definitions & a reality check

Malformation
A primary structural defect arising from a localized error in morphogenesis - results in the abnormal formation of an organ

Dysplasia
Refers to an abnormal organization of cells into tissues - results in abnormal tissues

The distinction of malformation from dysplasia is at best blurry – there is much overlap

repair
complete anatomic correction of congenital heart defect

palliation
provides physiologic correction of blood flow

normal cardiac circulation

intracardiac & extracardiac shunts
in utero – 2 normal shunts
foramen ovale
ductus arteriosus
postnatal abnormal shunts
ASD VSD PDA
 truncus arteriosus

patent ductus arteriosus

division & over-sewing 1938
Gross – Children’s Hospital Boston

triple ligation technique 1946
Bilalock – Johns Hopkins

pharmacologic closure 1976
Indomethacin

Catheter based coil or device closure 1993

Performed when he was Chief Resident & his surgical chairman was out of town!
atrial septal defect

- **closed technique**
  - late 1940s & early 1950s
  - Bailey & Sondergaard (separately)
- **open repair technique**
  - 1952
  - Gross – Children’s Hospital Boston
- **direct visualization**
  - 1953 – Lewis & Taufic
  - using cardiopulmonary bypass
  - 1954 – Gibbons
- **Catheter based device closure**
  - 1997 – Matsuoka

post-surgical findings, complications & re-op indications

- essentially none
- need for re-imaging
- virtually none
- residual ASD

ventricular septal defect

- **PA banding – palliation of VSD**
  - 1952 – Muller & Dammann
- **VSD closure**
  - 1954 – Lillehei – U of Minnesota
  - using a heart-lung machine
  - 1955 – Kirklin – Mayo Clinic
- **transatrial closure**
  - 1958 – Stirling
- **total circulatory arrest**
  - 1969 – Okamoto
- **deep hypothermia & arrest**
  - Barratt-Boyes
- **Amplatz closure device**
  - 1999 – Thanopoulos

post-surgical findings, complications & re-op indications

- essentially none
- need for re-imaging
- virtually none
- residual VSD

TOF – PA atresia – PA stenosis

- **Prosthetic conduit between subclavian & PA**
  - 1962
  - Kliner – refined by Leval
- **BT shunt ipsilateral to the aortic arch**
  - Laks and Castaneda
- **DAo to PA shunt**
  - 1946 – Potts
- **Central aortopulmonary shunt**
  - 1955 – Davidson
- **AAo to PA shunt**
  - 1962 – Waterston

in utero

- blood flow is supplied to the lungs via the ductus arteriosus
- pulmonary vascular resistance is high
  - requires arterial pressure to perfuse the lungs
  - ductus arteriosus closes . . . or . . . maintained opened with PGE

post natal

-
commonly used temporary shunt designed to palliate low pulmonary blood flow (TOF, PA atresia) directs arterial blood flow from a subclavian artery to pulmonary arteries.

- **Blalock-Taussig shunt – classic**
  - Developed for ‘blue babies’
  - 1945
  - Blalock & Taussig & Thomas
  - Johns Hopkins

- **Blalock-Taussig shunt – modified**
  - Developed for ‘blue babies’
  - 1945
  - Blalock & Taussig & Thomas
  - Johns Hopkins

**trans-annular patch**
- Augmentation of the RVOT & enlargement of the MPA
- 1986 – Kirklin

**trans-annular patch**
- Surgical complications
  - Inadequate relief of obstruction
  - Pulmonary insufficiency
- Need for re-imaging
- Restenosis of RVOT
- Branch PA stenosis
- RV failure due to PI

**Hypoplastic left heart syndrome**
- RPA – AAo anastomosis
  - 1970
  - Cayler
- Multiple modifications of this anastomosis
  - Doty
  - Levitsky
  - Behrendt
  - Norwood
- Stage 1 – proceeding to successful Fontan
  - 1983
  - Norwood – Children’s Hospital Boston

**HLHS – staged repair**
- Staged surgical procedures toward goal of Fontan circulation
- Palliation of HLHS
- Neo-aorta & BT shunt are created
- Anastomosis of MPA to AAo
- Limit pulmonary blood flow
- ASO – created or enlarged arterial pressure to the lungs
- Bidirectional cavo-pulmonary shunt
- Venous pressure to lungs
- Fontan circuit
- Completed circuit delivers SVC & IVC blood flow to the lungs
Norwood procedure – alternatives

Sano shunt
2003
Distal MPA is separated from the heart
MPA is used to create neo-aorta
shunt between the systemic RV and the PAs

Hybrid procedure
Akintuerk – 2002
2004 – Bacha & Hijazi
PA bands – regulate pulmonary blood flow
Stent maintains patent ductus arteriosus
ASD is made or enlarged

Norwood procedure – alternatives

Glenn shunt
1958
Glenn – Yale
1975 – Damus
1975 – Kaye
1975 – Stansel

Glenn shunt

circulatory bypass of the R heart
post-surgical findings, complications & re-op indications

• thrombosis
need for re-imaging
• confirming patency
• assessment of
pulmonary blood flow

Glenn shunt post-surgical findings, complications & re-op indications

• thrombosis
need for re-imaging
• confirming patency
• assessment of
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Glenn shunt post-surgical findings, complications & re-op indications

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Glenn shunt post-surgical findings, complications & re-op indications

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Dr. Glenn – Yale

Damus – Kaye – Stansel

Anastomosis of AAo & MPA & RV to PA conduit
1975 – Damus
1975 – Kaye
1975 – Stansel

Damus – Kaye – Stansel

correction of TGA with single ventricle physiology – or
single ventricle repair – HLHS

the MPA is transected and
anastomosed with the AAo

Damus – Kaye – Stansel

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correction of TGA with single ventricle physiology – or
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the MPA is transected and
anastomosed with the AAo
Multi-staged procedure to palliate tricuspid atresia, single ventricle syndromes [HLHS, HRV with PA atresia]

- Total cavopulmonary connection returns systemic venous blood flow to the lungs separate from right heart contractions.
- The R & L circulations are separate.
- Post-surgical findings, complications & re-op indications:
  - Thrombosis
  - Pleural effusions
  - Ascites
  - Need for re-imaging
  - Confirming patency
  - Assessment of pulmonary blood flow

Transposition of the great arteries

- D – TGA
  - AV concordance
  - VA discordance
  - Parallel circulations requires mixing - shunt

- L – TGA
  - AV discordance
  - VA discordance
  - 2 wrongs do not make a right

Jatene arterial switch

- Correction of D loop TGA at the arterial level
- Physiological correction of D-TGA
  - The aorta and MPA are switched and the coronary arteries are reimplanted into the neo aorta
- Post-surgical findings, complications & re-op indications:
  - Tension on great vessels & reimplanted coronary arteries
  - Need for re-imaging
  - Coronary artery origin stenosis
  - RV failure as it is not well suited to be the systemic ventricle
manner to minimize kinking of the coronary arteries which can be a complication of the arterial switch Jatene procedure. The pulmonary arteries are divided to transfer the PAs anterior to the AAo. The Le Compte maneuver is used for the correction of TGA with VSD and LVOT obstruction. RV – PA conduit is also used for PA atresia, TOF, DORV, or HLHS. The MPA is ligated and anastomosed to the RV via a bovine pericardial conduit or artificial graft material from the RV to the PAs. The tunnel connecting the LV to the aorta is created by Borromei.

Rastelli procedure

post-surgical findings, complications & re-op indications
- thrombosis
- pleural effusions
- ascites
- need for re-imaging
  - conduit stenosis
  - pulmonary insufficiency
  - RV hypertrophy & failure

PA banding

circumference of band (mm) = child's weight (kg) + 20

Trusler & Mustard

Mustard or Senning – atrial switch

interatrial baffle
- 1954 – Mustard using artificial pericardium
- 1959 – Senning using atrial tissue

double switch

Physiologic correction of congenitally corrected L-TGA

Senning or Mustard & Jatene or Rastelli
**Ross procedure**

Pulmonic valve moved to the aortic position

1962 – Ross
Guys Hospital – London

**Coarctation of the aorta**

First surgical repair of coarctation of the aorta
1944
Grafoord – Karolinska Institute

**Coarctation of the aorta – surgical**

End – to – end anastomosis
most often performed during the first year of life

tissues are more elastic
so bringing ends together easier

may be an oblique anastomosis

Patch repair
performed at any age

**Coarctation of the aorta – stent**

Interventional – catheter based repair

angioplasty & stenting to dilate coarctation of the aorta

beware of jailing of the left subclavian artery origin

**Aorto-aorto bypass graft**

used to palliate interrupted aortic arch

OR

to supplement repaired coarctation of the aorta

**Waterston & Potts shunts**

not currently performed
augments pulmonary arterial blood flow
sometimes excessively so

Waterston shunt
AAo – RPA

Pott’s shunt
DAo - LPA
Waldhausen procedure

Repair of aortic coarctation

- Left subclavian artery is ligated
- and used to augment the stenotic aorta

Complications
- reduced blood flow to the left upper extremity
- poor growth of the extremity
- no longer used

Common Surgical Procedures for Congenital Heart Disease

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