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Lucile Packard Children's Hospital Stanford

What's on the Horizon for Sleep and Down Syndrome

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What are some emerging treatment options?

- More access with Telemedicine
- Custom Fitting PAP masks
- High Flow Nasal Cannula
- Drug Induced Sleep Endoscopy directed surgeries
- Hypoglossal Nerve Stimulator
- Pharmacologic Treatments

Trisomy 21

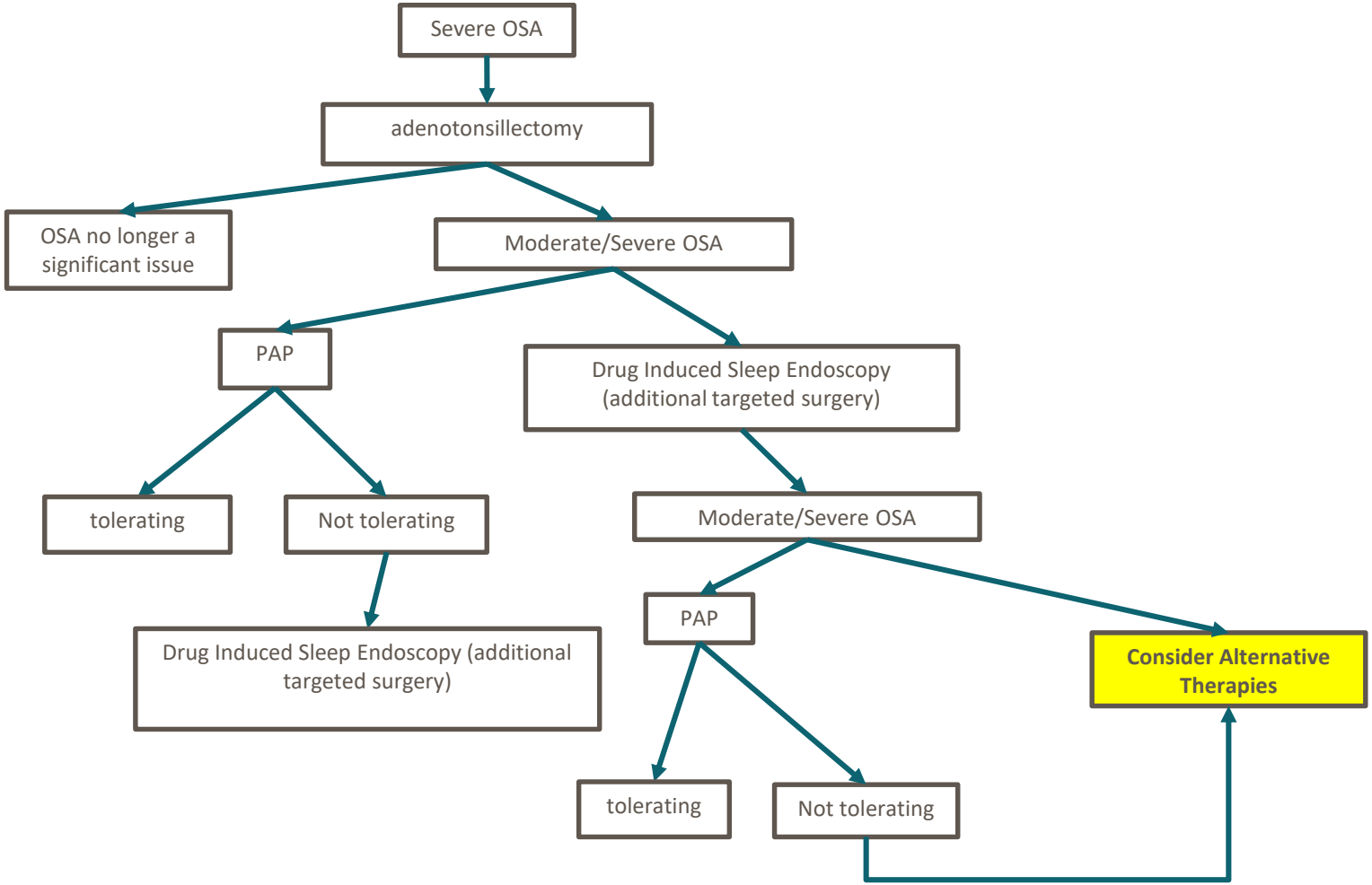
- Most common chromosomal disorder in children
- Up to 60% of children with Down Syndrome have OSA – and it tends to be severe
- 50-80% have residual disease after adenotonsillectomy

Telemedicine in Pediatric Sleep

Shalini Paruthi, MD[a,b,*]

Paruthi S. Telemedicine in Pediatric Sleep. Sleep Med Clin 2020;15(3S):e1–e7.

- Doctors should try synchronous or asynchronous modalities of telemedicine (phone, video, websites, audio recordings, social media, etc..)
- Use televisits when possible
- In person visits still important in evaluation of obstructive sleep apnea or CPAP mask fitting



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(a) (b)

Figure 1. Nasal Pillows and Nasal Cradle Mask Examples (**a**) Nasal pillows PAP mask, sealing the nares. (**b**) Nasal cradle mask, sealing under the nares.

(a) (b)

Figure 2. Nasal and Full Face Mask Examples (**a**) Nasal PAP mask, over the nose. (**b**) Full Face PAP mask covers nose and mouth.

- Nasal pillows
- Nasal cradle
- Nasal mask
- Full face mask
- Total Face Mask

CrossMark

ORIGINAL ARTICLE
SLEEP

Feasibility of three-dimensional facial imaging and printing for producing customised nasal masks for continuous positive airway pressure

Kelvin Duong[1], Joel Glover[2], Alexander C. Perry[3], Deborah Olmstead[4], Mark Ungrin[5], Pina Colarusso[2], Joanna E. MacLean[6] and Andrew R. Martin[1]

Original Article

Institution of MECHANICAL ENGINEERS

Journal of ENGINEERING IN MEDICINE

Proc IMechE Part H:
J Engineering in Medicine
2021, Vol. 235(1) 44–53

DOI: 10.1177/0954411920959879
journals.sagepub.com/home/pih
SAGE

Development of a smart-fit system for CPAP interface selection

Zhichao Ma, Philip Hyde, Michael Drinnan and Javier Munguia

- Using 3D face scans and engineering technology, customized masks are more feasible
- Consider exploring how to obtain a customized mask if needed

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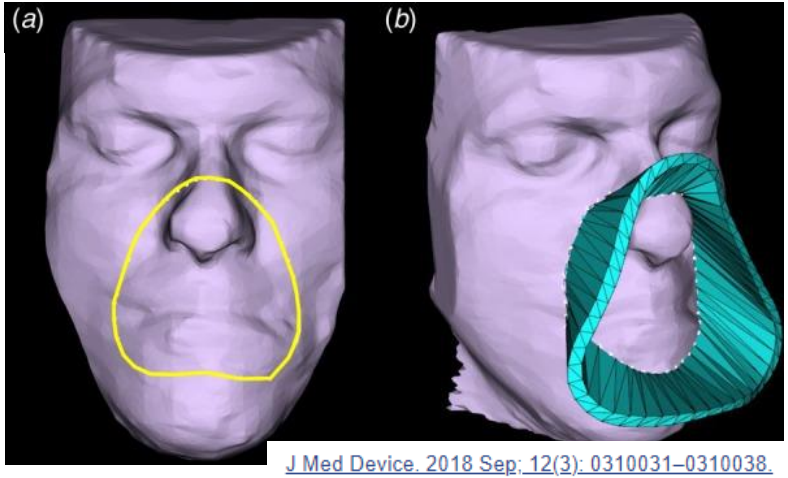
Custom-Fit Three-Dimensional-Printed BiPAP Mask to Improve Compliance in Patients Requiring Long-Term Noninvasive Ventilatory Support

Noninvasive ventilator support using bi-level positive airway pressure/continuous positive airway pressure (BiPAP/CPAP) is commonly utilized for chronic medical conditions like sleep apnea and neuromuscular disorders like amyotrophic lateral sclerosis (ALS) that lead to weakness of respiratory muscles. Generic masks come in standard sizes and are often perceived by patients as being uncomfortable, ill-fitting, and leaky. A significant number of patients are unable to tolerate the masks and eventually stop using their devices. The goal of this project is to develop custom-fit masks to increase comfort, decrease air leakage, and thereby improve patient compliance. A single-patient case study of a patient with variant ALS was performed to evaluate the custom-fit masks. His high nose bridge and overbite of lower jaw caused poor fit with generic masks, and he was noncompliant with his machine. Using desktop Stereolithography three-dimensional (3D) printing and magnetic resonance imaging (MRI) data, a generic mask was extended with a rigid interface such that it was complementary to the patient's unique facial contours. Patient or clinicians interactively select a desired mask shape using a newly developed computer program. Subsequently, a compliant silicone layer was applied to the rigid interface. Ten different custom-fit mask designs were made using computer-aided design software. Patient evaluated the comfort, extent of leakage, and satisfaction of each mask via a questionnaire. All custom-fit masks were rated higher than the standard mask except for two. Our results suggest that modifying generic masks with a 3D-printed custom-fit interface is a promising strategy to improve compliance with BiPAP/CPAP machines. [DOI: 10.1115/1.4040187]



J Med Device. 2018 Sep; 12(3): 0310031–0310038.
Published online 2018 Jul 13. doi: 10.1115/1.4040187

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High Flow Nasal Cannula?

ERS EUROPEAN RESPIRATORY SOCIETY
every breath counts

High flow nasal cannula for children with severe obstructive sleep apnea not compliant with continuous positive airway pressure

Alessandro Amaddeo, Sonia Khirani, Annick Frapin, Theo Teng, Lucie Griffon, Brigitte Fauroux
ERJ Open Research 2019 5: P132; **DOI:** 10.1183/23120541.sleepandbreathing-2019.P132

- Study: 8 patients (mean age 9yo, mean AHI 33±22)
- Results: after 1 month, 62% slept with HFNC 4+hrs, mean AHI 2±2
- But not tolerated in 3 older DS patients
- Conclusion: HFNC may be used as a rescue therapy for children not compliant with CPAP. Further studies are needed

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DOI: 10.1002/ppul.25009

ORIGINAL ARTICLE: SLEEP & BREATHING

WILEY

High flow nasal cannula treatment for obstructive sleep apnea in infants and young children

- Retrospective review of treatment with heated humidified high flow nasal cannula (HFNC) as treatment for OSA in young children intolerant of PAP, or at high risk for midface hypoplasia
- 22 children with OSA (AHI 4.8-89.2 events/hr),
- 19 patients received HFNC at home
  - by 12 months, 5 discontinued due to intolerance, 3 had resolved OSA
  - Common complications included: skin irritation, dry mucous membranes, increased central apneas
- Can serve as a bridge to surgery or spontaneous resolution of OSA

Ignatiuk, Daniel, Britta Schaer, and Brian McGinley. 2020. “High Flow Nasal Cannula Treatment for Obstructive Sleep Apnea in Infants and Young Children.” *Pediatric Pulmonology* 55 (10): 2791–98.

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ORIGINAL ARTICLE: SLEEP & BREATHING

WILEY

High flow nasal cannula treatment for obstructive sleep apnea in infants and young children

- HFNC appears to be an effective treatment for OSA with increased tolerance and less risk for mid face hypoplasia.
- Prospective needed:
  - Explore ventilation support
  - Compare to CPAP
  - Understand/address adherence issues

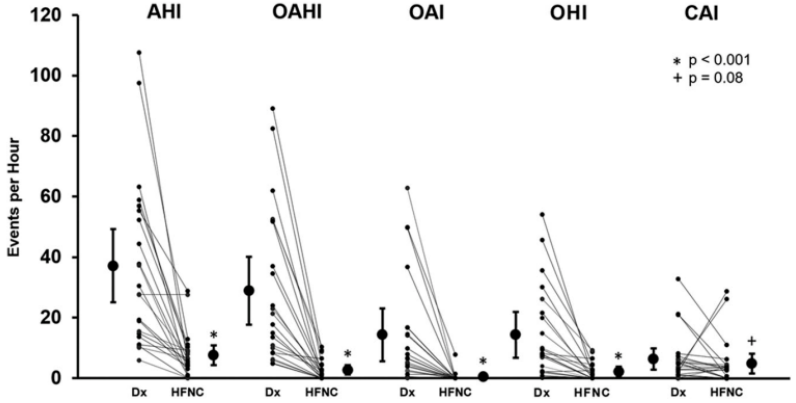

FIGURE 3 Comparison of apnea and hypopnea indices from diagnostic and HFNC titration sleep studies. Data presented are means [95% CI]. 95% CI, 95% confidence interval; AHI, apnea-hypopnea indices; CAI, central apnea index; Dx, diagnostic sleep study; HFNC, high flow nasal cannula titration study; OAHI, obstructive AHI; OAI, obstructive apnea index; OHI, obstructive hypopnea index

Ignatiuk, Daniel, Britta Schaer, and Brian McGinley. 2020. "High Flow Nasal Cannula Treatment for Obstructive Sleep Apnea in Infants and Young Children." *Pediatric Pulmonology* 55 (10): 2791–98.

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Taylor & Francis
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RESEARCH ARTICLE

Drug-induced sleep endoscopy directed surgery improves polysomnography measures in overweight and obese children with obstructive sleep apnea

- Retrospective analysis of PSG parameters following DISE-directed surgery in obese and overweight pediatric patients.
- 40 children (mean BMI 94% percentile, mean age 8).
  17 children had undergone previous T&A,
  Included children with comorbidities (Trisomy 21, developmental delay, etc…)
- Interventions performed included: T&A, supraglottoplasty, lingual tonsillectomy or a combination. *2 patients (prior T&A) did not have additional intervention*

- **DISE-directed interventions resulted in statistically and clinically significant improvements in PSG parameters. Improvement in oAHI, oxygen nadir, ODI noted.**

Research

JAMA Otolaryngology–Head & Neck Surgery | Original Investigation

Hypoglossal Nerve Stimulation in Adolescents With Down Syndrome and Obstructive Sleep Apnea

Gillian R. Diercks, MD, MPH; Carissa Wentland, DO; Donald Kearny, MD, MPH; Thomas Bernard Kinane, MD; Brian Skotko, MD, MPP; Vanessa de Guzman, MS; Ellen Grealish, RPSGT, REEGT; John Dobrowski, MD; Ryan Soose, MD; Christopher J. Hartnick, MD

- JAMA case series of 6 adolescents (12-18yo) with DS and severe OSA (AHI >10), after T&A.
- Device: Inspire
- Measures: AHI, QOL measures
- Conclusion: good tolerance, 56-85% reduction in AHI, clinical improvement, improved QOL

The Laryngoscope

Update on Hypoglossal Nerve Stimulation in Children With Down Syndrome and Obstructive Sleep Apnea

- Hypoglossal Nerve Stimulator (HGN) stimulation
- Case series of 20 children with Down Syndrome, ages 10-21 yo
- All participants had severe OSA (AHI >10, <50/hr) who failed CPAP therapy. All s/p DISE to confirm candidacy
- Patients underwent PSG HGN titration with reduction of AHI of 85% (though AHI was not fully normalized)
- Median nightly usage 9.21 hr/night. Caregivers reported improvement in QofL (OSA-18)
- 2 patients had interval adverse events requiring revision surgery that resolved problem
- 2 other patients underwent device interrogation to optimize tongue protrusion during DISE

- **HGN still appears to be a reasonable option for patients with severe OSA, at risk for severe sequalae from untreated OSA, who are intolerance of PAP therapy**
- Further study continues

Novel Medications for OSA

Combination medication Atomoxetine (a norepinephrine reuptake inhibitor) and Oxybutynin (an antimuscarinic) *Taranto-Montemurro, et al 2020*

- Increase genioglossus muscle responsiveness (upper airway dilator muscle)
- Reduced OSA severity by 63% in adults

New Studies suggest Long term use (>1 year) is effective *Chen, et al 2021*

Clinical Trial of Medication *ClinicalTrials.gov NCT04115878*

- Children with Down Syndrome, ages 6-17 yo
- Comparing high dose and low dose
- Primary outcome: oAHI
- Secondary outcomes: OSA QoL measures, arousal index, caregiver impression, sleep architecture

Taranto-Montemurro L, et al . Effects of the Combination of Atomoxetine and Oxybutynin on OSA Endotypic Traits. Chest. 2020 Jun;157(6):1626-1636. doi: 10.1016/j.chest.2020.01.012. Epub 2020 Jan 30.

Chen, TY.,. et al. Long-term atomoxetine-oxybutynin combination use may be beneficial for the prevention of obstructive sleep apnea. Sci Rep 11, 12526 (2021). https://doi.org/10.1038/s41598-021-91988-5

Be willing to think outside the box with your Trisomy 21 patients…