Neonatal Cardiac Anomalies

Karen Knuth, RNC, MN, NNP-BC, ARNP
Seattle Childrens Hospital

Objectives

• What is CHD?
• Normal anatomy and circulation
• Clinical presentation: signs and symptoms
• Diagnostics
• Common heart defects in the newborn period and in the NICU
• Treat / transport?
• Case studies

Congenital Heart Defects (CHD)

• A failure of fetal cardiac development
• Critical CHD: needs surgery or catheter intervention in the first year of life
• Incidence: ~1% or 6 -13:1000 live births
• Most common congenital disorder in newborns
• Second leading cause of death among children in the first year of life
• Failure to diagnose sooner: increased morbidity and mortality

Causes?

• Preterm infant
  o 2-3x term
• Chromosomal
  o Trisomy 21 – associated with VSD/AV canal
• Genetic
  o 3x risk if CHD in first deg relative
• Environment/maternal factors
  o Diabetes, lupus, obesity, hypertension, CHD, thyroid conditions, epilepsy/mood disorders, fever/flu/illness, smoking in the first trimester
• Recurrence risk
  o Higher if previous child, mother, then father
• Multifactorial

General Principles

• Blood flows in the path of least resistance
• Blood flows from higher pressure to lower pressure
• In utero blood flow
  o Functions
  o Shunts
• After birth
  o Shunts close, series circulation

Transitional Circulation

• Parallel becomes series
• Functional closure of ductus venosus and foramen ovale
• Anatomic closure ductus arteriosus at 48 – 96h of age
• Pulmonary vascular resistance decreases suddenly, then more slowly

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Cardiac Output - CO

\[ CO = HR \times SV \]

- **CO** = the amount of blood the heart pumps in one minute (litres)
- **SV** = preload, afterload, contractility
- **HR** = beats per minute

Normal Heart

Clinical Presentation

- Central cyanosis +/- crying
- Low/lower oxygen saturations
- Differing pre and post ductal oxygen saturations
- Respiratory distress – tachypnea common
- Pulses: weak vs bounding vs unequal vs absent
- Blood pressure: wide, differing upper/lower, left vs right
- Murmurs
- Poor feeding – tires out → failure to thrive?
- Hepatomegaly
- Congestive Heart Failure / Cardiogenic shock
- Death

Timing

- Prenatal screening <25%
- If not prenatally diagnosed may present during birth hospitalization
  - Circulation changes
  - Critical timing for ductal closure
- ~30% defects not diagnosed
- Examples of ductal dependent CHD missed: COA, TGA, HLHS, TAPVR
- Non-ductal dependent lesions missed (mild desats or tachypnea initially): TA, TOF, TAPVR
- Closure of ductus can lead to shock and death
- If not diagnosed during birth hospitalization, many within a couple of weeks
  - Mean age of death if missed was 13.5 days (HLHS and COA)
  - TGA is most commonly presented in the first 30 days
  - Tetralogy of Fallot is most common diagnosis after 1 month of age

Blue Babies / Cyanosis

- Cyanosis may or may not be related to CHD
  - See summary table from up to date, next slide
- Presence and timing of cyanosis are important in diagnosing CHD
  - Critical lesions are those that need accessory circulation (the ductus) to provide oxygen to body/organ
- Within the first few days profound cyanosis can occur with the following:
  - Critical pulmonary stenosis or atresia
  - Obstructive left heart lesions (HLH, Ao Valve Stenosis)
  - Transposition of the Great Arteries
Congestive Heart Failure - CHF

- Blood supply to the body is inadequate to meet the metabolic demands of the organs
- A manifestation of an underlying disease or defect
- Signs and symptoms:
  - Tachycardia
  - Cardiomegaly, active precordium
  - Gallop
  - Decreased peripheral pulses
  - Skin mottling
  - Decreased urine output
  - Sweating
  - Enlarged liver
  - Lethargy
  - Feeding problems
  - Failure to thrive

CHF - Onset

- At birth
  - HLHS
  - Severe tricuspid or pulmonic regurgitation
  - Systemic AV fistula
- Week one
  - PDA in preterm infants
  - TGV
  - TAPVR with pulmonary venous obstruction
- Weeks one to four
  - Coarctation Aorta
  - Critical aortic or pulmonary stenosis
  - All of above

Shock

- Inadequate tissue perfusion, ↓ oxygen delivery
- Several types
- Cardiogenic shock: pump failure
- Can lead to end organ damage, organ failure (multiple), and death
- Compensation: ↑ HR, ↑ SVR, ↑ venous tone
  - BP normal
  - Tissue perfusion poor
  - Tachycardia, tachypneic
- Hypotension late sign

Cardiogenic Shock

- Goal is to prevent this
- Compensation makes it difficult to treat before hypotension develops and irreversible downward spiral occurs
- Cardiomyopathy
  - Uncommon in children, especially neonate. Occurs in IDM, birth depression
- Arrhythmia
  - Too fast or too slow
- Obstructive
  - Tension pneumothorax, CHD (e.g.: COA, HLHS) – when duct closes
Signs of Shock

- See signs of CHF
- Hypotension late and ominous sign

Severe Pulmonary Edema

- Tachypnea
- Increased work of breathing
- Occurs with rapid and large increase in pulmonary blood flow when the PVR falls
  - I.e.: Truncus arteriosus
  - PDA in preterm infant
  - TAPVR with obstruction

Murmurs

- Absence of healthy heart
- Timing: systolic vs diastolic vs continuous
- Quality: harsh, blowing, musical
- Pitch: high vs low
- Location
  - Radiation?
- Intensity: grading

Grading
- I: barely audible
- II: soft, easily audible
- III: moderately loud, no thrill (palpation)
- IV: loud + thrill
- V: very loud + thrill, stethoscope just touching chest
- VI: very LOUD + thrill, stethoscope off chest wall

Persistent Pulmonary Stenosis

- Innocent or non-pathologic murmur
- Due to small size of branch pulmonary arteries and angle of pulmonary arteries
- Resolves in 3 – 6 months
- Heard best at base of heart, radiates to axillae, and back
- Can be very loud

Murmurs

- Absence ≠ no CHD
- Innocent
  - During first 48h of life
  - Usually Grade I – II
  - Usually systolic
  - No associated symptoms
- Pathologic
  - Persist, beyond 48h
  - Occur when PVR falls; day 3, 7, 14
  - May be louder than Grade II
  - May be diastolic
  - http://www.texasheart.org/education/cme/explore/events/eventdetail_5469.cfm

Diagnostic Studies

- Blood pressures:
  - Singular (hypotension)
  - Right upper extremity and lower extremity (differences)
- Pulse oximetry
  - Routine one extremity (right upper)
  - Pre/post: to detect differences “pre and post” ductaly
  - A.K.A. Congenital Heart Defect (CHD) Screening
    - Critical lesions (7) that cause hypoxia - HLHS, PA, TOF, TAPVR, TGA, TA, HAC 24h of age or prior to discharge (later better)
    - Calm baby
    - Good equipment and signal
    - Positive (not good) or negative
Diagnostic Studies

- **Hyperoxia Test:**
  - PaO2 on ABG >150mmHg with 100% FiO2, not CHD
- **Chest x-ray**:
  - Looking at lung fields, heart size, shape, and location
- **EKG**:
  - Adjunct: normal R side enlargement, with CHD would have severe L sided enlargement, rhythm disturbances
- **ECHO**:
  - Using ultrasonic waves to examine structure, function, pressures, and flow gradients
- **Cardiac Catheterization**:
  - More therapeutic (ie: balloon septostomy) than diagnostic these days
- **MRI**:
  - Rarely used

Lab Data

- **Electrolytes**: rule out other cause of heart disturbance: calcium, sugar, potassium
  - Worsening CHF - hyponatremia
- **ABG**: normal CO2: initially, then can rise
- **Metabolic**: lactic acidosis
- **CBC**: infection, polycythemia, anemia
- **Renal function**: BUN, Creatinine, Sodium

X-ray Appearance

- Size of heart
- Pulmonary vascularity
  - Heart Lesions with increased vascularity
  - Heart Lesions with decreased vascularity
- Position of the heart

Types

- Cyanotic versus acyanotic (BLUE vs PINK)
  - Mixed?
- Timing – of presentation
  - Increased and decreased vascularity
- Obstructive with and without pulmonary venous congestion

Acyanotic Heart Defects

- Passage between left and right heart
- Left to right shunting
- Pulmonary overcirculation and CHF
- Some respiratory distress
- Typically don’t present until Pulm resistance has (PVR) fallen at 4 – 6 weeks of age
  - Exception PDA in preterm and AV canal
- Examples
  - Patent Ductus Arteriosus (PDA)
  - Atrial Septal Defect (ASD)
  - Ventricular Septal Defect (VSD)
  - Endocardial Cushion Defect (ECD)

Patent Ductus Arteriosus - PDA

- In utero shunt
- Persistent vascular communication from the pulmonary artery to the aorta
- Blood flows from the aorta into the pulmonary artery
- May lead to pulmonary edema and respiratory distress
- Common cause of murmur after birth
Atrial Septal Defect - ASD

• Defect in the formation of the septum
  o primum, secundum, or partial ECD
• Results in a communication between right and left atria

Ventricular Septal Defect – VSD

• Imperfect division in fetal development results in one or more holes in the septum of the ventricles
• Anywhere
• Pinhole to entire septum
• Membranous most common
• Murmur

Endocardial Cushion Defect - ECD

• Abnormal development of endocardial cushion area or junction between the four chambers of the heart
• Extent varies
  o Partial or complete
  o Simple defect is a defect in the lower portion of the atrial septum
  o Extensive defect is known as a complete atrioventricular canal AV canal

Cyanotic Heart Defects

• 25% of heart defects
• Right to left shunting
• Arterial oxygen saturation unable to rise
• Results in:
  o chronic hypoxemia which causes
  o RBC production to increase
  o promoting increased oxygen carrying capacity (polycythemia)
  o blood is hyper viscous → poor tissue perfusion

Examples:
• Transposition of the Great Vessels (TGV)
• Truncus Arteriosus
• Total Anomalous Pulmonary Venous Return (TAPVR)
• Tetralogy of Fallot (TOF)
• Tricuspid Atresia

Transposition of the Great Vessels - TGV

• Position of great arteries are reversed:
  o Aorta originates from the right ventricle
  o Pulmonary artery originates from the left ventricle
  o Aorta is positioned anteriorly to the pulmonary artery
  →two separate circulations, a parallel circuit
• Compatible with life only when there is a shunt or communication between the two circuits that allows mixing of the blood
• Shunts:
  o PDA
  o ASD
  o VSD – best mixing
Truncus Arteriosus

- A single vessel arising from the heart that forms the aorta and pulmonary artery.
- Another congenital heart defect that occurs with truncus arteriosus is a ventricular septal defect.
- Several types

Total Anomalous Pulmonary Venous Return - TAPVR

- No connection of pulmonary veins with left atrium – they drain directly or indirectly into the right atrium via a systemic (body) channel.
- The connection occurs either directly to the right atrium or by way of systemic veins that empty into the vena cava above or below the heart.

Oxygenated blood from the lungs can’t get to the body

Four types:
- Most commonly supracardiac
- Cardiac
- Infracardiac
- Mixed
Tetralogy of Fallot - TOF
- Pulmonary stenosis
- VSD (large)
- Overriding aorta
- Hypertrophy of the right ventricle

Tricuspid Atresia
- Agenesis of the valve
- Associated with ASD/PFO
- Right ventricular is usually hypoplastic
- Can have complex variations

Obstructive Heart Lesions
- Lesions that obstruct blood flow from the heart may involve either the left or the right side of the heart
- Major clinical findings reflect the increased work of the heart against an obstruction, i.e.: right ventricular hypertrophy

Examples:
- Aortic Stenosis
- Pulmonary Stenosis
- Coarctation of the Aorta

Aortic Stenosis
- Four ways:
  1. Thickenings and fusion of the valve cusps (valvular stenosis)
  2. Localized constriction or diffuse narrowing above the valve
  3. Obstruction membrane or fibromuscular ring below the valve
  4. Thickening of the ventricular septum
Pulmonary Stenosis

- Narrow opening in pulmonary valve due to cusp fusions
- Can be above or below the valve because of tissue

Coarctation of the Aorta

- Narrowed lumen, usually by the site of the ductus arteriosus
- Bicuspid aortic valve occurs in about half the patients
- VSDs are common

Other defects

- Hypoplastic Left Heart Syndrome (HLHS)
- Ebstein’s Anomaly

Hypoplastic Left Heart Syndrome - HLHS

Spectrum of:
- Severe coarctation of the aorta
- Severe mitral stenosis or atresia
- Severe aortic stenosis or atresia
- L.V. hypoplasia
- Retrograde coronary artery flow

Ebstein’s Anomaly
CHDs - Present Later

- Often by 2 weeks
- Routine pediatrician follow up
- Most common presentation is difficulty with feeding
  - Intake poor, LONG feeding, choking, gagging, vomiting
  - Cyanosis, pallor, mottling
  - Excessive, unexplained irritability
  - Sweating
  - Poor growth/weight gain
  - Lethargy, sleeping a lot
  - Diminished pulses/cool feet/mottling lower/↓UOP
- At 6 weeks: murmur can lead to dx (VSD most common)
- Some coarctation (COA) not diagnosed until 3 mos

Evaluation

- CXR
- EKG
- Hyperoxia test if cyanotic +/- respiratory distress
- Echocardiogram
- Genetic labs (ie: chromosomes, trisomy, i.e.: 22q11)
- Electrolytes, C&B, blood gasses, lactate, electrolytes
- Arrange for transport

Treatment

- Depends on the lesion, size, degree of circulatory impairment
- PDA
  - Newborn: normal finding, wait
  - Clinical strategies with preterm infants
  - Ibuprofen/indomethacin/tylenol
  - May need surgery
- VSD
  - Small – most likely no treatment, cardiology follow up
  - Small-moderate: cardiology follow up
  - Large-cardiology, possible meds (diuretics, beta-blockers, digoxin). At risk for heart failure
  - May need to have surgical closure

Treatment - Critical CHD

- ABC’s always
- Ventilation strategies usually ‘gentle’
- Pre/post ductal saturations
- Provide oxygen to minimize hypoxia
  - may not alleviate cyanosis.
  - Can usually anticipate oxygen saturations less than 90% (70-80’s range may be acceptable).
  - Oxygen may not be indicated for some defects (Left sided lesions – HLHS, PA or PS), but is usually okay to use until exact defect is confirmed by ECHO (TAPVR).
- Consult cardiac center
- Stabilize prior to transport

Stabilizing Infant with CHD

- IV access
- Watch fluid intake: usually limited
- Maintain neutral pH: correct acidosis if necessary
- Sedation prn
- Provide hemodynamic support
  - 10ml/kg of N.S. or L.R.
  - Dopamine (3-10 mcg/kg/min) to improve cardiac output
  - Doutamine or milrinone
- Later, Digoxin and diuretics may be indicated
- Prostaglandins

PGE 1

- Administer to maintain a patent ductus arteriosus and prevent shock etc...
  - if ductal dependent lesion is suspected
  - Relaxes ALL arterioles
  - Dose is 0.05 – 0.1 mcg/kg/min via continuous infusion (0.03 common)
  - Max effect within 30 mins – several hours
  - Prime the tubing all the way down to the baby to ensure that the baby gets the drug ASAP.
  - Cutaneous flushing, inatibility, bradycardia
  - Fever and apnea occur in 12% of the cases
  - Be prepared to intubate
THANK YOU!