Air Travel Safety for Children with Critical Illness: Considerations for Voluntary Travel
Jessica Edgar, MSN, CPNP
Director of Medical Affairs
Make-A-Wish America

Learning Objectives
Cite three reasons for ensuring air travel is appropriate (or not) for critically ill children.

Disclosures
Jessica Edgar works for Make-A-Wish America and receives a salary.

1. No child chooses to be hospitalized on a voluntary air travel wish. Do no harm!
2. Hospitalization creates unnecessary stress on the child and family who are in an unfamiliar place. Do no harm!
3. Results in increased insurance claims and less money to grant wishes to other children with critical illness.
Novel Three Dimensional Optical Imaging Technique for the Evaluation of Pectus Excavatum

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Pediatric Nurse Practitioner, General Surgery
Manager, Advanced Practice
Lucile Packard Children's Hospital Stanford

Learning Objectives

• Describe the use of a novel 3D optical imaging technique to measure chest wall dimensions
• Discuss data correlation between the optical index (OI)/predictive Haller index (pHI) and the radiographic Haller index (rHI)
• Review clinical implications and opportunities for future studies
• Highlight nurse practitioner involvement in exploring the innovative imaging technique

Incidence

• Most common anterior chest wall deformity in children, resulting in a sunken appearance of the chest
• Affects 1 in 400 live births
• Male to female ratio 5:1
• ~46% have a family history of pectus excavatum
• May be present at birth, increases in severity during period of rapid growth in puberty
• Psychosocial impairment due to negative body image

Diagnosis

• History and Physical Examination
• Pulmonary function studies, if indicated
• Cardiac evaluation, if indicated

Treatment Options

• Imaging modalities have relied on the Haller index, which is calculated from a chest x-ray (lateral and anterior-posterior) or CT scan

Disclosures

No relevant financial disclosures or conflicts of interest
Methods

- 3D structure light imaging
- Correlation between optical index (OI) and radiographic Haller index (rHI)

Results

- **predicted Haller index (pHI)**

  \[
  pHI = (4.2396 \times OI) - (0.0291 \times \text{Height}) + (0.0226 \times \text{Weight}) - 0.9340
  \]

Conclusions

- Novel modality for imaging for adolescents with pectus excavatum can be made without radiation
- Optical index and predicted Haller index correlates well with radiographic Haller index
- Future studies: Track OI/pHI for non-operative and operative patients
- Develop predictive correction index
- Potential for volumetric quantification of chest wall deformities
- NP has direct role in evaluation and management of children adolescents with pectus excavatum and being a part of understanding innovative imaging modalities to provide anticipatory guidance

Acknowledgements and References

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- Stephanie Chao, MD, FACS
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- James Wall, MD, FACS
- Enrico Danzer, MD
- Dalis Szafer, BS
- Alison Pei, BS
- Sriraman Madhavan, MS
- Karen Barnaby, NP

References

It isn’t always asthma that takes your breath away!

Amanda C. Filippelli, MSN, MPH, APRN, AE-C
Pediatric Nurse Practitioner
Department of Pulmonary Medicine
Connecticut Children’s Medical Center

Learning Objectives

Describe how Vocal Cord Dysfunction (VCD) or Paradoxical Vocal Fold Movement (PVFM) is a possible etiology for shortness of breath and noisy breathing in the pediatric population.

Understand treatment options for VCD or PVFM.

What is VCD or PVFM?

• Abnormal closure of the vocal cords during inspiration – can happen at any time but often occurs during physical activity
• Official diagnosis requires flexible fiberoptic laryngoscopy during an episode however flat inspiratory loops on pulmonary function tests often can indicate VCD or PVFM.

Hallmark Characteristics of VCD or PVFM

• Difficulty getting air in
• Throat tightening
• Stridor or noisy breathing
• Shortness of breath that can be very scary, exacerbated by anxiety
• Little or no relief with bronchodilator

Management of VCD or PVFM

• Patients are often referred to Pulmonary or ENT specialists however if VCD is strongly suspected, can send directly to Speech therapy
• Speech and behavioral therapy
  • Breathing techniques
  • Biofeedback
  • Coping mechanisms
• Treatment of co-morbid conditions
  • Anxiety
  • Reflux
  • Allergies
  • Reactive airway disease

Disclosures

I have no disclosures.
Weaning High Flow: How slow do you go?

Dani Sebbens, DNP, CPNP-AC/PC

Learning Objectives
- Recognize the impact of high flow nasal cannula (HFNC) on intubation frequency
- Describe the benefit of using a weaning protocol for HFNC
- Consider using a weaning protocol in your practice setting

How did we get here?
- HFNC is generally comfortable and tolerated well by children
- In recent years, many authors report a significant decrease in the need for intubation associated with acute respiratory failure.
- Flow limits range from 0.5 liters/kg up to 2 liters/kg

Now what?
- Child has improved
- We avoided intubation
- Controversy remains regarding approach to weaning the respiratory support
- Lack of weaning impacts length of stay and patient discharge
- "Bronchiolitis HFNC Weaning Protocol"

Does patient have the primary diagnosis of viral bronchiolitis requiring high flow nasal cannula (HFNC)?

Are any of the following conditions present?
- Gestational age < 40 weeks
- Congenital heart disease
- Persistent apnea or periodic breathing

Excluded from guideline

Has child required change in HFNC settings in past 4 hours?
- FiO2 < 50%
- Flow less than 2 L/min/kg

Excluded from guideline

Start Weaning Phase

Excluded from previous step in 2 hours

Reassess from previous step in 1 hour

started by: Betters et al., 2017

Adapted from Betters et al., 2017
**Daily Test**

For children who were entered into weaning phase if flow is ≥ 3 L/min/kg.

- Observe the patient and score RSS (15 minutes).
- If RSS is less than 6, continue at the same flow rate for 15 minutes.
- If patient is on a ventilator, adjust to the same flow rate setting.
- If floor patient, reassess in 4 hours.

**Respiratory Score (RSS)**

<table>
<thead>
<tr>
<th>Item</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>RR</td>
<td>≤ 25</td>
</tr>
<tr>
<td>CVP</td>
<td>≤ 15</td>
</tr>
<tr>
<td>EpO2</td>
<td>≤ 90</td>
</tr>
<tr>
<td>PaO2</td>
<td>≥ 60</td>
</tr>
<tr>
<td>PaCO2</td>
<td>≤ 50</td>
</tr>
<tr>
<td>SaO2</td>
<td>≤ 90</td>
</tr>
</tbody>
</table>

- If RSS is ≥ 6, do not wean, then reassess in 2 hours.
- If RSS is 8 or greater, consider increasing flow and notify provider.

**Flow to Simple Nasal Cannula**

- If flow is greater than 3 L/min/kg, switch to simple nasal cannula directly.
- If flow is less than or equal to 3 L/min/kg, continue at the same flow rate settings.

**Flow Observation**

- Observe patient and score RSS (15 minutes).
- If RSS is less than 6, continue at the same flow rate for 15 minutes.
- If floor patient, reassess in 4 hours.

**References**

What’s old is new again: Use of non-invasive negative pressure cuirass ventilation

Mark A Washam, RRT, MSN, APRN (FNP, PNP-AC)
Nurse Practitioner – Pulmonary Division
Cincinnati Children’s Hospital Medical Center

Learning Objectives
• Understand what Negative Pressure ventilation is using Biphasic Cuirass Ventilation (BCV).
• Common uses for BCV
• Describe common settings for BCV.
• Identify the barriers to pediatric patient acceptance and demonstrate understanding in overcoming those barriers.
• Understand special considerations for routine nursing care and patient feeding.

Conventional mechanical ventilation
• In positive pressure ventilation (PPV), the gas pushed into the lungs naturally follows the path of least resistance, therefore best ventilating the already well-ventilated areas.
• Increases in pressures/volumes to aid ventilation of all areas of the lungs leads to barotrauma, volutrauma and possible development of a pneumothorax.
• These complications, along with those of ventilator associated pneumonia, are theorized to have no relevance with negative pressure ventilation.

For our discussion
• Cuirass, or chest cuirass will generically be referred as negative pressure ventilation (NPV).
• NPV is mode of non-invasive ventilation that utilizes a device (i.e., chamber, shell) over the patient or their chest and creates a lower than ambient pressure causing the patient’s chest to move and air to rush in or out of the lungs.

Disclosures
I have nothing to disclose
Photographs of patients in this presentation were used with permission of parents or guardians.

Warren E. Collins Iron Lung

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Does Negative Pressure Ventilation Still have a Role?

• YES!
• Advantages (some theoretically)
  • More physiologic, possibly (likely) better ventilation of the lungs than PPV
  • Better airway clearance (Dilatation of the airways improving expectoration)
  • Does not require artificial airway
  • Improvement in cardiac output vs positive pressure ventilation
  • Improvement in secretion management

Case Example

• Left lung atelectasis
• Mucus plugging
• Developing pneumonia
• Patient was sent to the PICU
• Placed on the PPV
• Increased airway clearance
• Close monitoring
• Could not get the L side to clear/open

After aggressive airway clearance, ABX, mucolytics and mechanical clearance with bronchoscopy, not significantly improved.
Complications & Issues

- Acceptance of device
- J-tube feeds
- Covered the J-tube stoma site
- BCV lack of portability.
- Unable to do PT with BCV.
- Cost for home use was high.
- Several different device failures.

Questions

You will go out in joy and be led forth in peace; the mountains and hills will burst into song before you, and all the trees of the field will clap their hands.

Isaiah 55:12

In fond memory of Caleb.
Drug-Induced or Iatrogenic Long QT Syndrome

Sarah S. LeRoy DNP, RN, CPNP-AC/PC
Assistant Clinical Professor
Wayne State University College of Nursing

Learning Objectives
• Define drug-induced or iatrogenic long QT syndrome
• Demonstrate the correct technique used to measure the QTc on the surface electrocardiogram
• Identify risk factors associated with drug induced long QT syndrome
• Identify a Smartphone App for QT prolonging drugs

Overview
• Numerous diverse pharmacologic agents that prolong the QT interval have been identified
• Drug-induced long QT syndrome (LQTS) is characterized by drug-induced QT interval prolongation and an increased risk for torsade de pointes
• Torsade de pointes (TdP) is a potential fatal ventricular tachyarrhythmia
• Due to the relatively rarity of this adverse effect, QT prolongation may not be identified until post market surveillance

Risk Factors: Drug Classifications
• Well over 100 pharmacotherapeutic identified
  • Antiarrhythmic agents
  • Antimicrobial agents
  • Psychoactive agents
  • Anti-emetic agents
  • Promotility agents

Disclosures
No relevant financial disclosure
Patient Risk Factors

Risk factors*
- Female gender (71%)
- Structural heart disease
- Hypokalemia
- Multiple QT prolonging drugs or agents interfering with their metabolism
- Prolonged baseline QTc > 450 ms
- Family history of LQTS

*Zeltzer, Justo, Halkin, Prokhorov, Heller, & Viskin, 2003

Guidelines for QT Measurement

- Measurements should be made manually from a 12-lead ECG
- Measure from the onset of the QRS complex to the end of T wave
- Prominent U waves should be included if they merge into the T wave
- Correct for heart rate by Bazett formula*

*Bazett Formula: \( QTc = \frac{QT}{\sqrt{R-R \text{ interval}}} \)

*Use the preceding R-R interval

Normal Values for QTc*

<table>
<thead>
<tr>
<th></th>
<th>Adult Males</th>
<th>Adult Females</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>&lt;440 ms</td>
<td>&lt;440 ms</td>
</tr>
<tr>
<td>Borderline</td>
<td>440-460</td>
<td>440-470</td>
</tr>
<tr>
<td>Prolonged</td>
<td>&gt;470</td>
<td>&gt;470</td>
</tr>
</tbody>
</table>


Resource for Drugs

- CredibleMeds https://crediblemeds.org/
- Multilingual Smartphone App for QT drugs
- QT drug list
- Drug-drug interactions
- Register so clinician can be contacted if change in QT drug list

References

How sweet it is: Infant Botulism
Heather Herrera, RN, CPNP-AC/PC

Objectives:
1. Identify presenting signs and symptoms of infant botulism.
2. Identify botulism resources available to both providers and parents.

Statistics
• Infant botulism
  • Rare but also the most common form
  • Infants 1-12 months are at risk, with the highest incidence observed among ages 1-6 months
  • Mortality rates have decreased

C. botulinum
• Gram-positive, spore-forming obligate anaerobe
  • Natural habitat – soil
  • Spores are commonly present on fresh fruits, vegetables, and other agricultural products such as honey
  • Botulinum toxin is the most poisonous substance known – listed as a potential biologic weapon agent

Pathophysiology
Sources of exposure

- Dust
- Honey (honey pacifiers)
- Oftentimes, exposure is unidentifiable

Presentation

- Signs and symptoms
  - Constipation
  - Listlessness, lethargy, poor feeding
  - Symmetric, descending paralysis
  - First signs of illness are found in the cranial nerves

Differential Diagnoses

- Sepsis
- Spinal muscular atrophy type 1
- Mitochondrial disorders
- Metabolic disorders
- Failure to thrive
- Idiopathic hypotonia
- Infectious disease disorders

Diagnosis

- C. botulinum in the feces of an infant with clinical signs consistent with the paralyzing action of botulinum toxin
- C. botulinum is NOT part of the normal flora of infants or adults
- If fecal specimen is obtained early enough in the illness course, it will contain botulinum toxin

Treatment

- BIG-IV (Baby-BIG)
  - Human botulism immune globulin intravenous
  - Reduces hospital stay and cost
  - Should be started as soon as possible to maximally neutralize the toxin (do not delay for confirmation of diagnosis)
  - Can be obtained from the California Department of Public Health as a public service orphan drug
  - Infantbotulism.org

Management

- Supportive care
  - Anticipation and avoidance of potentially fatal complications
    - Provide supportive care – may need PICU level of care
    - Endotracheal intubation is often necessary to maintain and protect the airway
NO ANTIBIOTICS

• Antibiotics
  • Should only be used to treat secondary infections
  • **antibiotics can result in lysis of intraintestinal C. botulinum with resulting release of intracellular neurotoxin into the gut lumen and absorption

Outcomes

• Recovery
  • Occurs through regeneration of the poisoned terminal unmyelinated nerve endings
  • New nerve twigs cause formation of new motor end-plates that are indistinguishable from the original ones

Patient Education

• If BabyBIG was administered:
  • Should have good recovery of muscle strength and tone before immunizations continue
  • Most live-virus vaccines (i.e., measles, mumps, rubella and varicella) will need to be delayed until 6 months after BabyBIG® treatment because the antibodies in BabyBIG® may interfere with the effectiveness of the vaccine.

Prevention

• Educate families – don’t feed honey to infants (only known preventative factor)
• Breast-feeding may help moderate the severity of illness and speed of onset

References

• https://www.slideshare.net/amanullah9803150/clostridium-56423432 (mechanism of action for botulism)
• www.tinypic.com (honey pacifiers)
• Infantbotulism.org
Neurotoxicity in Chimeric Antigen Receptor (CAR) T Cell Therapy
Jessica L. Spruit, DNP, CPNP-AC
Wayne State University College of Nursing
Detroit, MI
C.S. Mott Children’s Hospital Blood and Marrow Transplant
Ann Arbor, MI

Learning Objectives
• Review indications and utilization of CAR T cell therapy in pediatrics
• Describe neurotoxicity secondary to CAR T cell therapy
• Discuss screening and therapeutic intervention guidelines for neurotoxicity following CAR T cell therapy

Chimeric Antigen Receptor (CAR) T Cell Therapy
• Emerging therapy for B cell leukemia in children and lymphoma in adults
• Recently FDA approved
  • Tisagenlecleucel (Kymriah™) in August 2017 for patients up to 25 years of age
  • Axicabtagene ciloleucel (Yescarta™) in October 2017 for adults
• Evolving indications leading to increased utilization in our population
• Genetic modification of T cells to target and attack a specific antigen

Complications of CD19 CAR T Cell Therapy

<table>
<thead>
<tr>
<th>Cytokine Release Syndrome (CRS)</th>
<th>Neurotoxicity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fever</td>
<td>Headache</td>
</tr>
<tr>
<td>Hypotension</td>
<td>Tremor</td>
</tr>
<tr>
<td>Hypoxia</td>
<td>Lethargy</td>
</tr>
<tr>
<td>Organ dysfunction</td>
<td>Aphasia*</td>
</tr>
<tr>
<td></td>
<td>Agitation</td>
</tr>
<tr>
<td></td>
<td>Dysesthesia</td>
</tr>
<tr>
<td></td>
<td>Delirium</td>
</tr>
<tr>
<td></td>
<td>Encaphalopathy</td>
</tr>
<tr>
<td></td>
<td>Seizure</td>
</tr>
<tr>
<td></td>
<td>Cerebral edema (rare)</td>
</tr>
</tbody>
</table>

Immune Effector Cell-Associated Neurotoxicity (ICANS)
• Pathologic process involving the central nervous system following immune therapy
• Result of activation or engagement of endogenous or infused T cells or immune effector cells
• Symptoms may be progressive
• May present simultaneously with CRS or following CRS (rarely before)
• Less specific symptoms and signs excluded from definition
**ICANS Assessment and Grading**

**Children < 12 years**
- Cornell Assessment of Pediatric Delirium (CAPD) Score
  - Assess interactions throughout shift
  - Also validated in patients up to 21 years of age with developmental delay
  - CAPD score ≥ 9 is suggestive of delirium, consistent with grade 3 ICANS
  - Prolonged response during interactions

**Children > 12 years**
- Immune Effector Cell-Associated Encephalopathy (ICE) Score
  - Perform at least daily, and as clinically indicated
  - Scoring:
    - 10 = no impairment
    - 7‐9 = grade 1 ICANS
    - 3‐6 = grade 2 ICANS
    - 0‐2 = grade 3 ICANS
    - If score is 0 because patient is unarousable and unable to perform test, grade 4 ICANS

**ICANS Assessment and Grading**

**Children < 12 years**
- Cornell Assessment of Pediatric Delirium (CAPD) Score
- Level of consciousness
- Seizure
- Motor weakness
- Elevated intracranial pressure

**Children > 12 years**
- Immune Effector Cell-Associated Encephalopathy (ICE) Score
- Level of consciousness
- Seizure
- Motor weakness
- Elevated intracranial pressure

**Interventions for ICANS**
- Awareness of risk factors, prompt recognition
- Consideration of seizure prophylaxis
- Dexamethasone
  - Excellent CNS penetration
  - Ideal in the setting of neurotoxicity without CRS
- Concomitant CRS
  - IL-6 targeted therapy

**References**
To GRV or Not to GRV – That is the Question!

Ann-Marie Brown, PhD, CPNP-AC/PC, CCRN, CNE, FCCM
Assistant Clinical Professor, Emory University
Nurse Scientist, Children’s Healthcare of Atlanta

Disclosures
I have no commercial funding or other financial disclosures to report.
I am not presenting any off label medication recommendations.
The COBO2 study was funded by the Akron Children’s Hospital Foundation and the American Nurses Foundation

Learning Objectives
• Participants will be able to apply current evidence to monitor feeding intolerance in the critically ill pediatric patient.

Barriers to EN Delivery - Intolerance
• Fluid Overload
• Shock/gut hypoperfusion
• Vasopressors
• Lack of protocols
• Feeding Intolerance

State of the Science
• Systematic ROL reported FI most commonly defined as
  • Presence of GI symptoms and/or large gastric residual volumes (GRV)
  • Cessation of feeds due to GI symptoms
  • Inadequate delivery of EN
  • Median incidence of FI was 20% (IQR 7.4%-33%)
  • Large GRV, abdominal distension, diarrhea and emesis were most commonly reported GI symptoms
  • Authors recommended a standardized definition of FI as inability to achieve EN target intake in combination with presence GI symptoms/dysfunction

Guidelines

Definitions, predictors and outcomes of feeding intolerance in critically ill children: A systematic review
State of the Science

- Definition remains inconsistent, nebulous and arbitrary
- GRV remains the most common factor used to define FI, despite lack of evidence
- Emesis
- Bowel Sounds/Bowel movements
- Abdominal pain/distension
- Markers of systemic/gut perfusion

Continuous vs Bolus NG Feeds in Mechanically Ventilated Pediatric Patients (COBO2)

- **Secondary Outcomes:**
  - There is lower incidence and duration of avoidable feeding interruptions and feeding intolerance in a bolus compared to continuous gastric feeding protocol.
  - There is no relationship between gastric residual volume (GRV) or abdominal girth and the incidence of emesis.

Feed Interruptions by Intolerance Assignment

<table>
<thead>
<tr>
<th>Type of interruption</th>
<th>GRV Sites</th>
<th>Emesis Sites</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>P = 0.0035</em></td>
<td>0.9604</td>
<td>1.0000</td>
<td>0.6060</td>
</tr>
<tr>
<td>Emesis</td>
<td>20</td>
<td>18</td>
<td>2</td>
</tr>
<tr>
<td>procedures (non-surgical)</td>
<td>8</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>worsening clinical status</td>
<td>7</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>elevated GRV or connected measures</td>
<td>18</td>
<td>19</td>
<td>1</td>
</tr>
<tr>
<td>other</td>
<td>9</td>
<td>10</td>
<td>1</td>
</tr>
</tbody>
</table>

No emesis event was preceded by an elevated GRV
No Ventilator Associated Infection in either group
Thank you!

• Participating Sites
  - Akron Children’s Hospital, Akron, OH
  - Oklahoma Children’s Hospital, Oklahoma City, OK
  - Shands Children’s Hospital, Gainesville, FL
  - Cardinal Glennon Children’s Hospital, St. Louis, MO
  - Children’s Hospital of Philadelphia, Philadelphia, PA
  - Dartmouth-Hitchcock Children’s Hospital, Lebanon, NH
  - Children’s Hospital of Wisconsin, Milwaukee, WI

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References

Management of Pediatric Gynecological Complaints

Mary Schucker  MSN,MSHI,CPNP ,CRNP ,ENP-BC,PPCNP-BC
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Disclosures
None

Vaginal Bleeding
Pregnant or Not
Hemodynamically Stable or Not

History and Physical Exam
Oligomenorrhea

Vaginal Discharge

Joyce Adams Criteria

Injuries

Infection

Diagnostic Tests
Take Home Points

- Most children who are evaluated for suspected sexual abuse will not have physical signs of injury or infection.
- The child’s description of the events surrounding the incident is crucial.
- Emergency Evaluation is indicated:
  - Alleged assault may have occurred in the previous 72 hours
  - Prepubertal patients—evidence can be collected within 24 hours of assault
  - Within time frame for emergency contraception
  - Within time frame for STI testing/HIV Post Exposure Prophylaxis (HIV-PEP)
  - Report of genital injury, genital pain, or bleeding
  - Medical, psychological, or safety concerns
  - Concern for self harm or human trafficking
- Photo documentation is key
- Multidisciplinary team approach is crucial for success

Resources & References

- www.chop.edu/pathways/emergency
- QR CODE
  https://epdf.pub/textbook-of-pediatric-emergency-medicineb334df8b462c2876030a77612b3e1d699440.html
- https://www.jpagonline.org/article/S1083-3188(17)30542-9/fulltextInterpretation of Medical Findings in Suspected Child Sexual Abuse: An Update for 2018
Adams, Joyce A. et al.
Journal of Pediatric and Adolescent Gynecology, Volume 31, Issue 3, 225 - 231

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