

# Narcolepsy in Otolaryngology

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# Questions

- Is narcolepsy important to an otolaryngologist?
- How do narcolepsy patients present in the clinic
- Why is there a sleep domain on the SNOT 22?
- Do sleep disorders help to understand the subtypes of sinusitis?

# Narcolepsy

- A disorder of the sleep wake cycle
- Patients have disturbed sleep at night
- Patients are sleepy during the day

# Questionnaires

- Stop Bang
- Epworth Sleepiness scale
- SNOT 22

# Narcolepsy demographics

- More common than in literature
- Most patients do not have cataplexy
- Majority of my patients are women
- Often onset in middle school

# Narcolepsy History

- Daytime sleepiness out of proportion to severity of apnea
- Vivid dreaming , hypnagogic hallucinations
- Dreaming before asleep
- Dreaming during naps
- Sleep paralysis frequency
- cataplexy

# Narcolepsy History

- Hard to get out of bed in the morning
- Repeat sinus infections
- Repeat viral infections mononucleosis
- Traumatic brain injury
- Prior therapy
- Caffeine usage

# Sleep apnea

- Must be treated first before testing for narcolepsy



# Narcolepsy history

- School/ work performance
- Impact on family
- Caffeine use/ insomnia
- Sleep hygiene
- Sleep logs
- Near miss driving
- Naps restorative

# REM sleep

- 90 minutes till first REM episode
- Paralyzed
- Turbinate engorgement
- REM episode closer together in early morning

# Types of hyper-somnolence

- Narcolepsy Type 1
- Narcolepsy Type 2
- Idiopathic hyper-somnolence

# Narcolepsy Type 1

- EDS + Orexin/hypocretin deficiency 90%
- EDS + Cataplexy + MSLT findings
- HLA DQB1\*0602 94% in cataplexy
- MSLT < 8 minutes with 2 SOREMP or 1 Soremp and 1 Soremp on polysomnogram
- Cataplexy absent in 30% of patients with low hypocretin

# Narcolepsy Type 2

- No cataplexy
- Normal Orexin CSF
- MSLT findings
- Clinical History

# MSLT Interpretation

- Look for REM arising out of stage wake or stage 1

# MSLT Limitations

- False positives 6% of men 1 % of women
- False negatives 20% anxiety, psych medications, noise in lab
- MSLT not performed to guidelines- lack of actigraphy and sleep logs- sleep logs under estimate actigraphy by 1.5 hours
- Even guidelines 6-7 hours may not be enough to be satiated
- Poor test retest reproducibility in NT2/ IH- diagnosis changes in 50 %

# Test-Retest Reliability of MSLT in Narcolepsy

- MSLT 95% sensitivity in Type 1 narcolepsy
- 78% of Type 1 positive on test retest
- For type 2 narcoleptics only 18% positive on repeat study
- 2-5% of normals show + MSLT on a single study M>F



# Treatment

- Concerta- methylphenadate
- Provigil- modafinil
- Xyrem- Sodium Oxybate
- Ritalin
- Trazadone
- Baclofen
- Trilosant - H3 blocker

# SNOT 22

- Minimal clinically important difference
- Instructions on questionnaire
- Kaiser Permanente average surgeon improved 20 points
- Best surgeon improved by 30 points
- Office selected balloon sinuplasty improved by 40 points

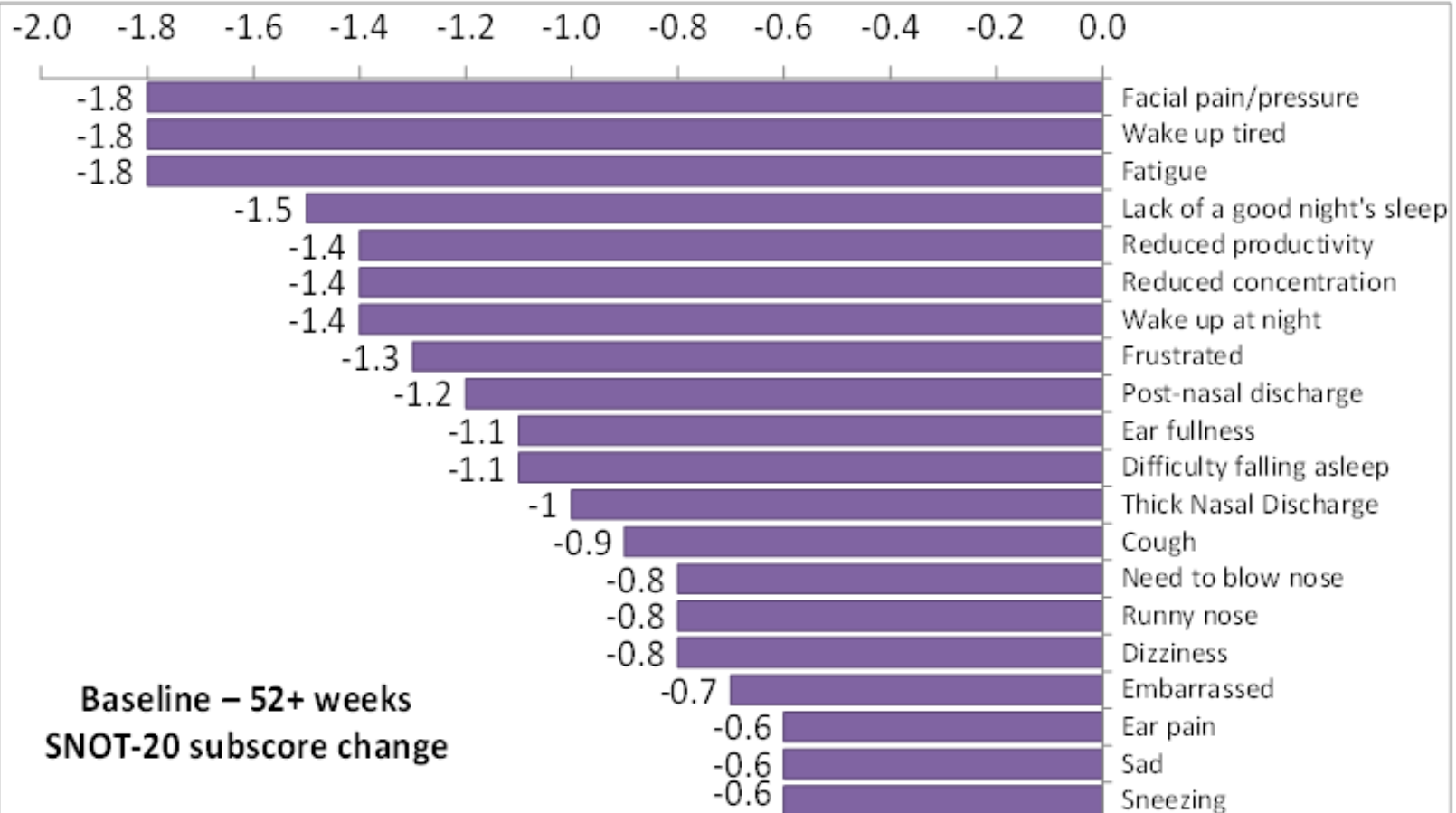
# Balloon sinuplasty Orios study

- Karanfilov B, et al. Office-based balloon sinus dilation: a prospective, multicenter study of 203 patients. *Int Forum Allergy Rhinol*, 2013; 3:404-411
- Sikand, et al. Office-Based Balloon Sinus Dilation 52+ week follow-up of a prospective multicenter study. Presented at American Rhinologic Society, April 2013, Orlando

# Orios study

- Safe as FESS in the operating room
- Similar reduction in SNOT 20
- Snot 20 most improves subgroups were facial pain, and slept better at night.

# Results : SNOT-20 Subscores



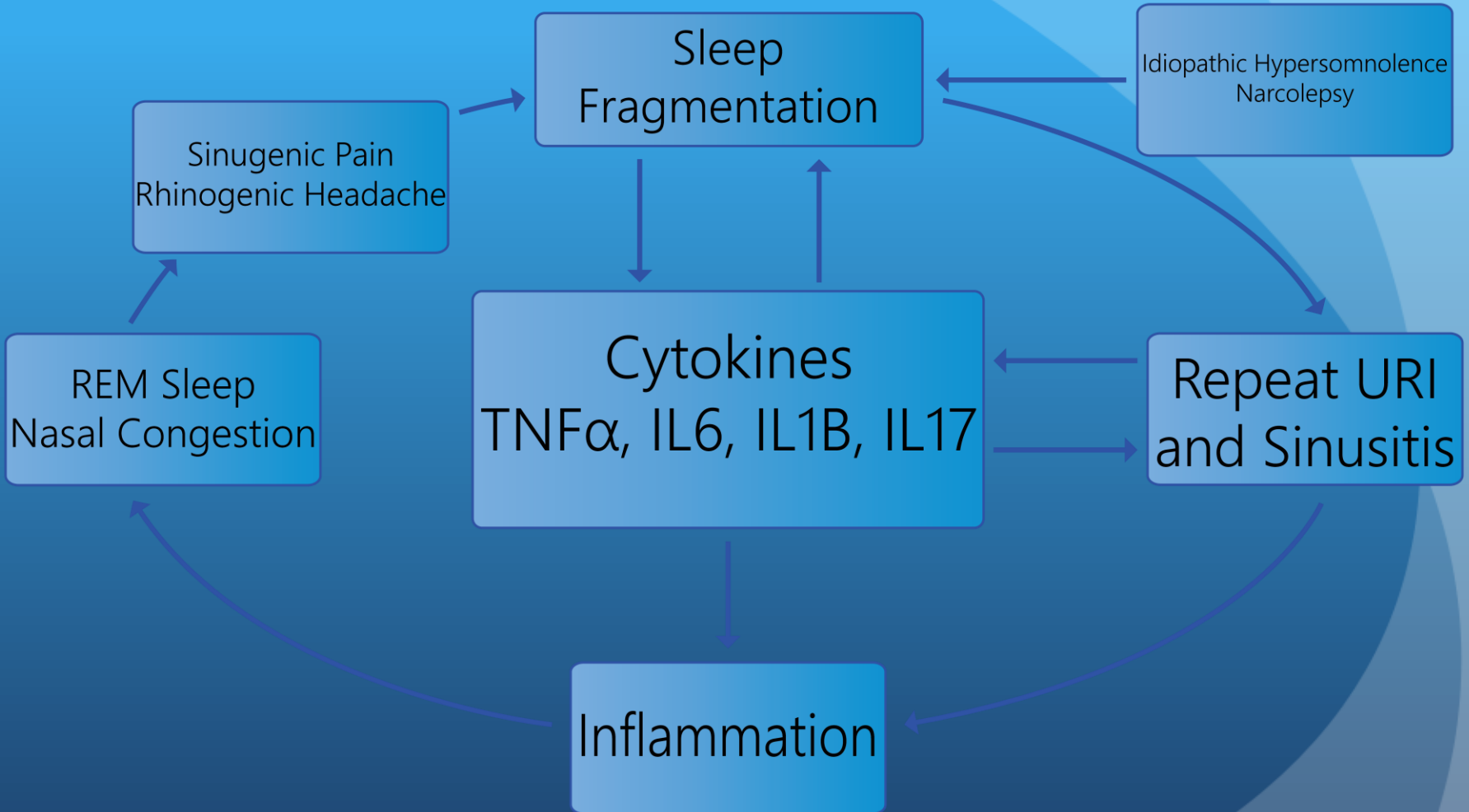
- All SNOT-20 subscore changes statistically significant (  $P < 0.0001$ ) at 52+ weeks post procedure (after multiplicity correction)
- Top ranking changes in fatigue-related symptoms consistent with prior studies of effects of traditional FESS <sup>1,2</sup>

# Index patient

- An engineer with facial pain worse at night, repeat URI, and sinusitis
- Deviated septal spur to the right
- Facial pain worse in the opposite maxillary sinus
- CT scan low Lund McKay score

# Index patient

- Excel spreadsheet of nocturnal pain every 90 minutes in the early night and every 60 minutes early in the morning
- Correlation with REM sleep cycles
- Complete resolution of symptoms and normal SNOT 22 with septoplasty and balloon dilation of the left maxillary sinus





# Sleep disruption chronic pain

- Increase IL1, c-reactive protein, TNF alpha, IL6
- These cytokines increase slow wave sleep and decrease REM sleep

# Pain

- TNF alpha and IL1 increase pain at sinus receptor site

# Sleep fragmentation

- Increases TNF alpha, IL1 and IL6
- Narcolepsy has decreased IGG1 and IGG2
- Idiopathic hyper-somnolence has increased TNF alpha and decreased IGG2

# Polymorphisms

- IL6 subtypes have increased chronic sinusitis
- Subtypes of TNF alpha, IL1B, IL6, and IL8 show differences in severity of influenza

# Sinusitis

- IL1B , IL6 and IL8 are elevated in acute sinusitis

# Decreased immune function

- IgG abnormality in Narcolepsy and idiopathic hypersomnia. Susumu Tanaka
- Some overlap with common variable immune deficiency
- Many patients with low pneumococcal titers, respond to pneumovax

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2. Kaushal N, Ramesh V, Gozal D. TNF $\alpha$  and temporal changes in sleep architecture in mice exposed to sleep fragmentation . *Plos One*. 2012;(9):e45610.
3. Clinton JM; Davis CJ; Jewett KA. Biological regulation on sleep biomarkers . *J Clin Sleep Med* 2011;7(5):Supplement S38-S42.
4. Tanaka S, Honda M. IgG abnormality in narcolepsy and idiopathic hypersomnia. *Plos One*. 2010 Mar 5;5(3):e9555
5. Mullington JM; Simpson NS; Meier-Ewert HK; Haack M. Sleep loss and inflammation. *Best Pract Res Clin Endocrinol Metab*. 2010 October; 24(5): 775-784
6. Min Yg; Lee KS. The role of cytokines in rhinosinusitis .*J Korean Med Sci*. 2000 Jun; 15(3):225-9.
7. Zhang M; Cai C; Chen N; Wang S. The association between genetic polymorphisms of IL-6 and the susceptibility of chronic rhinosinusitis. *Lin Chung ER Bi Yan Hou Tou Jing Wai Ke Za Zhi*. 2012 Mar;26(5):197-200,204.
8. Morales-Garcia G et al. Pandemic influenza A/ H1N1 virus infection and TNF, LTA, IL1B, IL6, IL8, and CCL polymorphisms in Mexican population: a case control study. *BMC Infect Dis*. 2012 Nov 13;12:299

# SNOT 20 domains

- Sinus
- Sleep
- Otology
- Social



# Subtypes of sinusitis

- Lund McKay not correlate to SNOT 20
- SNOT 20 domains
- Immunologic profiles

- ORIGINAL ARTICLE
- **SNOT-22–based clusters in chronic rhinosinusitis without nasal polyposis exhibit distinct endotypic and prognostic differences**
- [Devyani Lal MD Claire Hopkins BM, BCh, MA \(Oxon\), DM, FRCS \(ORL-HNS\)](#)  
[... See all authors](#)
- First published: 13 February 2018  
<https://doi.org/10.1002/alr.22101>  
Cited by:



# Four subgroups

- High scores all domains elevated, elevated eosinophil asthma, inflamed improved the most
- Moderate scores elevated sinus domain
- Moderate scores high in sleep domain, did not show stable improvement
- Mild scores

# Inflammation

- Inflammatory Endotypes and Microbial Association in Chronic Rhinosinusitis , Hoggard et al Front. Immunol, 19 September 2018
- 8 endotypes SC 3 IL-2, IL-4 LL-6, IL 17a, IFN, TNF alpha

# Summary

- Consider sleep disorders in elevated sleep domain on SNOT 22 and in patients with 5 or more servings of caffeine.
- Ask for narcolepsy history or evaluation by a sleep specialist. Most narcoleptics do not have cataplexy
- Evaluate immune status in narcolepsy patients

# Summary

- Look at instructions for SNOT 22
- Consider balloon sinuplasty in high SNOT 22 scores and low Lund McKay scores. Have Haller cells, obstructing concha bullosa, and blocking frontal sinus cells
- Ask about sleep disorders in tinnitus patients, recurrent or atypical mononucleosis
- SNOT 23 . Does your sinus pain disturb your sleep at night? Largest improvement in SNOT 22 with balloon sinuplasty

# Case 4

- 12 old male with chief complaint of dizziness so bad he is in a wheel chair. He gets so dizzy he falls out of his wheel chair. BVDQ is normal ( no double vision or motion sickness) Halpikes for Bppv are normal
- History of vivid dreams, sleep paralysis, and no classical cataplexy
- Orthostatic blood pressures show elevation of blood pressure and heart rate 90- 135 on standing with dizziness and patient falls

# Case 4

- PSG normal, MSLT 2 minutes with 4 Soremp
- Diagnosed with POTS ( positional orthostatic tachycardia syndrome)
- Treated with Concerta and a neuro- chiropractor for pots
- Attacks or Tumarkin



# Narcolepsy and Sinusitis

- Have treated many narcolepsy patients with improvement of their recurrent acute sinusitis and decreased upper respiratory infections
- Most have low pneumococcal titers and respond to pneumovax

# Measurement of Sinusitis

- SNOT 22 added nasal stuffiness and smell taste disturbance- Should you operate on people with low SNOT 22 and low Lund McKay scores?

# Patient Case 1

- 35 yo male presents with repeat sinusitis, bronchitis and 2 pneumonias, 6 caffeine a day
- ROS positive for snoring sleepiness and vivid dreams-dreams during naps, no cataplexy
- PSG OSA, on Pap still sleepy
- Pap MSLT 2 minutes with 4 Soremp
- Labs common variable immune deficiency, low pneumococcal titers

# Patient case 2

- 30 year old driver falling asleep while driving for work, normal BMI, normal stop bang, elevated ESS
- PSG was normal
- MSLT 2 minutes 4 Soremp
- Had allergic reactions sever side effects to all medicines
- On disability

# Case 3

- 15 yo old male out of school for 3 months- sent by psychologist for evaluation of sleep disorder
- Laying on the floor when I entered the exam room
- Played video games until 2 am, slept in late
- Had anxiety, difficulty reading, and motion sickness
- History of remote concussion
- Was treated with Adderall for ADHD with insomnia and palpitations

# Case 5

- 15 yo male presents with a large neck lymph node, very sleepy for 3 months, has acne, vivid dreams and dreams during naps
- CBC with diff atypical lymphocytes, positive monospot
- Plays video games until 2 am, has difficulty getting up for school. Has been out of school for a month
- MSLT 3 min with 3 Soremp
- Treatment Concerta and Trazadone

# Case 6

- 35 yo female with chronic sinus and viral infections, CT sinus normal
- Polysomnogram normal MSLT5 minutes with 1 Soremp
- Low pneumococcal titers
- Treatment Concerta, pneumovax, balloon sinuplasty

# Case 7

- 35 yo female after MVA with TBI treated with prism lenses still dizzy, and difficulty with rehabilitation, 6 caffeine a day, has tinnitus
- Balance testing was normal
- ROS positive for vivid dreaming, sleep paralysis, fatigue thought to be due to brain injury
- Audiogram with bilateral sensory-neural hearing loss
- Tinnitus improved with decreased caffeine



# Case 7

- Diagnosed with post traumatic narcolepsy treated with Concerta and a hearing aid for tinnitus, and therapy has progressed

# Starting Point Questions

- Surgery should not be performed on patients with low Lund McKay CT findings
- Surgery should not be performed on patients with sinus headache
- The eyes do not cause headache or dizziness
- Treating sleep disorders do not improve sinusitis

# Surgery for sinus pain

- ESS performed on 80/211 patients.
- ESS helped 66/80 that had surgery
- Medicines from ENT or neurology helped others for total positive outcome of 90%

# Indications Sinus pain

- Intermittent ostial occlusion
- Uncinate pain
- Barotrauma
- Whistling or squeaking from the sinuses
- Sinus pain that disturbs sleep

# Contraindications sinus pain

- Binocular vision dysfunction
- Chronic daily or rebound headaches



- Identification of Binocular Vision Dysfunction (Vertical Heterophoria) in Traumatic Brain Injury Patients and Effects of Individualized Prismatic Spectacle Lenses in the Treatment of Post-concussive Symptoms: A Retrospective Analysis
- Jennifer E. Doble, MD, Debby L. Feinberg, OD, Mark S. Rosner, MD, Arthur J. Rosner, MD
- *PM R 2010;2:244-253*