Surgical Management of Congenital Heart Disease – Part 1
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Group 4

Additional information from Batch Jax's notes were not included here (if may ara man gid, gamay lang). You are encouraged to read their novel-like notes from last year if you wish to.
SAVE SPACE SAVE PAPER SAVE TREES SAVE THE EARTH

General Principles of Treatment

- Most patients who have mild congenital heart disease require no treatment
- Ex. Small VSD because it will close spontaneously in some cases
- Need not be restricted in physical activities
- Discourage competitive sports
- Routine immunization should be given
- To prevent superimposed bacterial infection because they are more prone to develop bacterial endocarditis
- Childbearing and on use of contraceptives and tubal ligation be encouraged
- Specially in cyanotic congenital heart disease
- Bacterial infections should be treated vigorously (because of the danger of septic shock)
- Treatment of iron deficiency anemia (IDA)
- Hemodynamic changes is more pronounced when there is IDA or polycythemia
- Careful observation for polycythemia for danger of thrombosis
- Avoid sudden changes in temperature
- Careful monitoring during surgery and anesthesia
- Counseling on risks associated with pregnancy

PERIOPERATIVE CARE OF THE INFANT AND CHILD

PREOPERATIVE CONSIDERATIONS

- Be familiar with both the patient and the family
- Knowledge of the intended surgery, expected outcome, potential complications
- Physical examination, review of catheterization and other physiologic data
- Cardiac Catheterization (determine the oxygen pressure and saturation):
  - Right side – insert at the femoral vein → IVC RA → RV→ PA
  - Left side – femoral artery → aorta → LV →LA (unless your patient has VSD or ASD that you can measure from the right side to the left)
- Identify risk - cardiac failure, respiratory compromise, chronic cachexia
- Adjust medications and therapies
- Digitalis must be withheld prior to surgery because of the possibility of cardiac failure post operatively.
- Intravenous vasoactive medications
- Parenteral nutrition
- Transport and Initial Stabilization
- Often with intravenous catheters, endotracheal tube, foley catheter, chest tubes
- Ensure:
  - Ventilation - monitor movement of the chest, breath sounds
  - Circulation - adequate heart rate, palpable pulses

ASSESSMENT AND MONITORING

I. Cardiovascular System

A. Physical Sign:

- **Adequate perfusion** - child alert and breathing comfortable with warm extremities, and normal peripheral pulses
- **Cardiac Output (CO) mildly compromised** - slight increase in heart rate, cool distal extremities, diminished capillary refill, decreased urine flow
- **CO moderately impaired** - tachycardia, restlessness, oliguria, cool extremities, faint peripheral pulses
- **CO severely curtailed** - child agitated or somnolent, cool trunk, cold mottled extremity, faint pulse, very rapid heart rate, minimal urine output or anuria

II. Central Nervous System

- Awareness of whether drugs were used that might interfere with neurologic function, level of consciousness, brainstem reflexes and gross motor response

Risk for complications:

a. Right-to-left shunt – embolism
   Ex. in ASD, thrombus from the RA to LA to aorta leading to thromboembolism.
b. Low cardiac output - cerebral ischemia
c. Muscle relaxants and heavy sedation - limited ability to respond

III. Respiratory System

Risk for complications:

a. **Pulmonary hypertension** - distal emphysema or collapse pulmonary overcirculation
b. **Difficult intraoperative care** - atelectasis, pooling of secretions
c. **Muscle relaxant or narcotics** - depress cough, interfere with mucociliary action

What to monitor:
1. Breathing pattern
2. Breath sounds
3. Type of ventilation (controlled, intermittent, mandatory)
4. Specific ventilator settings (rate, FIO_2, tidal volume, inspiratory and end-expiratory pressures)

Note the following:
- Lung fields
- Heart size
- Mediastinal width
- Endotracheal tube position
  - Seen through xray if there is preferential flow to the left/right due to position
- Position of central lines, chest tubes
- Arterial blood gas

IV. Hematologic System
a. Coagulation
b. Blood balance

Problems:
1. Intraoperative heparinization
When you insert a foreign body such as valve replacement to prevent coagulation.
2. Stored blood contains agent that chelate calcium
Remedy: 1 mg calcium gluconate/ml of blood transfused (Ca is needed for coagulation)
3. Massive blood transfusion - large load of acidic blood
   - Packed red blood cells - small amount of clotting factors - bleeding diathesis/tendencies
   - Platelets - aggregate in the lungs - impair gas exchange
   - Since there is hemolysis of blood as it passes through the heart-lung machine.
4. Hematocrit - bleeding post-operatively - blood loss due to sampling

V. Renal system
Best Guide: urine output
- Sodium concentration (hyponatremia may cause seizures)
- Adequate renal perfusion if the serum creatinine normal and urine output is 0.5 to 1 ml/kg/hr

VI. Metabolism
- Hypothermia - slow metabolism of drugs
- Hemodilution - increased total body water
May result to hemolysis of blood
- Fluid losses - insensible losses from overheated heater, loss through chest tubes, and mobilization of excess water
- Electrolyte changes - loss of potassium

MANAGEMENT

- General Care - fluid balance, metabolism, respiration
- Temperature - heater, blanket
In the OR, if the surgeon is comfortable, it means that the pediatric patient is not hypothermic (can lead to lactic acidosis)
- Glucose - 4-5g/kg/day- fluids with 10% dextrose
- Crystalloid- one-half the usual maintenance fluid requirements
- Plasma and blood - if patient has chest tube, dressings, blood sampling
  - IF active bleeding or poor clotting: packed rbc, fresh frozen plasma
  - IF hematocrit normal (30-40%): then give whole blood or colloid
  - IF hematocrit low (less than 30%): give packed rbc
  - IF hematocrit high (greater than 40): give colloid or crystalloid
  - For active bleeding, give whole blood.

- Mechanical ventilation
Criteria for extubation:
  - Adequate control of ventilation - ABG while patient breathes spontaneously
  - Only maximum of 14 days for the endotracheal tube to be in place then tracheostomy should be used.
    - Wean patients by turning off ventilator every 5, 10 or 15 minutes.
  - Ability to protect the airway and clear secretions - gag and cough
  - Patent upper and lower airways - chest x-ray, breathing pattern
  - Mechanical ability to take breaths and respire without great effort
- Reintubate if with retractions, tachypnea, agitation, hypoxemia

- Inadequate Perfusion
I. Can demands for blood flow be lowered?
Factors increasing demands:
  - Thermal stress (fever or cold)
  - Anemia
  - Hypoxemia
  - Agitation
  - Excessive work of breathing
II. Can cardiac output be augmented?

a. Preload *(end volumetric pressure stretching the right or left ventricle prior to contraction)*
   - insufficient intravascular volume
   - infuse colloids while monitoring blood pressure, heart rate and filling pressure

   Increase preload by giving fluids

b. Heart rate - first defense against low cardiac output (*except in digitalis toxicity, cardiomyopathy chronic disese, cachexia)*

   Decreased urine output comes late
   - Increase heart rate by atropine, adrenergic drugs, pacemaker
   - Tachyarryhthmia - secondary to hypoxemia, acidemia
   - Support of circulation, oxygen, correction of acidosis, electrical conversion

c. Contractility
   - Monitor by palpation of precordium, arterial pulse tracing, echocardiogram
   - Drug of choice- inotropic medications

d. Afterload
   - Reduction in ventricular volume and lowering of systemic blood pressure

   Decrease the afterload using the diuretics

POSTOPERATIVE MANAGEMENT

- Depends on the duration of cardiopulmonary bypass

The longer, the more complications

- Body temperature to 280°C to as much as 180°C in complex cases using blanket roll (hooked to a machine that will lower the body temperature) or by pouring ice during the operation.
- To assess if intraoperatively you have preserved the heart: heart is soft
- Duration of aortic cross-clamping

SVC and IVC are cross clamped that blood from the UE and LE derives oxygen from the heart lung machine and connected to the clamped aorta then distributed to the body. At risk for hypoxemia on prolonged duration

- Duration of profound hypothermia

COMPLICATIONS

- Respiratory failure - major post-operative complication

Especially atelectasis since the lungs is not used during the operation.

- Change in heart rate - 1st indication of serious complication- could indicate hemorrhage(inadequate closure of the heart, internal mammary arteries could have been severed), hypothermia, hypoventilation or heart failure
- Arrhythmia- complete heart block (usually temporary)

Usually in operations involving the SA/AV node like ASD/VSD closure. Usually returns to normal, otherwise pacemaker is needed.

- Heart failure
  - serious arrhythmia
  - myocardial injury
  - blood loss
  - hypervolemia/hypovolemia
  - significant residual hemodynamic abnormality

Ex. multiple VSD muscular type – requires cardiac catheterization to locate multiple VSD

- Acidosis
  - low cardiac output
  - renal failure
  - hypovolemia
  - renal failure
- Neurologic abnormalities
  - seizures
  - thromboembolism
- Postpericardiotomy syndrome
  - febrile illness associated with pericarditis and pleurisy(inflammation of the pleura), decreased appetite, nausea and vomiting
  - cardiac tamponade(fluid accumulation in the pericardium)
  - give salicylates, steroids
- Hemolysis -secondary to unusual turbulence of blood at increased pressure
- Infection - infection of the lung (post-operative atelectasis), subcutaneous tissues at the incision site, sternum (sternal osteomyelitis because of the reaction of the body to the suture) and urinary tract.

DEFECTS WHERE REPAIR IS THE ONLY OR BEST OPTION

Definitions from Schwartz 9th ed

- Atrial Septal Defect (should be closed)
- Aortic Stenosis
- Patent Ductus Arteriosus
- Aortic Coarctation
- Truncus Arteriosus
- Total Anomalous Pulmonary Venous Connection - abnormal drainage of the pulmonary veins into the right heart
- Cor Triatriatum - presence of a fibromuscular diaphragm that partitions the left atrium into two chambers

- Aortopulmonary Window - characterized by incomeplete development of the septum that normally divides the truncus into the aorta and the PA

SHUNTING PROCEDURES

Superior Vena Cava-Right Pulmonary Artery Shunt

- first successful cavopulmonary anastomosis

- Bidirectional Glenn - end-to-side RPA-to-SVC anastomosis
- Classic Glenn shunt - end-to-side right pulmonary artery (RPA)-to-superior vena cava (SVC) anastomosis with ligation of SVC–right atrial junction

DEFECTS REQUIRING PALLIATION (FIRST STEP BEFORE TOTAL CORRECTION)

- Tricuspid Atresia
- Hypoplastic left-heart syndrome

DEFECTS THAT MAY BE PALLIATED OR REPAIRED

- Ebstein's Anomaly
- Transposition of the Great Arteries
- Double-Outlet Right Ventricle
- Taussig-Bing Anomaly with /without pulmonary stenosis
- Tetralogy of Fallot
- Ventricular Septal Defect
- Atrioventricular Canal defects
- Interrupted Aortic Arch

SHUNTING PROCEDURES

Cyanosis – manifestation of decreased pulmonary blood flow

Palliation - To increase or decrease pulmonary blood flow by creating a shunt.

Increased pulmonary blood flow – use pulmonary artery banding and diuretics

Decreased pulmonary blood flow - create shunts

I. SYSTEMIC ARTERY – PULMONARY ARTERY SHUNTS

Subclavian Artery-Left Pulmonary Artery Shunt

- Aka "BLALOCK-TAUSSIG"
- Done in patients with decreased pulmonary blood flow, TOF, TGA in the absence of PDA

Results to an increased pulmonary blood flow

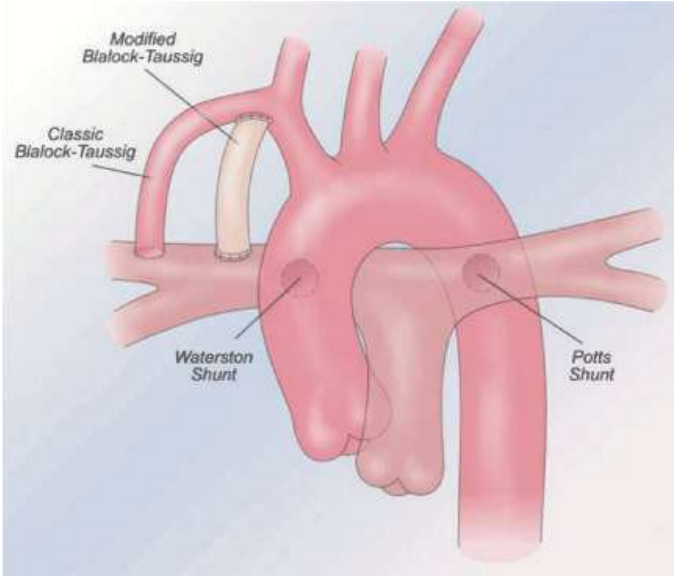
- Problems: Stenosis or thrombosis of shunt, pulmonary hypertension (uncommon)

Synthetic Interposition Grafts

- "MODIFIED BLALOCK-TAUSSIG"
- Use polytetrafluoroethylene (PTFE) graft from the subclavian artery to the pulmonary artery
- Problems: Congestive heart failure early, shunt later inadequate due to size restriction, kinking, thrombosis, growth of child

Advantage: if taken down for total correction, it will be easier to identify because it is a graft compared to classic type

- Disadvantage: Goretex (PTFE) does not grow, as the child grows, the graft does not grow. In later years, there will still be decreased pulmonary blood flow.



II. CENTRAL SHUNT

Ascending Aorta-Right Pulmonary Artery Shunt

- Aka "WATERSON"
- Problems: Kinking of pulmonary artery with obstruction of flow to lungs, enlargement of anastomosis, pulmonary hypertension in perfused lung and decreased flow to contralateral lung.

Descending Aorta-Left Pulmonary Artery Shunt

- Aka "POTTS"
- Feasible only if the aorta descends on the left side
- Associated with premature closure of shunt, enlargement of anastomosis, pulmonary hypertension earlier

III. SYSTEMIC VEIN – PULMONARY ARTERY SHUNT

Superior Vena Cava-Right Pulmonary Artery Shunt

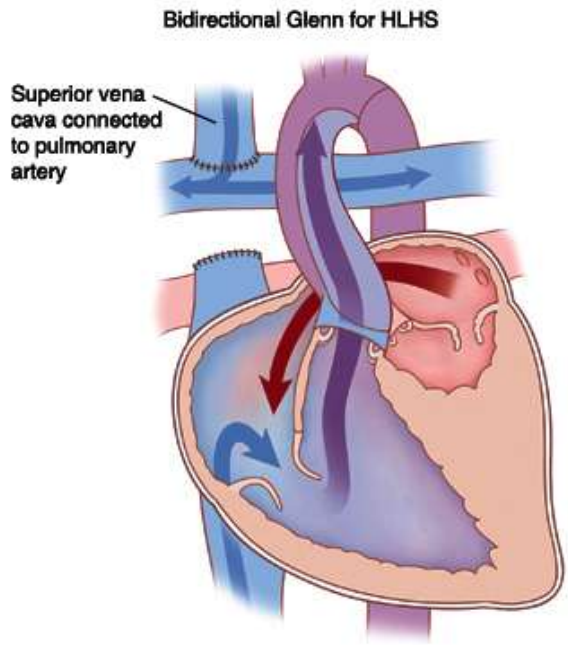
- Aka "GLENN SHUNT"

- Decreased flow to lung due to A-V fistulization in lower lobe, venous collaterals to IVC, decreased flow to left lung, polycythemia
- Problems: increased collateral circulation, difficulty taking down the shunt during the total correction
- Indication:
  - Absence or hypoplasia or evidence of obstructive pulmonary disease localized to the left PA
  - Where it is apparent that a combination of a stable arterial shunt to the left lung and vena cava – PA shunt to the right will produce optimum oxygenation

Seldom used as a palliative procedure. This shunt should be closed during total correction of the CHD

- **DIMINISH EFFECT**

- Development of collateral veins to the inferior vena cava
- Widening of A-V connection in the right lower lobe
- Decrease in blood flow to the left unshunted lung
- Fistulization at the vena cava – right atrial junction
- Polycythemia – *can result to thrombosis*



PULMONARY ARTERY BANDING

- Purpose: limit pulmonary blood flow
- Done in patients with Large VSD, Atrioventricular Canal Defects, Truncus Arteriosus, Tricuspid Atresia
- Material: Teflon Band
- Factors to consider:
  - Pulmonary & Systemic resistance
  - Performance of the myocardium
  - Postoperative care
  - Accuracy of the surgery
- Consequences:
  - Obliteration of the pericardial space by adhesions
  - Thickening of the pulmonary valve
  - Narrowing of one or both branches of the pulmonary artery
  - Closure of large VSD
  - Development of subaortic stenosis

Teflon band may migrate proximally, distally or cause stenosis leading to cyanosis.

EVALUATION OF THE INFANT OR CHILD WITH CHD

- Congenital defects can be divided into two major groups based on the presence or absence of cyanosis
- Subdivided whether the chest radiograph shows evidence of increased, normal or decreased pulmonary vascular markings
- Electrocardiogram determine whether right, left or biventricular hypertrophy axis

ACYANOTIC CONGENITAL HEART LESIONS

I. Lesions Resulting in Increased Volume Load

- ATRIAL SEPTAL DEFECT , AV CANAL, PATENT DUCTUS ARTERIOSUS

ASD 3 TYPES :
1. Sinus Venosus Defects -5-10% OF ALL ASD
2. Ostium Primum Defects
3. Ostium Secundum Defects

- Communication between systemic & pulmonary sides of the circulation resulting in shunting of fully oxygenated blood back into the lungs
- Before surgery: Calculate the ratio of pulmonary to systemic blood flow --- Qp:Qs (normally equal)
- Direction and magnitude depends on the size of the defect and the relative pulmonary and systemic pressures and vascular resistance
- Increased volume of blood in the lung → decreased pulmonary compliance → increases work of breathing → (+) fluid leaks into the interstitial space and alveoli → pulmonary edema
- Heart failure → tachypnea, chest retraction, nasal flaring *(manifested early in life)*
- Increased work of breathing → Increased total body oxygen consumption → sweating, irritability and failure to thrive

Remember that severe pulmonary hypertension is a contraindication to definitive surgery in patients with CHD

II. Lesions Resulting in Increased Pressure Load – obstruction to normal blood flow

- Obstruction to ventricular outflow:
  - ➢ PULMONARY STENOSIS
  - Critical Pulmonary Stenosis (very small orifice) in newborn presents as right-sided heart failure → hepatomegaly, peripheral edema, cyanosis (shunting across foramen ovale)
  - ➢ AORTIC STENOSIS
  - Critical Aortic Stenosis(very small orifice) in newborn presents as left-sided heart failure → pulmonary edema, poor perfusion, and right-sided heart failure
  - ➢ COARCTATION OF THE AORTA

- Obstruction to ventricular inflow:
  - ➢ TRICUSPID STENOSIS
  - ➢ MITRAL STENOSIS
  - As a congenital defect, it is seldom seen
  - More often a complication of rheumatic heart disease

CYANOTIC CONGENITAL HEART DISEASE

I. DECREASED PULMONARY BLOOD FLOW

- Obstruction to pulmonary blood flow and a pathway by which systemic venous blood can shunt from right to left
  - ➢ TRICUSPID ATRESIA
  - ➢ TETRALOGY OF FALLOT

II. INCREASED PULMONARY BLOOD FLOW

- Cyanosis caused by either abnormal ventricular-arterial connections or by total mixing of systemic venous and pulmonary venous blood within the heart
  - ➢ TOTAL ANOMALOUS PULMONARY VENOUS RETURN (TAPVR)
  - ➢ TRUNCUS ARTERIOSUS
  - ➢ TETRALOGY OF FALLOT

VENTRICULAR SEPTAL DEFECT

- Most common: 25% of CHD
- Defects occur in any portion if ventricular septum; the majority are of the membranous type
- Location: Anterior to the septal leaflet of tricuspid valve
  - Between supraventricularis and papillary muscle of conus
- Location: Superior to crista supraventricularis
  - Just beneath the pulmonary valve and may impinge on an aortic sinus
  - Midportion or apical region of the ventricular septum – muscular type – single or multiple(catheterization is indicated due to multiple VSD)

DETERMINANT OF THE SIZE OF SHUNT

- Size of VSD
- Level of PVR compared with SVR, *O_2 saturation*
- Small, <0.5 cm – aka restrictive
  - RV pressure is normal
  - *No indication for immediate medical intervention*
- >1.0 cm – aka nonrestrictive
  - RV & LV pressures are equalized
- Pulmonic Vascular Resistance (PVR): Systemic Vasuclar Resistance (SVR) = 1:1
  - The shunt becomes bidirectional
  - Signs of heart failure abate and patient becomes cyanotic
- Small Shunt
  - Qp:Qs <1.75
  - Cardiac chambers not enlarged
  - Pulmonary vascular bed are normal
- Large Shunt
  - Qp:Qs >2:1
  - Left atrial & ventricular volume overload occurs
  - Enlarged main pulmonary artery, left atrium, left ventricle

CLINICAL MANIFESTATION

- Small VSD
  - Asymptomatic
  - Found on routine physical exam
- Large VSD
  - Excessive pulmonary blood flow and pulmonary hypertension
  - Dyspnea, feeding difficulties, poor growth, profuse perspiration, recurrent pulmonary infection, cardiac failure in early infancy
  - Duskiness during infections or crying
  - Prominence of left precordium and palpable parasternal lift
  - Laterally displaced apical impulse
  - Increase pulmonic component of 2nd sound
    - Pulmonary hypertension
  - Less harsh holosystolic murmur

- Mid-diastolic low-pitched rumble at the apex – because increased blood flow across mitral valve

DIAGNOSIS

- Small VSD
  - Chest radiograph – normal
  - ECG – normal
- Large VSD
  - ➢ Chest radiograph
  - Cardiomegaly
  - Increased pulmonary markings
  - ➢ ECG
  - Biventricular hypertrophy
- 2D Echocardiogram
  - Position & size of VSD
  - Estimate shunt size
  - Associated lesions
  - Calculate pressure gradient
- Cardiac Catheterization

Important to identify multiple VSD

- Complications: premature rupture of balloon, air embolism

PROGNOSIS

- Small defect – 30-35% spontaneous closure
  - Small muscular defect are more likely (80%) to close than membranous (35%)
- Large defect – less common to close

Advise surgical intervention at an early age to prevent heart failure. If large defect, right away advise VSD closure. If small defect, observe and monitor. Advise not to become hypoxemic because VSD won't close.

TREATMENT

- Small VSD
  - No restrictions of physical activity
  - Surgery not recommended
  - Protection against infective endocarditis

Patient should have antibiotics before any interventionand dental clearance.

- Indications for surgery:
  - Large defects with clinical symptoms and failure to thrive which cannot be controlled medically
  - Infants between 6-12 months with large defects, with pulmonary hypertension
  - Patients older than 24 months with Qp:Qs > 2:1
  - Supracritical VSD – high risk of Aortic Insufficiency because of proximity.

- ✓ Sources: slides, Schwartz, audio
- ✓ By M. Prado, R. Gabor, K. Carvajal, N. Sameon