Tetralogy of Fallot: Hypercyanotic spell

Learning objectives

- Adequate use of Oxygen, Sedatives, Volume, in the acute setting of a Tet spell.
- Recognize a refractory tet spell and tailor an appropriate treatment strategy.

Introduction

Brief description of simulation activities and physical assessment of mannequin

Brief summary

Sara Johnson, 3 months old, 6.5 Kg, 62 cm, female.

Prenatal history

FT AGA
Section for fetal decelerations and maternal HTN
No abnormalities in prenatal US

Past Medical record

Patient APGAR 8/8, d/c home after 4 days with mother, remained in regular nursery; mother recalls occasional tachypnea and difficulty feeding

Patient description

2 days ago h/o poor feeding, loose stools and mild fever. Parents are bringing her to Emergency Department of Children’s Hospital of Pittsburgh because she is very irritable.

Physical Exam/General things

Airway/Breathing

- Airway: patent
- Tachypnea
- Sat 85% in right arm
- No retractions, clear lungs to auscultation.

Circulation

- Sinus tachycardia
Clinical Simulation Course
Pediatric Critical Congenital Heart Disease (PedCritHeart)

- BP 75/45 mmHg
- Strong symmetric pulses, including femoral
- Capillary refill 2 sec.
- No hepato-splenomegaly

**Auscultation**
II/VI systolic murmur over left precordium. → Murmur disappears at TOF tet spell box

**General Appearance/information:**
- Irritable and crying.
- Mucous membranes and lips are slightly dry
- Temperature 38°C

**Cognitive skills**
- Standard precautions.
- Communication skills.
- Clinical prioritization skills.
- Resuscitation skills.
- Respiratory, cardiology anatomy and physiology.
- Diagnostic and Pharmacology management of hypercyanotic spell in an Infant with undiagnosed Tetralogy Fallot

**Psychomotor Skills**
- Vital signs
- Physical exam (Airway, Breathing, Circulation).
- Lung and heart auscultation.
- Interpretation of X-Ray, Echocardiography, ECG.
- >Appropriate O2 delivery.
- Medication administration and dosage of appropriate sedative agents and Phenylephrine.

**Learning objectives**
- Demonstrate adequate diagnostic and therapeutic skills given a simulated case of Hypercyanotic Spell in an Infant with undiagnosed Tetralogy Fallot.
- Identify hemodynamics of a simulated infant with Tet spell and tailor an appropriate treatment strategy.
- Recognize importance of incremental approach: Supplemental O2, IV Fluid bolus, morphine, Bicarbonate, Phenylephrine.
- Use of supplemental O2 to decrease PVR.
- Identify need of I.V. fluid bolus to improve RV filling and pulmonary flow.
- Recognize a persistent refractory spell, consider intubation with proper sedation and paralysis. Use of phenylephrine to increase SVR. ECMO and surgery as last source.
Event/conditional list
See roadmap

Setting
ED – Tertiary care

Patient information

<table>
<thead>
<tr>
<th>Age</th>
<th>3 months</th>
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<tbody>
<tr>
<td>Medications</td>
<td>No</td>
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<tr>
<td>Allergies</td>
<td>NKAD</td>
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<tr>
<td>PCP</td>
<td>NA</td>
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</tbody>
</table>

Monitor settings

(Participants should ask for it, first patient without any monitor setting)

SpO2
O2
Primary ECG
Pulse
CVP
ABP
Temperature
Laboratories

Admission

- Lactate 1 mg/dl
- Glucose 100 mg/dl
- Na 145 mmol/l
- K 4.9 mmol/l
- Bun 11 mg/dl
- Cr 0.8 mg/dl
- Hgb 15 g/dl
- Hct 45%
- WBC 13 /nl
- Plt ct 200/nl
- SGOT/AST 35 U/l
- SGPT/ALT 30 U/l

ABG

- pH 7.35
- pO2 50 mmHg
- pCO2 30 mmHg
- HCO3 21 mmol/l
- **BE** -5 mmol/l

**Tet spell**
- Lactate 2.5 mg/dl
- ABG
  - pH 7.26
  - pO2 45 mmHg
  - pCO2 40 mmHg
  - HCO3 20 mmol/l
  - BE -10 mmol/l

**No action 1**
- Lactate 4 mg/dl
- Glucose 180 mg/dl
- ABG
  - pH 7.2
  - pO2 40 mmHg
  - pCO2 38 mmHg
  - HCO3 17mmol/l
  - BE -15 mmol/l

**No action 2**
- Lactate 7 mg/dl
- Glucose 200 mg/dl
- ABG
  - pH 7.11
  - pO2 30 mmHg
  - pCO2 50 mmHg
  - HCO3 15mmol/l
  - BE -15 mmol/l

**Arrest**
- Lactate 12 mg/dl
- Glucose 200 mg/dl
- ABG
  - pH 7.03
  - pO2 20 mmHg
  - pCO2 55 mmHg
  - HCO3 11 mmol/l
  - BE -17 mmol/l

**Labs Recovery**
- Lactate 2 mg/dl
- ABG
• pH 7.31
• pO2 50 mmHg
• pCO2 30 mmHg
• HCO3 21 mmol/l
• BE -8 mmol/l