Screening and Initial Stabilization of Critical Congenital Heart Disease

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Disclosures

- I have no relevant financial relationships with the manufacturers(s) of any commercial products(s) and/or provider of commercial services discussed in this CME activity
- I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation

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

SPECIAL ARTICLES

Strategies for Implementing Screening for Critical Congenital Heart Disease

AUTHORS: Alex R. Kemper, MD, MPH, MS,[a] William T. Mahle, MD,[b] Gerard R. Martin, MD,[c] W. Carl Cooley, MD,[d] Praveen Kumar, MBBS, DCH, MD,[e] W. Robert Morrow, MD[f] Kellie Kelm, PhD,[g] Gail D. Pearson, MD, ScD,[h] Jill Glidewell, RN, MSN, MPH,[i] Scott D. Grosse, PhD,[j] and R. Rodney Howell, MD[k]

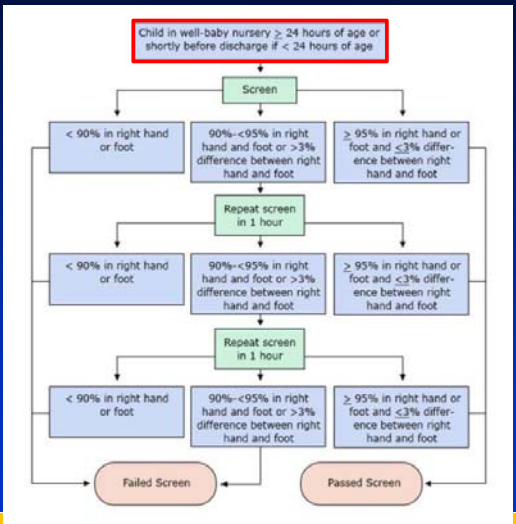
abstract

FREE

BACKGROUND: Although newborn screening for critical congenital heart disease (CCHD) was recommended by the US Health and Human Services Secretary's Advisory Committee on Heritable Disorders in

- 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***

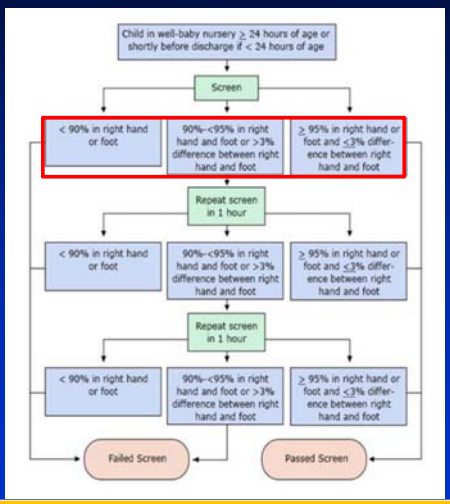
CCHD Screening Algorithm



https://www.cdc.gov/ncbddd/heartdefects/hcp.html
Kemper AR et. al. Pediatrics 2011

CCHD Screening Algorithm

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



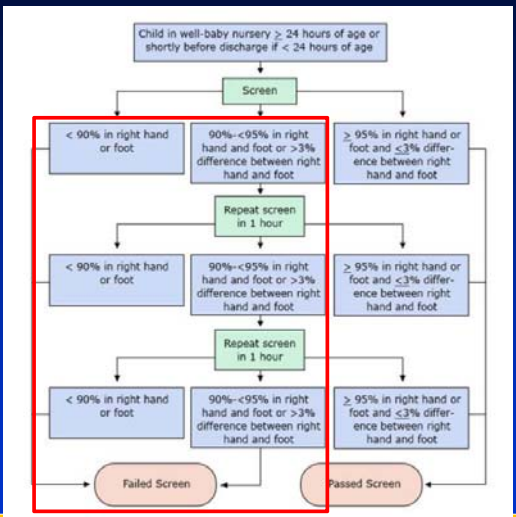
https://www.cdc.gov/ncbddd/heartdefects/hcp.html
Kemper AR et. al. Pediatrics 2011

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

Failed CCHD Screen



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 → 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

Why Screen?

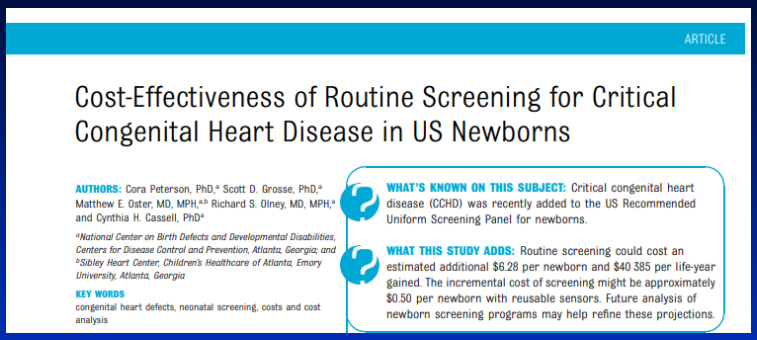

Articles

Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies: a systematic review and meta-analysis

Shakila Thangaratinam, Kiritrea Brown, Javier Zamora, Khalid S Khan, Andrew K Ewer

- 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?



ARTICLE

Cost-Effectiveness of Routine Screening for Critical Congenital Heart Disease in US Newborns

AUTHORS: Cora Peterson, PhD,[a] Scott D. Grosse, PhD,[a] Matthew E. Oster, MD, MPH,[a,b] Richard S. Olney, MD, MPH,[a] and Cynthia H. Cassell, PhD[a]

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KEY WORDS
congenital heart defects, neonatal screening, costs and cost analysis

WHAT'S KNOWN ON THIS SUBJECT: Critical congenital heart disease (CCHD) was recently added to the US Recommended Uniform Screening Panel for newborns.

WHAT THIS STUDY ADDS: Routine screening could cost an estimated additional $6.28 per newborn and $40 385 per life-year gained. The incremental cost of screening might be approximately $0.50 per newborn with reusable sensors. Future analysis of newborn screening programs may help refine these projections.

- **$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?

Pulse Oximetry and Auscultation for Congenital Heart Disease Detection

Xiao-jing Hu, PhD,[a] Xiao-jing Ma, MD,[a,b] Qu-ming Zhao, MD,[a] Wei-li Yan, PhD,[a,b] Xiao-ling Ge, MD,[a] Bing Jia, MD,[a] Fang Liu, MD,[a] Lin Wu, MD,[a] Ming Ye, MD,[a] Xue-cun Liang, MD,[a] Jing Zhang, MD,[a] Yan Gao, MD,[a] Xiao-wen Zhai, MD,[a] Guo-ying Huang, MD[a,b]

- **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**

CCHD Screening by State
Kansas and Missouri

What is CCHD Screening?

Critical Congenital Heart Defects (CCHD) Screening can detect heart defects in newborns that often have no other symptoms. It is a simple bedside test to determine the amount of oxygen in a baby's blood. Low levels of oxygen in the blood can be a sign of a congenital heart defect. CCHD screening is a part of the national Recommended Uniform Screening Panel (RUSP).

What is KDHE doing about it?

All Birthing Facilities and Midwives in Kansas are encouraged to screen and report CCHD results via the electronic birth certificate (EBC).

Kansas Department of Health and Environment (KDHE) launched a quality initiative and program to increase awareness of Critical Congenital Heart Defects (CCHD) and ensure that all babies in Kansas are being screened for CCHD after birth. The program's goals also include assurance of prompt care, connection to resources, long-term follow-up, and improvement of overall health outcomes for infants with CCHD.

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)

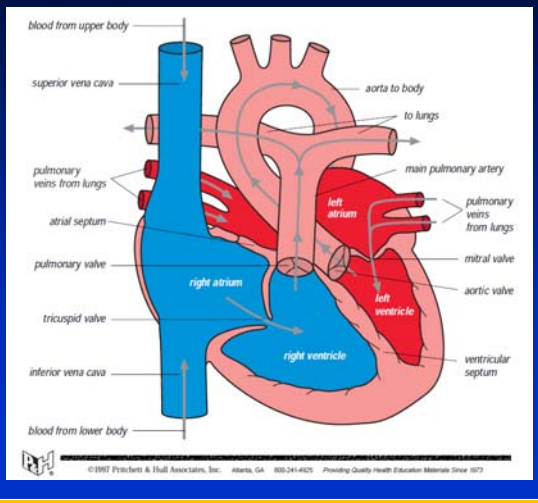
Obstructive Lesions

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

Obstructive Lesions

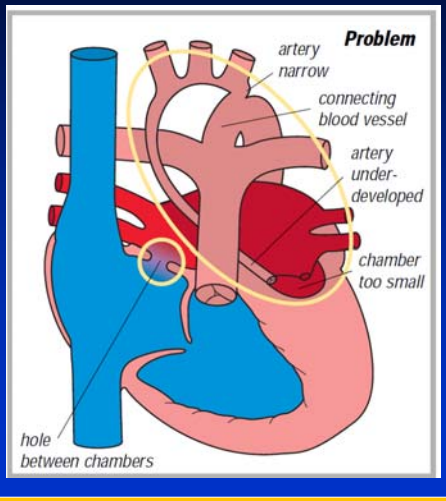
- Right ventricular obstructive lesion: ductal dependent for pulmonary blood flow (i.e. critical pulmonary stenosis)

Cyanotic CHD: 7 Primary Target Lesions



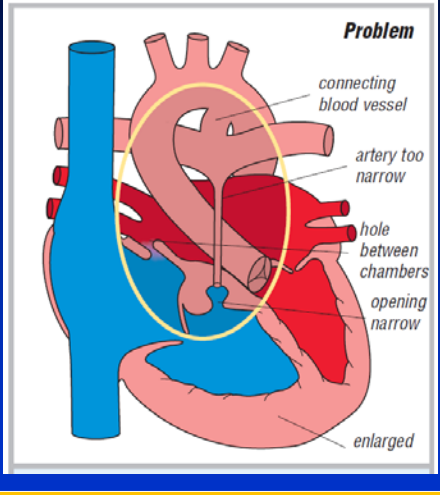
- Normal Heart

Hypoplastic Left Heart Syndrome



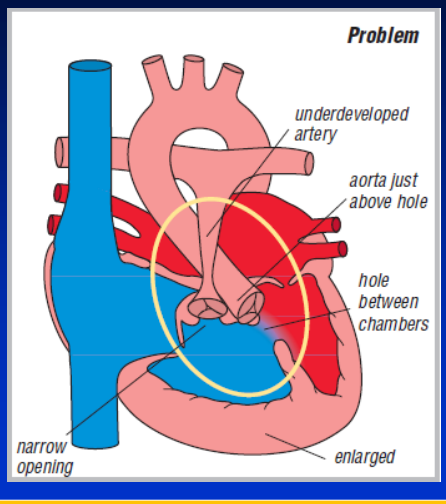
- LV hypoplasia and atresia/critical stenosis of the aortic/mitral valves
- PDA closure → marked decrease in systemic cardiac output → shock and metabolic acidosis
- Tachycardia, dyspnea, weak peripheral pulses
- Heart murmur usually absent

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

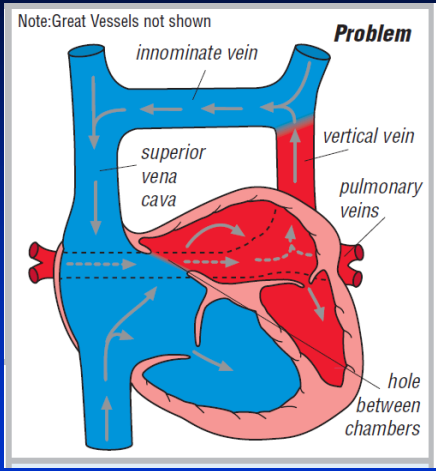
Tetralogy of Fallot



- 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

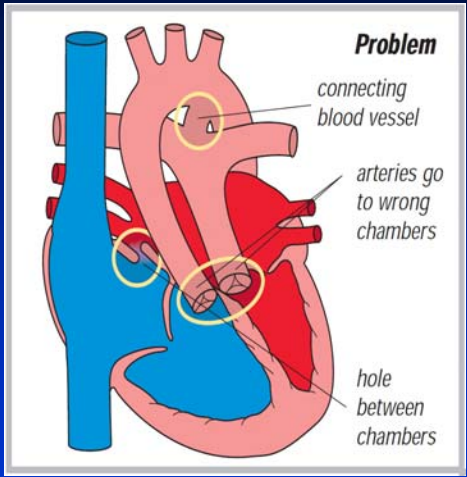
Tetralogy of Fallot

Total Anomalous Pulmonary Venous Return



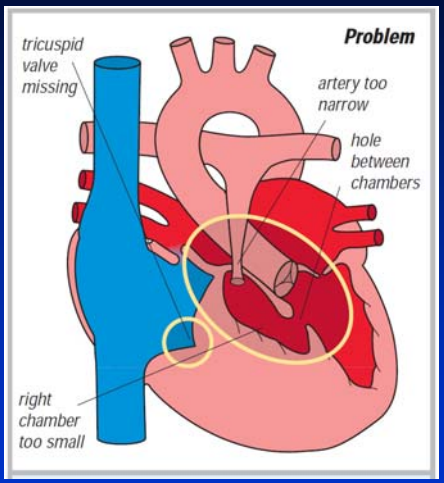
- No direct communication of pulmonary veins to LA
- 4 types: supracardiac, cardiac, infracardiac, mixed type
- Many have pulmonary HTN
- Obstructed versus unobstructed
  - Marked cyanosis/respiratory distress to mild cyanosis
- CXR: ↑ pulmonary vascular markings, "snowman" sign

Transposition of the Great Arteries



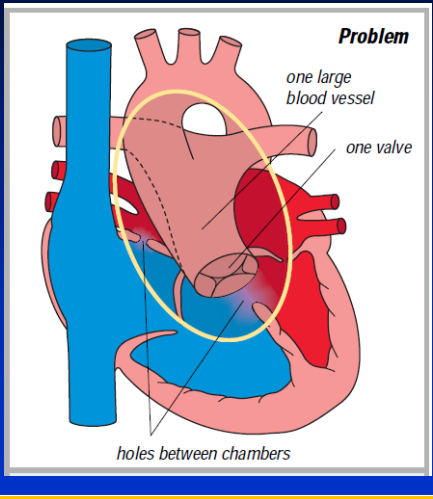
- Aorta from RV → desaturated blood to body, and PA from LV → oxygenated blood to lungs
- Mixing defects (ASD, PDA, VSD) are necessary for survival
- Cyanosis from birth
- Large, male newborns
- No heart murmur if no VSD
- CXR: ↑ vascular markings, "egg on a string"

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

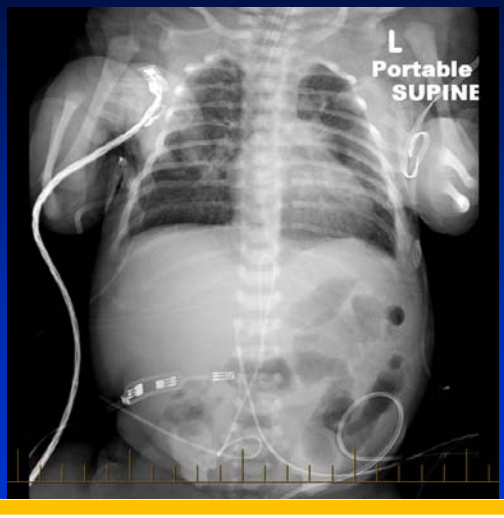


- Single common arterial trunk exits from the heart: coronary, pulmonary, systemic arteries
- Clinical manifestations depend on PBF: initially mildly cyanotic, then tachypnea, tachycardia, excessive sweating, poor feeding (pulmonary overcirculation) as PVR decreases
- CXR: ↑ pulmonary vascular markings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

1. Kemper AR, Mahle WT, Martin GR, Cooley WC, Kumar P, Morrow WR, Kelm K, Pearson GD, Glidewell J, Grosse SD, Lloyd-Puryear M, Howell RR. Strategies for implementing screening for critical congenital heart disease. *Pediatrics.* 2011; 128:e1-8.
2. Knapp AA, Metterville DR, Kemper AR, Prosser L, Perrin JM. Evidence review: Critical congenital cyanotic heart disease, Final Draft, September 3, 2010. Prepared for the Maternal and Child Health Bureau, Health Resources and Services Administration.
3. Mahle WT, Newburger JW, Matherne GP, Smith FC, Hoke TR, Koppel R, Gidding SS, Beekman RH, 3rd, Grosse, SD. Role of pulse oximetry in examining newborns for congenital heart disease: A scientific statement from the AHA and AAP. *Pediatrics.*2009;124:823-36.
4. Park, Myung K., Park's Pediatric Cardiology for Practitioners. Mosby, 2014.
5. Peterson C, Grosse SD, Oster ME, Olney RS, Cassell CH. Cost-effectiveness of routine screening for critical congenital heart disease in US newborns. *Pediatrics*. 2013; 132:e595-e603.
6. Thangaratinam S, Brown K, Zamora J, Khan K, Ewer AK. Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies: a systematic review and meta-analysis. *Lancet*. 2012; 379: 2459-64.
7. Hu X, Ma X, Zhao Q, Yan W, Ge X, Jia B, et. al. Pulse oximetry and auscultation for congenital heart disease detection. *Pediatrics*. 2017; 140 (4): e20171154.
8. https://www.cdc.gov/ncbddd/heartdefects/hcp.html