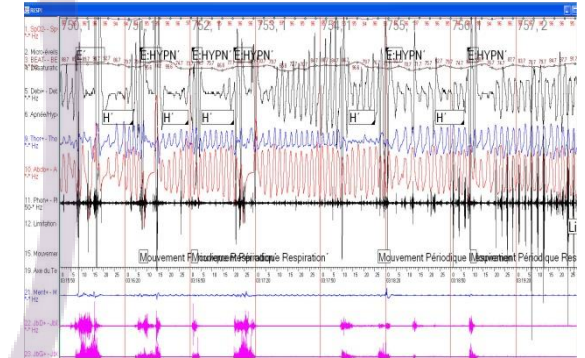


Dento-facial aspects of paediatric Obstructive Sleep Apnea-Hypopnea Syndrome (OSAHS) : Diagnosis and Management strategies



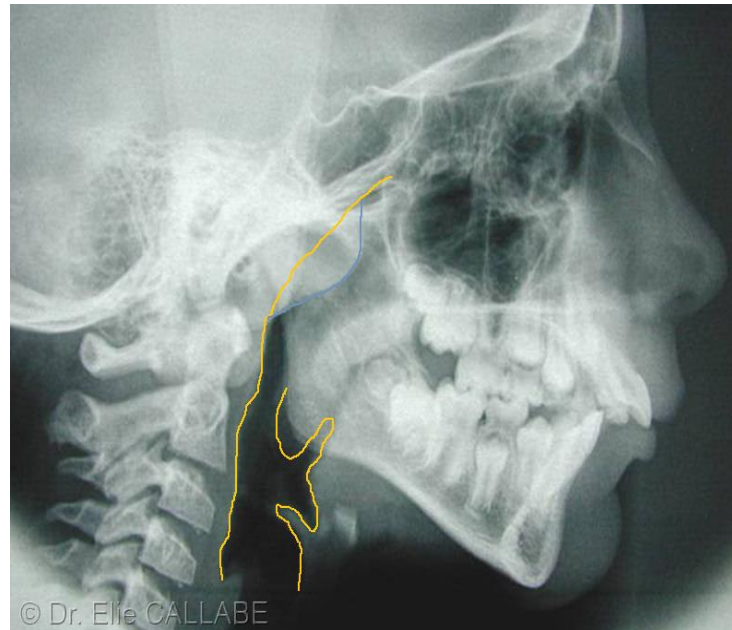
F CLAUSS, B GROLLEMUND, D WAGNER, U KILLIC-HUCK, J TEULADE, M-C MANIÈRE



DENTAL FACULTY OF STRASBOURG

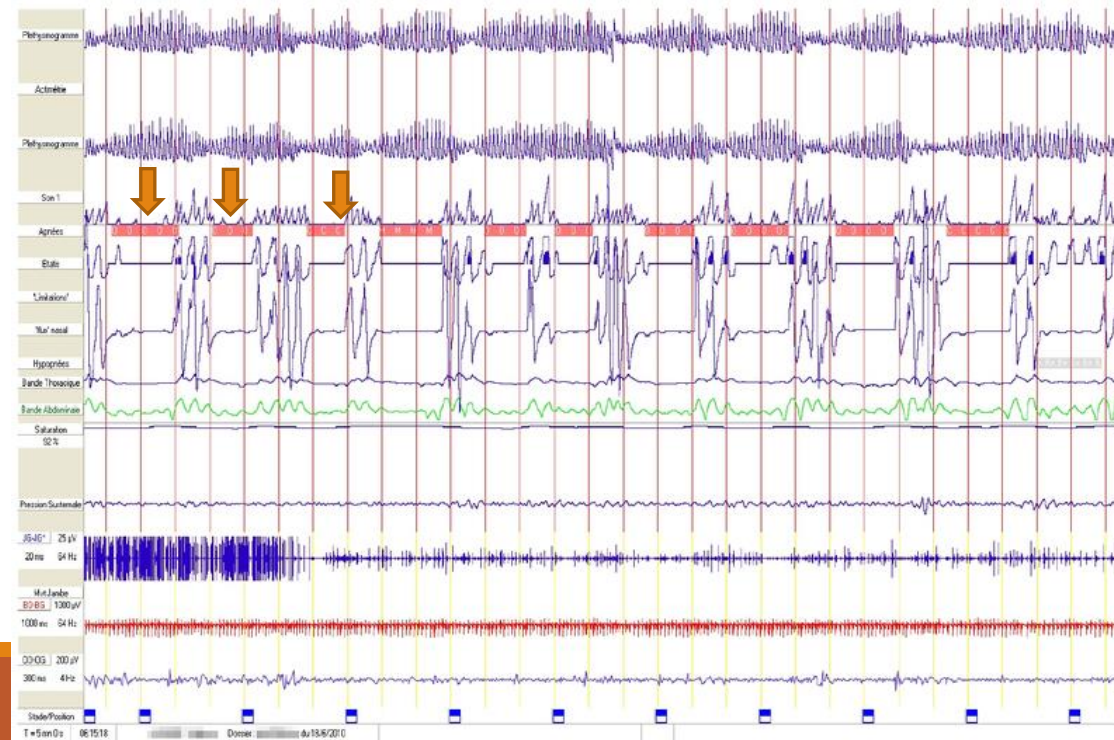
REFERENCE CENTER FOR DENTAL MANIFESTATIONS OF RARE DISEASE ORARES

Definition of paediatric OSAHS and classification



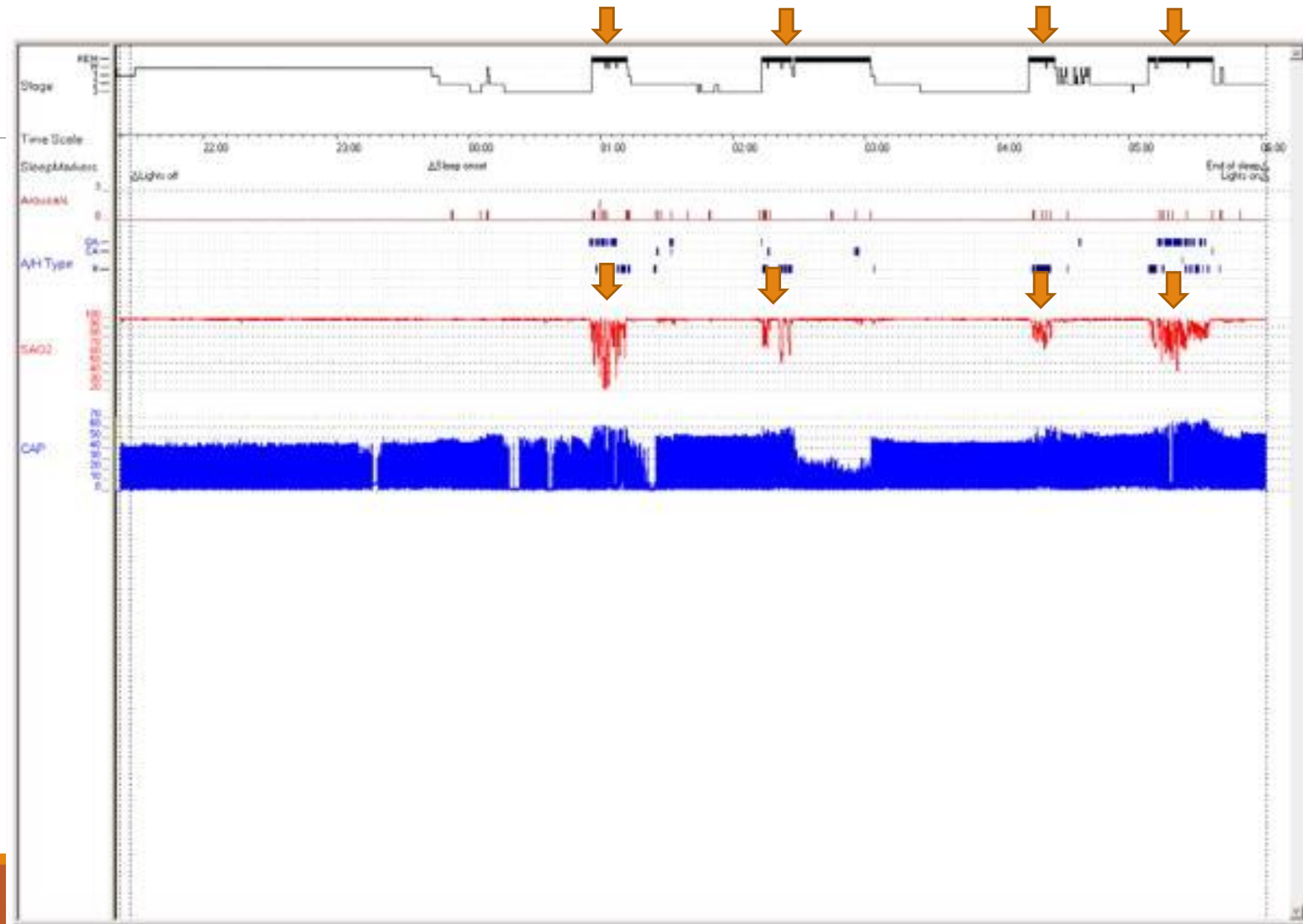
Definition and classification of OSAHS

- **OSAHS** : pathological events of temporary interruption of breathing during sleep with variable severity and chronic evolution
- Loco-regional and systemic major consequences (neuro-developemental, cardiac, metabolic..)
- Condition often linked to a chronic pharyngeal obstruction

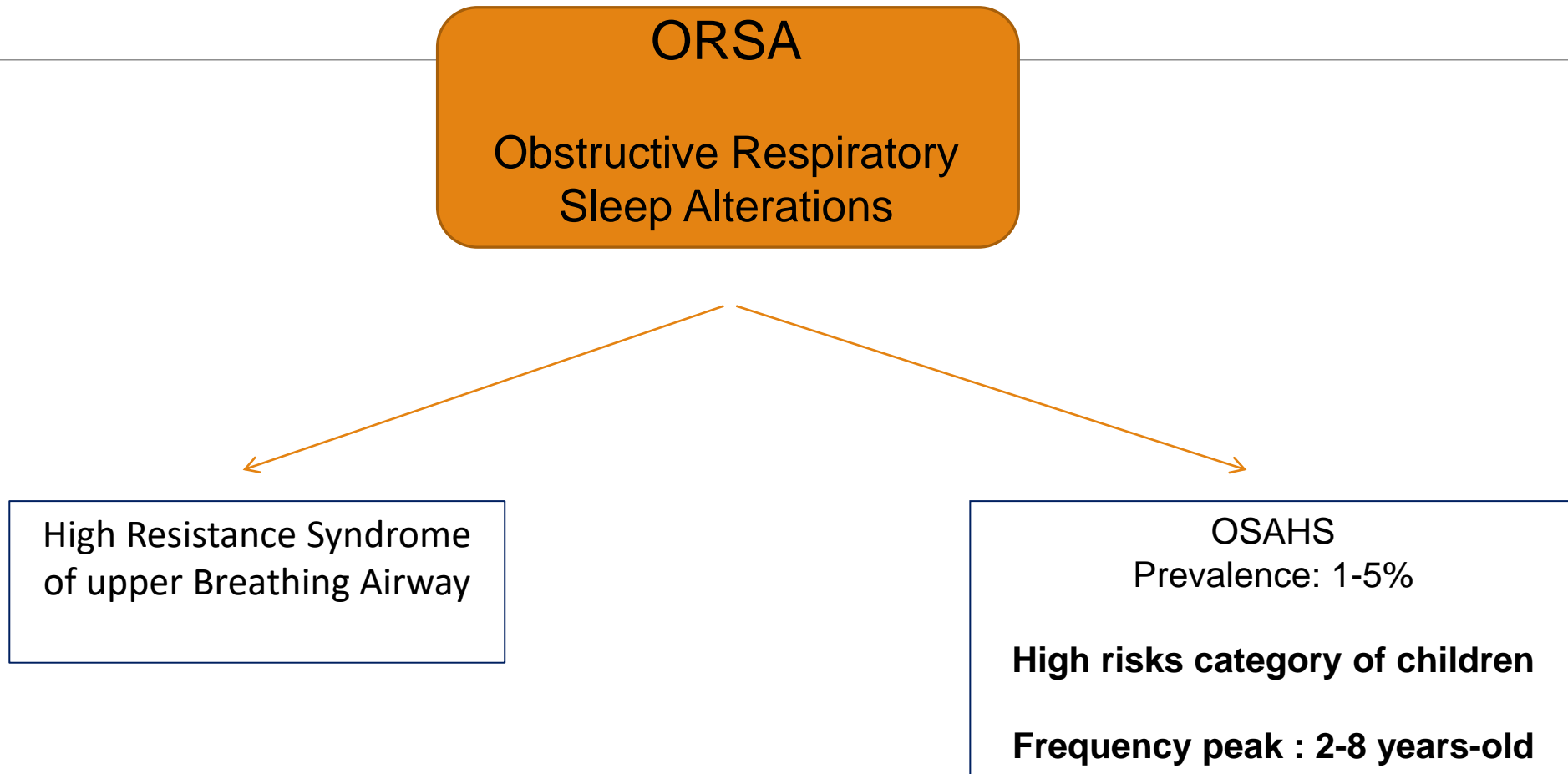


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Gold standard : polysomnographic diagnosis

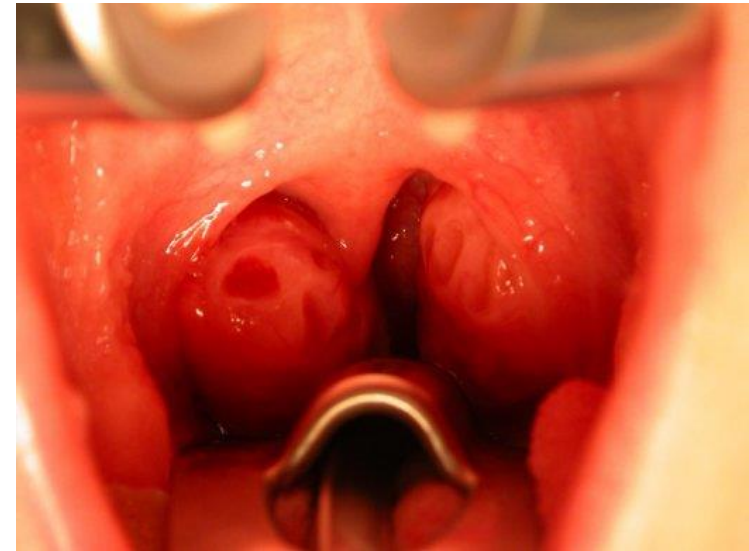


Definition and classification of OSAHS



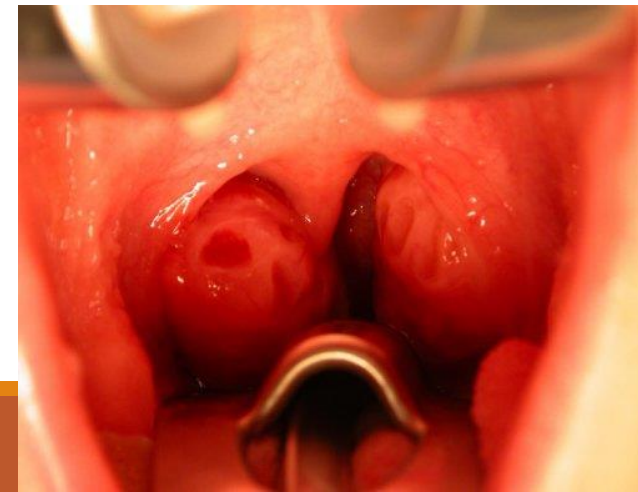
Definition and classification of OSAHS

- **Obstructive peripheral forms of OSAHS** : tonsils hypertrophy corresponds to 80% of cases.
- Central forms : neurological alteration of breathing regulation – idiopathic central apnea
- Mixed combined forms : peripheral and central etiologies
- Syndromic forms of OSAHS



Physiopathology of the obstructive peripheral form of SAHS

- Obstruction anatomical factors
- Reduction of breathing airways and pharyngeal dimensions
- 80% of cases: tonsils hypertrophy
- **Associations: mouth breathing-pediatric obesity and OSAHS**



Physiopathology of the obstructive peripheral form of SAHS

Anatomic and functional factors

- Dysmorphosis : micrognathia-retrognathia-maxillary hypoplasia
- Macroglossia
- Pharyngeal insufficiency
- Tonsil hypertrophy
- Mouth breathing



Paediatric obesity

- Adipose tissue in cervical and para-pharyngeal areas
- Inflammatory phenotype

Reduction in dimensions of upper breathing airway

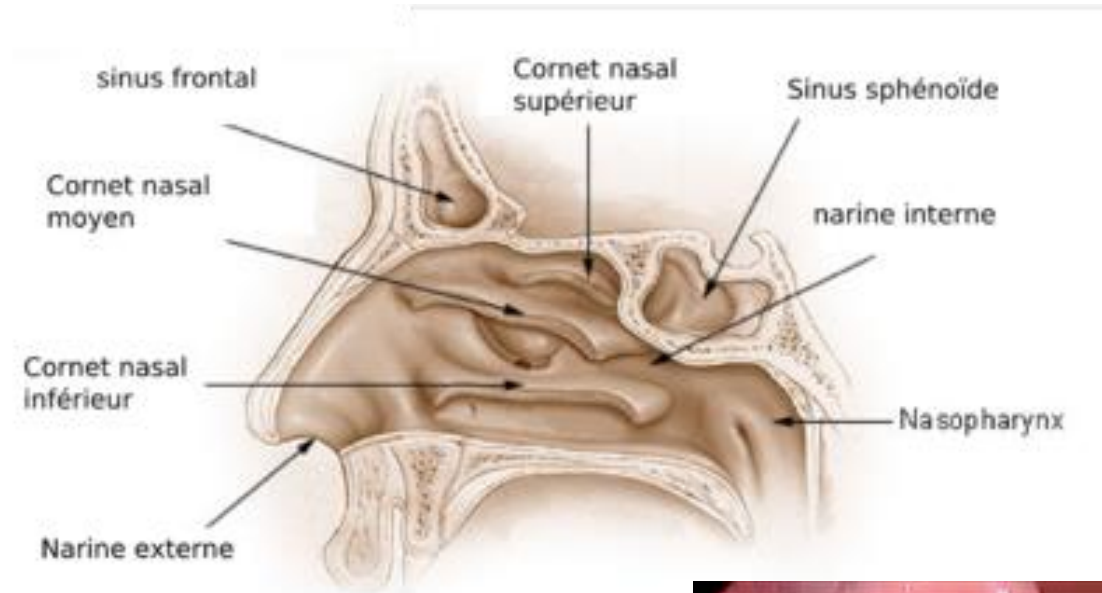
- Hypotonia
- Inflammatory pathology

Pediatric OSAHS



Physiopathology : implications of ENT congenital and acquired pathologies

- Tonsils hypertrophy
- Bilateral choanal atresia
- Supra-glottic stenosis
- Laryngomalacia
- Chronic rhinitis
- Anatomical anomalies of nasal cavities

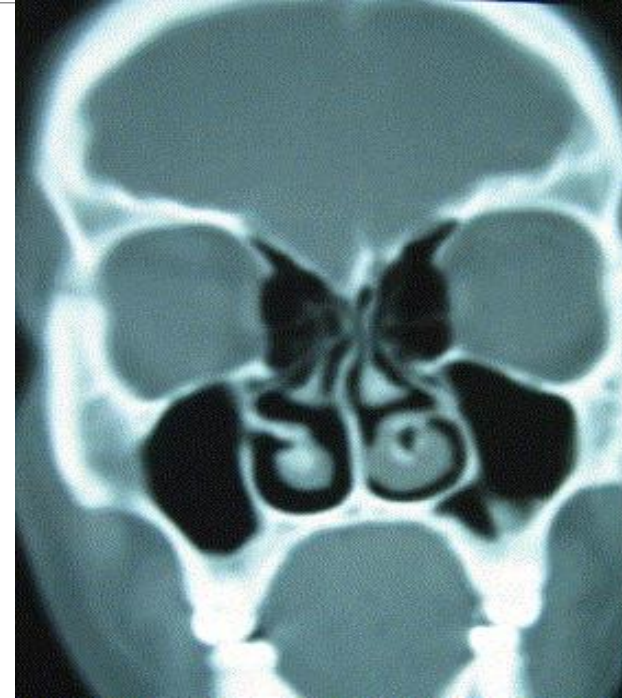


Close collaboration with ENT specialist



Anomalies and pathologies of nasal cavities in pediatric OSAHS

- Nasal dysmorphism, inflammation
- Septum deviation
- Congenital pyriform stenosis





Specific association between paediatric obesity and OSAHS

- Reduction in dimensions of the pharyngeal space
- **Specific clinical presentation** : older child, with no signs of tonsils hypertrophy or ENT pathology and **increased BMI**
- Lipid metabolism alterations, early insulino-resistance, neuro-endocrine modifications

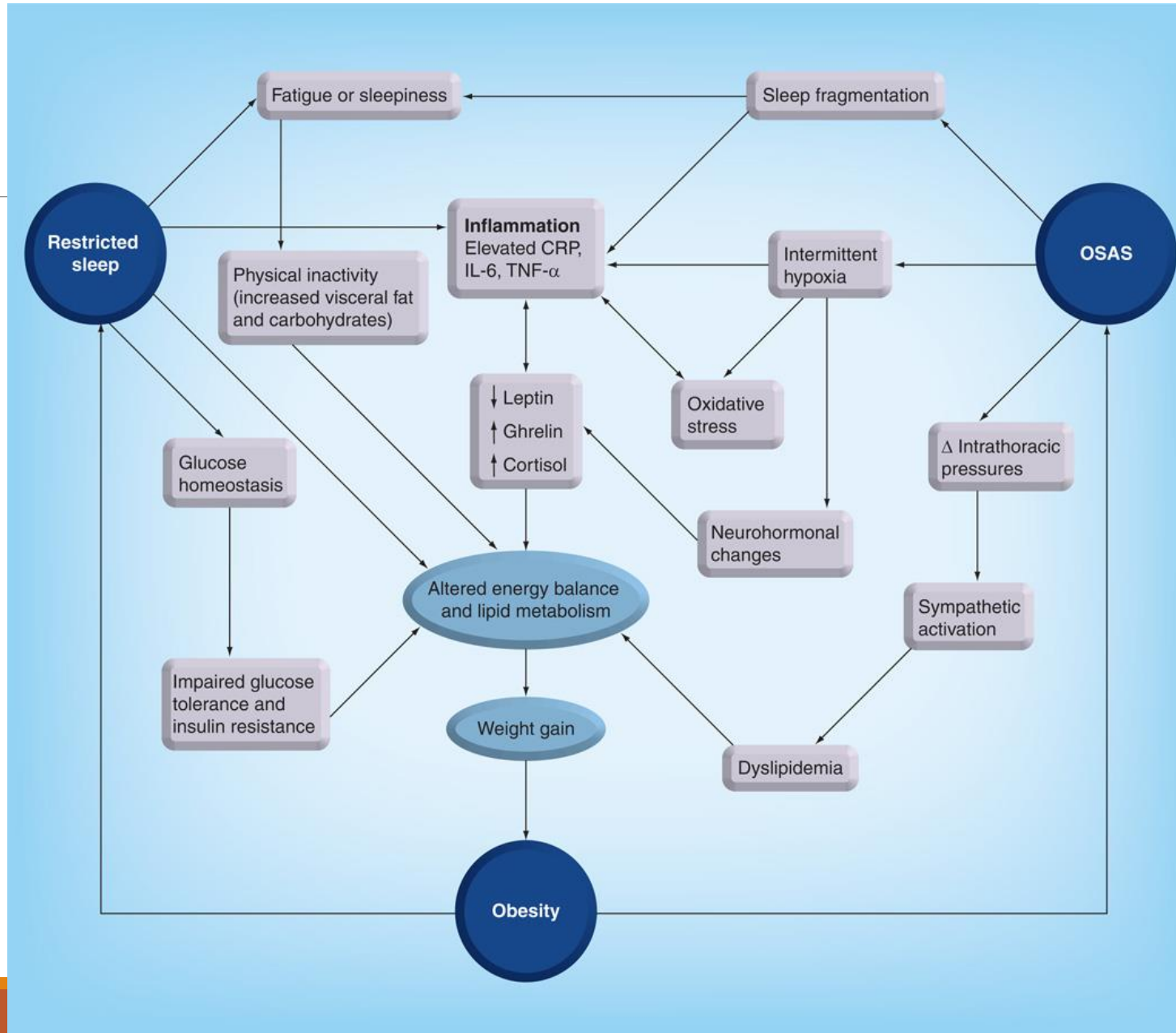
**Reciprocal and negative interactions
between OSAHS and obesity**

Systematic PSG recording

Van Eyck A, Van Hoorenbeeck K, De Winter BY, Van Gaal L, De Backer W, Verhulst SL. **Sleep-disordered breathing, systemic adipokine secretion, and metabolic dysregulation in overweight and obese children and adolescents.** Sleep Med. 2017 Feb;30:52-56.



Specific association between paediatric obesity and OSAHS



Local, loco-regional and systemic consequences of OSAHS



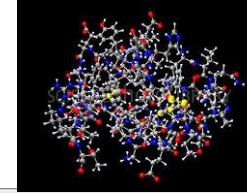
Dento-facial and periodontal pathologies

- Anterior gingivitis
- Increased caries risk
 - Xerostomia
- Mouth breathing
- Anterior infraclusion
- Maxillary hypoplasia
- Facial hyperdivergence



Cardio-vascular pathologies

- Endothelial and left ventricular dysfunctions
 - Diastolic hypertension
- Hypoxic pulmonary arterial hypertension
 - Atherosclerosis
- Blood pressure variability
- Alterations in Brain Natriuretic Peptid (**BNP**) plasmatic levels



Endocrine and metabolic modifications

- Insulino-resistance
- Inflammatory phenotype
- Increase in LDL-grehline plasmatic levels



Neuro-behavioral alterations

- **ADHD** : attention deficit and hyperactivity disorder
- Sleepiness
- Cognitive alterations



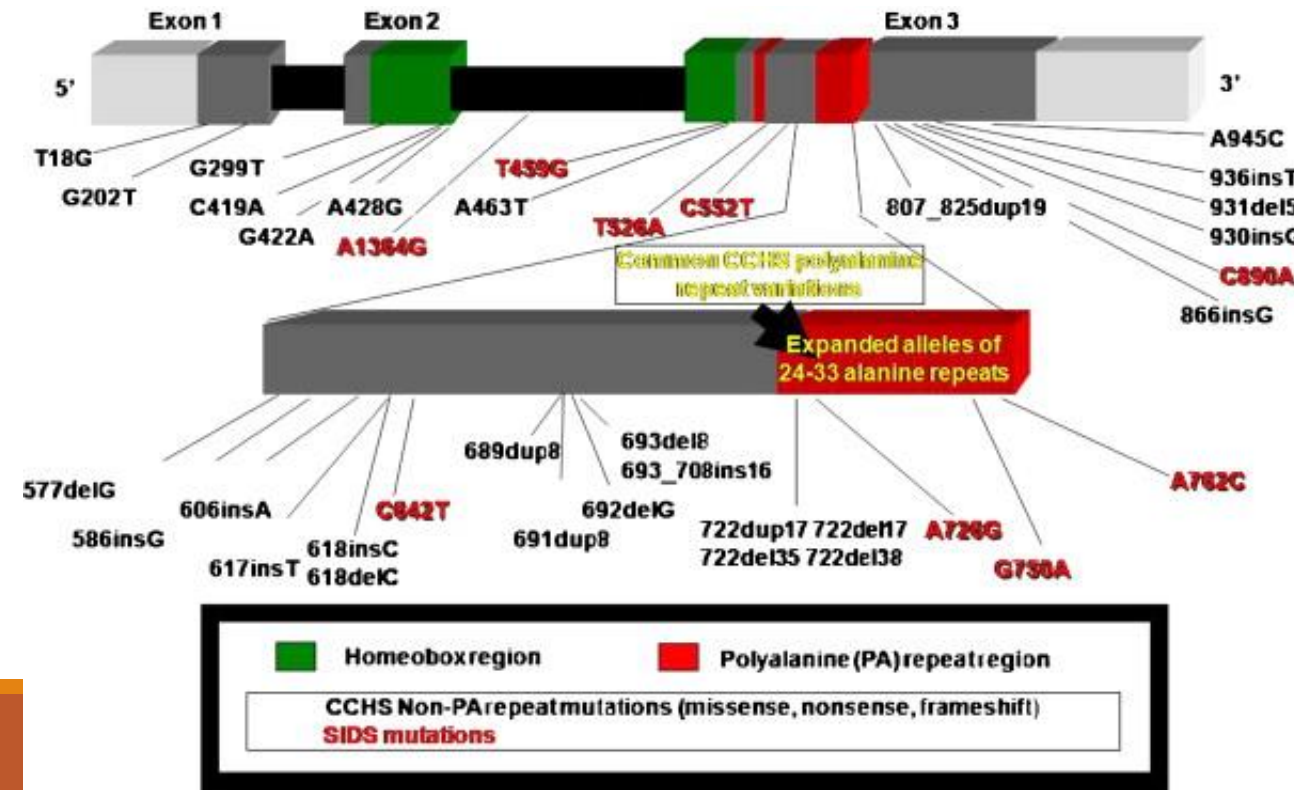
Central forms of paediatric OSAHS: example of Ondine syndrome

- **Congenital central alveolar hypoventilation syndrome : Ondine Syndrome**
- Autonomous central nervous system dysfunction of genetic origin
- Alteration of central breathing regulation
- Rare syndrome with a **prevalency of 1/200.000**
- Trachetomy Indication



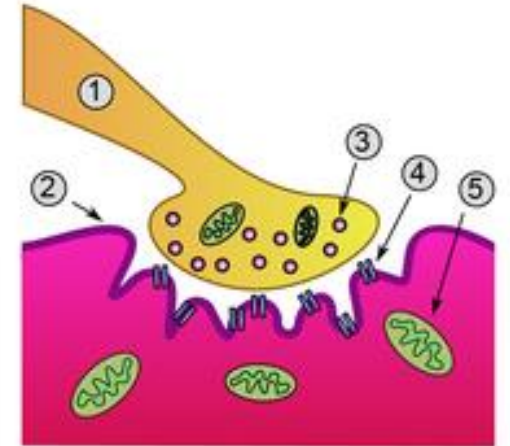
Molecular aspects of Ondine syndrome : heterozygous mutation of *PHOX2B* gene

- *PHOX2B* gene coding for a transcriptional factor involved in the development of autonomous nervous system
- Associated Hirschprung disease and increased risk of neuroblastoma



Physiopathology : neuro-vegetative and motor alterations

- Altered neurological control : acquired or congenital respiratory neurovegetative dystonia
- **Myasthenia gravis**: auto-immune neuro-muscular disease with muscle weakness (Auto-immune synaptopathy affecting the nerve-muscle junction)
- Congenital muscular dystrophy
- Laryngomalacia-tracheomalacia



Involvement of metabolic and respiratory diseases

- Asthma
- Broncho-pulmonary dysplasia
- Mucopolysaccharidosis : metabolic disorders of lysosomal enzymes affecting skeletal and connective tissues (Hurler, Hunter, Morquio syndromes)
- Mitochondrial metabolic diseases



Syndromic OSAHS

Association between OSAHS and several polymalformative syndromes

- Syndromes with cranio-facial or **skeletal defects**: achondroplasia (60% of OSAHS)-skeletal dysplasia
- Treacher-Collins and Prader-Willi syndromes
- Hemi-facial microsomia
- Cranyostosis



Tenconi R, Khirani S, Amaddeo A, Michot C, Baujat G, Couloigner V, De Sanctis L, James S, Zerah M, Cormier-Daire V, Fauroux B. **Sleep-disordered breathing and its management in children with achondroplasia.** Am J Med Genet A. 2017

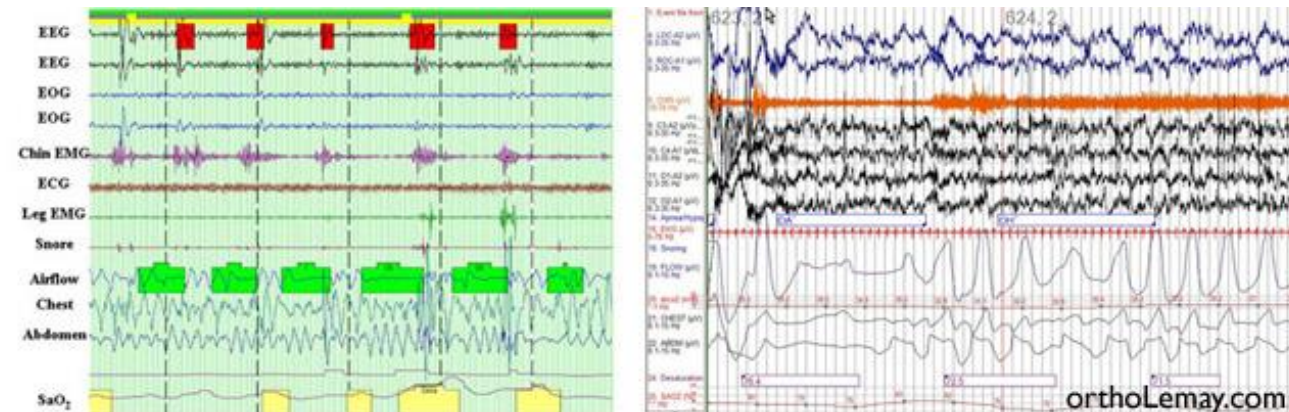




Syndromic forms : OSAHS associated with Down syndrome

- **Multi-factorial OSAHS in the context of Down syndrome**
- **Combined form of OSAH** with peripheric and central factors
- Muscles Hypotonia – facial dysmorphism
- Endocrine pathologies : thyroid dysfunction

Polysomnographic
recording systematic in
Down syndrome



Surgery.Mims M, Thottam PJ, Kitsko D, Shaffer A, Choi S. **Characterization of Sleep Architecture in Down Syndrome Patients Pre and Post Airway.** Cureus. 2017 Jan 17;9(1):e983.



Paediatric OSAHS and Prader-Willi syndrome

- Genetical disease linked to a 15q11-q13 deletion
- Altered growth
- Facial dysmorphism
- **Syndromic obesity:** neuro-hormonal changes with increased ghrelin* level
- Neuro-behavioural alterations
- Generalized muscular hypotonia

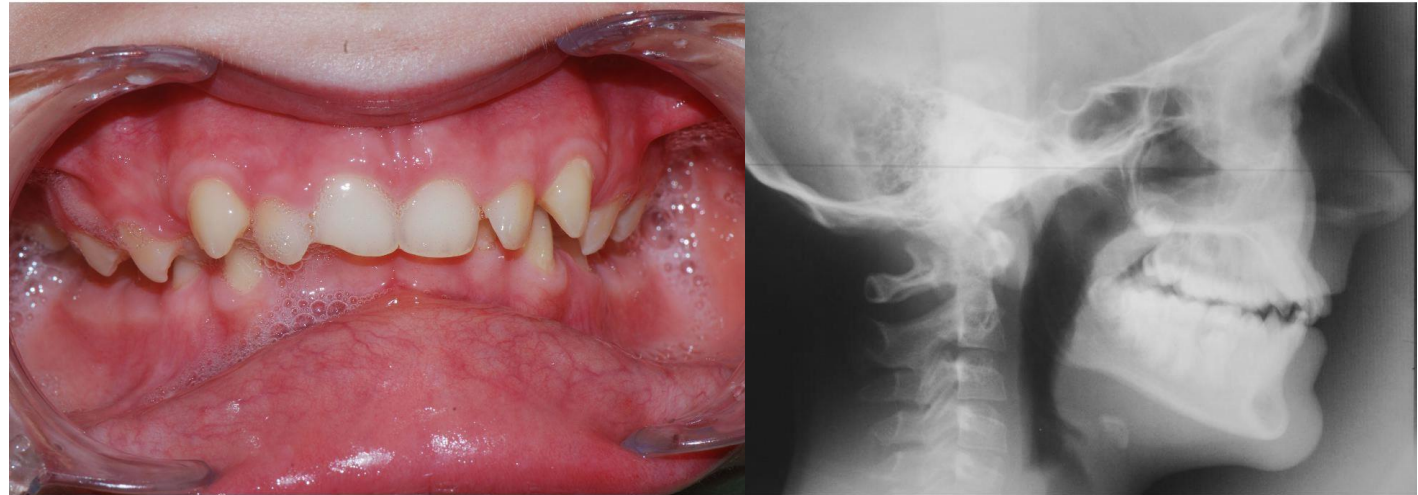
* **Ghrelin** is a hormone that is produced and released mainly by the stomach with small amounts also released by the small intestine, pancreas and brain. Ghrelin is also called the 'hunger hormone' because it stimulates appetite, increases food intake and promotes fat storage.



Paediatric OSAHS and Prader-Willi syndrome: patient treated in the Reference Center for Dental Manifestations of Rare Diseases - Strasbourg

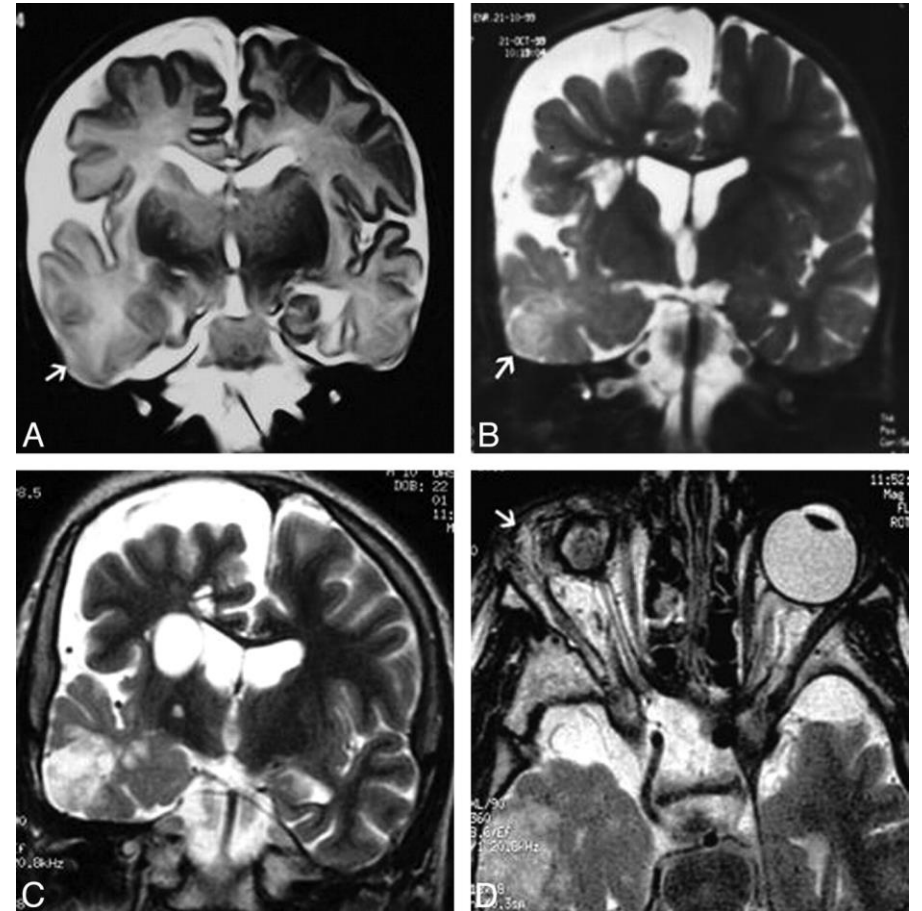
- Pediatric OSAHS often associated with Prader-Willi syndrome
- Altered sleep architecture and quality
- Sleepyness during daytime and **narcolepsy** in severe forms

Systematic diagnosis of sleep
pathologies based on
polysomnography for children
with Prader-Willi



Example of a syndromic mixed form of paediatric OSA : the Schimmelpenning syndrome

- Neurocristopathy
- Rare neuro-cutaneous syndrome with a prevalence of 1/10.000
- Cerebral tumors and enlargement of cerebral ventricles



Polysomnographic diagnosis in the Schimmelpenning syndrome

Nuit d'enregistrement polysomnographique du 14 décembre 2012	
Temps total de sommeil :	558 min
Latence d'endormissement :	56 min
Nombre de cycles :	6
Efficiencie du sommeil	
Index d'éveils + micro éveils / heure de sommeil :	8.27
Index d'apnées – hypopnées + résistances / heure de sommeil :	6.2
Dont index d'apnées + hypopnées	6
Dont index de limitation inspiratoire de débit	0.22
Dont index d'apnées centrales	0
SaO ₂ moyenne :	97.28 %
SaO ₂ minimale < 90 %	0.15 %
Index des mouvements périodiques du sommeil / heure de sommeil :	0



Etiological mechanisms of OSAHS in the Schimmelpenning syndrome



Combined peripheric and central etiologies of OSA



Alteration of central nervous system

Lesions of areas regulating autonomous breathing
Cerebral hemi-atrophy

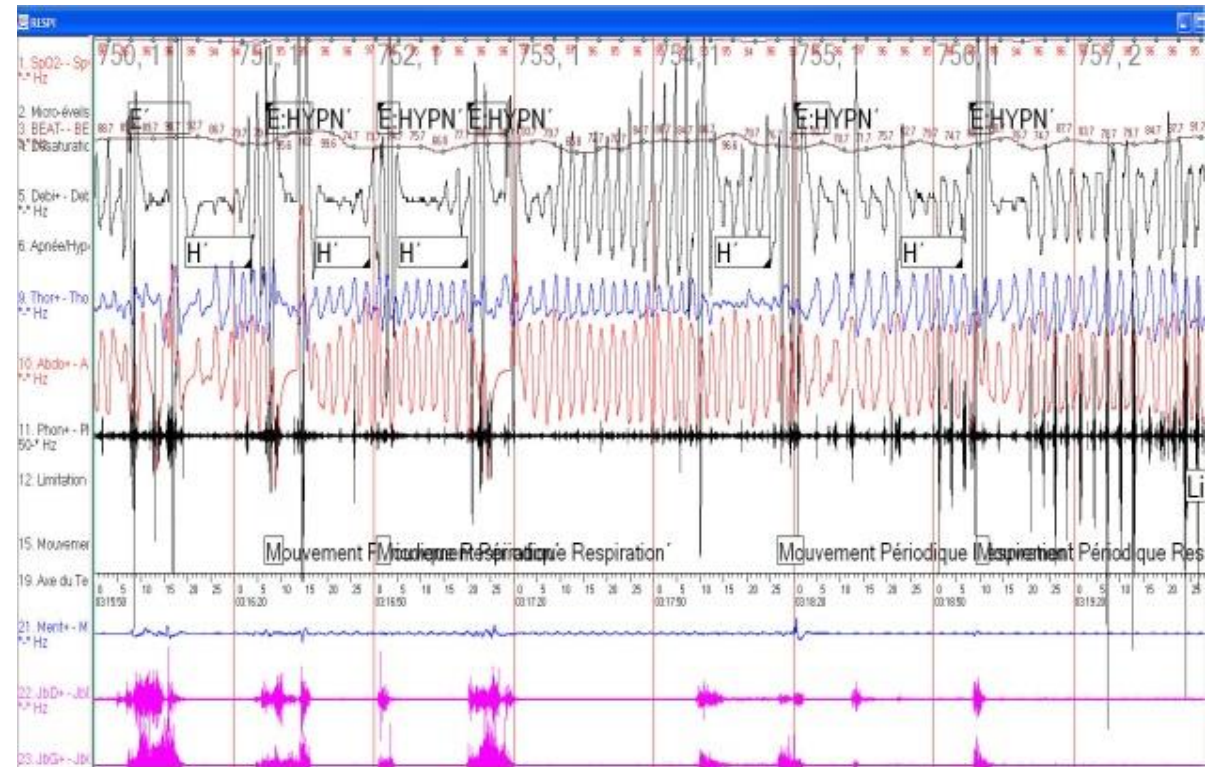
Peripheric obstructive and anatomical factors

Mandibular retrognathia
Lower tongue position
Tonsils hypertrophy





Multi-disciplinary diagnostic of paediatric OSA



The polysomnographic diagnostic of OSA

Recording of several parameters in a specialized sleep pathology department

- EEG
- EMG
- ECG
- EOG: electro-oculography
- PCO₂ (transcutaneous)-capnography
- SpO₂
- Nasal breathing recording
- Chest plathysmography
- Video
- Monitoring of body position



OSA & pediatric dentistry

Dento-facial diagnostic aspects



Systematic diagnosis



Medical anamnesis

Alterations in breathing
Sleep quality
Neuro-behavioral
changes

Extra-oral examination

Intra-oral examination

Ear-nose-throat diagnosis

Oro-facial functions

Complementary examinations

Polysomnographic recording

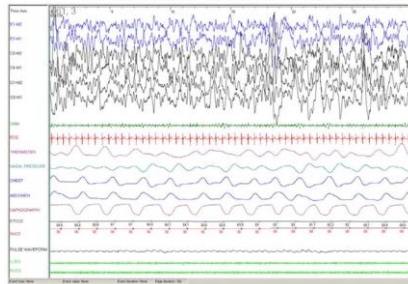
Rhinomanometry

Rhinofibroscopy

Rhinopharyngoscopy

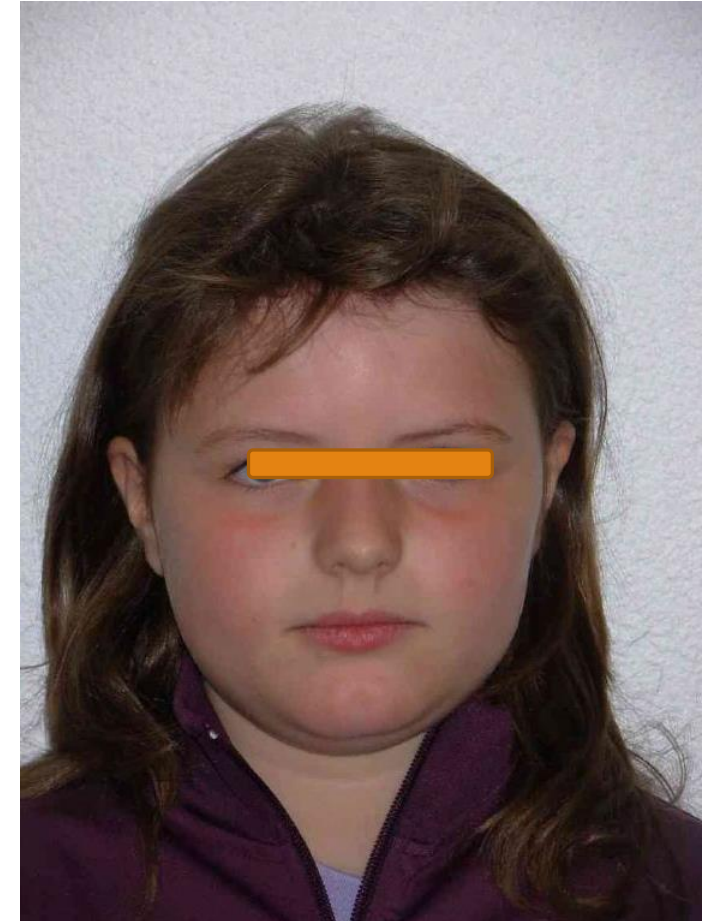
Profile Teleradiography

3D CBCT reconstructions of pharyngeal space



Dento-facial clinical aspects

- Increase in lower face height
- Nasal Collapsus
- Lip incompetency-mouth breathing
- Dark rings under the eyes



Dento-maxillary dysmorphosis associated to OSAHS

Maxillary Hypoplasia
Open bite



Dento-maxillary dysmorphism associated with OSAHS

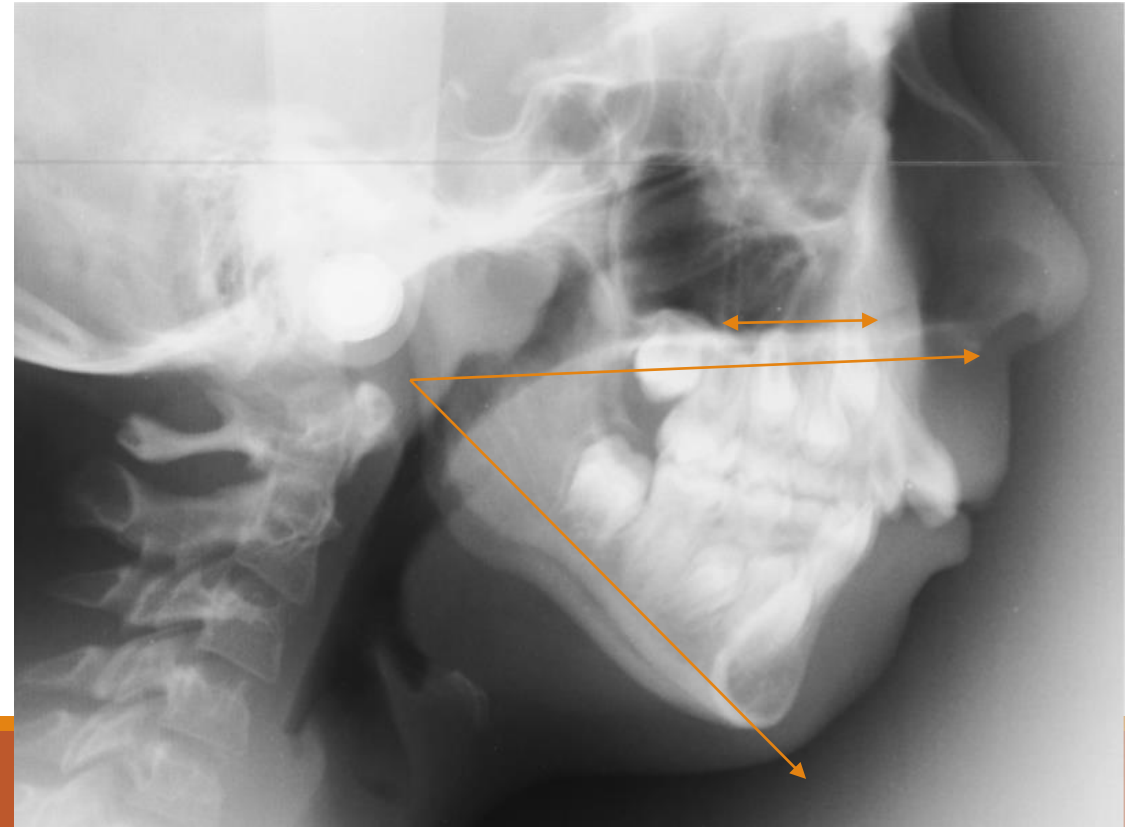


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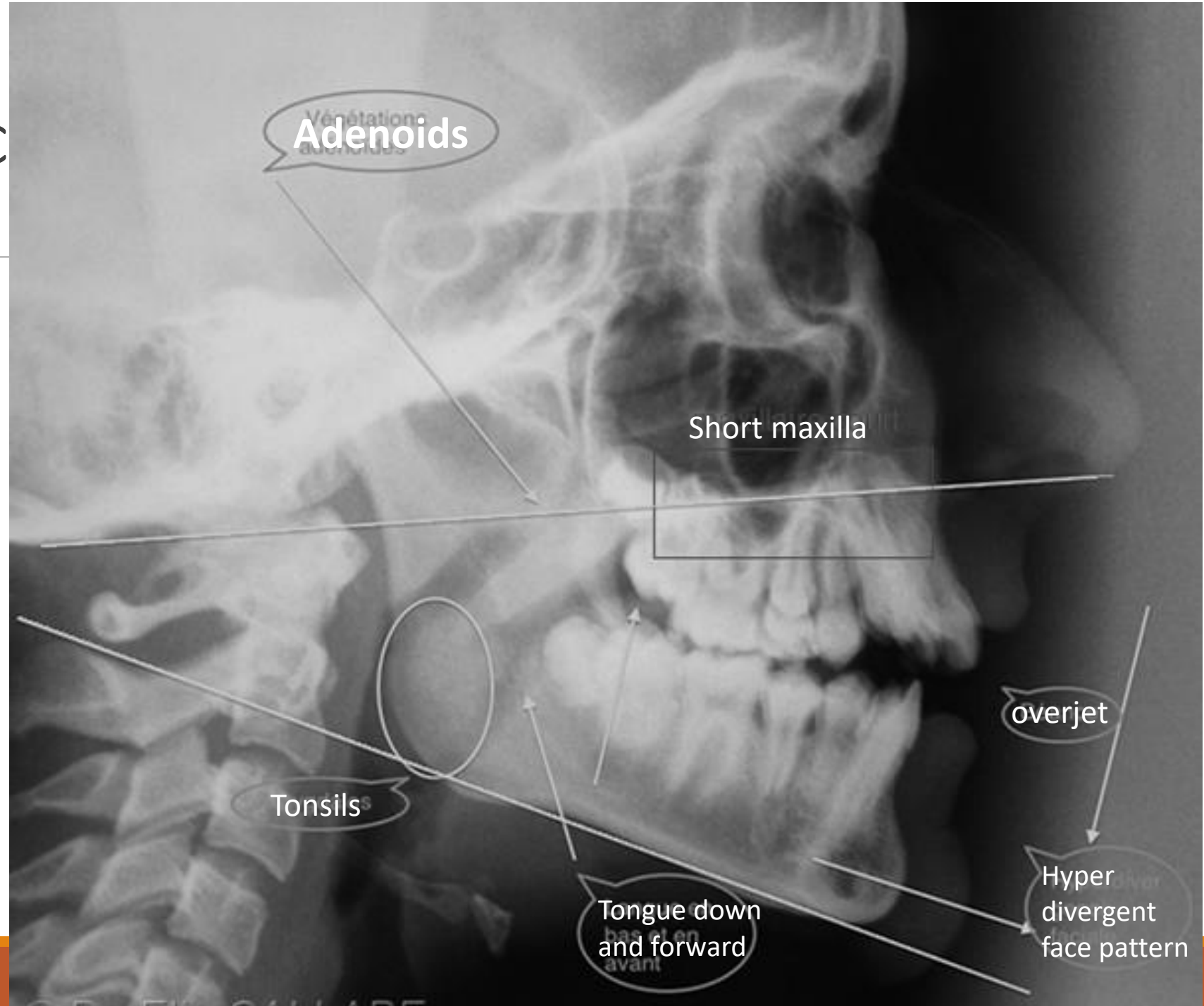


Dento-maxillary dysmorphosis associated with OSAHS

- Mandibular retrognathia
- Lower position of hyoid bone
- Facial hyperdivergence



Teleradiographic signs associated to OSAHS

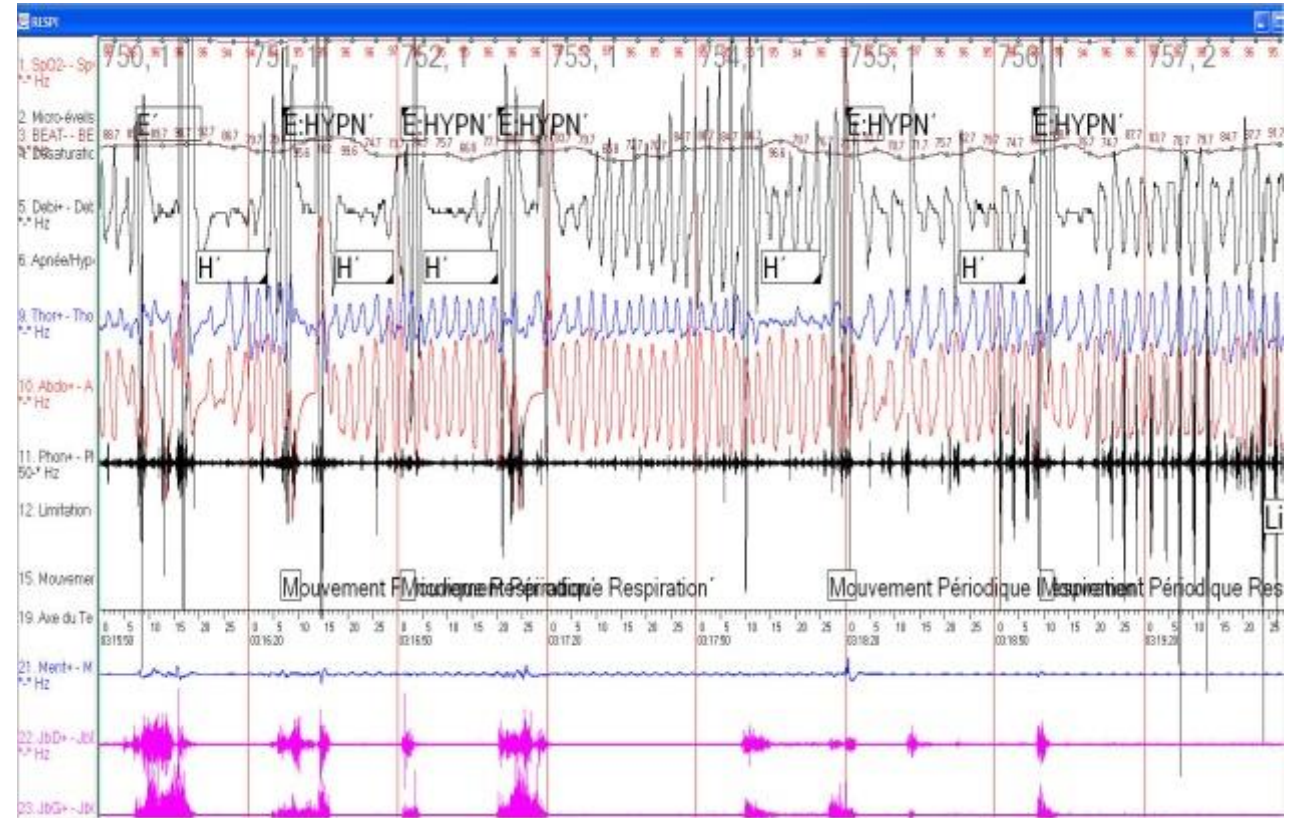


Cephalometric characteristics of children with OSA

- Alterations in maxillary and mandibular growth linked to mouth breathing and postural changes
- **Central role of paediatric dentists and orthodontists in early diagnosis and interception of OSA**



Therapeutic multi-disciplinary strategies of paediatric OSAHS



Multi-disciplinary medical and surgical management

- Obstructive peripheric forms : indication of **tonsillectomy**
- Importance of a post-operative ENT follow-up : **residual OSAHS**
- Post-operative polysomnographic recording
- Management of **associated pathologies**: rhinitis, asthma..



Multi-disciplinary medical and surgical management

Management of **paediatric obesity**

- Close collaboration with the **paediatric endocrinologist**
- **Weight reduction** and **orthopedic treatment** : maxillary transversal expansion
- **Dental follow-up** : increased caries risk



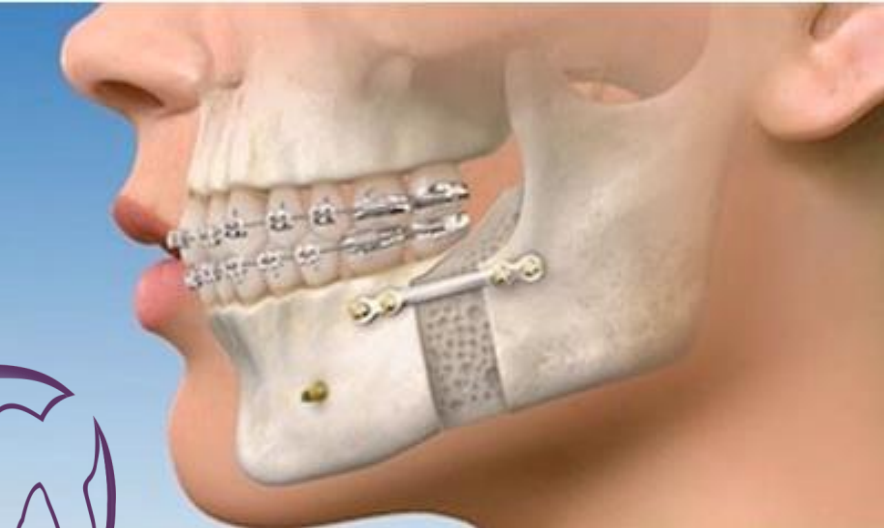
Orthopedic treatment in OSAHS

- Early maxillary transversal expansion
- Rapid palatal expansion (RPE)



Orthognathic surgery in paediatric OSAHS

- Indication of anterior mandibular displacement in cases of retrognathia:
bilateral ramus osteotomy
- Bi-maxillary osteotomy
- Close collaboration with **maxillo-facial surgery**



Multi-disciplinary medical and surgical management

Severe or residual forms of OSAHS

- Intra-nasal steroids administration
- Positive pressure (continuous positive air pressure CPAP)
- ENT surgery : pharyngoplasty-veloplasty



Conclusions

- Importance of multi-disciplinary diagnosis and treatment
- Organisation of the consultation devoted to paediatric OSAHS in Strasbourg:
 - Paediatric dentist
 - Orthodontist
 - Medical specialist in sleep pathology
 - ENT specialist performing a rhinoscopy
 - Maxillo-facial surgeon

