

Sleep apnea and Intellectual Disability

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Nov 15, 2010



Epidemiology

- ❑ Sleep disturbances are very common in children: up to 50% in children 0-6 years old, ~25% of all children at some time during childhood.
- ❑ In children with developmental disability – in up to 80%.
- ❑ The most important pediatric sleep disturbance is sleep apnea

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Specific syndromes with sleep apnea



Down Syndrome

- ❑ High prevalence of obstructive sleep apnea in children and adults with Down syndrome.
- ❑ In one study, 97% of non-obese children with DS had OSA, with average AHI of 13.

(Fitzgerald et. al, Arch Dis Child. 2007 May;92(5):423-5)

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Down Syndrome/...

- ❑ Evidence shows that adenotonsillectomy in Down syndrome improves OSA, however not as markedly as in non-Down syndrome patients.
- ❑ 73% of patients in one study required further treatment for OSA after the operation (CPAP, BiPAP or oxygen).

(Shete et. al., Int J Pediatr Otorhinolaryngol. 2010 Mar;74(3):241-4)

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Down syndrome – other sleep disturbances

- ❑ Greater bedtime resistance and sleep anxiety
- ❑ Higher prevalence of parasomnias and night awakenings
- ❑ Shorter duration of REM sleep
- ❑ High prevalence of bruxism

(Carter et. Al, Arch Dis Child. 2009 Apr;94(4):308-10)

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- Clinical vignette

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Prader-Willi syndrome

- Higher prevalence of sleep apnea (up to 100% in 2 studies) in children and adults with PWS compared to healthy controls (corrected for BMI).
- A positive correlation between BMI and sleep hypoxemia.
- Suspected mechanism: primary disorder of ventilatory control

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Prader-willi syndrome/...

And more:

- Excessive daytime sleepiness
- Abnormal sleep architecture: shortened sleep latency and REM latency and shortened NREM sleep stages 3+4 duration.

(Joo et. al, *J Sleep Res.* 2010 Mar;19:248-54,
Lin et. al, *Pediatr Pulmonol.* 2007 Oct;42(10):881-7)

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- ❑ Fig from leaflet
- ❑ Sleepontario.com

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Cranio-vertebral junction anomalies

- ❑ Achondroplasia
- ❑ Arnold- chiary malformation
- ❑ Syringomyelia
- ❑ Down syndrome (atlanto-axial instability)

Brainstem dysfunction can cause both central and obstructive sleep apnea (due to vocal cord paralysis)

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And more...

- ❑ **Rett syndrome** (female autism): central sleep apnea due to seizures
- ❑ **Leigh disease** (mitochondrial disease): central apnea or hypoventilation due to brainstem involvement
- ❑ **Hurler's syndrome** (lysosomal disease): obstructive apnea due to macroglossia

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□ Are you getting bored yet----14

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Known Risk Factors of OSA in Children

BODY

- ⌚ Slow physical development, including stunted growth

BRAIN

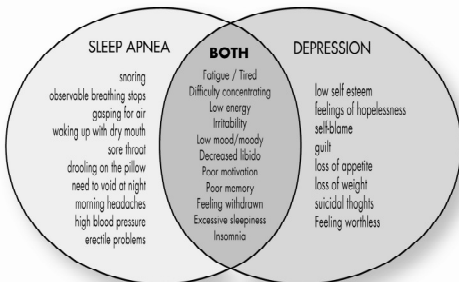
- ⌚ Behavioral problems: hyperactivity, aggression
- ⌚ Difficulties with learning and problem solving, memory problems and inattentiveness
- ⌚ Decreased school performance
- ⌚ Poor school discipline/attendance
- ⌚ Irritability, anxiety, low mood

CARDIOVASCULAR SYSTEM

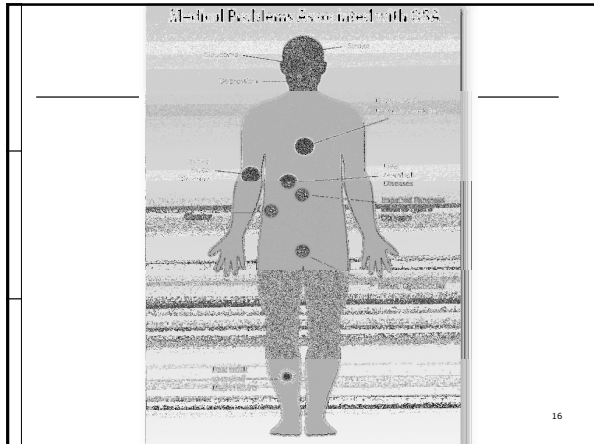
- ⌚ High blood pressure
- ⌚ High pressure in the vessels of the lung
- ⌚ Impaired heart function

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Symptoms



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Sleep Apnea Syndrome, Death and Heart



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Sleep apnea as an independent risk factor for cardiovascular (CV) disease

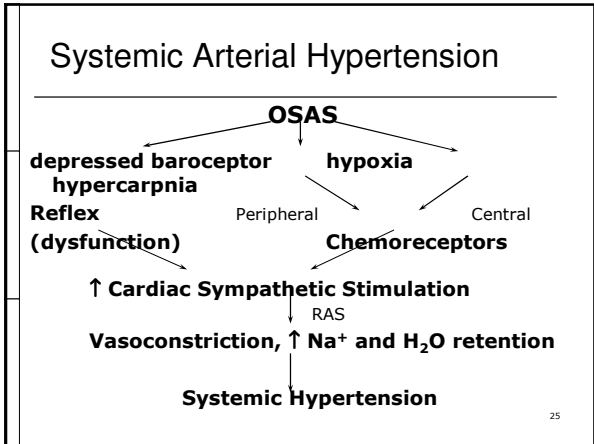
- Systemic arterial hypertension
- Coronary arterial disease
- Stroke
- Heart failure
- Atrial fibrillation
- Sudden cardiac death

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The pathogenesis of CV disease in obstructive sleep apnea (OSA)

- Not completely understood, likely to be multifactorial
- Sympathetic nervous system overactivity (originates from hypoxic or hypercapnic chemoreceptor stimulation)
- Endothelial dysfunction (nitric oxide, NO; or prostacyclin)
- Select activation of inflammatory molecular pathway (C reactive protein, CRP; Proinflammatory cytokines—IL-1 β , IL-6, TNF α)
- Abnormal coagulation
- Metabolic dysregulation—Insulin resistance and disordered lipid metabolism

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Ischaemic Heart Disease (CHD or CAD)

□ **Sleep Apnea as a risk marker in CHD**

OSA and CHD are both frequent in the middle ages, both disease share a similar spectrum or risk factor and attendant disease. OSA are more frequent in patients with CHD (30.6%) in comparison to patients without CHD (21.8%). Patients with CHD and OSA had a significantly higher frequency of a history of Myocardial infarction and a significantly lower left ventricular ejection fraction than patients without OSA (Schafer H, et al. 1996,1999)

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RDI: an independent predictor of mortality in CHD

□ **Conclusion:** untreated OSA is associated with an increased risk of CV mortality in CHD patients. RDI should be taken into consideration when evaluating secondary prevention models in CHD.

Peker Y, Am J Respir Crit Care Med. 2000 Jul; 162 (1): 81~6

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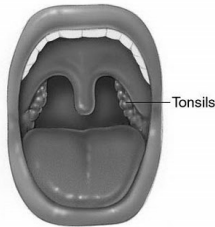
Ischaemic Heart Disease (CHD or CAD)

□ OSA is linked to Sudden death

- A family history of premature CHD and related mortality is an independent risk factor for the development of CHD. The authors hypothesized that OSA is associated with a family history of premature mortality from Ischemic heart disease.
- Methods: 588 subjects underwent PSG, Demographics, Comorbidities, family history of CV disease, ages and causes of death for 10 strata of family members were recorded for all subjects. Those subjects with known causes of Premature Cardiac Death, such as Hypertrophic Cardiomyopathy and Long Q-T Syndrome were excluded. OSA was defined as AHI > 5. Premature CHD mortality was defined as death due to ischemic heart disease or sudden cardiac death before 55 and 65 years of age for men and women respectively

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BIG TONSIL CAUSES BREATHING DISORDER IN CHILDREN (CLINICAL CASE)



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DEFINITION OF OSA IN CHILDREN:

Absence of oronasal airflow for at least 2 breaths with presence of respiratory effort. Associated with hypoxemia (<92%)/EEG arousal cannot be always present. In children an AHI of 1.5 clinically significant.

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HYPOPNEA:

Reduction of oronasal flow >50% for at least 2 breaths and associated with EEG arousal and/or oxygen desaturation $\geq 3\%$.

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OSA is estimated to occur in 1 percent to 3 percent in children. The peak prevalence is at two to six years of age, but it can be seen in neonates to adolescent.

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ETIOLOGY:

- ❑ Adenotonsillar Hypertrophy (most common, there is no relation between tonsils and adenoid size and degree of OSA)
- ❑ Disproportionate growth of oropharynx structure, soft palate, uvula,
- ❑ Nasal aperture stenosis
- ❑ Nasal polyp, micrognathia
- ❑ Nasal septal deviation
- ❑ Inflammation of upper airway muscles
- ❑ Turbinate hypertrophy etc

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SIGNS/SYMPTOMS:

Snoring (3% to 12% children), pause in breathing while asleep, bed wetting, perspiration, sleep in strange positions, learning problem, hyperactivity, developmental problem, failure to thrive or grow, chest retraction during sleep, mouth breathing, etc.

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CLINICAL CASE IN OUR LAB:

3-year-old boy came with his mother to the sleep lab.

□ C/C: mother reported her son has had breathing stops during the night for the last year. He breathes with difficulty and snores loudly, often stops breathing which interrupts his sleep. He wakes up once or twice in the night. He drools on his pillow and wakes up feeling thirsty in the morning. Mother usually puts him to bed around 7pm; it takes him about an hour to fall asleep.

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□ During the day, he has been alert and hyperactive, sometimes he feels tired and sleepy. Sometimes he dozes on the couch. He naps for a few minutes nearly every day.

□ He talks during his sleep, grinds his teeth and sometimes wakes up crying in the middle of the night.

□ Past Psychiatric history: NO

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- ❑ Medical History: He has asthma on treatment
- ❑ Medication:
 - ❑ Ventolin inhaler
 - ❑ Nasocort inhaler
 - ❑ Singular 4mg tab
- ❑ Personal and developmental history: he was born in Canada. His mother had normal pregnancy and normal spontaneous delivery. There is no delay development or milestone. He lives with his mother and step father. He has two brothers (4 years and 1 and half years old). He has no abnormal features.
- ❑ His throat was examined and showed size 3+ tonsils

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THE GRADING OF PALATINE TONSIL HYPERTROPHY WAS SUMMARIZED AS FOLLOWS:

- ❑ 0 – no enlargement;
- ❑ 1 – tonsils occupy less than half of the transverse diameter of oropharynx;
- ❑ 2 – tonsils occupy half of the transverse diameter of oropharynx;
- ❑ 3 – tonsils occupy more than half of the transverse diameter of oropharynx;
- ❑ 4 – tonsils occupy whole of the transverse diameter of oropharynx; i.e., kissing tonsils

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Grade 1+ Tonsil Grade 2+ Tonsil

Grade 3+ Tonsil Grade 4+ Tonsil

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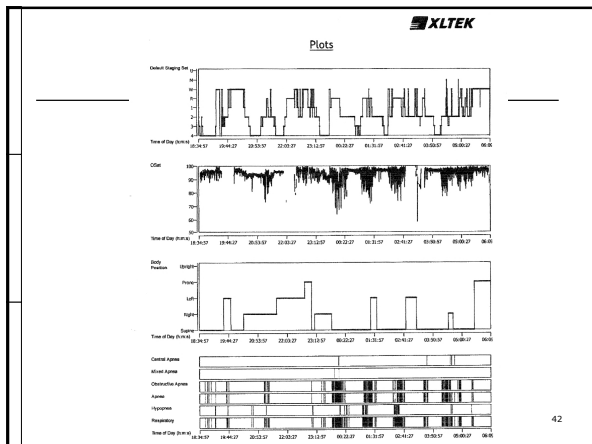
□ Height 3'1" and weight 28.

□ Sleep study was done on the same day (Oct 28, 2008) with pflor/naf but difficult to keep in position of pflor/naf whole night. During night he snored loudly, mouth breathing, desat and abnormal respiratory events noted, teeth grinding, sleep talking, sat up few times and cried, eating candy and waking up early. They left lab early morning. They live in Timmins.

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□ October 29, 2008: scored study urgently and diagnosed SOSA in NREM AHI 16.0/h and worse in REM AHI 98.1/h and TST AHI 29.8/h with desat 64.1% in REM. SE 80.0%, SL 0 min. Dr. Shapiro discussed the study over phone with his father, his family doctor and ENT doctor too within 24 hours of sleep assessment having been done in out lab.

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- His OSA is due to adenotonsillar hyperactivity also known as "enlarged tonsil and adenoid", is one of the main cause of OSA in children.

- November 04, 2009: (surgery day): left tonsillectomy was performed, rt tonsil was excised medializing. Upper pole adenoidectomy was done only because he has a bifid uvula, so lower pole was preserved. Bilateral myringotomies were also performed.

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- Repeat sleep on Feb 07, 2009: S/S report dramatically different to the original study. The comparison is that subsequent to his tonsillectomy AHI of 1/hr and in REM AHI 2.3/hr. mild snored. SL 0 min, SE 94.9%. advised mother to keep track of his ht and wt over the next year and repeat study again in March 2010.

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- S/S on March 15, 2010 with pfl0 and End tidal CO2:

- Height 3'6" and weight 39.7 lbs

- AHI 0/h in both REM and NREM. CO2 range between 35 to 50 (between 45 and 50 of 13.9% of TST – normal limits SL 43.1 min SE 72.1%

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Bariatric Surgery
Obesity related sleep disorder

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Obesity is defined as an excess of adipose tissue

- ❑ 10-25% of teenagers & 20-50% of adults are obese in Canada.
- ❑ Physical examination sufficient to detect excess body fat.
- ❑ Two methods commonly used for qualitative evaluation of obesity.
- ❑ Relative weight (RW)
- ❑ Body mass index (BMI)

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Sleep apnea- a problem associated with obesity

Side effects of sleep apnea are :

- ❑ Loud snoring, Morning headache, Non-refreshing sleep, Dry mouth upon awaking, High blood pressure, irritability
- ❑ Change in personality, Difficulty concentrating, Excessive perspiration.
- ❑ Heart burn, Reduced libido, frequent nocturnal urination, Restless sleep.
- ❑ Nocturnal snoring, gasping, choking.

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Treatment of obesity

- ❑ Conventional technique - multi disciplinary approach to weight loss with hypocaloric diets, behavior modification to change eating behavior, aerobic exercise & social support.
- ❑ Meal replacement diet can also be used effectively and safely to achieve weight loss.
- ❑ Eating behavior - Long term change in eating behavior are required to maintain weight loss.
- ❑ Exercise - Aerobic exercise ↑ daily energy expenditure and essential for long term weight maintenance
- ❑ Medications - amphetamine sometimes prescribed with risk of substance abuse
- ❑ Surgery - a *Last resort* of treatment

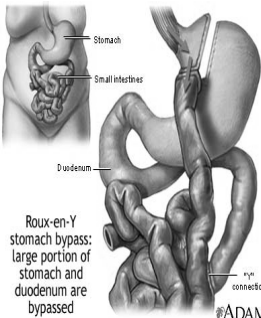
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Surgical treatment of obesity

- ❑ Bariatric surgery - gastric operations are considered the procedure of choice.
- ❑ Most popular is the roux-en-Y gastric bypass (GBP).
- ❑ Sleeve gastrectomy.
- ❑ Gastric Banding

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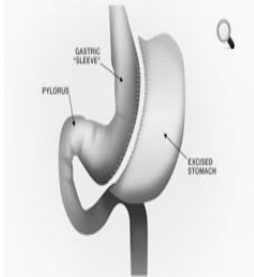
Roux-en-Y Gastric bypass



- ❑ Surgeon creates a small stomach pouch, then constructs a bypass for food. The bypass allows the outlet of the pouch to open into intestine rather than the rest of stomach
- ❑ Having a smaller stomach pouch causes patient to feel full sooner and eat less.
- ❑ Therefore the body cannot absorb excess calories or nutrients .
- ❑ It also reduces appetite helping patient feel satisfied with very small amount of food.

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Gastric sleeve



- Induces weight loss by restriction food intake, patient is going to eat smaller amount
- Surgeon removes 60% of stomach & it take the shape of a tube or sleeve, so patient can continue to eat but in much smaller amounts.
- Hormone that regulates appetite also diminished, causing less desire for food & eating.

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Results of bariatric surgery

Health problems(comorbidity)	Result after surgery
Obstructive sleep apnea	Resolved in 80.4% patients
Depression	Improved in 47% patients
High blood pressure	Resolved in 67.5% patients
High cholesterol	Improved in 94.9% patients
Osteoarthritis	Resolved in 41% patients
Acid reflux	Resolved in 98% patients
Ovulation & fertility	Restored in 100% patients
Stress urinary incontinence	Resolved in 44% of patients
Menstrual dysfunction due to polycystic ovarian syndrome	Resolved in 100% of patients
Type 2 diabetes	Resolved in 83.7% of patients
Patient's initial body weight	Reduced up to 50% according to some studies
Mortality	Low -0.5%, or 1 out of very 200 patients

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Complications of surgery

Complications occur up to 40% of subject undergoing bariatric surgery.

- Complications – peritonitis
- abdominal wall hernia
- staple line disruption
- gallstone
- neuropathy
- marginal ulcer
- stomal stenosis
- wound infection
- thromboembolic disease
- nutritional deficiency
- gastrointestinal symptoms

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Thank you. Questions?



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