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### Hot Topics in Pediatric Cardiology

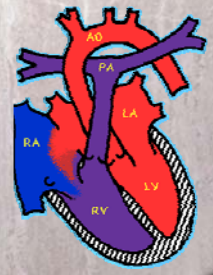
- Review of Major Defects
- Fetal Diagnosis of Congenital Heart Disease
- Neonatal Diagnosis of CHD
- Treatment in Cath Lab
- Heart Surgery Outcomes
- Primary Care Issues after Heart Surgery
- Adults with CHD
- Pregnant women with CHD

### 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

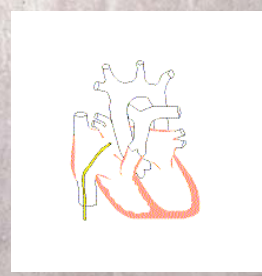
### Atrial Septal Defect

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



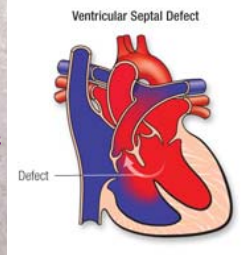
### Treatment in Cath Lab

ASD Device Closure



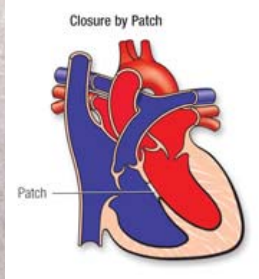
### Ventricular Septal Defect

- 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

Closure by Patch



Patch

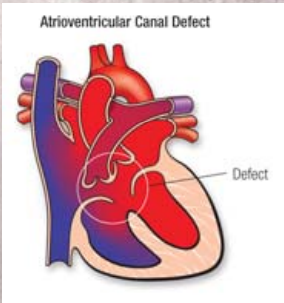
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Detailed description: A diagram of the heart showing a ventricular septal defect (VSD) being closed with a patch. The patch is a blue area placed over the hole in the septum between the right and left ventricles. Labels include 'Closure by Patch' and 'Patch'.

### Atrioventricular Canal

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

Atrioventricular Canal Defect



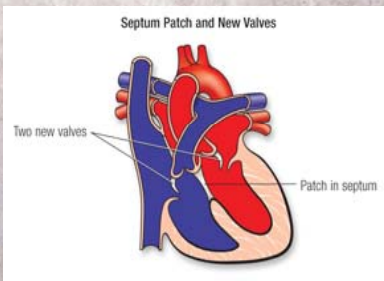
Defect

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Detailed description: A diagram of the heart showing an atrioventricular canal defect. A line points to a hole in the atrial septum. Labels include 'Atrioventricular Canal Defect' and 'Defect'.

### AV Canal Repair

Septum Patch and New Valves



Two new valves

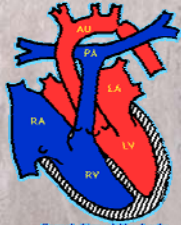
Patch in septum

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Detailed description: A diagram of the heart after AV canal repair. Two new valves are shown in the atrioventricular junctions, and a patch is visible in the septum. Labels include 'Septum Patch and New Valves', 'Two new valves', and 'Patch in septum'.

### 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

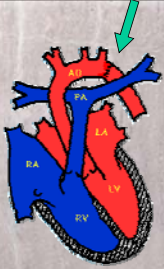


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Detailed description: A diagram of the heart showing a narrowing (coarctation) in the aorta. Labels include 'RA', 'PA', 'LA', 'LV', and 'RV'.

### 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)

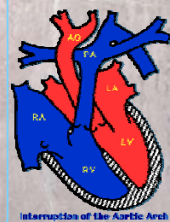


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Detailed description: A diagram of the heart showing the site of coarctation repair. A green arrow points to the area where the narrowing was resected and replaced with a patch. Labels include 'RA', 'PA', 'LA', 'LV', and 'RV'.

### 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.



Interruption of the Aortic Arch

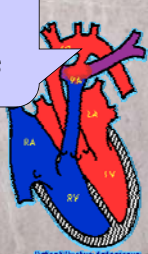
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Detailed description: A diagram of the heart showing a complete interruption of the aortic arch. Labels include 'RA', 'PA', 'LA', 'LV', and 'RV'.

### Patent Ductus Arteriosus

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

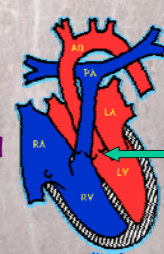
Flow from the Aorta to Branch PA's, floods lungs



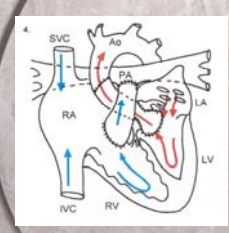
Patent Ductus Arteriosus

### Aortic Stenosis

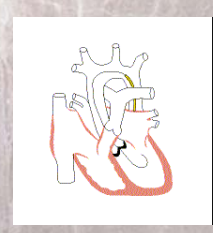
- Aortic valve is often small as well as stenotic, may be leaky (regurgitant).
- The LV has to work progressively harder and becomes hypertrophied.
- At risk for ventricular arrhythmias



### Aortic Valve Repair



Ross Procedure



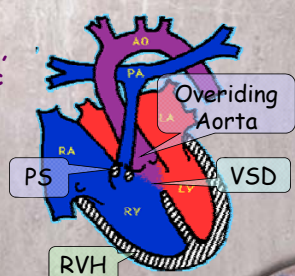
Balloon Valvuloplasty

### Cyanotic Defects

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

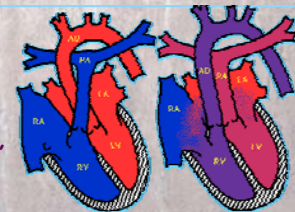
### Tetralogy of Fallot

- 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 compatible with life
- May have VSD, PFO, DA for mixing





### Arterial Switch

After 'Switch' operation

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### 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

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### HLHS Paliation

Aortic Arch Reconstruction

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### Glenn Anastamosis (Stage II ~6mos of age)

Bidirectional Glenn

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

Fontan

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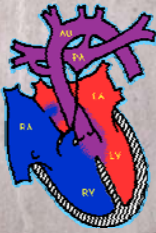
### Total Anomalous Pulmonary Venous Return

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

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### Pulmonary Atresia with VSD

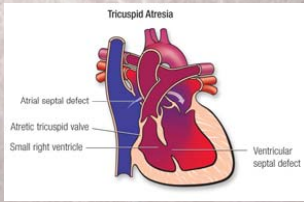
- Most severe end of TOF spectrum
- Cyanotic @ birth
- Ductal Dependant for pulmonary blood flow
- Often poor pulmonary branch tree development



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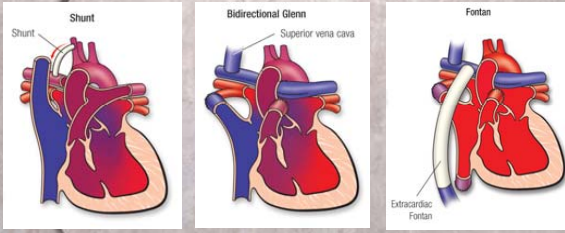
### Tricuspid Atresia

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



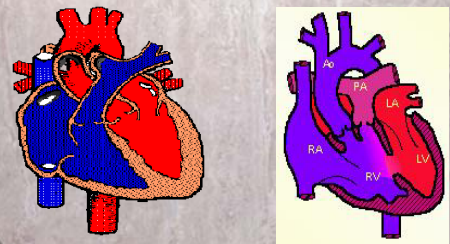
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### Tricuspid Atresia Palliation



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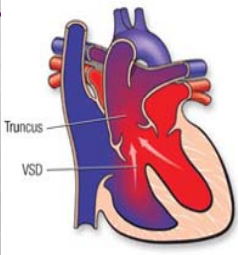
### Double Outlet Right Ventricle



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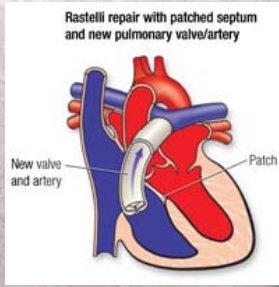
### Truncus Arteriosus

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



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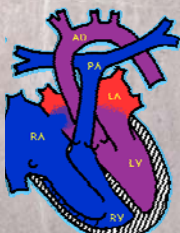
### Truncus repair



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### Ebsteins Anomaly

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



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### 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

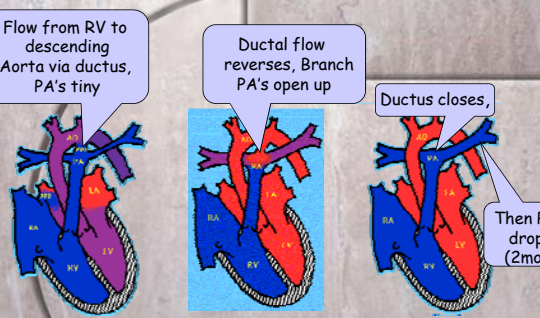
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### Neonatal Diagnosis

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, bicuspid AV)

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Flow from RV to descending Aorta via ductus, PA's tiny

Ductal flow reverses, Branch PA's open up

Ductus closes, Then PVR drops (2mos)

Fetal Circulation      8 Hours old      24 hrs

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### 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

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### Neonatal Diagnosis

**When to get consult on a newborn**

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



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### 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.

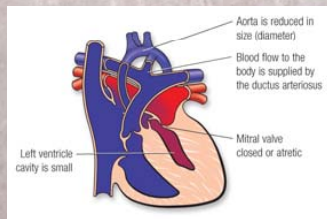
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- Went to pcp, he was noted to have poor perfusion (cool extremities & faint pulses) and cyanosis (saturation 84).
- Sent to ED nearby tele echo showed hypoplastic left heart syndrome

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### HLHS

- What caused symptoms to appear?



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### Scenario continued

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

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### Interstage Monitoring



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

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### Primary Care Issues



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

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### Primary Care Issues

#### Post-op Medical Visit

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

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### 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

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### 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

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### 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

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### 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



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## 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

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## Dental Care

- Good dental hygiene essential,
- Out of 5 outpatient endocarditis admits in last 5 years
  - 4 multiple caries with underlying CV dx
  - 1 multiple 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



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## 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

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## 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"

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## 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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- Kumar RK Comparison of outcome when hypoplastic left heart syndrome and transposition of the great arteries are diagnosed prenatally versus when diagnosis of these two conditions is made only postnatally. - *Am J Cardiol* 1999; 83
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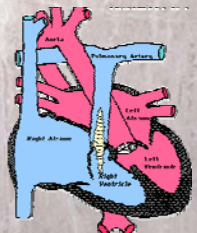
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### Surgical Repair of Tetralogy

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



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