#### Sleep in Children



#### CATHERINE KIER, MD

Professor of Clinical Pediatrics

Division Chief, Pediatric Pulmonary, and Cystic Fibrosis Center

Director, Pediatric Sleep Disorders Center







Hospital.

Stony Brook Children's

#### No disclosures



#### **Objectives**

- 1. Describe the development of sleep patterns from infancy through adolescence.
- 2. Review sleep disordered breathing (obstructive sleep apnea).
- 3. Summarize the approach to the evaluation and diagnosis of sleep disordered breathing (OSA).



#### Sleep Medicine Fellowship

- ACGME
- 1 year fellowship
  - Post-fellowship
  - Post-residency
- Virtual interview



# What is your <u>current</u> subspecialty fellowship training?

- A. Pediatric Pulmonology
- B. Pediatric Sleep
- C. Combined Pediatric Pulmonology and Pediatric Sleep
- D. Pediatric Critical Care
- E. Neonatology
- F. Other



### Does your current program also have a sleep medicine fellowship program?

- A. Yes, General Sleep Medicine Fellowship
- B. Yes, Pediatric Sleep (Sleep Medicine in a Pediatric program)
- C. No
- D. Other



# Are you going to a sleep fellowship training after your current fellowship?

- A. Yes
- B. No
- C. Already done with Sleep Fellowship
- D. Considering to go to Sleep Fellowship



# We are going to answer the following questions:

- 1. Why?
  - Why do we sleep?
  - Complications of sleep deprivation and sleep fragmentation
- 2. What?
  - Basics of sleep (physiology, diagnosis screening, testing sleep studies)
- 3. How?
  - Management of pediatric sleep disorders
  - The latest and evolving options of treatment
- 4. What else?
  - Special populations



#### Wish we all could have a good night sleep!

- Rechtschaffen (1971) "if sleep does not serve an absolute vital function, then it is the biggest mistake the evolutionary process ever made..."
- Neuroplasticity
- sleep linked to memory and learning
- pediatric sleep disorders in the first 5 years of life associated with special educational need at 8 years of age



Curr Opin Neurobiol 2017 Mar 15;44:43-49. Pediatrics 2012;130:634–642



#### Recommended hours of sleep in children

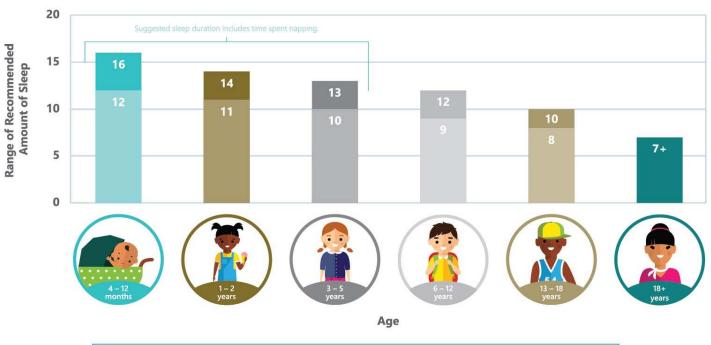
- Ages 4-12 months: 12-16 hours (including naps)
- Ages 1-2 years: 11-14 hours (including naps)
- Ages 3-5 years: 10-13 hours (including naps)
- Age 6-12 years: 9-12 hours
- Age 13-18 years: 8-10 hours
- a consensus statement of the American Academy of Sleep Medicine
- endorsed by the American Academy of Pediatrics
- Sleep is essential for optimal health



J Clin Sleep Med. 2016;12(6):785-786.

#### **Healthy Sleep Duration**

The American Academy of Sleep Medicine recommends that you get the following hours of sleep on a regular basis for optimal health at each stage of life.



SleepEducation.org

A sleep health information resource by the American Academy of Sleep Medicine





# Adverse consequences not just to immediate but to long-term health (adulthood)

- Cardiovascular
  - Hypertension
  - Pulmonary hypertension
  - Cor pulmonale
  - Endothelial dysfunction
- Nutrition and Metabolic
  - Obesity
  - Failure to thrive
  - Insulin resistance
  - Dyslipidemia
- Nocturnal enuresis
- Increased healthcare utilization

- Neuropsychiatric
  - Neurocognitive impairment
  - Hyperactivity
  - Attention deficits
  - Concentration difficulties
  - Impulsivity
  - Anxiety/depression
  - Developmental delay
- Excessive daytime sleepiness



# American Academy of Pediatrics Clinical Practice Guideline

TABLE 2. American Academy of Pediatrics 2012 Guideline
Recommendations for the Diagnosis and Treatment of OSA in
Children and Adolescents (3)

- 1. All children/adolescents should be screened for snoring.
- 2. Polysomnography should be performed in children/adolescents with snoring and symptoms/signs of OSA; if polysomnography is not available, then alternative diagnostic tests or referral to a specialist for more extensive evaluation may be considered.
- 3. Adenotonsillectomy is recommended as the first-line treatment for patients with adenotonsillar hypertrophy.
- 4. High-risk patients should be monitored as inpatients postoperatively.
- 5. Patients should be reevaluated postoperatively to determine whether further treatment is required. Objective testing should be performed in patients who are high risk or have persistent symptoms/signs of OSA after therapy.
- 6. Continuous positive airway pressure is recommended as treatment if adenotonsillectomy is not performed or if OSA persists postoperatively.
- 7. Weight loss is recommended in addition to other therapy in patients who are overweight or obese.
- 8. Intranasal corticosteroids are an option for children with mild OSA in whom adenotonsillectomy is contraindicated or for mild postoperative OSA

OSA=obstructive sleep apnea.

**Pediatrics** 2012 Sep;130(3):e714-55. Pediatrics in Review January 2019, 40 (1) 3-13.



# Pediatric Sleep Disordered Breathing (SDB)



#### Mechanisms of OSA

#### upper airway narrowing

- adenotonsillar hypertrophy
- obesity

#### upper airway collapsibility

- decrease in muscle tone: neuromuscular diseases/cerebral palsy
- airway inflammation: rhinitis
- obesity



Airway patency

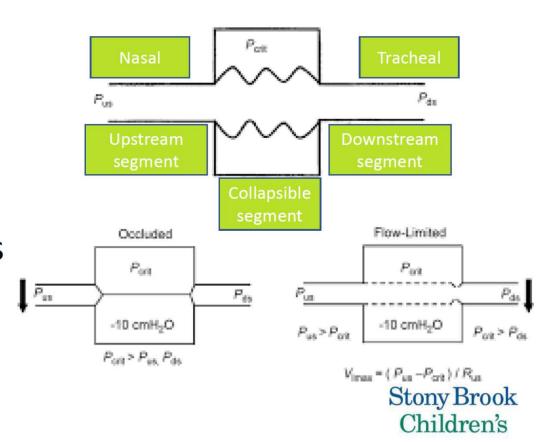




### Increased tone of the upper airway dilator muscles may compensate for a narrow airway

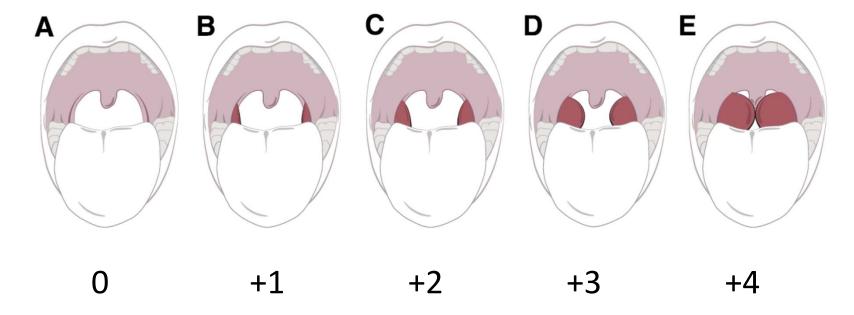
 Pcrit (pressure at which the airway collapse occurs)

 normal children able to maintain inspiratory airflow at subatmospheric pressures but OSA children have Pcrit value in the positive range



J Appl Physiol. 1994 Aug;77(2):918-24.

#### **Tonsil size**





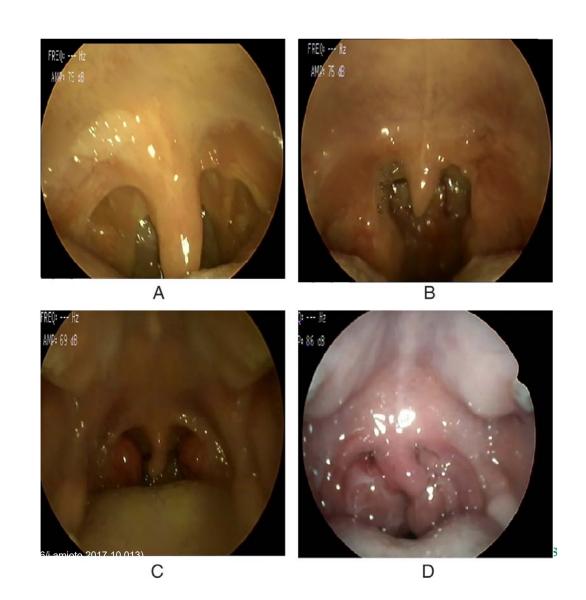
# Brodsky tonsillar grading scale

Grade 1: tonsil occupies ≤25% of the oropharyngeal width to midline

Grade 2: Tonsil occupies 26%–50%

Grade 3: Tonsil occupies 51%–75%

Grade 4: Tonsil occupies >75%

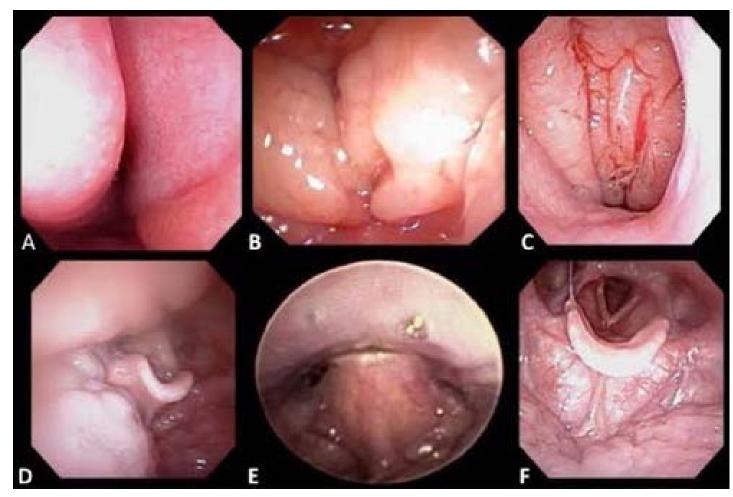


# Does your program/institution perform sleep endoscopy?

- A. Yes, Pediatric Pulmonary
- B. Yes, ENT
- C. Yes, both Pediatric Pulmonology and ENT could do
- D. No



#### Sleep endoscopy



Laryngoscope Investigative Otolaryngology. 2017;2(6):423-431.

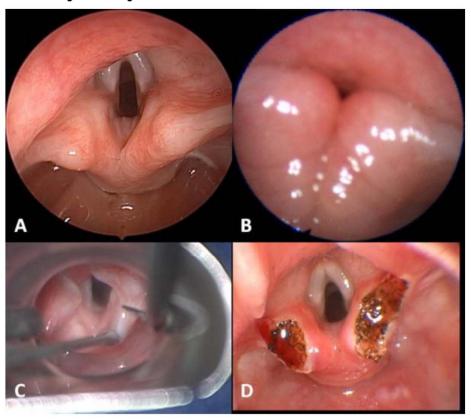
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5743164/



# Supraglottoplasty for laryngomalacia with obstructive sleep apnea

- polysomnography useful to identify OSA and/or to objectively measure outcomes
- mild improvement (10%), significant improvement (70%), and complete resolution (20%) of stridor and nocturnal snoring at 4 weeks after discharge
- obstructive apnea index, obstructive apnea/hypopnea index, respiratory disturbance index and O2 nadir improved

Laryngoscope. 2008 Oct;118(10):1873-7. Internat J of Ped Otorhinolaryn. 2011 Oct; 75(10): 1234-1239



### Does your program have a sleep center or sleep lab?

- A. Yes, combined adult and pediatric
- B. Yes, pediatric only
- C. Yes, adult only
- D. No



# Classification of OSA in Children (versus adults) based on Apnea Hypopnea Index

Severity	Pediatric	Adult	
Normal	<1	<5	
Mild	1-5	5-15	
Moderate	>5-10	>15-30	
Severe	>10	>30	



J Thorac Dis. 2016;8(2):224–235.

- Primary snoring
- Upper airway resistance syndrome (UARS)
- Obstructive hypoventilation
- OSA syndrome (OSAS)



Primary snoring

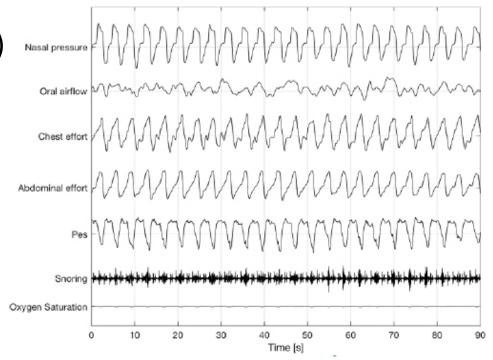
• habitual snoring for more than 3 nights per week without apneas, hypopneas, frequent arousals or

gas exchange abnormalities (prevalence 7.45%)

Upper airway resistance syndrome (UARS)

Obstructive hypoventilation

OSA syndrome (OSAS)

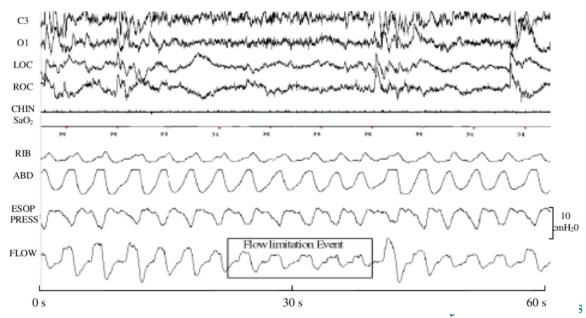


- Primary snoring
- Upper airway resistance syndrome (UARS)

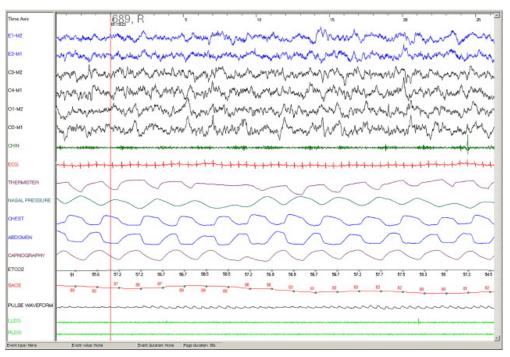
• snoring, increased work of breathing and frequent arousals, without recognizable obstructive

events or gas exchange abnormalities

- Obstructive hypoventilation
- OSA syndrome (OSAS)



- Primary snoring
- Upper airway resistance syndrome (UARS)
- Obstructive hypoventilation
  - snoring plus elevated end-expiratory carbon dioxide partial pressure in the absence of recognizable obstructive events
- OSA syndrome (OSAS)



#### Respiratory disturbance index (RDI)

- The AHI is the number of <u>apneas</u> and <u>hypopneas</u> per hour of <u>sleep</u>.
- The term (RDI) is often used as another term for the AHI
- AASM Scoring Manual recommends that the RDI be used for the sum of the AHI and the RERA index (number of RERAs per hour of sleep).



#### Obstructive hypoventilation

- Measurements:
- 1. through nasal cannula: endtidal  $CO_2$  (PET<sub>CO2</sub>)
- may be difficult to maintain, especially in young children
- 2. Through skin: transcutaneous CO<sub>2</sub> monitoring (Ptc<sub>CO2</sub>)
- -warming the surface of the skin so that capillary blood is arterialized and closely matches arterial CO<sub>2</sub> tension.







# Obstructive hypoventilation (CO2 parameters)

Severity	CO2> 50 mm Hg	
Normal	<10%	
Mild	10-25% TST	
Moderate	>25-50% TST	
Severe	>50%	

PET<sub>CO2</sub> > 45 mm Hg exceeds more than 60% of TST

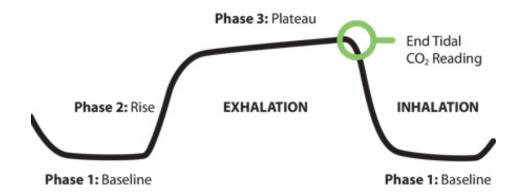
AASM guidelines recommend that obstructive hypoventilation can be diagnosed when more than 25% of TST is spent with a  $Pet_{CO2}$  of more than 50 mm Hg



Proc Am Thorac Soc. 2008 Feb 15; 5(2): 263-273.

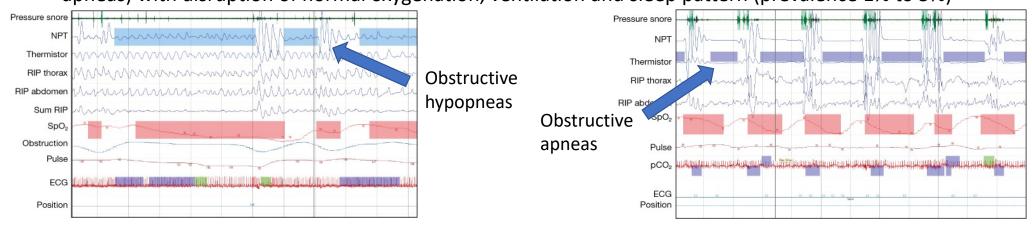
# Even obstructive hypoventilation alone (isolated hypoventilation)

- Can result in disruption of normal ventilation and sleep patterns neurocognitive deficits, and cardiovascular morbidities
- hypoventilation is a common in children with Down's syndrome and obese children with obstructive sleep apnea syndrome (OSAS)





- Primary snoring
- Upper airway resistance syndrome (UARS)
- Obstructive hypoventilation
- OSA syndrome (OSAS)
  - recurrent events of partial or complete upper airway obstruction (hypopneas, obstructive or mixed apneas) with disruption of normal oxygenation, ventilation and sleep pattern (prevalence 1% to 5%)



#### Central apnea is a real entity but...

- Post-arousal sighs
- central apneas are commonly seen following an arousal or after a sigh
   Herring-Breuer reflex or hypocapnia induced by hyperventilation caused by a sigh or arousal
- Central apneas in children how much is acceptable



#### Central apnea ranges per age group

Polysomnographic Characteristics in Normal Preschool and Early School-Aged Children

Hawley E. Montgomery-Downs, PhDa, Louise M. O'Brien, PhDb, Tanya E. Gulliver, MDc, David Gozal, MDb

aDepartment of Psychology, West Virginia University, Morgantown, West Virginia; bKosair Children's Hospital Research Institute and Division of Pediatric Sleep Medicine, Department of Pediatrics, University of Louisville, Louisville, Kentucky; Department of Paediatric Respiratory and Sleep, John Hunter Children's Hospital, Newcastle, New South Wales, Australia

The authors have indicated they have no relationships relevant to this article to disclose.

TABLE 5 Apnea, Apnea Subtypes, and Hypopnea Indices

542 children age range: 3.2 to 8.6 years

	151		
	Average	SD	Range
3- to 5-y-olds			
Total AHI	0.90	0.78	0-3.6
Total Al	0.86	0.75	0-3.6
Central Al	0.82	0.73	0-3.6
Obstructive Al	0.03	0.10	09
Mixed Al	0.01	0.05	060
Total hypopnea index	0.03	0.07	040
6- to 7-y-olds			
Total AHI	0.68	0.75	0-6.60
Total Al	0.50	0.52	0-3.60
Central Al	0.45	0.49	0-3.40
Obstructive Al	0.05	0.11	09
Mixed Al	0.01	0.06	0-1.1
Total hypopnea index	0.10	0.18	090

Pediatrics March 2006, VOLUME 117 / ISSUE 3





#### Normal Polysomnographic Values for Children and Adolescents

Carole L. Marcus 4, Kenneth J. Omlin , Daniel J. Basinki , Sandra L. Bailey , Adriana B. Rachal , Walter S. Von Pechmann , Thomas G. Keens , and Sally L. Davidson Ward

Received: June 24, 1991

50 normal children and adolescents (9.7 ± 4.6 SD yr) range 1.1 to 17.4 yr



### Proposed classification of pediatric OSA

- two types of OSA in children (Gozal, et al)
  - Type I adenotonsillar hypertrophy
  - Type II obese children and adolescents
  - analogy with type I and type II diabetes
- (Type III variety of craniofacial and neuromuscular disorders)
  - Crouzon and Apert syndromes, Pierre Robin sequence, Down syndrome, Goldenhar syndrome, achondroplasia, myelomeningocele, and cerebral palsy

Clin Sleep Med 2007;42:374–379.

Proc Am Thorac Soc; 2008 Feb 15; 5(2):274-282.



## Anatomical features $\rightarrow$ upper airway narrowing

- Micrognathia
- Macroglossia
- Midface hypoplasia

Treacher Collins syndrome,
Crouzon syndrome, Apert syndrome,
Pierre Robin sequence,
achondroplasia, trisomy 21,
Beckwith Wiedemann syndrome,
mucopolysaccharidoses







Stown Brook Children's 's

### Epidemic of obesity

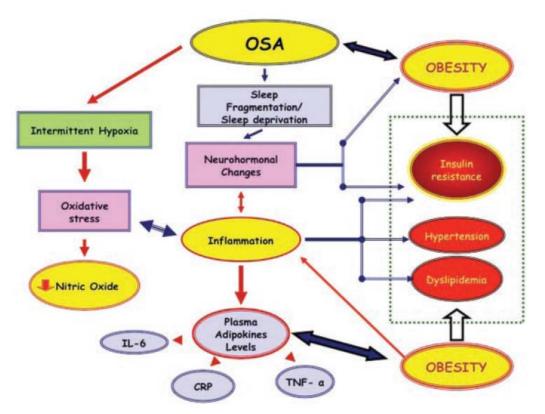
- About 50% of overweight/obese children have obstructive sleep apnea (OSA) compared to up to 6% of normal weight children
- Sleep 2014; 37:943-9
- every 1 kg/m2 increase in body mass index (BMI) above the mean for age and sex increases the risk of developing OSA by 12%
- Am J Respir Crit Care Med 2007;176:401e8.



## Two key issues that underpin the development of OSA-related morbidities

low grade systemic inflammation

increased oxidative stress



Pediatr Pulmonol. 2011; 46:313-323.

Fig. 1. Hypothetical interaction of obstructive sleep apnea syndrome and obesity in activating pathways leading to metabolic disease. TNF- $\alpha$ , tumor necrosis factor alpha; IL-6, interleukin-6; CRP, C-reactive protein.

#### OSA and inflammation evidences

- Nasal nitric oxide elevated in OSA and primary snoring (marker of airway inflammation)
- -Sleep Breath.2016 Mar;20(1):303-8.
- H2O2 in morning exhaled breath condensate elevated (marker of oxidative stress)
- Sleep Breath 2012;16:703-8.
- pro-inflammatory cytokines, TNF- $\alpha$ , IL-6, and IL-8 elevated in serum
- regulatory cytokines, IL-10 decreased
- Mediators Inflamm 2015;2015:493409.

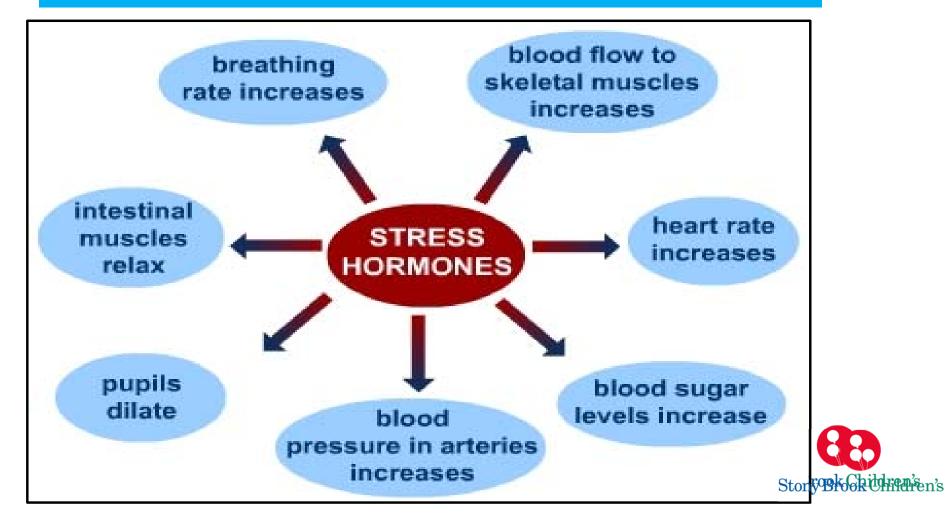


### Fight and flight

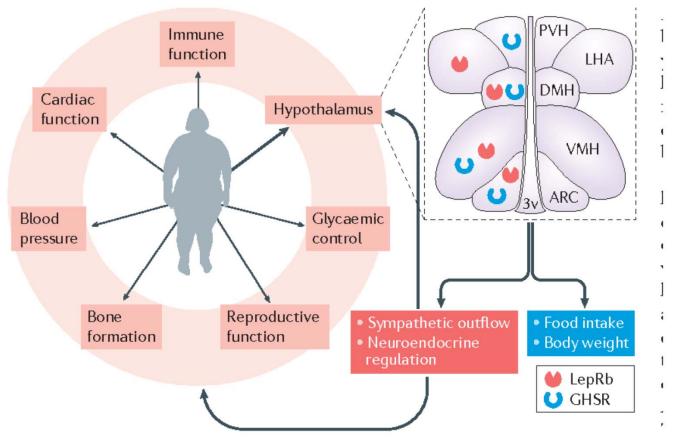
- an interruption of normal sleep architecture
- alterations in physiologic gas exchange
  - repetitive decreases in oxygen saturation followed by rapid re-oxygenation
  - episodic hypercapnia
- occlusion of the upper airway
  - large fluxes in intrathoracic pressure
  - recurrent brain arousals
- induces potent and sustained activation of sympathetic nervous system

StatenBrando@bildhebrein's

### Stress hormones during sleep!!!



#### Affects a wide range of physiological processes





#### SOMATIC AROUSAL = Sympathetic Tone

Reflects the sympathetic nervous system component of the stress response

#### Manifestation of stress:

- Increased heart rate
- Sweaty palms
- Dry mouth





### **Body Sensation Questionnaire**

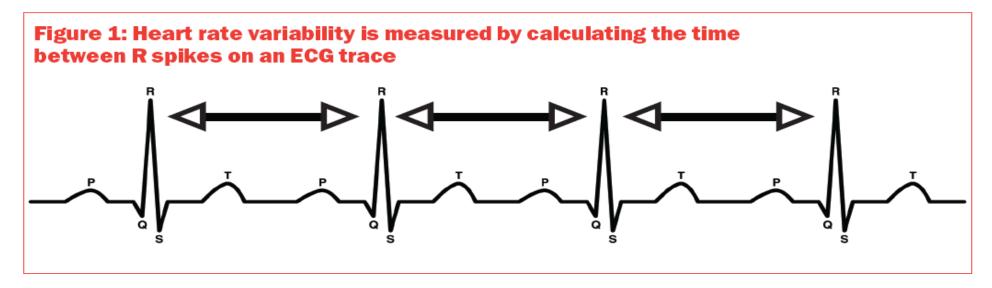
	Body	Sensations Q	uestionnaire		•
sometimes h best describ	list of feelings, nave. Read each i es how much you ding today. Use t	tem and use the that have felt or expe	f to 5 scale below rienced things this	to select a rating way during the	that
1 Not at all	2	3. Moderately	Quite a bit	5 : Extremely	-   .
Not at all	A little bit	Moderately	Quite a bit	Extraction	
<ol> <li>Startled</li> </ol>	easily				
2. Hands v	vere shaky				
3. Was sho	ort of breath		<b></b>		-
4. Felt fain	t .ii				-
5. Had hot	or cold spells				-1
6, Hands v	vere cold or sweat	y			
7. Was treat	mbling or shaking	er an hear with 5	(4 d		_
8. Had trou	ble swallowing				_
	y or lightheaded				_
	in my chest				_
	I was choking				
12. Muscles twitched or trembled					
13. Had a very dry mouth					
	id I was going to				
	t racing, pounding				_
	571.				_
	bness or tingling i				_
17. Had to u	rinate frequently.				
				SUM	4

- Somatic syndromes, insomnia, anxiety, and stress among sleep disordered breathing patients
- BSQ is a measure of somatic arousal and correlates with sympathetic tone measured by heart rate variability in adult patients with sleep-disordered breathing

Stony Brook Children's

Sleep Breath. 2016 May;20(2):759-68.

#### Heart Rate Variability



- It is measured by the variation in the beat-to-beat interval, also known as R-R intervals.
- The SA node receives several different inputs and the instantaneous heart rate or RR interval and its variation are the results of these inputs.
- The main inputs are the sympathetic and the parasympathetic nervous system (PSNS) and humoral factors.



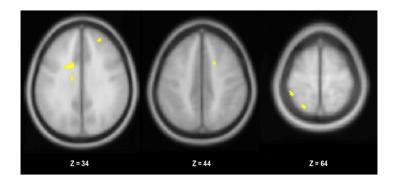
### **Heart Rate Variability**

- High HRV: indication of healthy autonomic and cardiovascular response.
- Low HRV: may indicate that the sympathetic and parasympathetic nervous systems aren't properly coordinating to provide an appropriate heart rate response.
- measured by linear parameters (time domain and frequency domain) and non-linear parameters.



## Neurocognitive and behavioral morbidities

- Probable mechanistic role?
- MRI data showed greater activity in regions of the brain implicated in cognitive control, conflict monitoring and attentional allocation for children with OSA (in order to perform same tasks at same level)

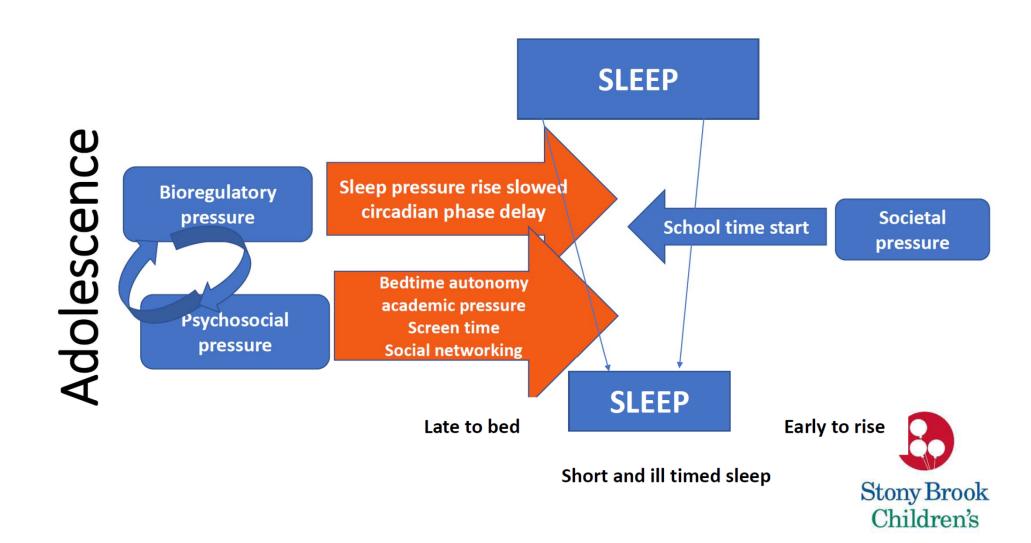




## Sleep-disordered breathing and school performance in children (Gozal)

- a prospective study in first-grade school children
- OSA disproportionally high in the lowest 10% of their class
- while children treated for OSA showed significant academic improvement versus children who did not receive treatment did not improve





#### Adolescents – the perfect storm

- maturation of sleep regulatory systems during adolescence in combination with psychosocial and societal pressures culminate in a "Perfect Storm" of short and ill-timed sleep and the associated consequences
- 14% higher among students who slept less than 7 hours on school night
- Hours of sleep ask sleep hygiene
- Media and sleep
- Task force/advocacy for teens



Journal of Adolescence 67 (2018) 55-65

## Management of pediatric OSA



#### Childhood Adenotonsillectomy (CHAT) Study

- the first ever randomized controlled trial to compare adenotonsillectomy with watchful waiting
- 464 children, 5 to 9 years of age
- primary outcome: change in the attention and executive-function score (Developmental Neuropsychological Assessment) did not differ significantly (7 months follow-up)
- Secondary outcomes: behavioral, quality-of-life, and polysomnographic findings and significantly greater reduction in symptoms in the early adenotonsillectomy
- Normalization of polysomnographic studies (79% vs. 46%)



## CHAT follow-up study — comparison of sleep study and symptoms

- did not undergo adenotonsillectomy
  - 42 % had resolution of OSA on follow-up PSG 7 months later (lower apnea-hypopnea index and and waist circumference <90 % percentile)
  - Only 15 % experienced symptomatic resolution (lower sleep questionnaire and snoring scores)







Chest. 2015 Nov;148(5):1204-13.

### Adenotonsillectomy

- Outcome or success rate of adenotonsillectomy
- true percentage? success rate?
- recent study (multicenter retrospective) : 30%-based on post-op PSG (AHI)
- 12 school-age children four years after T&A, 2/3 normalized their PSG findings
- 23 pre-school children three years after T&A, 61% had normal sleep

*Am J Respir Crit Care Med.* 2010;182:676–83. Sleep Med. **2013**,14, 440–448. J. Clin. Sleep Med. **2015**, 11, 1143–1151.



## Complications of T and A

- >600,000 T&A are performed annually in the US alone
- Complications of T and A (meta-analysis)
  - Children with OSA appear to have more respiratory complications
  - Most frequent early complications are respiratory compromise and secondary hemorrhage

JAMA Otolaryngol. Head Neck Surg. **2018**, 144, 594–603. *Pediatrics*. 2015 Oct;136(4):702-18.



### Risk factors for persistent OSA

- Obesity
- Severe OSA (AHI)>20/hr TST)
- Older age (>7 years)
- Asthma
- African ethnicity
- History of prematurity



#### Prematurity

- often associated with generalized muscle hypotonia
- atypical breathing pattern is associated with the development of mouth breathing and a high and narrow hard palate
- early premature infants often have abnormalities with feeding functions, such as suction, mastication, and swallowing
- weakness of orofacial muscles negatively alter craniofacial growth and lead to a small UA
- Higher risk of obesity at later age



# Medical management (pharmacotherapy)

- tonsillar cultures in OSA: CD3, CD4 and CD8 lymphocytes and proinflammatory cytokines TNF $\alpha$ , IL-1 $\alpha$  and IL-6 are increased
- In vitro experiments: corticosteroids were added to OSA tonsillar cell cultures, decreased proliferative rates and increased apoptosis, and reduction in the secretion of IL-6, IL-8 and TNF $\alpha$
- randomized crossover trial of intranasal budesonide for mild OSA (6 weeks of therapy)
  - reduction in OSA severity, decreased adenoidal size

Eur Respir J. 2009;33:1077–84.

Pediatrics. 2008;122:e149-55



### Leukotriene antagonists (LT)

- In vitro:
  - increased levels of leukotriene receptors 1 and 2 in tonsils from OSA
  - LT antagonists to tonsillar cell cultures with dose-dependent reductions in cellular proliferation and secretion of the cytokines TNF $\alpha$ ,IL-6 and IL-12
- Clinically:
  - double-blind placebo-controlled study
    - Improvement in sleep disturbances



#### When to use O2 in OSA

#### AMERICAN THORACIC SOCIETY DOCUMENTS

#### Home Oxygen Therapy for Children

An Official American Thoracic Society Clinical Practice Guideline

Don Hayes, Jr., Kevin C. Wilson, Katelyn Krivchenia, Stephen M. M. Hawkins, Ian M. Balfour-Lynn, David Gozal, Howard B. Panitch, Mark L. Splaingard, Lawrence M. Rhein, Geoffrey Kurland, Steven H. Abman, Timothy M. Hoffman, Christopher L. Carroll, Mary E. Cataletto, Dmitry Tumin, Eyal Oren, Richard J. Martin, Joyce Baker, Gregory R. Porta, Deborah Kaley, Ann Gettys, and Robin R. Deterding; on behalf of the American Thoracic Society Assembly on Pediatrics

This official clinical practice guideline of the American Thoracic Society was approved December 2018

 sleep-disordered breathing complicated by severe nocturnal hypoxemia who cannot tolerate positive airway pressure therapy or are awaiting surgical treatment home oxygen therapy be prescribed (conditional recommendation, very low-quality evidence)

Stony Brook

Children's

AJRCCM Volume 199 Number 3 February 1 2019

#### CPAP in children

- Indications:
  - minimally enlarged tonsils and adenoids
  - residual OSA after adenotonsillectomy
- effective CPAP treatment linked with improvements in cognitive performance
- Adherence can be extremely challenging in children
- Ramp pressure (start with lower pressure) or APAP (automatic positive airway pressure – pressure range)



#### Bariatric surgery

- After significant weight loss, OSA severity markedly decreased (median apnea-hypopnea index at baseline vs. after weight loss, 9.1 vs. 0.65)
- bariatric surgery reliably decrease weight in a sustainable fashion and reverse many of the comorbidities associated with obesity

Obes Res. 2005 Jul;13(7):1175-9. Arch Pediatr Adolesc Med. 2012;166(8):757-766.



## Does your program refer for rapid maxillary expansion?

- A. Yes
- B. No



## Rapid maxillary expansion (RME)

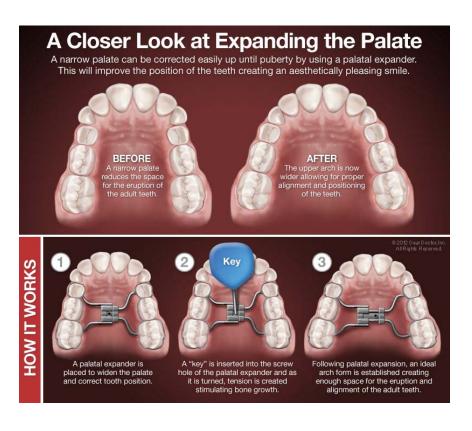
 orthodontic appliance deliver a lateral force to the upper posterior molars, opens the midpalatal suture transversely and therefore widens the nasal cavity







# Efficacy of RME (rapid maxillary expansion)



- decreases nasal resistance and improves nasal airflow.
- improves maxillary arch with increase in oropharyngeal space
- helps repositioning the high riding tongue appropriately thereby, resolving mouth breathing



# Metanalysis of RME (rapid maxillary expansion)

- Camacho et al., (2017) 17 studies and a total of 314 children with mean age of 7.6+/-2 years (transverse maxillary deficiency with high arched narrow palate with OSA)
  - At < 3 years follow up, AHI was noted to drop 70% from baseline</li>
  - At > 3yrs follow up, there was 79% reduction in AHI

- Sanchez-Sucar (2019) 9 studies
  - AHI dropped by 5.79 events/hour (CI-95% 9.06-2.5) and mean oxygen saturations increased by 2.54%(CI95% 0.28 to 4.8,6.7%)
- Machado- Junior et al (2016) 215 children



#### First Visit 1/30/2019



Stony Brook Children's

With permission

#### Treatment progress 4/10/2019























#### Retention 2/4/2020























#### First Visit 1/30/2019

#### Post-RME 2/4/2020











































#### Limitations of access to RME treatment

- poor insurance coverage
- a lack of adequate screening and referral by primary care and dental providers
- a multidisciplinary team involving pediatric sleep, otolaryngology, orthodontic, oromaxillary, speech pathology and pediatricians is highly encouraged
- Cost-analysis:



# Does your sleep practice/program refer for myofunctional therapy?

- A. Yes
- B. No
- C. Don't know
- D. We have no sleep practice/program



# Myofunctional therapy

- structured intervention
- specific oropharyngeal exercises:
  - improvements in labial seal and lip tone
  - use of nasal breathing as the preferred respiratory route
  - more favorable positioning of the tongue within the oral cavity
- strengthening of the tongue and orofacial muscles
- relatively easy to teach to patients, child co-operation and adherence are essential

Myofunctional Therapy: Role in Pediatric OSA. Sleep Med. Clin. **2019**, 14, 135–142.



## Hypoglossal nerve stimulator

- an implanted medical device electrically stimulating the hypoglossal nerve, which causes tongue movement
- timed with breathing to relieve upper airway obstruction
- Investigational phase in children; role in Down's syndrome and OSA





## Summary

- Pathophysiology of OSA mechanics of upper airway narrowing and increased airway collapsibility
- Yet, pediatric OSA spectrum is complex
- Phenotypic variations due to interplay of genetic, environmental, disease severity and individual susceptibility factors
- Is it time to look into personalized treatments for pediatric OSA -precision medicine, for improved outcomes?



# Special populations



## Down's syndrome

- Poor sleep
- reduced REM sleep and increased slow-wave sleep independent of OSA
- implications for learning, memory, and behavior
- Anatomy: maxillary hypoplasia and small nose with low nasal bridge midface
- Recurrent upper airway infections 

   adenotonsillar hypertrophy
- Hypotonia
- Obstructive hypoventilation is common Sleep Breath 19 (3), 1065-1071. 2014 Dec 12.



Stony Brook

Children's

#### Craniofacial anomalies

- clinical findings snoring, work of breathing, desaturations
- congenital craniofacial anomalies is strongly associated with inpatient diagnosis of OSA
- anatomic features maxillary or mandibular hypoplasia, crowded oropharynx, macroglossia, or poor motor tone

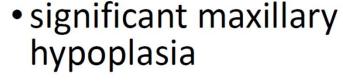


#### Craniosynostosis















 estimated prevalence of OSA of 53% in this population based on symptom report

Apert syndrome

Crouzon syndrome Pfeiffer syndrome



#### Oromaxillofacial procedures

 surgical advancement of the midface to enlarge the upper airway

Le Fort II or Le Fort III procedure (internal or external distraction devices)

- substantially reduced the AHI and improved oxygen saturation in a cohort of 11 children (Pfeiffer, Crouzon, or Apert syndrome and severe OSAS)
- external transfacial pin for distraction J Plast Reconstr Aesthet Surg. 2013 Sep;66(9):1206–1211.



## Pierre Robin Sequence





### Pierre Robin sequence surgery

- Tongue-lip adhesion and tongue repositioning can improve apnea/hypopnea index (AHI) and oxygenation saturations
- Systematic review (90 patients)
- Tongue-lip adhesion
  - AHI from 30 to 18 events per hour (50% reduction)
  - lowest oxygen saturation from 75% to 84%
- Tongue repositioning
  - AHI from 46 to 17 events per hour (62% reduction)
  - mean oxygen saturation from 90 to 95%

Laryngol Otol. 2017 May;131(5):378-383.





## Autism spectrum disorder (ASD)

Stony Brook Children's

- Sleep problems as high as 80% in children with ASD
- Sleep problems and insufficient sleep →
- daytime sleepiness, learning problems and behavioral issues
- hyperactivity, inattentiveness and aggression
- most common sleep problems
- difficulty falling asleep and repeated awakenings
- very prolonged awakenings or awaken very early for the day
- sleep of other family members is often impacted

#### Does Melatonin work?

- Melatonin used commonly in children with autism spectrum disorder
- role in regulating the circadian sleep cycle
- reported to be low in individuals with ASD
- 1–3mg of supplemental melatonin (comparable pharmacokinetic parameters with typically developing children



#### Possible actions/role of melatonin

- hypnotic independent of a deficiency state (with normal endogenous melatonin levels)
- chronobiotic (circadian phase-shifting for delayed sleep onset)
- antianxiolytic (comorbid anxiety)
- mitigate hyperarousal-related insomnia through its effects on the hypothalamic pituitary adrenal axis (suppress ACTH in animal model)
- non-responders or exhibit disappearing effectiveness may be related to dysfunction of CYP1A2 genes (slow metabolizers)

Stony Brook

Children's

J Nat Sci. 2015; 1(7): e125.