Surgical Management of Congenital Heart Disease: Cyanotic Lesions

Dr. T.K Susheel Kumar
Assistant Professor,
Pediatric Cardiothoracic Surgery,
Le Bonheur Children's Hospital
University of Tennessee

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It’s 2:00 A.M.

Your first night on call on your Pediatric rotation.

The R.N. in the newborn nursery calls you to see a blue baby.

You run.
Scenario

- $O_2$ sat=58%
- RR=82
- Mild respiratory distress

What will you do next?
Cyanosis

- Bluish discoloration of skin and mucous membranes
- Noticeable when the concentration of deoxy-hemoglobin is at least 5g/dl
  - \((O_2 \text{ sat} < 85\%)\)
Cyanosis: peripheral vs central

**Peripheral**
- Sluggish blood flow in capillaries
- Involves extremities (acrocyanosis)
- Spares trunk & mucous membranes

**Central**
- Abnormalities of lungs or heart that interfere with O2 transport
- Involves trunk & mucous membranes
Cyanosis and Hemoglobin

![Graph showing hemoglobin levels]

Anemia (Hgb. 6 gm%): 45-50%
Normal (Hgb. 15 gm%): 75-80%
Polycythemia (Hgb. 20 gm%): 80-85%
Evaluation of cyanosis: Hyperoxia test

*pAO*₂ measurement on room air and 100% O₂

**Lung disease:**
1. On room air, *pAO*₂ is 30 mmHg with O₂ sat=60%.
2. On 100% O₂, *pAO*₂ is 110 mmHg with O₂ sat=100%.

**Cardiac disease:**
1. On room air, *pAO*₂ is 30 mmHg with O₂ sat=60%.
2. On 100% O₂, *pAO*₂ is 40 mmHg with O₂ sat=75%.

- *pAO*₂ >150 mmHg suggests lung disease
- Little or no change in *pAO*₂ (< 100 mmHg) suggests cyanotic heart disease

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Chronic cyanosis causes **clubbing of the digits**
Effects of cyanosis

- Right to left shunting of blood due to obstruction to pulmonary blood flow
- Hypoxemia and cyanosis
- Polycythemia
- Altered hemostasis
- Microvascular thrombosis
- Poor tissue perfusion
- Myocardial dysfunction
- Growth retardation
- Acidosis
- Renal and cerebral thrombosis

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Prostaglandin (PGE$_1$)

- Stable derivative of endogenous compound that maintains ductal patency in utero.
- Prevents postnatal ductal closure, improves pulmonary blood flow, improves oxygenation.
- Stabilizes cyanotic neonate so that corrective surgery can be performed “electively.”
- PGE$_1$ revolutionized the fields of pediatric cardiology and cardiovascular surgery.

The basis of congenital heart disease is rooted in an arrest of or deviation in normal cardiac development.
Physiologic classification

Acyanotic
- Increased pulmonary blood flow
- Normal pulmonary blood flow

Cyanotic
- Increased pulmonary blood flow
- Decreased pulmonary blood flow
CLASSIFICATION OF CONGENITAL HEART DISEASE

Acyanotic

↑ Pulmonary blood flow
- Atrial septal defect
- Ventricular septal defect
- Patent ductus arteriosus
- Atrioventricular canal

- Coarctation of aorta
- Aortic stenosis
- Pulmonic stenosis

Obstruction to blood flow from ventricles
- Tetralogy of Fallot
- Tricuspid atresia

↓ Pulmonary blood flow

Cyanotic

Mixed blood flow

- Transposition of great arteries
- Total anomalous pulmonary venous return
- Truncus arteriosus
- Hypoplastic left heart syndrome
The 5 T’s of cyanotic heart disease

- Tetralogy of Fallot
- TGA (d-transposition of the great arteries)
- Truncus arteriosus
- Total anomalous pulmonary venous return
- Tricuspid atresia / single ventricle
- Pulmonary atresia
- Ebstein’s malformation of tricuspid valve
Tetralogy of Fallot

1. Pulmonary stenosis
2. Large VSD
3. Overriding aorta
4. Right ventricular hypertrophy

Étienne-Louis Arthur Fallot
Tetralogy of Fallot

- 6% of all congenital heart disease
- 1:3600 live births
- most common cause of cyanosis in infancy/childhood
- Severity of cyanosis proportional to severity of RVOT obstruction
TOF occurs as a result of abnormal cardiac septation
Tetralogy of Fallot

- Anterior deviation of the outlet ventricular septum is the cause of all four abnormalities seen in tetralogy of Fallot.
Tetralogy of Fallot - CXR

- Typical “boot-shaped” heart secondary to RVH and small main pulmonary artery segment
- Pulmonary vascular markings are decreased
Pathophysiology

- Pathophysiology of TOF depends on
  - The degree of RVOTO
  - Ratio of SVR to PVR
R – L shunt: Balance between SVR and PVR

Right to left shunt increases

Pulmonary vascular resistance

Systemic vascular resistance

High
Pulmonary vascular resistance

Low
Systemic vascular resistance

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Hypercyanotic “tet” spell

- Paroxysmal hypoxemia due to acute change in balance between PVR and SVR
- ↓ SVR causes an increase in R→L shunt, increasing cyanosis
- ↓ SVR (hot bath, fever, exercise)
- Agitation →dynamic subpulmonic obstruction
- Life-threatening if untreated
Management of “tet” spell:
goal is to ↑ SVR and ↓ PVR

1. Knee-chest position (↑ SVR)
2. Supplemental O₂
3. Fluid bolus i.v. (↑ SVR)
4. Morphine i.v. (↓ agitation, ↓ dynamic RVOT obstruction)
5. NaHCO₃ to correct metabolic acidosis (↓ PVR)
6. Phenylephrine to ↑ SVR
7. β-blocker to ↓ dynamic RVOT obstruction
Surgical Approach

Palliative procedures

Increase PBF by creating a shunt between systemic & pulmonary circulations.

Systemic-pulmonary arterial shunts

- Classic Blalock-Taussig shunt
- Pott’s shunt
- Waterston shunt
- Modified Blalock-Taussig shunt (MBTS)
Thomas-Blalock-Taussig Shunt

Alfred Blalock

Vivien Thomas

Helen Taussig

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Cardiopulmonary Bypass

- 1954. Lillehei
  1st surgical closure of VSD under controlled cross-circulation
- Used in 45 patients between 1954 to 1955
- VSD
  TOF
  AVSD
Surgical repair: TOF
Surgical repair: TOF
D-Transposition of the Great Arteries

- Ao is anterior, arises from right ventricle
- PA posterior, arises from left ventricle
- Systemic venous (blue) blood returns to RV and is ejected into aorta
- Pulm venous (red) blood returns to LV and is ejected into PA
d-TGA results from abnormal formation of aortico-pulmonary septum
D-transposition of great arteries

- Systemic and pulmonary circulations are in parallel, rather than in series
- Mixing occurs at atrial and ductal levels
- Severe, life-threatening hypoxemia
D-transposition of great arteries

- 5% of all congenital heart disease
- Most common cause of cyanosis in neonate
- Male:female 2:1
d-TGA CXR: “egg on a string”

- Narrow mediastinum due to anterior-posterior orientation of great arteries and small thymus
- Cardiomegaly is present w/ increased pulmonary vascular markings
Initial management of d-TGA: goal is to improve mixing

1. Start PGE$_1$ to prevent ductal closure
2. Open atrial septum to improve mixing at atrial level (Rashkind procedure).
Balloon atrial septostomy (Rashkind procedure)
Surgical management of d-TGA: Arterial switch procedure

The arterial trunks are transected and “switched” to restore “normal” anatomy.

The coronary arteries are harvested and re-implanted.
Surgical management of d-TGA: Arterial switch procedure
Truncus arteriosus

- Aorta, pulmonary arteries, and coronary arteries arise from single vessel.
- Truncus sits over large ventricular septal defect.
- Failure of septation of embryonic truncus.
- Uncommon (1.4% of CHD)
Truncus Arteriosus

RAA  Tr  LAA

RV  LAA

PA  Tr

Ao  PA

LV  T
Truncus arteriosus: aortico-pulmonary septum fails to develop
Conotruncal development is dependent upon normal migration of neural crest cells.

Neural crest tissue is required for formation of:

- conotruncus
- aortic arches
- facial structures
- thymus
- parathyroid
DiGeorge Syndrome (22q11 deletion)

- Deletion of a portion of chromosome 22 results in abnormal neural crest development
- **Conotruncal defects** (truncus arteriosus, interrupted aortic arch)
- **Thymic aplasia** (T-cell dysfunction)
- **Parathyroid aplasia** (hypocalcemia)
- **Facial dysmorphic features**
Surgical correction: Truncus Arteriosus
Surgical correction: Truncus Arteriosus
Total anomalous pulmonary venous return (TAPVR)

- Failure of pulmonary veins (PV) to fuse with developing left atrium

- PV drainage occurs through embryological remnants of systemic veins

- Incidence: rare
Embryological basis of TAPVR

Systemic venous connections reabsorb as PV confluence fuses with left atrium

Systemic venous connections fails to fuse with left atrium
Venous connections in TAPVR

Supracardiac: Ascending vertical vein most common

Cardiac: Right atrium or coronary sinus

Infracardiac: Descending vein to portal system
Clinical manifestation of TAPVR

Obstructed

- Severe pulmonary edema, cyanosis, shock
- Surgical emergency

Unobstructed

- Mild to moderate congestive heart failure and cyanosis
- Surgery in first 6 mo.
Supracardiac TAPVR

- Superior vena cava
- Ascending vertical vein
- Pulmonary vein
- Ascending aorta
TAPVR - Supracardiac
Supracardiac TAPVR - CXR

“Snowman” appearance secondary to dilated vertical vein, innominate vein and right superior vena cava draining all the pulmonary venous blood
Infracardiac TAPVR
TAPVR - Infracardiac

Lungs

Descending vertical vein

Liver

heart
TAPVR - CXR
Infracardiac = Obstructed = Surgical Emergency
Surgical repair of TAPVR
Tricuspid atresia

• Absent communication from RA to RV
• Obligate R to L shunt at atrial level
• GA normally related (70%)
• GA transposed (30%)
Tricuspid atresia

- Pulmonary valve may be normal, stenotic or atretic
- Degree of cyanosis proportional to degree of pulmonary stenosis
- Necessity for PGE$_1$ related to degree of pulmonary stenosis
Tricuspid atresia
Surgical management of Tricuspid atresia

- Stage 1: BT shunt/ PA banding/ Norwood procedure
- Stage 2: Glenn/ Hemifontan operation
- Stage 3: Fontan operation
Norwood procedure
Norwood Sano operation

Stage I

- Neo-aorta
- Pulmonary artery
- Shunt
Bidirectional Glenn operation
Fontan operation

Extracardiac Fontan

Superior vena cava and Gore-tex® conduit attached to pulmonary artery

Inferior vena cava attached to Gore-tex® conduit

A fenestration (small hole) allows blood flow between right atrium and conduit

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Pulmonary atresia

- Atretic pulmonary valve
- Pulmonary arteries often normal in size
- Hypoplastic RV, RVH
- Hypoplastic TV
Pulmonary atresia

- Ductal-dependent lesion
- Requires PGE\textsubscript{1} to maintain oxygenation
- Therapy directed at opening atretic valve in cath lab or surgery
- Prognosis depends upon size and compliance of hypoplastic RV
Pulmonary atresia
Ebstein’s malformation of tricuspid valve

- TV leaflets attach to RV wall, rather than TV annulus
- Tethered leaflets create 2 chambers w/in RV: large “atrialized” RV, small noncompliant functional RV
- Severe tricuspid regurgitation
Ebstein’s malformation of TV

- Massive RA dilation due to severe tricuspid regurgitation
- R to L shunt at atrial level causes cyanosis
- Degree of cyanosis related to size and compliance of functional RV
- Cyanosis usually decreases as PVR falls shortly after birth
Ebstein’s malformation of TV

RA

LV

aRV

RV

RV

nRV

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Surgical correction of Ebstein’s malformation
Scenario

- Room air pAO$_2$ = 29  sat=58%
- 100% O2 pAO$_2$ = 35  sat=67%

What will you do next?

Start PGE$_1$
Call cardiology to do echocardiogram
Questions

1. The minimum concentration of reduced or deoxygenated hemoglobin in the cutaneous veins for detection of cyanosis is

   a) 3 g/dL

   b) 4 g/dL

   c) 5 g/dL

   d) 2 g/dL
Questions

2. Management of Tet spell includes everything except

a) Oxygen

b) Fluids

c) Phenylephrine

d) Epinephrine
Questions

3. Most common cause of cyanosis in neonate is

a) Transposition of great arteries.

b) Tetralogy of Fallot.

c) Truncus arteriosus.

d) Total anomalous pulmonary venous return.
Questions

4. The following is a true cardiac emergency operation

a) Tetralogy of Fallot with 'tet' spells.

b) Transposition of great arteries with poor oxygen saturation.

c) Infradiaphragmatic obstructed total anomalous pulmonary venous return.

d) Truncus arteriosus with interrupted aortic arch.
Thank you