How to Recognize a Suspected Cardiac Defect in the Neonate

Purpose and Goal: CNEP # 2092

- Understand the signs of congenital heart defects in the neonate.
- Learn to recognize and detect heart defects in the neonate.

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Requirements for successful completion:

- Successfully complete the post-test
- Complete the evaluation form
Date

- December 2018 – December 2020

Learning Objectives

- Describe the risk factors for congenital heart defects.
- Describe the clinical features of suspected heart defects.
- Identify 2 approaches for recognizing congenital heart defects.

Introduction

- Congenital heart defects may be seen at birth
- They are the most common congenital defect
- They are the leading cause of neonatal death
- Many neonates present with symptoms at birth
  - Some may present after discharge
- Early recognition of CHD improves outcomes

Congenital Heart Defects

- Congenital heart defects are also known as CHD
- CHD occurs in 6-13/1000 live births
  - 15% occur as cyanotic defects
  - 25-33% occur as critical defects
- Up to 58% of CHD may be diagnosed prenatally
  - Prenatal ECHO is highly variable
  - Its sensitivity depends on:
    - Operator expertise
    - Gestational age
• Fetal position
• Type of defect
• CHD may be referred to as:
  • Cyanotic CHD
  • Ductal-dependent CHD
  • Critical CHD
• When the diagnosis of CHD is delayed:
  • The risk of morbidity increases
  • The risk of mortality increases

Types of Congenital Heart Defects

• CHD may be classified as:
  • Cyanotic CHD
  • Ductal-dependent CHD
  • Critical CHD
• Cyanotic heart defects
  • Intra or extra cardiac shunting
  • Circulate deoxygenated blood
• Ductal-Dependent heart defects
  • Dependent on a patent ductus arteriosus
  • To allow mixing of blood
    • Oxygenated
    • Deoxygenated
  • Many cyanotic defects are ductal dependent
• Critical heart defects
  • Require intervention
    • Catheter intervention
    • Cardiac surgery

Risk Factors for Congenital Heart Defects

• There are several risk factors for CHD
  • Family history
  • Multiple fetuses
- Genetic syndromes
  - In 7-12% of CHD
  - Most common in:
    - Trisomy 21
    - Turner syndrome
    - DiGeorge Deletion 22q

- Maternal factors
  - Obesity
  - Diabetes
  - Epilepsy
  - Hypertension
  - Preeclampsia
  - Thyroid disorders
  - Phenylketonuria
  - Mood disorders
  - Connective tissue disorders
  - Advanced age >40
  - Alcohol or substance use
    - Amphetamines
  - First trimester tobacco use

- Medications
  - NSAIDs
  - ACE inhibitors
  - Retinoic acid
  - Thalidomide
  - Phenytoin
  - Lithium

- In utero infections
  - Rubella
  - Coxsackie virus
  - Cytomegalovirus
  - Ebstein-Barr virus
  - Toxoplasmosis
  - Parvo virus B19
  - Herpes simplex virus
  - Flu-like illness

- Assisted reproductive technology
- There are several risk factors for ↑ survival
• Earlier diagnosis
• Lower birth weight
• Maternal age <30

Clinical Features of Congenital Heart Defects

• Some infants may present without symptoms
• Some present with immediate onset of symptoms
  • Shock
  • Cyanosis
  • Tachypnea
  • Pulmonary edema
• Shock may be seen in several types of CHD
  • Hypoplastic left heart syndrome
  • Critical aortic valve stenosis
  • Critical coarctation of the aorta
  • Interrupted aortic arch
• When infants present with shock:
  • Septic shock must be ruled out
  • Cardiogenic shock is suggested when:
    • Cardiomegaly is present on x-ray
    • Volume resuscitation is unsuccessful
• Cyanosis may be seen in several types of CHD
  • Pulmonary atresia
  • Ebstein’s anomaly
  • Truncus arteriosis
  • Tetralogy of Fallot
  • Ductal dependent lesions
  • Pulmonary valve atresia
  • Critical pulmonary valve stenosis
  • Hypoplastic left heart syndrome
  • Transposition of the great arteries
  • Total anomalous pulmonary venous return
• When infants present with cyanosis:
  • Non-cardiac causes must be ruled out
    • Sepsis
• Pulmonary
• Hypoglycemia
• Dehydration
• Hypoadrenalism
• Rare causes:
  • Methemoglobinemia
  • Metabolic disorders
• Cyanosis is suggested when:
  • Pulse oximetry saturations are <80s%
  • Pre and post-ductal saturations are different
    • A >3% difference is abnormal
• Tachypnea may be seen in several types of CHD
  • Truncus arteriosus
  • Patent ductus arteriosus
  • Large ventricular septal defects
  • Total anomalous pulmonary venous connection
• When infants present with respiratory distress:
  • Non-cardiac causes must be ruled out
    • Sepsis
    • Pulmonary
    • Hypoglycemia
    • Dehydration
    • Abnormal forms of hemoglobin

**When to Be Suspicious of a Heart Defect**

• CHD should be suspected with:
  • Family history of CHD
  • Abnormal fetal ECHO
  • Failed CCHD screens
• CHD should also be suspected with:
  • Heart murmur
  • Central cyanosis
  • Comfortable tachypnea
  • Comfortable desaturations
  • Increased CRT >3 seconds
• Associated anomalies
  • Trisomy 21
  • Skeletal anomalies
    • Hand and arm
  • CHARGE syndrome
  • Ear anomalies
  • Renal anomalies

Initial Diagnosis of Congenital Heart Defects

• Initial diagnosis is based on:
  • History
  • Physical findings
  • Chest x-rays
  • Hyperoxia test
  • Echocardiogram

• History
  • Risk factors
  • Poor feeding
  • Color changes
  • Excessive irritability
  • Excessive sweating
  • Poor weight gain
  • Excessive sleeping

• Physical findings include:
  • Abnormal heart rate
  • Comfortable tachypnea
  • Abnormal heart sounds
  • Abnormal precordial activity
  • Abnormal oxygen saturation
    • <90% in any extremity
  • Oxygen saturation gradient
    • >3% difference in extremities
  • Blood pressure gradient
    • >10 mmHg higher in arms
    • >10 mmHg lower in legs
• Abnormal femoral pulses
  • Weakened pulses
  • Absent pulses
• Hepatomegaly
• Chest x-rays
  • Heart size and shape
    • Dextrocardia
    • Enlarged heart size
      • >60% of the chest
    • Boot shaped heart
    • Egg shaped heart
    • Snowman shaped heart
• Pulmonary vascular markings
  • Decreased markings
    • ↓ pulmonary blood flow
  • Asymmetric markings
  • Pulmonary congestion
    • ↑ pulmonary blood flow
• Site of the aortic arch
  • Left sided arch is normal
  • Right sided arch abnormal
• Hyperoxia test
  • Useful in ruling out pulmonary causes
    • Obtain an ABG in room air
      • Right radial artery
    • Provide 100% oxygen for 10 minutes
    • Obtain an ABG in oxygen
      • Right radial artery
      • An ↑ O2 should be seen
      • pO2 should be >150
    • No significant ↑ in O2 is abnormal
• Electrocardiograms
  • A large right ventricle is normal
    • Right ventricular hypertrophy
  • Other presentations suggest CHD
    • A small right ventricle is abnormal
    • A large left ventricle is abnormal
      • Left ventricular hypertrophy
Classification of Abnormal Heart Sounds

- Murmurs may or may or be associated with CHD
- Murmurs associated with CHD include:
  - ≥ grade 3 intensity
  - Holosystolic timing
  - Maximum intensity
    - At upper left sternal border
    - With upright positioning
  - Diastolic murmur
  - Harsh or blowing quality
- Grading of murmurs:
  - Grade 1 murmur
    - Faint sound detected
    - Often only audible to cardiologists
  - Grade 2 murmur
    - Soft murmur
    - Readily detected
  - Grade 3 murmur
    - Louder than grade 2
    - Not associated with palpable thrill
  - Grade 4 murmur
    - Easily detected murmur
    - Associated with palpable thrill
  - Grade 5 murmur
    - Very loud murmur
    - Easily audible with stethoscope
  - Grade 6 murmur
    - Extremely loud murmur
    - Easily audible with stethoscope off chest

Specific Congenital Heart Defect Care

- Specific care is indicated for infants who are:
• Cyanotic
• Fail a hyperoxia test
• Do not have PPHN
• Do not have lung disease on x-ray
• In most cases, these infants have:
  • Cyanotic heart disease
  • A ductal dependent heart defect
  • An increased risk of significant morbidity
  • An increased risk of death
• The ductus arteriosus must be kept open
  • To ensure mixing of oxygenated blood
  • To ensure mixing of deoxygenated blood
• Prostaglandin should be started prior to an ECHO
  • The initial dose should be 0.01 mcg/kg/min
  • Dosing may be ↑ to 0.05 mcg/kg as needed
  • A Cardiology consult should be obtained
• An ECHO is not needed to treat these infants
• Transfer to a referral center should be immediate

**Echocardiogram Examination**

• Echocardiogram imaging is definitive
• It provides information on cardiac:
  • Anatomy
  • Function
• It can also evaluate for pulmonary causes
• The ECHO should include:
  • Cardiac imaging
  • Pulsed Doppler flow
  • Color Doppler flow
• ECHOs should be done in consultation with a cardiologist
• All ECHOs should be interpreted by a pediatric cardiologist

**Summary**

• CHDs are the most common defect in the neonate
• They are a leading cause of morbidity and mortality
• Early recognition and identification is critical
• Several clinical signs are suspicious for CHD
• Alert neonatal nurses can identify infants at risk

References


