Pediatric Cardiology 2016
Congenital Heart Disease

Incidence

- 8% of live births
- 32,000 new cases/yr in the U.S.
- 1.5 million worldwide
- Rate of increase in adults with CHD is approximately 5%.

Genetics

Chromosomal disorders:
- caused by absent or duplicated chromosomes - trisomy 21 (Down syndrome), 22q11 deletion (DiGeorge syndrome), and 45X deletion (Turner syndrome).
- From 5% to 8% of congenital heart disease patients have a chromosomal disorder.
- Recurrence risk in an offspring is that of the chromosomal disorder.
**Landmarks in congenital heart disease surgery**

- **Ligation of patent ductus** - 1938
- **Resection of aortic coarctation** - 1944
- **Subclavian to pulmonary artery shunt** - 1945
- **Closure of ASD using hypothermia** - 1952
- **Closure of ASD using cardiopulmonary bypass** - 1953
- **Routine use of cardiopulmonary bypass in repair of congenital heart defects** – late 1950s
- **Use of deep hypothermia and circulatory arrest** – late 1960s

**Landmarks in cardiac diagnosis**

- **First cardiac catheterization** – 1929
- **Diagnostic catheterizations are performed in children** – 1950s
- **Use of PGE1 for ductal patency** - 1977
- **2-dimensional echocardiography makes it possible to diagnose defects** – 1980s
- **Utilization of MRI and CT** – 1990s

**Treatment in the cath lab**

- **Balloon atrial septostomy** - 1966
- **First ductal occlusion** - 1967
- **Stainless steel coils were introduced in the 1970s for the embolization of small PDAs and collateral vessels.**
- **Catheter closure of ASD** – 1976
- **Balloon valvuloplasty of pulmonary valve** - 1982
- **Amplatzer ductal occluder** - 1997
- **First transcatheter pulmonary valve replacement** - 2000
Congenital Defects

- Acyanotic
  - left to right shunts
- Cyanotic
  - right to left shunts
- Complex

Acyanotic heart defects
Left to right shunts
Left to right shunts

- Patent ductus arteriosus
- Atrial septal defect
- Ventricular septal defect
- Atrioventricular canal

A cyanotic congenital lesion
Patent ductus arteriosus (PDA)

Patent ductus arteriosus

- Premature infants
- Term infants
- Children
Incidence

- 5-10% of all types of congenital heart disease.
- PDA commonly occurs in cases of exposure to Rubella.

Patent Ductus Arteriosus

Ductal Occluding Devices
Secundum ASDs account for 7% of all congenital cardiac defects. Excluding MVP and BAV it is the most common form of CHD in adults. Most patients with isolated secundum ASDs are females, by a factor of 2 to 1.
Anatomy

- Defects of the Fossa Ovalis
- Sinus Venosus Defect
- Coronary Sinus Defects
- Ostium Primum
Risks

- Embolization of the device
  - early - to the systemic circulation
  - late - to the pulmonary circulation
- Cardiac perforation
- Bleeding
- Arrhythmias
- Transient headaches

Long term concerns

Device erosion – seen in larger defects where the rim of tissue holding the device is deficient.

Natural History

- Children are asymptomatic.
- Adolescents may have some degree of exercise intolerance.
- By age 40 many patients develop CHF.
- Atrial arrhythmias ↑ with advancing age.
- PVOD is rare.
Accounts for ~20% of patients in a large pediatric cardiology practice.

The incidence of congenital heart defects is ~8/1000 births and 29 – 50% have VSD.

The reported incidence has increased in recent years because of the increased sensitivity of color flow Doppler.

Slightly more common in females.

The most common lesion in many chromosomal lesions.

95% of VSDs are not associated with a chromosomal abnormality.

A multifactorial etiology.
Anatomy

- Inlet septum
- Outlet septum
- Trabecular septum
- Membranous septum

Anatomy

- Supracristal defects (subpulmonic)
- Perimembranous defects
- Inlet defects
- Muscular defects

Physiology

The physiologic consequences depend chiefly on the size of the defect and the pulmonary vascular resistance.
Early Clinical Manifestations

- Small defects – no symptoms
- Moderate-size defects – tachypnea, poor weight gain, and sweating.
- Large defects – Tachypnea, poor weight gain, sweating, retractions, and frequent lower respiratory tract infections.

Pulmonary Artery Banding
Cine fluoroscopic images in a seven-month-old female baby with single muscular ventricular septal defect (VSD), demonstrating the steps of percutaneous device closure.


Atroventricular canal

- Aorta
- Pulmonary artery
- Left atrium
- Right atrium
- Right ventricle
- Left ventricle

Incidence

- 4-5% of persons born with CHD.
- CAVC is common in patients with Downs syndrome.
- Partial AVC has been associated with DiGeorge’s syndrome and Ellis-van Creveld syndrome.
Most centers recommend surgical repair in the neonatal period. Residual coarctations are usually treated with balloon dilatation and stent insertion in older patients.
Pulmonary valve stenosis

- Usually asymptomatic as infants unless severe.
- The vast majority are now treated with balloon valvuloplasty.

Valve Replacement

- Congenital aortic valve disease – bicuspid aortic valve with stenosis/or insufficiency
- Postoperative patients for pulmonary atresia, severe PS, or Tetralogy of Fallot with residual conduit obstruction or free pulmonary insufficiency
Cyanotic heart defects

- Tetralogy of Fallot
- Transposition of the Great Arteries
- Tricuspid Atresia
- Total Anomalous Pulmonary Venous Connection
- (Truncus Arteriosus)
- Pulmonary Atresia with Intact Ventricular Septum
1. Tetralogy of Fallot

2. Epidemiology

- 7% of congenital heart disease
- Most common cyanotic defect
- Associated malformations: 35%
  - Major organ defect: 13%
  - Syndrome/genetic disease: 20%
  - Chromosome 22q11 deletion syndrome
First surgery was palliative.

The condition was initially thought untreatable until surgeon Alfred Blalock, cardiologist Helen B. Taussig, and lab assistant Vivien Thomas at Johns Hopkins University developed a surgical procedure, which involved forming an anastomosis between the subclavian artery and the pulmonary artery.

The first Blalock-Thomas-Taussig shunt surgery was performed on 15-month old Eileen Saxon on November 29, 1944 with dramatic results.

Currently, Blalock-Thomas-Taussig shunts are not normally performed on infants with TOF except for severe variants such as TOF with pulmonary atresia.
The first total repair was performed by C. Walton Lillehei at the University of Minnesota in 1954 on a 10-month boy. Total repair initially carried a high mortality risk which has consistently improved over the years. Surgery is now often carried out in infants 1 year of age or younger with a <5% perioperative mortality.

- Complete repair has very low risk today
- 30 year follow up
  - 85 – 90% survival
  - Expect better results for repairs done recently
  - Most have good quality of life
- Important complications
  - Pulmonary stenosis and insufficiency
  - Ventricular tachycardia/sudden death
- Reintervention improves long-term outlook
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TGA
- 5-7% of all patients with congenital heart disease.
- Sporadic and nonfamilial.
- Male : female = 2:1
- Increased incidence in siblings of affected patients.

Transposition of the Great Arteries
Physiology

- Two parallel circuits versus one series circuit.
- Survival depends on shunting at the atrial and ductal levels.

Series vs parallel circuits

- 1950, Blalock Hanlon septectomy
- 1964, Mustard procedure
- 1966, Rashkind balloon atrial septostomy
- 1975, Arterial Switch procedure
- 1977, Prostaglandin E₁
**Cardiac Catheterization**

- Balloon atrial septostomy
- Coronary artery anatomy

**Balloon Atrial Septostomy**

**Surgical Repair**

- Atrial Baffle Procedures – Senning and Mustard
- Arterial Switch Procedure
Success first reported in 1975.
Early mortality initially ranged from 5-15%.
Now in most surgical centers the early operative mortality is < 5%.

Kinked or obstructed coronary arteries.
"unprepared" left ventricle
Hemorrhage from multiple suture lines.
Late Mortality

- Myocardial ischemia
- Pulmonary vascular obstructive disease
- During reoperation for supravalvar obstruction.

Complications

- Supravalvar pulmonary stenosis
- Supravalvar aortic stenosis
- Neoaoartic root enlargement
- Neoaoartic valve regurgitation
- Coronary artery occlusion

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Complex congenital defects

- Single ventricle
  - tricuspid atresia
  - hypoplastic left heart syndrome
- Truncus arteriosus
- Tetralogy of Fallot, PA with MAPCAs
Hypoplastic left heart syndrome

- Fatal diagnosis until the 1980s
- Dr. William Norwood introduced the first palliative procedure and it still being performed with modifications.
Stage II – cavopulmonary anastamosis Fontan procedure


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Truncus Arteriosus
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Tetralogy of Fallot, PA with MAPCAs

Collateral Arteries feeding Pulmonary Circulation (C)

The Pulmonary arteries are present but very small

Tetralogy of Fallot, PA with MAPCAs
About one in 300 boys and one in 333 girls will develop cancer before the age of 20 (≈0.33%)

186,300, or 0.22% of children have Type diabetes

About one in every 400 to 600 children and adolescents has type 1 diabetes

2 million adolescents (or 1 in 6 overweight adolescents) aged 12-19 have pre-diabetes

Incidence of CHD, 0.8% - 0.3% for complex defects
0.6% for moderate defects
0.3 -2% for simple defects

“Would you like a bypass with that?”

No thanks, I quit.
“What fits your busy schedule better, exercising one hour a day or being dead 24 hours a day?”