

Pediatric Hypoglossal Nerve Stimulation: What, why, and for whom?

Christopher Hartnick MD, MS Epi
Professor

Department of Otolaryngology
Massachusetts Eye and Ear Infirmary
Harvard Medical School

Christopher_hartnick@meei.harvard.edu

A Very Quick Overview...

- Down Syndrome:
 - Most common chromosomal disorder (1/660-800).
- Higher risk of OSA (30-60% of patients)
 - Large tongue
 - Maxilla underdeveloped
 - Adenotonsillar hypertrophy
 - Decreased neuromuscular tone
- OSA persists in 30-50% despite adenotonsillectomy.
- Craniofacial surgeries, tracheotomy, and CPAP are invasive and poorly tolerated.



We needed the “Pediatric STAR trial”...

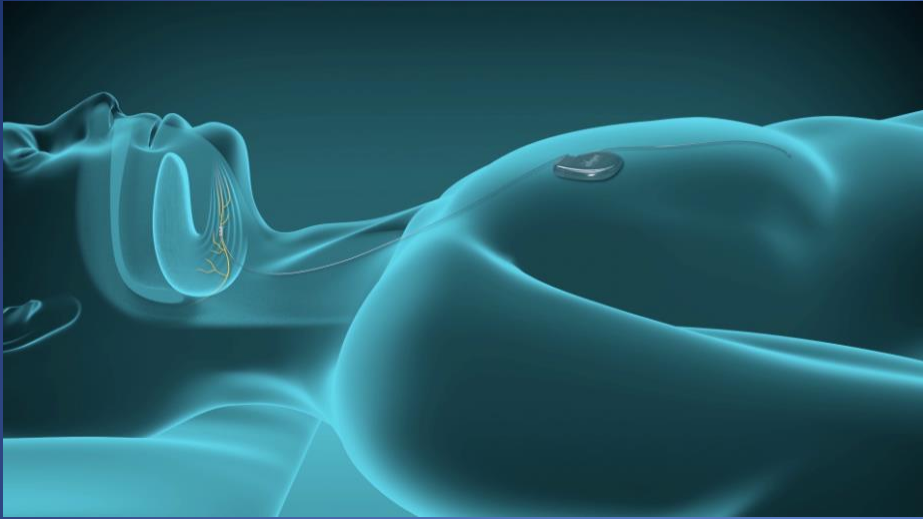
The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Upper-Airway Stimulation for Obstructive Sleep Apnea

Patrick J. Strollo, Jr., M.D., Ryan J. Soose, M.D., Joachim T. Maurer, M.D.,
Nico de Vries, M.D., Jason Cornelius, M.D., Oleg Froymovich, M.D.,
Ronald D. Hanson, M.D., Tapan A. Padhya, M.D., David L. Steward, M.D.,
M. Boyd Gillespie, M.D., B. Tucker Woodson, M.D., Paul H. Van de Heyning, M.D., Ph.D.,
Mark G. Goetting, M.D., Olivier M. Vanderveken, M.D., Ph.D., Neil Feldman, M.D.,
Lennart Knaack, M.D., and Kingman P. Strohl, M.D., for the STAR Trial Group*

Strollo et al. Upper-airway stimulation for obstructive sleep apnea. NEJM 2014; 270 (2): 139-149.



Could HGN Stimulation Work in the Pediatric Down Syndrome Population?

Would it be safe ?

Would it be effective?

This was the rationale to our first multi-institutional FDA IDE approved 42 patient Pediatric HGN study and where these pearls and pitfalls come from!

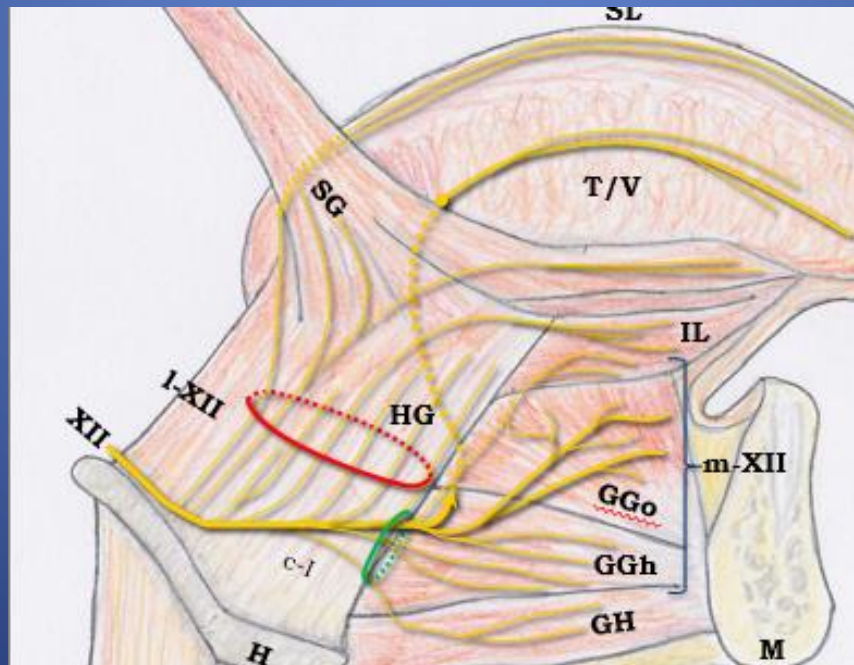
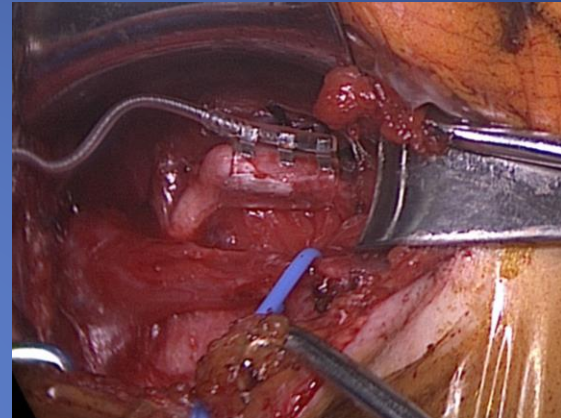
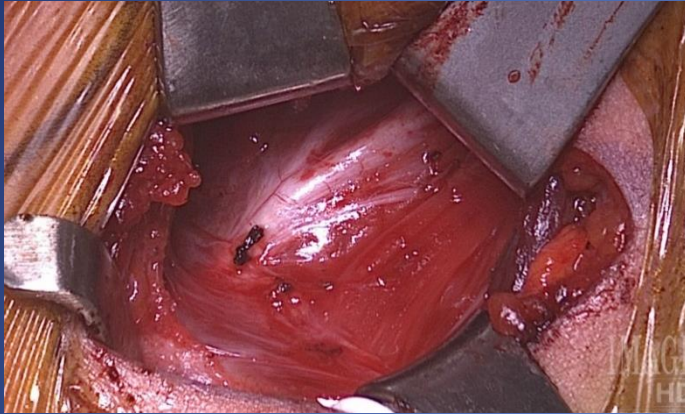
Multi-Center Safety and Efficacy Trial

- Multi-institutional study
- 10-21 years of age with Down syndrome
- Severe OSA; $10 < \text{AHI} < 50$ events/hr
- BMI < 95TH percentile
- Persistent OSA despite T&A
- Intolerance of CPAP or reliant on tracheotomy
- Appropriate DISE

Pre-Op DISE is critical (Pearl discussion later, **but experience matters!**)



Cuff Placement: Potential Pitfall



Overall Details/ Results: first 42 patients

Characteristic	Frequency
Gender	
Male	28 (66.7%)
Female	14 (33.3%)
Age	
10–13	13 (31.0%)
14–17	19 (45.2%)
18–21	10 (23.8%)
BMI Percentile	
Normal (<85 th percentile)	23 (54.8%)
Overweight (85-95 th percentile)	19 (45.2%)

Overall Details/ Results: first 42 patients

Table 3: Polysomnographic outcomes

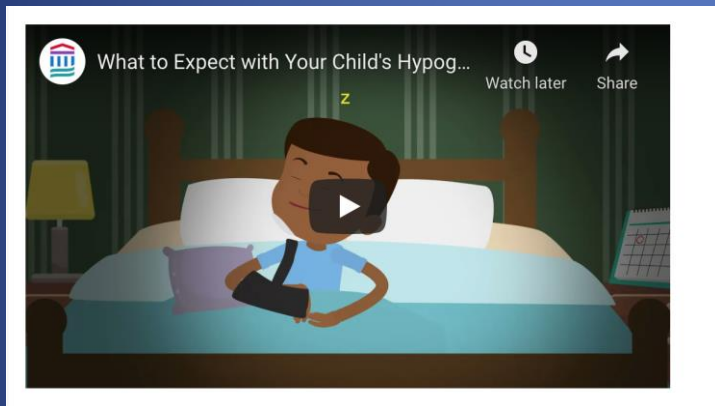
Characteristic	Baseline (SD)	12-Month (SD)	Change (SD)	P-value
Respiratory Events				
AHI	23.5 (9.7)	11.0 (13.4)	-12.9 (13.2)	<0.0001
Obstructive AHI	22.0 (9.8)	10.1 (12.9)	-12.2 (12.7)	<0.0001

There was a mean decrease in AHI of 12.9 ($p < 0.0001$). Using a therapy response definition of a 50% decrease in AHI, the 12-month response rate was 65.9%, and 73.2% of patients had a 12-month AHI under 10.

The average nightly therapy use was 8.96 hours

Complications

Basic Information about what this all is!



https://youtu.be/RXpDR1e_6gc



<https://youtu.be/CBiP9TomjGc>

Website and Facebook

massgeneral.org/children/pediatric-aerodigestive-center

Mass General Brigham
Mass General for Children

About Us Patients & Families Health Care Pr

Hypoglossal Nerve Stimulation on Children with Down Syndrome and Severe Sleep Apnea

We have a NIH-funded grant to work together with the [Down Syndrome Program](#) to evaluate the possible effect of [Hypoglossal Nerve Stimulation on children with Down Syndrome and severe sleep apnea](#). This represents a follow up for pioneering work on this surgical technique in children with Down Syndrome and severe OSA. [36-41] MEE and MGIC are the lead sites in a multi-institutional prospective trial across the country.

What to Expect with Your Child's Hypoglossal Nerve Stimulation

MORE VIDEOS

Switch groups

Adolescents with Down Syndrome and Hypoglossal Nerve Stimulator

Parent Sharing Page

Adolescents with Down Syndrome and Hypoglossal Nerve Stimulator

Private group · 137 members

Join group

News and Stories

Press Coverage

- **STAT:** [For kids with Down syndrome, a new device offers hope for treating difficult sleep problems](#) - *interview with Dr. Hartnick*
- **Healthline:** [A New Device May Help Children With Down Syndrome Get a Better Night's Sleep](#) - *interview with Dr. Hartnick*
- **Medscape:** [Implant May Alleviate Sleep Apnea in Teens With Down Syndrome](#) - *interview with Dr. Hartnick, Dr. Skotko quoted as well from press release*
- **Medpage Today:** [Upper Airway Stimulation Helps Sleep Apnea in Teens With Down Syndrome](#)
- **HealthDay:** [Upper Airway Stimulation Effective in Teens With Down Syndrome, OSA](#)

Post-Op Pain Management!!!

- First night
- Second night
- First week

- Medication
- Route to allow for feedback

Focusing on pearls learned from the outcomes from our first 42 patients

There was a mean decrease in AHI of 12.9 ($p < 0.0001$). Using a therapy response definition of a 50% decrease in AHI, the 12-month response rate was 65.9%, and 73.2% of patients had a 12-month AHI under 10.

Lessons learned from first 42 outcomes

- 73% reduction to AHI is good, but
 - Why not better?
 - Issue with how we titrated sleep studies for first 42?
 - What should be shooting for?
 - AHI < 10? < 5? < 2?
 - Should AHI be the primary outcome we should be looking at?

Pearl: Listening to our patients and their parents

“She’s a different, better kid. She just has a sense of **connecting with other people, connecting with things around her,** being able to not be so exhausted that she can’t deal with transition and frustrations. Those things seem small but as a part of a whole, they’re huge!”

“He will be a senior in high school next year, he never ever would have made it without HGN and sleep, his speech has improved enough he can be understood in school.”

Restore vs Restory

- 16 out of caregivers reported subjective assessments of positive neurocognitive changes and 14 of these 16 reported concomitant expressive speech improvement.
- Questions beget questions:
 - Was/Is this real?

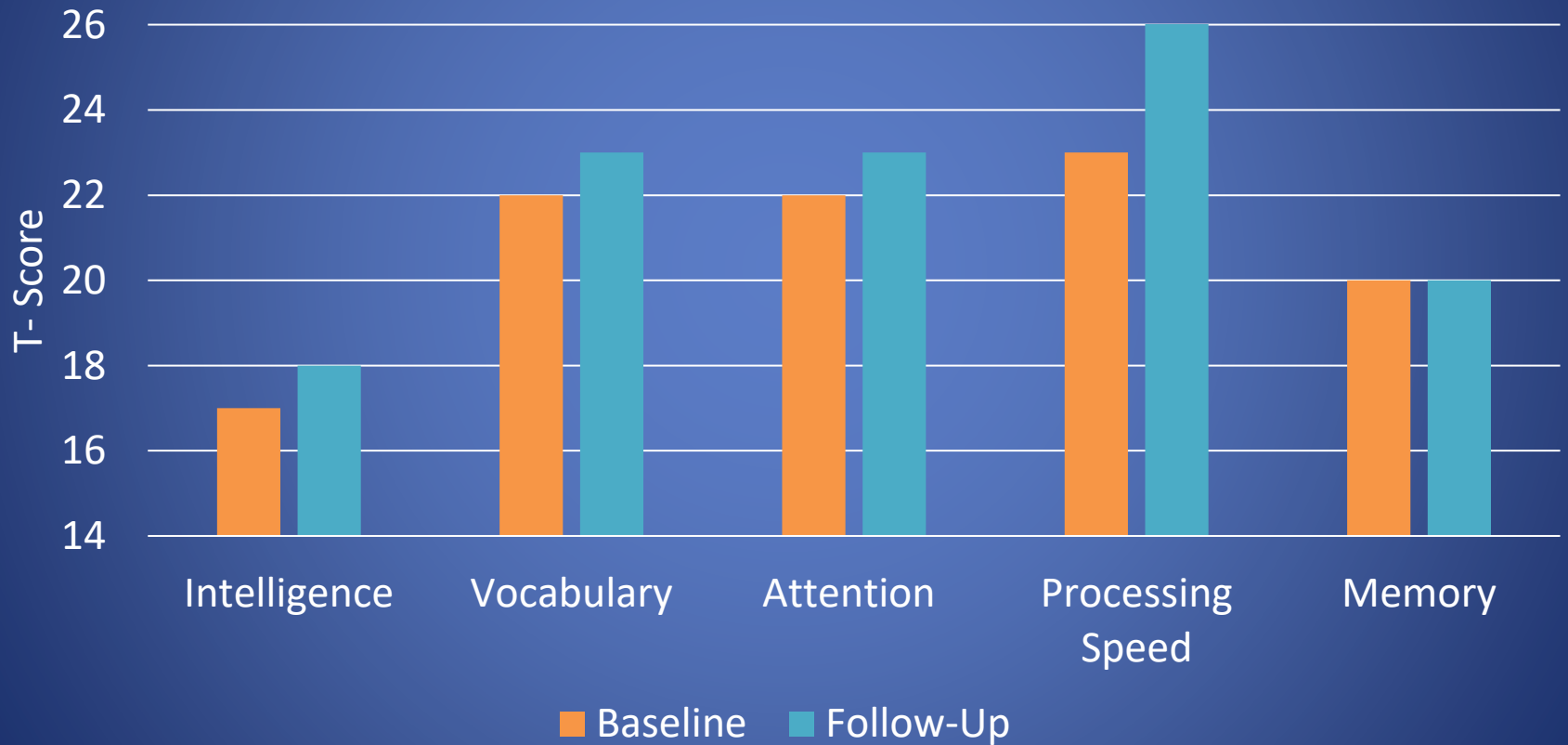
Nested Pilot Study within first 42

- Last 6 children implanted at MEEI within these 42
- Pre and post neurocognitive and expressive language testing
- For these 6, interestingly
 - AHI for these 6 did decrease their AHI by more than 50% and 4/6 has 1 yr AHIs under 10 but
 - None under 2 or for that matter none under 5
 - What happened to their speech and to the measures of cognition?

NCT/ELS of first 6 “nested” children

- General Cognition
- Language (receptive/expressive)
- Visuospatial/motor
- Learning and Memory
- Attention and Executive Functions
- Fine Motor
- Adaptive/Daily Living
- Emotional/Behavioral
- Social Engagement
- Quality of Life
- Articulation and Narrative Sampling

Intelligence and Language



Expressive Language Sampling

- First 6 patients
- Mean (range) change in total number of utterances
+12.3 (-6 to +51)

Why did the results of this nested study matter?

Implication:

- Should we be shooting for a AHI < 2 in all children with DS?
- or, does the change score matter more than the final AHI?
- Are there other factors (such as NCT and EL) that might even be more important?

Next steps

- R01 1R01DC019279-01
 - 09/15/2020 – 08/31/2025
 - NIH-NIDCD
 - Effects of Hypoglossal Nerve Stimulation on Cognition and Language in Down Syndrome

Study Design

- This study aims to test if the treatment of severe OSA with HGS significantly improves the neurocognition (by improving sleep and thereby facilitating memory consolidation) as well as expressive language skills .
- 57 children with DS ages 10-21 across 6 sites nationally
 - MEE (Boston)
 - King's Daughter Norfolk, VA
 - Children's Hospital (Pittsburgh)
 - Emory (Atlanta)
 - CHOP (Philadelphia)
 - UT southwestern (Houston, TX)

Pearl or Pitfall?

- This is why we do research!
 - We don't know the answer to the questions we pose
 - Else they would not be interesting questions
 - How does NCT and EL change with change in AHI following HGNS?
 - Is it a linear relationship? Is there a plateau?
 - Should we be shooting for $AHI < 2$ or something else?
 - So many other questions to ask and answer?
 - I certainly don't have the answers as of yet!
 - Thank you

The team

- MEE
 - Dr Christopher Hartnick
 - Dr. Donald Keamy
 - Dr Gillian Diercks
 - Dr Phoebe Yu
 - Dr John Dobrowski (DSMB)
 - Dr Noah Siegal (DSMB)
 - Elizabeth Grimm and Matt Stenerfson (CCRO)
- MGH
 - **Dr Brian Skotko (Chief DS Clinic and co-PI)**
 - Dr Bernard Kinane (chief, sleep lab)
 - Dr Margaret Pulsifer (NCT)
 - Julie Grieco (NCT)
 - Dr Mark Vangel (Statistician)
 - Dr . Shannon Fracchia
 - Dr Ben Nelson
- UC Davis
 - Dr Len Abbeduto (Expressive Language)
- DISE reviewers
 - Dr. Ryan Soose (Pittsburgh)
 - Dr Raj Deda (Philadelphia)
- MEE fellows :
 - Dr Gillian Diercks
 - Dr Carissa Wentland
 - Dr Sarah Bowe
 - Dr Nikhila Raol
 - Dr. Sarah Bouhabel
 - Dr. Asitha Jayawardena
 - Dr Matt Partian
- Cincinnati
 - Dr Stacey Ishman
 - Dr Sally Shott
- Pittsburgh
 - Dr Alison Tobey
 - Dr. Jeffrey Simons
- Atlanta
 - Dr Nikhila Raol
- Norfolk VA
 - Dr Christina Baldassari
- UT Southwestern
 - Dr Ron Mitchell
- CHOP
 - Dr Lisa Elden

Citations

- Diercks GR, Keamy D, Kinane TB, Skotko B, Schwartz A, Grealish E, Dobrowski J, Soose R, **Hartnick CJ**. Hypoglossal Nerve Stimulator Implantation in an Adolescent with Down Syndrome and Sleep Apnea. *Pediatrics* 2016 May; 137(5). pii: e2015366. doi:10.1542/peds.2015-3663
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