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Pediatric Cardiology/Cardiac Surgery

Hot Topics in Pediatric Cardiology

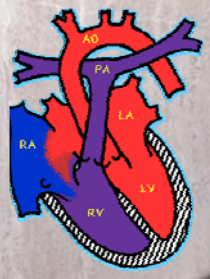
- Review of Major Defects
- Fetal Diagnosis of Congenital Heart Disease
- Neonatal Diagnosis of CHD
- Treatment in Cath Lab
- Heart Surgery Outcomes
- Primary Care Issues after Heart Surgery
- Adults with CHD
- Pregnant women with CHD

Non-cyanotic defects

- Don't get blue,
- May have trouble gaining weight
- May have congestive heart failure and pulmonary overcirculation
- May have exercise/ exertion difficulties

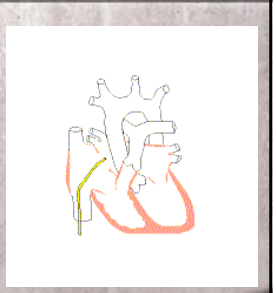
Atrial Septal Defect

- Often doesn't show up for 1 yr.
- Pulmonary Vascular DZ risk 10% if not corrected
- Usually corrected at age 3-4



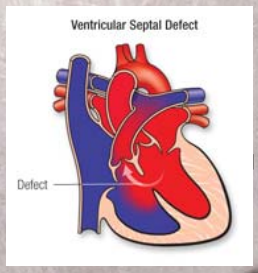
Treatment in Cath Lab

ASD Device Closure

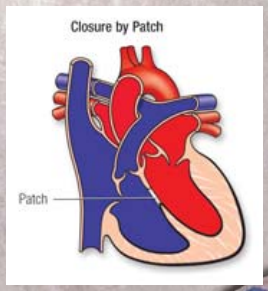


Ventricular Septal Defect

- Effect of VSD depends on Size, Location, PVR
- Moderate to Moderately severe often no S/S until 6-8 wks of age because of high PVR of newborn

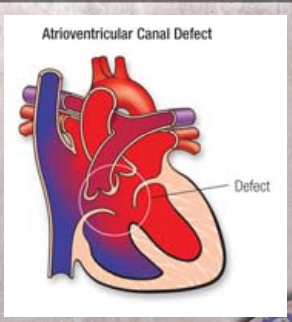


VSD Closure

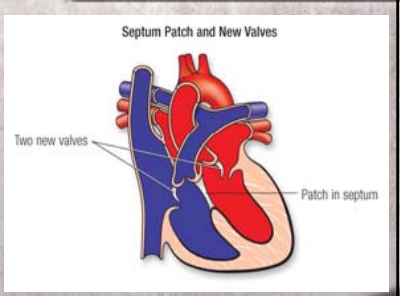


Atrioventricular Canal

- Often have Downs Syndrome
- Partial or Complete Types
- Usually have large Left to Right shunts

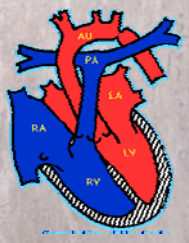


AV Canal Repair



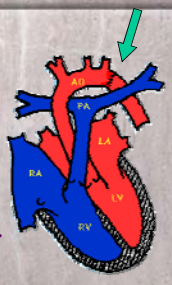
Coarctation of the Aorta

- Severe coarctation is dependant on ductus for systemic blood flow—shock when DA closes
- Can be diagnosed much later in less severe cases



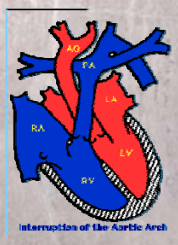
Coarctation Repair

- Resection of narrowing with patch augmentation in some cases for initial coarct
- For restenosis balloon aortoplasty (cath lab)
- Hypertension not uncommon after and before repair
- Side effects of surgery r/t cross-clamp time (below clamp ischemia)



Interruption of the Aortic Arch

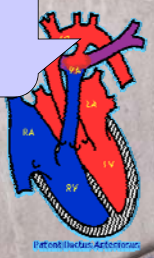
- Severe form of coarctation with no continuation of the aortic arch to the descending aorta.
- If the ductus closes the child has no flow to the lower extremities and may become severely ill and die.



Patent Ductus Arteriosus

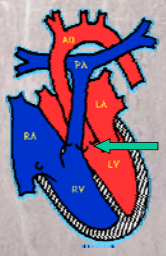
- Hard on little premies
- Big DA (term baby) big failure, can be disastrous
- Endocarditis risk

Flow from the Aorta to Branch PA's, floods lungs

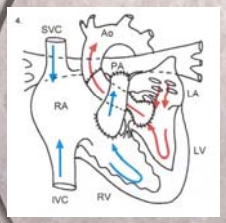


Aortic Stenosis

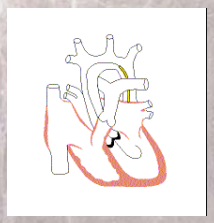
- Aortic valve is often small as well as stenotic, may be leaky (regurgitant).
- The LV has to work progressively harder and becomes hypertrophied.
- At risk for ventricular arrhythmias



Aortic Valve Repair



Ross Procedure



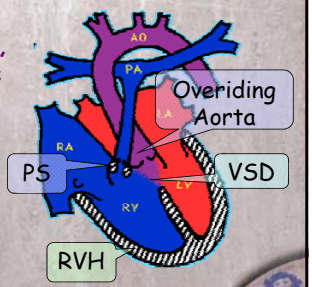
Balloon Valvuloplasty

Cyanotic Defects

- Are blue, get bluer with cry, exertion
- Can grow ok sometimes
- If not repaired-At greater risk of
  - endocarditis
  - Stokes/abcesses
  - Immune problems

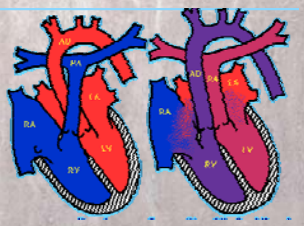
Tetrology of Fallot

- Often not cyanotic at birth, becomes cyanotic over time especially with crying.
- Usually repaired surgically 3-6 months of age

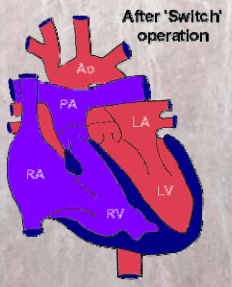


D-Transposition of Great Arteries

- Great arteries reversed, not compatable with life
- May have VSD, PFO, DA for mixing

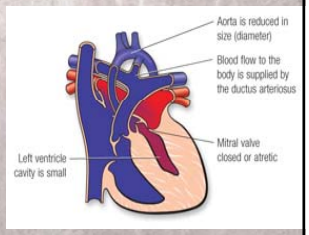


Arterial Switch

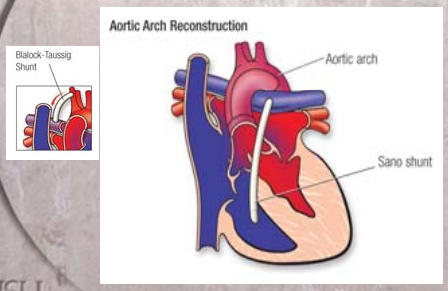


Hypoplastic Left Heart Syndrome

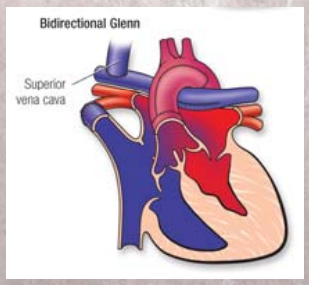
- Often the LA, mitral valve, aortic valve, and aortic arch are also small.
- Dependant on the Ductus to maintain perfusion, shocky and acidotic when closes



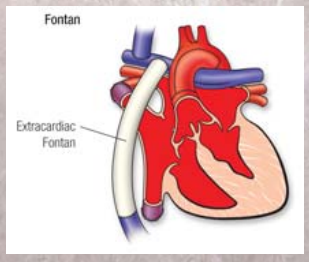
HLHS Paliation



Glenn Anastamosis (Stage II ~6mos of age)

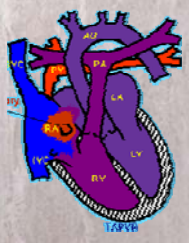


Fontan Procedure (Stage III ~ 2-4 years of age)



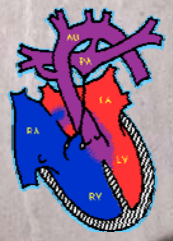
Total Anomalous Pulmonary Venous Return

- Can have supra-diaphramatic or infra-diaphragmatic drainage, if obstructed quite cyanotic



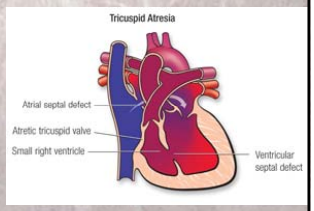
Pulmonary Atresia with VSD

- Most severe end of TOF spectrum
- Cyanotic @ birth
- Ductal Dependant for pulmonary blood flow
- Often poor pulmonary branch tree development

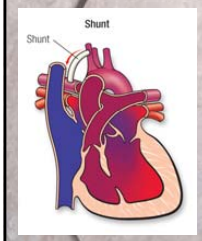


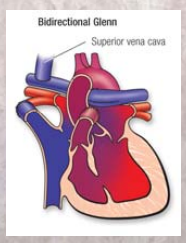
Tricuspid Atresia

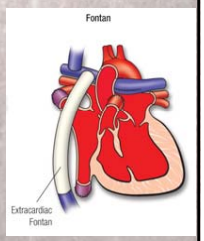
- Hypoplastic right heart
- 50% have transposition of great arteries
- Cyanotic early



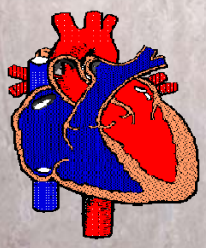
Tricuspid Atresia Palliation

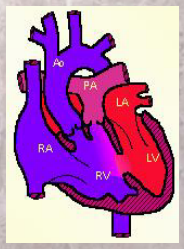






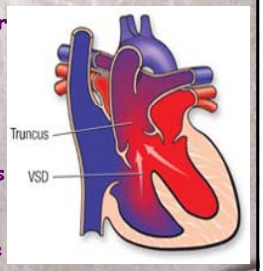
Double Outlet Right Ventricle



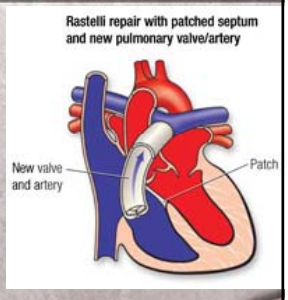


Truncus Arteriosus

- Fetal truncal artery never completed dividing into the aorta and pulmonic artery.
- Various types
- If pulmonary artery takeoff narrow then lungs protected
- Otherwise-pulmonary overflow and vasc disease

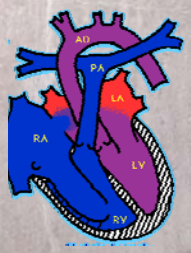


Truncus repair



Ebsteins Anomaly

- Tricuspid valve displaced low in RV
- Very regurgitant
- Shunting R→L thru Foramen Ovale
- Sometimes inadequate pulm flow without ductus
- SVT



Fetal Diagnosis

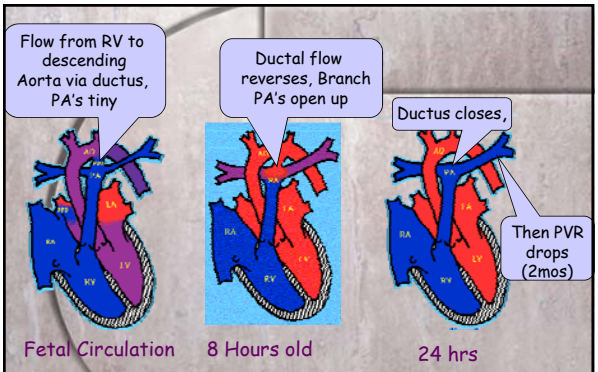
Fetal Echocardiograms

- Some CHD easier to diagnose via fetal echo
  - HLH,HRH, other single ventricles
  - Ebstein's anomaly
  - AV canal
- More difficult to diagnose
  - TGA, TAPVR, COARCT, VSD, ASD, TETS
- Impossible—
  - Ductus, small VSD, ASD, mild valve problem
  - In Oregon ~ 60% of children who need neonatal surgery were diagnosed in utero

Neonatal Diagnosis

Nursery is the ideal time to diagnose congenital heart disease in order to assure early appropriate care (if not prenatally diagnosed)

- Many problems very subtle in early NB period
  - Some present after ductus closes (8-48? Hours)
  - Some present when PulmonaryVascular Resistance drops (2-6 weeks)
  - Some very minor findings won 't be obvious for years (minor coarct, ASD, bicusp AV)



Neonatal Diagnosis

- 50 % of babies with murmur in first few days of life have CHD
- 25 % of babies with murmur at 6 weeks have CHD
- Diagnoses most likely to lead to death soon after discharge: HLH, IAA, Coarctation (they look pink until ductus closes)
- Some get irreversible pulmonary vascular disease and can't be repaired- shortened life

Neonatal Diagnosis

When to get consult on a newborn

- Pathological Murmur
- Cyanosis (sats less than 95)
- Poor pulses/perfusion

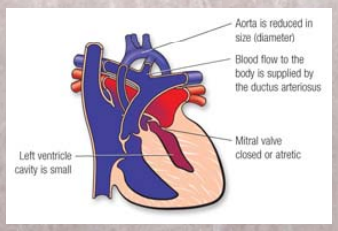
Case present

- Term Baby born in southern Oregon, went home day 2. On day 5 mom noted fussiness, appearing pale and cold and decreased feeding. Mom states that his stools on day of presentation were "frothy" in appearance, a decreasing PO intake,
- Mom also describes increased work of breathing for two days leading to presentation.

- Went to pcp, he was noted to have poor perfusion (cool extremities & faint pulses) and cyanosis (saturation 84).
- Sent to ED nearby tele echo showed hypoplastic left heart syndrome

HLHS

- What caused symptoms to appear?

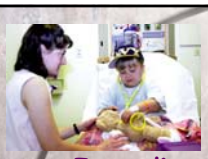


Scenario continued

- Child started on prostaglandin,
- Transported to DCH
- Had Norwood procedure and was eventually discharged home with Interstage Monitoring

Interstage Monitoring

- For children with single ventricle or very severe palliated anatomy
- Mortality between initial surgery and 2nd stage ~ 16%
- To reduce this we send home with
  - OXYGEN SATURATION MONITOR
  - BABY SCALE
  - GUIDELINES TO CALL US



Primary Care Issues

- Immediate post-op complications
- vaccines
- Synagis
- Endocarditis prophylaxis
- Dental care
- Neuro-developmental issues

Primary Care Issues

Post-op Medical Visit

- Assess wound healing, pain, feeding
- Assess medications compliance/complications (Digoxin, Lasix, Aldactone, Enalapril, sildenafil, propranol,).
- Assess ability to obtain medications
- Assess for arrhythmias, post-cardiotomy syndrome.

Vaccines postop

- Can restart regular vaccines except:
- No live virus 6 mo after surgery if received blood.
- Should get flu vax if older than 6 months

Synagis/Numax

Palivizumab (Synagis) Recommendations

- Give to children under 2 years of age with serious congenital heart disease (Nov-May)
  - Cyanotic heart disease
  - Acyanotic disease requiring medications
  - Administer next dose when medically stable following surgery
    - Adjust monthly timetable accordingly

Endocarditis prophylaxis

Risks for Endocarditis

High---Need prophylaxis for life

- Prosthetic Valves
- Complex cyanotic CHD (Tet, TGA, single ventricle)
- Shunts & Conduits
- Epicardial pacers

Moderate—need prophylaxis for 6 months after surgery

- All other congenital heart surgery

Endocarditis prophylaxis

Procedures with Endocarditis Prophylaxis Recommended

- Dental extraction
- Cleaning
- T &A, Bronch w\ rigid

Not Recommended

- Vaginal Delivery
- C-section, Hysterectomy
- Ear tubes, intubation

Endocarditis prophylaxis

- Amoxicillin 50mg/kg po 1hr before procedure
- For Penicillin Allergic or GI/GU procedures see recommendations
- NO 6 hr post procedure dose anymore

Dental Care

- Good dental hygiene essential,
- Out of 5 outpatient endocarditis admits in last 5 years
  - 4 multiple caries with underlying CV dx
  - 1multiple piercings of risky nature with underlying CV dx
  - None related to dental procedures
  - We try to bring it up at visits
  - Getting dental care hard in Oregon
    Especially if need sedation

Neurodevelopment

Boston study: circ arrest vs bypass in neonatal heart surgery

- At 8 years old—Both groups had ↓academic, fine motor, visual spatial, attention and higher order thinking than expected for general population. 1/3 in special ed
- TCA-worse manual dexterity, apraxia, V-M tracking, Handwriting
- Low flow bypass--↑impulsiveness, worse behavior

- These findings have been duplicated with many different heart infant surgeries

Bellinger et al. *J Thoracic Cardiovascular Surgery* 2003

Adults with CHD

Adults with Congenital Heart Disease

- There are as many adult congenitals as pediatric congenital heart patients now
  - Often not in any cardiac care
    - Thought they were fixed
    - Often don't understand heart disease, parents dealt with it.
    - Few specialists who know disease (adult cards-no training in CHD)
    - Insurance issues
- Very few truly "fixed"

Pregnant Women with CHD

Canadian Study

- 562 pregnant women with CHD/13 Canadian Hospitals. Minor to severe. 28% had either maternal and/or neonatal event
- Most common maternal events were arrhythmias and pulmonary edema, 4 CVA's, 3 deaths
- Most common neonatal events-prematurity, SGA, 15 fetal or neonatal deaths. 7% CHD

Siu S, Circ 104 2001

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Surgical Repair of Tetrology

- Definitive repair: patch closure of VSD, resection of sub-PS, patch enlargement of RVOT and ± main pulmonary artery
- Post surgical risks for arrhythmias and heart blocks in addition to usual open heart surgery complications