Adult Congenital Heart

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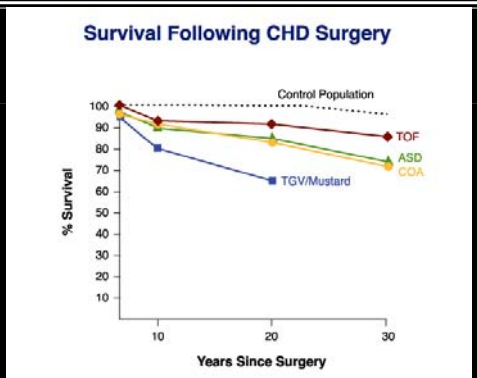
Overview

- US: 1,000,000 adults with congenital heart dz
- 20,000 more patients reach adolescents yearly
- Cardiologists must
  - Have detailed knowledge of congenital dz, both repaired and unrepaired
  - Clearly define each patients surgical and corrective procedures (read surgical notes)

*All figures from ACCSAP V unless otherwise noted

Survival Following CHD Surgery



Daniels, CJ. Congenital Heart Disease. ACCSAP V

Surgical Terminology in Adult Congenital Heart Disease

Surgical Shunts in CHD

Shunt	Procedure
Classic Blalock-Taussig	Subclavian artery to pulmonary artery as end to side anastomosis.
Modified Blalock-Taussig	Gore-Tex tube graft from subclavian artery to pulmonary artery.
Bidirectional Glenn	SVC to RPA. End to side. Second stage to fontan.
Fontan Procedure	IVC to RPA to complete systemic venous circulation to PAs. Gore-Tex tube that may be in the heart (lateral tunnel) or outside the heart (extracardiac).
Waterson	Ascending aorta to RPA. A punch hole between the vessels.
Potts	Descending aorta to LPA. Punch hole between the vessels.

Surgical Procedures

Procedure	Surgery
Rastelli Procedure	Transposition with a VSD whereby the VSD is closed baffling LV to Ao and a RV to PA conduit is placed.
Norwood Procedure	Hypoplastic LV syndrome. The pulmonary valve and artery are used to create neo-aorta and a BT shunt is placed to provide pulmonary blood flow.
Mustard Procedure	Transposition of the great vessels. Baffle the SVC/IVC to the MV - LV – PA and baffle the pulmonary veins to TV – RV – Ao.
Arterial Switch	TGV where the aorta and MPA are switched and coronary arteries re-implanted.

Congenital Heart Disease. ACCSAP V

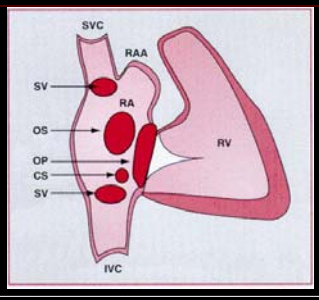
Adult Congenital Heart Disease

- Atrial Septal Defect
- Coarctation of Aorta
- Tetralogy of Fallot
- Transposition of Great Arteries
- Common Ventricle/Fontan Procedure
- Ebstien's Anomaly
- Eisenmenger Syndrome
- Pregnancy

Atrial Septal Defect

- 1/1500 live births
- Secundum
  - most common ACHD (6-10%)
  - RAD
- Primum
  - associated with other endocardial cushion defects (cleft AV valves, inlet type VSD)
  - LAD
- Sinus Venosus
  - large, associated with anomalous pulmonary venous drainage (usually R superior PV)
- Coronary sinus (rare)
  - associated with unroofed coronary sinus

ASD- Anatomy/Prevalence

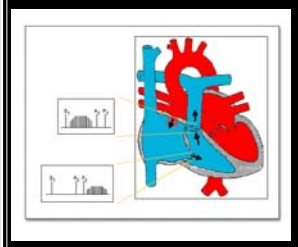


- Secundum 75%
- Primum 15%
- Sinus Venosus 10%
- Cor Sinus (rare)

ASD - Clinical

- Majority repaired in childhood, but may present in adolescence/adulthood
- Asymptomatic
  - murmur, abnl ECG/CXR
- Symptomatic
  - dyspnea/CHF
  - CVA/emboli
  - Atrial Fibrillation

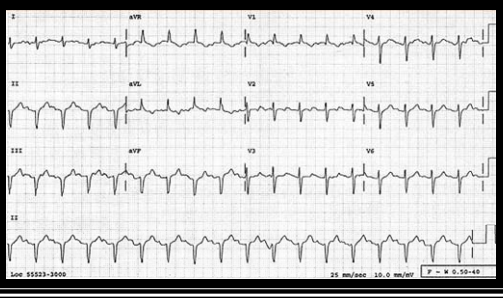
Auscultation in ASD



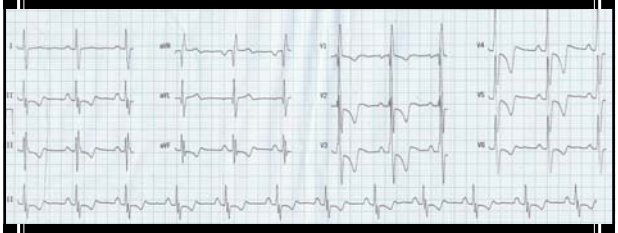
Increased flow across the pulmonary valve produces a systolic ejection murmur and fixed splitting of the second heart sound.
Fixed splitting of S2 may in part be due to delayed right bundle conduction.
Increased flow across the TV produces a diastolic rumble at the mid to lower right sternal border.

- Older pt loses pulm ejection murmur as shunt becomes bidirectional
- signs of pulm HTN/ CHF may predominate

ECG of ASD, Secundum



ECG of ASD, Primum

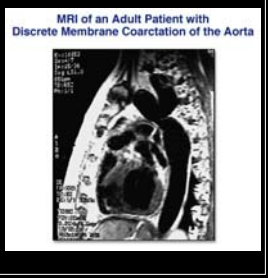


ASD: Therapy

- Percutaneous Closure
  - only for secundum (contra in others)
  - adequate superior/inferior rim around ASD
  - no R-L shunting
- Surgical Closure
  - Good prognosis:
    - closure age < 25, PA pressure <40
    - If >25 or PA>40, decreased survival due to CHF, stroke, and afib

Coarctation of Aorta

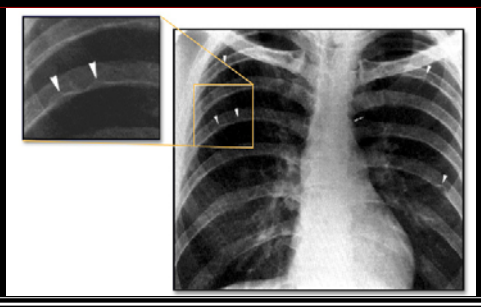
- Narrowing in proximal descending aorta
- May be long/tubular but most commonly discrete ridge
- Natural hx: poor prognosis if unrepaired
  - Aortic Aneurysm/dissection
  - CHF
  - Premature CADz



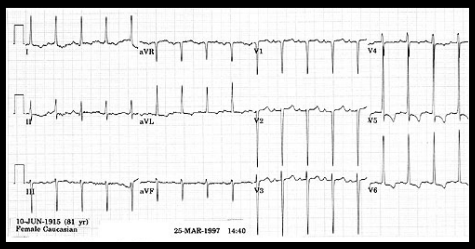
Coarctation of Aorta: Clinical

- Most repaired, but adult presentation may be:
  - HTN
  - murmur (continuous or systolic murmur heard in back or SEM/ejection click of bicuspid AV)
- weak/delayed LE pulses
- Rib notching on CXR pathognomonic

Rib notching

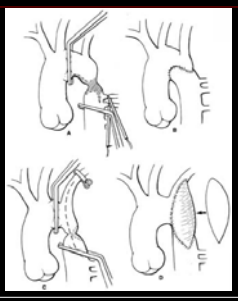


Coarctation of Aorta: ECG



Coarctation Repair

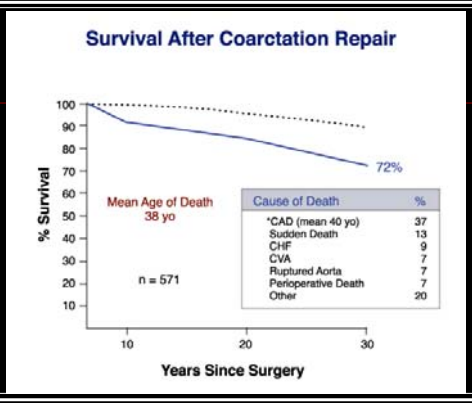
- Surgical correction
  1) Patch aortoplasty with removal of segment
  and end to end anastomosis or subclavian flap repair
  2) bypass tube grafting around segment



Edmunds' Cardiac Surgery in the Adult, Ch 47

Coarcation: Treatment

- Despite surgery, patients still have significant morbidity/mortality with average age 38
- Up to 70% of repaired patients still go on to develop HTN, pathology not well understood
- Recurrence in 8-54% of repairs, can undergo repeat surgery or balloon angioplasty
- Aortic Aneurysm/ruputure may occur despite successful repair and correction of HTN (freq around anastomosis site on patch repair – 30% in one study)

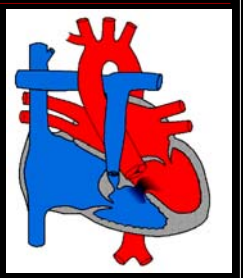


Coarctation: Followup

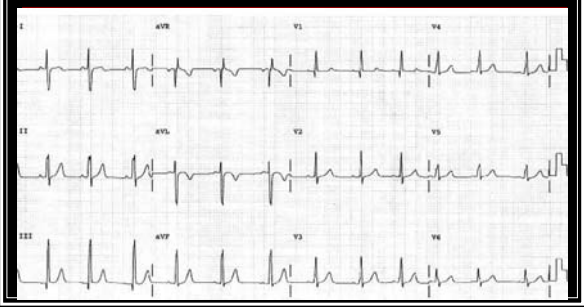
- Every 1-2 years
  - Document arm/leg BP
  - Screen/treat CAD risk factors
  - HTN: rest, provoked by exercise or seen on ambulatory monitoring
  - ECHO/doppler to eval recurrent
  - MRI for aneurysm

Tetralogy of Fallot

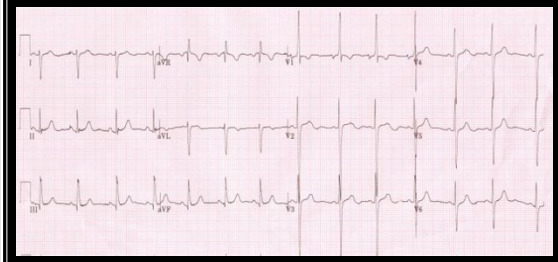
- 4 features
  - Malalignment VSD
  - Overriding Aorta
  - Pulmonic Stenosis
  - RVH
- Variability correlates with degree of RVOT obstruction and size/anatomy of PA



Tetralogy of Fallot: ECG



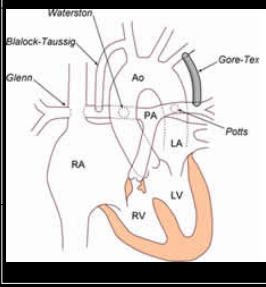
Tetralogy of Fallot: ECG



Tetralogy: Surgical Treatment

- Systemic – Pulmonary Shunt
  - Blalock-Taussig
  - Waterston (RPA)
  - Potts (LPA)
- Complete Repair
  - takedown of prior shunt
  - patch VSD
  - resection of subpulmonic obstruction
  - transannular patch around pulm valve annulus (usually leads to severe PI)

Systemic – Pulmonary Shunt



A classic Blalock-Taussig shunt (BT shunt) connects the right or left subclavian artery directly to the left or right pulmonary artery.

A modified BT shunt consists of a Gore-Tex graft that then connects the subclavian artery to the pulmonary artery. The advantage to a modified BT shunt is having more controlled blood flow.

A Waterston shunt connects the ascending aorta to the main or right pulmonary artery.

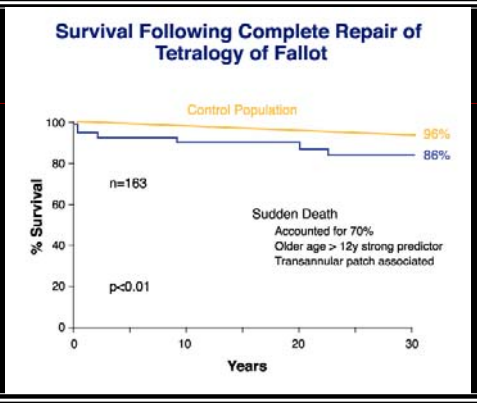
The Potts shunt connects the descending aorta to the pulmonary artery.

Tetralogy: Treatment/complications

- Systemic-Pulm shunt
  - leads to high flow through PA, elevated PVR and branch PA distortion
  - survival after repair worse in pt with prior Waterston or Potts shunt (?higher flow); some pt with Blalock-Taussig shunts may survive unrepaired into adulthood
  - these pt should be evaluated for pulm artery stenosis and Pulm HTN

Tetralogy: Treatment/complications

- Prior pulmonary valve atresia or anomalous LAD may have had prosthetic or homograft conduit ± valve placed between RV and PA
- Conduits can undergo endothelial overgrowth and obstruction of "pseudo RVOT" – can Rx with balloon angioplasty or operative conduit replacement

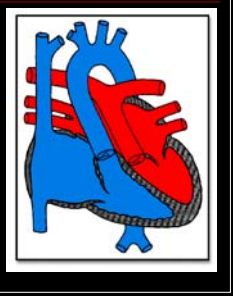


Tetralogy: Risk/followup

- SCD ↑ 25-100 fold
  - risk can occur 2 decades after correction
  - related to QRS duration> 180msec
  - ? Due to PI/RV conduction defect
  - atrial arrhythmias also common
- Hemodynamic effects of PI
  - Chronic RV volume overload, RV dysfunction and exercise intolerance
  - Pulmonic Valve Replacement can decrease QRS duration and stabilize RV fxn; timing unclear but earlier better than later
  - RV fxn: ECHO or MRI

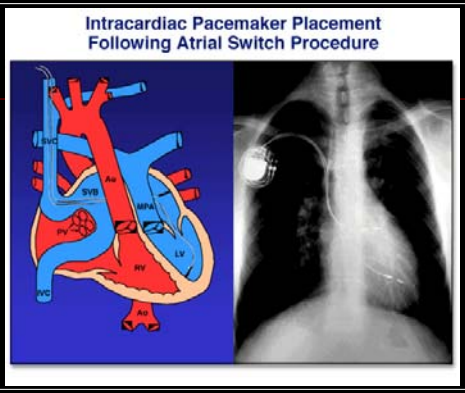
Transposition D-type

- PA arises from LV, Aorta from RV and anterior/right of PA
- cyanosis
- corrected initially with prostaglandin to keep ductus open and balloon atrial septostomy to improve systemic saturation
- repair via "atrial switch" Mustard procedure
  - SVC/IVC baffled to LA/LV/PA
  - Pulm Veins baffled to RA/RV/Ao
  - Symptom free survival until 2nd-3rd decade of life
- repair via "arterial switch"
  - long term data ?
  - pulmonic valve (neo-aortic valve) competence?, reimplanted coronaries may develop ostial stenoses



D-Transposition complications

- Complications
  - arrhythmias/SCD
    - Only 18% maintain SR; most go on to SSS/Afib/ Aflutter; pacemaker often needed
  - systemic (tricuspid) atrioventricular valve regurgitation
    - ? TVR
  - systemic (RV) ventricular failure
    - 15% have CHF sxs by 2nd-3rd decade
    - Rx transplant or staged Arterial switch (pulm banding to "train" LV)
  - baffle obstruction
    - Rare (5%) but serious complication; venous more common
    - Suspect if new upper extremity edema (venous) or new CHF sxs (pulm venous)
    - ECHO or Cath to eval, pulm venous obstruction Rx with surgery, systemic venous with angioplasty/stents

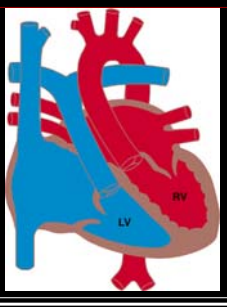


Pacer wire must go to LV via SVC baffle

- Patients surviving arterial switch

L-type Transposition

- Atrial-ventricular AND ventricular-arterial discordance
- Physiologically correct, anatomically incorrect
- "congenitally corrected" transposition
- RV is systemic ventricle, TV is systemic AV valve
- Asymptomatic for many years, often into adulthood

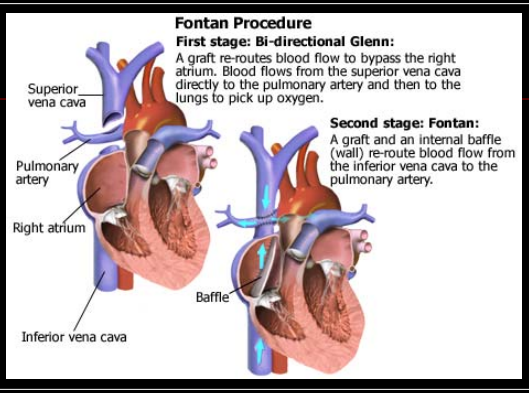


L-type transposition: complications

- Although seemingly benign, survival is reduced with one study showing 25% of patients died by mean age 38
- Progressive Heart Failure
- Arrhythmias
  - SCD
  - AV block
  - Atrial arrhythmias
- Severe AV (tricuspid) regurgitation – TVR
  - difficult to image using conventional ECHO
  - MRI becoming test of choice for RV function

The Fontan Patient

- Any congenital anomaly with an effective "single" or "common" ventricle may lead to a Fontan procedure
  - Tricuspid Atresia – also any other form of right sided hypoplasia or atresia.
  - Double Inlet LV.
  - Hypoplastic Left Heart.
  - Some variations of Double Outlet RV
- Staged Procedure
  - Basic concept is to provide systemic venous return directly to PA and bypass ventricle
  - systemic-pulm shunt to stabilize pulm blood flow
  - bi-directional Glenn or hemi-Fontan procedure
    - SVC flow directed to PA and sys-pulm shunt ligated
  - Finally, Fontan procedure with IVC directed to PA
  - Older Fontan: includes RA in circuit; newer methods bypass RA



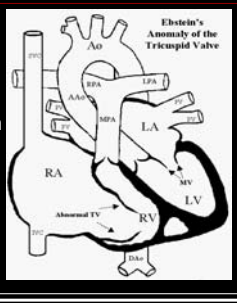
www.heartcenteronline.com

Fontan: complications

- Arrhythmias
  - most pt develop SSS/tachy-brady
- Heart Failure
- RA may become enlarged and source for thrombus (with older Fontan), can undergo Fontan revision with bypass of RA/extracardiac graft
- Uncorrected patients develop polycythemia and treatment becomes palliative at this point

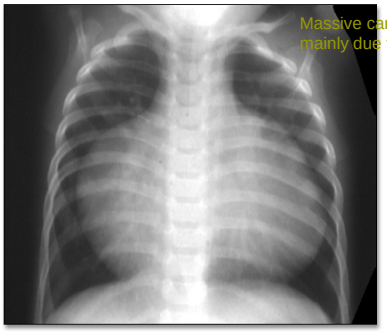
Ebsteins Anomaly

- Atrialization of RV, sail-like TV, TR
- 50% ASD/PFO
- 50% ECG evidence of WPW
- Age at presentation varies from childhood→adulthood and depends on factors such as severity of TR, Pulm Vascular resistance in newborn, and associated abnormalities such as ASD



www.uoch.org

CXR of a Patient With Ebstein Anomaly



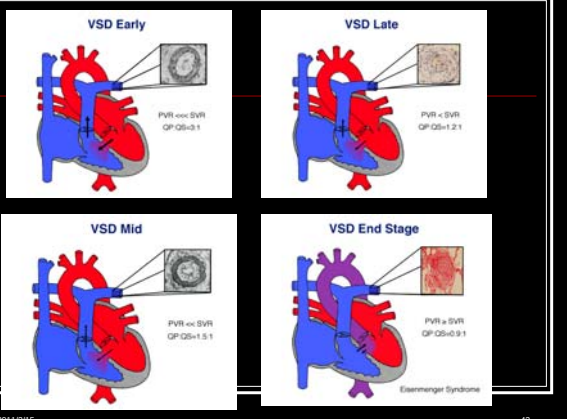
Massive cardiomegaly, mainly due to RAE

Ebsteins: Clinical Presentation

- Pediatric
  - murmur
- Adult (unrepaired with ASD)
  - atrial arrhythmias
  - murmur
  - cyanosis
    - R→L shunt NOT due to PulmHTN but TR jet directed across ASD
  - exercise intolerance
- Surgery in pts with significant TR/sxs

Eisenmenger's Syndrome

- Final common pathway for all significant L→R shunting in which unrestricted pulmonary blood flow leads to pulmonary vaso-occlusive disease (PVOD); R→L shunting/cyanosis develops
- Generally need Qp:Qs >2:1



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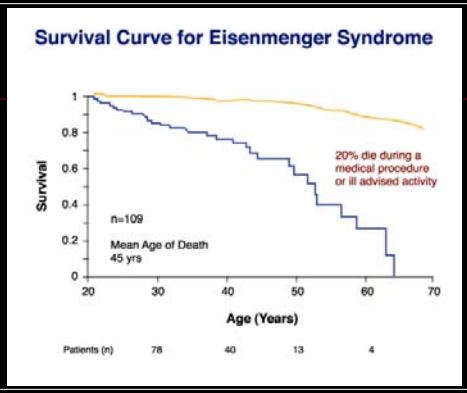

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Eisenmenger Complications

- Coagulopathy/platelet consumption
- Brain abcesses
- Cerebral microemboli
- Airway hemorrhage
  - especially moving from lower→higher altitudes (air travel, mountains)

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Eisenmenger: Treatment

- Sxs +polycythemia → phlebotomy
  - Careful if microcytosis, strongest predictor of cerebrovascular events
- RULE OUT CORRECTABLE DISEASE
- Once diagnosis established, avoid aggressive testing as many patients die during cardiovascular procedures
- Diuretics prn, oxygen
- Definitive: Heart Lung transplant
  - Prostacyclin therapy may delay, expensive

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Pregnancy

Hemodynamic Changes During Normal Pregnancy

Parameter	1st Trimester	2nd Trimester	3rd Trimester
Blood Volume	↑	↑↑	↑↑↑
Cardiac Volume	↑	↑↑to↑↑↑	↑↑↑to↑↑↑
Stroke Volume	↑	↑↑↑	↑,↔, or ↓
Heart Rate	↑	↑↑	↑↑to↑↑↑
Systolic Blood Pressure	↔	↓	↔
Diastolic Blood Pressure	↓	↓↓	↓
Pulse Pressure	↑	↑↑	↔
Systemic Vascular Resistance	↓	↓↓↓	↓↓

Hemodynamic Changes During Labor and Delivery

Anxiety, pain, and uterine contractions all alter hemodynamics.

Cardiocirculatory Effects of Uterine Contraction*

Parameter	Change	Comments
Blood Volume	↑	300-500 ml
Cardiac Output	↑	30-60% with cumulative increase between contractions
Heart Rate	↑↓	Variable responses
Blood Pressure	↑	Significant rise of both systolic and diastolic blood pressures, return to baseline between contractions
Peripheral Resistance	↔	
Oxygen Consumption	↑	Increased gradually to average of 100%

* Hemodynamic effects of uterine contractions are less pronounced in lateral recumbency than in the supine position.
↑, increase; ↓, decrease; ↔, no significant change

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Pathophysiologic Categories

Shunt (L→R)
- ASD
- VSD
- PDA

Left Heart Obstruction
- AS
- MS
- HOCM
- Coarctation of Aorta

Right Heart Obstruction
- TS
- PS (TOF)

Prior Arrythmia
- Sustained/Symptomatic Brady or Tachyarrhythmia

Left Heart Regurgitation
- AI
- MR (AVSD)

Right Heart Regurgitation
- PI (TOF)
- TR (TGV, Ebstein's)

Myocardial Dysfunction
- Dilated CMP
- Ischemic CMP (Kawasaki's Disease)
- Systemic RV (TGV)
- Single Ventricles

Pulmonary Hypertension
- L→R Shunt
- PPH

Shunt (R→L)
- Cyanotic (Eisenmenger)

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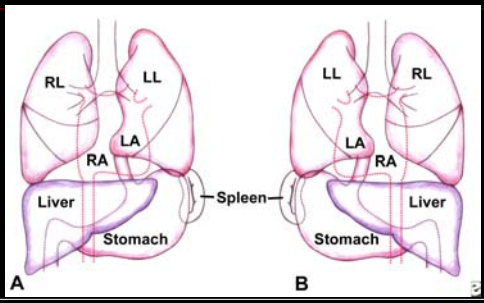
Pregnancy

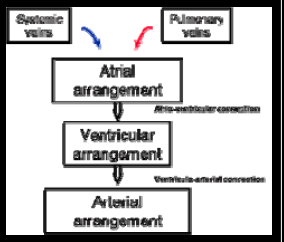
- Shunts
  - generally handled pretty well unless Pulm vascular obstructive dz; use same standards to decide if closure warranted as in non-preg
- L sided obstructive lesions
  - AS, MS, Coarctation carry significant risk
  - AS: can tolerate peak grad<50
  - Coarc: repaired needs MRI to eval anastomosis sites before pregnant, if aneurysm need repair before pregnant
  - Physiology more impt than type of lesion
  - balloon valvuloplasty if necessary (best to dx/fix before pregnancy)

Sequential Segmental Analysis



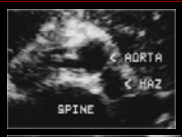
Sequential Segmental Analysis

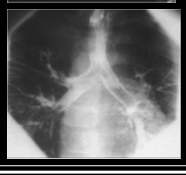
- Situs (Atrial Arrangement)
- Ventricular Morphology
- Atrioventricular Connection
- Aterial Morphology
- Ventricular Arterial Connections
- Cardiac Position



Sequential Segmental Analysis

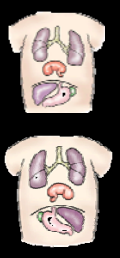
- Situs (Atrial Arrangement)
  - TEE: the relationship of the aorta and IVC to the spine
  - High KV CXR: early-branched morphologically right bronchus
  - ECG: P wave axis





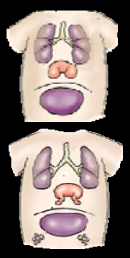
Sequential Segmental Analysis

- Situs (Atrial Arrangement)
  - Situs Solitus
  - Situs Inversus
    - Kartagener's syndrome



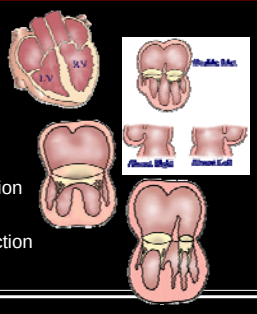
Sequential Segmental Analysis

- Situs (Atrial Arrangement)
  - RA Isomerism
  - LA Isomerism



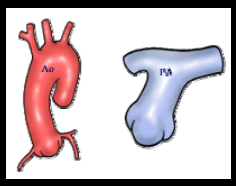
Sequential Segmental Analysis

- Atrioventricular Connection
  - Biventricular
  - Univentricular
  - Common Connection
  - Overriding Connection



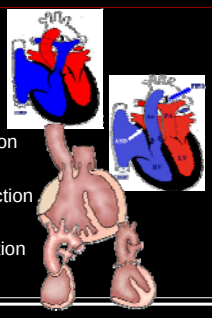
Sequential Segmental Analysis

- Arterial Morphology
  - Aorta
  - Pulmonary Artery



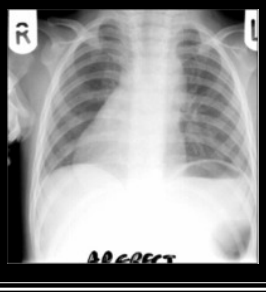
Sequential Segmental Analysis

- Ventricular Arterial Connections
  - Concordant
  - Transposed Connection
  - Double Outlet Connection
  - Single Outlet Connection



Sequential Segmental Analysis

- Cardiac Position
  - Dextrocardia
  - Laevocardia



Sequential Segmental Analysis

- Ventricular Morphology
  - Morphologically right ventricle: triangular shape, heavy apical muscle bands and septal attachments of the valve leaflets
  - Morphologically left ventricle: banana shaped with a smooth wall and valvar attachments only to the free wall.

Question 1

- A 24-year-old Indian man is seen after a syncopal episode that occurred while he was watching a football game on TV. His wife noticed that after a particularly exciting play, the patient suddenly slumped over. She shook him hard, and, after about 30 seconds, he woke up and said that he remembered nothing of the incident. This has never happened before. Up until this time, he has had no limitation of physical activity. His past medical history is significant in that he had repair of tetralogy of Fallot at age 4, at which time a VSD was patched and a right ventricular infundibulectomy was done.

  Physical examination finds no cyanosis. Blood pressure is 100/70 mmHg, and pulse is 65 per minute with an occasional premature contraction. The lungs are clear to auscultation and percussion. Neck veins are 4cm. There is a mid sternal incision that is well healed. There is a slight precordial systolic lift. S2 is single. There is a Grade II/VI systolic ejection murmur with a short Grade II/VI diastolic low-pitched murmur along the left sternal border. There is no S3 or S4.

  The ECG shows right bundle branch block with left anterior hemiblock. The PR interval is 0.12 seconds. The echocardiogram reveals a slightly dilated right ventricle and paradoxical motion of the interventricular septum. Doppler gradient across the right ventricular outflow tract is 35 mmHg. There is evidence of moderately severe pulmonic regurgitation, and there are no left-to-right or right-to-left shunts.

  What is the most important diagnostic test needed for this patient?
- A. TEE.

  B. Electrophysiology study.

  C. Cardiac catheterization and angiography.

  D. Tilt table test.

  E. 24-hour ambulatory ECG.

Question 1 Answer

- **Comment**
  The correct answer is B.

- This young man had a sudden, apparently unprovoked, syncopal episode. His childhood repair of a tetralogy of Fallot resulted in a right ventricular scar. He has the usual postoperative physical findings of a repaired tetralogy, with residual right ventricular obstruction and pulmonic valve insufficiency. Sudden death due to ventricular tachycardia-fibrillation is a danger to these patients. They deserve an electrophysiologic study since, in this case, syncope is equivalent to aborted sudden death.

Question 2

- Catheter-delivered balloon expansion techniques are now the treatment of choice for which one of the following lesions in adults?

  A. Valvular pulmonic stenosis.

  B. Valvular aortic stenosis.

  C. Coarctation of the aorta.

  D. Ebstein's anomaly of the tricuspid valve.

  E. Severe mitral stenosis/regurgitation.

Question 2 Answer

- **Comment**
  The correct answer is A.

- Although catheter balloon valvuloplasty and aortoplasty have been attempted in all these conditions, only pulmonary valvotomy has achieved a success level consistent with being the treatment of choice in adults. Aortic stenosis responds initially to balloon expansion and may serve as a bridge to valve replacement surgery, but is associated with rapid restenosis. Success rates with coarctation and Ebstein's anomaly are not uniform enough to displace surgery except in selected patients.

  Mitral stenosis in the absence of severe subvalvular disease can be successfully treated by balloon valvuloplasty, but the presence of moderate to severe regurgitation is an indication for surgery.

Question 3

- An important predisposing cause for late atrial fibrillation following closing of an atrial septal defect is:

  A. Patch versus suture closure of the defect.

  B. The age of the patient at the time of surgery.

  C. Right ventricular dysfunction.

  D. Sinus venosus defect.

  E. Concomitant mitral regurgitation.

Question 3 Answer

- **Comment**
  The correct answer is B.

  Late atrial fibrillation may occur in as many as 25% of patients with atrial septal defect and is directly related to the age of the patient at time of surgery as well as the age of the patient at late follow-up. It is also influenced by whether the patient had atrial fibrillation early in the preoperative period and may represent interruption of interatrial pathways.

Question 4

- What determines the physiology in tetralogy of Fallot?

  A. The size of the ventricular septal defect.

  B. The position of the ventricular septal defect.

  C. The presence of an atrial septal defect.

  D. The degree of RV outflow tract obstruction.

  E. The presence of a left superior vena cava.

Question 4 Answer

- **Comment**
  The correct answer is D.

- The size of the ventricular septal defect in tetralogy of Fallot is quite uniform. The presentation of the patient as to whether he or she is acyanotic or cyanotic is determined by the degree of the right ventricular outflow tract obstruction (RVOT). RVOT obstruction determines the amount of right-to-left shunting.

Question 5

- A 36-year-old man is referred for suspected ASD. He is employed, active, and asymptomatic. ECG shows a normal axis and incomplete right bundle branch block. Chest x-ray shows an enlarged right heart silhouette and increased pulmonary vessels throughout the lungs. Echocardiography confirms a 3cm diameter secundum ASD with a large shunt. There is a mild tricuspid regurgitation jet of 2 m/sec.

  Your recommendation is which one of the following?

  A. Yearly follow-up.

  B. Digoxin.

  C. Warfarin.

  D. Angiotensin converting enzyme inhibitor.

  E. Cardiac surgical repair.

Question 5 Answer

- **Comment**
  The correct answer is E.

- Patients with large ASDs should have them closed because the natural history of ASD is a shortened life span due to eventual right heart failure from the volume overload. Paradoxical embolism and pulmonary hypertension are additional concerns as are atrial fibrillation and its sequelae. The recommended closure technique today is surgical. Catheter-delivered devices are promising but have not been perfected. Digoxin and ACE inhibitors are of no known value. Anticoagulation is not indicated unless atrial fibrillation or other indications for it are present.

Question 6

- The best approach for the adult patient with a calcified ductus is:

  A. Medical management.

  B. Closure of the defect at cardiac catheterization.

  C. Surgical closure of the defect utilizing cardiopulmonary bypass.

  D. Left thoracotomy and surgical closure.

Comment
The correct answer is B.

The calcified ductus in the adult must be handled very carefully. In the past, the treatment of choice was surgical closure, and, because of the pliability of the ductus, to have it done on cardiopulmonary bypass. Now, however, the capability of closing the ductus in the cath lab negates the need for surgery.

Question 7

- A 50-year-old Asian woman is seen because of the onset of palpitations for the past 24 hours. She had finished a 2-day bus ride just prior to the onset of the symptoms. She reports that for the past year, she has had to stop after one flight of stairs because of fatigue and shortness of breath. She denies chest discomfort. On questioning, she admits heavy alcohol intake.

  Physical examination reveals a healthy appearing woman with blood pressure 150/70 mmHg and pulse irregularly irregular at 140 per minute at the apex. The lungs are clear to auscultation and percussion. The neck veins are 8cm with a predominant V wave. S2 is widely split with little respiratory variation. There is no S3 or S4. There is a Grade III/VI systolic ejection murmur at the second interspace at the left sternal border. There is no peripheral edema.

  The chest x-ray is shown (Figure 1). The ECG shows atrial fibrillation with left axis deviation and right bundle branch block.

  The most probable diagnosis is:

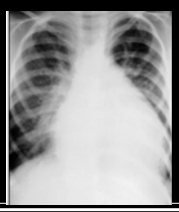
  A. Pulmonary emboli.

  B. Alcoholic cardiomyopathy.

  C. Ostium secundum atrial septal defect.

  D. Primary pulmonary hypertension.

  E. Ostium primum atrial septal defect.



Question 7 Answer

- **Comment**
  The correct answer is E.

  Patients with significant atrial septal defects often develop atrial fibrillation when they get older. The clue to the atrial septal defect on exam is the systolic ejection murmur at the base, accompanied by a widely split second heart sound. The chest x-ray, with increased pulmonary vascular markings, is typical. The ECG with left axis deviation is characteristic of ASD of the ostium primum type. The more common secundum ASD would be expected to be associated with right axis deviation.

Question 8

A 48-year-old man with known Eisenmenger's syndrome (ventricular septal defect with high pulmonary vascular resistance) is seen for his annual visit. He continues to work full-time as a computer operator. During the past year he has had chest discomfort "a few times." It's vague, visceral in character, and substernal in location. It may be at rest or with activity, and never lasts more than a few minutes. He has no gastrointestinal complaints and doesn't think this is related to eating. He has also had minimal hemoptysis with severe coughing paroxysms--"maybe when I had a little cold." He has also had vague ankle and toe aching periodically--"maybe I need a new style of shoe!"

His examination is not changed from previous years. He is well nourished and well muscled. He has mild clubbing, and minimally evident jugular veins with an A wave made more prominent by abdominal compression. He has a striking left peristernal lift. On auscultation, S2 is split, P2 is strikingly increased, and there is a Grade II decrescendo diastolic murmur along the left sternal border. There are several ejection clicks but no systolic murmur.

The electrocardiogram shows striking RVH with STT abnormalities (no change) and his hematocrit is 65 (has varied from 63 to 67 during the past few years).

Which of the following do you believe should be done in this patient?

A. Perform several careful phlebotomies in the next few weeks to reduce his hematocrit to less than 60.

B. Administer intravenous prostacyclin.

C. Refer to specialized center for work-up and consideration for future heart/lung transplant.

D. Begin home O2.

E. Commence anticoagulation with warfarin.

Question 8 Answer

Comment
The correct answer is C.

Patients with adult congenital heart disease now constitute a sizable population (many have had operative procedures) and are approaching a million patients in the United States. In this patient, and in all patients with complex congenital heart disease, serious consideration should be given to referring him to a specialized center where a broad comprehensive approach to these patients is available.

The patient should not be given phlebotomies. (A) Unless patients are clearly suffering from hyperviscosity syndrome (e.g., transient visual disturbances, headaches, paraesthesia, fatigue), phlebotomy should not be considered--certainly not with a hematocrit under 70%. Because the effects of phlebotomy are only transient and iron deficiency will result, an increased incidence of stroke occurs related to alter RBC shape with Fe deficiency. All patients with polycythemia should be reminded that they must avoid dehydration.

Options B and D are not appropriate for this patient since he is quite stable and the natural history of the Eisenmenger syndrome is extremely variable. Patients may remain stable for many years and survival into the late 50s and beyond is not rare in patients such as the one described here. Prostacyclin (B) has been used with some enthusiasm in patients with primary pulmonary hypertension but there are a variety of problems and complications associated with this treatment. In addition, data including patients with Eisenmenger syndrome are extremely limited and responses have been varied.

His symptoms are almost certainly "right ventricular angina," and it is possible that he does have small hemoptyses related to his pulmonary hypertension. These symptoms alone, in the absence of the development of right-sided failure, do not constitute strong evidence of significant progression of his pulmonary vascular disease.

His uric acid should be measured since he will have hyperuricemia due to his polycythemia, and gout occurs in cyanotic patients, particularly as they age. The use of warfarin (E) is controversial and ordinarily would not be given.

Question 9

Which one of the following is most consistent with the findings in the figure?

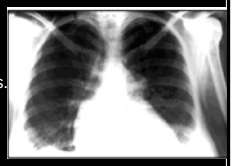
A. Congenital pulmonic stenosis.

B. Primary pulmonary hypertension.

C. Massive "saddle" pulmonary embolus.

D. Tetralogy of Fallot.

E. Patent ductus arteriosus.



Question 9 Answer

Comment
The correct answer is A.

Dilatation of the main pulmonary artery on chest x-ray without evidence of generalized pulmonary artery dilation is characteristically observed in congenital pulmonic stenosis and is believed to represent the phenomena of poststenotic dilatation. It can also be seen in idiopathic dilatation of the main pulmonary artery, in which an ejection sound and pulmonic flow murmur can also occur, making clinical differentiation from pulmonic stenosis difficult.

Primary pulmonary hypertension results in dilation of all proximal pulmonary artery branches with distal caliber reduction (pruned tree appearance). Patent ductus arteriosus would appear similar to primary pulmonary hypertension depending on the level of hypertension. Tetralogy of Fallot and saddle pulmonary embolus do not significantly change main pulmonary artery caliber.

Question 10

Which of the following conditions are amenable to repair by the Fontan operation?

A. Ostium primum defect.

B. Tetralogy of Fallot.

C. Tricuspid atresia.

D. Corrected transposition of the great vessels.

Comment
The correct answer is C.

The ability to connect the right atrium directly to the pulmonary artery and utilize a single ventricular chamber has revolutionized surgical care for conditions where there is essentially one ventricle, such as tricuspid atresia.

Question 11

A common difficult management problem following the Fontan operation is:

A. Unrelenting congestive heart failure.

B. Severe mitral regurgitation.

C. Progressive left ventricular dysfunction.

D. Atrial arrhythmia, particularly atrial flutter.

E. Ventricular tachycardia.

Comment
The correct answer is D.

Atrial arrhythmias are extremely common after the Fontan operation and probably relate to the effect of surgery done within the atrium. Atrial flutter can be a particularly difficult rhythm to control in these patients and may severely depress cardiac output.

Question 12

- In which of the following diseases is pregnancy difficult, but not highly risky to mother and fetus?

  A. Eisenmenger's syndrome.

  B. Primary pulmonary hypertension.

  C. Hypertrophic obstructive cardiomyopathy.

  D. Prior peripartum cardiomyopathy with heart failure.

  E. The Marfan syndrome with dilated aortic root.

Question 12 Answer

- **Comment**
  The correct answer is C.

  The cardiovascular system must be able to handle a doubling of cardiac output during pregnancy. Thus, cardiopulmonary diseases that obstruct blood flow are usually contraindications to pregnancy because both the mother and fetus get inadequate blood flow. Thus, obstruction to pulmonary flow due to the Eisenmenger reaction or primary pulmonary hypertension fits into this category, but hypertrophic cardiomyopathy does not. The increased cardiac output increases venous return to the left heart resulting in left ventricular enlargement and less obstruction. In fact, during pregnancy the murmur of hypertrophic obstructive cardiomyopathy may lessen or even disappear, causing the diagnosis to be missed.

  Prior peripartum cardiomyopathy with heart failure is a contraindication to pregnancy because of the high incidence of recurrent failure and death.

  Hormonal changes during pregnancy alter vascular walls, making them more distensible. This is a normal mechanism to adapt to higher cardiac output; however, in the patient with the Marfan syndrome and an enlarged aortic root, it can lead to increased wall stress and aortic rupture or dissection.

Question 13

- In long-term follow-up of patients after surgical repair of tetralogy of Fallot, the most common dysrhythmia observed is:

  A. Sinus bradycardia.

  B. Atrial fibrillation.

  C. Atrial tachycardia.

  D. Ventricular tachycardia.

  E. Junctional tachycardia.

Question 13 Answer

- **Comment**
  The correct answer is D.

- Complex ventricular arrhythmias often occur during long-term follow-up of patients with tetralogy of Fallot. The incidence correlates with age at repair and with higher residual postoperative right ventricular systolic and end-diastolic pressures. Sudden death accounts for a significant proportion of the late mortality among these patients. In patients with ventricular tachycardia, the site of origin is typically found to be in the right ventricular outflow tract related to the previous ventriculotomy and infundibular resection.

  Bundle branch block and AV block are also observed in some patients after repair of tetralogy of Fallot. Sinus bradycardia and atrial dysrhythmias are common problems found in long-term follow-up after surgical repair for transposition of the great vessels.

Question 14

- A 42-year-old man is referred for evaluation of a systolic murmur. Your exam shows normal carotid pulses, a prominent apical impulse, an early systolic sound, and a grade III/VI mid-systolic murmur at the base. Respiration did not change the character of these auscultatory findings. After an extrasystole, the systolic murmur increased in intensity. Handgrip did not alter the systolic murmur. Valsalva decreased the intensity of the murmur, and it returned to baseline intensity after seven heart beats.

  Which one of the following diagnoses is most likely?

  A. Congenital pulmonic stenosis.

  B. Innocent murmur.

  C. Mitral valve prolapse.

  D. Hypertrophic obstructive cardiomyopathy.

  E. Bicuspid aortic valve.

Question 14 Answer

- **Comment**
  The correct answer is E.

- A systolic murmur that increases in intensity in the beat following an extrasystole is usually due to turbulent flow out of the ventricles. Mitral regurgitation is less likely because this murmur does not change following an extrasystole. The murmur of hypertrophic obstructive myopathy usually decreases with handgrip exercise. An innocent flow murmur is less likely because of the presence of an early systolic sound and grade III intensity. With pulmonic stenosis, there are characteristic changes in the intensity of the murmur and the ejection sound during respiration. The ejection sound establishes the diagnosis of an abnormal aortic valve, a bicuspid valve being the most common abnormality.

Question 15

- A 30-year-old woman with inoperable cyanotic congenital heart disease is scheduled for total abdominal hysterectomy for uterine cancer. You are asked to see her for preoperative cardiac evaluation. Her hematocrit is 66%. This is unchanged from previous values over several years. The patient denies any bleeding tendencies or hyperviscosity symptoms such as fatigue, headache, or lethargy.

  Which of the following is most appropriate?

  A. The patient should not undergo phlebotomy.

  B. The patient should undergo phlebotomy preoperatively.

  C. The patient should undergo phlebotomy postoperatively.

  D. The patient should undergo phlebotomy on an ongoing basis, beginning preoperatively.

  E. The patient should undergo phlebotomy on an ongoing basis, beginning postoperatively.

Question 15 Answer

- **Comment**
  The correct answer is B.

- In patients with unoperated or palliated cyanotic congenital heart disease, secondary erythrocytosis is triggered by tissue hypoxia. This increases the hematocrit and improves the blood's oxygen-carrying capacity, however, symptoms of hyperviscosity may occur. Long-term cyanosis also affects platelets and coagulation parameters. This results in abnormal hemostasis manifested as increased bleeding tendency.

  For management of secondary erythrocytosis, phlebotomy is advised under two conditions. Phlebotomy is recommended for patients with symptoms of hyperviscosity who are not dehydrated and who have hematocrit values over 65%. Phlebotomy is also recommended prior to surgery in asymptomatic patients with hematocrit values over 65% in order to minimize surgical bleeding. Therefore, phlebotomy would be recommended preoperatively in this patient. Because this patient does not have hyperviscosity symptoms, postoperative phlebotomy or ongoing phlebotomy would not be recommended.

Question 16

- All but one of the following have a < 1% risk of maternal mortality during pregnancy. Which has a higher risk?

  A. Atrial septal defect.

  B. Patent ductus arteriosus.

  C. Mild mitral stenosis.

  D. Marfan syndrome.

  E. Mild pulmonic stenosis.

Question 16 Answer

- **Comment**
  The correct answer is D.

- Since pregnancy results in an approximately 50% increase in cardiac output, the major lesions with significant pregnancy-related cardiac mortality are those that obstruct the circulation, such as significant aortic stenosis or lesions where parts of the heart and vasculature are weakened such that the increased cardiac volumes and stroke volume could cause rupture of the structure. Such is the case with the aorta and Marfan syndrome, so that even patients with normal-sized aortas can experience marked aortic dilation and rupture during pregnancy. Lesions that are volume loads on the heart, such as atrial septal defect, patent ductus arteriosus, and valvular regurgitation are usually well tolerated during pregnancy with knowledgeable perinatal care. Mild stenotic lesions are also well tolerated.

Question 17

- Which of the following results in decreased pulmonary vascularity on chest x-ray?

  A. ASD with patent ductus arteriosus.

  B. ASD with restrictive VSD.

  C. ASD with tricuspid atresia and restrictive VSD.

  D. ASD with partial anomalous pulmonary venous drainage.

Question 17 Answer

- **Comment**
  The correct answer is C.

- ASD with tricuspid atresia and restrictive VSD. In tricuspid atresia, venous blood shunts across the ASD from right-to-left and mixes with arterial blood. Blood can reach the pulmonary circuit through the VSD, however this flow is limited because the defect is restrictive (i.e., small). The pulmonary system is undercirculated (Qp to Qs is < 1), and the patient is cyanotic. The other four answers all lead to left-to-right shunting. ASD, patent ductus arteriosus, VSD, and partial anomalous pulmonary venous drainage all overcirculate the pulmonary system.

Question 18

A 25-year-old Caucasian man presents complaining of chest discomfort, occurring intermittently for the past 2 months, at times severe. It lasts 5-10 minutes and is exacerbated by taking a deep breath or heavy lifting, which he frequently does as a warehouseman.

Family history: mother has hypertension.

Physical Examination: The patient is obese, weighing 220lb at 5' 6". BP 150/100 mmHg. P 74/min. Neck veins 5cm. Fundi: Narrowing of arterioles, no hemorrhages or A/V nicking. There is a prominent suprasternal pulsation. Lungs are clear to auscultation and percussion. PMI is sustained in the left 5th intercostal space in the midclavicular line. There is a systolic ejection click and a grade II/VI systolic ejection murmur at the 2nd intercostal space, left sternal border. The murmur can be heard in the back, loudest in the interscapular region to the left of the spine. There is a grade II/VI diastolic blowing murmur loudest in the 3rd intercostal space at the left sternal border. No S3 or S4 gallop. Pulses: Carotids 3+, brachials 3+, femorals 1+.

Laboratory: ECG: LVH. Chest X-ray: Normal-sized heart, prominent ascending aorta. Echo-Doppler: LV posterior wall 12mm and ventricular septal wall of 13mm, LV end diastolic diameter 4.8cm and estimated EF 55%. Turbulence in diastole under the aortic valve, which extends 3cm into the LV cavity, and a systolic jet across the aortic valve of 2.5 m/sec.

Which of the following is the most likely diagnosis?

A. Severe congenital valvular aortic stenosis.

B. Bicuspid aortic valve with severe aortic regurgitation.

C. Coarctation of the aorta.

D. Hypertrophic cardiomyopathy.

E. Essential hypertension with chest wall pain.

Question 18 Answer

Comment
The correct answer is C.

A is incorrect. The jet of 2.5cm is a gradient of only 25 mmHg.

B is incorrect. Although there is a bicuspid valve in up to 80% of patients with coarctation of the aorta, and the patient has an ejection click with an AR murmur, the pulse pressure is not wide and the LV end diastolic diameter is not increased, making severe chronic AR unlikely.

C is correct. The patient has coarctation of the aorta. He has a systolic murmur heard posteriorly and femoral pulses that are less palpable than the brachial pulses, both of which are highly likely in this age group to be consistent with aortic coarctation.

D is incorrect. Although there is an aortic systolic ejection murmur, a systolic gradient across the aortic valve, and LVH by ECG and echo, there is no asymmetric hypertrophy, and AR is rare with HOCM.

E is incorrect, even with a family history of hypertension, since there is a better likelihood of another etiology for the hypertension. He did come in with musculoskeletal chest pain.