



# **Initiation of CPAP: for which children, when and how?**

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# **INTRODUCTION**

# Obstructive sleep disordered breathing in 2- to 18-year-old children: diagnosis and management

Athanasios G. Kaditis<sup>1</sup>, Maria Luz Alonso Alvarez<sup>2</sup>, An Boudewyns<sup>3</sup>, Emmanouel I. Alexopoulos<sup>4</sup>, Refika Ersu<sup>5</sup>, Koen Joosten<sup>6</sup>, Helena Larramona<sup>7</sup>, Silvia Miano<sup>8</sup>, Indra Narang<sup>9</sup>, Ha Trang<sup>10</sup>, Marina Tsaoussoglou<sup>1</sup>, Nele Vandenbussche<sup>11</sup>, Maria Pia Villa<sup>12</sup>, Dick Van Waardenburg<sup>13</sup>, Silke Weber<sup>14</sup> and Stijn Verhulst<sup>15</sup>

ERS Statement on obstructive sleep-disordered breathing  
in 1- to 23-month-old children

## Adenotonsillectomy to Treat Obstructive Sleep Apnea: Is It Enough?

A. Boudewyns, MD, PhD,<sup>1\*</sup> F. Abel, FRCPCH, MD,<sup>2</sup> E. Alexopoulos, MD,<sup>3</sup>  
M. Evangelisti, MD,<sup>4,5</sup> A. Kaditis, MD,<sup>6</sup> S. Miano, MD,<sup>7</sup> M.P. Villa, MD, PhD,<sup>4,5</sup>  
and S.L. Verhulst, MD, PhD<sup>8</sup>

**Step 1:** Recognize the child at risk for obstructive SDB

**Step 2:** Identify SDB-related morbidity or conditions co-existing with SDB (probably common pathogenesis)

**Step 3:** Recognize factors predicting persistence of SDB

**Step 4:** Assess severity of SDB objectively

**Step 5:** Determine indications for treatment

**Step 6:** Stepwise treatment approach for SDB

**Step 7:** Follow-up, diagnosis and management of persistent SDB



# **COMPLEX OSA**

# The prevalence of obstructive sleep apnea in symptomatic patients with syndromic craniosynostosis☆

G. Inverso, K.A. Brustowicz, E. Katz, B.L. Padwa: The prevalence of obstructive sleep apnea in symptomatic patients with syndromic craniosynostosis. *Int. J. Oral Maxillofac. Surg.* 2016; 45: 167–169. © 2015 International Association of Oral and Maxillofacial Surgeons. Published by Elsevier Ltd. All rights reserved.

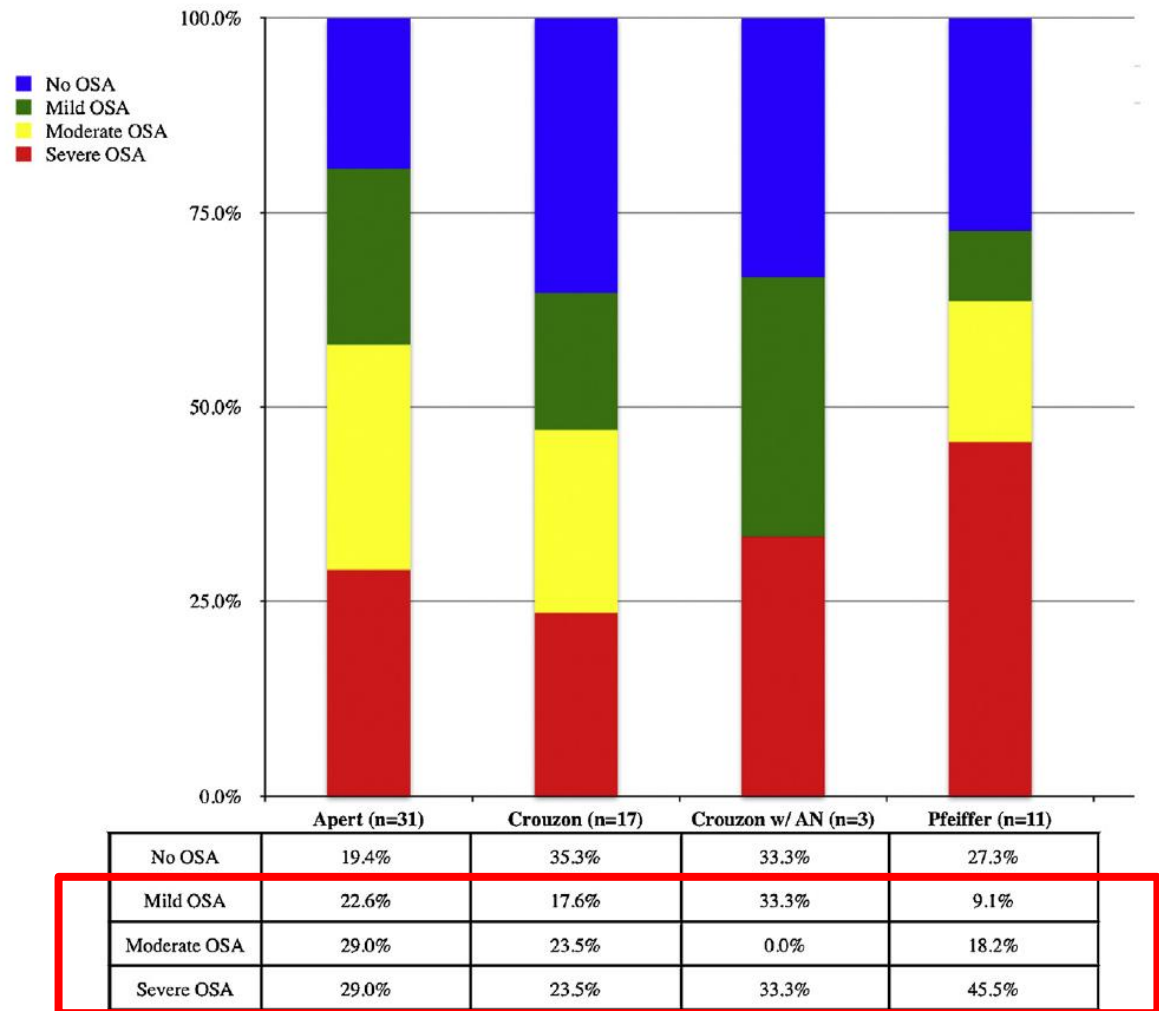


Fig. 1. Prevalence and severity of OSA by phenotypic diagnosis.

# Prader Willi Syndrome and Obstructive Sleep Apnea: Co-occurrence in the Pediatric Population

Karim Sedky, M.D., M.Sc.<sup>1</sup>; David S. Bennett, Ph.D.<sup>2</sup>; Andres Pumariega, M.D.<sup>1</sup>

*J Clin Sleep Med* 2014;10(4):403-409.

- Overall OSAS prevalence: 80% (most patients without symptoms)
- Mild OSAS (apnea-hypopnea index-AHI >1 to <5 episodes/h): 53.1%
- Moderate OSAS (AHI 5-10 episodes/h): 22.3%
- Severe OSAS (>10 episodes/h): 24.6%

# Prevalence of Obstructive Sleep Apnea in Children with Down Syndrome

Mieke Maris, MD<sup>1</sup>; Stijn Verhulst, MD, PhD<sup>2</sup>; Marek Wojciechowski, MD<sup>2</sup>; Paul Van de Heyning, MD, PhD<sup>1</sup>; An Boudewyns, MD, PhD<sup>1</sup>

*SLEEP, Vol. 39, No. 3, 2016*

- 122 children with DS underwent PSG.
- Overall prevalence: 66%
  - Majority had severe OSA
  - Younger age was associated with more severe OSA
- Prevalence in children + symptoms: 76%
- Prevalence in children – symptoms: 54%



# Obstructive Sleep Apnea in Young Infants with Down Syndrome Evaluated in a Down Syndrome Specialty Clinic

AMERICAN JOURNAL OF  
medical genetics

Aida Goffinski,<sup>1</sup> Maria A. Stanley,<sup>2\*\*</sup> Nicole Shepherd,<sup>3</sup> Nichole Duvall,<sup>3</sup> Sandra B. Jenkinson,<sup>3</sup> Charlene Davis,<sup>4</sup> Marilyn J. Bull,<sup>4</sup> and Randall J. Roper<sup>3\*</sup>

	All infants (n = 59) <sup>a</sup>
Average age at diagnosis (days) <sup>b</sup>	44 ± 48
Total sleep time (minutes)	150 ± 69
End-tidal CO <sub>2</sub> mean (mm Hg)	44 ± 8
Mean Apnea Hypopnea Index	19 ± 14
OSA <sup>c</sup> Criteria Not Met	3
Mild OSA (AHI 2–5)	7
Moderate OSA (AHI 6–10)	9
Severe OSA (AHI ≥10)	40 <sup>a</sup>



# Complex OSA

- Children with complex OSA:
  - High prevalence of severe OSA
  - Increased risk of complications
  - Limited coping abilities.

Child with  
complex OSA

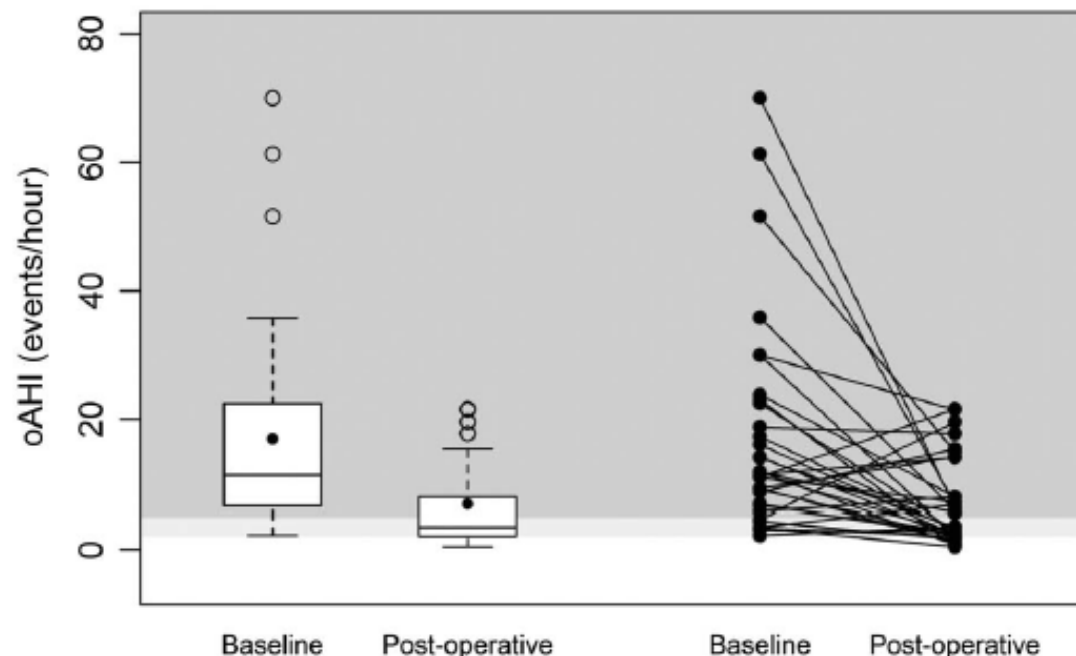
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graph TD; A[Child with complex OSA] --> B[Adenotonsillectomy]; B --> C[Routine PSG for residual OSA];
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Adenotonsillectomy

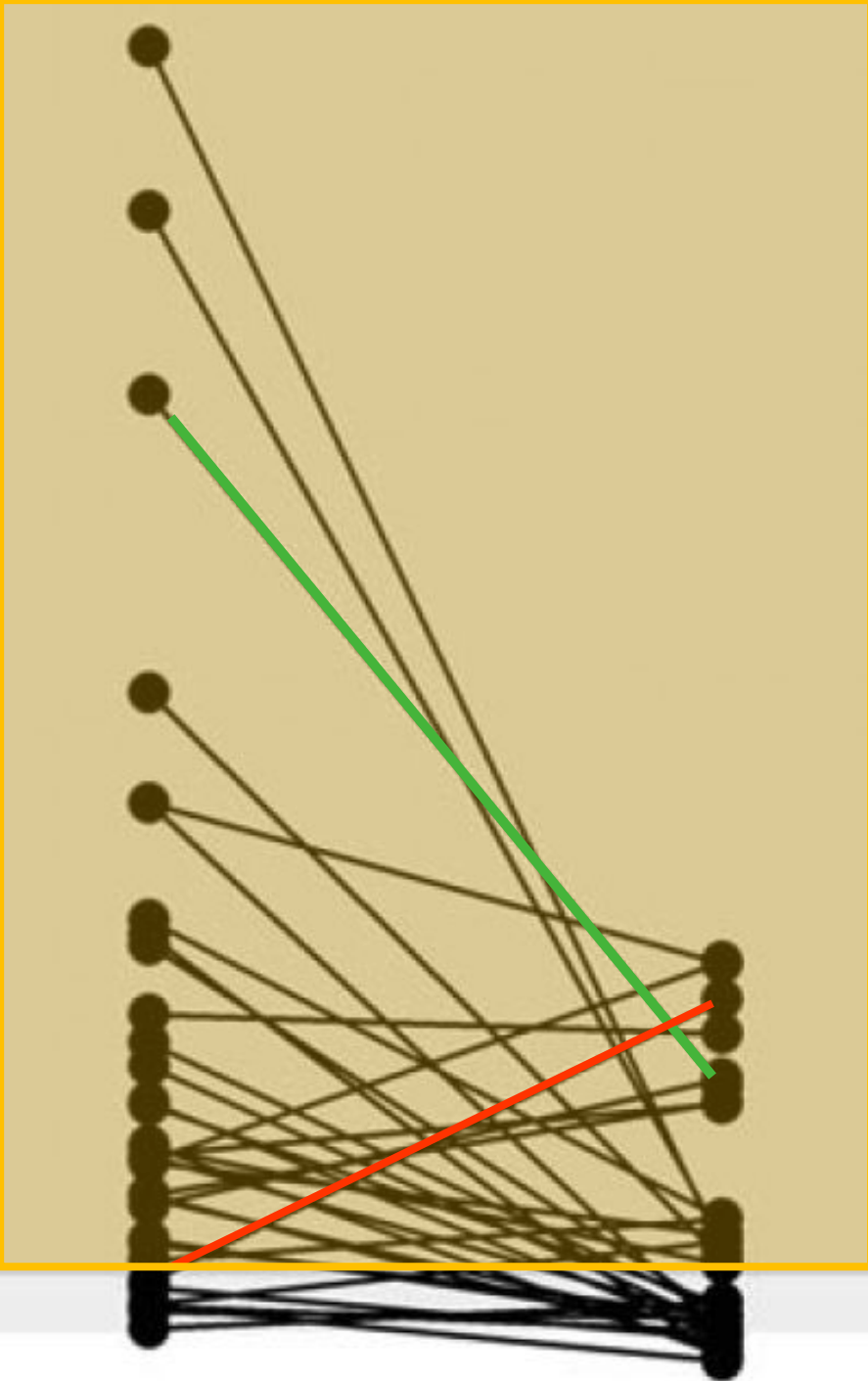
Routine PSG for  
residual OSA

# Outcome of adenotonsillectomy in children with Down syndrome and obstructive sleep apnoea

Mieke Maris,<sup>1</sup> Stijn Verhulst,<sup>2</sup> Marek Wojciechowski,<sup>2</sup> Paul Van de Heyning,<sup>1</sup> An Boudewyns<sup>1</sup>



**Figure 1** Pre- and postoperative obstructive apnoea-hypopnoea index (oAHI) values in 34 patients with Down syndrome. Left-hand plot shows median values with upper and lower quartile. Right hand plot shows individual values.



- *High prevalence of residual OSA after AT.*
- Responders
- Non-responders
- Increased risk of post-operative complications



# Complex OSA

- The indication for surgery should be set correctly.
- Individualize treatment = identify markers that could identify the success of a treatment before the actual intervention



# Complex OSA

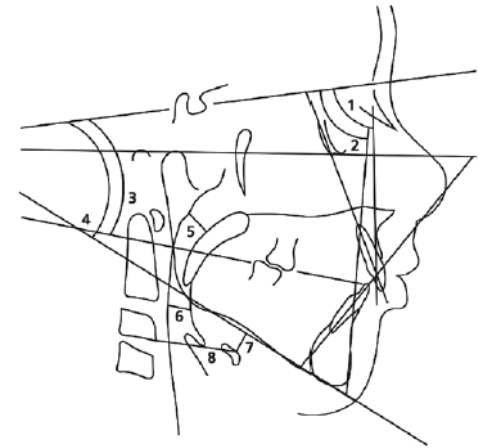
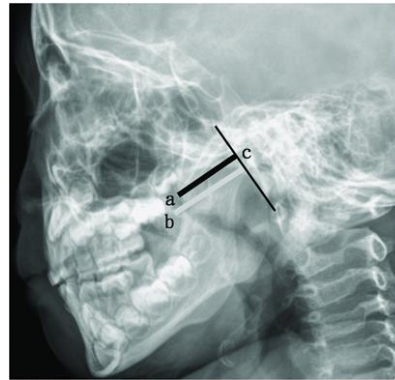
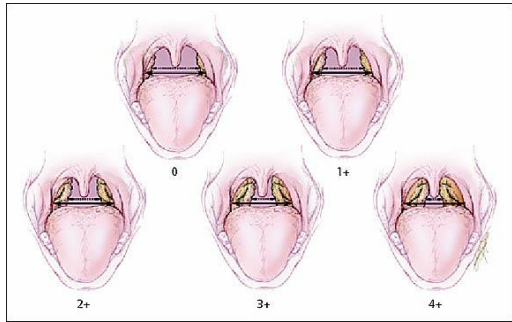
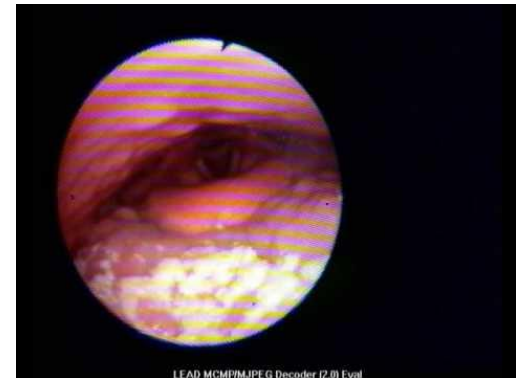
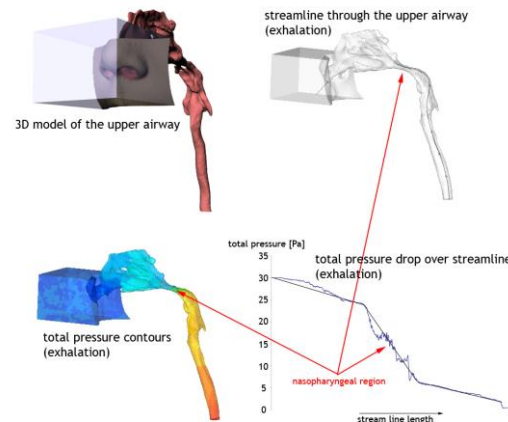
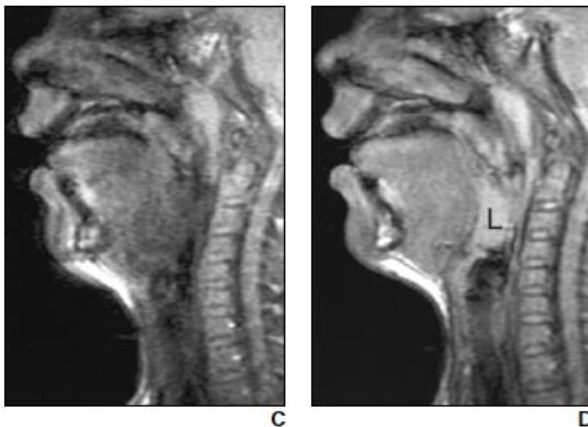


Figure 1—Anatomic drawing, linear measurements, and angles traced for the determination of the cephalometric variables: 1, SNA; 2, SNB; 3, NSPIO; 4, NSGoGn; 5, SPAS; 6, PAS; 7, MPH; 8, C<sub>3</sub>H.



# Drug-induced sedation endoscopy in surgically naive children with Down syndrome and obstructive sleep apnea

Mieke Maris <sup>a</sup>, Stijn Verhulst <sup>b</sup>, Vera Saldien <sup>c</sup>, Paul Van de Heyning <sup>a</sup>, Marek Wojciechowski <sup>b</sup>, An Boudewyns <sup>a,\*</sup>

surgery. Seven patients were non-surgically treated, and three received a combined treatment. A multilevel collapse was present in 85.4%. Tongue base obstruction was present in ten patients (24.4%) and epiglottic collapse in 48.8%. Pre- and postoperative PSG data were available for 25 children (adenotonsillectomy, n = 16; tonsillectomy, n = 7; adenoidectomy, n = 2). A significant improvement in oAHI from 11.4/h (range, 7.7–27.0) to 5.5/h (range, 2.1–7.6) was found. Persistent OSA was present in 52% of the children. No significant association between different DISE findings and persistent OSA could be found.

*Conclusion:* Most patients with Down syndrome and OSA present with multilevel collapse on DISE. Adenotonsillectomy results in a significant improvement of the oAHI; however more than half of the patients had persistent OSA, probably due to multilevel collapse. Upper airway evaluation may provide more insights into the pattern of UA obstruction in patients with persistent OSA.



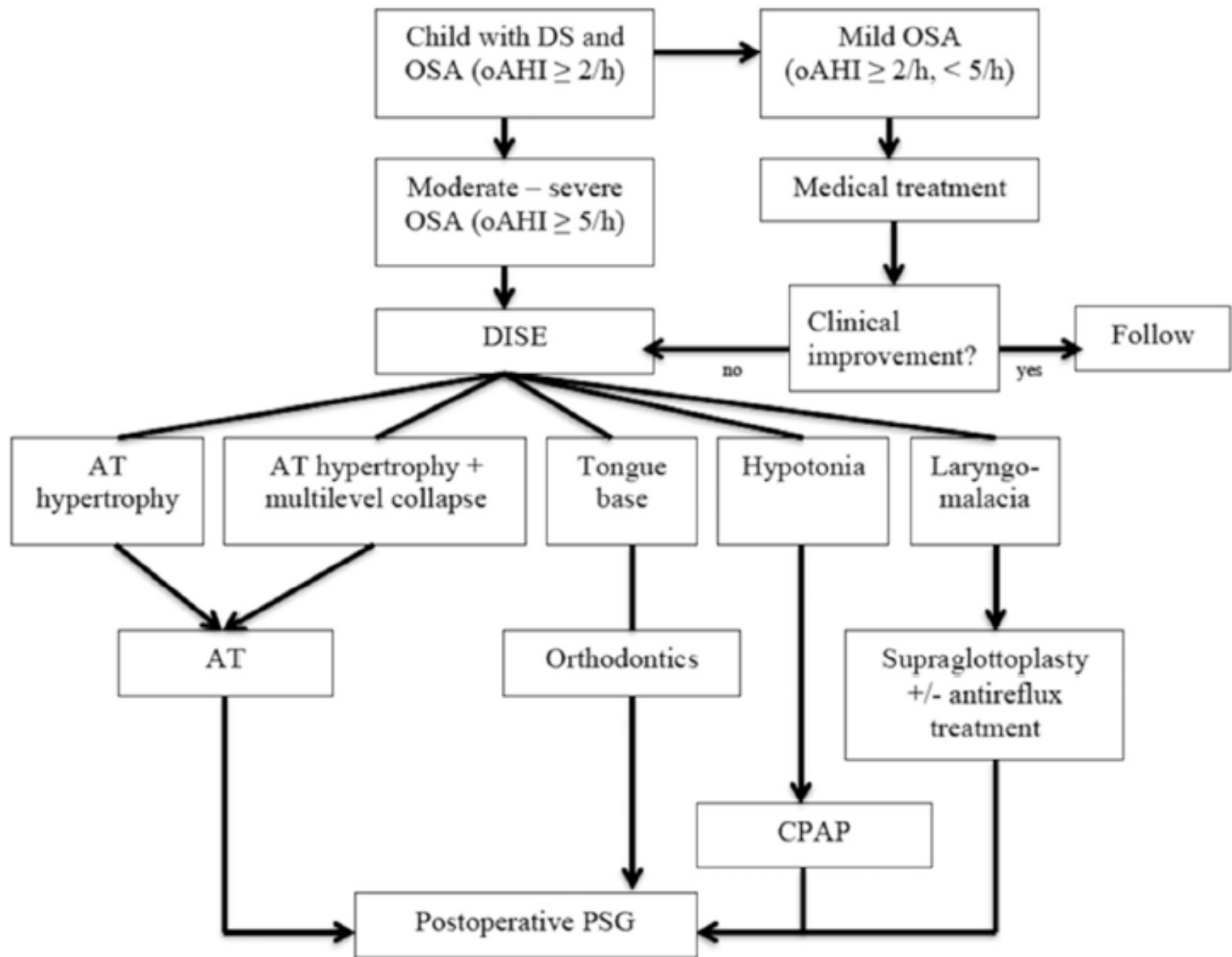


Fig. 1. Overview of the treatment decision-making process.



# **CPAP IN THE ERS STATEMENT**



## Topic 5: stepwise treatment approach for OSAS in young children

### 5.1. What is the hierarchy of treatment interventions for OSAS in young children?

#### *Evidence summary*

- a) Interventions for OSAS in young children are individualised according to aetiology, severity and morbidity.
- b) Nasopharyngoscopy or drug-induced sleep endoscopy may be used to determine the type and sequence of treatment interventions.
- c) In infants with OSAS due to multiple causes, surgical treatment is overall more effective in reducing symptoms than oxygen administration, use of antireflux medications or continuous positive airway pressure (CPAP) application. CPAP or non-invasive positive pressure ventilation (NPPV), tracheostomy and supraglottoplasty are equally effective in reducing the AHI.

## Obstructive Sleep Apnea in Infancy: A 7-Year Experience at a Pediatric Sleep Center

Sriram Ramgopal, MD,<sup>1</sup> Sanjeev V. Kothare, MD,<sup>1</sup> Mandeep Rana, MD,<sup>1</sup> Kanwaljit Singh, MD, MPH,<sup>2</sup>  
and Umakanth Khatwa, MD<sup>3\*</sup>

In comparing the types of interventions with each other, we found that 67% percent (16/24) patients who received medical management, 40% (10/25) patients who received O<sub>2</sub> or CPAP treatment, and 76% (19/25) who received surgical management had symptom resolution. Also, 30% (7/16) patients who received no intervention had symptom resolution. The odds of symptom resolution increased if a patient had surgical management as compared to O<sub>2</sub>/CPAP therapy (OR = 4.75, *P* = 0.01), but the odds of symptom resolution did not differ significantly between medical management and CPAP/O<sub>2</sub> (*P* > 0.05).

# Evaluating the Management of Obstructive Sleep Apnea in Neonates and Infants

*Rachel L. Leonardis, BS; Jacob G. Robison, MD, PhD; Todd D. Otteson, MD, MPH*

**Conclusions:** Anti-gastroesophageal reflux disease treatment is the most common intervention in each age group. Although adenoidectomy is the most common surgical intervention overall, the prevalence increases with age. Supraglottoplasty is the most common surgical intervention in neonates and infants aged 0 to 3 months and offers the greatest objective improvement in this age group. Overall, the use of CPAP/BiPAP is associated with the greatest objective improvement.

# ERS statement on obstructive sleep disordered breathing in 1- to 23-month-old children

## *5.5. What are the efficacy and risks of CPAP or NPPV for OSAS in young children?*

### *Evidence summary*

- a) In children aged <24 months with moderate-to-severe OSAS who are not candidates for or do not improve after adenotonsillectomy or other surgical interventions: i) CPAP initiated at 4–6 cmH<sub>2</sub>O and titrated up to 10 cmH<sub>2</sub>O is an effective and well-tolerated treatment; ii) it can be applied as a temporary intervention while waiting for craniofacial surgery; iii) NPPV has been used in cases of OSAS coexisting with hypoventilation (*e.g.* spinal muscular atrophy type 1).
- b) Most studies report minor complications. Occurrence of midface flattening has been described, but long-term consequences and potential reversibility of this finding are unknown.
- c) A nasal mask is the most common interface for CPAP or NPPV; an oronasal mask is used for uncontrollable mouth leak or severe nasal obstruction.

# The use of nasal continuous positive airway pressure to treat obstructive sleep apnoea

F Massa, S Gonzalez, A Lavery, C Wallis, R Lane

Arch Dis Child 2002;87:438-443

**Table 2** Comparison between patients that accepted and patients that refused nCPAP therapy

	nCPAP accepted (n=42)	nCPAP refused (n=22)
Age at diagnostic sleep study	5.3 (5.2)	4.3 (5.7)
Age at nCPAP trial	5.9 (5.1)	5.2 (5.5)
Sex	62% male 38% female	54.5% male 45.5% female
<i>Diagnoses</i>		
Congenital malformations of the upper airway	21 (50%)	12 (54.5%)
Conditions predisposing to OSAS	10 (23.8%)	9 (41%)
Lower airway structural abnormalities	3 (7.2%)	0
OSAS	4 (9.5%)	0
Associated medical conditions	4 (9.5%)	1 (4.5%)

Data are expressed as mean (SD) or n (%). The sum of patients is 64 because two patients in which nCPAP was not effective were excluded from this comparison. No statistically significant differences between the groups were found for any of the variables.  
OSAS, obstructive sleep apnoea syndrome.

# ERS statement on obstructive sleep disordered breathing in 1- to 23-month-old children

## *5.8. What is the management of OSAS in young children with complex conditions?*

### *Evidence summary*

In children with OSAS secondary to complex conditions a combination of interventions may be required.

- a) Achondroplasia: OSAS may improve after adenotonsillectomy, but nasopharyngeal airway or CPAP are necessary if upper airway obstruction persists post-operatively.
- b) Chiari malformation: adenoidectomy and/or tonsillectomy, CPAP or NPPV may be required to treat OSAS, central sleep apnoea and nocturnal hypoventilation; surgical decompression of the cervicomedullary junction is accompanied by decrease in the frequency of central apnoeas.
- c) Down syndrome: adenoidectomy or adenotonsillectomy for adenoidal or tonsillar hypertrophy and supraglottoplasty for laryngomalacia have been reported. CPAP has been applied for persistent OSAS post-operatively or as first-line treatment in the absence of adenotonsillar hypertrophy. Some infants with Down syndrome outgrow OSAS within several months.
- d) Mucopolysaccharidoses: adenotonsillectomy and CPAP are used to relieve upper airway obstruction, while enzyme replacement and haemopoietic stem cell transplantation target the metabolic disorder.
- e) Prader–Willi syndrome: adenotonsillectomy for OSAS; oxygen therapy for central sleep apnoeas which decrease in frequency with age.



# Obstructive sleep disordered breathing in 2- to 18-year-old children: diagnosis and management

Athanasios G. Kaditis<sup>1</sup>, Maria Luz Alonso Alvarez<sup>2</sup>, An Boudewyns<sup>3</sup>, Emmanouel I. Alexopoulos<sup>4</sup>, Refika Ersu<sup>5</sup>, Koen Joosten<sup>6</sup>, Helena Larramona<sup>7</sup>, Silvia Miano<sup>8</sup>, Indra Narang<sup>9</sup>, Ha Trang<sup>10</sup>, Marina Tsaoussoglou<sup>1</sup>, Nele Vandebussche<sup>11</sup>, Maria Pia Villa<sup>12</sup>, Dick Van Waardenburg<sup>13</sup>, Silke Weber<sup>14</sup> and Stijn Verhulst<sup>15</sup>

## 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 *p.o.*
- 6.4 Adenotonsillectomy
- 6.5 Unclear whether adenoidectomy or tonsillectomy alone are adequate
- 6.6 Rapid maxillary expansion or orthodontic appliances
- 6.7 CPAP or NPPV (for nocturnal hypoventilation)
- 6.8 Craniofacial surgery
- 6.9 Tracheostomy

## STEP 7: Recognition and management of persistent SDB:

### 7.1

- a) Outcomes monitored after intervention (6 weeks-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  $\geq$ 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

# Obstructive sleep disordered breathing in 2- to 18-year-old children: diagnosis and management

*6.7. What are the indications, efficacy and potential complications of CPAP or NPPV in children with obstructive SDB?*

*Summary*

- a) Usual indications for CPAP are: residual OSAS after adenotonsillectomy ( $AHI >5$  episodes·h<sup>-1</sup>) and OSAS related to obesity, craniofacial abnormalities or neuromuscular disorders. If nocturnal hypoventilation occurs (*e.g.* end-tidal carbon dioxide tension ( $PCO_2$ )  $>50$  mmHg for  $>25\%$  of total sleep time or peak end-tidal  $PCO_2 \geq 55$  mmHg) NPPV is preferred.
- b) Positive airway pressure ventilation is accompanied by improvements in gas exchange, attention deficits, sleepiness, behaviour and quality of life.
- c) Complications of CPAP and NPPV include: nasal congestion, rhinorrhoea, epistaxis, facial skin erythema related to the mask and midface retrusion.
- d) Adherence to CPAP or NPPV should be monitored using the device software. Management of complications and behavioural modification may improve patient adherence.

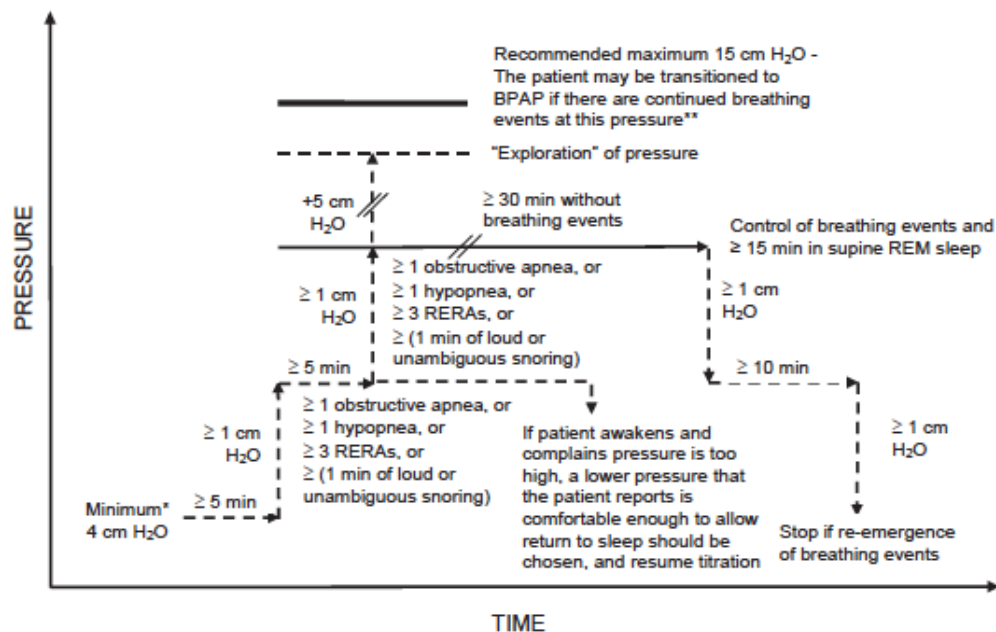


# **CPAP TITRATION**

# Clinical Guidelines for the Manual Titration of Positive Airway Pressure in Patients with Obstructive Sleep Apnea

Positive Airway Pressure Titration Task Force of the American Academy of Sleep Medicine

Task Force Members: Clete A. Kushida, M.D., Ph.D., RPSGT (Chair)<sup>1</sup>; Alejandro Chediak, M.D. (Vice-Chair)<sup>2</sup>; Richard B. Berry, M.D.<sup>3</sup>; Lee K. Brown, M.D.<sup>4</sup>; David Gozal, M.D.<sup>5</sup>; Conrad Iber, M.D.<sup>6</sup>; Sairam Parthasarathy, M.D.<sup>7</sup>; Stuart F. Quan, M.D.<sup>8</sup>; James A. Rowley, M.D.<sup>9</sup>



**Figure 1**—CPAP Titration Algorithm for Patients <12 years During Full- or Split-Night Titration Studies. Note: Upward titration at  $\geq 1$ -cm increments over  $\geq 5$ -min periods is continued according to the breathing events observed until  $\geq 30$  min without breathing events is achieved.

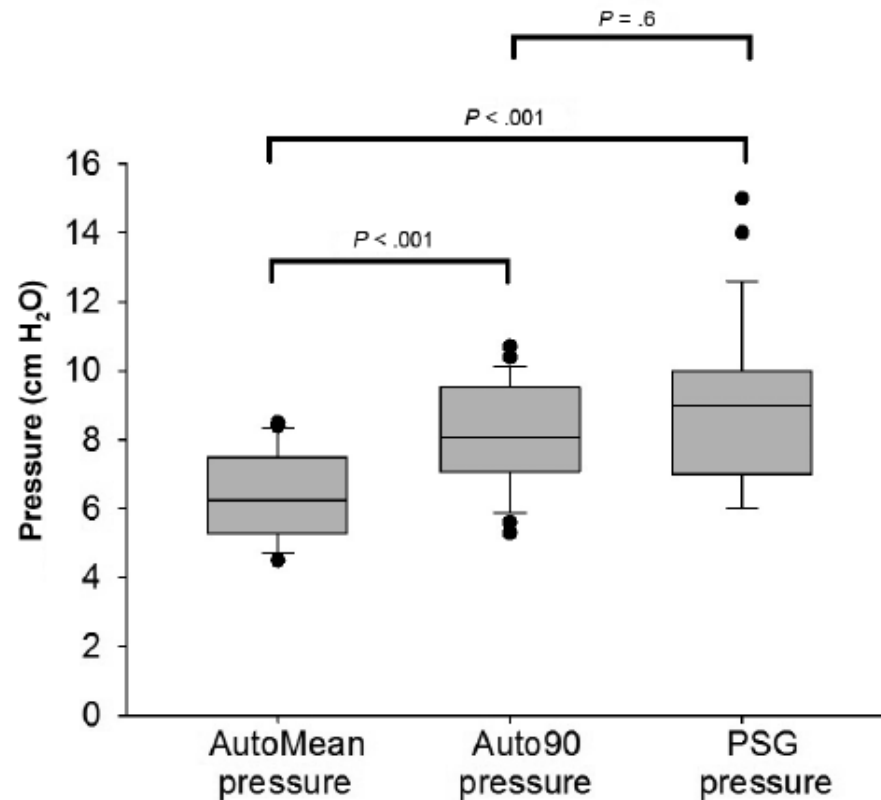
\* A higher starting CPAP may be selected for patients with an elevated BMI and for retitration studies

\*\* The patient should also be tried on BPAP if the patient is uncomfortable or intolerant of high CPAP

# Autotitrating CPAP as a Tool for CPAP Initiation for Children




Rebecca Mihai, BAppSc (Hons)<sup>1</sup>; Moya Vandeleur, MB BCH, BAO<sup>1</sup>; Sally Pecoraro<sup>1</sup>; Margot J. Davey, MBBS<sup>1,3</sup>; Gillian M. Nixon, MD<sup>1,2,3</sup>

Figure 2—AutoMean, Auto90, and PSG pressure for 26 patients.



AutoMean pressure = mean autoPAP pressure, Auto90 pressure = average device pressure  $\leq$  90% of time, PSG pressure = pressure determined by attended polysomnography.

# Outpatient initiation of long-term continuous positive airway pressure in children

Alessandro Amaddeo MD, PhD<sup>1,2,3</sup>  | Annick Frapin MSN<sup>1</sup> | Samira Touil BSc<sup>1</sup> |  
Sonia Khirani PhD<sup>1,4</sup>  | Lucie Griffon MD<sup>1</sup> | Brigitte Fauroux MD, PhD<sup>1,2,3</sup> 

**TABLE 2** Continuous positive airway pressure (CPAP) compliance and equipment (n = 27)

Duration of follow-up, months (median, range)	12.3 (2.2-25.2)
Objective CPAP compliance over the last month (n = 27)	
Average use per night, h:min (median, range)	08:21 (05:45- 12:20)
Percentage of nights with CPAP use >4 h, % (mean ± standard deviation)	83 ± 17
Average nights use per month, nights (median, range)	25 (18-30)
Constant CPAP pressure, cmH <sub>2</sub> O (mean ± SD) (n = 15)	8.5 ± 1.0
Interface (n = 27)	
Nasal mask	22
Facial mask	3
Nasal prongs	2



# **CONCLUSIONS**



# Conclusions

- CPAP is mainly used in complex OSAS.
- The etiology of OSAS in the individual child should be determined.
- Role of CPAP:
  - More prominent role in infants according to the etiology
  - Second or third-line treatment in older children
- CPAP titration protocol – outpatient initiation.