Update on severe OSAS

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Overview

• Introduction and definitions

• How to diagnose OSA in children and classify its severity?

• Why is an objective diagnosis important?

• Treatment algorithm of severe OSA
Introduction

- Pediatric obstructive sleep apnea (OSA) affects 2-5% of the general pediatric population.
- The prevalence is much higher in children with specific risk factors:
  - Obesity
  - Neuromuscular disease
  - Craniofacial abnormalities
- Pediatric OSA is associated with significant complications affecting the developing central nervous and cardiovascular systems.
Introduction

• Continuum of obstructive breathing disorders:
  - Normal
  - Primary snoring
  - Upper airway resistance syndrome
  - Obstructive hypoventilation
  - Obstructive sleep apnea
    • Mild
    • Moderate-to-severe
HOW TO DIAGNOSE OSA IN CHILDREN AND CLASSIFY ITS SEVERITY?
Obstructive Sleep-Disordered Breathing in 2-18 Year Old Children: Diagnosis and Management

An ERS Document
ERS Task Force 2012-09

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Stepwise Approach to the Diagnosis and Management of Obstructive SDB (2-18 y.o.)

STEP 1. Child at risk for SDB if (one or more):
1.1 Symptoms of upper airway obstruction (snoring, apnoea, restless sleep, oral breathing)
1.2 Findings on exam (tonsillar hypertrophy, obesity, midface deficiency, mandibular hypoplasia, neuromuscular disorders, Down syndrome, Prader-Willi syndrome)
1.3 Objective findings related to SDB (lateral neck X-ray, flexible nasopharyngoscopy, cephalometry, upper airway MRI or CT)
1.4 Prematurity or family history of SDB

STEP 2. Recognition of morbidity and conditions co-existing with SDB:
2.1 Morbidity
   - Cardiovascular system
     a. Elevated blood pressure
     b. Pulmonary hypertension and cor pulmonale
   - Central nervous system
     a. Excessive diurnal sleepiness
     b. Inattention/hyperactivity
     c. Cognitive deficits/academic difficulties
     d. Behavioural problems
       - Enuresis and somatic growth delay or growth failure
       - Decreased quality of life
   2.2 Conditions co-existing with SDB (probably common pathogenesis)
     a. History of recurrent otitis media or tympanostomy tube placement
     b. Recurrent wheezing or asthma
     c. Metabolic syndrome
     d. Oral-motor dysfunction

STEP 3. Recognition of factors predicting long-term persistence of SDB:
3.1 a. Obesity
       b. Male gender
       c. Obstructive apnoea-hypopnoea index-AHI > 5 episodes/h (step 4)
       d. Black race
       e. Untreated tonsillar hypertrophy, narrow mandible

STEP 4. Objective diagnosis and assessment of SDB severity:
4.1 Polysomnography (PSG) or polygraphy if child at risk for SDB (see steps 1 and 2)
4.2 OSAS-Definition 1: obstructive AHI ≥2 episodes/h or obstructive apnoea index ≥1 episode/h;
OSAS-Definition 2: AHI ≥1 episode/h (including central events)
4.3 If AHI >5 episodes/h: SDB unlikely to resolve spontaneously and child at risk for morbidity
4.4 If PSG or polygraphy not available: ambulatory PSG, nocturnal oximetry, Paediatric Sleep Questionnaire or Sleep Clinical Record
Diagnosis

- Limited diagnostic value of clinical symptoms and findings on physical examination.
- No correlation with OSA severity.
Diagnosis

• Overnight, attended, in-laboratory polysomnography is the gold standard test.

• However, polysomnography has many disadvantages and may not always be possible or feasible in which case alternative diagnostic methods should be used.
  - Previously untreated, otherwise healthy, non-obese children at risk for OSA due to adenotonsillar hypertrophy may be of low priority for polysomnography.
Diagnosis

- Children with symptoms of OSA and prior to adenotonsillectomy especially in the presence of obesity, craniofacial deformities, neuromuscular disorders, complex abnormalities or when the need for treatment is unclear.

- Post-adenotonsillectomy in patients with: persistent symptoms of OSAS despite surgery, moderate-to-severe OSAS preoperatively, obesity, craniofacial deformities, neuromuscular disorders or complex abnormalities.
Diagnosis

- Prior to and after rapid maxillary expansion or application of orthodontic appliances, continuous positive airway pressure (CPAP) and noninvasive positive pressure ventilation (NPPV) for treating OSAS.
Diagnosis

- Different scoring rules in pediatrics:
  
  - American Academy of Sleep Medicine Manual for the Scoring of Sleep and Associated Events.

  - Can be used in children and adolescents up to 18 year. However, a sleep specialist can decide to use the adult rules from 13 years or older depending on the clinical context.
Diagnosis

• Obstructive apnea/hypopnea:
  - lasts for at least two missed breaths or the duration of two breaths as determined by the baseline breathing pattern.

• Central apnea:
  - Lasts for at least 20 s or of at least two missed breaths, but associated with arousal, awakening or ≥ 3% desaturation.
Diagnosis

• Sleep-related hypoventilation is defined when CO2 levels are $> 50$ mmHg as measured by transcutaneous or end-tidal CO2 sensors for $> 25\%$ of total sleep time.
Diagnosis

• Normative data:
  - In healthy infants, central apneas frequently occur but are of short duration and are not followed by bradycardia or oxygen desaturation.
  - Obstructive and mixed apneas are less frequently seen.
  - Obstructive apnea or mixed apnea appear to be more common in premature infants and decreases in frequency over the first year of life.
  - In general, the obstructive apnea index is below 1 in the first year of life both in full-term and in pre-term infants.
Diagnosis

• Normative data:
  - During the first year of life, the number of central apneas decreases.
  - Periodic breathing normally disappears in the first 6 months of life.
  - During childhood and adolescence, central apneas can still occur although at a lower frequency that decreases with age.
  - Obstructive and mixed events are rarely seen.
  - Mean saturation during sleep is approximately 97% with a mean nadir of ≥ 90%.
Reference Values for Sleep-Related Respiratory Variables in Asymptomatic European Children and Adolescents

**TABLE 4—Comparison of Previously Published Sleep Architecture and Respiratory Events Data**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Current study</th>
<th>Marcus et al.(^6,7)</th>
<th>Uliel et al.(^8)</th>
<th>Traeger et al.(^9)</th>
<th>Montgomery-Downs et al.(^10)</th>
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<tbody>
<tr>
<td>N</td>
<td>60</td>
<td>50</td>
<td>70</td>
<td>66</td>
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<td>Age group</td>
<td></td>
<td></td>
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<tr>
<td>Mean ± SD</td>
<td>11.7 ± 2.6</td>
<td>9.7 ± 4.6</td>
<td>7.9 ± 4.4</td>
<td>6.6 ± 1.9</td>
<td>6.8</td>
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<td>Range</td>
<td>7.1–16.6</td>
<td>1.1–17.4</td>
<td>1.0–15.0</td>
<td>2.5–9.4</td>
<td>6.0–8.6</td>
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<td>TST (hr)</td>
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<tr>
<td>Mean ± SD</td>
<td>7.8 ± 0.8</td>
<td>6.0 ± 1.6</td>
<td>6.5 ± 1.2</td>
<td>7.7 ± 0.9</td>
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<td>Sleep efficiency (%)</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Mean ± SD</td>
<td>80.5 ± 8.5</td>
<td>NP</td>
<td>90.8 ± 6.5</td>
<td>89.0 ± 8.0</td>
<td>90 ± 7.0</td>
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<td>Mean ± SD</td>
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<td>NP</td>
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<td>NP</td>
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<td>CAI(^2)</td>
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<tr>
<td>Mean ± SD</td>
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<td>NP</td>
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<tr>
<td>Mean ± SD</td>
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<td>0.10 ± 0.50</td>
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<td>0.01 ± 0.03</td>
<td>0.05 ± 0.11</td>
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<td>0.00–3.10</td>
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<td>oAHI</td>
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<td>Mean ± SD</td>
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<td>RDI</td>
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<tr>
<td>Mean ± SD</td>
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<td>0.00–6.60</td>
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<td>Total arousal index</td>
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<tr>
<td>Mean ± SD</td>
<td>6.1 ± 1.8</td>
<td>NP</td>
<td>5.29 ± 3.49</td>
<td>11.2 ± 4.3</td>
<td>9.5 ± 5.3</td>
</tr>
<tr>
<td>Range</td>
<td>2.7–10.9</td>
<td></td>
<td>5.4–21.5</td>
<td></td>
<td>NP</td>
</tr>
</tbody>
</table>

\(^{6,7}\) Marcus et al., \(^8\) Uliel et al., \(^9\) Traeger et al., \(^10\) Montgomery-Downs et al.
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4.4 If PSG or polygraphy not available: ambulatory PSG, nocturnal oximetry, Paediatric Sleep Questionnaire or Sleep Clinical Record

• OSAS definition 1:
  - CHAT trial: more normalisation after adenotonsillectomy than in control group

• OSAS definition 2:
  - Most commonly used in research studies
  - Modest improvement in behavioural difficulties after adenotonsillectomy
Diagnosis

• OSA severity is most commonly defined as:
  - Mild: 2 < AHI < 5
  - Moderate-to-severe: AHI > 5

• No information on gas exchange abnormalities.

• No information on symptoms, morbidity.
  - No clear-cut correlation between AHI and morbidity.
• Polygraphy:
  - “best” alternative to polysomnography.
  - AHI > 1 on polygraphy predicts AHI > 1 on polysomnography with moderate sensitivity (88%) and specificity (71%).
Diagnosis

- Oximetry:
  - Widely used.
  - An abnormal oximetry (three or more clusters of desaturation events ≥ 4% and at least three desaturations to <90% - McGill criteria) may be used for the diagnosis and treatment of the most severe OSAS cases but the rate of false negative or inconclusive results is high.
  - The value in children with specific risk factors remains little studied.
Diagnosis

• Oximetry:

- A child with negative/inconclusive oximetry undergoing adenotonsillectomy for SDB has low risk of postoperative respiratory complications.
- Oxygen desaturation (≥4%) index > 2 in children with snoring may be a predictor of AHI > 1.
WHY IS AN OBJECTIVE DIAGNOSIS IMPORTANT?
Objective diagnosis

- OSA severity is positively associated with risk of morbidity:
  - Elevated blood pressure
  - Nocturnal enuresis
  - No clear association between OSA severity and behaviour problems, cognitive deficits or sleepiness.
  - Improvement in OSA severity does not explain improvement in neurobehavioral outcomes after adenotonsillectomy.
Objective diagnosis

- Moderate-to-severe OSA is less likely to resolve without treatment compared to mild OSA:

  - CHAT study
Objective diagnosis

- The frequency of respiratory complications in the immediate post-adenotonsillectomy period increases with OSA severity.

- The frequency of postoperative respiratory complications in school-aged children without comorbidities is low irrespective of the OSAS severity.
Objective diagnosis

• The likelihood of residual OSA after adenotonsillectomy increases in parallel with preoperative disease severity.

• Prioritize treatment
  - Prioritize for upper airway surgery
  - Start NIV before surgery
  - Better prepare for postoperative care
TREATMENT OF SEVERE OSA
Treatment

- Prioritize treatment:
  - Major craniofacial abnormalities
  - Neuromuscular disorders
  - Achondroplasia
  - Chiari malformation
  - Down syndrome
  - MPS
  - Prader-Willi syndrome

- Higher risk of pulmonary HT, positive effects on symptoms and QoL, higher rate of residual OSA
STEP 5. Indications for treatment of SDB:
5.1 a. AHI >5 episodes/h irrespective of the presence of morbidity
      b. Treatment may be beneficial if AHI 1-5 episodes/h especially in the presence: of i) morbidity from the cardiovascular system (see 2.1); ii) morbidity from the central nervous system (see 2.1); iii) enuresis; iv) somatic growth delay or growth failure; v) decreased quality of life; vi) risk factors for OSAS persistence (see 3.1)
      c. If at risk for OSAS and PSG or polygraphy not available, treatment is considered when positive oximetry or SDB questionnaires (see 4.4) or morbidity present
5.2 No evidence for treating primary snoring (evaluation yearly)
5.3 OSAS treatment is a priority if:
      a. Craniofacial abnormalities
      b. Neuromuscular disorders
      c. Achondroplasia
      d. Chiari malformation
      e. Down syndrome
      f. Mucopolysaccharidoses
      g. Prader-Willi syndrome

STEP 6. Stepwise treatment approach to SDB:
6.1 A stepwise treatment approach (from 6.2 to 6.9) is usually implemented until complete resolution of SDB
6.2 Weight loss if the child is overweight or obese
6.3 Nasal corticosteroids and/or montelukast po
6.4 Adenotonsillectomy
6.5 Unclear whether adenoidectomy or tonsillectomy alone are adequate
6.6 Rapid maxillary expansion or orthodontic appliances
6.7 Continuous positive airway pressure (CPAP) or noninvasive positive pressure ventilation (for nocturnal hypoventilation)
6.8 Craniofacial surgery
6.9 Tracheostomy
For details on indications, efficacy, adverse effects or complications of different treatment interventions see text

STEP 7. Recognition and management of persistent SDB:
7.1 a. Outcomes monitored after intervention (6 wks-12 months): symptoms, PSG, quality of life, cardiovascular or central nervous system morbidity, enuresis, growth rate
      b. If PSG not available: polygraphy, oximetry/capnography
      c. PSG ≥6 weeks after adenotonsillectomy (persistent SDB symptoms or at risk of persistent OSAS preoperatively); after 12 weeks of montelukast/nasal steroid
      d. PSG after 12 months of rapid maxillary expansion (earlier if symptoms persist) and after 6 months with an oral appliance
      e. PSG for titration of CPAP, NPPV and then annually; PSG as predictor of successful decannulation with tracheostomy
      f. Airway re-evaluation by nasopharyngoscopy, drug-induced sleep endoscopy, MRI
Treatment

- Individualize treatment:
  - Significant nocturnal gas exchange abnormalities
  - Significant comorbidities
  - Complex airway abnormalities
  - Careful planning and preparation – multidisciplinary team:
    - ENT surgery
    - Maxillofacial surgery
    - Pediatric Pulmonology
    - Anaesthesiology
    - PICU
Treatment

• Diagnosis

- Multidisciplinary discussion
- Investigations to further characterize OSA:
  • Cardiac ultrasound – pulmonary hypertension
  • ENT evaluation
  • Start NIV before possible surgery
Treatment

- **Treatment**
  - Identify the site(s) of obstruction
    - Drug induced sleep endoscopy
    - Rigid and/or flexible bronchoscopy
    - Imaging
  - Decision on further treatment
    - Surgery
    - Non-invasive ventilation
    - Tracheostomy
    - ...

- ...
Drug-induced sedation endoscopy in pediatric obstructive sleep apnea syndrome

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Fig. 2. Drug-induced sedation endoscopy findings for upper airway obstruction at the level of the adenoids, tonsils, and tongue base.

Fig. 3. Drug-induced sedation endoscopy findings for dynamic upper airway collapse at the level of the palate, epiglottis, presence of laryngomalacia and hypotonia.

- Increased success percentage of 91%
Treatment

Future research

Verhulst et al., ATS, 2010.
Treatment

- Different treatment modalities
  - Non-invasive to very invasive
  - Combination of treatments
  - Stepwise treatment plan
Treatment

- Weight loss
- Adenotonsillectomy
- Orthodontics
- Non-invasive ventilation
- Upper airway surgery: supraglottoplasty, tongue reduction surgery, etc.
- Craniofacial surgery
- Tracheostomy
Treatment

- High risk of residual OSA after treatment.
- Follow-up is critical.
- This includes repeated objective evaluation for OSA.
CONCLUSIONS
Conclusions

• Objective diagnosis of OSA is critical
  - Take the whole syndrome into account

• These patients should be treated in a multidisciplinary team especially children with underlying conditions

• Treatment should be individualized