Long case

Frédéric Proulx PGY-5

Sainte-Justine Hospital, Montreal

March 15th, 2018

Disclosure

- This is not a rare case

Case presentation

- ♀ 14 year old referred by endocrinology for snoring

Case presentation

♀ 14 year old referred by endocrinology for snoring

- **Past medical history:**
  - Trisomy 21
    - Mild pulmonary stenosis and aortic insufficiency
  - Obesity
  - Asthma
  - Obstructive sleep apnea (AHI 6,8 and OAHI: 4,3) in 2004 (2 years old)

Case presentation

- **Medication:**
  - Fluticasone 100 mcg twice daily, salbutamol as needed
- **Allergy**: None
- **Family history**:
  - Father: Type 2 diabetes, OSA
- **Past surgical history:**
  - T&A at 2 years old

Case presentation

- She is sleeping in front of the respirologist while he is talking to parents

Case presentation

History:

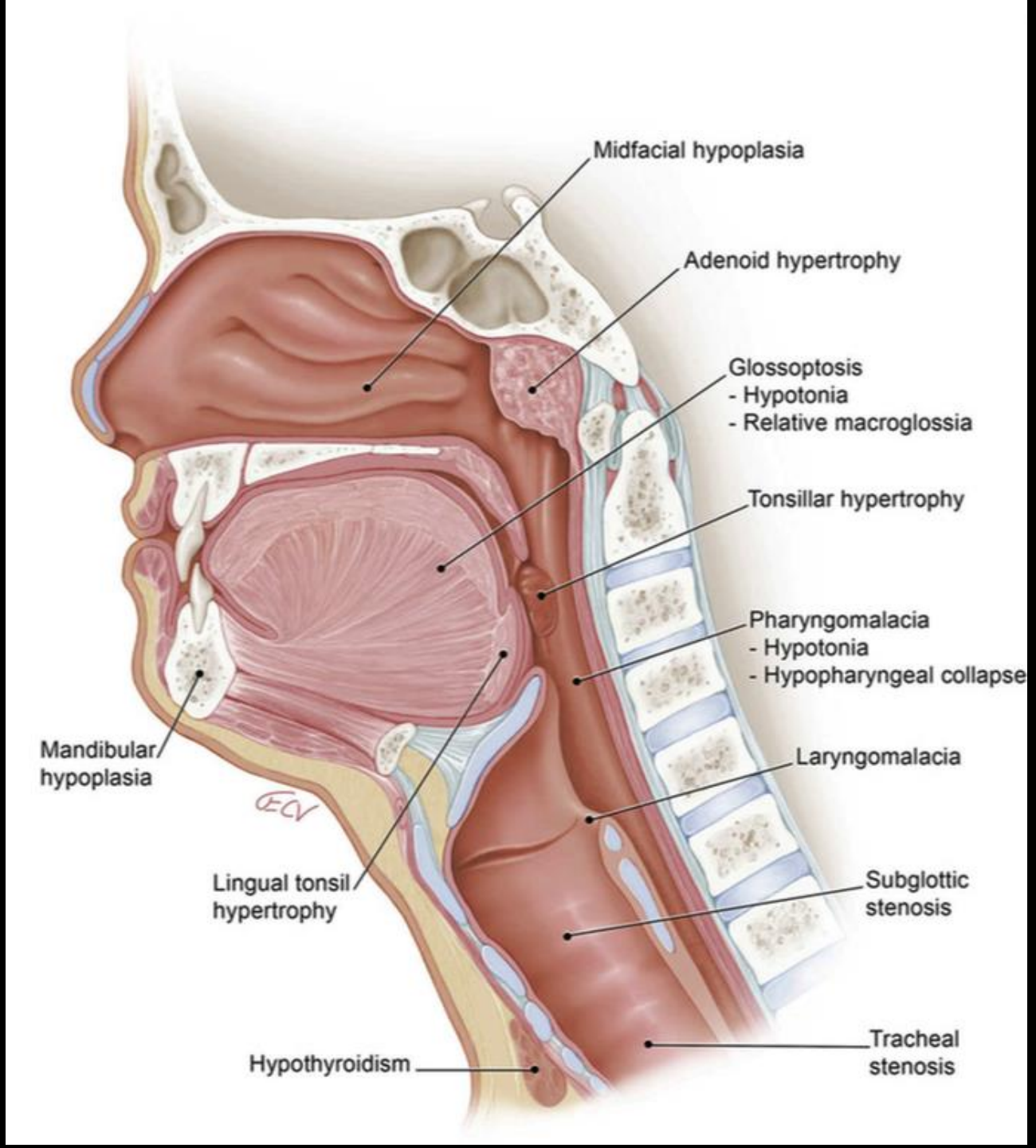
- Snoring all night with frequent gasping episodes and awakenings
- Had witnessed apneas
- Sleeps in seated position
- Agitated sleep
- No behavior problems
- Daytime sleepiness and daytime fatigue

- Parents don't have any private insurance

Case presentation

- **Physical exam**:
- Very calm and cooperative
  - BP: 104/62
  - Weight: 81kg Height: 137 cm BMI: 44
  - Acanthosis nigricans
  - ENT: lingual tonsil hypertrophy, mallampati 4
  - Clear heart and lung sounds are obscured by chest wall obesity
  - No clubbing

What predispose Trisomy 21 to SDB?


Midfacial hypoplasia
Adenoid hypertrophy
Glossoptosis
- Hypotonia
- Relative macroglossia
Tonsillar hypertrophy
Pharyngomalacia
- Hypotonia
- Hypopharyngeal collapse
Laryngomalacia
Subglottic stenosis
Tracheal stenosis
Hypothyroidism
Lingual tonsil hypertrophy
Mandibular hypoplasia

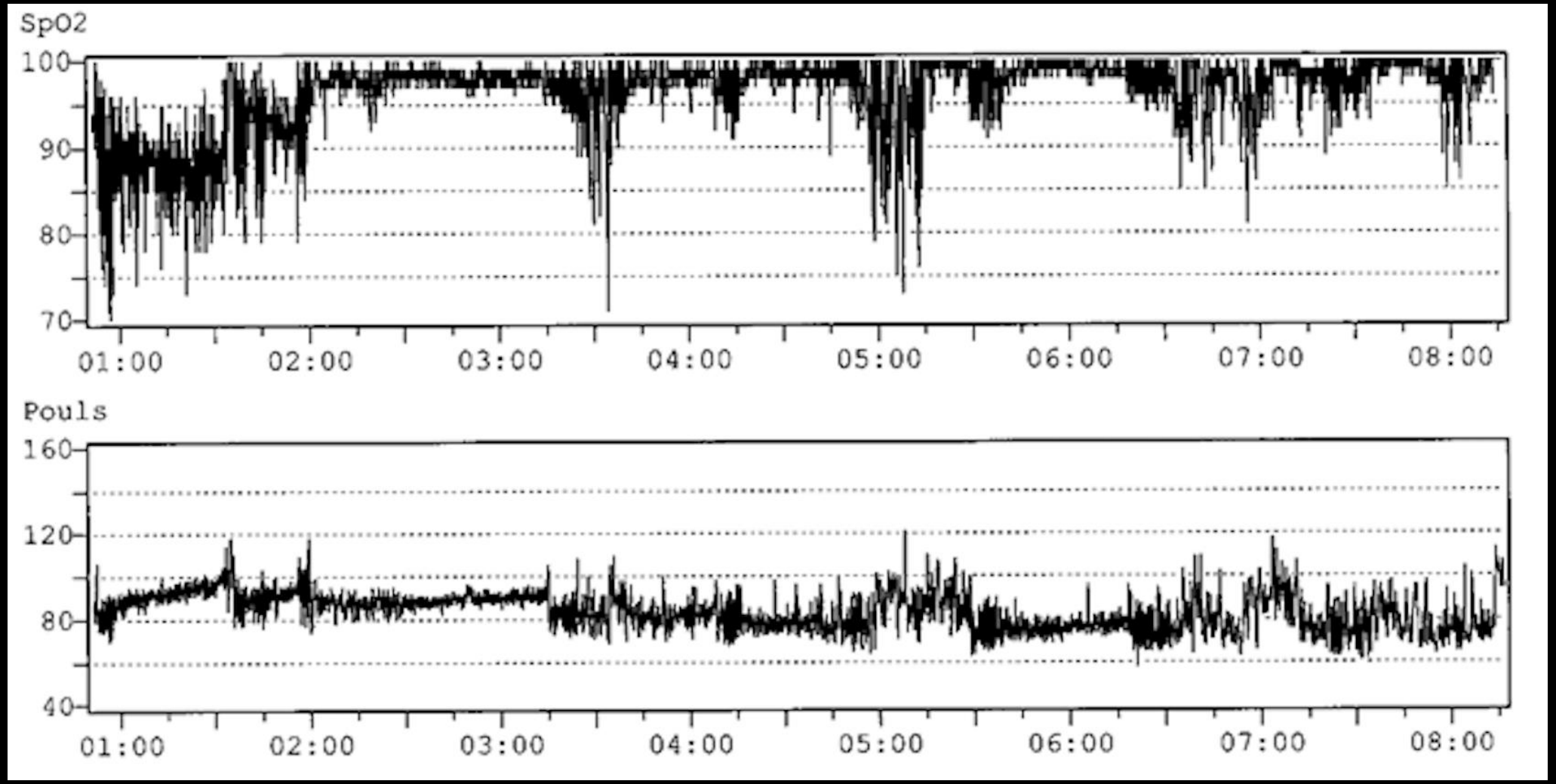
Investigations

February,2013

Overnight home oximetry

Overnight home oximetry

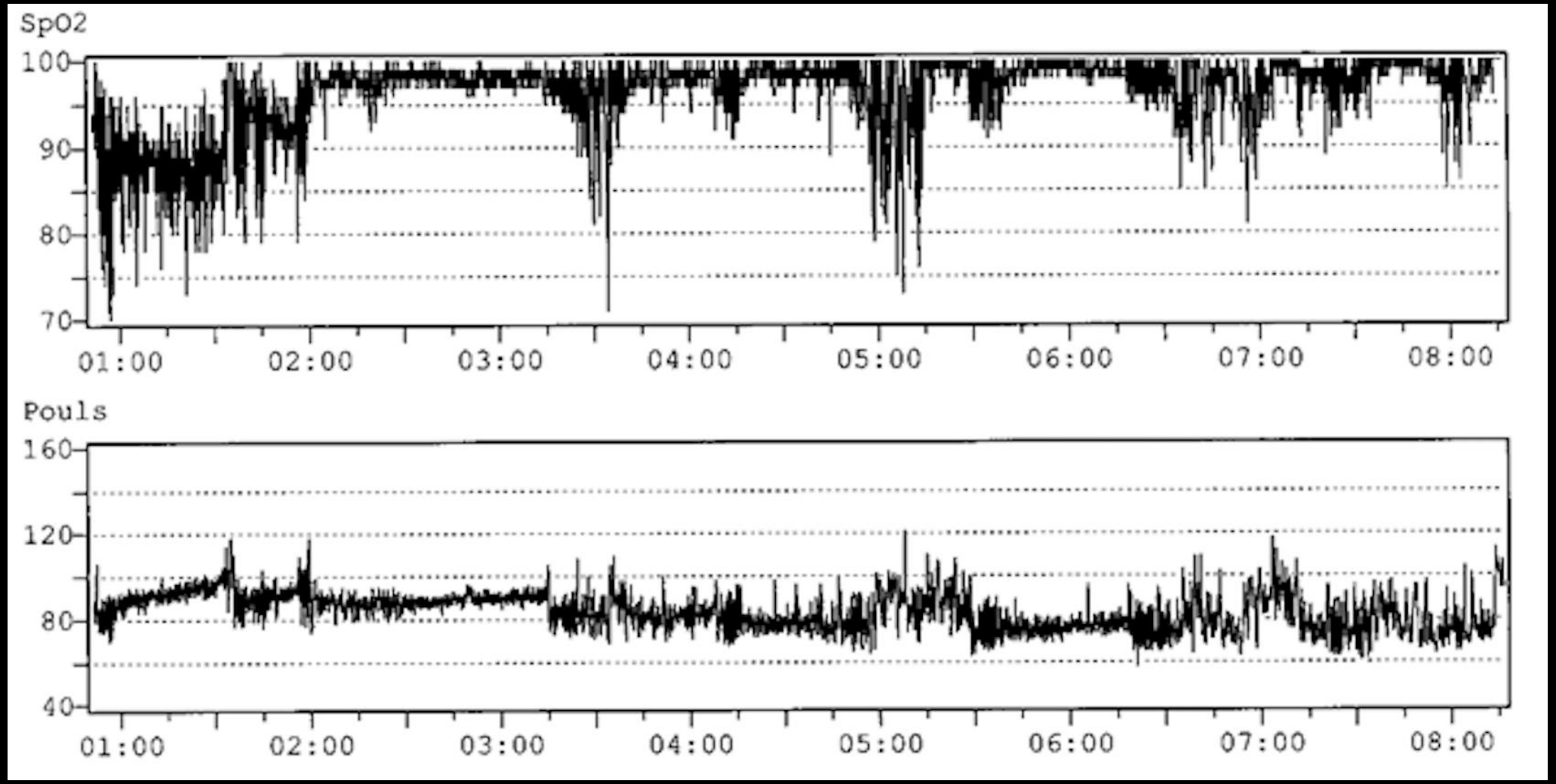
July, 2017



Min SpO2: 70% Mean SpO2: 96,4%
Time with SpO2 ≤ 90% : 11,7%
4 % desaturation index/h (events ≥ 0 sec): 60,9
3% desaturation index/h: 78,9

Overnight home oximetry

July, 2017



Interpretation:

Oximetry suggestive of OSA with cluster of severe desaturations

McGill oximetry score of 4

Elevated baseline heart rate

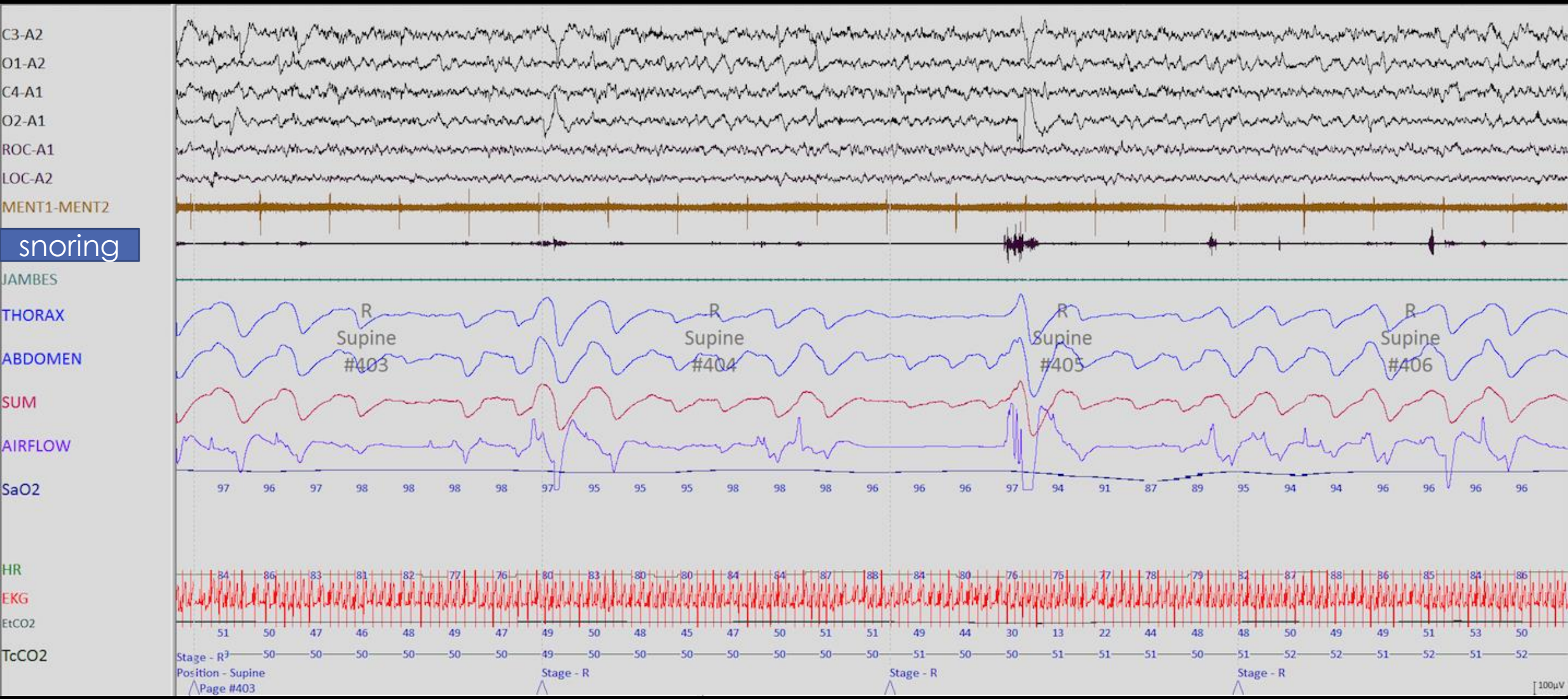
ENT F/U – 1 month later

- On flexible laryngoscopy: adenoids occupy 50% of the space
- Lost 6 kg in the last 3 months
- Rhinitis
- Decreased lingual tonsils
- 'GERD'
- Recommendations:
  - Start on mometasone
  - Continue to loose weight
  - F/U in 3 months

What will be your next steps in evaluation?

PSG

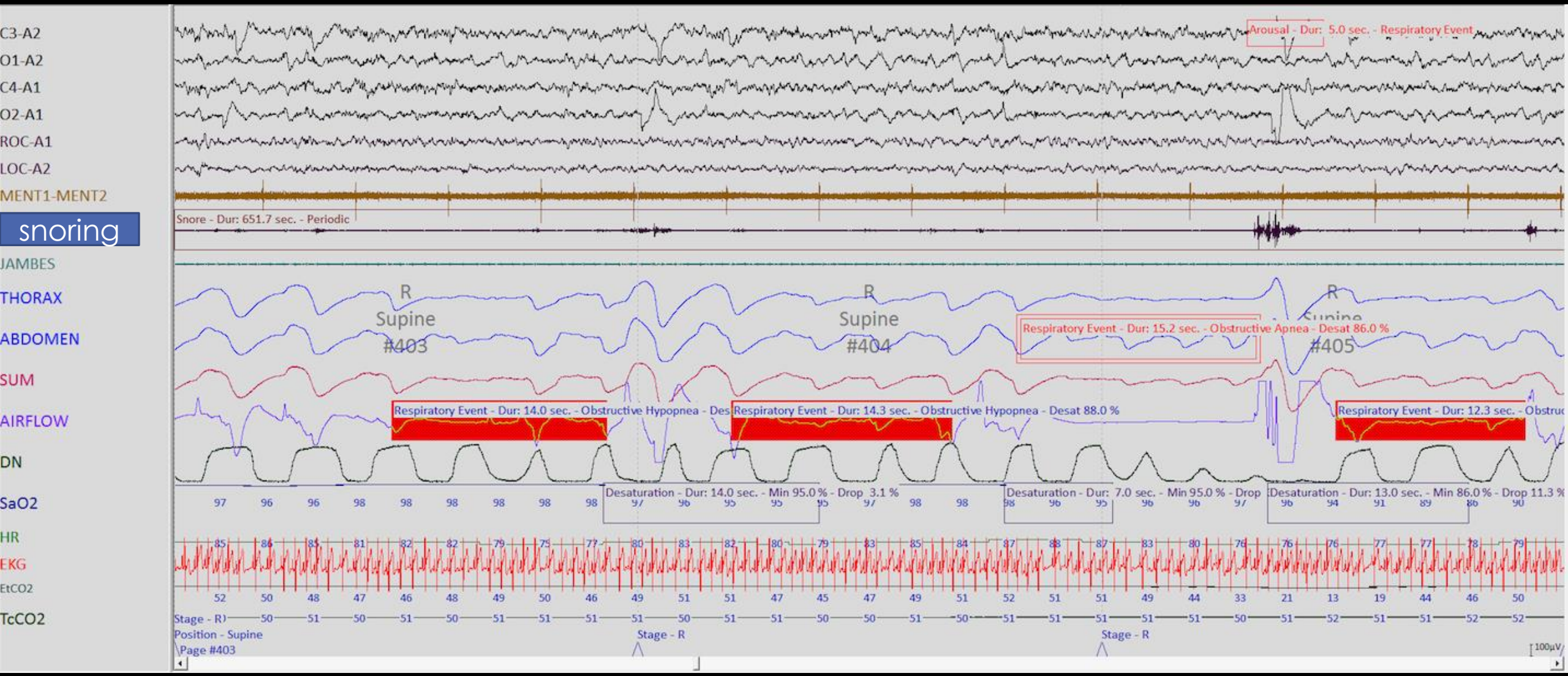
PSG


C3-A2
O1-A2
C4-A1
O2-A1
ROC-A1
LOC-A2
MENT1-MENT2
snoring
JAMBES
THORAX
ABDOMEN
SUM
AIRFLOW
SaO2
HR
EKG
EtCO2
TcCO2
R
Supine
#403
R
Supine
#404
R
Supine
#405
R
Supine
#406
Stage - R
Position - Supine
Page #403
Stage - R
Stage - R
Stage - R
100µV

PSG - Results

- Total sleep time: 408 minutes
- AHI: 22 (OAHI: 21 CAHI: 1)
- 7 central apneas (max 20 sec)
- 60 mixed apneas (max 27 sec)
- 80 obstructive hypopneas (max 48 sec)
- Min desaturation : 81% 0,3% of the time <90%
- Mean saturation : 95,7%
- TcCO2 > 50 mmHg for 26% of the total sleep
- Morning capillary gas: 7,31/52/25

PSG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snoring
JAMBES
THORAX
ABDOMEN
SUM
AIRFLOW
DN
SaO2
HR
EKG
EtCO2
TcCO2
Arousal - Dur: 5.0 sec. - Respiratory Event
Snore - Dur: 651.7 sec. - Periodic
R
Supine
#403
R
Supine
#404
Respiratory Event - Dur: 15.2 sec. - Obstructive Apnea - Desat 86.0 %
#405
Respiratory Event - Dur: 14.0 sec. - Obstructive Hypopnea - Des
Respiratory Event - Dur: 14.3 sec. - Obstructive Hypopnea - Desat 88.0 %
Respiratory Event - Dur: 12.3 sec. - Obstruc
Desaturation - Dur: 14.0 sec. - Min 95.0 % - Drop 3.1 %
Desaturation - Dur: 7.0 sec. - Min 95.0 % - Drop
Desaturation - Dur: 13.0 sec. - Min 86.0 % - Drop 11.3 %
Stage - R
Position - Supine
Page #403
Stage - R
Stage - R
100µV

AASM (2017)

D. Scoring of Apneas

1. Score a respiratory event as an apnea when ALL of the following criteria are met: N1,N2,N3,N4,N5,N6 RECOMMENDED

a. There is a drop in the peak signal excursion by ≥90% of pre-event baseline using an oronasal thermal sensor (diagnostic study), PAP device flow (titration study), or an *alternative* apnea sensor (diagnostic study).

b. The duration of the ≥90% drop in sensor signal lasts at least the minimum duration as specified by obstructive, mixed, or central apnea duration criteria.

c. The event meets respiratory effort criteria for obstructive, central or mixed apnea.

2. Score an apnea as <u>obstructive</u> if it meets apnea criteria for at least the duration of 2 breaths during baseline breathing AND is associated with the presence of respiratory effort throughout the entire period of absent airflow. RECOMMENDED

AASM (2017)

E. Scoring of Hypopneas

1. Score a respiratory event as a hypopnea if ALL of the following criteria are met: N1,N2,N3 RECOMMENDED

a. The peak signal excursions drop by ≥30% of pre-event baseline using nasal pressure (diagnostic study), PAP device flow (titration study) or an *alternative* hypopnea sensor (diagnostic study).
b. The duration of the ≥30% drop in signal excursion lasts for ≥2 breaths.
c. There is a ≥3% oxygen desaturation from pre-event baseline or the event is associated with an arousal.

2. If electing to score obstructive hypopneas, score a hypopnea as obstructive if ANY of the following criteria are met: RECOMMENDED

a. There is snoring during the event.
b. There is increased inspiratory flattening of the nasal pressure or PAP device flow signal compared to baseline breathing.
c. There is an associated thoracoabdominal paradox occurs during the event but not during pre-event breathing.

AASM (2017)

G. Scoring of Hypoventilation

Monitoring hypoventilation in children is recommended during a diagnostic study and optional during a PAP titration study.

1. **Score as hypoventilation during sleep when >25% of the total sleep time as measured by either the arterial PCO_2 or surrogate is spent with a PCO_2 >50 mmHg.**[N1,N2] RECOMMENDED

PSG - Results

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- TcCO2 > 50 mmHg for 26% of the total sleep

Severe OSA with nocturnal hypoventilation

ENT F/U following PSG

- On Naso-pharyngo-laryngoscopy: adenoids occupy 40% of the space
- No sign of GERD
- Decrease lingual tonsils size

ENT F/U following PSG

- On Naso-pharyngo-laryngoscopy: adenoids occupy 40% of the space
- No sign of GERD
- Decrease lingual tonsils size

- No surgical indication at this moment

Trisomy 21 and SDB

Trisomy 21 and SDB

- Trisomy 21: 1 in 770 live births (MSSS 2018)
- OSA is highly prevalent in children with T21 with prevalence rates between 30 to 60% (J.Nehme et al. 2017)
- OSA prevalence in normal developing children ranging from 1-5% (J.Nehme et al. 2017)

Article

Sleep Apnea and Hypoventilation in Patients with Down Syndrome: Analysis of 144 Polysomnogram Studies

Zheng Fan [1,*], Mihye Ahn [2], Heidi L. Roth [1], Leping Li [3] and Bradley V. Vaughn [1]

Children **2017**, *4*, 55; doi:10.3390/children4070055

(Fan z. et al., 2017)

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- Obstructive apnea was positively correlated with age and BMI
  - High obstruction associated with increased age
- Central sleep apnea was associated with younger age (0-3 years)
- Hypoventilation was common occurring in more than 22% of patients
  - Positive correlation between maximum CO2 and BMI

(Fan z. et al., 2017)

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Children **2017**, *4*, 55; doi:10.3390/children4070055

- Patients with T21 have significant sleep related hypoxemia
- Subjects spent an average time of 14,3 min with oxygen saturation <88%
- Patients with T21 have been reported to have prolonged postoperative desaturations.

(Fan z. et al., 2017)

T21 and SDB

- Studies have indicated a high prevalence of sleep disorders in T21
- Bedtime resistance, sleep anxiety, night waking, parasomnias, and daytime sleepiness are commonly reported sleep disorders and behaviors amongst children and adolescents with T21. (Breslin et al. 2011)
- Sleep deprivation in parents with T21, resulting in reduced capacity to manage and maintain positive feelings towards children

Complications of SDB

- Complications in the cardiovascular, metabolic and central nervous systems
- Pulmonary hypertension and *cor pulmonale*
- Cognitive development, behavior, daytime function and quality of life also adversely affected

(Carter et al., 2008)

Health supervision for children with T21 (AAP 2011)

- **Discuss symptoms of OSA with parents at least once during the first 6 months of life:**
  - Heavy breathing, snoring, uncommon sleep positions, frequent night awakening, daytime sleepiness, apneic pauses, and behavior problems that could be associated with poor sleep.

- Refer to a physician with expertise in pediatric sleep disorders for examination and further evaluation of a possible sleep disorder if any of the above mentioned symptoms occur.

Bull and Committee on Genetics, 2011

Health supervision for children with DS (AAP 2011)

- Recommends universal screening for OSA with a polysomnogram (PSG) by **4 years age regardless of symptoms**

- Their evidence to support this recommendation based on 2 small case series
  - Poor correlation between parent report and polysomnogram results
  - Parent reports have been found to underestimate the presence of OSA in children with T21 (Lin et al., 2014)

- One recent review of a large pediatric tertiary care center (Cincinnati), found only 47.7% (455/954) of T21 patients had undergone at least one PSG and 39,1% were diagnosed with a sleep disorder (Esbensen AJ. et al. 2016)

Bull and Committee on Genetics, 2011

American Academy of Otolaryngology-Head and Neck Surgery Foundation 2011

- Clinical practice guideline :
  - Strong recommendation: PSG prior to adenotonsillectomy for these children

- Rationale:
  - Physical examination and history are poor predictors of the presence and severity of OSA
  - Those with severe OSA may require additional preoperative evaluation and a higher level of inpatient monitoring
  - Children with T21 have **higher perioperative risks**

? Higher perioperative risk

- Goldstrein et al. compared the postoperative complications of T&A between 87 T21 children and 64 non-T21 patients.
- The length of hospitalization significantly increased for T21 children (1.6 vs 0.8 days; P=0.001)
- 25% of the T21 children required airway management or PICU admission compared with no children in non-T21 patients (P<.001)
- Respiratory complications requiring intervention were 5 times more likely in T21 patients (P<.001)

(Goldstrein et al., 1998)

? Higher perioperative risk

- Peri-operative complications after adenotonsillectomy in a UK pediatric tertiary referral centre was review from 1627 patients between 2003 and 2010

- The OR of PICU admission in all children with T21 (4/99) versus those with no comorbidities was 5,67 (p=0,025)

(Tweedie D.J. et al. 2012)

? PSG for all at 4 years

Paediatric sleep resources in Canada

- There is a significant lack of resources and services for paediatric sleep medicine care for SDB across Canada
- The demand for polysomnography is 10-fold the capacity
- If only affected children were tested with a PSG – assuming a low prevalence of 1% of OSA.
  - This represent 7.5 times more children than current testing capacity

(Katz et al. 2014)

Time lag from symptom onset to diagnosis

- Diagnosis limited by current resources to obtain PSG
- Lack of simple effective screening tools
- Limited access to sleep specialists
- PSG more expensive, labour-intensive and cumbersome in paediatric patients

(Katz et al. 2014)

TABLE 1
Types of patients seen in sleep clinics and sleep laboratories for assessment of sleep-disordered breathing

	Overall	Sleep clinic	Sleep laboratory
Suspected obstructive sleep apnea needing tonsillectomy and adenoidectomy	45.4	41.1	45.4
Neuromuscular hypoventilation	7.4	7.7	7.4
Central hypoventilation due to brain stem lesion	1.9	2.0	1.9
Invasively ventilated (via tracheostomy)	2.2	2.4	2.2
Obesity	14.2	15.6	14.2
Congenital central hypoventilation syndrome	1.6	1.7	1.6
Down syndrome	9.4	10.3	9.4
Craniofacial abnormality	5.5	6.0	5.5
Continuous positive airway pressure/bilevel therapy titration	2.4	2.6	2.4
Stridor	2.0	2.2	2.0
Cardiac condition	0.5	0.0	0.5
Other: Sleep apnea	7.5	8.2	7.5

Data presented as mean %. In cases in which respondents' answers totaled >100, they were rescaled to add to 100

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(Katz et al. 2014)

How to prioritize who will get a PSG?

How should we prioritize

Predicting the presence of sleep-disordered breathing in children with Down syndrome

Joy Nehme [a], Robert LaBerge [a, b], Mary Pothos [a, c], Nick Barrowman [a, c], Lynda Hoey [a], Andrea Monsour [a], Madelaine Kukko [a], Sherri Lynne Katz [a, b, d, *]

[a] *Children's Hospital of Eastern Ontario, Ottawa, Ontario, Canada*
[b] *Department of Pediatrics, Faculty of Medicine, University of Ottawa, Ottawa, Ontario, Canada*
[c] *Department of Pediatrics, Faculty of Medicine, Ottawa, Ontario, Canada*
[d] *Children's Hospital of Eastern Ontario Research Institute, Ottawa, Ontario, Canada*

Article history:
Received 20 January 2017
Received in revised form
10 March 2017
Accepted 11 March 2017
Available online 31 May 2017

Predicting the presence of SDB among T21

- Physicians most often refer children with T21 for PSG testing when the children report snoring
- Neither the presence of snoring nor the number of nights/week with snoring was associated with an increased prevalence of SDB
- «Struggling to breath at night» was significantly associated with AHI >5 (p=0.05)
- Obesity was not significantly associated with AHI > 5 or OAHI > 5

Predicting the presence of SDB among T21

- Some symptoms may mimic those of SDB
- In their reference population, parent-reported mouth breathing and GERD comorbidity were inverse predictors of SDB
- None of the clinical symptoms investigated were predictive of SDB and OSA
- The absence of symptoms was still associated with a high prevalence of SDB

Screening

Screening: Parent perception

- In a 5-year longitudinal study in which the ENT problems seen in T21 were evaluated, parental history was unable to predict PSG findings
- 54% (13 of 24) had no OSA symptoms and their PSG results were abnormal
- 64% (7 of 11) with OSA symptoms had normal PSG results

(Shott et al., 2006)

Accuracy of Parental Perception of Nighttime Breathing in Children with Down Syndrome

Norman R. Friedman, MD[1], Amanda G. Ruiz[1], Dexiang Gao, PhD[2], and David G. Ingram, MD[3]

Otolaryngology–
Head and Neck Surgery
2018, Vol. 158(2) 364–367

Study aimed to assess parent's accuracy of their children's breathing patterns as compared with PSGs in a cohort of children with T21 (113)

Parents were unable to predict the presence or absence of OSA by nighttime symptoms (P=0.60)

The risk of OSA for children with frequent symptoms versus those with infrequent symptoms was 1.04 (95% CI, 0.89- 1.3)

Parents of T21 children are unable to predict the presence of absence of OSA, nor are they able to determine its severity

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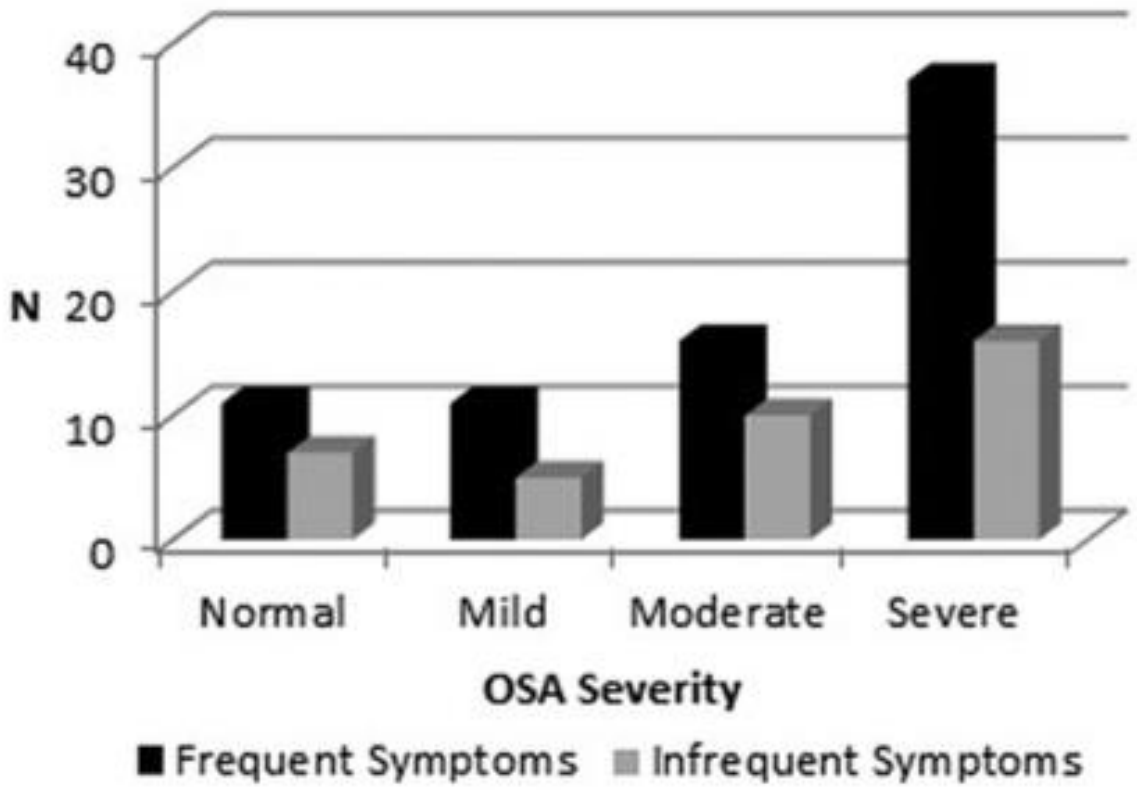

Figure 1. Obstructive sleep apnea (OSA) severity by symptom frequency.

Screening: Oximetry

Overnight Pulse Oximetry for Evaluation of Sleep Apnea among Children with Trisomy 21

Andrea M. Coverstone, M.D.[1]; Merielle Bird, M.S.N.[2]; Melissa Sicard, M.S.N.[1]; Yu Tao, M.B., M.S.[3]; Dorothy K. Grange, M.D.[1]; Claudia Cleveland[2]; David Molter, M.D.[4]; James S. Kemp, M.D.[1]

[1]Department of Pediatrics, Washington University School of Medicine, St. Louis, MO; [2]Sleep Medicine, St. Louis Children's Hospital, St. Louis, MO; [3]Department of Biostatistics, Washington University School of Medicine, St. Louis, MO; [4]Department of Otolaryngology/Head and Neck Surgery, Washington University School of Medicine, St. Louis, MO

Journal of Clinical Sleep Medicine, Vol. 10, No. 12, 2014

- Retrospective cohort of 119 children with T21 who had PSG done to evaluate OSDB and also had PSG oximetry results

Screening: Oximetry

Table 2—Median Age, BMI, and PSG Results for McGill Groups 1-4.

	McGill 1 (n = 59)	McGill 2 (n = 43)	McGill 3 (n = 6)	McGill 4 (n = 11)	p value
Age (months)	65.0	73.0	95.5	95.0	0.52
Body mass index	17.0	18.5	24.4	26.9	< 0.001
Total sleep time (min)	423.0	437.5	360.5	392.0	0.08
AHI (events/h)	2.8	6.5	15.1	23.9	< 0.001
OAHI (events/h)	1.0	4.5	11.8	19.8	< 0.001
CAI (events/h)	0.9	1.6	3.9	1.6	0.03
Lowest SpO_2 (%)	90	85	78	69	< 0.001

AHI, apnea-hypopnea index; OAHI, obstructive AHI; CAI, central apnea index.

(Coverstone A. M. et al., 2014)

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Screening: Oximetry

Specificity and positive predictive value of McGill score for obstructive sleep disordered breathing

- Using OAHI ≥ 2.5 as indicative of abnormality and McGill Scores of 3 or 4 as a positive test
- Oximetry score was 98% specific with a positive predictive value of 94% for obstructive SDB

Screening: Oximetry

- **Based on these results, early ENT referral for surgery :**
  - McGill Score of 3 or 4
  - McGill score of 2, with elevated BMI or AT hypertrophy

- **McGill score of 1 or 2, without associated obesity or AT hypertrophy:**
  - likely require full PSG

(Coverstone A. M. et al., 2014)

Outcome of T&A among T21 patients

Systematic Review

Adenotonsillectomy Outcomes in Patients With Down Syndrome and Obstructive Sleep Apnea

Zachary Farhood, MD; Jonathan W. Isley, MS; Adrian A. Ong, MD; Shaun A. Nguyen, MD, MA; Terence J. Camilon; Angela C. LaRosa, MD; David R. White, MD

Laryngoscope, 127:1465–1470, 2017

- Can decrease AHI by more than 50% in children with T21and OSA (Nation J., et al. 2017)
- Up to 80% of T21 patients have persistent OSA after adenotonsillectomy
- Given the multifactorial nature and complexity of OSA in children with DS, monotherapy with T&A is likely insufficient to cure these patients

Systematic Review

Adenotonsillectomy Outcomes in Patients With Down Syndrome and Obstructive Sleep Apnea

Zachary Farhood, MD; Jonathan W. Isley, MS; Adrian A. Ong, MD; Shaun A. Nguyen, MD, MA; Terence J. Camilon; Angela C. LaRosa, MD; David R. White, MD

Laryngoscope, 127:1465–1470, 2017

- Preoperative characteristics, such as BMI, tonsil size, and OSA severity were not found to correlate with change in AHI
- Existing evidence is heterogeneous but suggest that T&A does improve OSA in this population and can be recommended as a first-line treatment
- They recommend that all children with T21and OSA who undergo T&A should have a postoperative PSG to quantify the degree of residual disease

What to do when T&A fails?

The Laryngoscope

Down Syndrome and Pediatric Obstructive Sleep Apnea Surgery: A National Cohort

Adrian A. Ong, MD; Carlyn M. Atwood, BS; Shaun A. Nguyen, MD, FAPCR; Ronald J. Teufel II, MD; Chitra Lal, MD; Angela C. LaRosa, MD; David R. White, MD

From 1997 to 2012, a total of 48,301 and 2,991 sleep surgeries were identified in a non-syndromic and T21 groups, respectively

T&A the most common procedure in both groups

Proportion of tonsillectomy with adenoidectomy decreased over time ($P<.01$)
Proportion of palatal surgery and tracheostomy also decreased significantly

Increase in proportion of lingual tonsillectomies, tongue-base reduction procedures, and supraglottoplasties performed in both groups over time (higher in T21 groups)

What's next when T&A fail? Lingual tonsillectomy

Polysomnographic Outcomes Following Lingual Tonsillectomy for Persistent Obstructive Sleep Apnea in Down Syndrome

J. Drew Prosser, MD; Sally R. Shott, MD; Oscar Rodriguez, MD; Narong Simakajornboon, MD; Jareen Meinzen-Derr, PhD; Stacey L. Ishman, MD, MPH

***Laryngoscope*, 127:520–524, 2017**

- 21 patients included

What's next when T&A fail? Lingual tonsillectomy

- The median AHI was 9.1 events/hour (3.8 to 43.8 events/hour) before surgery and 3.7 (0.5 to 24.4 events/hour)after surgery
- The median improvement in overall AHI and the oAHI were 5.1 events/hour (-2,9 to 41 events/hour) and 5.3 events/hour (-2,9 to 41 events/hour), respectively (P<.0001)
- After the surgery:
  - 19% had complete resolution of OSA
  - 28.5% had moderate OSA defined as 5 to <10/h
  - 14% had severe OSA defined ≥ 10/h

When T&A fail?
Continuous positive airway pressure

When T&A fail?
Continuous positive airway pressure

- It is less clear whether CPAP is both feasible or beneficial in this patient population that already has high level comorbidities and intellectual disability and require a high burden of care

- Although CPAP is consider the mainstay of nonsurgical management of OSA, we might be reluctant to start it in this population patient

Correlates of Pediatric CPAP Adherence

Stephen M.M. Hawkins, MD[1,2]; Emily L. Jensen, BS[3,4]; Stacey L. Simon, PhD[1,2]; Norman R. Friedman, MD[3,4]

[1]Department of Pediatric Pulmonology, University of Colorado School of Medicine, Aurora, CO; [2]The Breathing Institute, Children's Hospital Colorado, Aurora, CO; [3]Department of Otolaryngology, University of Colorado School of Medicine, Aurora, CO; [4]Department of Pediatric Otolaryngology, Children's Hospital Colorado, Aurora, CO

J Clin Sleep Med 2016;12(6):879–884.

- From 140 pediatric OSA patients, who had a documented in-laboratory titration with adherence data available:

Table 3—Study population CPAP use.

	Adherent (n = 69)	Non-Adherent (n = 71)
Hours per night	7.4 (5.5–9.3)	1.7 (0.3–3.1)
% nights	94 (86.2–101.7)	52 (24.9–78.1)

Adherence is defined as an average of both > 4 h per night and use on > 70% of nights. Values presented as mean (mean − 1 SD–mean + 1 SD). Total n = 140.

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- Female sex was associated with adherence (60,9% vs 39,5% of males, OR=2,41, 95%CI=1.20-4.85;p=0,01)
- No differences in adherence were found by patient age, ethnicity, or insurance status
- Severity of OSA, therapeutic pressure, and residual AHI did not impact CPAP adherence (p>0.05)
- A diagnosis of developmental delay was associated with CPAP adherence (OR=2.55, 95% CI =1.27-5.13; p=0.007)

SCIENTIFIC INVESTIGATIONS

Correlates of Pediatric CPAP Adherence

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- Broad spectrum of developmental delay (mild learning disability or speech delay to global developmental delay or profound intellectual disability)

- **Potential reasons:**
  - Increased dependence upon caregiver support
  - Decreased ability to remove mask due to either physical or intellectual disability
  - Increase parental perception of CPAP necessity for a child with neurodevelopmental disabilities

- Children with developmental delays in addition to OSA are an at-risk population and are expected to benefit from therapeutic intervention

OSA and cognition

Cognitive impairment, T21 and OSA: A triple threat

- Studies have found that decreased slow-wave sleep is seen in children and adolescents with T21 who have OSA

- OSA might explain some of the variability associated with verbal IQ, memory, and executive function in a T21 community cohort

(Chitra Lal et al., 2015)

OSA and cognition in T21

- The presence of OSA in individuals with T21 can result :
  - Poorer school performance
  - Limited social interactions,
  - Impairment in activities of daily living
  - Significantly decreasing quality of life

(Chitra Lal et al., 2015)

OSA and cognition in T21

- Patients with T21 are particularly predisposed to developing early-onset Alzheimer disease after the age of 35 years (Fernandez F. et al., 2013)
- Oxidative stress has been implicated in the pathogenesis of premature AD in patients with T21 (Muchova J, et al., 2014)
- Oxidative stress is also a major player in OSA, with the intermittent, episodic hypoxemia and hypoxia-reperfusion injury associated with it (Lavie L, et al., 2003)

(Chitra Lal et al., 2015)

Treatment effects on cognition

Review

Neurocognitive and Behavioural Outcomes Following Intervention for Obstructive Sleep Apnoea Syndrome in Children

Sofia Konstantinopoulou MD[1], Ignacio E. Tapia MD, MS[1,*]

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Paediatric Respiratory Reviews 20 (2016) 51–54

Treatment effects on cognition

- Although CPAP is now being used commonly in children, only a handful of studies have evaluated its efficacy regarding neurocognitive and behavioral outcomes

(Konstantinopoulou S. et al., 2016)

Treatment effects on cognition

- PAP use was associated with significant improvements in attention deficits, sleepiness, behavioral and caregiver and child reported quality of life, after 3 months of use

- A significant correlation between improvement and PAP adherence was found only with regards to daytime sleepiness, but not between other behavioral outcomes and adherence

(Marcus et al., 2012)

Back to our case

Back to our case – 4 months later

- CPAP titration was done on an outpatient basis

CPAP titration

CPAP	Sleep (min)	OA	CA	Hyp	AHI (/h)	SpO2 Min%	SpO2 Max%	TcCO2 mmHg
4cm H2O	12,8	7	0	7	37,5	88	98,5	50-54
5cm H2O	5,1	0	0	0	11,7	89	94,2	50-54
6cm H2O	17,6	2	0	2	13,7	91	94,4	50-54
7cm H2O	11,5	0	0	5	26	91	94,8	50-54
8cm H2O	16,9	0	0	3	10,6	93	95,2	50-54
9cm H2O	10,3	1	0	3	23,2	93	95,2	50-54
10cmH2O	21,5	4	0	8	22,3	92	96,8	50-54
11cmH2O	117,1	3	1	4	4,6	92	97,7	39-44
12cmH2O	174,6	0	0	0	0,3	93	97,8	36-42

CPAP titration

CPAP	Sleep (min)	OA	CA	Hyp	AHI (/h)	SpO2 Min%	SpO2 Max%	TcCO2 mmHg
4cm H2O	12,8	7	0	7	37,5	88	98,5	50-54
5cm H2O	5,1	0	0	0	11,7	89	94,2	50-54
6cm H2O	17,6	2	0	2	13,7	91	94,4	50-54
7cm H2O	11,5	0	0	5	26	91	94,8	50-54
8cm H2O	16,9	0	0	3	10,6	93	95,2	50-54
9cm H2O	10,3	1	0	3	23,2	93	95,2	50-54
10cmH2O	21,5	4	0	8	22,3	92	96,8	50-54
11cmH2O	117,1	3	1	4	4,6	92	97,7	39-44
12cmH2O	174,6	0	0	0	0,3	93	97,8	36-42

Clear improvement of obstructive events and resolution of nocturnal hypoventilation with use of CPAP peep 12 cmH2O

Respirology f/u – Morning after titration

- She's now awake seating in front of the respirologist
- She slept well
- The mask was well tolerated
- Parents already appreciated a difference on energy level

- CPAP could be obtained later through a donation

Challenges

- Cost
- Coverage of CPAP in QC
  - Only by private insurance for the time being
- Bipap cover by National Program for Home Ventilatory Assistance
  - Failure of CPAP must be demonstrated
- Tolerance – Introduction as outpatient or inpatient?

Conclusion

- Children with Trisomy 21 have a higher OSA incidence (30-60%)
- Increased prevalence is due to multiple factors
- T&A is administered as a first-line treatment for OSA but often is not sufficient
- T21 should be monitor closely for recurrence of SDB
- May require further surgical assessment and/or treatment
- Giving the fact that OSA cause multiples complications and have an impact on cognition the treatment is of paramount importance

Discussion

- Beside T&A what is the trend in your surgical management for those patient in your center?
- How do you initiate CPAP among those patient?

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