Screening and Initial Stabilization of Critical Congenital Heart Disease

KAAP Progress in Pediatrics Spring 2019

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Objectives

- Review the AAP critical congenital heart disease (CCHD) screening algorithm and rationale
- Review classification of critical congenital heart disease and specific lesions
- Discuss initial stabilization of a newborn with CCHD
- Follow a case example throughout the presentation

Critical Congenital Heart Disease

- ~1% of all newborns have a congenital heart defect (CHD)
- 18 per 10,000 newborns have critical CHD (CCHD)
  - Life threatening CHD requiring neonatal catheter-based intervention or heart surgery
- AAP recommends that all newborns be screened for CCHD
Case Example

- Well appearing term male infant, 3.6 kg
- Normal newborn course
- At ~24 hours of age, CCHD test was performed
  - Right hand saturation 80%, right foot saturation 84%

CCHD Screening Algorithm

- Work group of US Health and Human Services Secretary’s Advisory Committee on Heritable Disorders in Newborns and Children, AAP, American College of Cardiology Foundation, American Heart Association
- **Goal:** to identify newborns with CHD usually associated with neonatal hypoxia that could have significant early morbidity/mortality with PDA closure
CCHD Screening Algorithm

- Screening should occur in right hand and either foot
- Rarely a baby will require 3 screens, separated by 1 hour
Failed CCHD Screen

- Any oxygen saturation <90% (initial or repeat screens)
- Oxygen saturation <95% in right hand and foot on 3 measures, each separated by 1 hour
- A >3% absolute difference in oxygen saturation between right hand and foot on 3 measures, each separated by one hour

https://www.cdc.gov/ncbddd/heartdefects/hcp.html

Kemper AR et al. Pediatrics 2011
If a baby fails the CCHD screen…

- First, examine the infant to ensure hemodynamic stability
- Next, evaluate for any non-cardiac causes of hypoxemia
  - Sepsis
  - Pneumonia
- Any signs or symptoms of CHD should prompt rapid evaluation \(\rightarrow\) possible urgent transfer

If a baby fails the CCHD screen…

- If no obvious cause for hypoxemia, consult neonatology or cardiology
  - Echocardiogram should be performed
- Newborns should not be discharged until the underlying reason for hypoxemia identified or resolved
  - May require transfer
Why Screen?

- Cyanosis of cardiac origin must be diagnosed early
- Detection of mild cyanosis is difficult
- Acrocyanosis is normal in newborns → confusion
- Cyanosis due to lung disease/CNS disorders → crying may improve cyanosis
- Cyanosis in CHD → crying may worsen the cyanosis

13 eligible studies; data for 229,421 newborns
Sensitivity 76.5%, specificity 99.9%, false positive rate 0.14%
Conclusion: pulse oximetry is highly specific for detection of critical CHD with moderate sensitivity, that meets criteria for universal screening
Why Screen?

- $6.28 per newborn, incremental costs of $20,862 per newborn with CCHD detected at birth hospitals, and $40,385 per life-year gained
- 1189 more newborns with critical CHD identified at birth hospitals; 20 infant deaths averted annually with screening
- Conclusion: critical CHD screening in the US could be reasonably cost-effective

Why Screen?

- 15 hospitals in Shanghai; 167,190 asymptomatic newborns screened
- Sensitivity 95.5% for critical CHD; 92.1% for major CHD
- Conclusion: pulse oximetry plus auscultation significantly improved the detection rate of major CHD, with a high sensitivity and reasonable false positive rate
Primary Targets for Screening: 7 lesions

- Hypoplastic left heart syndrome
- Pulmonary atresia
- Tetralogy of Fallot
- Total anomalous pulmonary venous return
- Transposition of the great arteries
- Tricuspid atresia
- Truncus arteriosus
CHD Classification

- Left to right shunt lesions (ASD, VSD, PDA, AVSD)
- Valvular regurgitant lesions (MR, TR, AI, PI)
- Obstructive lesions (AS, PS, COA, AV valve stenosis)
- Cyanotic CHD (right to left shunts, “mixing” lesions)
  - HLHS, PA, TOF, TAPVR, TGA, TA, Truncus

Obstructive Lesions

- Obstruction to ventricular output
- Three pathophysiologic changes:
  - Systolic ejection murmur (by auscultation)
  - Ventricular hypertrophy (by ECG)
  - Post-stenotic dilatation (by CXR)
Obstructive Lesions

- Left ventricular obstructive lesion: ductal dependent for systemic blood flow (i.e. critical aortic stenosis)

- Right ventricular obstructive lesion: ductal dependent for pulmonary blood flow (i.e. critical pulmonary stenosis)
Cyanotic CHD: 7 Primary Target Lesions

- Normal Heart

Hypoplastic Left Heart Syndrome

- LV hypoplasia and atresia/critical stenosis of the aortic/mitral valves
- PDA closure → marked decrease in systemic cardiac output → shock and metabolic acidosis
- Tachycardia, dyspnea, weak peripheral pulses
- Heart murmur usually absent
Pulmonary Atresia (Intact Ventricular Septum)

- Interatrial communication/PDA necessary for survival
- Severe cyanosis, tachypnea
- Heart murmur usually absent (soft TR or PDA murmur may be present)
- ECG: LVH usually present (in contrast to normal neonatal RVH)
- CXR: ↓ vascular markings/dark lung fields

Tetralogy of Fallot

- Four components: large malalignment type VSD, overriding aorta, RVOT obstruction/pulmonary stenosis, right ventricular hypertrophy
- Variable presentation depends on severity of RVOT obstruction/PS → atresia
  - Normal O2 levels with L→R shunt to severe cyanosis
  - Systolic murmur present at birth
  - CXR: concave MPA with upturned apex ("boot-shaped"), ↓ vascular markings
  - Association with DiGeorge Syndrome
Total Anomalous Pulmonary Venous Return

- No direct communication of pulmonary veins to LA
- 4 types: supracardiac, cardiac, infracardiac, mixed type
- Many have pulmonary HTN
- Obstructed versus unobstructed
  - Marked cyanosis/respiratory distress to mild cyanosis
- CXR: ↑ pulmonary vascular markings, “snowman” sign
Transposition of the Great Arteries

- Aorta from RV → desaturated blood to body, and PA from LV → oxygenated blood to lungs
- Mixing defects (ASD, PDA, VSD) are necessary for survival
- Cyanosis from birth
- Large, male newborns
- No heart murmur if no VSD
- CXR: ↑ vascular markings, “egg on a string”

Tricuspid Atresia

- Absent tricuspid valve, hypoplastic RV
- Classified according to the presence/absence of PS/great artery relationship, associated COA/IAA
- Severe cyanosis from birth
- ECG: “superior” QRS axis, LVH
- CXR: vascular markings usually decreased (depend on PS/TGA)
Truncus Arteriosus

- Single common arterial trunk exits from the heart: coronary, pulmonary, systemic arteries
- Clinical manifestations depend on PBF: initially mildly cyanotic, then tachypnea, tachycardia, excessive sweating, poor feeding (pulmonary overcirculation) as PVR decreases
- CXR: ↑ pulmonary vascular markings
- Association with DiGeorge syndrome

Case Example

- Well appearing term male infant, failed CCHD screen
- Initial vital signs: O2 saturations in the upper 70s to low 80s, HR 160s, RR low 70s, BP 60/30, MAP 40
- Physical exam: mildly tachypneic, soft systolic murmur, pulses equal, CR <3 sec but feet cool
Case Example Chest Xray

- Increased pulmonary vascular markings
- Narrow superior mediastinum
- “Egg on a string”

Case Example

- Labs
  - CBC WBC 15.7, Hbg 14.7, Hct 42, Platelets 256
  - Blood cultures obtained
  - VBG 7.43/41/23/22/-4
- Saturations decreased to 60s, infant on CPAP 30%, FiO2 increased to 100% with sats into low 70s
- Prostaglandins initiated at 0.05 μg/kg/min, baby intubated for transport
Case Example Echocardiogram

D-transposition of the great arteries

Initial Stabilization: Evaluation

- Complete physical exam:
  - Vital signs: hemodynamic instability?
  - General: distress? any dysmorphic features?
  - Resp: tachypnea/increased work of breathing?
  - CV: murmur? Pulses and perfusion?
  - Abdomen: hepatomegaly?
Initial Stabilization: Evaluation

- Chest xray: evaluate pulmonary vascular markings, heart size, shape, non-cardiac causes of cyanosis
- 12 lead electrocardiogram: exclude associated cardiac arrhythmia
- Labs: blood gas and lactate (if able), CBC (sepsis evaluation), BMP, LFTs
- Echocardiogram (within the hospital or transport)

Initial Stabilization: Evaluation

- Hyperoxia test:
  - Test the response of arterial PO2 to 100% oxygen
  - Differentiate cyanosis caused by cardiac disease from that caused by pulmonary disease
  - Administer 100% FiO2 for 10 minutes
  - Repeat the arterial PO2
    - >100 mmHg → pulmonary disease, <100 mmHg → intracardiac right to left shunt
Initial Stabilization: Prostaglandins

- Prostaglandin E1 IV infusion should be started as soon as diagnosis of CCHD is suspected or established
- Starting dose 0.05 to 0.1 μg/kg/minute
- When increased PaO2/saturation, increased systemic BP, improved pH, dose can be weaned to 0.02 μ/kg/min

Initial Stabilization: Prostaglandins

- Side effects:
  - Apnea: consider intubation
  - Fever, flushing
  - Tachycardia/bradycardia, hypotension: consider vasoactive inotropic support
- If arterial O2 saturation cannot be raised (i.e. inadequate mixing in D-TGA), cardiology consultation
Case Example

- Diagnosis: d-transposition of the great arteries
- Infant continued to have low sats/PaO2s despite PGE (inadequate mixing at the atrial level)
- Taken to the cath lab for balloon atrial septostomy
- Ultimately underwent arterial switch operation

Conclusion

- AAP recommends that all newborns be screened for CCHD
- Screen at approximately 24 hours of age
- Any signs or symptoms of CHD should prompt rapid evaluation → possible urgent transfer
- Seven primary target lesions for screening (remember additional critical obstructive lesions)
References


