The Ill-Appearing Infant: Emergencies in the Early Months of Life

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Department of Emergency Medicine
The Reading Hospital
2 May 2017
Disclosures

• Nothing to disclose
One Hour Into Your Shift...
Goal and Objectives

Goal:
• Develop a thoughtful approach to neonatal emergencies

Objectives:
• Discuss key components of the history and physical exam to consider when an ill-appearing neonate presents to care
• Describe the differential diagnosis for an ill-appearing neonate
• Discuss the initial management of a neonate in shock
Why Do Neonates Scare Us?
Why Do Neonates Scare Us?

• Neonates often present with non-specific signs and symptoms
• Guardians may not appreciate the severity of presentation
• Rapid deterioration
• Most in shock have sepsis, *but we must consider other possibilities*...
  • ....*And there are many other possibilities!*
Case Presentation: Triage History

• 7 day-old male brought in by parents for poor feeding
• Mom was initially breast-feeding → switched to formula 2 days ago
• Now taking < 1 ounce at a time
• No fevers
• No vomiting
Case Presentation: Triage History

• 7 day-old male brought in by parents for poor feeding
• Mom was initially breast-feeding → switched to formula 2 days ago
• Taking < 1 ounce at a time
• No fevers
• No vomiting

• Triage nurse notes rapid breathing and calls the ED physician in
Case Presentation: History

Prenatal and Birth History:
- G1P1 healthy mother; normal prenatal care
- Uncomplicated SVD; ROM <4hr; birth weight 7lb 4oz; 2 days in nursery

Interim History:
- Saw PCP on DOL 4; weight 7lb 1oz
- Transitioned from breast to formula 2 day ago

History of Present Illness:
- Decreased intake
- Brief period a rapid breathing after last feed, no color changes or apnea
- No fever, cough, vomiting, or rash
Case Presentation: Physical Examination

Vital signs: T 98.4F; HR 182; RR 68; BP 68/36; SpO2 92%; Weight 6lb 11oz
General: moderate respiratory distress with inter-/sub-costal retractions and nasal flaring
HEENT: fontanel sunken, eyes open, mucous membranes tacky, no cyanosis
CV: tachycardia, regular rhythm, no murmur or gallop, weak femoral pulses, CR 3 sec
Resp: increased work of breathing, equal and clear breath sounds
Abdomen: soft and non-distended, liver down 2 cm
Skin: no rash, cool extremities
Case Presentation: Problem List

- Respiratory distress
- Shock
Case Presentation:
Problem List

Respiratory distress

Shock

Temperature retaken → 96.4F rectally
Case Presentation:
Problem List

Respiratory distress

Shock

Temperature retaken → 96.4°F rectally = Hypothermia
Case Presentation: Differential Diagnosis

Infection – sepsis; viral
Cardiac disease
Trauma (non-accidental)
Metabolic and electrolyte disorders
GI disorders
Endocrine disorders
Neurologic disorders
Feeding failure/problems

T – Trauma
H – Heart disease; Hypovolemia
E – Endocrine
M – Metabolic
I – Inborn errors of metabolism
S – Sepsis
F – Formula dilution/overconcentration
I – Intestinal catastrophes
T – Toxins
S – Seizures

Sharieff GQ, McCollough M, Emerg Med Clin North Am, 2002
Causes of the Ill-Appearing Neonate: “Neo-secrets”

N – i”N”-born errors of metabolism
E – Electrolyte abnormalities
O – Overdose
S – Seizure
E – Enteric emergencies
C – Cardiac
R – Renal, Recipe errors
E – Endocrine
T – Trauma
S – Sepsis

History: Key Considerations

• Prenatal care
• Birth history
• Feeds
  • *If formula, how is it mixed?*
• Wet diapers, bowel movements
• Activity level
History: Key Considerations

• Fever OR hypothermia
  • *Lack of fever does not exclude infection*

• Associated symptoms

• Newborn screen
  • *May not be available*

• Family history

• Social history
  • *All caregivers, exposures*
Physical Examination: Key Considerations

- Vital signs
  - 4-extremity BP’s
  - Pre-/post-ductal SpO2

- Respiratory
  - Increased WOB often non-specific
  - Usually no crackles

- Cardiovascular
  - May not have a murmur in CHD
  - Gallop, pulses, perfusion; liver edge
Causes of Neonatal Emergencies: SEPSIS, SEPSIS, SEPSIS

By far the most common, and you should (almost) ALWAYS treat an ill-appearing neonate for potential sepsis

BUT - keep an open mind and remember the mnemonics
Neonatal Emergencies:
Neonatal Emergencies: Initial Management of Shock

Airway
  • BVM ventilation
  • Intubation
    • Use caution with sedatives
    • 3.0 – 3.5 mm ETT secured 10 – 10.5 cm at lip

Breathing
  • Attempt synchronous breaths, about 20 per minute

Circulation
  • IV and/or IO access
  • 0.9% NaCl 20cc/kg IV push, reassess after each one
  • Anticipate pressors
Neonatal Emergencies: IO Lines

- Tibial plateau is preferred
- Distal femur is an option
- Can give anything through an IO you would give through an IV
- If full arrest \(\rightarrow\) place IO as IV access is attempted
Neonatal Emergencies: Initial Management of Shock

**Labs:**
- Check blood glucose early!
- If possible: CBC, blood culture, A(V)BG, BMP, LFTs, urine (u/a, culture, gram stain)
- Often too ill for LP initially

**Ancillary tests:**
- ECG, CXR, lactate, ammonia, abdominal imaging

**Medications:**
- Ampicillin 200 – 300 mg/kg/day
- Cefotaxime 100 – 200 mg/kg/day
- Acyclovir 60 mg/kg/day (if critically ill or < 3 weeks)
- **Pressors:** Dopamine, epinephrine
Causes of Neonatal Emergencies: Cardiac Disease

- Significant variation in rates of prenatal diagnosis of CHD in all states
- Overall prenatal detection occurred in 34%

<table>
<thead>
<tr>
<th>TABLE 2</th>
<th>PDRs by Fundamental Diagnosis</th>
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<td>Fundamental Diagnosis</td>
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<td>Pulmonary stenosis and pulmonary atresia with IVS</td>
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<td>Hypoplastic left heart syndrome</td>
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</table>

Quartermain M, Pasquali S, Goldberg D, Pediatrics, 2015
Causes of Neonatal Emergencies: Cardiac Disease

Cyanotic and Obstructive Lesions
• Usually present within first week
• Precipitous deterioration as PDA closes

Coronary artery abnormalities

Arrhythmias
• SVT
Causes of Neonatal Emergencies: Cardiac Disease

Cyanotic Lesions
- Tricuspid atresia
- Pulmonic atresia
- Transposition of the great arteries

Obstructive Lesions
- Hypoplastic left heart
- Critical aortic stenosis
- Critical coarctation of the aorta or interrupted aortic arch

*Most other congenital heart disease patients present at several months of age in heart failure*
Causes of Neonatal Emergencies: Cardiac Disease

Presentation:
- Slow feeding, *tachypnea* and *tachycardia*, murmur, gallop; *hepatomegaly*, cyanosis, shock

Workup:
- ECG
- CXR
- 4-extremity blood pressures
- Labs: ABG, CBC, electrolytes
- ECHO
Causes of Neonatal Emergencies: Cardiac Disease

Management:

• Supplemental oxygen

• (Small) fluid boluses → reassess after each one

• Prostaglandin E<sub>1</sub> may be life-saving

• Early discussion with pediatric cardiologist
Causes of Neonatal Emergencies: Trauma

Child abuse is common
  • 119,000 victims annually
  • 600 deaths/year
  • $124 billion in total costs/year

It is often missed
  • 30% of abusive head trauma
  • 20% of abusive fractures

Causes of Neonatal Emergencies: Trauma

• Most common chief complaint is respiratory distress or apnea
• May not see external signs of trauma
• Testing ranges from 40% - 90% with high-risk injuries

Skeletal Survey Use in Infants <1 y.o. with Femur Fractures
Skeletal Survey Use in Infants <1 y.o. with Femur Fractures at Pediatric Hospitals

### Occult Fracture Screening (%)

<table>
<thead>
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<th>Percent</th>
<th>95% CI</th>
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<td>40 Hospitals</td>
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Causes of Neonatal Emergencies: Trauma

• Have a high index of suspicion
• May see a bulging fontanel or increased head circumference
• Retinal hemorrhages are pathognomonic, but not always present
• If hemodynamically unstable → look for other sources of bleeding
Causes of Neonatal Emergencies: Electrolyte Derangements

Hypoglycemia
- Several factors make neonates at high risk
- Consider metabolic, endocrine, toxins, and infections

Hyponatremia
- Water intoxication
- Aspirin toxicity
- CAH
  - May be the initial presentation of cystic fibrosis

Hypernatremia
### TABLE 1: Status of Newborn Screening in the United States

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<th>Endocrine</th>
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### TABLE 2: Core Conditions Detected by MS/MS

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<thead>
<tr>
<th>State</th>
<th>Fatty Acid Disorders</th>
<th>Organic Acid Disorders</th>
<th>Amino Acid Disorders</th>
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Kay C, Committee on Genetics, AAP, 2006
Newborn Screening Mandated Screening Panel and Mandated Follow-up Panel

### Conditions Mandated for Screening and Follow-up by NSFP

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<th>PKU</th>
<th>Phenylketonuria</th>
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<td>Maple Syrup Urine Disease</td>
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<td>CAH</td>
<td>Congenital Adrenal Hyperplasia</td>
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<td>GAL T</td>
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<td>GAA</td>
<td>Glycogen Storage Disease Type II (Pompe Disease)</td>
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<td>MPS I</td>
<td>Mucopolysaccharidosis type I (Hurler Syndrome) – <strong>Effective 2/1/17</strong></td>
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<td>X-ALD</td>
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### Acylcarnitine Disorders

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### Fatty Oxidation Disorders

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### Amino Acid Disorders

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<tr>
<td>TYR I</td>
<td>Tyrosinemia Type I</td>
</tr>
</tbody>
</table>

### Hemoglobinopathies

| Hb SC-Disease | Sickle-C Disease |
| Hb S | S-Beta Thalassemia |

### Lysosomal Storage Disorders

<table>
<thead>
<tr>
<th>GLA</th>
<th>Fabry Deficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASM</td>
<td>Niemann-Pick A/B</td>
</tr>
<tr>
<td>GBA</td>
<td>Gaucher Deficiency</td>
</tr>
<tr>
<td>GALC</td>
<td>Globoid cell leukodystrophy (Krabbe Disease)</td>
</tr>
</tbody>
</table>

### Others

<table>
<thead>
<tr>
<th>BIO</th>
<th>Biotinidase Deficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td>CF</td>
<td>Cystic Fibrosis</td>
</tr>
<tr>
<td>SCID</td>
<td>Severe Combined Immunodeficiency</td>
</tr>
</tbody>
</table>

### Point of Care Testing

<table>
<thead>
<tr>
<th>CCHD</th>
<th>Critical Congenital Heart Defects</th>
</tr>
</thead>
<tbody>
<tr>
<td>HEAR</td>
<td>Newborn Hearing Screening</td>
</tr>
</tbody>
</table>

Kay C, Committee on Genetics, AAP, 2006
Causes of Neonatal Emergencies: In-born Errors of Metabolism

<table>
<thead>
<tr>
<th>Subtle findings:</th>
<th>Overt findings:</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Abnormal tone</td>
<td>• <strong>Acidosis</strong></td>
</tr>
<tr>
<td>• Irritability</td>
<td>• Temperature instability</td>
</tr>
<tr>
<td>• Poor feeding</td>
<td>• Dehydration</td>
</tr>
<tr>
<td>• FTT</td>
<td>• Shock</td>
</tr>
<tr>
<td>• Vomiting</td>
<td>• Persistent <strong>hypoglycemia</strong></td>
</tr>
<tr>
<td>• Unusual odor to urine</td>
<td>• Seizures</td>
</tr>
<tr>
<td></td>
<td>• <strong>Hepatomegaly</strong>, jaundice</td>
</tr>
<tr>
<td></td>
<td>• May see gram-negative infections in Galactosemia</td>
</tr>
</tbody>
</table>
Causes of Neonatal Emergencies: In-born Errors of Metabolism

Workup:
- Blood glucose
- Electrolytes
- ABG
- Ammonia
- Lactate
- Urine (ketones, reducing substances)
- Specific testing can be sent, but rarely helpful in the ED
  - Urine organic acids
  - Plasma amino acids

Management:
- Fluid resuscitation
- Often, large amounts of glucose needed to turn off catabolism (D10)
- Early metabolism specialist or pediatric referral center
Bonus Case

• 5 week-old with 2-3 days of vomiting and diarrhea
• No fever
• Had previously been well
• T37.4  P 176  RR 42  BP 75/43  SpO₂ 87%
• Ill-appearing and dusky, sunken anterior fontanel
• Dry mucous membranes
• 1/6 systolic murmur upper sternal border radiating to back
• Soft, non-tender abdomen, liver edge at costal margin
Case

• Differential:
  • Shock (sepsis, cardiogenic, hypovolemic)
  • Pneumonia, bronchiolitis
  • Congenital heart disease (R → L shunting)
  • Methemoglobinemia
What is the most important next intervention?
What is the most important next intervention?

- Administration of 100% FiO₂ via non-rebreather leads to no change in pulse ox reading
  - This makes pulmonary etiology less likely
Next steps?

• 4 extremity BPs, 4 extremity $O_2$ Sat, EKG, CXR, labs
Next steps?

• EKG sinus tachycardia
• Labs normal
• Fluids are administered
• $O_2$ still 88%
What is the diagnosis?

• EKG sinus tachycardia
• Labs normal
• Fluids are administered
• $O_2$ still 88%
What is the diagnosis?  Methemoglobinemia

• Ferrous iron (Fe$^{2+}$) oxidized to ferric (Fe$^{3+}$) state which cannot bind oxygen

• Causes: infections/diarrhea/acidosis, well-water contaminated with nitrates, medications (benzocaine, lidocaine, sulfonamides)
  • Particular susceptibility due to lower cytochrome B$_3$ reductase, ease of oxidation of HbF and increased nitrate → nitrite converting intestinal flora

• Diagnosis: MetHb level by co-oximetry
  • Chocolate brown blood may be a clue

• Treatment: Methylene blue 1mg/kg when levels > 30%
Causes of Neonatal Emergencies: Gastrointestinal Disease

Malrotation with midgut volvulus
- 90% cases in first 3 months
- Consider in any neonate with emesis that does not look like digested milk
- Often do not have significant abdominal distention

Duodenal atresia
- Also with bilious emesis
- Look for “double bubble” on x-ray

Necrotizing enterocolitis
- Suspect in premies but 10% cases occur in full term infants
- Irritability or lethargy, +/- fever, abdominal distention, vomiting or diarrhea
- Look for pneumatosis intestinalis on x-ray
Causes of Neonatal Emergencies: Malro with volvulus

Normal anatomy

Whirlpool sign on US

Abnormal UGI
Causes of Neonatal Emergencies:
Duodenal Atresia
Causes of Neonatal Emergencies: Necrotizing Enterocolitis
Causes of Neonatal Emergencies: Necrotizing Enterocolitis
Causes of Neonatal Emergencies: Endocrine – Congenital Adrenal Hyperplasia

• >95% from 21-hydroxylase deficiency
• Classic: 1 per 16,000
• Look for clitoromegaly in females
• Males may be missed

Adrenal crisis
• Vomiting; diarrhea; lethargy; shock
• ↑ Na⁺ and ↓ K⁺, hypoglycemia
Causes of Neonatal Emergencies: Endocrine – Congenital Adrenal Hyperplasia

• Managing the adrenal crisis is key
• Avoid hypotonic solutions, including dextrose replacement
• May see arrhythmias with hyperkalemia
• Hyperkalemia typically improves rapidly with steroids
• Hydrocortisone stress dose: 100mg/m² (25mg)
Causes of Neonatal Emergencies: Neurologic Disorders – Seizure

• In first days of life → commonly due to hypoxic-ischemic events
• Often subtle
• Check d-stick early

Other considerations:
• Hypocalcemia, sodium derangements, in-born errors, congenital brain anomalies (CMV, toxo), infection, drug withdrawal, pyridoxine deficiency
  • Give pyridoxine if seizures refractory to phenobarbital and benzos
Bonus Case

• 4 month-old with increased sleep over the past three days
• No fever, vomiting or diarrhea
• Last bowel movement was 3 days ago
• T37.5   P 136   RR 42   BP 86/52   SpO₂ 97%
• Slightly pale, decreased tone
• Normal heart and lung exam, normal perfusion
• Weak suck and gag
Causes of Neonatal Emergencies: Neurologic Disorders - Infant Botulism

- Clostridium botulinum
- Approximately 110 cases/year (72% infant botulism)
- Most from ingestion of environmental dust and soil containing spores
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Causes of Neonatal Emergencies: Neurologic Disorders - Infant Botulism

Presentation:
- Can look ill, but generally well perfused and normal hemodynamics
- Afebrile
- **Constipation, feeding difficulties, poor suck, weak cry**, descending or global hypotonia, drooling, respiratory failure

Management:
- Botulinum toxin recovered from stool
- Call California Department of Health Services
  - Human antitoxin: botulism immune globulin
- May need intubation and critical care services
- Avoid aminoglycosides
Back to Our Patient: Summary

- 7 day-old male with hypothermia, respiratory distress, and shock
- Decreased femoral pulses, hepatomegaly
- Patient intubated with a 3.5 mm ETT taped at 10.5 cm at the lip
- Unable to obtain IV → IO line placed in right tibia
- 20 cc/kg 0.9% NaCl pushed through line
- Blood glucose 120
- Cefotaxime and Ampicillin given
Back to Our Original Patient: Further Interventions

- CXR obtained
  - ETT in proper position
  - Normal cardiac silhouette
- Labs
  - Severe metabolic acidosis; WBC 14k, Na 137, K 4.8
- Suspect obstructive cardiac disease
- PGE$_1$ started
- Cardiology called for bedside ECHO
  - Severe coarctation of the aorta
- Transferred to ICU
Summary

• Think broadly for reasons why a neonate may appear ill (THE MISFITS; NEO SECRETS)

• Keep in mind key components of the history and physical examination to help differentiate etiologies of septic-appearing newborns

• Address signs of shock early as neonates often decompensate quickly


Thank you!

Questions?