Non-invasive Ventilation in Children

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Objectives

- Identify positive outcomes with NIV use
- Identify indications for CPAP in children with obstructive sleep apnea.
- Identify the indications and proper mode of non-invasive ventilation in children with neuromuscular disorders
- Recognize complications of non-invasive ventilation in children
- Identify strategies to improve compliance in children requiring non-invasive ventilation
Cases
Snoring in an Overweight Child

- My 14-year-old son has obstructive sleep apnea as documented on an overnight sleep study.
- On physical exam:
  - Tonsils have been surgically removed OR
  - 1+ tonsils
  - BMI = 34 kg/m2
- What treatment options are available?
My 15-year old son has Trisomy 21. He used to have symptoms of obstructive sleep apnea but is status post a T&A. During his annual physical, we let his PMD know that the snoring had recurred.

A sleep study confirmed the diagnosis of recurrent obstructive sleep apnea.

What treatment options are available?
Trisomy 21

- Risk factors
  - Adenotonsillar hypertrophy
  - Large tongue
  - Midface hypoplasia
  - Pharyngeal hypotonia

- Health supervision per AAP
  - Discuss symptoms of OSA
  - Referral for sleep study by 4 years of age
  - Referral for sleep study after 4 years if symptoms

- May have more post-operative complications
- Increased risk for persistent/recurrent OSA

Bull. Pediatrics 2011;128
Rosen. Pediatrics 1994;93
Trisomy 21 – Increased risk of OSA

~1/2 had sleep-disordered breathing

~1/3 had persistent OSA

Amos, APSS 2014
Sleep-disordered breathing in a teen with Duchenne muscular dystrophy

- A 17-year-old young man presents with concerns for poor sleep.
  - Snoring
  - Frequent nocturnal awakenings that require repositioning
  - Morning headaches

- Physical examination
  - Wheel-chair bound
  - SpO2 94% in room air
  - ETCO2 48 mmHg
  - No ATH
  - BMI = 15
  - Status post PSF surgery

- Diagnostics
  - FVC = 27%
  - MIP = -20 cm
Correlation between daytime lung function and nocturnal hypoventilation

FEV1 <40% associated with nocturnal hypercapnia

Hukins, Amer J Respir Crit Care Med 2000;161:166
Central hypopneas in REM – No desaturations
Central hypopneas in REM – Mild desaturations
Nocturnal Hypoventilation in a Child with Congenital Central Hypoventilation Syndrome

- **Diagnosis:** 1 mo old
- **Trach placed at time of diagnosis**
- **Decannulation = 8 years**
- **Device = Trilogy**
- **Mode: AC/VC**
- **Mask: full face mask**

<table>
<thead>
<tr>
<th>Services involved in transition from IV to NIV</th>
<th>Key roles</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pediatric Intensive Care</td>
<td>ICU management during hospitalization</td>
</tr>
<tr>
<td>Pediatric Pulmonology</td>
<td>Bronchoscopy, IV and NIV management</td>
</tr>
<tr>
<td>Pediatric Otolaryngology</td>
<td>Bronchoscopy, stoma leak management, TCF closure</td>
</tr>
<tr>
<td>Pediatric Sleep Medicine</td>
<td>Polysomnogram(s) on NIV</td>
</tr>
<tr>
<td>Respiratory Therapy</td>
<td>Mask fittings, equipment teaching, equipment management</td>
</tr>
<tr>
<td>Advanced Practice Nursing</td>
<td>Oversight of transition process, Pulmonary provider support</td>
</tr>
<tr>
<td>Durable Medical Equipment</td>
<td>Provide/exchange equipment as needed during transition</td>
</tr>
<tr>
<td>Social Work</td>
<td>Help keep necessary services and equipment in the home</td>
</tr>
<tr>
<td>Home Nursing Agency</td>
<td>Provide continued home nursing support if needed after decannulation</td>
</tr>
</tbody>
</table>
Demographics

- Incidence of children managed with NIV is UNKNOWN
- Most common medical conditions managed with NIV:
  - OSA – 29% of studies
  - Spinal muscular atrophy – 8%
  - Sleep-disordered breathing – 6%
  - Neuromuscular disease – 5%
- Mean age of NIV initiation: 8.06 ± 3.08 years

Castro-Codesal, Sleep Med Reviews, 2017, in press
Demographics (2)

• Medical conditions by age:
  › Infants:
    • Upper airway disorders (52% of studies)
    • Neuromuscular disorders (33%) (SMA most common)
    • CCHS (4%)
  › Children:
    • Upper airway disorders (41%)
    • Neuromuscular disorders (20%)
    • SDB related to obesity (10%)
    • Pulmonary disorders (8%)

Castro-Codesal, Sleep Med Reviews, 2017, in press
Demographics (3)

• NIV Equipment
  › CPAP – 25% of studies
  › Bi-level – 21%
  › Auto-Pap – 2%
  › Combined CPAP and Bi-level – 22%
  › Other – 30% (not specified, negative pressure)

• NIV Equipment based on disease
  › CPAP: Upper airway disorders with/without obesity
  › Bi-level: Musculoskeletal or pulmonary disorders

Castro-Codesal, Sleep Med Reviews, 2017, in press
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Children's Hospital of Wisconsin
Demographics in Milwaukee

- 294 children
- Age range: 3 – 24 yrs
- Gender:
  - 194 M and 100 F
- Device
  - CPAP – 119
    - AutoCPAP - 73
  - BiPAP - 88
  - Ventilator - 14
- Primary sleep diagnosis
  - OSA – 216
  - Hypoventilation – 60
  - CSA – 4
  - Altered pulmonary mechanics – 14
- Co-morbid diagnoses
  - Obesity
  - Neuromuscular weakness
  - Trisomy 21
  - CCHS
Outcomes with the Use of NIV

• Improve clinical symptoms
• Improve neurobehavioral functioning
• Improve PSG parameters
• Assist with weight management
• Improve pulmonary health
• Reduction in healthcare encounters
• Improved quality of life
• Improved longevity

Marcus, Pediatrics 2006; 117:e442-e451
Non-invasive ventilation: Pros and Cons

• Advantages
  ✓ Avoids perioperative complications of adenotonsillectomy.
    › But does have localized/temporary effects from equipment such as nasal irritation, skin breakdown.
  ✓ May serve as a “bridge” preoperatively until surgery to reduce morbidity.
  ✓ Can be used postoperatively for residual OSA.
    › Useful in obese and complex patients.

• Disadvantages
  ✓ Poor compliance
    › ≥3-4h of use per night is considered good compliance.
    › Children require more sleep than adults; therefore such limited use may not be adequate.
  ✓ Requires training for parents as well as patients.
  ✓ Lifelong use required.
  ✓ Risk for aspiration of stomach contents.
    › Very young.
    › Significant GERD.
    › Neuromuscular weakness.
Age and NIPPV

• NIPPV may be implemented at any age. However, certain age groups are particularly problematic:
  ✓ Preemies or infants <4 kg
  ✓ “Terrible two-year-old”
  ✓ Adolescents
  ✓ Developmental delays
Indications for CPAP

• Persistent OSA following T&A
  ✓ Abnormalities in up to 25-50% of children
  ✓ Risk factors include:
    › Underlying condition
      • Craniofacial abnormality
      • Obesity
    › Severe preoperative OSA
• Patient not a candidate for airway surgery
• Perioperative management of severe OSA
Other Indications for NIV

- Hypoventilation
  - Related to obesity
  - Related to neuromuscular weakness
  - Abnormalities in central respiratory drive
- Central sleep apnea
  - Brainstem malformation
  - Brainstem tumor
- Altered pulmonary mechanics
  - Chronic lung disease
  - Skeletal abnormalities
  - Congenital heart disease
  - Diaphragm paralysis
NIV in Neuromuscular Patients

• Indications:
  ✓ Correct chronic respiratory insufficiency
  ✓ Prevent recurrent pneumonia or atelectasis
  ✓ Improve sleep quality if increased work of breathing or respiratory arousals

• Questions:
  ✓ Nocturnal vs. Continuous?
  ✓ Positive pressure vs. Negative pressure?
  ✓ Noninvasive vs. Invasive?
  ✓ Elective vs. Urgent
Mask
Features
Accessories
Settings
Noninvasive ventilation

- Modes
  - CPAP
  - AutoCPAP
  - Bilevel
  - Ventilator

- Interfaces
  - Nasal pillows
  - Nasal mask
  - Full face mask
  - HFNC
  - Mouth piece (daytime)
Which Mask is the Best Mask?

- The one that the child will wear
- Be prepared to change a few times
  - Average 2 times in first 3-6 months
- Don’t avoid full face masks
  - But recognize the concerns
  - Troubleshoot for safety
Philips Respironics Wisp Pediatric
- Silicone nasal mask
- 3 sizes designed for young children
- FDA approved for 22 lbs and up

Sleep Net MiniMe2®
- Gel nasal mask, with adjustable wire frame
- 2 sizes
- FDA cleared for ages 2-12 years old

AG Industries Nonny™
- Pediatric nasal mask
- 4 sizes.
- FDA approved for ≥1 year and older. Fits toddlers to adolescents
ResMed Pixi™
- Silicone nasal mask,
- One size
- Authorized for children age $\geq$ seven years

Philips Respironics Wisp Youth
- Silicone nasal mask
- Authorized for children age $\geq$ seven years

Circadiance Sleep Weaver ® Advance Pediatric
- Cloth mask
- One size
- FDA cleared for children 2-7 years old.
- Especially effective when used with patients with skin breakdown or contact dermatitis
Full and Total Face Masks

- Philips Respironics
  - FitLife
  - Approved for children 7 years and up

- ResMed Quattro™ FX for Her

- Respironics
  - ComfortGel Blue

- Respironics
  - Amara View
Extended Features

- **I time**
  - I time min and I time max
- **C-flex® or EPR**
  - Exhalation relief
- **Ramp**
Accessories

- Chin strap
- Soft collar
- Battery pack
- Heated wire circuit
- Airview/MyAir
- Heated Humidification

Children's Hospital of Wisconsin

Medical College of Wisconsin
• No significant difference in CPAP levels among different age groups
• No correlation between AHI and CPAP level

O’Donnell, Sleep 2006;29:651-658
Settings: AutoCPAP

- 26 children with autoCPAP
- Started CPAP at home but came to lab for titration study
- PSG pressure > AutoMean pressure
- Interestingly, 21 children changed to fixed CPAP
  - Adherence similar

Mihai, J Clin Sleep Med 2017;13:713-718
Settings: Bilevel
Neuromuscular patients

- IPAP: 10-20 cmH2O
- EPAP: 2-6 cmH2O
- Mode: ST (spontaneous timed)
- Rate: Similar to patient’s rate
- Inspiratory time: 1 second
Settings: Ventilator

- Trilogy ventilator has two settings:
  - Primary: Night time settings
  - Secondary: Day time settings
High-flow Nasal Cannula

- 10 school-age children with OSA and intolerant of CPAP
- OAHI = 11.1 events/hr (interquartile range 8.7-18.8)
  - OAI = 2.2 events/hr
  - OHI = 9.9 events/hr
- HFNC titration
  - Started 5 or 15 L/min, pediatric or adult cannula
  - Increased by 5 or 15 L/min increments
  - Maximum 20 or 50 L/min
- Final OAHI = 2.1 events/hr

NIPPV during the day

- Mouthpiece ventilation
  ✓ “Sip and Puff”
- Settings:
  › Assist control/pressure control
  › Higher PIP to = “large sigh”
  › IMV = 0-1
Complications of NIV

- Skin irritation or breakdown
- Eye irritation
- Nasal dryness or congestion
- Aerophagia or gastroesophageal reflux
  › Be aware of g-tubes
- Chest discomfort
- Mid-face hypoplasia
Complications of NIV mask

- At age four, only about 60% of the adult facial bone structure is fully developed.
- Bones in the mid-face area are malleable during childhood.
- Prolonged use of nasal positive airway pressure masks can displace the teeth and upper jaw bone inward.

Patrick R. Sleep Review. November 2006
Kasey K, Chest. 2000;117
15 Year Old With Mid-Facial Bone Displacement Secondary to CPAP
Mid-Facial Bone Displacement and Correction

Before                            After
Clinical Practice Guidelines

- Adenotonsillectomy is first line of treatment for most children.
  - CPAP is an option for those who are not candidates for surgery or do not respond to surgery.
  - Treatment for select cases includes weight loss, management of allergies, craniofacial surgery, tracheotomy.

AAP, Pediatrics, 2012; 130:576-584
Practice Parameters: Respiratory Indications for PSG - Standards

• PSG is indicated when clinical assessment suggests OSA
• Children with mild OSA pre-operatively should have clinical assessment after T&A. PSG should be performed if there are residual symptoms
• PSG is indicated post-operatively if child had moderate to severe OSA, or other risk factors (e.g., obesity, craniofacial anomalies, neuromuscular weakness)
• PSG is indicated for PAP titration
Adherence
Adherence to NIV in Children

• Adherence is variable…..although overall pretty good

• Children and parents tend to overestimate adherence
  › 5.8 hours (download) vs 7.6 hours (report)

• Adherence improves over time
  › 39% of children accepted NIV immediately
  › 66% of children accepted NIV within 3 months
  › 82% of children accepted NIV by 46 months (!)

Marcus, Pediatrics 2006;117;e442-e451
O’Donnell, Sleep 2006; 29:651-658
Adherence to NIV in Children (2)

- 76% of children use NIV at least half of the days
  - Half of children used NIV 75% of the time
- Children without associated medical disorders were slightly more likely to accept NIV
- Children using a full-face mask took longer to be compliant
# Milwaukee Adherence Data

**Demographics**

<table>
<thead>
<tr>
<th>Demographics</th>
<th>N=111</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Gender</strong></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>60%  (N=67)</td>
</tr>
<tr>
<td>Female</td>
<td>40%  (N=44)</td>
</tr>
<tr>
<td><strong>Race</strong></td>
<td></td>
</tr>
<tr>
<td>Caucasian</td>
<td>48.6% (N=54)</td>
</tr>
<tr>
<td>African-American</td>
<td>36.9% (N=41)</td>
</tr>
<tr>
<td>Hispanic</td>
<td>11.7% (N=13)</td>
</tr>
<tr>
<td>Multiracial</td>
<td>0.9%  (N=1)</td>
</tr>
<tr>
<td>Not documented</td>
<td>1.9%  (N=2)</td>
</tr>
</tbody>
</table>

**Primary SDB**

- Obstructive sleep apnea, N=78
- Hypoventilation, N=17
- Altered respiratory mechanics, N=14
- Central sleep apnea, N=2

**Age**

<table>
<thead>
<tr>
<th>Age</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Median age at set-up</td>
<td>14.1 yrs (range 0.05-33.1 yrs)</td>
</tr>
<tr>
<td>&lt; 7 years old at set-up</td>
<td>11.7% (N=13)</td>
</tr>
</tbody>
</table>

Amos, APSS 2015
Baughn, APSS 2012
Symptom improvement at phone follow-up carried over to 1st PAP clinic visit

<table>
<thead>
<tr>
<th>Phone follow-up</th>
<th>1st PAP clinic visit</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Improved daytime energy</td>
<td>Improved daytime symptoms</td>
<td>0.042</td>
</tr>
<tr>
<td>Improved daytime sleepiness</td>
<td>Improved daytime symptoms</td>
<td>0.016</td>
</tr>
</tbody>
</table>

Symptom Improvement

- Nighttime symptoms
- Daytime symptoms

- Phone follow-up (SLEEP 2015), N=98
- 1st PAP clinic visit (SLEEP 2015), N=82
- 1st PAP clinic visit (SLEEP 2012), N=199
- 2nd PAP clinic visit (SLEEP 2015), N=50
- 2nd PAP clinic visit (SLEEP 2012), N=138
Adherence at phone follow-up carried over to 1st PAP clinic visit

*Objective adherence = >70% nights used > 4 hours by data download
**Subjective adherence = reported all night use 6-7 nights per week
## Adherence Outcomes

<table>
<thead>
<tr>
<th>Adherence (≥70% nights used &gt; 4 hours) was more likely with:</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Phone follow-up</strong></td>
<td></td>
</tr>
<tr>
<td>Altered respiratory mechanics vs. OSA</td>
<td>0.033</td>
</tr>
<tr>
<td>Inpatient vs. outpatient set-up</td>
<td>0.004</td>
</tr>
<tr>
<td><strong>1st PAP clinic visit</strong></td>
<td></td>
</tr>
<tr>
<td>Caucasian vs. African-American</td>
<td>0.015</td>
</tr>
<tr>
<td>Hypoventilation vs. OSA</td>
<td>0.006</td>
</tr>
<tr>
<td>Inpatient vs. outpatient set-up</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Bilevel PAP vs. CPAP</td>
<td>0.013</td>
</tr>
</tbody>
</table>
Steps to Achieve Adherence: Pre Sleep Study

- Pre-CPAP clinic
  - ✓ Meet with APN and RRT
    - › Discuss preconceived concerns
      - • Sadness about diagnosis
      - • Benefits if use NIV
    - › Identify possible barriers
    - › Partnership with child and parents
      - • Model expectations using family members
      - • Prepare bedroom
  - ✓ Mask fitting
- • Desensitization program for home
Interface Desensitization

• Interface desensitization done before a titration sleep study
• Child wears the interface during the day, while doing something they enjoy, like a favorite movie or game
  ✓ Gradually increase time
  › Maximum time = 20-30 minutes
  ✓ Can use positive rewards

Don’t put that scary mask on me!
Early Identification of Barriers to Adherence: Post Sleep Study

- Early identification
  - 1 week after starting NIV:
    - Phone call
    - Compliance download
- Early intervention
Steps to Achieve/Improve Adherence: Post Sleep Study

- 4-6 weeks after starting NIV
  - 1st CPAP multidisciplinary clinic visit
  - Compliance Download
- Behavioral interventions as necessary
  - Verbal and Visual
    - Reminder >4 hours for >70% of nights
  - Review barriers and problem solve
  - Engage child and parent
    - Results are better when parents participate
- Positive reinforcement in clinic and at home
## Compliance Report

**Usage**

<table>
<thead>
<tr>
<th>Description</th>
<th>Days</th>
<th>Hours</th>
</tr>
</thead>
<tbody>
<tr>
<td>Usage days</td>
<td>79/80 (99%)</td>
<td>655 hours 30 minutes</td>
</tr>
<tr>
<td>&gt;= 4 hours</td>
<td>79 days (99%)</td>
<td></td>
</tr>
<tr>
<td>&lt; 4 hours</td>
<td>0 days (0%)</td>
<td></td>
</tr>
</tbody>
</table>

**AirCurve 10 ST**

- **Serial number**: 22161389163
- **Model**: Seept Timed
- **IPAP**: 12 cmH2O
- **EPAP**: 5 cmH2O
- **Respiratory rate**: 14 bpm

**Therapy**

- **Leaks - L/min**: Median: 0.1, 95th percentile: 6.2, Maximum: 12.2
- **Events per hour**: Alt: 0.0, HP: 0.0, Al+HP: 0.0

**Usage - hours**

![Usage chart graph]
Compliance Download - Ideal

Patients can surprise you
Compliance Download – Not so Ideal

### Compliance Report

**Usage**

<table>
<thead>
<tr>
<th>Days</th>
<th>Hours</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-4</td>
<td>56/60 days</td>
<td>85%</td>
</tr>
<tr>
<td>&gt; 4</td>
<td>91 days</td>
<td>80%</td>
</tr>
<tr>
<td>&lt; 4</td>
<td>5 days</td>
<td>8%</td>
</tr>
</tbody>
</table>

**Usage hours**

- Average usage (total days): 7 hours 0 minutes
- Average usage (days used): 7 hours 20 minutes
- Median usage (days used): 7 hours 45 minutes

#### AirSense 10 AutoSet For Her

- **Serial number**: 23162164004
- **Mode**: AutoSet for Her
- **Min Pressure**: 9 cmH2O
- **Max Pressure**: 15 cmH2O
- **EPR**: Off
- **EPR level**: 2

**Therapy**

- **Pressure - cmH2O**: Median: 9.1, 95th percentile: 10.4, Maximum: 10.7
- **Leaks - L/min**: Median: 12.6, 95th percentile: 40.7, Maximum: 80.3
- **Events per hour**: Total: 4.8, High: 0.0, AH1: 4.8
- **Apnea Index**: Central: 0.8, Obstructive: 0.2, Unknown: 3.7
- **RERA Index**: 0.0

**Usage - hours**

![Graph of usage hours]
Compliance Summary

Date Range: 2/7/2017 - 5/7/2017 (90 days)
Percent Days with Device Usage: 70.0%
Average Usage (All Days): 4 hrs. 38 mins. 17 secs.
Average Usage (Days Used): 6 hrs. 37 mins. 33 secs.
Percent of Days with Usage >= 4 Hours: 58.9%
Assessment of Treatment Barriers

- Medical
  - Nasal congestion
  - Eye irritation
  - Dry mouth
  - Skin breakdown

- Comfort
  - Mask
  - Pressure
  - Humidity
  - Alarms

- Life-style
  - Glasses
  - Orthodontics
  - Fall asleep outside bedroom
  - No parent supervision
  - Sleepovers
  - Two homes
Gradual Exposure to Improve Adherence

- Tolerate mask without device attached
- Begin with afterschool practice times then advance to part of bedtime routine
  - Application, positive reinforcement, reassurance modeled with parents
  - Gradually increase compliance expectations so that child experiences success
- Add NIV device
  - Can use a low pressure (e.g., 4 cm)
- Utilize ramp feature
Follow-Up

• Clinic visits:
  ✓ Twice yearly if doing OK
  ✓ Every 3-4 months if struggling
    › With intermittent downloads

• Sleep studies:
  ✓ No “hard and fast” rules
    › If new symptoms
    › If significant weight change
    › If family thinks SDB has improved/resolved
    › With milestones
      • Adolescence
      • Before transition to adult sleep specialist
Good Night, Sleep Tight!