CYANOTIC CONGENITAL HEART DISEASE: TEST YOUR KNOWLEDGE

Mohamed Sulaiman, MD
Pediatric cardiologist

Kidsheart: American Fetal & Children's Heart Center
Dubai Healthcare City, Dubai-UAE
Normal Cardiac Physiology

**PRESSURES**
- RA: 3-7 mmHg
- RV: 25/0 mmHg
- PA: 25/10 mmHg
- LA: 5-10 mmHg
- LV: 100/10 mmHg
- AO: 100/70 mmHg

**SATS**
- RA: 70 +/- 5%
- RV: 70 +/- 5%
- PA: 70 +/- 5%
- LA: 97 +/- 3%
- LV: 97 +/- 3%
- AO: 97 +/- 3%
Mixing everything in real live
TACHYPNEIC POOR FEEDER: H&P (1)

- You are called to see a 24 hour-old infant who initially was doing well, but now is not feeding and is mildly tachypneic (but not in respiratory distress)

- On exam,
  
  His precordium is quiet, $S_2$ is normally split, and you heard no heart murmurs.
  
  His liver is mildly enlarged, abdomen soft.
  
  Femoral pulses are present.
TACHYPNEIC POOR FEEDER: PE & LABS

- Saturation by pulse oximeter is 90%:
  - with administration of 100% FiO₂ it increases to 99%.
- Blood pressures are equal in the arms and legs.

- Electrolytes, CBC are normal
- You suspect either sepsis or CHD.
TACHYPNEIC POOR FEEDER: PE & LABS

- Saturation by pulse oximeter is 90%:
- with administration of 100% FiO₂ it increases to 99%.
- Blood pressures are equal in the arms and legs.

- Electrolytes, CBC are normal
- You suspect either sepsis or CHD.
TACHYPINEIC POOR FEEDER: PE & LABS

What is the one test that would lead you to the correct diagnosis?

- A. CXR
- B. 12 Lead Electrocardiogram
- C. Simultaneous hand and foot pulse oximetry determinations
- D. Arterial blood gas on room air
  On 100% FiO₂
**TACHYPNEIC POOR FEEDER: PE & LABS**

What is the one test that would lead you to the correct diagnosis?

- A. CXR
- B. 12 Lead Electrocardiogram
- C. Simultaneous hand and foot pulse oximetry determinations
- D. Arterial blood gas on room air
  On 100% FiO₂
Think for a minute

- Cyanosis: NOT PROMINENT
- Equal blood pressures
- The findings of femoral pulses
TACHYPNEIC POOR FEEDER: MANAGEMENT

The most appropriate management of this infant is:

- A. STAT ECHO
- B. Start PGE, intubate, hyperventilate
- C. Start PGE, put in head hood with 40% FiO₂
- D. Start PGE, leave in room air
- E. Start PGE, leave in room air plus AB
**TACHYPNEIC POOR FEEDER: MANAGEMENT**

The most appropriate management of this infant is:

- **A.** STAT ECHO
- **B.** Start PGE, intubate, hyperventilate
- **C.** Start PGE, put in head hood with 40% FiO₂
- **D.** Start PGE, leave in room air
- **E.** Start PGE, leave in room air plus AB
** RR

HIGH

Aneurysm size

** DESAT

100

93

59

87

Preducatal

Postducatal
Why is that: CAVC with discrepancy in the sat
TACHYPNEIC POOR FEEDER - PULSE OXIMETRY
In this infant, which pulse oximetry findings are consistent with critical coarctation of the aorta?

A
99%
99%
97%
97%

B
80%
80%
95%
95%

C
95%
95%
94%
92%

D
85%
85%
90%
90%
Cyanosis
Which patient has HLHS out of triplet?

- A
- B
- C
Next Case:

- Next day: does not want to eat, saturation: 95%, breath fast, Lactate 7.
HYPOPLASTIC LEFT HEART SYNDROME

1% of all CHD, 9% of all CHD detected in nursery

Most common cause of death from CHD in neonatal period
HYPOPLASTIC LEFT HEART SYNDROME

Clinical Presentation

Initially look ok.
Dyspnea, cyanosis may not be obvious, usually tachypneic
Second heart sound- loud single
Murmur is usually absent
Tachycardia, hepatomegaly, gallop, pulmonary rales, weak pulses, cold extremities
HYPOPLASTIC LEFT HEART SYNDROME

Therapy
PGE1
RA!, no O2
Maintain NL pH-7.35, PCO2 40-45, Sats 75-85%
Need UAC, UVC
Avoid intubation and do not intubate for transfer
Next Case:
Cyanotic CHD:

- Is it cyanosis.
- Is pt comfortable.
- Does he need PGE
- Does he need oxygen
Big Blue BOY
24 hours old
Sat 92 %, mild tachypnea
X Ray
MORE CYANOTIC NEWBORNS: CXR 1

The ECG is “unremarkable for a newborn”

This CXR demonstrates:

A. Transposition of the great vessels
B. Tetralogy of Fallot
C. Total anomalous pulmonary venous return
D. Tricuspid atresia
E. Pulmonary atresia with intact septum
MORE CYANOTIC NEWBORNS: CXR 1

The ECG is “unremarkable for a newborn”

This CXR demonstrates:

A. Transposition of the great vessels
B. Tetralogy of Fallot
C. Total anomalous pulmonary venous return
D. Tricuspid atresia
E. Pulmonary atresia with intact septum
The best way of mixing is:

1. Large PDA so PGE will save the baby
2. Large VSD
3. Moderate size ASD
Cyanotic CHD: Lesions with increased pulmonary blood flow

- D-Transposition of the great arteries
- Truncus arteriosus
- Total anomalous pulmonary venous return
- DORV
- Single ventricle
Next Case
7 MONTH OLD WITH A MURMUR

- 7 months old.
- Mom stated he was blue when he wake up in the morning for few minutes.
- Murmur at 3 months old, seen by adult cardiologist (Small Premembranous VSD).
7 MONTH OLD WITH A MURMUR

- P/E and EKG: sat 85%, increase to 92 with 100% oxygen
  - RV impulse at the LLSB
  - A Systolic ejection click at the LUSB
  - A GR III/VI SEM LUSB.

- THIS ECG WAS OBTAINED.
You conclude:
1. The PE and EKG match the diagnosis
2. Send him to same cardiologist to avoid confusion
7 MONTH OLD WITH A MURMUR

- The next step for this patient is:
  - Send him home and follow him in 3 months.
  - Tell the mom the VSD will close by 1 year
  - Call your friend pediatric cardiologist and ask him to see the pt urgently
  - Admit the pt to the hosp and treat him with oxygen
7 MONTH OLD WITH A MURMUR

- The next step for this patient is
  - Send him home and follow him in 3 months.
  - Tell the mom the VSD will close by 1 year
  - Call your friend pediatric cardiologist and ask him to see the pt urgently
  - Admit the pt to the hosp and treat him with oxygen
Echo finding

Diastole
Interventricular septum
Left ventricle
Mitral valve
Right ventricle
VSD
Aorta
Aortic valve
Left atrium
ECG
Questions?
Which country is this?