Interesting Cases from the Road

Patricia A. Scott, DNP, APN, NNP-BC, C-NPT
Advanced Practice Coordinator
Pediatrix Medical Group of Tennessee, Nashville, TN

The speaker has signed a disclosure form and indicated she has no significant financial interest or relationship with companies or the manufacturer(s) of any commercial product/service that will be discussed as part of this presentation.

Session Summary

Although we deal with a specific population, the babies do not always read the same textbook that we do. This presentation will discuss unusual neonatal case presentations and the management provided.

Session Objectives

Upon completion of this presentation, the participant will be able to:
- discuss differential diagnoses for free air in the pleural space, atypical apnea, respiratory distress and hypoglycemia;
- describe free pleural air due to tracheal/subglottic tears, congenital central hypoventilation syndrome, congenital diaphragmatic hernia, and septo-optic dysplasia.

References


Session Outline

See handout on the following pages.
Interesting Cases from the Road

Patricia A. Scott, DNP, APN, NNP-BC, C-NPT

Pediatrics Medical Group of Tennessee
Advanced Practitioner Coordinator
Nashville, Tennessee

Vanderbilt University School of Nursing
Faculty, NNP and DNP Programs
Nashville, Tennessee

Case #1

- Term infant: 40 weeks / 3 kg

- Maternal info:
  - 38 years old
  - G8
  - O positive / prenatal labs negative

- Pregnancy history: unremarkable

Case #1 - continued

- Labor and delivery history:
  - Spontaneous labor
  - AROM 6 hours PTD
  - VAD secondary to fetal decelerations
  - Shoulder dystocia
  - Apgars: 8 / 7 / 7

Case #1

- Initial Exam:
  - Good cry initially, progressed to ↑ WOB
  - Neck swelling – soft, large, with slight bluish hue
  - Swelling more prominent on the right side
  - Required intubation at – 8-10 minutes of life due to increased WOB, cyanosis, neck swelling, and ↓ air entry bilaterally
  - Poor perfusion

Sign: Respiratory Distress and Neck Swelling

- Differential Diagnosis
  - Cystic hygroma
  - Hemangioma
  - Teratoma
  - Goiter
  - Third branchial cleft cyst
  - Sarcoma
  - Neuroblastoma
  - Subcutaneous emphysema
Case #1 - continued
- Chest x-ray made the diagnosis
- Needle aspiration performed emergently
- Chest tube placed
- Continued airway management

Case #1 - continued
- Enteral feeds started DOL#7
- Antibiotics for 7 days
- RA on DOL #12
- Discharged DOL #17

Case #1 - continued
- Discharge diagnosis: Subcutaneous emphysema secondary to dissecting air from pneumomediastinum possibly related to a spontaneous tracheal and/or subglottic tear.
Case #1 - continued

- Kacmarynski, D., Sidman, J., Rimell, F. & Hustead, V. (2002) reported four cases of spontaneous subglottic or tracheal rupture during a five year period of time at a Children’s Hospital.

- Ammari et al (2002) also published a similar case presentation.
- Perforated or injured trachea is not always associated with intubation attempts.

Case #2

- Term infant: 37.4 weeks / 2.3 kg

- Maternal Info:
  - 39 years old
  - G₂
  - O positive/prenatal labs negative

- Pregnancy history:
  - Insulin dependent diabetes mellitus
  - Hypothyroidism

Case #2 - continued

- Labor and delivery history:
  - Spontaneous labor
  - AROM –6 hours PTD
  - C/section secondary to FTP
  - Apgars: 7/9

Case #2 - continued

- Initial hospital course:
  - Respiratory:
    - Mechanical ventilation and surfactant therapy
    - Failed wean to NCPAP several times with respiratory insufficiency and apnea
    - Treated for laryngeal edema
    - Treated pharmacologically for apnea

- Cardiovascular:
  - Stable throughout

  - FEN:
    - Initially received TPN/IL
    - Enteral feeds initiated and advanced
    - Poor po feeder
Case #2 - continued

- Hematologic:
  - MBT: O positive
  - BBT: A positive / negative coombs
  - Physiologic jaundice resolved without intervention

- ID:
  - Initial septic work-up negative
  - Additional septic work-up – bacterial and viral coverage
  - Work-up, including LP and CSF PCR for HSV, negative

- Neurologic:
  - Development of hypotonia and encephalopathy noted
  - Continued apnea, requiring intervention (12 day old, former 37.4 weeker)

- Sign: Apnea
  - Differential Diagnosis – by system
    - Head and CNS
      - Perinatal asphyxia, IVH, SAH, meningitis, hydrocephalus, cerebral infarct, seizures, central hypoventilation syndrome
      - Respiratory
      - Hypoxia, airway obstruction, lung disease, inadequate ventilation

- Cardiovascular
  - CHF, PDA, congenital heart block, TOGV, HLHS
  - Gastrointestinal
  - NEC, GER, feeding intolerance
  - Hematologic
  - Anemia, polycythemia
  - Other
    - Temperature instability, infection, vagal reflex, drugs

- Many of these differentials could be excluded due to baby’s age

- Referring neonatologist was fairly certain of diagnosis based on one fact
  - Nurses reported apneas occurred during sleep

- Referring neonatologist was fairly certain of diagnosis based on one fact
  - Nurses reported apneas occurred during sleep
Case #2 - continued

- Infant transferred to the area Children’s Hospital for consults:
  - Genetics
    - Normal 46 XY
    - Inborn Errors – ruled out at referral facility
  - Pediatric Neurology
    - EEG – WNL
    - MRI – WNL
    - Hypercapnia test – pCO₂ 112

Congenital Central Hypoventilation Syndrome

- First reported in 1970 in an infant
- Characterized by progressive hypercapnia and hypoxemia during sleep
- These infants lack an arousal response to hypoxemia and hypercapnia
- Mechanical ventilation required long-term

Specific genetic test for Congenital Central Hypoventilation Syndrome (CCHS) sent – confirmed
- Identified mutation in the PHOX2B gene

Preparation for discharge
- Gastrostomy
- Fundoplication
- Tracheostomy
- Home ventilator care

Case #3

- Term infant: 39.6 weeks / 3.2 kg
- Maternal Info:
  - 22 years old
  - G2
  - O positive/prenatal labs negative
- Pregnancy history:
  - Hypothyroidism
Case #3 - continued
- Labor and delivery history:
  - Induced labor
  - AROM ~6 hours PTD
  - SVD
  - Apgars: 7 / 9

Sign: Respiratory Distress
- Differential Diagnosis
  - Respiratory Distress Syndrome
  - Transient Tachypnea of the Newborn
  - Pneumonia
  - Aspiration
  - Pneumothorax
  - Airway Obstruction
  - Diaphragmatic Hernia

1st CXR

Second Chest X-Ray

Case #3 - continued
- Initial hospital course:
  - Respiratory:
    - Mild respiratory distress – RA SaO2 60-70's
    - Placed under oxygen hood
    - Respiratory distress continued to progress
- UAC inserted
  - 7.25/56/51/24/-4.2
  - Physical exam remains consistent with respiratory distress
  - CXR for line placement and as comparison for lung fields
Next CXR

Case #3 - continued
- On initial physical exam
  - Abdomen soft, rounded
  - Decreased air entry bilaterally
  - Prenatal care including normal anatomy ultrasound

Case #3 - continued
- Intubation – mechanical ventilation
- Repogle tube to ILWS
- Sedation
- Transfer to Children’s Hospital

4th CXR
- Stable transport
- ECMO
- Did well

Congenital Diaphragmatic Hernia
- Abnormal communication between the peritoneal and pleural cavities allowing herniation of the intestine into the pleural space
  - Left sided: ~85%
  - Right sided: ~13%
  - Bilateral: ~2%
- Commonly, these infants are placed on ECMO prior to surgery
- Surgical repair required

Case #4
**Case #4**
- Term infant: 40.3 weeks / 3.6 kg

- Maternal Information:
  - 22 year old G1
  - A positive/prenatal labs negative

- Pregnancy History:
  - Uncomplicated

**Case #4 - continued**
- Labor and Delivery:
  - Labor induced

  - Complicated by elevated BP and non-reassuring FHR pattern with multiple variable decelerations

  - Vaginal delivery – nuchal cord x 1

  - Apgars: 9/5/9

**Case #4 - continued**
- Initial hospital course:
  - Transferred to NICU on DOL #2 secondary to hypoglycemia and temperature instability
  - Parenteral fluids started
  - Enteral feeds continued
  - Septic work-up obtained and antibiotics started
  - Phototherapy started

**Sign: Hypoglycemia**
- Differential Diagnosis
  - Transient hypoglycemia
    - Decreased glycogen stores
    - Hyperinsulinemia
    - Increased utilization of glycogen

  - Recurrent/persistent hypoglycemia

**Sign: Temperature Instability**
- Differential Diagnosis
  - Iatrogenic causes
  - Environmental
  - Equipment
  - End-user error

  - Infection

  - Central Nervous System Abnormalities

**Case #4 - continued**
- Other clinical signs:
  - Low borderline / baseline heart rate – 95-105 bpm

  - Head CT Scan to rule out SAH
    - No bleed
    - Absent septum pellucidum

  - Possible agenesis of corpus callosum

  - Head US
    - Agenesis of the corpus callosum confirmed
Sign: Absence of Corpus Collosum
- Isolated condition
- In combination with other cerebral anomalies
  - Arnold-Chiari malformation
  - Dandy-Walker syndrome
  - Andermann syndrome
  - Schizencephaly
  - Holoprosencephaly
  - Aicardi’s syndrome

Sign: Absent Septum Pellucidum
- Differential Diagnosis
  - Septo-optic Dysplasia (de Morsier Syndrome)

Case #4 - continued
- Ophthalmologic exam
  - Revealed optic nerve hypoplasia
  - Confirming the diagnosis of Septic-Optic Dysplasia (de Morsier Syndrome)
- Endocrine work-up
  - Diagnosed with hypothyroidism – placed on medication
  - ACTH stress test – levels low – placed on supplementation prior to discharge

de Morsier Syndrome
- Also called Septo-Optic Dysplasia
- Characterized by:
  - Abnormal development of the optic disk
  - Pituitary deficiencies
  - Absence of the septum pellucidum
  - Jaundice
- Treatment related to symptoms
  - Hormone replacement
  - Vision, physical, and occupational therapies
  - Prognosis dependent on severity of symptoms

Acknowledgements
- Ashraf Hamdan, MD
- Kathy Harrison, MSN, APN, NNP - BC
- Sarah Hassell, MD
- Delinda Pearson, MD
- Nurses and Medical Record Office staff at various hospitals