UNDERSTANDING COMMON CONGENITAL HEART DEFECTS

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Learning Objectives

- Define epidemiology, risk factors, and pathophysiology of common congenital heart defects (ASD, VSD, PDA, Tetralogy of Fallot, and Aortic Coarctation).
- Discuss initial presentation and diagnostic work-up of ASD, VSD, PDA, Tetralogy of Fallot, and Aortic Coarctation.
- Describe therapeutic management and complications of ASD, VSD, PDA, Tetralogy of Fallot, and Aortic Coarctation.
Figure 2: Total CHD Birth Prevalence Over Time

Time course of reported total congenital heart disease (CHD) birth prevalence from 1930 until 2010. The blue line shows the time trend, and the squares represent the calculated birth prevalence values for each time period.
General Information

Figure 5

Birth Prevalence of CHD Subtypes

Reported birth prevalence of the 8 most common CHD subtypes per continent. Distribution of subtypes within total CHD is mentioned as percentages above bars.

*Reported PS and TOF birth prevalence in Asia was significantly higher than in Europe (p < 0.001) and North America (p < 0.001). †Reported Coarc birth prevalence in Asia was significantly lower than in Europe (p < 0.001). ‡Reported TGA and AoS birth prevalence in Asia was significantly lower than in Europe (p < 0.001), North America (p < 0.001) and Oceania (p < 0.001). §No data on TOF and AoS birth prevalence in Africa were available. Abbreviations as in Figure 4.
Patent Ductus Arteriosus (PDA)
PDA: Epidemiology & Risk Factors

- Incidence 1/2000 to 1/5000 live births
- Female to male 2:1
- Risk factors: Prematurity, prenatal infection, (maternal rubella 1st 4 weeks gestation), Trisomy 21, family Hx (parent), valporate, low oxygen tension at delivery (asphyxia), high altitude.
Normal Fetal and Neonatal Circulation

http://www.pted.org/?id=fetal1
Pathophysiology

L→R shunt

- ↑pulmonary blood flow
- ↑LVEDP (volume overload)
- ↑PV mmHg

- Diastolic runoff/hypotension
  - ↓ perfusion
  - Subendocardial Ischemia
    - LV Dysfunction
      - ↓ CO

- Hypoxemia
- Pulmonary edema

Effect of L-to-R Shunt on Cardiac Output

<table>
<thead>
<tr>
<th>Shunt</th>
<th>↑LV output</th>
<th>Qp:Qs</th>
</tr>
</thead>
<tbody>
<tr>
<td>25%</td>
<td>1.3x</td>
<td>1.3:1</td>
</tr>
<tr>
<td>50%</td>
<td>2x</td>
<td>2:1</td>
</tr>
<tr>
<td>75%</td>
<td>4x</td>
<td>4:1</td>
</tr>
<tr>
<td>80%</td>
<td>5x</td>
<td>failure</td>
</tr>
</tbody>
</table>
Clinical Presentation

Premature:
- No lung disease (birth weight >1500g) – PDA typically close near term,
- Recovering lungs (Birth weight 1000-1500g)
- Lung disease (Birth weight <1000g) – vent dependent (shunt ↑ vent days)

Small PDA (QP:QS <1.5:1):
- Silent PDA, found on routine ECHO, MRI, CT
- Asymptomatic detected by murmur
- Benign unless endarteritis occurs

Moderate PDA (QP:QS 1.5-2.2:1):
- Progressive pulmonary arterial changes → Pulmonary HTN
- LA/LV dilation → CHF
- Present in 3rd decade

Large PDA (QP:QS >2.2:1):
- Typically present weeks - months after birth
- May present as FTT, feeding difficulties, frequent URI’s, and CHF
Physical Exam

- Premature: Systolic murmur extends to diastole as PVR↓
- Small - Grade 2-3 continuous machinery-like murmur best at left infraclavicular radiates to LUSB and left interscapular
- Moderate - Grade 4 Murmur with palpable thrill, widened pulse pressure, bounding pulses, displaced PMI
- Large - Above findings + 3rd heart sound with apical diastolic rumble
- Elevated PAP: Diastolic murmur disappears 1st then PDA murmur, PR and TR murmur develop with split S2 and loud pulmonary portion
PDA: Diagnosis

- ECG: Normal (small) vs. LAE/LVH (mod-large) vs. RAD/RVH (irreversible PH)
- CXR: Normal (small), Cardiomegaly, increased pulmonary vascular markings, prominent ascending aorta, LA, LV (mod-large), Duct calcification in adult patients
- ECHO: quantify flow, direction, anatomy (R→L or bidirectional = increased PAP). Small – normal to mild enlargement. Moderate to large shunt: dilated LA/LV
- CT or MRI may been needed in adults patients
Management

Premature:

- Supportive care: Mechanical Ventilation, Surfactant, Vasopressors, Diuretics, Fluid restriction
- Indomethacin
- Surgical Ligation/division (left thoracotomy)
  - Indications: Medical failure, heart failure, PHTN
  - Contraindications: Cyanotic heart defect (ductal dependent)
  - Complications: Laryngeal or phrenic nerve injury, chylothorax

Term & Adult:

- Spontaneous closure: delayed 3 months, after 3 months rate 0.6%/yr
- Small shunt without HF monitor every 3-5 years
- Indications: Volume overload (LA/LV dilation), reversible PHTN, Endarteritis
- Therapeutic catheterization: treatment of choice in Adults
PDA: Complications & Prognosis

Premature:
- ↑ morbidity & mortality
- Prolonged ventilatory support → BPD
- Association with NEC, IVH, Renal Failure

Term & Adult:
- CHF & Pulmonary HTN
- Endarteritis
Atrial Septal Defect (ASD)
Epidemiology & Risk Factors

- Prevalence 1.64/1000 live births
- Female:Male 2:1
- Most cases are sporadic
- RF: Holt-orm syndrome (heart-hand), trisomy 21, family hx
- Associated defects: AVSD, T/PAPVR, MV prolapse
Development of the Atrial Septum

A
- Septum primum
- Ostium primum
- Endocardial cushion

A*
The septum primum grows caudally from the superior portion of the common atrium towards the endocardial cushion.

B
- Septum primum

B*
The septum primum fuses with the endocardial cushion closing the ostium primum.

C
- Septum secundum
- Ostium secundum

C*
A second orifice, ostium secundum, develops within the septum primum.
A second septum, septum secundum, develops on the right atrial side of the septum primum.

D
- Septum secundum
- Ostium secundum
- Septum primum

D*
The septum secundum covers the ostium secundum but does not completely divide the atria.

E
- Foramen ovale

E*
The persisting orifice within the septum secundum is the foramen ovale. In approximately 70 to 75 percent of individuals, the septa fuse by age two closing the foramen ovale. In the remaining 25 to 30 percent, the foramen ovale remains patent.

- **Septum primum**
- **Septum secundum**
Pathophysiology

- L-to-R shunt (↑ w/ age)
  - ↑ Pulm circulation
    - ↑ PV return
      - ↑ PV mmHg
      - Pulmonary Edema
        - ↑ LVEDP
          - RA/RV dilation
            - Chronic dilation
              - Atrial arrhythmias
              - CHF
                - ↑ Pulm circulation
                  - ↑ PV return
                    - ↑ PV mmHg
                      - Pulmonary Edema
                        - ↑ PAP progressive
                          - ↑ RV mmHg
                            - RVH
                              - ↑ RA mmHg
                                - Shunt flow reversal
                                  - Eisenmenger Syndrome
Clinical Presentation

- Asymptomatic Murmur
- Recurrent respiratory infections and FTT
- CHF (rare during infancy, typically 30-40’s)
- Atrial arrhythmia (flutter or fibrillation) >40yrs
- Cryptogenic stroke/Paradoxical embolism
- Symptoms: Palpitations, dyspnea, exercise intolerance, fatigue
- Exam:
  - Loud first heart sound (wider opening of tricuspid valve from overload)
  - 2nd sound wide and fixed split
  - Symptomatic - soft (grade 1-2) crescendo-decrescendo mid systolic murmur over LUSB (increased pulmonary valve flow)
  - Soft mid-diastolic murmur LLSB (tricuspid flow, very subtle)
  - PHTN: PR and TR murmur
  - CHF: hepatomegaly, JVD, rales
  - Classic: Systolic ejection murmur at LUSB with wide fixed splitting of left heart sound
Diagnosis

- ECG: NSR, Atrial flutter/fib, First degree AV block, incomplete RBBB, RVH, RAE, If LAD likely ostium primum (from incompetent left AV valve)
- CXR: cardiomegaly, enlarged RA/RV, increased pulmonary vascular markings,
- ECHO: size, location, degree of shunting, associated defects
- MRI: helpful to define PAPVR
- Catheterization: interventional closure
Spontaneous Closure

250 patients

50 excluded

40 CHF

7 ↓<3mm

200

4-5mm
81 (40%)

6-7mm
56 (28%)

8-10mm
41 (21%)

>10mm
22 (11%)

Hanslik 2006
ASD Growth

- Small 3-6mm: 34 (33%)
  - 7 ↑ moderate
  - 3 ↑ large
  - 27 (26%); 3 SC
  - 0.63mm/yr

- Moderate 6-12mm: 40 (38%)
  - 3 ↓ small
  - 32 (31%)
  - 0.67mm/yr

- Large >12mm: 30 (29%)
  - 12 ↑ large
  - 45 (43%)
  - 1.44mm/yr

McMahon 2002
Management

Medical Management:
- Watch and wait (spontaneous closure)
- Symptom relief with Diuretics and afterload reduction (controversial)

Elective Closure
- Moderate to large asymptomatic ASD closure recommended 4-5 years

Surgical/Interventional Closure:
- QP:QS < 1.5:1 – Typically <5mm, if no RV overload, no need to close
- QP:QS 1.5:1 to 2:1 – closure indicated based on signs of overload
- QP:QS >2:1 – closure to prevent PHTN, CHF, Arrhythmias

Adult AHA guidelines:
- ASD closure w/ or w/o sx’s if RA or RV enlargement (class I)
- Primum or sinus venosus ASD surgical closure (class I)
- Reasonable to close for paradoxical embolism (class IIa)
- Small ASD (<5mm) without RV overload, no need to close unless embolism
ATRIAL SEPTAL DEFECT WITH AN INDICATION FOR CLOSURE

Secundum septal defect

All Must Be Present:
- Defect size 36 mm or less
- Adequate rim
- Safe distance from adjacent structures
- Adequate atria to device size
- Absence of associated pathology requiring cardiac surgery.

Percutaneous or Surgical closure

Any May Be Present:
- Defect size >36 mm
- Inadequate rim
- Significant proximity to adjacent structures
- Inadequate atria to device size
- Failed percutaneous closure
- Associated pathology requiring cardiac surgery.

Surgical closure

- Primum septal defect
  - Sinus venosus septal defect

Surgical closure

Vasquez 2013
Procedural Differences

Surgery:
- Median sternotomy, atriotomy, cardiopulmonary bypass, direct suture vs. pericardial/Dacron patch

Interventional catheterization:

Complications

Un-repaired ASD

- Paradoxical embolism
- Pulmonary Hypertension
- CHF
- Arrhythmia

Table III. Severe transient complications

<table>
<thead>
<tr>
<th></th>
<th>Group A (533 patients)</th>
<th>Group B (751 patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe anemia—blood</td>
<td>33 (6.1)</td>
<td>/</td>
</tr>
<tr>
<td>transfusion</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pneumothorax requiring</td>
<td>6 (1.1)</td>
<td>/</td>
</tr>
<tr>
<td>surgical drainage</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pleural effusion</td>
<td>5 (1)</td>
<td>/</td>
</tr>
<tr>
<td>requiring surgical</td>
<td></td>
<td></td>
</tr>
<tr>
<td>drainage</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>13 (2.4)</td>
<td>1 (0.2)</td>
</tr>
<tr>
<td>requiring surgical</td>
<td></td>
<td></td>
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<tr>
<td>drainage</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transient heart failure</td>
<td>1 (0.2)</td>
<td>/</td>
</tr>
<tr>
<td>Transient AV block</td>
<td>3 (0.6)</td>
<td>/</td>
</tr>
<tr>
<td>requiring pacemaker</td>
<td></td>
<td></td>
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<tr>
<td>Reoperation</td>
<td></td>
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<tr>
<td>Severe bleeding</td>
<td>2 (0.4)</td>
<td>/</td>
</tr>
<tr>
<td>Patch detachment</td>
<td>1 (0.2)</td>
<td>/</td>
</tr>
<tr>
<td>Arrhythmias</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Atrial fibrillation—DC</td>
<td>10 (1.8)</td>
<td>4 (0.5)</td>
</tr>
<tr>
<td>shock</td>
<td>/</td>
<td></td>
</tr>
<tr>
<td>Ventricular fibrillation</td>
<td>1 (0.2)</td>
<td>/</td>
</tr>
<tr>
<td>Thrombus formation on</td>
<td>/</td>
<td>3 (0.4)</td>
</tr>
<tr>
<td>the left side of the</td>
<td></td>
<td></td>
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<tr>
<td>device</td>
<td>/</td>
<td>/</td>
</tr>
<tr>
<td>Malposition/embolization</td>
<td>/</td>
<td>14 (1.9)</td>
</tr>
<tr>
<td>needing surgery and</td>
<td></td>
<td></td>
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<tr>
<td>surgical ASD closure</td>
<td>/</td>
<td></td>
</tr>
<tr>
<td>Vascular injury of the</td>
<td>/</td>
<td>4 (0.5)</td>
</tr>
<tr>
<td>femoral vessels</td>
<td>/</td>
<td></td>
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<tr>
<td>Surgery due to left</td>
<td>/</td>
<td>2 (0.2)</td>
</tr>
<tr>
<td>atrium</td>
<td>/</td>
<td></td>
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<tr>
<td>free wall and aortic</td>
<td>/</td>
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<tr>
<td>perforation with</td>
<td>/</td>
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<tr>
<td>pericardial effusion with</td>
<td>/</td>
<td></td>
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<tr>
<td>or without cardiac</td>
<td>/</td>
<td></td>
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<tr>
<td>tamponade</td>
<td>/</td>
<td></td>
</tr>
<tr>
<td>Seizures</td>
<td>1 (0.2)</td>
<td>/</td>
</tr>
<tr>
<td>Systemic thromboembolism</td>
<td>1 (0.2)</td>
<td>/</td>
</tr>
<tr>
<td>with stroke</td>
<td>/</td>
<td>/</td>
</tr>
<tr>
<td>Cardiac failure</td>
<td>1 (0.2)</td>
<td>/</td>
</tr>
<tr>
<td>Thrombus on the atriotomy</td>
<td>8 (1.4)</td>
<td>/</td>
</tr>
<tr>
<td>treated with anticoagulant</td>
<td></td>
<td>/</td>
</tr>
<tr>
<td>Total</td>
<td>86 (16)</td>
<td>27 (3.6)</td>
</tr>
</tbody>
</table>

Butera 2006
Long term prognosis

Late post-operative complications

<table>
<thead>
<tr>
<th></th>
<th>Death</th>
<th>SVT</th>
<th>Pacemaker</th>
<th>Re-operation</th>
<th>RV dilation</th>
<th>↓RVEF</th>
<th>Stroke</th>
<th>HF</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>5 (4%)</td>
<td>14 (16%)</td>
<td>6 (6%)</td>
<td>2 (1.5%)</td>
<td>11 (20%)</td>
<td>17 (31%)</td>
<td>1</td>
<td>0</td>
</tr>
</tbody>
</table>

Cuypers 2013
Ventricular Septal Defect (VSD)
Epidemiology & Risk Factors

- Prevalence 3.94/1000
- Most common congenital heart defect (40%)
- RF: Untreated maternal metabolic disease (phenylketonuria, gestational diabetes), Parental hx, prematurity, Chromosomal abnormalities, Holt-Oram syndrome
- Solitary lesion or combined with complex cardiac lesion (TOF, AVSD, TGA) or CoA or AS
Embryology

- 7-8 week gestation
- Type 1 – membranous
- Type II – Outlet
- Type III – Inlet
- Type IV – Muscular

Angelini 1995
Pathophysiology

L-to-R shunt (unrestrictive)
- ↑ Pulm circulation
- ↑ PV return
- ↑ CO
- ↑ PV mmHg

Pulmonary Edema
Tachypnea/dyspnea

LVEF
Fluid retention
↑ SVR
↑ Metabolic activity
↑ oxygen demand
Hypoxia

RAS Activation & Vasopressin
↑ Catecholamines
↑ Metabolic activity
Neonatal Anemia
Metabolic Acidosis

Systemic Hypotension

Effect of L-to-R Shunt on Cardiac Output

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<th>Shunt</th>
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</tr>
<tr>
<td>75%</td>
<td>4x</td>
<td>4:1</td>
</tr>
<tr>
<td>80%</td>
<td>5x</td>
<td>failure</td>
</tr>
</tbody>
</table>

HEART FAILURE

http://www.cvphysiology.com/Cardiac%20Function/CF003.htm
Clinical Presentation

Small restrictive:
- After birth mild tachycardic and tachypneic
- Asymptomatic murmur (Low pitch harsh Holosystolic best at LUSB grade 2-3/6) +/- thrill

Moderate to Large: CHF
- Symptoms: Respiratory distress and sweating w/ feeding, poor feeding, hungry/irritable, failure to thrive
- Cardiac Exam: Hyperdynamic precordium, systolic thrill LSB, Normal S1/S2, Mid-diastolic murmur best apex (↑ Mitral flow ~Qp:Qs >2:1), Normal pulses
- CHF Findings: Tachypnea, tachycardia, grunting, respiratory distress, rales, displaced PMI, hepatomegaly, palor, mottled, poor pulses
- Severe PHTN: parasternal heave + Loud Split S2 + Diastolic murmur
Diagnosis

- **CXR:** Cardiomegaly, increase pulmonary vascular markings, diffuse pulmonary edema
- **ECG:**
  - Small: NSR to Tachycardia
  - Large: LVH, LAD, Deep Q waves
  - PHTN: RVH
- **ECHO:** Location, size, RV mmHg, degree shunting, associated defects
- **MRI:** helpful to define complex lesions
- **Catheterization:** if PHTN evaluate responsiveness
Conservative Management

Spontaneously closure:
- Variation between type, size, and age
- Small VSD >50% closure by age 5, 80% by adolescence

Watch & Wait:
- Small VSD, Asymptomatic, <50% shunt, no LV volume overload, no aortic insufficiency, normal PAP
- Event free survival 96+-2% at 8 yrs

### Long-Term Outcomes For Non-Surgical Pediatric VSD (N=222)

<table>
<thead>
<tr>
<th>Complication</th>
<th>Spontaneous Closure</th>
<th>Death</th>
<th>SVT</th>
<th>Endocarditis</th>
<th>AVR 2/2 Endocarditis</th>
<th>Normal LV size</th>
</tr>
</thead>
<tbody>
<tr>
<td>N (%)</td>
<td>14 (6%)</td>
<td>0</td>
<td>2 (1%)</td>
<td>4 (2%)</td>
<td>2 (1%)</td>
<td>191 (89%)</td>
</tr>
</tbody>
</table>
Management

- **Medical:**
  - Diuretics (Lasix, Diuril, Spironolactone)
  - +/- Digoxin for very large VSD
  - Optimizing nutrition
  - **Closure Indications:** Closure recommended <6-12mo (Earlier if Trisomy 21)
    - Qp:Qs >2:1 + LV volume overload
    - FTT
    - Aortic insufficiency
    - Infective endocarditis
    - Near systemic RV/RA mmHg

- **Contraindication:** Severe irreversible PHTN

- **Surgical options:**
  - Median sternotomy, CPB, Trans-atrial vs. trans-ventricular, Primary vs. Patch
  - PA banding
  - Single ventricle palliation

- **Interventional Catheterization:**
  - Transcatheter closure (Amplatzer): Muscular device FDA approved

- **Hybrid technique** (transventricular device placement – difficult apical VSD)
### Early Post-op Complications

**Table 3. Event rates by operative indication**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Overall (n = 285)</th>
<th>Category Q (n = 232)</th>
<th>Category P (n = 22)</th>
<th>Category O (n = 31)</th>
<th>Fisher’s exact P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Any reoperation</td>
<td>14</td>
<td>10 (71.4)</td>
<td>0 (0)</td>
<td>4 (28.6)</td>
<td>.094</td>
</tr>
<tr>
<td>Wound infection</td>
<td>4</td>
<td>4 (100)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>.999</td>
</tr>
<tr>
<td>Postpericardiotomy syndrome</td>
<td>3</td>
<td>2 (66.7)</td>
<td>0 (0)</td>
<td>1 (33.3)</td>
<td>.466</td>
</tr>
<tr>
<td>Chylous effusion</td>
<td>4</td>
<td>2 (50.0)</td>
<td>2 (50.0)</td>
<td>0 (0)</td>
<td>.045†</td>
</tr>
<tr>
<td>Chest tube</td>
<td>10</td>
<td>7 (70.0)</td>
<td>2 (20.0)</td>
<td>1 (10.0)</td>
<td>.254</td>
</tr>
<tr>
<td>Heart block</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transient</td>
<td>10</td>
<td>8 (80.0)</td>
<td>2 (20.0)</td>
<td>0 (0)</td>
<td>.197</td>
</tr>
<tr>
<td>Permanent, pacemaker placed</td>
<td>6</td>
<td>5 (83.3)</td>
<td>0 (0)</td>
<td>1 (16.7)</td>
<td>.717</td>
</tr>
<tr>
<td>Seizure</td>
<td>5</td>
<td>5 (100.0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>.999</td>
</tr>
<tr>
<td>Extracorporeal membrane</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Oxygenation</td>
<td>5</td>
<td>3 (60.0)</td>
<td>0 (0)</td>
<td>2 (40.0)</td>
<td>.143</td>
</tr>
<tr>
<td>Death</td>
<td>5</td>
<td>4 (80.0)</td>
<td>0 (0)</td>
<td>1 (20.0)</td>
<td>.650</td>
</tr>
<tr>
<td>Rehospitalization</td>
<td>1</td>
<td>1 (100)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>.999</td>
</tr>
</tbody>
</table>

Data are presented as n (%). *Indication for operation was classified in 1 of 3 mutually exclusive groups: failure to thrive/congestive heart failure (Category Q: “Flow”); right ventricular obstruction, aortic insufficiency, or double chamber right ventricle (Category O: “Obstruction”); and elevated pulmonary vascular resistance (Category P: “Pulmonary”). †P > .01 to .05.
### Device vs. Surgical Closure

<table>
<thead>
<tr>
<th></th>
<th>Device group (n = 852)</th>
<th>Surgery group (n = 1,326)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Success, n</td>
<td>850 (99.8%)</td>
<td>1,326 (100%)</td>
<td>NS</td>
</tr>
<tr>
<td>Major complications, n</td>
<td>16 (1.9%)</td>
<td>33 (2.5%)</td>
<td>NS</td>
</tr>
<tr>
<td>Minor complications, n</td>
<td>5 (0.6%)</td>
<td>85 (6.4%)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Blood products needed, n</td>
<td></td>
<td>136 (10.3%)</td>
<td></td>
</tr>
<tr>
<td>Volume of blood transfusion, ml</td>
<td>510 ± 130</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ICU stay, days</td>
<td>3.2 ± 1.5</td>
<td>12.9 ± 3.7</td>
<td>&lt;0.01</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th></th>
<th>Device group (n = 852)</th>
<th>Surgery group (n = 1,326)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Major complications</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Residual shunt</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>Severe arrhythmia</td>
<td>9</td>
<td>16</td>
</tr>
<tr>
<td>Secondary thoracotomy</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>Valvular insufficiency</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Minor complications</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiac insufficiency</td>
<td>0</td>
<td>23</td>
</tr>
<tr>
<td>Hemolysis</td>
<td>3</td>
<td>16</td>
</tr>
<tr>
<td>Vessel injury</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Infection</td>
<td>0</td>
<td>18</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>Pneumothorax</td>
<td>0</td>
<td>12</td>
</tr>
<tr>
<td>Hydrothorax</td>
<td>0</td>
<td>9</td>
</tr>
</tbody>
</table>

Zheng 2009
## Pediatric Surgical Long-term Results

### Long Term Operative Outcomes for Pediatric VSD Repair (N=176)

<table>
<thead>
<tr>
<th>Complication</th>
<th>Early Mortality</th>
<th>Late Mortality</th>
<th>Aortic Insufficiency</th>
<th>Endocarditis</th>
<th>Pacemaker</th>
<th>Re-operation</th>
<th>Residual VSD</th>
<th>NYHA Class I/II</th>
</tr>
</thead>
<tbody>
<tr>
<td>Precent</td>
<td>13%</td>
<td>4%</td>
<td>16%</td>
<td>1%</td>
<td>4%</td>
<td>10%</td>
<td>8%</td>
<td>100%</td>
</tr>
</tbody>
</table>

*Fig. 1 (Event-free) survival after VSD closure.*

Roos-Hesselink 2004
## Adult Surgical Long-term Results

### Long Term Operative Outcomes for Adult VSD Repair (N=42)

<table>
<thead>
<tr>
<th>Complication</th>
<th>Early Mortality</th>
<th>Late Mortality</th>
<th>Aortic Insufficiency (moderate)</th>
<th>Arrhythmia</th>
<th>Pacemaker</th>
<th>Re-operation</th>
<th>Residual VSD</th>
<th>NYHA Class I/II</th>
</tr>
</thead>
<tbody>
<tr>
<td>Precent</td>
<td>0%</td>
<td>5%</td>
<td>17%</td>
<td>28%</td>
<td>9%</td>
<td>5%</td>
<td>23%</td>
<td>97%</td>
</tr>
</tbody>
</table>

Mongeon 2010
Aortic Coarctation (CoA)
Epidemiology & Risk Factors

- Incidence 1/12,000 live births
- Risk Factors: Male > female 1.7:1, Turner syndrome, DiGeorge syndrome (22q11 deletion)
- Associated defects: VSD 11%, Bicuspid aortic valve 30-66%, Hypoplastic left heart syndrome
Anatomy

http://commons.wikimedia.org/wiki/File:Coarctation_and_PDA.png
Pathophysiology

Aortic Coarctation

Critical Stenosis

Ductus Closure

↓Diastolic Coronary flow

↓Stroke Volume

↓CO

↓LV strain

↑afterload

↑upfront load

↑LVEDP

↑PV mmHg

Hypertension

Sub-critical Stenosis

Develop Collateral Circulation

Hypotension

LV dysfunction

Metabolic Acidosis

Hypotension

Renal Failure

Heart Failure

PV mmHg

Pulmonary Edema

LV dysfunction

LV dysfunction

CHF

Aortic Rupture

Intracranial Hemorrhage

Necrotizing Enterocolitis
Clinical Presentation

Critical Coarctation:
- Collapse vs. poor feeding, sweating, respiratory distress, failure to thrive
- Present in 1st week after DA closure or within 3 months
- Exam: Tachypnea, Tachycardia, Respiratory distress, rales, continuous systolic murmur at LSB, gallop, delayed/weak/absent femoral pulses, delayed cap refill, mottling, peripheral cyanosis, hepatomegaly

Sub-critical Coarctation:
- Detected by murmur, weak femoral pulses, or hypertension
- Symptoms: Headaches, epistaxis, cold feet, Exertional calf pain
- Exam: Displaced PMI, 4th heart sound, continuous murmur best at infraclavicular fossa radiates to back
- Extremity BP Gradient (≥20mmHg) + weak/absent femoral pulses = CoA
Diagnosis

- **CXR:** Early – cardiomegaly, ↑pulmonary vascular markings; Late - Figure 3 sign, Rib notching
- **ECG:** Early – RAD; Late - LVH
- **ECHO** – best for infants
- **MRI** – children/adults 2/2 poor acoustic windows
- **CT** – best for patients with stents
- **Fetal ECHO** – 2/3rd false positive
Acute Management

- Prostaglandin E1 – maintain PDA (maximal response in 15 min – 4 hr (open DA and decrease CoA)
  - SE’s: ↓ respiratory drive, hypotension, diarrhea
  - No response need emergent surgical repair
- Transfer to surgical center
- Supportive Care: Mechanical ventilation and inotropes (dopamine)
Surgical vs. Endovascular Surgery: Infant Treatment of Choice

- Extended end-to-end repair (#1)
- Subclavian flap aortoplasty (aneurism and restenosis more common)
- Prosthetic patch aortoplasty
- Interposition graft (no risk to outgrow graft)

Interventional Catheterization:

- Balloon angioplasty (frequent Aneurism & restenosis)
- Balloon angioplasty + stent placement (Treatment of choice in older children/adults)

Gaca 2008
Complications & Prognosis

- Unrepaired prognosis: Mean age survival 31 yrs
  - CHF 25%, aortic rupture 21%, endocarditis 18%, intracranial hemorrhage 11% (Padua 2012)
- Early Operative Complications:
  - Bleeding, Chylothorax, hypertension, nerve injury (recurrent laryngeal, phrenic, Horner’s syndrome, spinal paraplegia)
- Late Repaired Complications:
  - Hypertension
  - Re-coarctation
- Endovascular Complications:
  - Aneurism (4-7% stent)
  - Dissection
  - Re-coarctation

Fuster 1989
Tetralogy of Fallot
(TOF)
Epidemiology and Risk Factors

- Most common cyanotic congenital heart lesion
- Prevalence: 3.9/10,000 live births
- 7-10% of congenital heart disease
- 15% have associated syndromes: Trisomy 21, (Chromosome 22q11 deletion - DiGeorge or Velocardiofacial), and Alagille syndrome (JAG1 mutation)
- 5-10% have pulmonary atresia with VSD
Embryology

[Images of embryological stages and anatomical diagrams]


Angelini 1995
Pathophysiology

- **RV Infundibular Spasm**
  - ↑ RVOT obstruction

- **Pain/Agitation**
  - Crying (prolonged expiration)
    - ↑ PVR
    - ↑ R→L shunt
    - ↓ Pulm Perfusion
    - ↑ hypoxemia/cyanosis
    - Metabolic acidosis
    - Hypotension

- **fever**
  - vasodilation
    - ↓ SVR

- **↓ RV preload**
  - Tachycardia

- **↑ RV filling**
  - ↑ Pulm Perfusion

- **↑ venous return**
  - Hyperpnea

- **Seizures/Stroke/Death**
  - syncope/bradycardia
  - ↑ Pulm Perfusion
**Typical Presentation**

**TOF:** A spectrum disorder in which severity of anatomy predicts timing of presentation

Degree of pulmonary obstruction:
- **Severe:** profound cyanosis in neonatal period
- **Moderate:** Found in clinic for murmur evaluation
- **Minimal:** Increased pulmonary circulation and HF

Prenatal ultrasonography has significantly improved early detection and treatment (Prenatal US 90% accurate)

Diagnosis

- ECG: NSR, RVH, RAD, RBBB (post repair)
- CXR: Boot shaped heart
- ECHO: Pulmonary stenosis/atresia, Overriding aorta, VSD, RVH, degree of shunting/stenosis, RV mmHg
- MRI: rare pre-op, evaluate extracardiac lesion
- Catheterization: rare, with complicated TOF
Acute Management

Hypercyanotic Spell:

1st line:
- Knee to chest (increase SVR)
- Fluid bolus & morphine (increase pulmonary flow)
- B-blockade (esmolol/propranolol) RVOT relaxation, HR and RV contractility↑SVR

2nd line:
- Phenylephrine (Increase SVR)
- Emergent BT shunt if intractable to medical management
Surgical Options

*Single stage complete repair within 1st year of life*

Approach: trans-atrial vs. trans-ventricular
1. Annular patch vs. pericardial valvuloplasty vs. RV-PA conduit
2. Resection RVOT obstruction
3. VSD patch closure

**Palliation:** Poor candidates for intracardiac repair
- BT shunt – premature, hypoplastic pulm art, coronary artery anatomy
- Preserve L→R shunt (PFO vs fenestrated VSD patch)

http://www.pediatricheartspecialists.com/articles/detail/tetralogy_of_fallot_surgery
Complications and Prognosis

- **Early complications:**
  - Bleeding, pericardial/pleural effusion, pneumothorax, cardiac dysfxn, arrhythmias, heart block, renal failure
  - RVIS (TCH): trans-atrial/PA approach (minimal to no RV incision); Event free 7 yr survival: w/o genetic d/o 96% and w/ genetic d/o 97% (Morales 2009)

- **Late Complications:**
  - Residual PR or RVOTO → RV dilation → RV failure & Arrhythmias
  - Trans-annular patch: ↑ risk of re-operation, PR or PS, RV dysfunction
References

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