentration is measured peripherally. Indocyanine green is suitable due to its low toxicity and short half-life. A curve is achieved, which is replotted semi-logarithmically to correct for recirculation of the dye. CO is calculated from the injected dose, the area under the curve (AUC) and its duration. (Short duration indicates high CO). Lithium has also been used as an alternative to indocyanine green. It is injected via a central venous catheter and measured by a lithium-sensitive electrode incorporated into the radial arterial cannula.

Thermodilution technique:

It was first demonstrated by Fegler in 1954.

Principle of measurement (5)

It uses a special thermistor tipped swan gantz catheter that is inserted from a peripheral vein into the PA. A cold saline of known temperature and volume is injected into RA from the proximal port of the catheter. This saline mixes with blood as it passes through ventricle into the PA and this cools the blood. The temperature of the blood is now measured by the thermisitor at the catheter tip which lies in the pulmonary artery. The change in temperature of blood in the PA causes a change in the thermistor resistance which allows calculation of the area under the thermodilution curve.

The thermodilution curve peaks rapidly and then follows an exponential decay, until there is recirculation or delayed cooling from the residual indicator in the PA catheter. To avoid this last portion of recirculation, a cut off point is determined in the thermodilution curve before recirculation occurs. The computer now analyses the data and quantifies the change in blood temperature as it flows over the thermisitor surface. The cardiac output is then calculated. Injection is repeated and an average of values is taken. As cardiac output changes with respiration it is important to inject saline at a constant point of the respiratory cycle – normally end expiration. Time activity curve shows only a primary peak unlike indocyanine green which shows a recirculation peak. Advantages of the thermodilution technique are it is not expensive, easy to perform and does not require arterial sampling or blood withdrawal. There is no recirculation so a computer analysis of primary curve is simple.

Drawbacks (6)

1. **Rewarming of injectate** – temperature of iced injectate in a 10ml syringe held in a warm hand will increase 1°C for every 13 sec -this could result in overestimation of CO by 2.86% for each °C of rewarming.

2. **Timing of injection and respiration** – Animal experiments show that baseline temperature in PA at expiration increases during spontaneous breathing and decreases during IPPV. CO will be underestimated during spontaneous breathing and over estimated during IPPV, in the face of large variations in PA temperature when end expiration is used to time indicator injection. RV output may vary due to cyclical alterations in venous return and RV after load during IPPV or kussmaul breathing.

3. **Speed and mode of injection** – when the injectate volume is 5 – 10 ml, an injection time of upto 4 seconds may be acceptable. If injection time is too long then thermodilution curves are of poor quality and CO values become unreliable.

4. **IV fluid administration** – Rapid peripheral IV infusion of 90 – 220ml fluid at room temperature prior to measurement may underestimate CO. Infusion of fluid should be maintained at a constant rate or discontinued for atleast 30 sec before CO measurement.

5. **Low flow** – in low CO states the appearance of the thermal curve is delayed and its decay is prolonged. Recirculation of indicator may occur before computer analysis is completed. CO is usually overestimated due to excessive heat exchange due to slow passage of injectate.

6. **Intra & extra cardiac shunts** – in presence of a right to left shunt some of the indicator bypasses the thermistor to reach the left side of the heart and this results in overestimation of cardiac output.

7. **Valve regurgitation** – CO values estimated by thermodilution is unreliable in patients with tricuspid regurgitation. Similarly also in aortic and mitral regurgitation

Angiographic Technique:

The Stroke volume corresponds to the volume of blood ejected with each heart beat. This can be determined by left ventriculography. Stroke volume multiplied by heart rate gives the cardiac output. The Angiographic method of estimating cardiac output is unreliable in patients with significant wall motion abnormalities, deformities in ventricular shape and valve regurgitation. When there is valvar

regurgitation like MR or AR the regurgitant volume does not enter the systemic circuit. The calculated Angiographic cardiac output will exceed the forward output.

Non-invasive methods:

Doppler – Transesophageal Echo (TEE) can measure the length of blood in the ascending aorta in unit time. This is multiplied by the cross sectional area of aorta to give stroke volume.
The time velocity integral which is the integral for instantaneous blood flow velocities during one cardiac cycle is obtained for the blood flow in the left ventricular outflow tract. This is multiplied by the cross sectional area and heart rate to give cardiac output.

Main disadvantage is 1) skilled operator is required 2) probe cannot be kept fixed to give continuous cardiac output recordings

Aortovelography – use of a Doppler ultrasound probe in the suprasternal notch to measure blood velocity and acceleration in the ascending aorta is generally an inaccurate method of measuring cardiac output.

Disadvantages

Doppler estimate of cardiac output is based on the square of the measured radius of aorta. Any error in measurement will be multiplied and affect the final value.

Transthoracic impedance – impedance can be measured across externally applied electrodes. Impedance changes with cardiac cycle. The rate of change of impedance is a reflection of the cardiac output. Since cyclical changes in transthoracic impedance of about 0.5% is noted during contraction of heart, it gives a low signal to noise ratio.

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VASCULAR RESISTANCE

Dr. S.R. Ramkumar

Estimation of Vascular Resistance:

- Calculation of vascular resistance is usually applied to both pulmonary and systemic circulations.
- Not only the arteriolar but multiple factors determine vascular resistance.
- In 1842, French physician, Jean Leonard Marie Poiseuille derived an equation describing flow of fluids through cylindrical tubes
  Poiseuille Law is showed as:

$$Q = \pi(Pi\text{-}Po)\ r4 / 8nl$$

Where,
Q- Volume of flow.
Pi-Po- Inflow pressure – Outflow pressure.
r- Radius of the tube.
l- Length of the tube.
n- Viscosity of the fluid.

- Poiseuille law is applicable in circumstances of steady state laminar flow of homogeneous fluid through a rigid tube.
- Hydraulic resistance 'R' is defined by analogy to Ohms Law as the ratio of mean pressure drop Delta P to flow Q, across the vascular circuit

$$R = \frac{Pi\text{-}Po}{Q} = \frac{8nl}{\pi r4}$$

- Resistance to flow depends on the dimension of the tube and viscosity of the fluid.
- Resistance if very sensitive to changes in the radius of the tube varying inversely with its fourth power.
- In clinical and physiological settings vascular resistance derived from haemodynamic measurements made during cardiac catheterization has acquired empiric patho physiologic meaning and is often an important factor in clinical decision making.

Formulae for calculation

To calculate vascular resistance the pressure differential across the pulmonary and systemic circuits and the respective blood flow through them are required.

Systemic vascular resistance is = $\frac{AO - RA}{QS}$

Pulmonary Vascular Resistance = $\frac{PA - LA}{QP}$

Total Pulmonary Resistance **= PA/QP**

Where,
PA – Mean Pulmonary Arterial Pressure
AO - Mean Systemic Arterial Pressure
RA - Mean Right Atrial Pressure
LA – Mean Left Atrial Pressure
QS – Systemic Blood Flow
QP – Pulmonary Blood Flow

- The flow and volume flow are expressed in liters per minute and pressure is in millimeters of mercury.
- The equation yields resistance in units called R Unit or wood unit as they are first introduced by Dr. Paul Wood.
- They may be converted to metric resistance units expressed in Dynes – Seconds-Cm-5 by use of conversion factor 80.
- When vascular resistance calculated in normalized body surface area it is expressed as resistance index. Thus,

PVRI = PVR multiplied by body surface area.
SVRI = SVR multiplied by body surface area.

Normal Values for Vascular Resistance in Adults Is Given In the Table

SVR	**1170+/- 270 dynes –sec-cm-5**
SVRI	**2130+/-450 dynes –sec-cm-5**
PVR	**67+/- 30dynes –sec-cm-5**
PVRI	**123+/-dynes –sec-cm-5**

PVR: The pulmonary vascular system in subject to many mechanical, neural and biochemical influences.

The factors are as follows:

Mechanical:

- Change in pulmonary venous pressure
- Change in pulmonary Blood flow
- Change in pulmonary Blood volume
- Change in Alveolar pressure
- Change in Intrathoracic pressure
- Pericapillary edema
- Changes in size of pulmonary vascular bed.

Neural:

- Autonomic nervous system
- Intravascular chemoreceptor
- Intravascular neuroreceptors
- Changes in neuroregulation of ventilation.

Biochemical and hormonal:

- Changes in oxygen tension
- Acute hypercapnia
- Acute acidosis
- Catecholamine.

  - The reversibility of elevated PVR and pulmonary hypertension can be tested by oxygen inhalation, infusion of acetylcholine, tolazine and inhaled nitric oxide
  - The patients who show a significant drop in PVR and pulmonary area of pressure following administration of the agents are called as pulmonary vascular hyper reaction who responded to increased pulmonary blood flow with pulmonary vasoconstriction.
  - When permanent obilerative changes in pulmonary vasculature occur there may not be any reversible changes upon testing with these agents.
  - The decision as to whether patient with congenital heart disease could benefit from corrective surgery hinges on calculated PVR.
  - The ratio between PVR and SVR are used as criteria for assessing operability
  - Normal PVR/SVR ratio is less then equal to 0.25
  - 0.25 -0.50 indicates moderate pulmonary vascular disease, more than 0.75 severe pulmonary vascular disease.
  - Wood has showed that in patients with pulmonary blood flow less then 1.75 times systemic flow or total PVR greater then 12 wood units, surgical repair of defects should not be done.
  - Surgical repair should be done when the net shunt in left to right and PVR is less then SVR with ratio of less than 0.50.

DETERMINATION OF VALVULAR STENOSIS

Dr. Anand Gnanaraj

Estimation of valve area in patients with valvular heart disease remains an important decision making tool. Gradients across the valve generally correlate with valve area and clinical severity. Various determinants play a role during the measurements of gradients across valves and during the calculation of valve area.

After the advent (extensive use) of echocardiography in patients with valvular heart disease, the number of occasions that compel the clinician to measure valve area in the cath lab has diminished. It is also known that peak instantaneous gradients are more relevant clinically than peak-to-peak gradients. Despite these challenges, direct hemodynamic measurements of valve area, especially in stenotic valves, remains the 'gold standard' for valve area calculations.

Basic principles:

The fluid dynamics that exist in the heart are complex with multiple factors playing their roles. To complicate things further, these factors play variable roles in different situations and cardiac status.

The important factors that alter (determine) the valve area are:

1. **Velocity of flow:** Velocity of flow varies with heart rate, adrenergic drive and peripheral vasodilatory state. These inturn are dependant on the hemoglobin content, the O2 saturation, myocardial perfusion, adrenergic stimuli, drugs and other mediators.
2. **Flow duration:** since the cardiac flow is pulsatile, the flow occurs only during a particular time in the cardiac cycle. This duration is dependant on the heart rate and the contractile force. Systolic ejection time being more or less constant, the majority of time compensation takes place at the cost of diastolic filling time.
3. **Cardiac output:** the effective cardiac output determines the amount of blood that would pass through an orifice. Despite this simplistic concept, the actual measurement of cardiac output is complicated. The lower the cardiac output, the more inaccurate the measurements become. Once the cardiac output becomes inaccurate, other calculations based on cardiac output also become inaccurate. This warrants meticulous care in calculating cardiac output and awareness to all the potential pitfalls.
4. **Instrumentation:** the advances in hardware in the form of better catheters, pressure measuring devices and electronics have made values more reliable. In spite of such advancements, meticulous set up of hardware and 'zeroing' of pressure transducers are crucial. No amount of technological innovation can overcome this procedure.

Evaluation of the formula

The basic parameters (information) required to calculate any vale area are:

1. Duration of flow

The seconds per minute during which there was flow across the valve and is calculated by

Average systolic ejction / diastolic filling period

$$= \frac{\text{average length (mm)}}{\text{paper speed (mm/sec)}}$$

2. Flow across the orifice

This is calculated as cardiac output and expressed as cm^3/min. Cardiac output is calculated using Fick principle or indicator dilution method.

Fick principle

$$CO = \frac{O2 \text{ consumption (ml/min)}}{A\text{-}VO2 \text{ difference (vol\%)} \times 10}$$

Indicator dilution:

$$CO = \frac{\text{amount of indicator injected (mg)} \times 60 \text{ sec/min}}{\text{mean indicator conc. (mg/ml)} \times \text{curve duration}}$$

3. Valve constants:

Constants are added to compensate for variables in area calculations that cannot be controlled. An empirical constant was derived by comparing the actual mitral valve area and the calculated valve area. This was initially thought to be 0.7 and later has been revised to 0.85.

Constants for other valves have not been calculated. These valves are derived (rather assumed) to be 1.0. A value of 44.3 is obtained by calculating the effect of gravity (980 cm/sec^2) and converting cm of H_2O to mm of Hg. Hence the constant is (1.0 x 44.3) = 44.3

Valve constant for mitral valve = 37.7

Valve constant for aortic valve = 43.3

4. Mean gradient:

Mean gradient is the area enclosed between the two curves of the two chambers that are measured. This is automatically calculated by most cath lab machines and the value is reliable.

With the above variables, the valve area calculation is derived by

$$\text{Valve area} = \frac{CO / (HR \times \text{avg.diastolic filling / systolic ejection period})}{\text{Valve constant} \times \sqrt{\text{mean gradient}}} \ cm^2$$

Valve area index = valve area / BSA (cm^2/m^2)

Specific issues with calculations:

Aortic valve:

The flow across the aortic valve determines the gradients and the calculated valve area. This flow is partly determined by the heart rate contrary to mitral hemodynamics, the aortic flow and gradients increase with slower heart rate. Since slower rate increase the cardiac stroke volume per beat, the flow is faster and gradients higher. A valve area of 0.7cm^2 may give a gradient of 33mmHg at 5 L/min. Doubling the cardiac output to 10L /min will increase the gradient to 132 mmHg (equivalent to 250mmHg LV systolic pressure). Significance of valve area also depends on the size of the patient. A larger valve area may be critical for large patients.

Common pitfalls

1) PCWP is not equal to LA pressure

   a) Swan – Ganz catheter: when Swan – Ganz catheter is used the PCWP is 3.3 to 3.5 mmHg more than LA pressure.

   b) Woven Dacron catheter: Using a stiff woven Dacron catheter minimizes the different to 1.7 ± 0.6 mmHg.

   The exception to this are patients with veno occlusive disease and cor-triatriatum.

2) Alignment mismatch:

The time delay between LA / LV pressure curves and PCWP pressure curves can be upto 50 – 70 msec. Awareness of this is usually enough to avoid errors. Using a tracing paper, the pressure tracings can be drawn manually and reapplied over the LV / LA pressure tracing. This is a simple solution and is very effective.

3) Calibration Errors:

A careful zero reference should be made. To cross check the accuracy, the left and right heart catheters can be switched and checked.

4) Early diastasis:

Mitral flow may continue even after diastasis (equal LV / LA pressures). So the time period for calculating the area should include the non isovolumic diastolic phase also.

5) Pull back hemodynamics:

A significant aortic stenosis shows more gradient with the catheter across the valve (e.g. 0.6 cm^2 valve area with a 7F catheter).

If the peripheral systolic pressure is augmented by 5mmHg during pull back, it indicates significant aortic stenosis. This sign is positive in >80% of patients with valve area of 0.5cm^2 or less.

6) Low cardiac output

Valve area calculation made using Gorlins formula are flow dependant. Calculated area increases with increased cardiac output and decreases with decreased cardiac output.

This can be overcome by using dobutamine or nitroprusside in the lab. Increased cardiac contractility and vasodialtation decreases the valve area if it is not significant but does not change or increase in significant aortic stenosis.

It is safer to use dobutamine in patients with severe AS as long as the coronaries are normal. Nitroprusside, despite the risk of hypotension, is preferred over dobutamine in the presence of significant CAD.

Simple formula:

Hakki et al proposed a simplified formula for calculating stenotic valve area. This formula may not be accurate during tachycardia.

$$\text{Valve area} = \frac{\text{cardiac output (L/min)}}{\sqrt{\text{pressure gradient}}}$$

CARDIAC CATHETERISATION AND ASSESSMENT OF OPERABILITY IN LEFT TO RIGHT SHUNTS

Sreeja Pavithran , R. Suresh Kumar

Detection, localization and quantification of intra cardiac shunts form an integral part of the hemodynamic evaluation of patients with congenital heart disease.

With advances in 2D and colour Doppler echocardiography, cardiac catheterization for left to right shunts is today done to quantify the shunt and to assess associated pulmonary hypertension.

Catheterization study

- A right heart catheterization is performed, measuring O2 saturation in the SVC, IVC and recording pressures and saturations in the right atrium (RA), right ventricle (RV), main, right and left pulmonary arteries (PA) and pulmonary capillary wedge positions.
- An arterial catheter is used to record pressure, oxygen saturation and blood gas values in the descending and then ascending aorta.
- These data are used in computation of pulmonary and systemic flow and resistance values. If the pulmonary artery pressures are very high, the reversibility of pulmonary hypertension is assessed by administering 100% oxygen by mask to the patient for 10 minutes. The oximetry run and pressure measurements are then repeated. Other drugs used for vasodilatory testing are nitric oxide, adenosine, calcium channel blockers, tolazoline and prostacyclin.
- Angiography may be done to assess the number and location of the defects if required.

Pressure Data

- Pressure measurements are the most important hemodynamic data obtained during cardiac catheterization.
- Before the catheterization is started, the pressure transducer is calibrated at the level of the heart so that it reads zero at atmospheric pressure.

NORMAL INTRACARDIAC PRESSURES IN CHILDREN

	Average	Range
Right atrial, mean	1 – 4	3 – 8
Right ventricular, systolic	24 – 29	13 – 42
Right ventricular, end-diastolic	3 – 5	0 – 10
Pulmonary arterial, systolic	20 – 22	11 – 36
Pulmonary arterial, diastolic	7 – 10	3 – 21
Pulmonary arterial, mean	11 – 12	6 – 22
Pulmonary arterial wedge or left atrial, mean	6 – 9	2 – 14
Left ventricular, systolic	100 – 106	72 – 133
Left ventricular, diastolic	7 – 10	3 – 14
Systemic arterial, systolic	108	77 – 150
Systemic arterial, diastolic	61 – 64	50 – 89
Systemic arterial, mean	70 – 81	67 – 105

If there is a large VSD or PDA, the PA systolic pressure equals the aortic systolic pressure, but PA diastolic pressure depends on pulmonary vascular resistance.

Blood flow measurement: The cardiac output and shunts

Cardiac output is measured by application of Fick's principle, which states that the total uptake or release of any substance by an organ is the product of blood flow to the organ and the arteriovenous concentration difference of the substance. This involves measurement of oxygen consumption and the arterio-venous oxygen difference across the lungs. Based on the Fick's principle,

$$\text{Cardiac output} = \frac{\text{oxygen consumption } (VO_2)}{\text{Arterio-venous oxygen difference}}$$

Definition of important terms:

- **Cardiac output** - quantity of blood delivered to the systemic circulation per unit time, expressed as (L/min)
- **Cardiac index (CI)** - cardiac input indexed to body surface area and is expressed as L/min/m^2. The normal cardiac index is 2.4 – 4.4 L/min/m^2
- **Oxygen consumption (VO_2)** - the amount of oxygen taken up by the tissues and is usually expressed in millilitres of oxygen consumed by the body each minute (ml/minute).

  The normal oxygen consumption for 0-5 years old is about 140-200 ml/min (per m^2) and for children 5-15 years old ranges from 110-160ml/min (per m^2). Oxygen consumption can be measured by using the polarographic method and the paramagnetic method. As this is a cumbersome process, for practical purposes, oxygen consumption is assumed based on the table of Lafarge and Miettinen (see Appendix).
- **Oxygen capacity** – maximal amount of oxygen that can be taken up by hemoglobin in blood. When the hemoglobin level in g/100ml is multiplied by 1.36, the oxygen capacity is derived, expressed as ml/dl, as each gram of hemoglobin taken up 1.36ml O_2.
- **Oxygen saturation** – a measure of the proportion of oxygen actually combined with hemoglobin to the total amount of oxygen that can be taken up by hemoglobin in a blood sample.
- **Oxygen content** – amount of oxygen present in any blood sample in question. It is expressed as ml/L of blood and refers to the total quantity both combined with Hb and dissolved in plasma. Oxygen content is usually determined from the measurement of hemoglobin concentration and oxygen saturation and addition of the amount of dissolved oxygen.

  The terms oxygen saturation and oxygen capacity concern only oxygen attached to hemoglobin and do not consider dissolved oxygen.
- **Arterio venous oxygen (AVO_2) difference** – the amount of oxygen extracted from the circulation as the blood flows through a vascular bed. Usually expressed in volumes percent (ml/100ml)

The relationship between oxygen saturation, oxygen content and oxygen capacity is:

$$O_2 \text{ saturation (\%)} = \frac{O_2 \text{ content} - \text{dissolved } O_2}{O_2 \text{ capacity}} \times 100$$

Detection of L→R Intracardiac shunts

Oximetry run

The oxygen content or percent saturation is measured in blood samples drawn sequentially from the pulmonary artery, RV, RA, SVC and IVC. If there is an interatrial communication, a pulmonary venous saturation may be obtained. Otherwise, in the presence of a normal systemic saturation, it is assumed to be equal to the systemic saturation in the absence of a right to left intracardiac shunt. A left to right shunt may be detected and localised if a significant step up in blood oxygen saturation or content is found in right heart chambers

Normal Oxygen Saturation

Site	Average	Range
Superior vena cava	74%	67 – 83%
Inferior vena cava	78	65 – 87
Right atrium	75	65 – 87
Right ventricle	75	67 – 84
Pulmonary artery	75	67 – 84
Left atrium	95	92 – 98
Left ventricle	95	93 – 98
Systemic artery	95	92 - 98

Criteria for significant step up

Atrial level >9%
Ventricular level >6%
Great artery level >6%

Possible causes of step-up

RA - ASD, PAPVC, Ruptured Sinus of Valsalva to RA, VSD with TR ,coronary fistula to RA

RV - VSD, PDA with PR, Coronary fistula to RV

Great Vessel (RV to PA) – PDA, Aortopulmonary window

A step down of >2% or an aortic saturation of <92% is suggestive of R à L shunt.

- If the oximetry run reveals that a significant step up is present, the pulmonary blood flow, systemic blood flow and magnitude of left to right and right to left shunts can be calculated to decide whether surgery is necessary
- A pulmonary to systemic flow ratio of greater than 2:1 usually indicates a large left to right shunt and a defect that should be surgically closed because of the volume of the shunt.

Calculations in L →R lesions

Calculation of oxygen content and A-V oxygen difference

- Step 1: Oxygen carrying capacity: Hemoglobin (gm/dl) x 1.36(ml of O_2/gm of Hb) x 10 = _____mlO_2/L of blood
- Step 2: Saturation of arterial (BA,FA,AO) blood = _____%
- Step 3: Oxygen content of arterial blood:
  O_2 capacity x % saturation = ———ml/L
  (step 1) (step 2)
- Step 4: Saturation of mixed venous blood = $\frac{[3\text{SVC } O_2 \text{ sat\%} + \text{IVC } O_2 \text{sat\%}]}{4}$
- Step 5: O_2 content of mixed venous blood:
  O_2 capacity x % saturation = _____ml/L
  (step 1) (step 4)
- Step 6: AVO_2 difference:
  Arterial O_2 content – Venous O_2 content = _____ml/L
  (step 3) (step 5)

Calculation of pulmonary and systemic blood flow

- Qp is the amount of blood flowing through the pulmonary capillary bed (pulmonary flow)

$$\text{Qp (L/min)} = \frac{O_2 \text{ consumption (VO2) (ml/min)}}{PVO_2 \text{ content (ml/L)} - \text{PA } O_2 \text{ content (ml/L)}}$$

$$= \frac{VO_2}{(\text{PVsat} - \text{PAsat})(O_2 \text{ capacity})}$$

· Qs is the amount of blood flowing through the systemic capillaries (systemic flow) (VO2) (ml/min)

$$\text{Qs} = \frac{O_2 \text{ consumption (VO2) (ml/min)}}{SaO_2 \text{ content (ml/L)} - MVO_2 \text{ content (ml/L)}}$$

$$= \frac{VO_2}{(\text{SAsat} - \text{MVsat})(O_2 \text{ capacity})}$$

$$MVO_2 \text{ content} = \frac{3(SVCO_2 \text{ content}) + 1(\text{IVC } O_2 \text{ content})}{4}$$

- Calculation of Lt to Rt shunt

– L →R Shunt (L/min) = Qp – Qs

- Flow Ratio

– The ratio Qp/Qs gives important physiologic information about the magnitude of left to right shunt. A simplified formula for calculation of flow ratio is

$$\frac{QP}{QS} = \frac{SA\ sat - MV\ Sat}{PV\ Sat - PA\ Sat}$$

Calculation of flow during oxygen administration

- Measurement of oxygen content in the blood samples is most reliable.
- If O_2 saturations are measured, it is imperative that PO_2 be measured to correct for dissolved oxygen. Dissolved O_2 is about 0.3 ml/dl or 3ml/L at a PO_2 of 100mmHg
- For e.g. when a patient is breathing room air and systemic arterial or pulmonary venous PO_2 is about 100mmHg, then O_2 sat is almost 100% and dissolved O_2 is only about 3ml/L
- If there is moderately large left to right shunt, with a relatively high O_2 saturation of 85% in pulmonary artery and if VO2 is 150ml/min and Hb 14.5 g/dl, then

O_2 capacity = 14.5 x 1.36 x 10 = 200ml/L

PVO_2 content = $\frac{100}{100}$ x 200 + 3 (dissolved) = 203 ml/L

PaO_2 content = $\frac{85}{100}$ x 200 + 2.5 = 172.5ml/L

$$Qp = \frac{150}{203 - 172.5} = 4.9\ L/min$$

If dissolved O_2 not taken into account,

PVO_2 = 200ml/L and $CPaO_2$ = 170ml/L
Qp = 5.0 L/min

Thus little error is produced during breathing of room air.

If patient is breathing 100% O_2 and pulmonary arterial saturation is 92%,
PO_2 is pulmonary venous blood is 550mmHg and PO_2 in pulmonary arterial blood is 100mmHg
PVO_2 content = $\frac{100}{100}$ x 200 + 3x5.5 (dissolved) = 216.5ml/L
PAO_2 content = $\frac{92}{100}$ x 200 + 3x1 (dissolved) = 187ml/L
Qp = 150/216.5 – 187.0 = 5.1 L/min
If dissolved O2 is not taken into consideration,
PVO_2 content = 200ml/L, PAO_2 content = 184ml/L
Qp = 150/200-184 = 9.4L/min
This represents an overestimate of pulmonary blood flow of almost 100%

Limitations and errors in calculating flows and shunts by the Fick method

1. Absence of a steady state during the collection of blood samples
2. A mixed venous sample may not be obtainable when there is a shunt of systemic arterial or pulmonary venous blood into the systemic veins. When one or more pulmonary veins drain into SVC / IVC, venous blood must be collected before their entry.

3. Oxygen saturations in the ascending and descending aorta may be different when there is a right to left shunt through the ductus arteriosus.
4. Pulmonary arterial oxygen saturation may be different in the left and right pulmonary artery in patients with patent ductus arteriosus.
5. Assumed values for oxygen consumption.
6. When the arteriovenous difference is large, the errors inherent in measuring oxygen content or saturation do not result in major errors in calculation of flow. However when arteriovenous difference is small, small errors in measurement may result in large errors of flow measurement.
7. Calculations of flow during oxygen administration – Here if the dissolved oxygen is not considered, it leads to an overestimation of pulmonary blood flow.
8. Lacks sensitivity – small shunts are not consistently detected
9. Influenced by the blood Hb concentration.
10. Improper collection of mixed venous blood sample (e.g. air bubbles) or dilution of the sample with too much heparinised saline solution.

Resistance

The calculation for resistance for blood flow is based on Poiseuille's law:

$$R = \frac{P_{in} - P_{out}}{Q}$$

Q is the flow, P_{in} is the mean inflow pressure, P_{out} is the mean outflow pressure and R is the resistance

The applicability of laws derived from steady state fluid mechanics in assessing vascular resistance is somewhat ambiguous because blood flow is pulsatile, blood is a nonhomogenous fluid and the vascular bed is a nonlinear elastic, frequency dependent system.

Normal systemic vascular resistance is 20 mmHg/L/min/m^2 in children, but varies markedly between 15 – 30 mmHg/L/min/m^2. Normal pulmonary vascular resistance is 1 –3 mmHg/L/min/m^2 (Wood Units).

Formula for calculating resistance

1. Systemic vascular resistance (SVR)

$$= \frac{\text{Aortic mean pressure - RA mean pressure}}{Qs}$$

2. Pulmonary vascular resistance (PVR)

$$= \frac{\text{PA mean pressure - LA/PCW mean pressure}}{Qp}$$

In children, it is conventional to normalize vascular resistances for body surface area (BSA), thus giving a resistance index.

SVR index (SVRI) = SVR x BSA
PVR index (PVRI) = PVR x BSA

Please note that the indices are obtained by multiplying the calculated resistance by the BSA.

Assessment of operability

1. A Qp/Qs <1.5 → a small left to right shunt , may be left alone.
   ≥2.0 → a large left to right shunt → surgical repair to prevent late pulmonary vascular disease as well as other complications of prolonged circulatory overload.
   A Qp/Qs <1 indicates that there is a resting right to left shunt, which may preclude surgical repair

2. The ratio between PVR and SVR and the absolute PVRI are used criteria for assessing operability in L → R shunts.

PVRI	PVR / SVR	INTERPRETATION
<8 WU	0.25 – 0.5	Operable (moderate pulmonary vascular disease)
8 – 12 WU	0.5 – 0.75	Borderline operability (high risk)
>12 WU	>0.75	Inoperable (severe pulmonary vascular disease)

Also, on oxygen administration, if the diastolic pressures fall significantly (by about 10 mmHg) and mean pressure falls by about 5mmHg, it indicates reversible pulmonary artery pressures and hence operability.

Representative data of common L → R shunts

1. Large VSD – severe PAH with calculation of shunt and vascular resistances.

height – 112 cm, weight – 15kg, Hb – 13gm%, BSA – 0.68m^2, VO_2 (assumed) – 97ml/mt

Hemodynamic and oximetry data

Pressures	On room air	On oxygen
Descending aorta	90/50 (68)	94 / 55 (74)
LPA	88 / 46 (66)	86 / 45 (65)
PCWP (mean)	3	2
RA	a^5v^3 (1)	a^5v^3 (1)
RV	90 / 5	84 / 3

Saturations (PO_2)	On room air	On oxygen
SVC	78.7 (41.5)	85.8 (47.2)
IVC	79.3 (41.8)	87.1 (47.7)
LPA	83 (48.1)	88.8 (57.2)
DA	96.8 (84.8)	99.6 (154.6)
PV (assumed)	98	100 (300)

Calculation of shunts and resistances

On room air

$$Qp = \frac{VO_2}{\text{Pul. Venous } O_2 \text{ content} - \text{pul. Artery } O_2 \text{ content}}$$

$$= \frac{97}{Hb \times 1.36 (PVO2sat\% - PA\ O2\ sat\%)}$$

$$= \frac{97}{Hb \times 1.36\ (0.98\text{-}0.83) \times 10}$$

$$= 3.66$$

$$Qs = \frac{VO_2}{\text{Aortic } O_2 \text{ content} - \text{mixed venous } O_2 \text{ content}}$$

$$= \frac{97}{Hb \times 1.36\ (\text{Aortic sat\%} - \text{mixed venous sat\%})}$$

$$\text{Mixed venous saturation} = \frac{(3SVC\ sat + IVCsat)}{4}$$

$$= \frac{97}{Hb \times 1.36\ (0.96\text{-}0.78) \times 10}$$

$$= 3.05$$

$$Qp / Qs = 1.19 : 1$$

$$PVR = \frac{PA\ mean - PCWP\ mean}{Qp} = \frac{66 - 3}{3.66} = 17.2$$

$$PVRI = 17.2 \times BSA = 17.2 \times 0.68 = 11.7$$

$$SVR = \frac{Aortic\ mean - RA\ mean}{Qs} = \frac{68 - 1}{3.05}$$

$$= 21.96$$

$$SVRI = 21.96 \times BSA = 21.96 \times 0.68 = 14.9$$

$$PVR / SVR = 0.78:1$$

Similarly, On oxygen

$$Q_p\ (L/min) = \frac{VO_2\ m1/min}{(\text{Pulm. Venous } O_2 \text{ Content} - \text{PA } O_2 \text{ Content})\ (m1/L)}$$

$$O_2 \text{ Content} = \text{Theoretic Capacity} \times \%\ \text{Sat} + \text{Dissolved } O_2$$

$$= (Hb \times 1.36 \times 10 \times \text{Saturation}) + (0.03 \times PO_2)$$

Pulm. Venous O_2 Content

$$= (1 \times 13 \times 1.35 \times 10) + (0.03 \times 300)$$

$$= \underline{185.8}$$

PA O_2 Content $= (0.88 \times 13 \times 1.36 \times 10) + (0.03 \times 57.2)$

$$= \underline{158.61}$$

$Q_P = \frac{97}{185.8 - 158.61} = \frac{97}{27.19} = 3.56$

$Q_P = 3.56$

$Q_S = \frac{VO_2}{AO\ O_2\ Content - \frac{(3SVC + IVC)\ O_2\ content}{4}}$

AO O_2 Content = 0.99 x 13 x 1.65 x 10 + 0.03 x 154.6
176 + 4.63 = 180.63

Syst. Venous O_2 Content = 0.86 x 13 x 1.36 x 10 + 0.03 x 47.5

$\frac{[3\ SVC + 1\ IVC\ ]}{4} = 86.10$

152.8 + 1.42 = 154.22

$Q_S = \frac{97}{180.63 - 154.22} = \frac{97}{26.41} = 3.67$

$Q_S = 3.67$

$Q_P/Q_S = 0.97:1$

PVR = $\frac{\text{Mean PA Pressure – LA Pressure (PCW)}}{Q_P}$

$= \frac{65 - 2}{3.56} = 17.6$

PVRI = 17.6 x 0.68 (BSA) = 11.9

PVRI = 11.9

SVR = $\frac{\text{Mean Aortic Pressure – RA Pressure}}{Q_S}$

$= \frac{74\text{-}1}{3.67} = 19.89$

PVR / SVR = 17.6 / 19.89 = 0.88
PVR/SVR = 0.88

Impression: Large VSD with irreversible PA pressures.

2. ASD

	On room air	
	O_2 saturation	Pressure
SVC	72	
IVC	74	
RA	85	a^5v^4 (5)
RV	88	30/6
PA	89	30/15 (20)
PAW (LA)	100	a^5v^4 (5)
LV	98	100/12
Ao	97	100/70 (85)
FA	98	110/70 (92)

Note the step up in the right atrium and identical RA & LA phasic and mean pressures. The pulmonary resistance in a large ASD is often as low as 0.8 – 1 wood units. If it is greater than 3 wood units, it denotes pulmonary vascular disease.

PDA

	On room air	
	O2 saturation	Pressure
SVC	73	
IVC	75	
RA	75	
RV	75	40 / 5
MPA	84	42/20 (27)
LPA	87	42/20 (27)
RPA	84	42/20 (27)
PAW		13
DA	97	120/56 (70)
FA	98	

In case of PDA, it is usually not possible to obtain a sample that is truly representative of mixed pulmonary arterial blood. The saturation in the LPA tends to be higher than in RPA because blood shunted through the ductus passes preferentially to the left side.

References:

1) Congenital Diseases of the Heart : Clinical Physiological Considerations . Fully Revised and Updated Second Edition – Abraham M. Rudolph MD.

2) Grossman's Cardiac catheterization, Angiography and Intervention, Sixth Edition - D.S. Baim and W. Grossman 2000.

TABLE 43.8. Oxygen Consumption per Body Surface Area in (mL/minute)/M² by Sex, Age, and Heart Rate

	Heart Rate (beats per minute)												
Age	*50*	*60*	*70*	*80*	*90*	*100*	*110*	*120*	*130*	*140*	*150*	*160*	*170*
Male patients													
3				155	159	163	167	171	175	178	182	186	190
4			149	152	156	160	163	168	171	175	179	182	186
6		141	144	148	151	155	159	162	167	171	174	178	181
8		136	141	145	148	152	156	159	163	167	171	175	178
10	130	134	139	142	146	149	153	157	160	165	169	172	176
12	128	132	136	140	144	147	151	155	158	162	167	170	174
14	127	130	134	137	142	146	149	153	157	160	165	169	172
16	125	129	132	136	141	144	148	152	155	159	162	167	
18	124	127	131	135	139	143	147	150	154	157	161	166	
20	123	126	130	134	137	142	145	149	153	156	160	165	
25	120	124	127	131	135	139	143	147	150	154	157		
30	118	122	125	129	133	136	141	145	148	152	155		
35	116	120	124	127	131	135	139	143	147	150			
40	115	119	122	126	130	133	137	141	145	149			
Female patients													
3				150	153	157	161	165	169	172	176	180	183
4			141	145	149	152	156	159	163	168	171	175	179
6		130	134	137	142	146	149	153	156	160	165	168	172
8		125	129	133	136	141	144	148	152	155	159	163	167
10	118	122	125	129	133	136	141	144	148	152	155	159	163
12	115	119	122	126	130	133	137	141	145	149	152	156	160
14	112	116	120	123	127	131	134	138	143	146	150	153	157
16	109	114	118	121	125	128	132	136	140	144	148	151	
18	107	111	116	119	123	127	130	134	137	142	146	149	
20	106	109	114	118	121	125	128	132	136	140	144	148	
25	102	106	109	114	118	121	125	128	132	136	140		
30	99	103	106	110	115	118	122	125	129	133	136		
35	97	100	104	107	111	116	119	123	127	130			
50	94	98	102	105	109	112	117	121	124	128			

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LEFT VENTRICULAR PRESSURE VOLUME LOOP

Prof. G. Gnanavelu

Pressure volume loop (PV loop) is a time independent representation in the form of a loop generated by plotting pressure against volume at many time points during a complete cardiac cycle. Pressure volume loop can be generated practically for any chamber i.e. atrium and ventricle both right and left. The ventricular pressure volume loop is a powerful tool to analyse ventricular function. Various aspects of left ventricular pressure volume loop will be discussed in this article.

The left ventricular PV loop for a single cardiac cycle typically represents four basic phases. Phase 'a' – diastole (ventricular filling), phase 'b' isovolumic contraction; phase 'c' – systole (ejection) and phase 'd' isovolumic relaxation. (Fig.1)

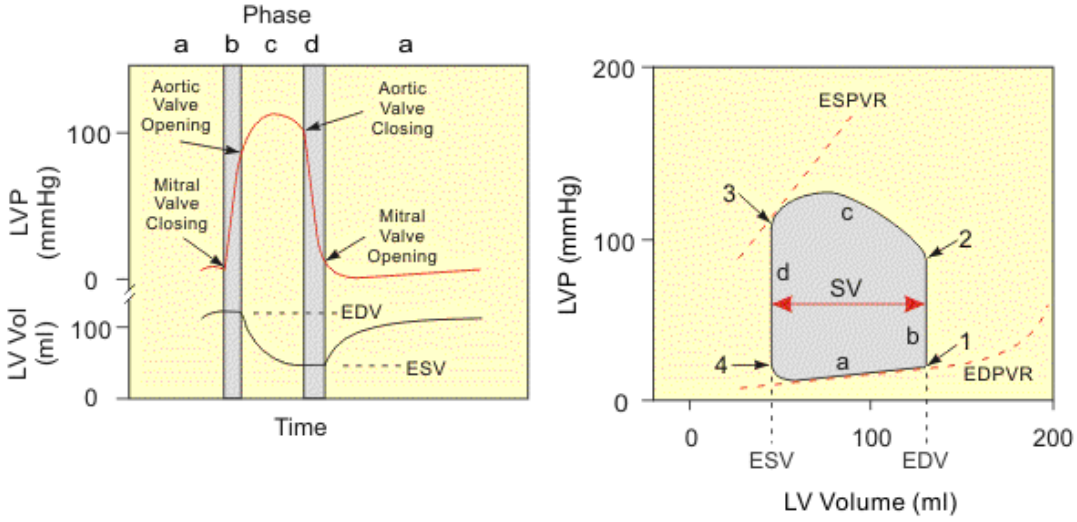


Fig. 1: *Left ventricular pressure volume loop. Explanation is given in the text. ESPVR – End systolic pressure volume relationship; EDPVR – End diastolic pressure volume relationship.EDV – End diastolic volume; ESV – End systolic volume; SV – Stroke volume; LVP – Left ventricular pressure*

Point 1 on the PV loop is the pressure and volume at the end of diastole and therefore represents end diastolic pressure and end diastolic volume for the ventricle. As the ventricle begins to contract isovolumetrically (phase b), the left ventricular pressure increases but the volume remains the same, therefore resulting in a vertical line (all the valves are closed). Once left ventricular pressure exceeds aortic diastolic pressure, the aortic valve opens (point 2) and ejection (phase c) begins. During this phase the left ventricular volume decreases as left ventricular pressure increases to a peak value and then decreases as the ventricle begins to relax. When the aortic valve closes (point 3) ejection ceases and the ventricle relaxes isovolumetrically ie. the left ventricular pressure falls but the volume remains unchanged, therefore the line is vertical (all the valves are closed). The left ventricular volume at this time is the end systolic volume i.e. residual volume. When the left ventricular pressure falls below left atrial pressure, the mitral valve opens (point 4) and the ventricle begins to fill. Initially the left ventricular pressure continues to fall as the ventricle fills because the ventricle is still relaxing. However once the ventricle is fully relaxed, the pressure gradually increases as the volume increases. The width of the loop represents the difference between end diastolic volume and end systolic volume, which is the stroke volume. The area within the loop is the ventricular stroke work.

Effects of preload, afterload, inotropy and heart rate on ventricular pressure volume loop:

The major determinants of ventricular function are

1. Preload
2. Afterload
3. Inotropic state
4. Heart rate

Ventricular pressure volume loops represent excellent images to visualise changes in ventricular function in response to changes in preload, afterload, inotropy and heart rate.

All the pathophysiologic conditions manipulate the pressure volume loop by altering one or more of these determinants. Proper understanding of pressure volume loop is facilitated by understanding Frank- Starling relationship, Force velocity relationship, Pressure volume relationship during diastole and systole.

Frank- starling relationship:

Frank-Starling mechanism relates venous return and stroke volume for the ventricle. When venous return to the heart is increased, ventricular filling increases i.e preload increases. This stretches the myocytes more leading to increase in force of contraction. This enables the ventricle to eject the additional venous return. (Fig. 2)

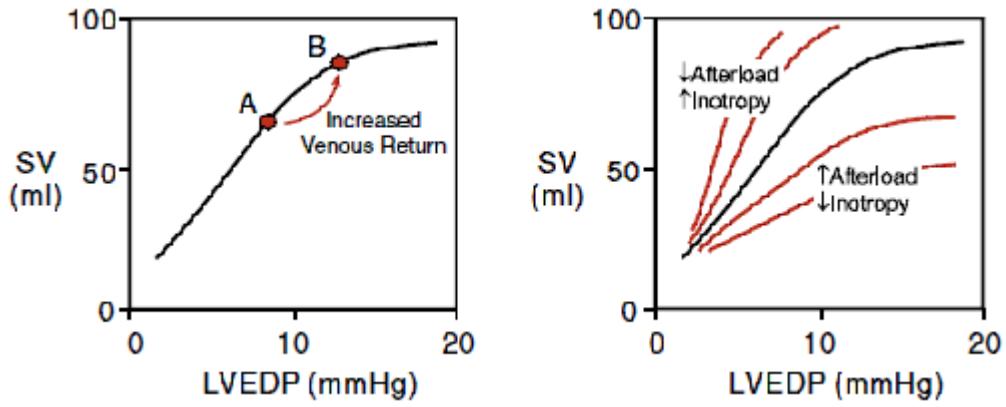


Fig.2: Frank Starling curve: Left panel : Increasing venous return to left ventricle increases left ventricular end diastolic pressure by increasing ventricular volume, this increases ventricular preload, resulting in an increase in stroke volume from point A to B. Right panel: A family of Frank- Starling curves generated at different afterloads and inotropic states. Increased afterload or decreased inotropy shifts the curve downward. Decreased afterload or increased inotropy shifts the curve upward.

Frank Starling mechanism can be represented by Frank Starling curve otherwise called ventricular function curve. There is no single Frank Starling curve for the ventricle but a family of curves with each curve defined by the afterload imposed on the heart and the inotropic state of the heart. Increasing afterload and decreasing inotropy shifts the curve down and to the right, whereas decreasing afterload and increasing inotropy shifts the curve up and to the left. In summary, changes

in venous return cause a ventricle to function along a Frank Starling curve that is defined by the existing conditions of afterload and inotropy. (Fig.2)

Frank Starling mechanism plays an important role in balancing the output of the two ventricles. When venous return increases to the right side of the heart eg. during physical activity, this mechanism enables the right ventricular stroke volume to increase, thereby matching its output to the increased venous return. This increases the venous return to the left side of the heart and the same mechanism increases the left ventricular stroke volume thereby matching the outputs of the two ventricles over time.

However the Frank Starling curves, do not show how changes in venous return affect end diastolic and end systolic volumes. These changes in ventricular volumes are best illustrated by pressure volume relationships.

Force velocity relationship:

Force velocity curve for ventricle describes the relationship between afterload and velocity of fiber shortening. Force here is afterload and the velocity is the velocity of muscle shortening. When afterload is reduced the velocity of shortening increases, thus increasing stroke volume. When afterload increases the stroke volume decreases due to impaired emptying of ventricle. However, increasing the preload enables the ventricle to contract faster against a given afterload. This shifts the force velocity relationship to the right. Therefore an increase in preload helps to offset the reduction in velocity that occurs when afterload is increased. (Fig. 3)

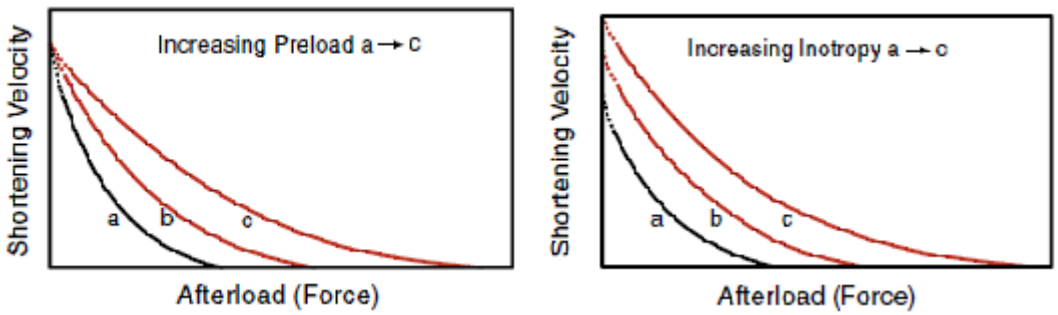


Fig. 3 : *Left panel : At a given afterload, increasing the preload increases the velocity of shortening. Increasing preload shifts the curve from a to c. Right panel : Increased inotropy increases the velocity of shortening at any given afterload. Increasing inotropy shifts the curve parallel from curve a to c .*

Left ventricular pressure volume relationship during diastole:

The end-diastolic pressure-volume relationship has been used to describe the filling or compliance properties of the ventricle. The filling phase moves along the end diastolic pressure volume relationship or passive filling curve for the ventricle. The relationship is a curve rather than a straight line. The slope of this curve (delta P/delta V) represents the stiffness or technically called elastance of the ventricle. The compliance or distensibility at any point is the inverse of the slope of the line at that point (delta V/delta P). (Fig. 4)

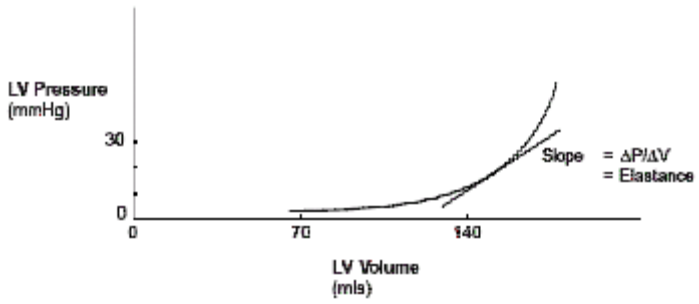


Fig. 4 : *Filling curve or compliance curve for left ventricle.*

The curve is fairly flat and only curves slowly upward within physiological range i.e the left ventricular volume increases without much increase in left ventricular pressure. The curve begins to rise steeply outside the typical value for left ventricular end diastolic volume i.e above 140 ml. In other words, the ventricle is easy to fill within physiologic range and difficult to overfill as the increase in pressure tends to impede excess increase in left ventricular end diastolic volume. To put in simple terms 'stiffer ventricle is harder to fill'.

The *preload* of the left ventricle describes the intraventricular pressure and volume immediately prior to contraction. Ideally, end diastolic volume is the best measure of preload. Clinically, it is easier to measure end diastolic pressure than end diastolic volume. So, the filling pressure otherwise called end diastolic pressure is used clinically as a measure of preload.

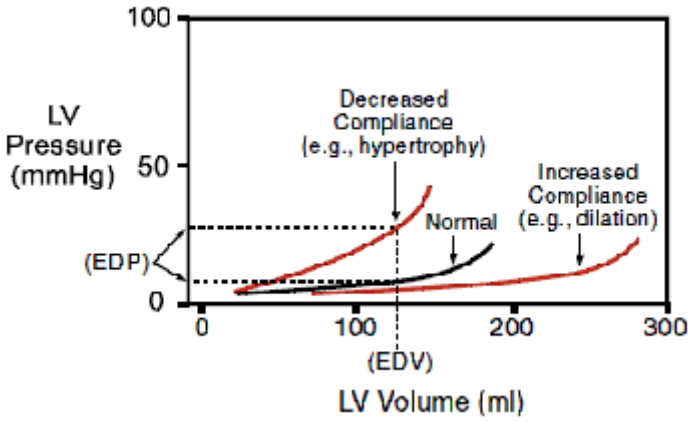


Fig. 5: *Ventricular compliance curves or filling curves.*

The slope of the normal filling curve is increased by a decrease in ventricular compliance (eg.) ventricular hypertrophy, whereas the slope of the compliance curve is reduced by an increase in ventricular compliance (eg. Ventricular dilatation). (Fig. 5) Decreased compliance increases the end diastolic pressure at a given end diastolic volume, whereas increased compliance decreases end diastolic pressure at a given end diastolic volume.

Left Ventricular Pressure Volume Relationship During Systole:

The maximal pressure that can be developed by the ventricle at any given left ventricular volume is defined by the endsystolic pressure volume relationship (ESPVR), which represents the inotropic state of the ventricle. The pressure-volume loop therefore cannot cross over the ESPVR, because that relationship defines the maximal pressure that can be generated under a given inotropic state. (Fig. 6)

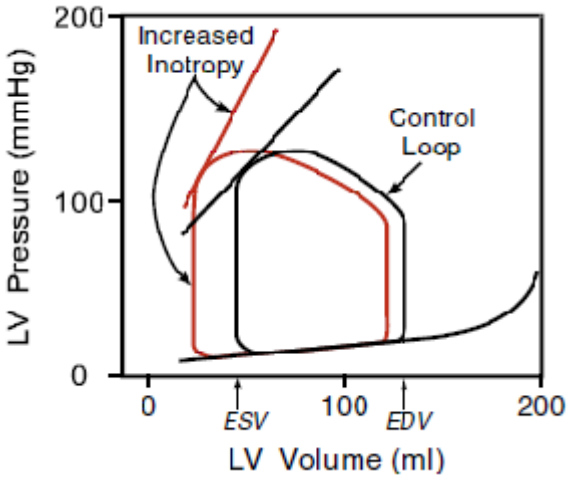


Fig. 6 : *Increasing inotropy on the end systolic pressure volume relationship shifts the slope up and to the left, thereby decreasing endsystolic volume. A secondary but smaller decrease in end diastolic volume follows. The net effect is an increase in stroke volume.*

Independent effects of preload on pressure volume loop:

To study the independent effects of preload on PV loop, the aortic pressure and inotropic state of the ventricle are assumed constant. If preload is increased by increasing the end diastolic volume eg. with intravenous fluids, the ventricle develops greater pressure and ejects blood more rapidly by Frank Starling mechanism. So the ventricle ejects blood to the same end systolic volume. The net effect will be an increase in stroke volume, shown by the width of the PV loop. If the preload is decreased eg. during dehydration or blood loss, opposite changes occur ie. stroke volume is decreased but the end-systolic volume is unchanged. (Fig.7)

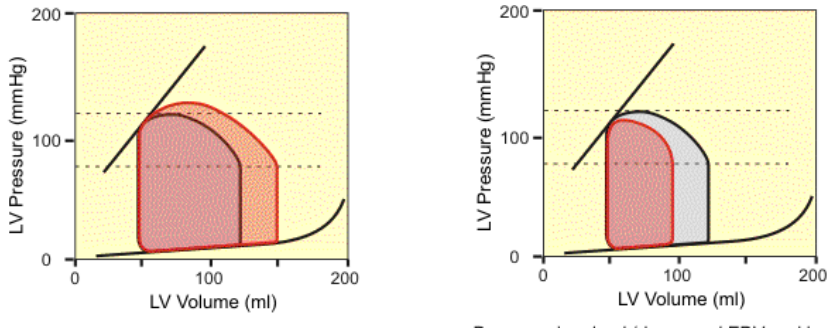


Fig. 7 : *Left panel : Increased preload (EDV) (dark shaded loop) at constant aortic diastolic pressure and inotropy. Stroke volume increases and ESV remains unchanged. Right panel : Decreased preload (EDV) (dark shaded loop) at constant aortic diastolic pressure and inotropy. Stroke volume decreaes and ESV remains unchanged.*

Independent effects of afterload on pv loop:

If afterload is increased by increasing aortic diastolic pressure eg. high blood pressure and if the preload and inotropy are held constant, this will result in less stroke volume and an increase in end systolic volume. Stroke volume is reduced because the increased afterload reduces the velocity of muscle fiber shortening and the velocity of ejection of blood. The reduced stroke volume at the

same end diastolic volume reduces the ejction fraction. If afterload is reduced eg. Hypotension, by decreasing aortic diastolic pressure, the opposite occurs ie. the stroke volume and ejection fraction increase and end systolic volume decreases. (Fig. 8)

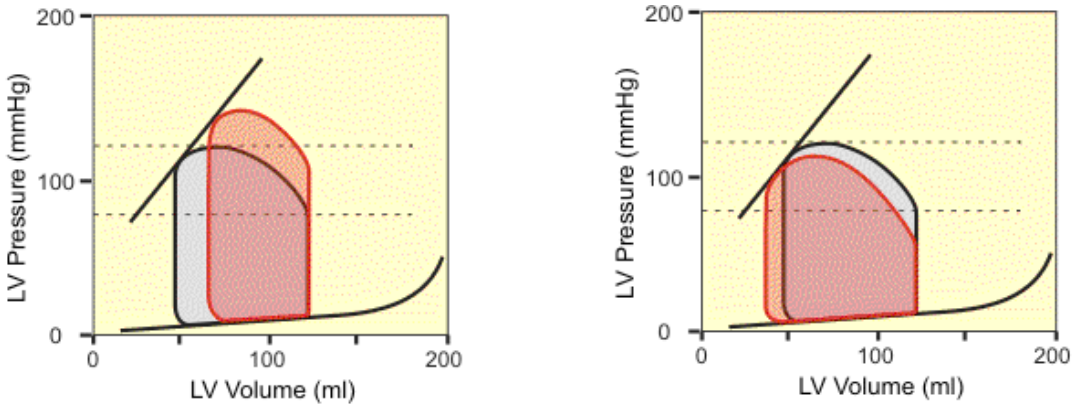


Fig. 8 : *Left panel : Increased afterload (aortic pressure) (dark shaded loop) at constant preload and inotropy. Stroke volume decreases and ESV increases. EF decreases. Right panel : Decreased afterload (aortic pressure) (dark shaded loop) at constant preload and inotropy. Stroke volume increases and ESV decreases. EF increases.*

Independent effects of inotropy on PV loop:

Increasing inotropy increases the velocity of fiber shortening at any given preload and afterload. This enables the ventricle to increase the rate of pressure development and ejection velocity, leading to an increase in stroke volume and ejection fraction and a decrease in end systolic volume. In PV loop, increased inotropy increases the slope and shifts the end systolic pressure-volume relationship to the left, which permits the ventricle to generate more pressure at a given volume. Decreasing inotropy has the opposite effects, namely increased end systolic volume and decreased stroke volume and ejection fraction. The effects of inotropy on PV loop are shown in Fig. 9.

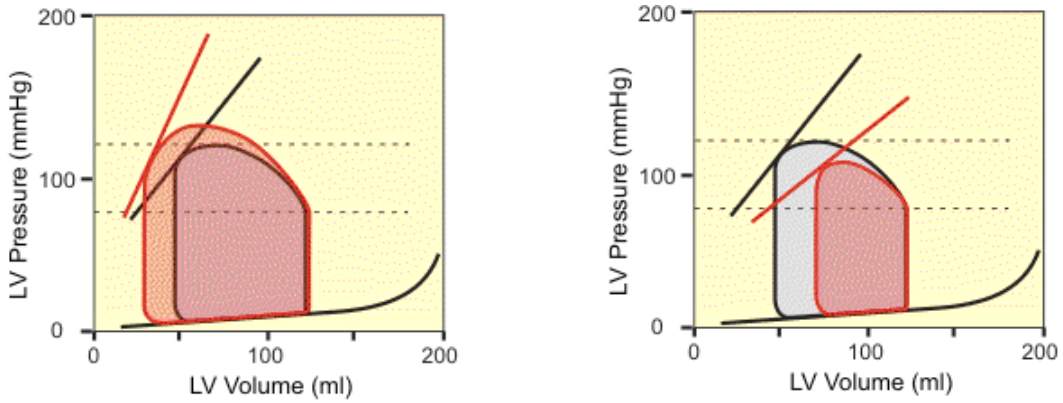


Fig. 9 : *Left panel : Increased inotropy (dark shaded loop) at constant preload and aortic diastolic pressure (afterload). Stroke volume increases and ESV decreases. EF increases. Right panel : Decreased inotropy (dark shaded loop) at constant preload and afterload. Stroke volume decreases and ESV increases. EF decreases.*

Independent effects of heart rate on PV loop:

Heart rate has no independent effect on pressure-volume loops. The rate dependent increase in contractility tends to shift the end systolic pressure volume line up and to the left, but the decreased diastolic filling time tends to simultaneously reduce preload.

Interdependent effects of preload, afterload and inotropy:

All the determinants of ventricular function are interdependent and a change in one parameter changes others. The interdependent effects and interactions between afterload, preload and inotropy are shown below. The above described independent effects of the three important variables namely preload, afterload and inotropy do not represent what happens in the intact ventricle. However it is easy to combine the loops and understand the interactions between each variable better, when the independent effects are understood well.

Interactions between preload and afterload at constant inotropy:

An increase in preload i.e. end diastolic volume leads to an increase in stroke volume according to Frank Starling curve. If afterload and inotropy do not change, then the endsystolic volume will not change. The heart simply ejects all of the extra blood that it receives. However if the increased stroke volume leads to an increase in cardiac output and arterial pressure, then the afterload on the ventricle increases, which partially offsets the increased stroke volume by increasing end systolic volume. The reason for this is that the increased afterload reduces the velocity of fiber shortening and therefore the ejection velocity (Force velocity relationship). Conversely, a decrease in preload reduces stroke volume, but this reduction is partially offset by the decreased afterload so that the end systolic volume decreases slightly. The effects of changes in preload when arterial pressure changes are illustrated in Fig. 10.

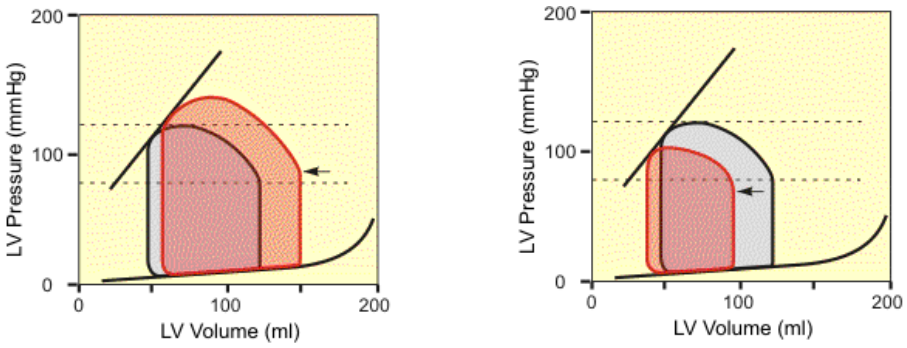


Fig. 10 : *Left panel : Increased preload (increased EDV – dark shaded loop) at constant inotropy. Stroke volume increases and ESV increases slightly because afterload (aortic diastolic pressure – arrow) increases. Right panel : Decreased preload (decreased EDV – dark shaded loop) at constant inotropy. Stroke volume decreases; ESV decreases slightly because afterload (aortic diastolic pressure – arrow) decreases.*

Interdependent effects of changes in afterload:

If afterload is increased, the stroke volume is reduced and the end systolic volume is increased. The increased end systolic volume, however leads to a secondary increase in end diastolic volume because more blood is left inside the ventricle following ejection and this extra blood is added to the venous return thereby increasing ventricular volume. This secondary increase in preload, by Frank Starling mechanism

partially offsets the reduction in stroke volume caused by the initial increase in afterload. Consequently, in a normal heart, changes in aortic pressure have little effect on stroke volume. However in heart failure patients in which the end diastolic volume is already maximal, an increase in aortic pressure can significantly reduce stroke volume. If afterload is reduced, the opposite changes occur ie. stroke volume increases due to the decrease in end systolic volume, accompanied by a smaller reduction in end diastolic volume. This forms the basis of administering vasodilators in patients with heart failure. (Fig. 11)

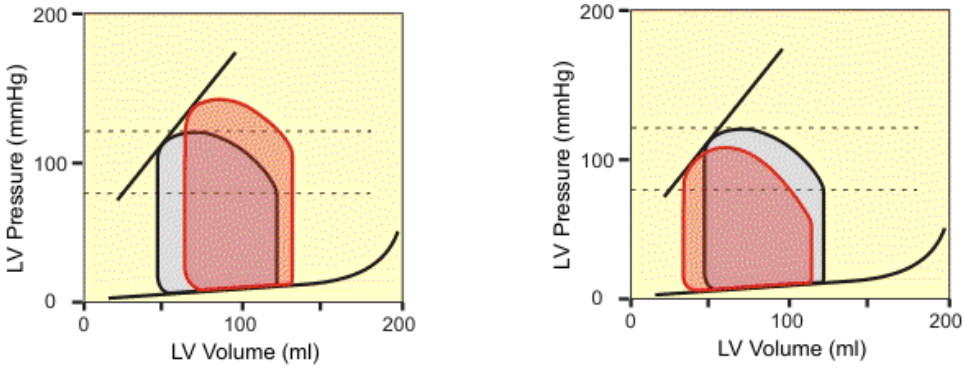


Fig. 11 : *Left panel : Increased afterload (increased aortic pressure – dark shaded loop) at constant inotropy. Stroke volume slightly decreases. ESV increases and EDV slightly increases secondarily. Right panel : Decreased afterload (decreased aortic pressure – dark shaded loop) at constant inotropy. Stroke volume slightly increases. ESV decreases and EDV slightly decreases secondarily.*

Interdependent effects of changes in inotropy:

Increased inotropy increases the slope and shifts the end systolic pressure volume relationship to the left, which permits the ventricle to generate more pressure at a given volume. Increased inotropy also increases the rate of pressure development and ejection velocity, which increases stroke volume and ejection fraction and decreases end systolic volume. With less end systolic volume the ventricle fills to a smaller end diastolic volume during diastole. Decreasing inotropy has the opposite effects namely, end systolic volume increases and stroke volume and ejection fraction decrease accompanied by a small increase in end diastolic volume. The effects of inotropy on PV loop are shown in Fig. 12.

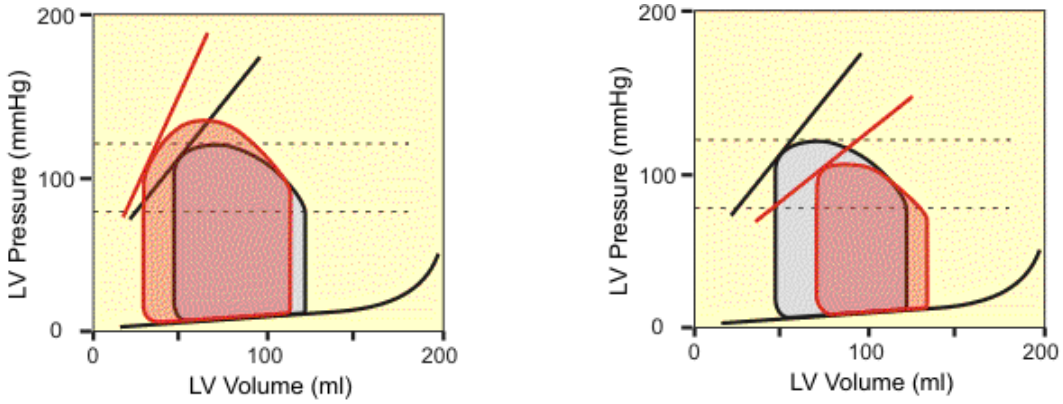


Fig. 12 : *Left panel : Increased inotropy (dark shaded loop). Stroke volume increases. ESV decreases and EDV decreases secondarily a small amount. EF increases.*
Right panel : Decreased inotropy (dark shaded loop) Stroke volume decreases. ESV increases and EDV increases secondarily a small amount. EF decreases.

The changes in PV loop in some of the pathophysiologic states will be discussed in the following sections:

Systolic dysfunction:

Systolic dysfunction implies impaired ventricular contraction ie. loss of inotropy. Loss of inotropy causes a downward shift in the Frank Starling curve. This leads to a decrease in stroke volume, increase in end systolic volume and a compensatory rise in preload. The increased end systolic volume is added to the normal venous return filling the ventricle, increasing the diastolic pressure and volume. The rise in preload is considered compensatory because it helps maintain stroke volume despite the loss of inotropy. If preload did not rise, the decline in stroke volume would be greater for a given loss of inotropy. (Fig. 14).

However the compensatory increase in end diastolic pressure proves deleterious in due course, raising the left atrial pressure and pulmonary venous pressure resulting in pulmonary congestion and edema. In case of right ventricular systolic dysfunction, the rise in right ventricular end diastolic pressure leads to raised right atrial pressure and systemic venous congestion, resulting in peripheral edema and ascites.

In a PV loop, the loss of inotropy decreases the slope of the end systolic pressure volume relationship. This leads to an increase in end systolic volume. There is also a compensatory increase in end diastolic volume, but this increase is not as great as the increase in end systolic volume. Therefore the net effect is a decrease in stroke volume (note the reduced width of the PV loop). As stroke volume decreases and end diastolic volume increases there is a reduction in ejection fraction. (Fig. 13)

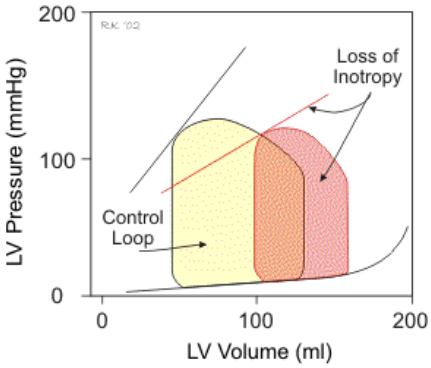


Fig. 13. *The effects of loss of inotropy on PV loop .*

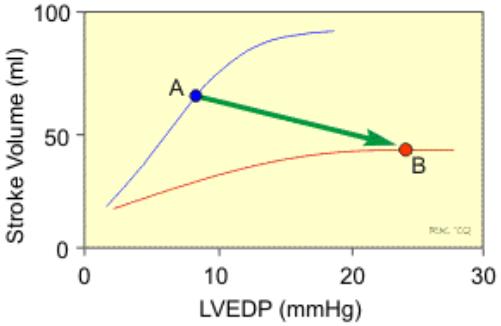


Fig. 14. *Frank Starling relationship showing effects of systolic dysfunction on stroke volume and preload. Point A – control point; Point B – systolic dysfunction*

The treatment of systolic dysfunction typically involves inotropic drugs eg. digitalis which increases contractility, afterload reducing agents ie. arterial vasodilators which by reducing afterload increases ventricular ejection (see Force velocity relationship) and preload reducing drugs ie. diuretics which reduce preload and venous pressures.

Diastolic dysfunction:

Ventricular function is very much dependent on preload according to Frank Starling law. Preload depends on venous return and compliance of the ventricle during diastole. In diastolic dysfunction eg. ventricular hypertrophy, the compliance decreases resulting in reduced ventricular filling or filling occurs at higher end diastolic pressure. This is shown in the PV loop in Fig. 15. Diastolic pressure volume curve moves up as compliance decreases, leading to a decrease in stroke volume, increase in end diastolic pressure. The increase in end diastolic pressure is a deleterious effect of diastolic dysfunction resulting in raised atrial pressures and venous congestion. If the left ventricle is involved the left atrial pressure increases resulting in pulmonary venous congestion and pulmonary edema. If the right ventricle is involved the right atrial pressure increases resulting in systemic venous congestion and ascites. The increase in venous pressure leads to activation of renin-angiotensin-aldosterone system resulting in renal retention of sodium and water.

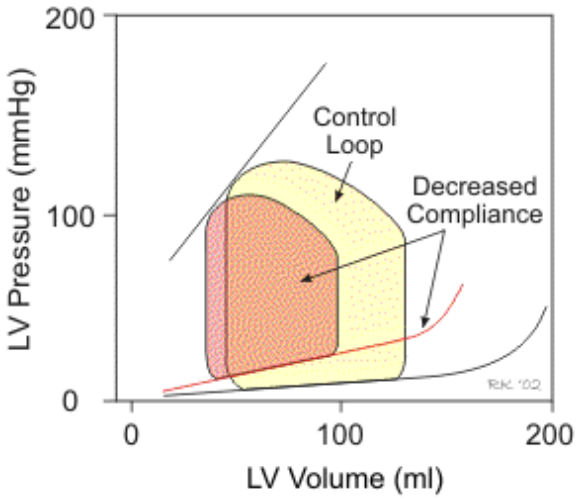
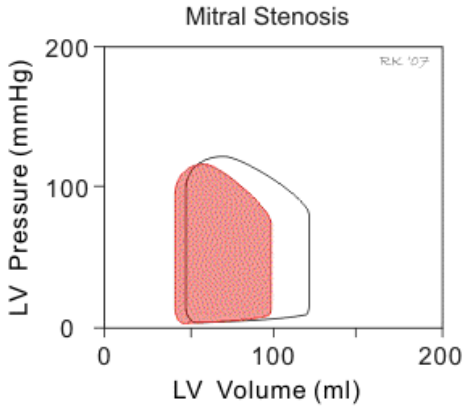


Fig. 15 *Effects of left ventricular diastolic dysfunction due to reduced compliance on left ventricular PV loop*

Diuretics are given to reduce blood volume in this situation, however too much of diuretics will be harmful, since some degree of elevated venous pressures are necessary to fill the less compliant ventricle.

Mitral stenosis:

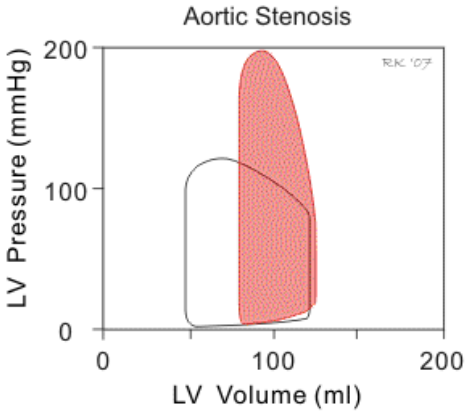
The changes observed in the PV loop with isolated mitral stenosis are shown in the shaded PV loop below. Normal PV loop (unshaded) is also shown.



There is a decrease in end diastolic volume due to reduced ventricular filling. This leads to decrease in stroke volume by Frank Starling mechanism and a fall in cardiac output and aortic pressure. Therefore there is a reduction in afterload, which enables the end systolic volume to decrease slightly but not enough to overcome the decrease in end diastolic volume. Since end diastolic volume decreases more than end systolic volume, the stroke volume decreases.

Aortic stenosis:

The following changes occur in the PV loop (shaded loop) in isolated aortic stenosis.

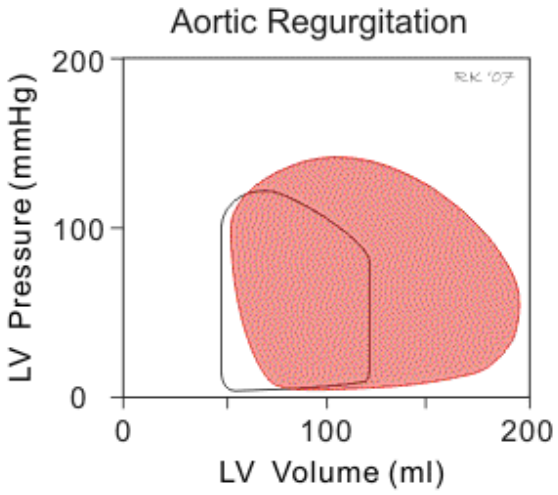


The left ventricular systolic pressure increases very much to overcome the outflow resistance caused by aortic stenosis leading to large pressure gradient across the aortic valve. This leads to an increase in ventricular afterload, a decrease in stroke volume and an increase in end systolic volume. The stroke volume decreases because the velocity of fiber shortening is decreased by the increased afterload (Force velocity relationship). Since end systolic volume is elevated, the residual volume is added to the incoming venous return increasing end diastolic volume also. This increases preload and activates Frank Starling mechanism to increase the force of contraction to help the ventricle overcome the increased outflow resistance.

In mild aortic stenosis this increase in preload and Frank Starling mechanism may be adequate to maintain normal stroke volume. But in moderate to severe aortic stenosis, the stroke volume may fall significantly because the end systolic volume increases substantially more than the increase in end diastolic volume. The stroke volume may further decrease if the ventricle develops systolic and diastolic dysfunction. Compensatory increase in end diastolic volume will be limited by ventricular hypertrophy that occurs in chronic increase in afterload. However this hypertrophy can lead to increase in end diastolic pressure resulting in pulmonary congestion.

Aortic regurgitation:

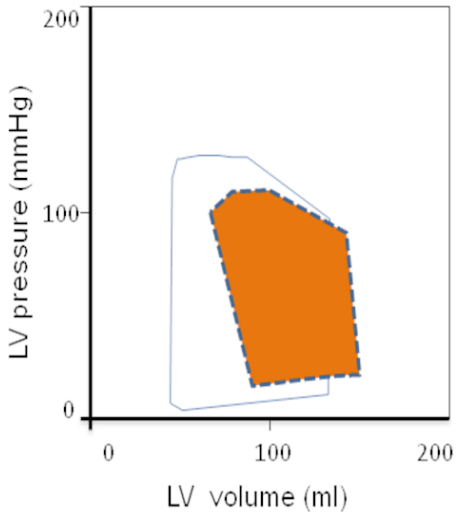
The following changes occur in PV loop (shaded loop) in chronic aortic regurgitation.



The aortic valve does not close completely at end diastole in aortic regurgitation. As the ventricle starts relaxing, blood flows from aorta back into ventricle even before mitral valve opens, hence filling occurs immediately. So there is no true phase of isovolumic relaxation. Once mitral valve opens, left ventricular filling occurs from left atrium also. The aortic regurgitation continues as aortic diastolic pressure is higher than ventricular diastolic pressure. This greatly increases end diastolic volume. When the ventricle starts contracting, blood is still entering the ventricle from the aorta because aortic pressure is higher than ventricular pressure, therefore there is no true isovolumic contraction because volume continues to increase. Once the ventricular pressure exceeds aortic diastolic pressure, ejection of blood starts. The increased enddiastolic volume (increased preload) increases force of contraction, ventricular peak systolic pressure and stroke volume which is shown by the width of PV loop. As long the ventricular inotropy is preserved there is only a slight increase in end systolic volume due to increased afterload. Once systolic dysfunction sets in, then the end systolic volume increases progressively and the peak systolic pressure and stroke volume fall.

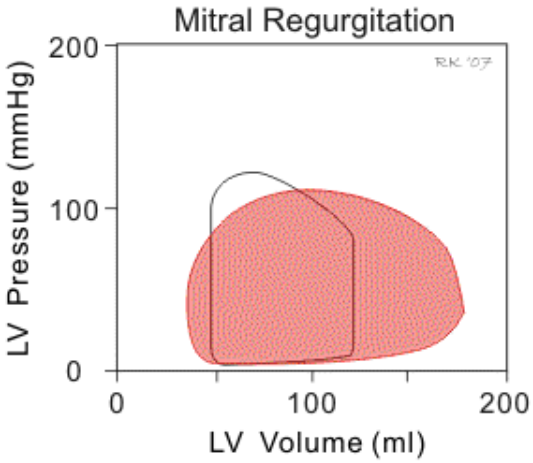
Acute aortic regurgitation:

In acute aortic regurgitation, the ventricular volume increases suddenly and the PV loop appears small. The end diastolic volume increases and end diastolic pressure increases abruptly. There is no true isovolumic relaxation.



Chronic mitral regurgitation:

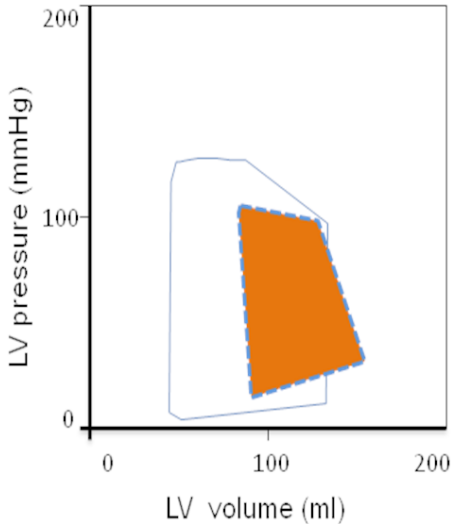
The following changes are noted in PV loop (shaded loop) in chronic mitral regurgitation.



In mitral regurgitation, during systole blood is ejected into aorta as well as into left atrium. This causes left atrial volume and pressure to increase during ventricular systole. There is no true isovolumic contraction phase because blood begins to flow immediately into left atrium as left ventricular pressure exceeds left atrial pressure, before aortic valve opens. Afterload on the ventricle is reduced so that end systolic volume can be smaller than normal. There is no true isovolumic relaxation also because when aortic valve closes and the ventricle begins to relax, the mitral valve is not completely closed so blood flows into left atrium as long as ventricular pressure is greater than left atrial pressure. During diastole, the elevated pressure within left atrium is transmitted to the ventricle during filling, so that the left ventricular end diastolic volume increases. The net effect of these changes is that the width of the PV loop is increased ie. stroke volume is increased, however this includes volume ejected into the aorta as well as volume ejected back into the left atrium. When systolic dysfunction sets in, the end systolic volume increases, the peak systolic pressure falls and forward stroke volume decreases. As a consequence the left atrial pressure progressively increases leading to pulmonary congestion and edema.

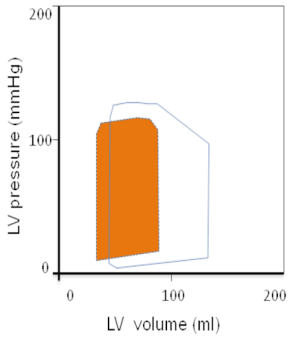
Acute mitral regurgitation:

In acute mitral regurgitation, the ventricular volume increases abruptly and PV loop appears small. There is no true isovolumic contraction. The left atrial pressure rapidly increases despite the compensatory increase in heart rate and initial increase in inotropy, leading to acute pulmonary edema.



Cardiac tamponade:

The pressure volume loop (shaded loop) appears unique, the preload is greatly decreased, the end systolic volume is also decreased leading to reduced stroke volume.



Further reading:

1. Cardiovascular physiology concepts, Richard E. Klabunde, Lippincott Williams & Wilkins.
2. Heart Physiology, From cell to circulation, Lionel H. Opie; III edition; Lippincott-Raven
3. Braunwald's Heart diseases, VIII edition.

CATHLAB HARDWARES

Dr. S. Vijayakumar

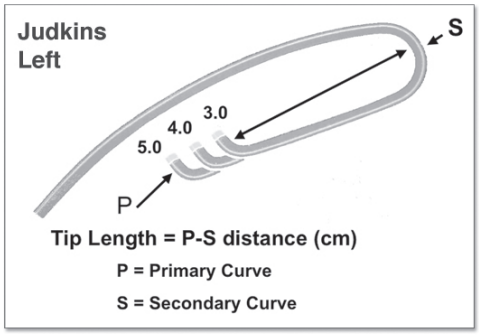
Catheters

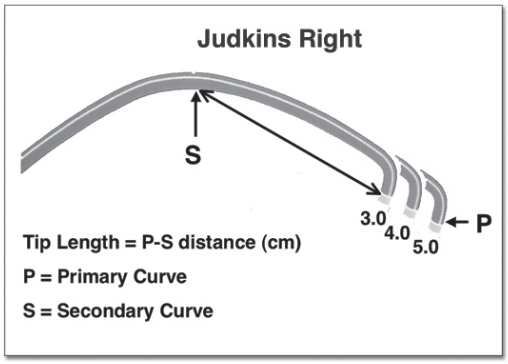
Ever since invasive cardiology started in the 1940 by Andre Cournand and Dickinson Richards, the catheter was the key in the lock. Certainly the has unlocked the door to expand the therapeutic interventions. Catheters are available in various shapes, sizes and lengths for different purposes. This chapter will review some of the commonly catheters in our practice.

Coronary Catheters :

- Coronary angiographic catheters include a body, a tip and various curves.
- Curves are classified as primary, secondary, and tertiary starting from the tip.
- The hub has a female Luer – Lok that allows attachment to a syringe or manifold, winged tips to facilitate catheter rotation, and labeling of the size of the catheter.
- Diagnostic catheters are sized by their external diameter.
- 4F – 8F sized catheters available. 6F catheters – most commonly used.
- Usually 100-110 cm in length.

Judkins' Catheters





Judkins left:

- 100 cm, polyurethane catheter with tapered tip and end hole

- The catheter bend size is measured between the steepest portions of the primary and secondary curves

- 90° Primary curve - allows the catheter to enter the coronary ostium

- 180° Secondary curve - provides backup support from the opposing aortic wall

Curve selection depends on size of the aortic root

Curve	Arm length
JL 4	4.2 cm
JL 5	5.2 cm
JL 6	6.2 cm

JL 4- Normal aortic roots, commonly used
JL 3.5 – Small aortic root
JL 5 &6- Larger aortic roots
Use- Selective left coronary angiography

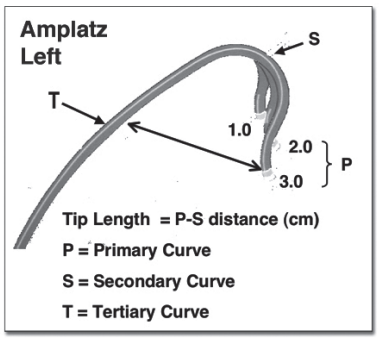
Judkins Right

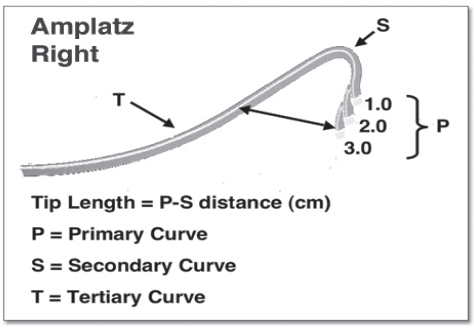
100 cm, polyurethane (PU), tapered tip, single end-hole braided steel catheter
90° Primary curve - allows the catheter to enter the coronary ostium
30° Secondary curve - provides backup support from the opposing aortic wall
Use- Selective right coronary angiography

Non-coronary uses

- To cross PDA from aortic side
- To encage collaterals
- To cross VSD from LV
- To cross the RVOT in PS
- Encaging BT shunt

Amplatz catheters:





Amplatz Left

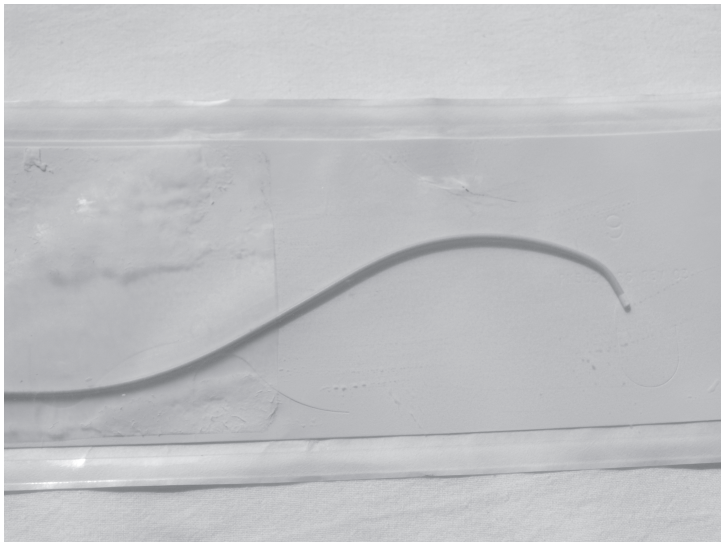
- polyurethane end-hole braided steel catheter.
- usually the second choice if a Judkins left catheter is unsuccessful
- secondary curve is shaped to fit the sinus of Valsalva.
- secondary curve comes in 4 sizes (L1-L4) all of which are larger than the Amplatz Rt. curve
- L1 and L2 – normal aortic roots
- L3 and L4 – dilated aortic roots

Amplatz Right:

- usually the second choice if a Judkins left catheter is unsuccessful
- Smaller than the Amplatz left curves.
- Only 3 secondary sizes (R1-R3)
- R1 – normal aortic roots
- R2 and R3 – dilated aortic roots

Right coronary bypass catheter:

- Similar to right Judkins catheter
- Primary curve (tip) is 110-120^0
- Prevents excessive caudal travel down the graft
- for grafts to right coronary tributaries



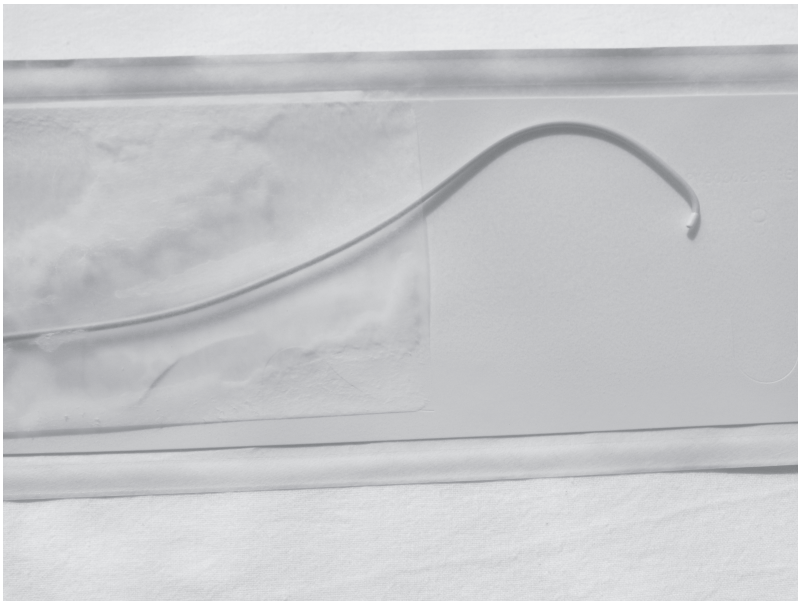
Left coronary bypass catheter:

- Similar to right Judkins catheter

- Primary curve – 90^0

- Secondary curve - 70^0

- For canulating left coronary grafts

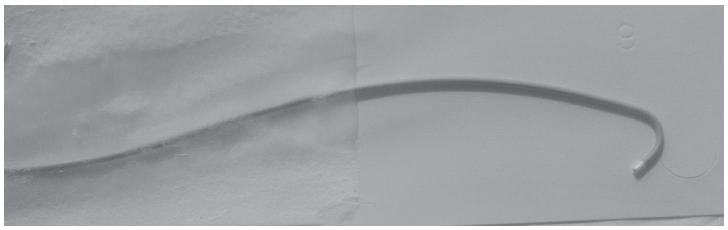


Internal mammary artery catheter

- Similar to right Judkins catheter

- Primary curve is - $<90^0$

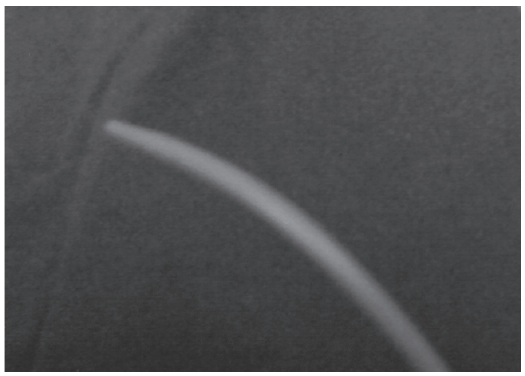
- Tip length – 1.5 – 2 cm instead of 1 cm
- For selective LIMA cannulation



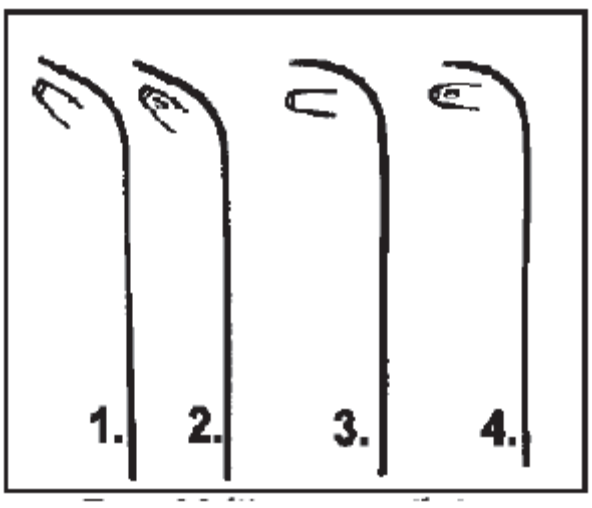
Catheters for Right heart study:

Cournand Catheter:

- End hole radiopaque woven Dacron catheter with an outer coating of polyurethane
- Gradual distal curve
- Tapered tip
- All –purpose right heart catheter



Multipurpose catheters



1. A-1 MP: Polyurethane (PU) with incorporated wire braid. The A bend is like a hockey stick with a straight tip. The 1 refers to one end-hole only.

2. A-2 MP (King): Same as A-1 except it has 2 sideholes and an end-hole.

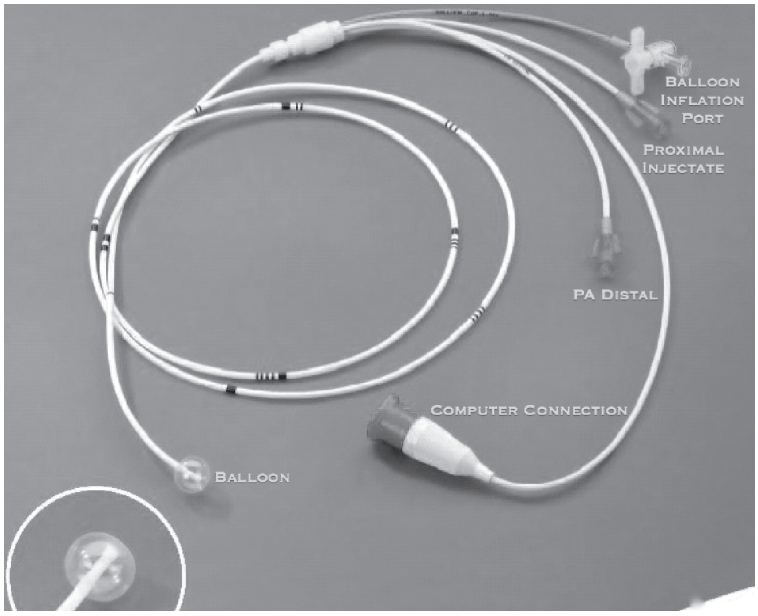
3. B-1 MP: The B bend is a gradual 90o curve up to the tip. The 1 refers to one end-hole only.

4. B-2 MP: The same as B-1 except has 2 sideholes and end-hole

Uses

- For crossing different lesions
- PDA & MAPCA coiling
- Angiography

Wedge pressure catheter (Swan-Ganz catheter)



Most widely used catheter for right heart hemodynamics

Balloon tipped – Float through the right side of the heart safely and easily

Available with two to five lumens

Two lumen catheter: distal port for pressure monitoring and one lumen for the balloon

Three lumen catheter: additional proximal port for RAP monitoring

Four lumen catheter: additional thermistor and computer connecting cable

Five lumen catheter: additional proximal port for fluid and drug infusion

Other use: Balloon occlusion catheter for large PDA

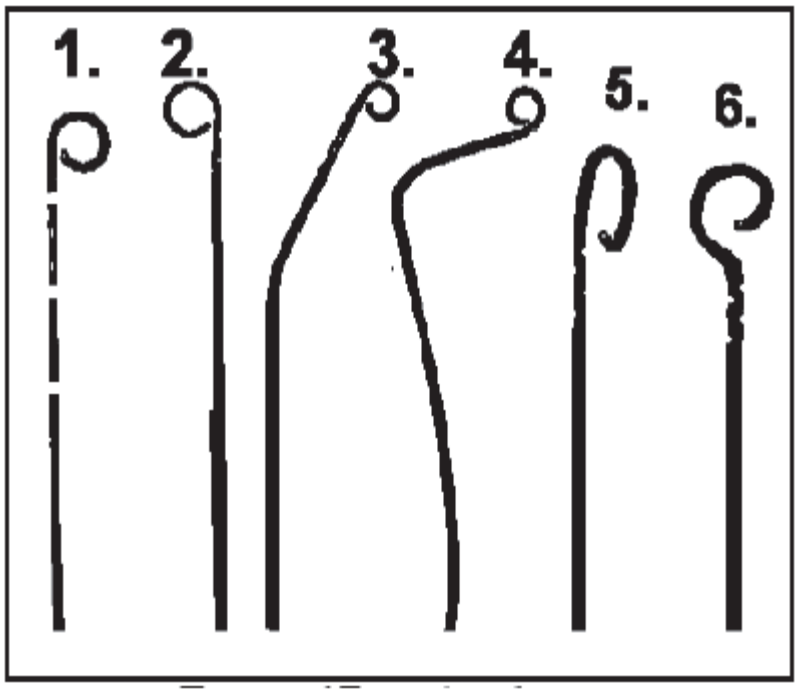
Angiographic catheters

Pigtail catheters

Made of polyurethane or polyethylene with a tapered tip, the terminal 5 cm of which is coiled back on itself in a tight loop (pigtail)
Open or closed end
4 to 12 non-laterally opposed side holes located in the terminal 5 cm of the catheter

Use:

Left ventricular and aortic angiography



1. **Quanticor (Cardiomarker pig):** This is a standard pigtail with radiopaque markers set 2 cm apart. These act to calibrate distance for quantitative angiography. Exact LV distances, volumes and stroke volume can then be calculated using these markers as a "ruler."
2. **Pigtail Angiographic:** The pigtail catheter is the most commonly used LV gram catheter. With up to 12 sideholes it evenly disperses the contrast within the LV.
3. **Van Tassel** angled pigtail: This is a Nylon core or woven steel high-flow pigtail. The 145-155 degree angle is 7 cm from the tip. This angle lifts the catheter off the inferior LV wall for a more centrally located LV gram. Also, it is useful for dilated aortas.
4. **Grollman PA:** is an angled pigtail catheter with the curve generally on the reverse side. It is designed for RV and selective PA angiography by the femoral approach.
5. **Elliptical or Oval:** Designed to pass small aortic valves or vessels with the curve intact

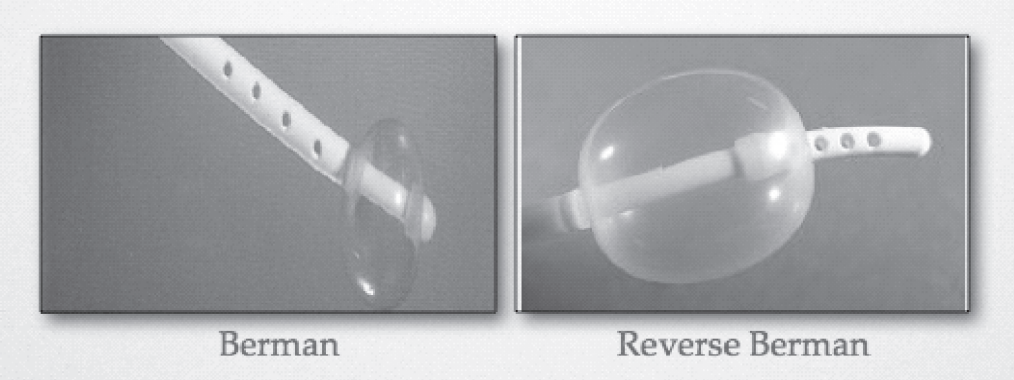
6. Tennis Racquet: The central shaft was designed to reduce the risk of vessel wall extravasation.

Berman Angiographic catheter

Large lumen – flow rates similar to the pigtail catheter
Balloon tipped – increases the catheter stability during angiography
No end hole – can't measure wedge pressure
Multiple side-holes proximal to the balloon

Uses:

Measurement of pulmonary artery pressure
Right ventriculography



Reverse Bermon catheter

Multiple side-holes distal to the balloon

Use: Pulmonary angiography

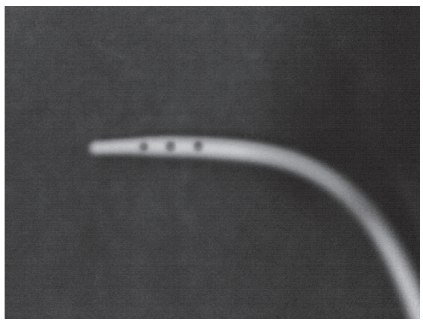
Gensini catheter:

Woven Dacron catheter with polyurethane coating
Taperded tip
Three pairs of laterally opposed oval side holes within 1.5 cm of its open tip

Use: Right and left heart angiographic studies

Disadvantages:

- Straight tip more arrhythmogenic
- Catheter recoil during injection at high flow rates
- Risk of intramyocardial injection and myocardial injection



NIH catheter:

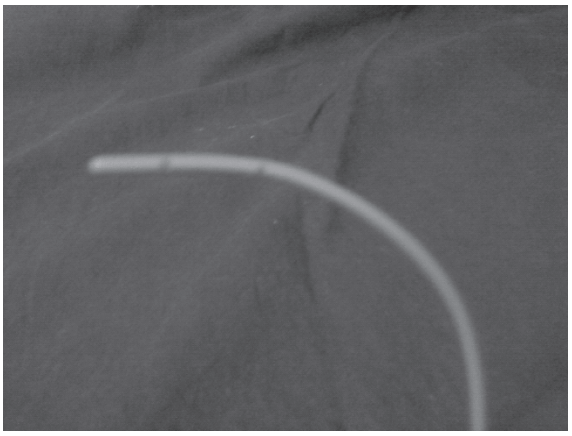
Constructed of woven dacron reinforced with nylon core – injection at high flow rates

No endhole

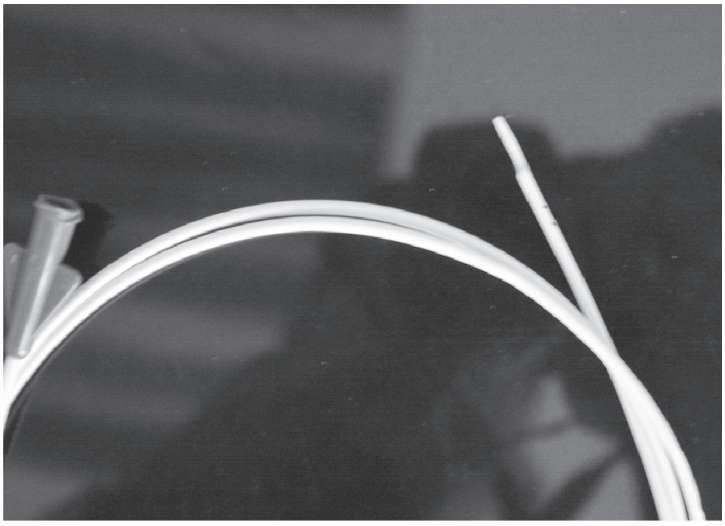
Six sideholes

Gentle curve

used for right and left heart angiography

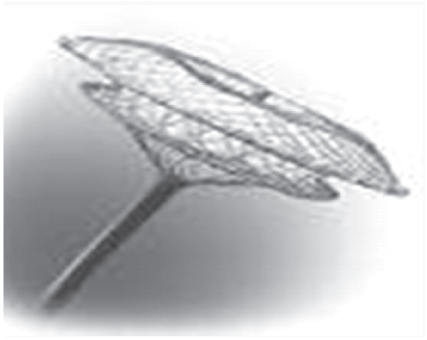


Multitrack catheter



- End hole and side hole catheter
- To record pressure while wire inside (Pull back gradient across valvular or arterial stenoses)
- Angiography while wire inside

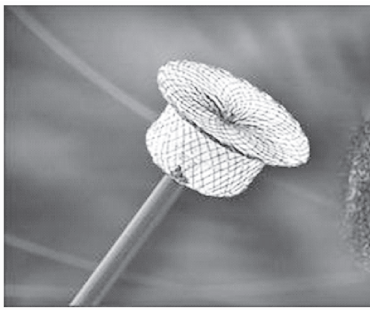
Amplatzer Septal occluder



Made up of Nitinol

Double disc, self centering device
Waist corresponds to the size of the ASD
LA disc 2 cm larger than RA disc
Available in size : 4 -40 mm
Used for closure of fossa ovalis ASD & Fontan fenestration

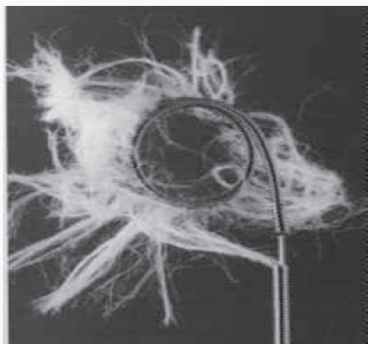
Amplatzer ductal occluder



Made up of nitinol

A retention skirt on the aortic side provides secure positioning in the ampulla of the ductus
Used for percutaneous closure of PDA, Coronary AV fistula, Pulmonary AV fistula and paravalvular leak (aortic and mitral)

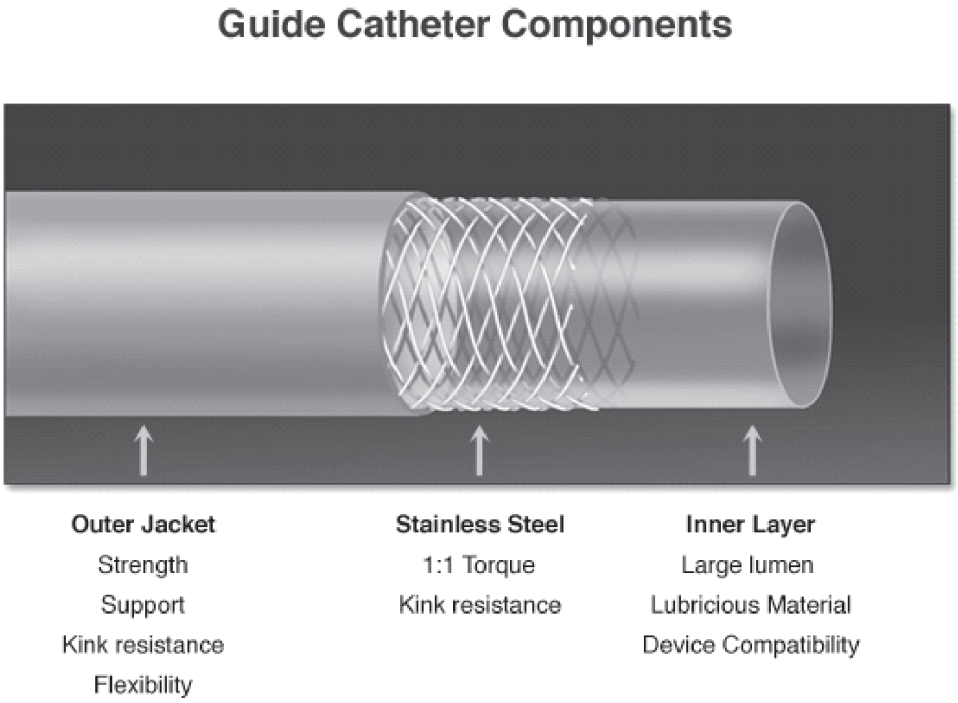
Gianturco coil:



Made up of stainless steal with polyester fabric
Used for percutaneous closure of PDA, Collaterals, AV fistula (coronary, pulmonary, systemic)

Guiding catheter:

Large lumen soft tipped catheters designed for coronary intervention



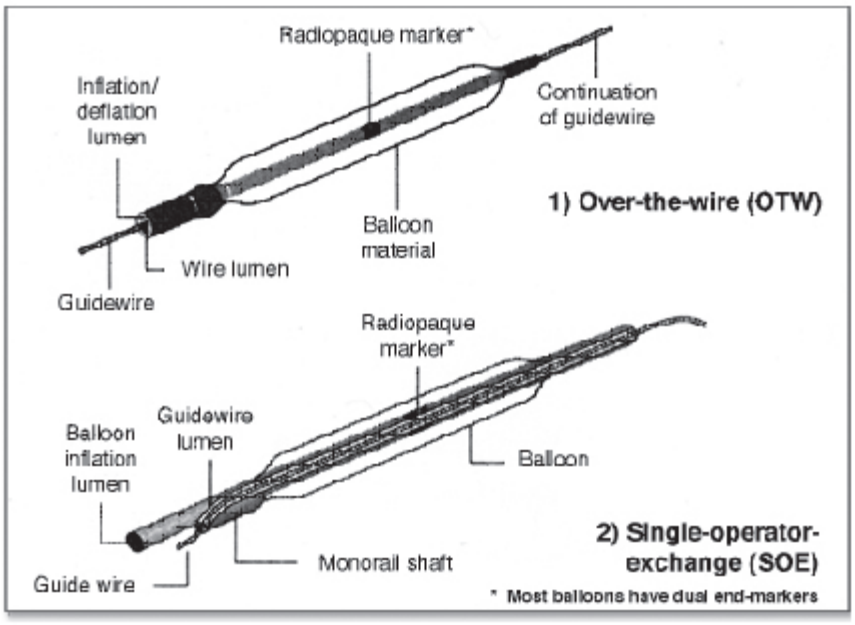
1. **Teflon:** This smooth slippery plastic forms the inner lumen or core of the catheter. It makes it easier to slide the balloon catheter through the lumen.
2. **Polyurethane polymer forms** the body of the catheter in which the wire or fiber braid is imbedded. Many new guiders include Nylon within the thermoplastic.
3. **Braid:** Stainless steel imbedded within the plastic jacket makes the catheter strong and torquable.
4. **Coating:** The slippery hypothrombogenic surface that coats many catheters may include Silicone, heparin, etc.

Range of Internal diameters of the guiding catheters

Range Among Manufacturers of Lumen Diameters for Coronary Guide Catheters

French Size	Minimum ID (inches)	Maximum ID (inches)
6	0.061	0.066
7	0.071	0.076
8	0.079	0.086
9	0.089	0.096
10	0.099	0.108

Balloon dilatation catheter



Contain two lumens for guide wire and for balloon inflation respectively and variable length balloon in the distal end

Balloon characteristics:

- **Compliant** : made of polyolefin polymer and with high pressure, the balloon becomes 20% bigger in diameter than the labeled balloon diameter
- **Noncompliant** : made of polyethylene terephtalate and balloon diameter does not change even at high pressures
- **Semi – compliant** : made of polyether blockamide, nylon, polyurethane elastomer and grows < 10% at high pressure

Two types of balloon catheter systems

Over the wire system:

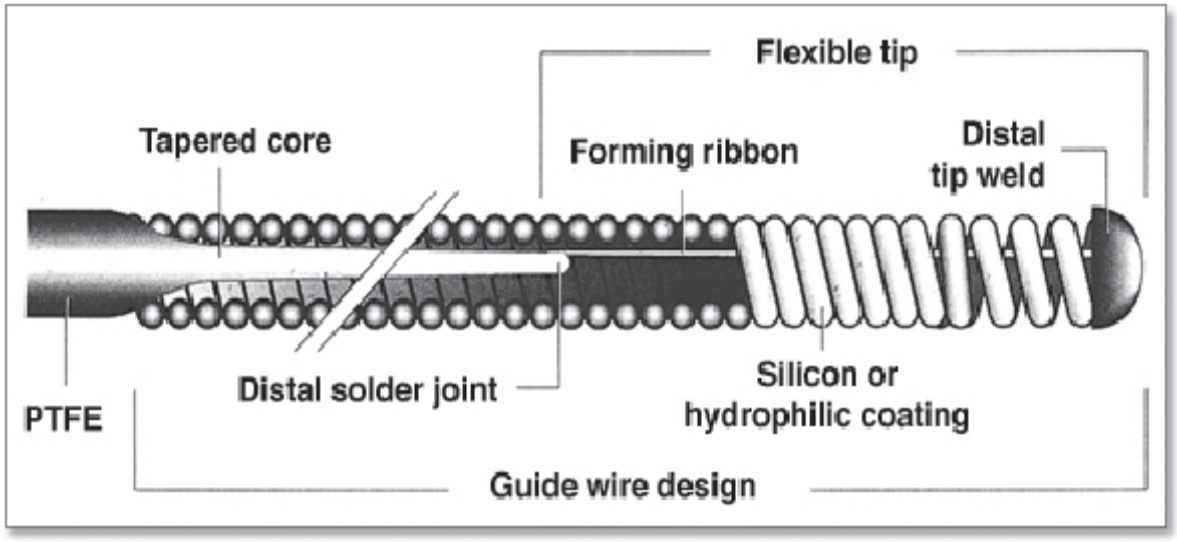
- Wire lumen through the entire length
- Better guide wire support
- Guide wire exchange possible
- Need for 300 mm guide wire, larger lumen catheter and two operators

Monorail (Rapid exchange) system:

- Shorter guide wire lumen (only 20 – 30 cm from the tip)
- Easier to use
- 180 cm guide wire, single operator, smaller lumen guiding
- Poor wire support with complex lesions, Wire exchange not possible

Guide wire

- used to advance the angioplasty equipment into the coronary artery.



Guide wire consists of three components:

- **Central core** : made of stainless steel or nitinol
  Tapers distally
  Provides support for device advancement
- **Flexible Tip**: Made up of platinum or tungsten coil
  Shapable into various and ratainable curves (Forming ribbon)
  Radiopaque
  Variable stiffness for varying lesion morphology
- **Coating** : Decreases friction and improves wire and device tracking
  Can be silicone, Teflon, polytetrafluoroethylene or hydrophilic Polymer

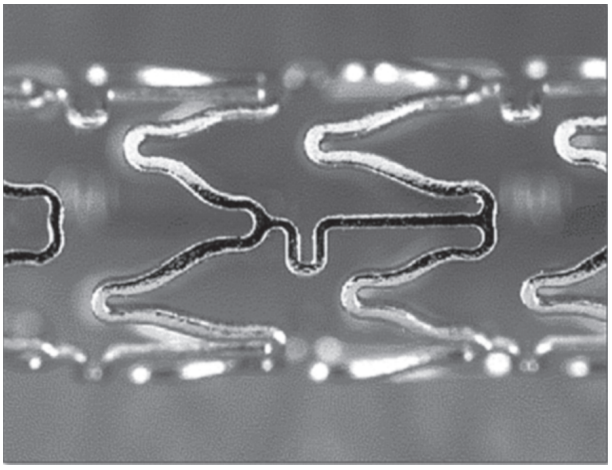
Length – standard (175 to 190 cm) ; exchange length (270 to 400 cm)

Diameter - 0.014"

Types:

1. All purpose – soft floppy tip, light to moderate support core – Simple to complex lesions- eg. Balance middle weight wire
2. Extra-support – High to extra-high support core wire – vessel straightening – eg. CholCE Extra Support wire
3. Stiff tipped/tapered tip – Added pushability – Severe stenoses, total occlusions – eg. Hi- torque floppy intermediate wire
4. Hydrophilic coated – Total occlusions, severe tortuosity - eg. Hi-torque Wisper
5. Firm tip and core – CTO , uncrossable lesions - eg. Asahi MiracleBros line wires

Stents :



- Stents are metallic scaffolds that are deployed within a diseased coronary segment to maintain wide luminal patency
- Commonly made of 316L stainless steel / tantalum or cobalt chromium alloys
- Strut thickness varies from 75 ìm to 150 ìm
- Open cell / closed cell design
- Length varies from 8 to 38 mm
- Diameters from 2.25 to 6.0mm
- Commonly used types:
  - bare metel stents: eg. Vision stent
  - Drug coated stents: eg. Taxus (paclitaxel)
    Cypher (Sirolimus)
    Endeavor (Zotorolimus)
    Xience V (Everolimus)
- Biodegradable stents: Igaki- Tamai stent (biodegradable polylactic acid polymer)

Balloon mitral valvotomy hardwares

Figure 1

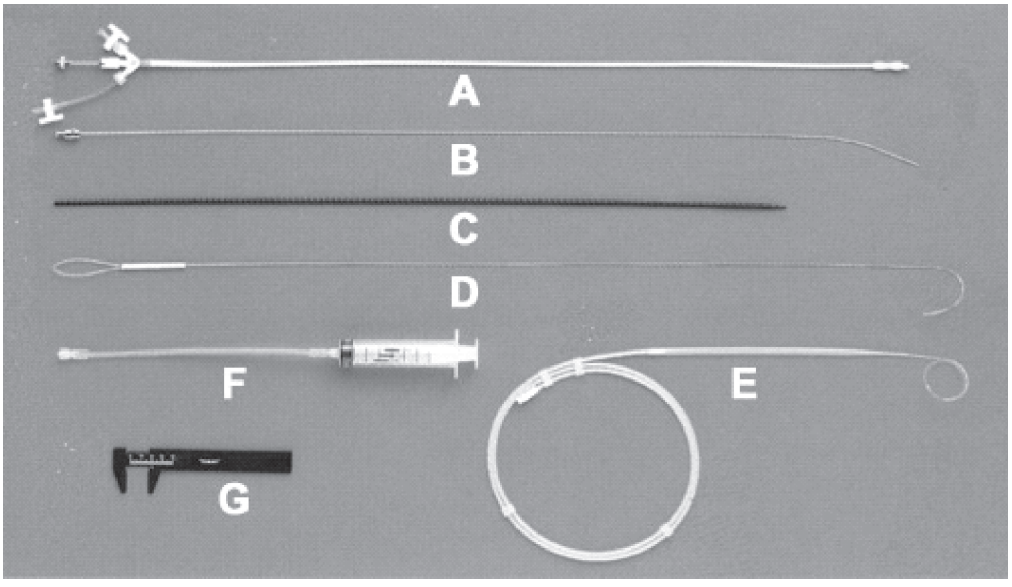
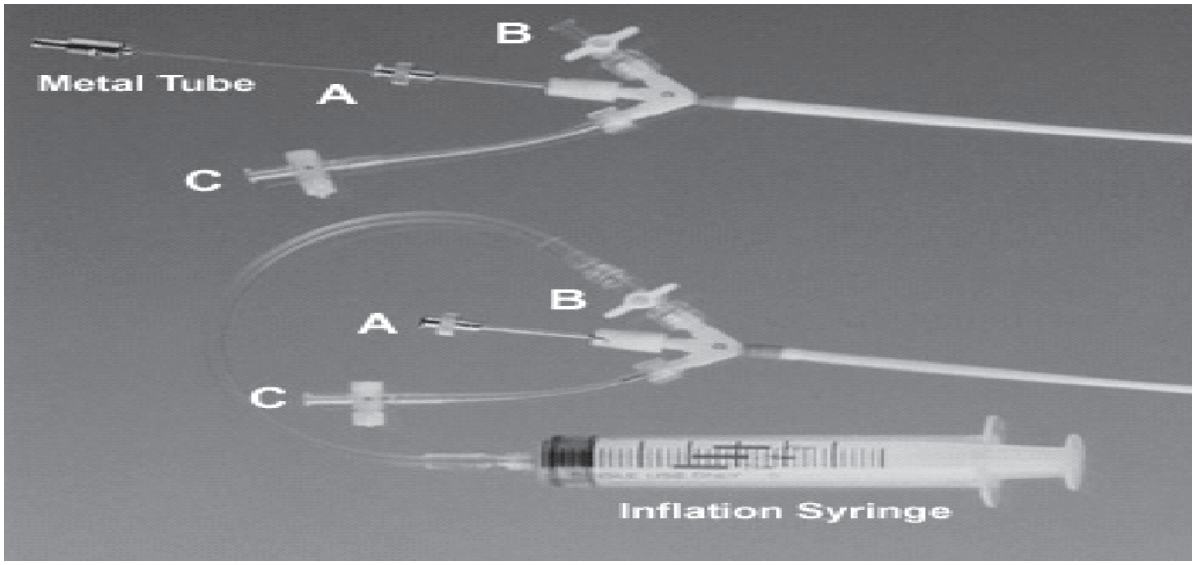


Figure 2 **Figure 3**



Inoue Balloon Catheter

The coaxial, double-lumen catheter with a 12F polyvinyl chloride tube shaft.

The inner lumen - permits pressure measurements, blood sampling or insertion of a metal tube, a guide wire, or a stylet (**Figure 2, A**).

The outer lumen - connects proximally with a two-way stopcock (**B**), and a vent (**C**).

The balloon - made of double layers of latex tubing, is mounted at the distal end of the catheter shaft.

There are **four types** of balloon catheters - each designated by a symbol followed by its maximally inflatable balloon diameter (in mm): PTMC-30, PTMC-28, PTMC-26, and PTMC-24.

The balloon can be transformed to various shapes from its natural form (**Figure 3**) to serve different functions. The balloon segment stiffens and slenderizes when the latex balloon is stretched by inserting a metal tube (**A**).

The synthetic mesh of the balloon is wound in such a way that the balloon changes shapes from its natural form (**B**) in three stages, depending on the extent of inflation. Initially, only the distal half inflates (**C**); then the proximal half inflates (**D**), with a constriction remaining in the middle. Finally, at full inflation, the constriction disappears (**E**) and the balloon assumes a more barrel-like shape with a maximal length of 45 mm.

The auxiliary instruments (Figure 1):

1) An 80-cm *18-gauge metal tube* (Figure 1, B)

The tube is inserted to lock with the inner lumen tube, thereby stiffening the catheter tip.
2) A 70-cm 14F ***polyethylene dilator*** (**Figure 1, C**)

It is used to dilate the opening of the femoral vein and the atrial septum.

3) A 80-cm 0.038-inch high-torque ***J-tipped spring wire stylet*** (**Figure 1, D**)

Helps in LV entry

4) A 180-cm **0.025-inch *stainless steel guide wire*** with coiled floppy tip (**Figure 1, E**)

To guide the balloon catheter to the left atrium.

5) A **30-cc *plastic syringe and a connecting tube*** (**Figure 1, F**)
The extent of balloon inflation is controlled by adjusting the volume of diluted contrast material in the syringe, which is injected manually into the catheter through a two-way stopcock

6) A ***ruler*** (**Figure 1, G**)
used to measure the diameter of inflated balloons in pretestings before insertion of the catheter into the patient.

BASIC ELECTROPHYSIOLOGY STUDY

Dr. Ulhas M. Pandurangi

NORMAL INTRACARDIAC ELECTROPHYSIOLGY INTERVALS

AH	60-120 ms
HV	35 – 55 ms
CSNRT	< 550 ms
SACT	<100 TO <170 ms
AERP	150 TO 360 ms
AVNERP	230 TO 425 ms
RVERP	170 TO 290 ms

CYCLE LENGTH (ms) = 60,000 / HEART RATE (BEATS PER MINUTE)

HEART RATE (BPM) = 60,000 / CYCLE LENGTH (ms)

NORMAL SNRT = 1500 ms

THIS IS MEASURED BY PACING ATRIUM FOR 30 SECS AND LOOKING AT THE INTERVAL WHEN SINUS NODE RECOVERS.

AV NODAL REENTRANT TACHYCARDIA:

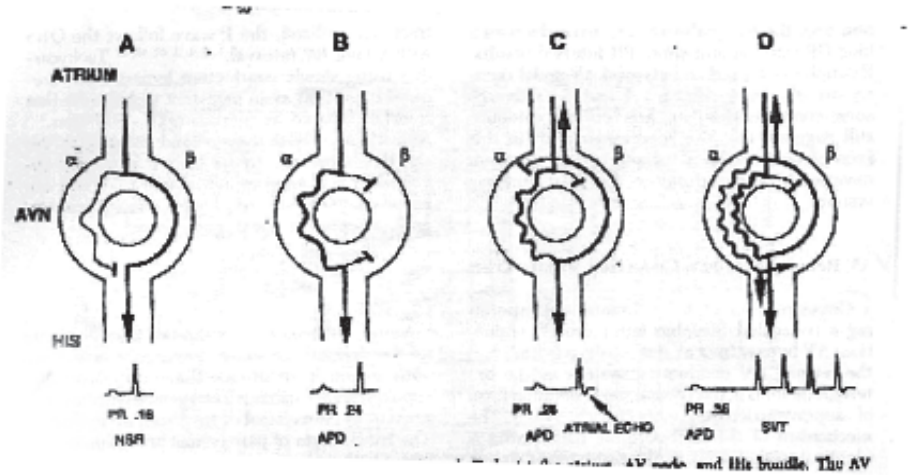


Figure 1. Mechanism of typical AV nodal reentry. A through D depict the atrium, AV node, and His bundle. The AV node is schematically divided with a slowly conducting alpha pathway and a rapidly conducting beta pathway. The beta pathway has a longer refractory period than the alpha-pathway. During sinus rhythm (NSR) the impulse preferentially conducts over the beta pathway. In panels B to D, progressively premature atrial premature depolarization (APD) block in the beta pathway and go down the alpha pathway, producing long PR. When the PR reaches 0.28seconds (C), a single atrial echo results. When conduction down the alpha pathway is sufficiently slow, producing a PR of 0.36second, sustained AV nodal reentry results.

- Most common cause of regular narrow QRS tachyarrhythmia in adults.
- The heart rate usually is between 120 to 250 bpm and is typically quite regular.
- In the common type of AVNRT (called Typical AVNRT, comprising more than 90% of all AVNRTs), the P wave is obscured by the QRS complex or may be present in its terminal portion, mimicking terminal delay ('pseudo r' or 'pseudo s' wave) (Figure 2). Very rarely the P wave precedes the QRS by a rather very short PR interval of <80 ms and may distort initial portion of the QRS.
- The P wave in the uncommon form (called Atypical AVNRT) occurs late (i.e., in or after the T wave), is relatively easy to recognize and produces a pattern of long RP and short PR. This form of tachycardia cannot be differentiated from PJRT or AT on the basis of ECG alone.
- AVNRT reentry circuit is localized to peri AV nodal region. The fast pathway of the re-entry circuit runs superiorly and anteriorly in the triangle of Koch, whereas the slow pathway runs inferiorly and posteriorly close to the coronary sinus ostium.
- In typical AVNRT the atrial activation spreads from fast pathway so that atria and ventricles are simultaneously depolarized and hence P and QRS occur near simultaneously. In atypical AVNRT atrial activation spreads from slow pathway so that atria and ventricles are depolarized at different times separated usually by more than 180ms. Thus in atypical AVNRT, P waves are separated from QRS such that RP interval is longer than PR interval.
- Very rarely AVNRT can be associated 2:1 A-V or V-A block implying that the tachycardia is not dependent on AV node.

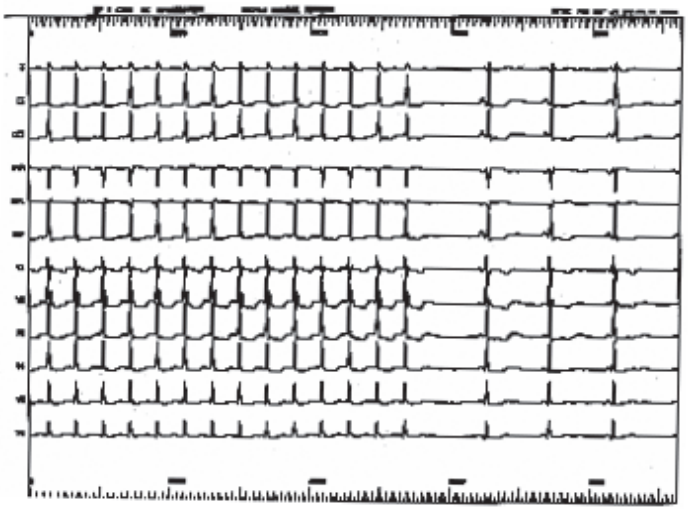


Figure 2. *AVNRT. Note characteristic pseudo 'r' in V1 during tachycardia and its absence during sinus rhythm.*

ATRIO-VENTRICULAR REENTRANT TACHYCARDIA:

- Second commonest cause regular narrow QRS tachyarrhythmia in adults.
- The commonest cause of tachyarrhythmia in children.
- In AVRT, an extranodal accessory pathway connects the atrium and ventricle. The reentry circuit involves atrium, AV node and the accessory pathway. When the reentry circuit involves accessory pathway in the retrograde direction during tachycardia the tachycardia is defined as orthodromic tachycardia. The tachycardia involving the reversal of the circuit is defined as antidromic tachycardia. AVRT when not further described is considered orthodromic AVRT

and usually it is a narrow QRS regular tachycardia. Accessory pathways may exhibit antegrade and retrograde conduction, or either only antegrade (rare) or only retrograde (concealed pathways) conduction. Most often the accessory pathways are concealed.

- More than 90% of AVRT are narrow irrespective of whether sinus rhythm ECG shows ventricular pre-excitation (Delta wave) due to the accessory pathway or not.
- The reentry direction during narrow QRS AVRT (described as orthodromic tachycardia) is along the AV-node, His bundle and ventricles antegradely and along the accessory pathway and atrium retrogradely. The reverse of the reentry direction results in wide QRS AVRT (described as antidromic tachycardia). In the presence of underlying Bundle branch block orthodromic tachycardia may also result in wide QRS.
- In AVRT, there is typically a short RP interval with the timing and morphology of the P-wave dependent on the site and conduction velocity of the accessory pathway.
- During AVRT atrium and ventricles are activated at different times separated at least by 90-100ms usually. Atrial depolarization coincides with ST segment and hence distorts or depresses the segment. Greater is the depression of ST segment more certain is the diagnosis of AVRT. The ST depression irrespective of severity should not be considered as a sign of ischemia. Conversely during AVRT the ST segment elevation in aVR can also be explained by the same mechanism.
- ST segment depression >3 mm and its presence in >5 leads, especially affecting precordial and limb leads together are strongly indicative of AVRT (Figure 3).

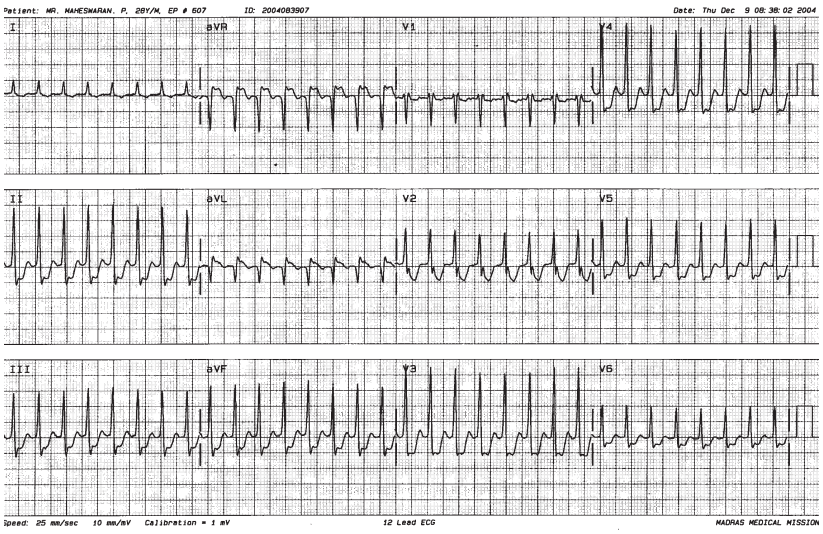


Figure 3. *Note significant ST depression in most of the leads and ST elevation in aVR.*

- Several patterns of repolarization changes (ST changes) have been shown to be specific for accessory pathway location, for example, ST-segment depression in lead V3-V6 for left lateral accessory pathway, ST-segment depression and negative T wave in inferior leads for posteroseptal or posterior accessory pathway and a negative or notched T wave in lead V2 for anteroseptal pathway.
- In AVRT, the ventricle is an obligate part of the circuit, and thus AV block cannot occur.

BIRD'S EYE VIEW OF PEDIATRIC CARDIAC SURGERY

Dr. Roy Varghese

I. Introduction:

While it would be extremely difficult to do justice to a specialty by attempting to summarize the entire subject into a talk that lasts only half an hour, it would be equally daunting to acknowledge the contributions made by the pioneers of the specialty.

The field of pediatric cardiac surgery is what it is today largely due to the pioneering efforts of these two surgeons who defied odds to treat congenital anomalies of the heart; an organ that was considered irreparable if afflicted by anomalies.

Dr Blalock and Dr Lillehei performed these operations at a time when it was considered "foolish" to perform operations on the heart and it would also mean "they would lose the respect of their colleagues". They and many others following them have over the years brought to us an understanding of the hemodynamics and surgical repair techniques that we now use routinely to correct congenital anomalies of the heart.

II. Classification of lesions:

It would be simplifying the topic to classify congenital anomalies that affect the heart as:

a. Shunt lesions

b. Left ventricular outflow tract obstructions

c. Right ventricular outflow tract obstructions

d. Single ventricle lesions

e. Other lesions

III. Normal anatomy:

Soon after the fetal circulation is abolished in favor of the adult circulation, the right and left sides of the heart pump as a circulation in "series" (reverting from the earlier circulation in "parallel") thereby dealing with the de-oxygenated and oxygenated blood for the right and left sides of the heart respectively (rather than a combined – left and right - ventricular output as was evident in the fetus).

Immediately following birth, with the expansion of the lungs, the increase in partial pressure of oxygen in the blood and the increase in left atrial pressures, the circulatory changes associated with the "Transitional Circulation" are established. These changes obliterate the several intercommunications (shunts) between the two circulations and lead to the establishment of the "Circulation in series".

IV. Patent ductus arteriosus:

Persistent ductus arteriosus (PDA) is unique in its history and scope:

It was a serious lapse on my part to omit the contribution made by perhaps the very first surgeon who paved the way for the specialty to develop. Dr Robert E. Gross was 33 years old in 1938 when he ligated the PDA in a 7 year old girl at the Boston Children's Hospital and this landmark operation ushered in the modern era of congenital heart surgery.

PDA was also the first congenital cardiac anomaly to be treated by transcatheter technique.

Looking into the future, PDA may well be the first congenital cardiac anomaly to be addressed through genetic modification.

Being a normal fetal structure, the PDA develops from the left 6th aortic arch. Functional closure occurs in the first few hours of birth though anatomic closure is complete by approximately six weeks.

V. Though in the fetal circulation, the ductus carries blood from the pulmonary artery to the aorta, after birth, with the fall in pulmonary vascular resistance, the PDA shunts blood into the pulmonary arteries resulting in increased pulmonary blood flow. This would result in raised pulmonary vascular resistance over a varying period of time depending on the magnitude of the shunt.

VI. Treatment consists of interruption of the ductus arteriosus. A variety of treatment options are available:

a. Pharmacological: Indomethacin (prostaglandin synthetase inhibitor) in the dose 0.1- 0.2 mg/ Kg intravenously every 12 – 24 hours. A schedule of three doses is recommended.

b. Surgical ligation / division & suturing

c. Transcatheter interruption

d. VATS

VII. Atrial septal defects are shunts across the inter atrial septum and result from varying degrees of failure of fusion of the septum secundum, septum primum and the endocardial cushions. The main anatomic types are:

a. Ostium secundum atrial septal defect

b. Sinus venosus atrial septal defect

c. Ostium primum atrial septal defect

d. Common atrium

e. Unroofed coronary sinus

f. Patent foramen ovale

These may co- exist with other congenital cardiac anomalies – simple & complex.

VIII. The magnitude of the shunt depends on the size of the interatrial communication and the compliance of the ventricles. A left to right shunt of varying magnitude therefore occurs resulting in dilatation of the right ventricle and enlargement of the pulmonary arteries.

IX. Though obstructive pulmonary vascular disease is uncommon, closure is recommended between 2 – 5 years.

Surgical closure involves cardio pulmonary bypass and closure. The approach for these has evolved over the years and many units around the world advocate cosmetic incisions resulting in partial sternotomy and right thoracotomy. Anomalously draining pulmonary veins and anomalies of atrio ventricular valves have also to be addressed at the time of the repair.

Trans catheter techniques are also widely practiced.

X. Ventricular septal defects are shunts across the inter ventricular septum and constitute one of the commonest congenital cardiac anomalies that require intervention in early life.

A ventricular septal defect occurs due to a failure of closure at the interventricular muscular partition, endocardial cushions and the bulbar ridges that separate the great vessels.

The interventricular communication may be located at almost any location in the septum and are therefore classified as:

a. Type I: Supra cristal/ conal/ infundibular/ sub arterial

b. Type II: Para/perimembranous

c. Type III: Atrioventricular canal type/ inlet septal

d. Type IV: Muscular

The size of the VSD and the pulmonary vascular resistance are the major determinants of the magnitude of the shunt. The shunt is left – right and results in increase in pulmonary vascular resistance which if left uncorrected could lead on to obstructive pulmonary vascular disease. Repair is therefore recommended by about one year of life if not earlier.

XI. Surgical repair entails cardio pulmonary bypass and a patch closure in the majority of cases. The approach depends on the location of the VSD and is right atrial in the majority of cases. Other approaches include trans pulmonary, trans aortic and right ventricular.

Risks are negligible in the modern era of cardiac surgery. These risks include damage to the conduction system of the heart and damage to the aortic valve.

Bacterial endocarditis and aortic regurgitation are complications of un repaired VSD's as is the incidence of Eisenmenger syndrome.

XII. Atrio ventricular canal defects are also called endocardial cushion defects and result from failure of development of the endocardial cushions at the inlet to the ventricles.

The complete atrio ventricular canal (CAVC) defect consists of a large ventricular septal defect below the level of the atrioventricular valves, an atrial septal defect above the level of the atrioventricular valves and a common atrio ventricular valve. A partial Atrioventricular canal defect results in having only the atrial component of the anomaly together with a cleft in the mitral valve resulting in varying degrees of mitral regurgitation.

Hemodynamics and repair depends on the balance between the two ventricles.

Left to right shunt at the atrial and ventricular levels together with regurgitation of the atrio ventricular valve results in excessive pulmonary blood flow, progressive cardiomegaly and significant pulmonary hypertension. These patients have a particularly rapid progression of pulmonary vascular disease. Down's syndrome is associated commonly and appears to accelerate the development of pulmonary hypertension.

XIII. Repair consists of closing the inter atrial and inter ventricular communications with a single or double patch and repair of the common atrio ventricular valve thereby attaching the valves to the patches. The resultant repair makes it possible to have two separate atrio ventricular valves that are non regurgitant.

XIV. Truncus arteriosus is a congenital malformation in which a single arterial trunk arises from the heart, over rides the inter ventricular septum and supplies the systemic, pulmonary and coronary arterial circulations.

XV. Embryologically, this is the result of a failure of the embryonic truncus to separate and spiral into an anterior pulmonary artery and a posterior aorta. Anatomically there is varying degrees of over riding of this truncus arteriosus. The Collett and Edwards classification is the one that is commonly followed and it relies on the origin of the pulmonary artery from the truncus.

The resultant large left to right shunt depends on the pulmonary vascular resistance. Truncal valve regurgitation compounds the problem.

Untreated patients have a 65% 6 month mortality and 75% one year mortality.

XVI. Surgical repair is indicated early and involves separation of the pulmonary arteries from the common arterial trunk, closure of the VSD to direct the left ventricular blood to the aorta (which was the original truncus) and establishment of right ventricular to pulmonary arterial continuity with a valved conduit (homograft or bovine jugular vein). Additional procedures like truncal valve repair for regurgitation may be necessary.

XVII. Tetralogy of Fallot is unique among the intra cardiac anomalies in that not only was it among the first of the major anomalies to be palliated by the Blalock-Taussig shunt but it was also among the first to be undergo complete correction through the pioneering efforts of Drs. Lillehei and Varco at the University of Minnesota. It therefore occupies a most unique place among the anomalies. The repair techniques employed in those years are still followed largely unaltered and the improvements in the results are mainly due to advancements in perfusion technology and ancillary care of these children.

The basic components of the "tetrad" consist of:

a. The large sub aortic VSD

b. RVOT obstruction (infundibular stenosis)

c. Dextroposed (overriding) aorta

d. Right ventricular hypertrophy

Each of these components however results from one basic abnormality: anterior and leftward deviation of the infundibular (conal) septum.

XVIII. The clinical picture depends on the delicate balance in maintaining the pulmonary circulation in the presence of the RVOT obstruction and whenever this requirement is not met, the child develops the typical cyanotic "spells". Squatting episodes and these troublesome spells demand urgent intervention; otherwise the consequences may well be life threatening

XIX. Surgical repair focuses on the closure of the VSD and opening up of the RVOT to maintain gradient free ante grade perfusion of the pulmonary arteries.

The trans atrial approach to address both these objectives is usually the norm nowadays in most units. It is particularly important during the repair to respect the tricuspid valve apparatus in these children because of the incompetence that may invariably result in some form or the other to the valve mechanism of the RVOT (pulmonary insufficiency).

XX. Transposition of great arteries is a congenital cardiac malformation wherein the anatomic relationships of the great arteries are reversed. Unlike the normal, the aorta arises anterior to the pulmonary artery and from the right ventricle while the pulmonary artery lies posterior to the aorta and arising from the left ventricle; ventriculo arterial discordance in the presence of atrio ventricular concordance.

Subdivided into three common variants; those with intact ventricular septum (50%), those with a co-existent ventricular septal defect (25%) and those with pulmonary stenosis (LVOTO) (25%).

XXI. This anatomic arrangement results in a circulation in "parallel" unlike the circulation in "series" that the normal heart is designed for. Survival beyond infancy is largely dependent on the degree of mixing these babies desperately require.

The embryogenesis of this anomaly has been the subject of speculation;

a. The straight truncoconal septum hypothesis incriminating the abnormal septation of the aorta and the pulmonary artery.

b. The abnormal fibrous skeleton hypothesis in which a pulmonary artery – mitral fibrous continuity occurs instead of the normal aorto-mitral continuity.

XXII. Surgical correction of TGA evolved since the early 1950's. The Blalock- Hanlon atrial septectomy was probably the first; the principle behind the procedure being to promote mixing at the atrial level. This has largely been replaced by balloon atrial septostomy or open septectomy.

The Mustard and Senning operations were devised to correct the anomaly at the atrial level viz: direct the right atrial blood to the left ventricle and the left atrial blood to the right ventricle and there by to the aorta. These procedures though successful and having good short term results, were not anatomic repairs and therefore were not associated with long term survival benefit for the patients.

The arterial switch operation was devised with these in mind and when performed in 1972 for the first time held a lot of promise as the procedure of choice. Despite a slow learning curve and initially poor results, the procedure has now come to be the gold standard for the correction of transposition of great arteries.

XXIII. The procedure involves coronary translocation to the neo aortic root and reconstruction of the neo pulmonary artery root using a "pantaloon patch" of autologous pericardium. The ductus arteriosus requires division at the beginning of the procedure.

XXIV. The "French Maneuver" facilitates completion of the neo aortic and neo pulmonary artery reconstruction.

XXV. At the completion of the operation the circulation is like a normal circulation in series – anatomic correction.

XXVI. The arterial switch operation besides providing an excellent repair for these neonates, has also over the years contributed to improvement in the care of the neonate and now many units worldwide have excellent results with this operation.

XXVII. Hearts with the single ventricle physiology mainly fall into one of these broad subsets.

Further, they are classified on the basis of adequacy of pulmonary blood into those with increased blood flow and those in whom the blood flow is decreased. The former would present with PAH and the latter with cyanosis.

These anomalies are also classified as those with normally related great arteries and those with transposed/ malposed great arteries.

Each of these sub classifications would influence the management.

XXVIII. Single ventricle with increased pulmonary blood flow requires to be palliated initially by restriction of their blood flow and pulmonary artery banding is done in these patients to prepare them for an eventual single ventricle repair.

XXIX. The hypoplastic left heart syndrome is a separate entity within the single ventricle spectrum of anomalies. Besides being a single ventricle as per the definition, HLHS further has hypoplasia of all left sided structures culminating in hypoplasia of the ascending aorta and the arch with severe coarctation of the aorta.

XXX. These would need to undergo the Norwood operation as the initial palliative operation wherein the ascending aorta and the arch are reconstructed using the pulmonary root. The pulmonary circulation is then maintained by means of a systemic to pulmonary artery shunt or a right ventricle to pulmonary artery non valved conduit. They would then qualify for the same pathway as the other single ventricle anomalies.

XXXI. The single ventricle pathway culminates in the Fontan operation. Many units worldwide would favor staging the Fontan operation and the first stage in these situations is the Bidirectional Cavo Pulmonary (Glenn) Shunt. The superior vena cava is disconnected and anastomosed to

right pulmonary artery. The pulmonary circulation is therefore maintained through a passive venous shunt that is dependent on the superior vena caval pressure as the driving force.

XXXII. These babies are necessarily nursed in the semi reclining position to promote superior caval flow into the pulmonary arteries. It is necessary that pulmonary pressures be low to facilitate this flow.

XXXIII. The Fontan operation is the completion of this sequence by conveying the inferior vena caval blood to pulmonary arteries as well. The procedure followed in many units worldwide now is by using an extra cardiac conduit for this purpose.

XXXIV. A "Bird's eye view" of this specialty would not be complete without the advances made in the field in the recent past.

Faced with peer competition and advances made in trans catheter interventions, minimally invasive procedures have evolved.

Hybrid procedures have been the result of close co- operation with the cardiological colleagues and have been instrumental in expanding the horizons of the interventionists and reducing the surgical morbidity associated with many procedures.

Pharmacological adjuncts have contributed to expand the surgical armamentarium and the scope of surgical procedures in many in operable or otherwise high risk operative procedures.

XXXV. Minimally invasive procedures have in mind the basic concept of cosmesis. The idea has been to provide the patient with a cosmetically acceptable scar while at the same time providing the same degree of safety and long term benefits as the conventional surgical procedure. This has been a direct result of the improvement in surgical expertise acquired over the years.

Awareness among patients has also been on the rise and more patients are actually opting for and in many instances demanding cosmetic incisions.

Peer pressure among colleagues has also contributed to the surge of cosmetically superior incisions.

XXXVI. The "simpler" operations can now be performed through these cosmetic/minimally invasive incisions.

XXXVII. Cosmetic incisions fall into three main types:

(i). Thoracotomy approach which may be the anterior or the posterior incisions

(ii). Sternotomy which may also be the upper or the lower sternotomy

(iii). Video Assisted Thoracoscopic Surgery

XXXVIII. The anterior thoracotomy incision has the advantages of being sub mammary in the female patient in whom breast development is complete. It is easy to approach the right atrium through this incision. The scar will also have good healing with less tendency for keloid formation unlike the sternotomy incisions.

XXXIX. The disadvantages are that in pre pubertal girls it may interfere with the development of the breast and may result in altered sensation around the areola.

XL. The posterior thoracotomy approach shares all the advantages of the anterior thoracotomy with the added benefit of not interfering with development of the breast in the future.

XLI. Theoretical disadvantages do exist though these are not significant in any large series that have experience in these operations.

XLII. Mini sternotomy likewise is also a well-suited approach in appropriately selected patients. Poor access to the persistent left superior vena cava, PDA and a partially anomalously draining pulmonary vein may limit its use in these circumstances.

XLIII. Taking advantage of the expertise acquired by the interventional cardiologists in trans catheter interventions, multiple ventricular septal defects may be closed by means of hybrid procedures wherein the surgeon and the cardiologist would occlude defects in areas of the heart that are difficult to access surgically like the apical septum.

XLIV. Pharmacological adjuncts that benefit the patient by expanding the scope of surgical interventions are noteworthy of mention in this discussion. Nitric oxide, the phosphodiesterase inhibitors (milrinone) and sildenafil are "magic" drugs that selectively act on the pulmonary vasculature and help the right ventricle. Their benefits range from reducing the pulmonary hypertension in the large ventricular septal defects to aiding the transplanted heart in the immediate postoperative period.

I sincerely hope this short presentation has been of benefit to you and I have done justice in summarizing a topic that has taken approximately six decades to develop and is still in its infancy.

SURGERY IN CYANOTIC CONGENITAL HEART DISEASE

Dr. Krishna Manohar

Cyanotic Heart Surgery

Introduction

Diagnosis of a cyanotic heart in a child is always an ominous sign and that unfortunate child will not have a normal life span.Uncorrected,sooner or later it will disappear from this world depending upon the seriousness of the lesion and luck of the suffering parents.

Normal human circulation needs **two ventricles** and two unobstructed circulations.Though most cyanotic children coming for surgical intervention have two ventricles, a good number will have only one functional ventricle (**Single Ventricle**). A few of them have only one circulation connected to the heart(**Single Circulation)** while the other circulation is supplied through the duct (duct dependent circulation)

More than half of them will have Tetralogy of Fallot or other variants of RVOT obstructions. Other conditions include Transposition of Great Vessels, Total anomalous pulmonary venous connection, Single ventricle, Single Circulation(Duct Dependent) , Persistent Truncus, Hypoplastic left heart syndrome and rarely Ebsteins anomaly.

Principles of Management

The type of repair depends on the availability of ventricles and connections of both circulations to the heart. When there are two balanced ventricles and normally connected two circulations, a biventricular repair is possible as in Tetralogy of Fallot, TGA and TAPVC which can give the baby a nearly normal heart for the rest of life.

When there is a functional single ventricle with both circulations normally connected , the pulmonary circulation is disconnected from the ventricle in stages , first the SVC and later the IVC so that the single ventricle needs to support only the systemic circulation offering the child the maximum possible palliation for a longer time. This is the Fontan principle.

A lethal situation develops when the systemic circulation is duct dependent and not connected to the available functional ventricle which is RV.(Hypoplastic Left Heart Syndrome) . The surgeon then needs to connect the available ventricle to systemic circulation (reconstructing the aorta) after disconnecting the pulmonary circulation. The pulmonary circulation is then maintained by a small BT shunt and this is the principle of Norwood repair in Hypoplastic left hearts as the first stage of a three staged Fontan procedure.

Early Primary Repair In Infancy

The minimum requirements for a normal life span are a normal heart, lungs and brain. Surprisingly these three organs are still developing in a neonate as the RV in foetal life is normally systemic.There is pulmonary hypertension of the newborn with elevated PVR. The neonatal brain is very underdeveloped and is only 25% as compared to it's adult weight.

The normal maturation of the circulation takes place during early infancy as the PVR regresses, the RV becomes a thin walled ventricle to support a low pressure pulmonary circulation for the rest of the life.Unlike LV ,the RV has less provision for angiogenesis and hyperplasia and continued hypertrophy of RV leads to ischaemic fibrosis and late dysfunction.

Both increased and decreased pulmonary blood flow in cyanotics can lead to abnormal development of the lung or irreversible pulmonary hypertension.

The brain is a fast growing organ in the infant reaching double the weight by six months and almost three fourths of the adult weight by the first birth day. A normal cardiac output without hypoxia is very essential for the crucial brain development at this time.In cyanotics with chronic hypoxia and cerebro vascular insults, brain development at this time will be affected which will cause problems in achieving intellectual and professional goals later when the child grows up.

In cyanotics where the RV continues to hypertrophy (TOF, TGA) early RV decompression can prevent further hypertrophy and late RV dysfunction.Restoration of pulmonary artery pressure and pulmonary blood flow to normal (TAPVC, Truncus) can promote normal lung development.

Early correction of cyanosis can prevent major cerebro vascular insults and promote normal development of the brain which can assure a near full productive life for the child when he or she reaches adulthood.

Early primary repair of the cyanotic heart prevents irreversible changes in right ventricle , lungs and brain and assures the maximum possible lifespan for the baby as decided by the type of lesion. Therefore we can say that the best first birthday gift we can offer to a cyanotic new born is a repaired heart, which will not be 100% normal , but will help the baby achieve at least the minimum possible expectations of the otherwise unfortunate parents.

Types Of Cyanotic Heart Repair With Predicted Outcome

Biventricular Repair ("Cure")

In most biventricular repair in cyanotics a near normal life span can be expected approaching a cure
. Eg. TGA - Arterial Switch , TAPVC Repair

TOF - Intra Cardiac Repair without free PR (Transanular

Patch

Biventricular repair (" Cure")

Biventricular repair using RV to PA valved conduits cannot be considered as

cure as these children need reoperations for conduit changes as they grow up.Some cases of Tetralogy repair with transanular patch may also belong to this group.

Eg. PA –VSD, DORV Repair, Turncus Arteriosus Repair

TOF – Absent pulmonary Valve Repair.

Single Ventricle Repairs - Palliation Only

Successful and timely unloading of pulmonary circulation from the single ventricle can offer a definite palliation for the unlucky cyanotic baby with a missing ventricle.

If the available ventricle is Left Ventricle, Eg (.Tricuspid Atresia), the parents can expect an excellent palliation.

If the available ventricle is Right Ventricle (eg. Hypolpastic Left Heart Syndrome), long term results may not be very good. Sometimes only a transplant can save the child.

Techniques of cyanotic heart repair

(Cartoon Models)

Cyanotic Heart Surgery In Neonates

1 . TGA - Arterial Switch Operation

2. TAPVC – Correction

3. Pulmonary Atresia with Intact IVS – Neonatal palliation

4. Pulmonary Atresia with VSD - BT shunt

5. Hypoplastic Left Heart Syndrome - Stage I Norwood / Sano procedure

6. Interrupted Aortic Arch – Repair.

7. Single Ventricle with increased PBF - PA Band

8. TA + TGA + VSD - Damus Kaye Stansel Procedure

9. TOF – Absent Pulmonary valve:- Conduit Repair

Cyanotic Heart Surgery During Infancy

1. Tetralogy of Fallot - Intra Cardiac Repair
2. Truncus Arteriosus - Conduit Repair
3. Single ventricle – Bi-Directional Glenn Procedure
4. Single ventricle - Hemi Fontan Procedure
5. DORV –Conduit / Tunnel Repairs

Cyanotic Heart Surgery Beyond Infancy

1. PA VSD MAPCAS – Conduit Repair
2. Double Outlet Right Ventricle – Conduit Repair
3. Single Ventricle -Lateral Tunnel Fontan
4. Single Ventricle -Extra cardiac Conduit Fontan
5. Ebstein's Anomaly – Repair.

Cyanotic Children Are Remarkable They Just Spring Back To Life

(Magdi Yacoob)

INTRODUCTION TO NUCLEAR MEDICINE IN CARDIOLOGY

Dr.K.Thirumurthi

Nuclear Medicine refers to a discipline, which employs radioactive isotopes for purpose of diagnosis and treatment. The term "Radioactive" implies that isotopes of elements that emit particulate matter (such as Alpha and Beta rays) and electromagnetic radiation (Gamma rays). Technetium-99m (Tc-99m), Iodine-131 (I-131), Thallium-201 (Tl-201) etc., are commonly employed radioactive isotopes in medicine.

In nuclear imaging usually a gamma ray emitting isotope is administered orally or parentally. The radioactive substance is administered as such or it is tagged to a particular vehicle or transporting agent and administered into the body. It is carried to the organ of interest by the blood stream. It gets distributed in the organ and emits Gamma rays. The distribution of isotope (thereby its radioactivity) in an organ of interest is then mapped out by employing a Gamma camera.

Normally the distribution of radioactivity is uniform in an organ and maps the functional image of the organ studied. Areas of reduced function within the organ concentrates less of isotope, giving rise to areas of filling defects referred to as cold spots in the image. Hyperfunctioning and sometimes malfunctioning areas take up more of tracer, which are appreciated as hot spots on the image.

Similar to X-ray tomography (CT Scan), the gamma camera can also be rotated around the patient and sectional images can be obtained. This procedure known as SPECT (Single Photon emission tomography) is particularly useful in studying Myocardial perfusion, Cerebral perfusion, bone etc.

Myocardial perfusion studies :

Perfusion of myocardium at rest and at stress can be studied non-invasively by using SPECT.

Tl-201 or Tc-99m labeled isonitrile compounds (Sestamibi, Tetrofosmin etc) distributes in healthy myocardium in perfusion. Areas of infarction do not concentrate the tracer and hence stand out as cold areas. Injecting these tracers during stress will show the disparity

in perfusion in an ischemic zone distal to a stenotic coronary vessel and areas of normal perfusion supplied by a healthy vessel. When rest images are taken both areas can be seen to have good perfusion. This method of assessing tissue perfusion is invaluable in detection and management of coronary artery disease. Tl-201 studies are also very useful in determining the amount of viable tissue present in the infarcted area.

Indications:

* To assess the extent of infarction.
* To detect and assess the amount of viable tissue present in the infarcted area.

Stress and rest study:

* To detect coronary artery disease.
* To assess the patency of by-pass graft.
* Risk assessment after myocardial infarction.

Blood Pool study:

To assess the left ventricle function accurately this study is carried out. It is also termed as MUGA (Multiple gated acquisition). Tc-99m is labeled to the RBC. An ECG gated study records all phases of cardiac cycle. Change in ventricular volume will be seen as change in radioactivity.

By this technique the ejection fraction of ventricles, stroke volume and cardiac output can be measured with precision. The movement of the heart can be observed in cinematic display. Any abnormal movement of the wall such as hypokinesia, akinesia or dyskinesia can be detected, quantified and documented.

Indications:

* To assess left ventricle function in coronary artery disease,

  cardiomyopathy etc.

* To assess right ventricle function.

* To study wall motion of left ventricle.

Lung Imaging:

* TC-99m labeled MAA (Macro aggregated albumin) are particles slightly bigger than the capillary lumen. Once injected into a peripheral vein the first capillary bed they meet will be the pulmonary capillary bed. They get stuck there and can be imaged. Areas of absent perfusion do not receive the tracer and appear cold. The studies are combined with ventilation studies to differentiate airway disease from true vascular problems.

Indications:

* To detect pulmonary embolism.

* To asses residual functional lung tissue.

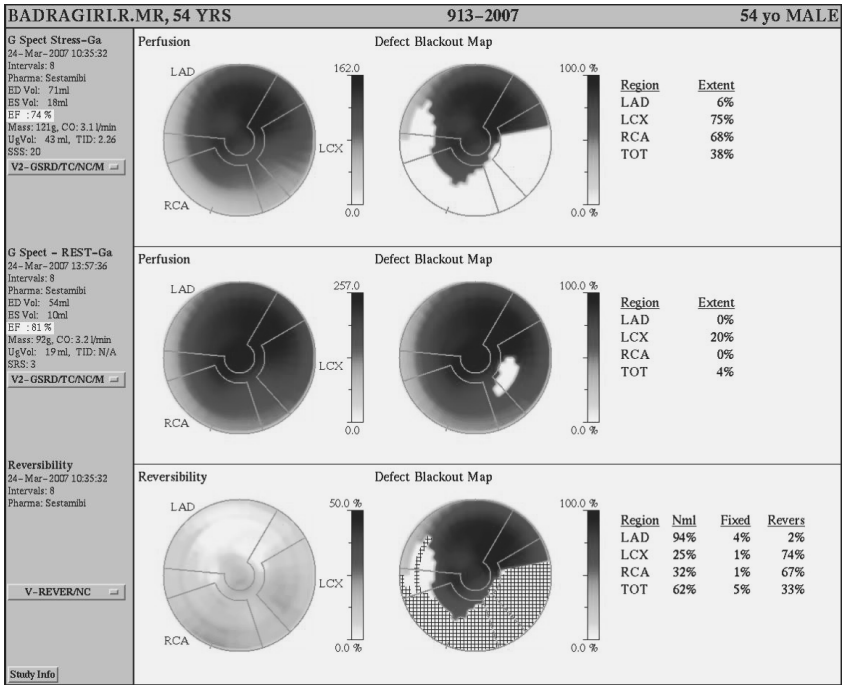
Positron Emission Tomography (PET):

Radioisotopes emitting positrons are now available as cyclotrons get established. These are isotopes having very short half-life and can be used at the site of production or in their close vicinity. These isotopes emit a positron (beta +) which interacts in matter to produce two photons(rays) of 511 kev while the other isotopes (like Tc-99m) are single emit single photon. The commonly used isotopes are F-18,0-15,N-13 etc. These are mainly used to study perfusion and metabolism of organs

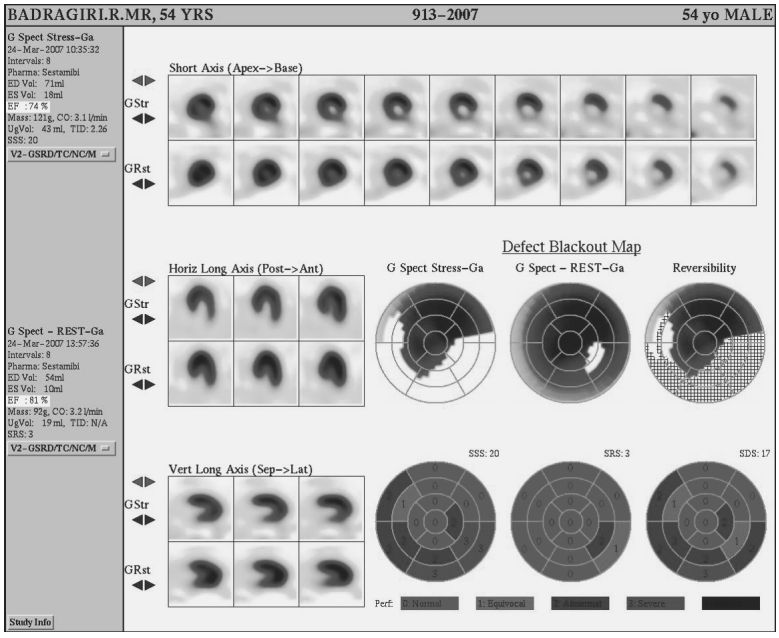
F-18 with a half life of 108 mins is labeled to glucose to study metabolism of ischemic myocardium. Flurodeoxyglucose(F-18 glucose) injected at rest distributes also in ischemic myocardium. As glucose is the main substrate of metabolism in ischemic myocardium, hibernating and stunned myocardium which were partly visible with perfusion tracers (like Tl-201 and Tc-99m Sestamibi) can be fully evaluated and their viability determined. An F-18 glucose study predicts the restoration of function after revascularization and now is considered as the gold standard for estimation of viability. Very bad myocardial segments(nearly 30%) thought to be not viable by SPECT studies were proved to be viable by PET tracers.

N-13 as ammonia (T 1/2 of 13 secs) is used as flow tracer to determine myocardial perfusion. Since there is very less attenuation due to 511 Kev emission and absence of neighboring organ uptake there is good visulasiation of the myocardium. Combining a perfusion study with N-13 ammonia and a metabolic study with F-18 glucose, both stunned and hibernating myocardium can be accurately quantified.

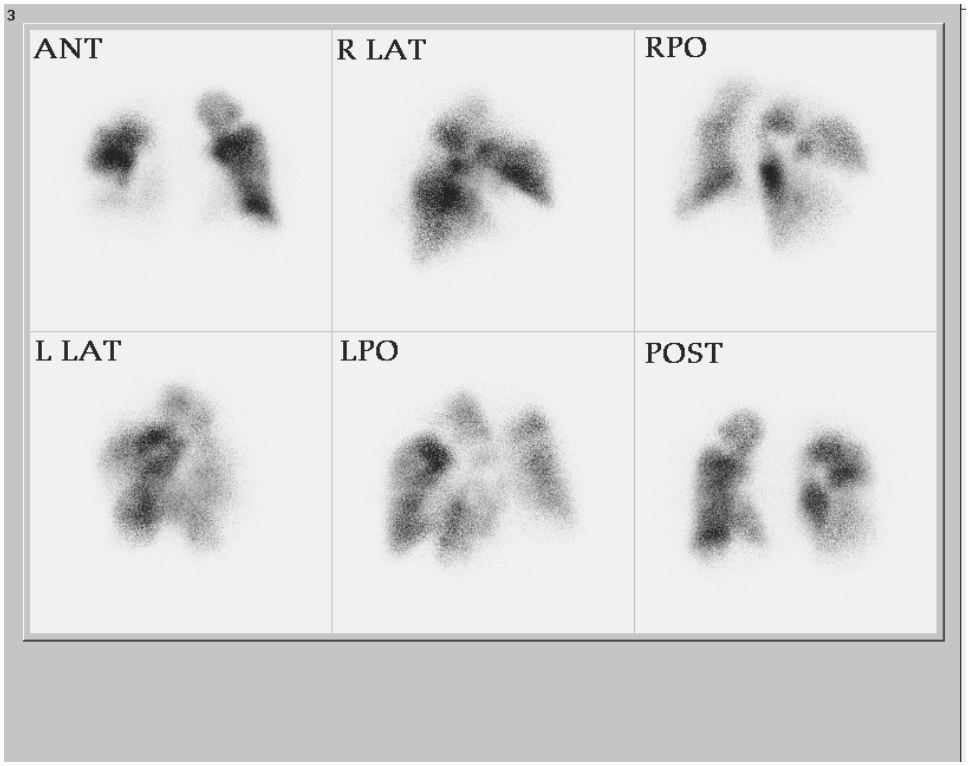
Figures- Contd-

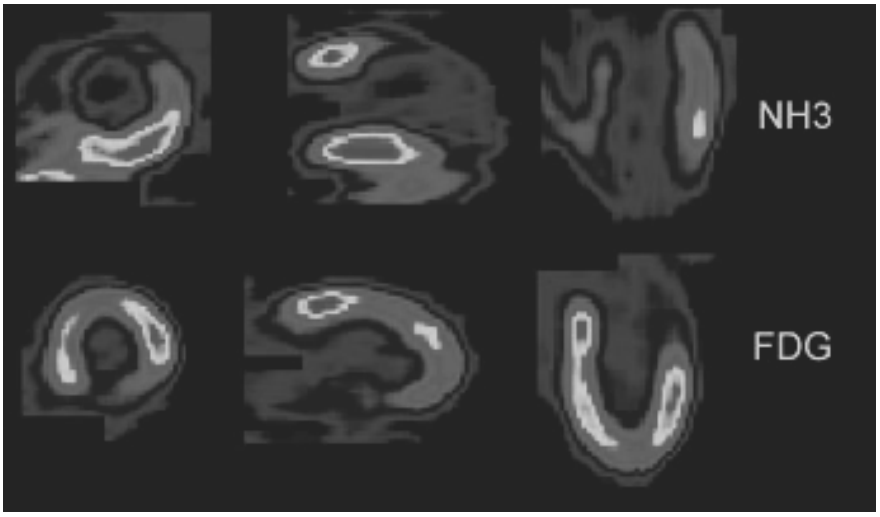


Tc-99m Mycocardial perfusion study at stress and rest showing reversible ischemia in LCX and RCA territories.



Tc-99m lung perfusion scan showing multiple emboli in both lungs





PET scan showing antero-septal infarction with reduced flow in NH3 scan and good FDG uptake indicating viable myocardium

ECHO MEASUREMENTS

Dr. S.R. Ramkumar

Normal values of the left heart chambers by TTE and TEE

MEASUREMENT	TTE	TEE
LV AP diastole (cm)	3.5 – 5.7	3.3 – 5.5
LV AP systole (cm)	2.5 – 4.3	1.8 – 4.0
LV volume diastole (ml)	59 - 157	
LV volume systole (ml)	18 - 68	
LV area diastole (sq cm)	18 – 47	
LV area systole (sq cm)	8 - 32	
LV fractional shortening (%)	30 - 35	30 - 35
LV ejection fraction (%)	> 55	> 55
LV IVS diastole cm	0.6 – 1.1	0.6 – 1.1
LV PW diastole (cm)	0.6 – 1.0	0.6 – 1.0
LV mass (g)	< 294 in men , < 198 in women	
LVOT systole (cm)	1.8 – 3.4	
LA AP (cm)	2.2 – 4.1	2.0 – 5.2
LA volume (ml)	20-77 in men , 15-59 in women	
LA area (sq cm)	9 - 23	
LA appendage length (cm)		1.5 – 4.3
LA appendage diameter (cm)		1.0 2.8

Normal values of right heart chambers by TTE and TEE

MEASUREMENT	TTE	TEE
RV AP diastole (cm)	2.5 – 3.8	
RV AP systole (cm)	2.0 3.4	
RV mediolateral diastole (cm)	2.1 – 4.2	
RV mediolateral systole (cm)	1.9 – 3.1	
RV area diastole (sq cm)	11 – 36	
RV area systole (sq cm)	5 – 20	
RV ejection fraction (%)	> 40	
RV free wall thickness (mm)	2 - 5	
RV out flow systole (cm)	1.8 – 3.4	1.6 – 3.6
RA AP systole (cm)	2.9 – 4.6	2.8 – 5.2
RA mediolateral (cm)		2.9 – 5.3
RA volume (ml) RA area (sq cm)	15 -58 in men , 14 \44 in women 8.3 – 19.5	

Normal measurements of left heart valves and great vessels

MEASUREMENT	TTE	TEE
Mitral valve area (sq cm)	4 - 6	4 – 6
Mitral annulus diastole (cm)	2.0 -3.4	2.0 - 3.8
Mitral leaflet thickness (mm)	<4	0.7 - 3
Pulmonary veins (mm)	8 – 15	7 - 16
Aortic valve area (sq cm)	3 - 5	3 - 5
Aortic annulus systole (cm)	1.4 – 2.6	1.8 – 2.7
Aortic sinus diastole (cm)	2.1 – 3.5	
Aortic root tubule (cm)	1.7 – 3.4	
Aortic arch (cm)	2.0 – 3.6	
Descending aorta (cm)	2.0 2.5	1.4 – 3.0

Normal measurements of right heart valves and great vessels

MEASUREMENT	TTE	TEE
Tricuspid valve area (cm)	4 – 6	4 - 6
Tricuspid annulus diastole (cm)	2.0 – 4.0	2.0 – 4.0
Tricuspid leaflet thickness (mm)	<4	0.7 - 3
SVC		0.8 – 2.0
IVC	1.8 – 2.3	
HEPATIC VEIN	0.5 – 1.1	
Coronary sinus		0.4 – 1.0
Pulmonary valve area (sq cm)	3 - 5	
Pulmonary valve annulus (cm)	1.0 - 2.2	
RVOT systole (cm)	1.8 – 3.4	1.6 – 3.6
MPA (cm)	1.0 – 2.9	
RPA or LPA	0.7 – 1.7	1.2 – 2.2

Normal aortic and pulmonary valve velocities in adults < 50

PARAMETER	AORTIC VALVE	PULMONARY VALVE
Peak velocity (cm/s)	72 - 120	44 - 78
Ejection time (ms)	265 – 325	280 - 380
Acceleration time (ms)	83 – 118	130 – 185

Normal left and right doppler values

Normal left and right doppler values	**Left ventricle**		**Right ventricle**	
Parameter	< 50 yrs	> 50 yrs	< 50 yrs	> 50 yrs
Peak E cm/s	72 ± 14	62 ± 14	51 ± 7	41 ± 7
Peak A cm/s	40 ± 10	59 ± 14	27 ± 8	33 ± 8
E / A ratio	1.9 ± 0.6	1.1 ± 0.3	2.0 ± 0.5	1.3 ± 0.4
Deceleration time ms	179 ± 20	210 ± 36	188 ± 22	198 ± 23
IVRT ms	76 ± 11	90 ± 17	76 ± 11	90 ± 17
Pulmonary vein peak S cm/s	48 ± 9	71 ± 9	41 ± 9	42 ± 12
Pulmonary vein peak D cm/s	50 ± 10	38 ± 9	22 ± 5	22 ± 5

RESTRICTIVE PHYSIOLOGY CONSTRICTION OR RESTRICTION ?

Dr.S.Shanmugasundaram

Restrictive physiology is characterized impediment to ventricular filling leading to elevation and equalization of diastolic pressures in both the ventricles and atria. Early diastolic filling is preserved or exaggerated in these conditions and filling ceases vitually in midlate diastole. Though in cardiac tamponade, the ventricular pressures are elevated and equal, impediment to ventricular filling occurs throughout diastole and thus the hemodynamics is different from the classical restrictive physiology. In normals, ventricular filling in early diastole is determined mostly by a minimal LA - LV gradient that occurs soon after the AV valves open and to a certain extent by the suction effect of relaxing compliant ventricle. In early diastole, nearly 80% of filling is completed. In diastasis the LA – LV gradient is abolished or even reversed and hence no or minimal (< 5%) filling occurs during this phase. During atrial contraction, the gradient is reestablished and the driving force of active atrial contraction contributes to 15% of filling. When ventricular filling is restricted either by a thick non yielding pericardial shell of constriction or by a stiff ventricular myocardium of restrictive cardiomyopathy , the filling pressures in the ventricles is elevated and this in turn leads to elevated atrial pressures. When constriction or restriction involves both the ventricles to a similar extent, the ventricular filling pressures of both ventricles are elevated to similar levels. Since the atria and ventricles function as a common chamber when the AV Valves are open, the diastolic pressures in all the four chambers are equal.

Differentiation between constrictive pericarditis and restrictive cardiomyopathy is crucial in terms of disease management, as the former is potentially curable by surgery. In general, restrictive physiology is suspected whenever symptoms and signs of right ventricular failure occur alone or out of proportion to that of left heart disease. Most of the common causes of right heart failure, namely primary and secondary pulmonary hypertension (mitral valve disease, LV dysfunction, pulmonary embolism etc) and right ventricular infarction can be diagnosed at the bedside and echo often resolves the diagnosis in such conditions. However the hemodynamics and clinical presentation of constrictive pericarditis and restrictive cardiomyopathy resemble each other and even the echo and invasive parameters might have some overlap so that the differential diagnosis between these two conditions becomes a difficult, but interestingly challenging task. Though pericardial thickness can be easily measured by CT scan, there are important caveats. In certain disease states pericardium may be thickened but not imposing restriction to filling. In some cases of postoperative and post irradiation constrictive pericarditis, the pericardial thickness may be normal.

An insight into the hemodynamics is very essential to understand how the clinical features and the findings in imaging studies help in differentiating between the conditions. In both the conditions, the ventricular filling is interfered – in constriction the limitation to filling is extrinsic, offered by the thickened pericardium and in restrictive cardiomyopathy the impediment to filling is intrinsic to myocardium. In the former, the early diastolic relaxation is unaffected and the elevated atrial pressure accentuates the early diastolic filling. The early diastolic filling occurs at a higher velocity in shorter period of time and comes to an abrupt cessation when further ventricular distension is halted by the constricting pericardium. In restrictive cardiomyopathy the active process of relaxation is defective and the ventricular myocardium is stiffer. Inspite of it, the early diastolic filling is rapid because elevated atrial pressure overcomes the delayed relaxation of myocardial disease. The stiffened myocardium terminates filling at the end of rapid filling phase. Because of the commonness of occurrence of rapid early diastolic filling which comes to abrupt halt, in both the conditions, the y descent in atrial

pressure wave form and in JVP is steeper, the ventricular pressure wave form reveals square root sign and the AV Valve flow Doppler reveals the so called restrictive pattern (E/A > 2 ; shortened deceleration time).

The hemodynamics of constrictive pericarditis (CP) differs from that of restrictive cardiomyopathy (RCM) in two ways – In CP, ventricular interaction is enhanced and there is dissociation between intracardiac pressures and intrathoracic pressures. The cardiac chambers are intrapericardial and the intrapericardial pressures reproduce the changes in intrathoracic pressure. The systemic and the pulmonary veins are extrapericardial, the former being mostly extrathoracic and the later being intrathoracic. In normals, inspiration decreases the intrathoracic pressure which is reflected on to the cardiac chambers and the pulmonary veins. The pressures in the extrathoracic systemic veins is unaltered and hence there is an increased gradient between systemic veins and right heart. Thus the systemic venous return increases (as much as 56%) and the increased RV filling shifts the interventricular septum slightly toward LV and may decrease LV filling slightly. The pulmonary venous pressure and the left heart pressures follow the changes in intrathoracic pressure to a similar extent and hence may not be responsible for decreased LV filling in inspiration in normals. In CP, the pericardial shell prevents the respiration induced changes in intrathoracic pressures from getting reflected on to the cardiac chambers. Inspiration reduces the pressure in pulmonary veins but does not alter the left heart pressures. Thus the gradient between the pulmonary veins and left atrium decreases in CP during inspiration contributing to decreased LV filling. Decreased LV filling lowers LV pressures slightly allowing the ventricular septum to bounce towards LV which in turn augments RV filling. Since both the ventricles are encased in the common pericardial shell, there is an accentuated ventricular interaction. Thus the respiration induced reciprocal changes in LV and RV filling are exaggerated in CP. In RCM, there is no dissociation between intrathoracic and intracardiac pressures. Even the normal increase in venous return in inspiration is not accommodated by the diseased RV. Hence the respiratory changes in RV and LV filling are not exaggerated and may even be attenuated. These hemodynamic features and certain patho anatomic features help in differential diagnosis between CP and RCM which are tabulated below.

DIFFERENTIAL DIAGNOSIS

S. No.		RCM	C.P
(1)	**PAST HISTORY**	-	**SURGERY, RADIATION, DRUGS, TRAUMA, CTD, MI, CKD**
(2)	**JVP**	**Steep XY**	**Steep XY**
		? a, ? v	
	KUSSMAUL	+	+
(3)	**PULSUS PARADOXUS**	**ABSENT**	**MAY BE PRESENT**
(4)	**PRECORDIUM**	**? API, PSH**	**APICAL RETRACTION**
		RVOT IMPULSE	

(5)	ADDITIONAL SOUNDS	S_4, S_3 (120-180 ms)	P. KNOCK (60 – 120 ms)
(6)	MURMURS	TR, MR	ABSENT
(7)	ECG : RHYTHM P wave QRS	ST / AF BROAD, TALL, LA/ BAE LOW VOLTAGE; BBB (25%) Q, VENTR. HYPERTROPHY	ST / AF BROAD; SHALLOW;LAE LOW VOLTAGE; Q waves T flat or inverted
(8)	CHEST X RAY : CTR CALCIFICATION PULMONARY EDEMA CHAMBER ?	 ?? INTRAMURAL ± +	 N? PERICARDIAL NO NO
(9)	BNP (pg / ml)	> 600	< 200
(10)	PERICARDIAL THICKNESS(CT/MRI)	N	?
(11)	2D & M MODE ECHO : PERICARDIAL FLUID THICKNESS IVC PLETHORA ATRIAL ENLARGEMENT MYOCARDIUM MR / TR SEPTAL NOTCH SEPTAL BOUNCE VENTRICULAR DIMENSION I/E	 ++ N ++ +++ Altered thickness & texture ++ ABSENT ABSENT -	 ± ? ++ NO / LAE Normal NO / MINIMAL PRESENT PRESENT RECIPROCAL

12)	**DOPPLER :** **DD Grade III** *(E/A > 2 ; DT < 160 ms ; IVR < 60 ms)*	+	+
	RESPIRATORY VARIATION OF **MITRAL / TRICUSPID FLOW**	**NO / MINIMAL** **(~ 5 %)**	**EXAGGERATED** **(> 25 %)**
	PULM. VEIN FLOW	**S/D ~ 0.5** **? AR**	**S/D = 1** **EXPIRATORY ? OF D** **(27 – 35 %)**
	TR VELOCITY I / E	**NO CHANGE**	**INSP.? OF TR – v & d**
	DIASTOLIC MR / TR	++	±
	E' ; E / E'	**< 8 cm/sec ; > 15**	**> 10 cm/sec ; < 15**
	VP IN COLOR M MODE	**< 45 cm/sec**	**> 45 cm/sec**
	MYOC.VEL.GRADIENT	**?**	**N ?**
	LV LONGITUDINAL STRAIN	**Decreased**	**Normal**
	LV CIRCUMFERENTIAL STRAIN	**Decreased**	**Decreased**
	LV TORSION	**Normal**	**Decreased**
(13)	**NUCLEAR PFR**	**?**	**? ~**
(14)	**CARDIAC CATHETERIZATION** **RA** **RV / LV PRESSURE** **RVSP (PASP)** **RVEDP** **CORONARY FLOW**	**XY (M/W)** **Prominent a & v** **LVEDP > RVEDP by 5mm** **Resp. CONCORDANCE** **> 50 mmHg** **< 1/3 OF RVSP** **Dip & PLATEAU** -	**XY (M/W)** **LVEDP = RVEDP** **DISCORDANCE** **< 50 mmHg** **> 1/3 OF RVSP** **Dip & PLATEAU** **Mid late systolic reversal**