Objectives

• Review the history of how cardiac surgery evolved in congenital heart disease
• Discuss the anatomical issues of the most common cardiac defects
• Discuss how each of these defects are repaired and the degree of difficulty of the surgery
• Discuss the outcomes of the various lesions with surgical repair

Disclosures

• I have no financial relationships with any manufacturer of any commercial product and/or provider of commercial services discussed in the CME activity.
• I do not intend to discuss an unapproved/investigative use of commercial product or device in my presentation.

History of Cardiac Surgery

• In 1938, the first pediatric heart surgery was performed with Dr. Gross ligating a patent ductus arteriosus.
• In 1945, after urging from Dr. Helen Taussig, Dr. Alfred Blaylock and Vivien Thomas performed the first palliative repair in Tetralogy of Fallot
• 1953, Lewis and Taufic used surface cooling with deep hypothermic arrest to close an ASD in a 5 year old girl
• In 1954, Dr. C.W. Lillehei with the ability to provide cross-circulation repaired a child with TOF
• In 1955, John Kirklin at the Mayo Clinic published a series of cases using a pump oxygenator to repair VSDs
• 1958, the first combined deep hypothermia and CPB surgery

Patent Ductus Arteriosus

• Diagnosed after birth
• Commonly seen in the premature infant
• Surgically addressed by ligating the vessel.
• In the older child may be closed in the cath lab
• Survival to discharge is over 96%
• Surgical complexity level: 1
Atrial Septal Defect (ASD)
- Defect between the wall of tissue between the 2 atria
- May be able to be closed with a device in the cath lab if anatomy is favorable
- If not, surgical closure is performed mandating CPB with either suture or patch closure
- Survival rate: Over 99%
- Surgical complexity level: 1

Ventricular Septal Defect (VSD)
- Defect in the wall between the 2 ventricles
- VSDs are named based on location
- Device closure possible for certain types
- Survival to discharge: 99.5%
- Surgical complexity level: 2

Coarctation of the Aorta
- Narrowing in the main body artery
- May be discrete or extended area of narrowing
- Survival to discharge over 98%
- Surgical complexity level 2-3- depending on type

Aortic Stenosis (AS)
- Thickening of the valve of the aorta
- May be anywhere from 1-3 leaflets
- Balloon dilation is usually attempted in the cath lab, before any surgery
- Surgical replacement may be with autologous, or bioprosthetic material, or mechanical valve
- Survival to discharge- post surgery 94-98%
- Surgical complexity level: 2-3
**Pulmonary Stenosis (PS)**

- Thickening of the valve in the pulmonary artery
- Balloon dilation is usually attempted in the cath lab
- Surgical valvotomy performed if balloon of the valve is not successful, or there is extended narrowing above or below the valve.
- Replacements are with bioprosthetic material
- Survival to discharge >98%
- Surgical complexity level: 1

**Atrioventricular Canal Defect (AV Canal)**

- Embryologically derived from abnormality in the formation of the endocardial cushions
- Surgery must close ASD/VSD and create 2 AV valves from a common valve
- Most commonly found in children with trisomy 21
- Survival to discharge: 97.5%
- Surgical complexity level: 3

**Tetralogy of Fallot (TOF)**

- One of the most common forms of cyanotic heart disease
- Anatomically occurs from asymmetric division of the truncus and failure of the infundibular muscle to migrate into the superior portion of the ventricular septum
- Associated with 22q11 deletions
- Survival to discharge: 98%
- Surgical complexity level: 3

**Transposition of the Great Arteries (DTGA)**

- Aorta arises from the RV and the pulmonary artery from the LV
- Surgery of choice is an arterial switch procedure
- More difficult as coronaries must be moved
- Survival to discharge: 97.6%
- Surgical complexity level: 4

**Transposition Surgeries**

**Double Outlet Right Ventricle (DORV)**

- Both great arteries arise from the RV with VSD as the outlet for the LV
- Different forms present which effect surgical repair complexity
- Survival to discharge: 95.2%
- Surgical complexity level: 4
**Truncus Arteriosus**
- Embryologically derived from the single great artery not septating into Ao and PA
- Frequently associated with an abnormal semilunar valve and 22q11 deletions
- Survival to discharge depends on type: 66-91.6%
- Surgical complexity level: 4

**Interrupted Aortic Arch (IAA)**
- Ascending aorta is not in continuity with Descending aorta. The lower body is supplied only by the ductus
- Most often associated with a posterior- malalignment type of VSD (Type B)
- Most commonly seen in 22q11 deletions
- Survival to discharge: 92.2%
- Surgical complexity level: 4

**Total Anomalous Pulmonary Venous Return (TAPVR)**
- Pulmonary veins do not connect the LA, but usually return to a common confluence
- Blood drains via a vertical vein back to the right side of the heart via other venous channels
- Surgery anastomoses the confluence to the LA
- Survival to discharge: 89.4%
- Surgical complexity level: 3

**Single Ventricles**
- Lesions in which there is only one ventricle to the heart or in which the anatomy is such that the ventricles present will have to function as a single ventricle.
- Surgery here is not “repairative” but “palliative”
- Usually requires at least 3 surgeries to get the blood directed so that “blue blood” goes straight to the lungs and “red blood” goes to the body.

**Tricuspid Atresia**
- No tricuspid valve forms and the right heart is small with usually pulmonary stenosis. Flow is supplied by a VSD.
- Will be a form of a single ventricle surgery
- Initially may need additional pulmonary blood flow
- Will need 3 other surgeries to get blood flow “plumbed” to “normal”
- Survival to discharge: 93-94%
- Surgical complexity level: 2

**Pulmonary Atresia (PA/IVS)**
- Small right heart with no flow across the pulmonary outflow
- Ductal dependant initially
- Amount of hypoplasia of the right heart and coronaries determine whether a 1 or 2 ventricle repair can be achieved
- If SV approach, initial surgery is a shunt
- Survival to discharge: 93-94%
- Surgical complexity level: 2
Hypoplastic Left Heart Syndrome (HLHS)
- Failure of the left heart to develop to the degree where it can provide adequate systemic output
- Single ventricle physiology
- Ductal dependant at birth
- Stage 1 is initially performed with reconstruction of the aorta and some form of pulmonary flow
- Overall survival Stage 1: @83%
- Surgical complexity level:4

Caval (Stage 2)
- Survival: >98%
- Complexity: 2

Fontan (Stage 3)
- Survival: 98-99%
- Complexity: 3

Summary
- Surgery for congenital heart defects first began about 75 years ago.
- Gradually over time, more and more lesions were tackled—first palliation and then repairs
- Lesions that were uniformly fatal 30 years ago, are now routinely survivable.
- Single ventricles still tend to fair less favorably than hearts with 2 working ventricles

Summary
- There may still be ongoing cardiac anatomic issues to be dealt with and followed and subsequent surgeries may be need in some lesions.
- But overall most children are able to survive far past the initial state, and most well into adulthood, and many are now having children of their own
- We continue to work to improve the surgical and postoperative care techniques and increase the survivability and quality of life for these patients

References
- Historical congenital surgery information taken from: “Pediatric Cardiac Surgery” Constatine Mavroudis, editor. Copyright 1994, Mosby, St. Louis, Missouri.
- Images are from the American Heart Association’s website: www.heart.org You can find these images under Conditions; Congenital Heart Defects Tools and Resources.

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