the Child Dental Patient with

Physical impairment: LARSEN Syndrome

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ORares MALADIES RARES ORALES ET DENTAIRES
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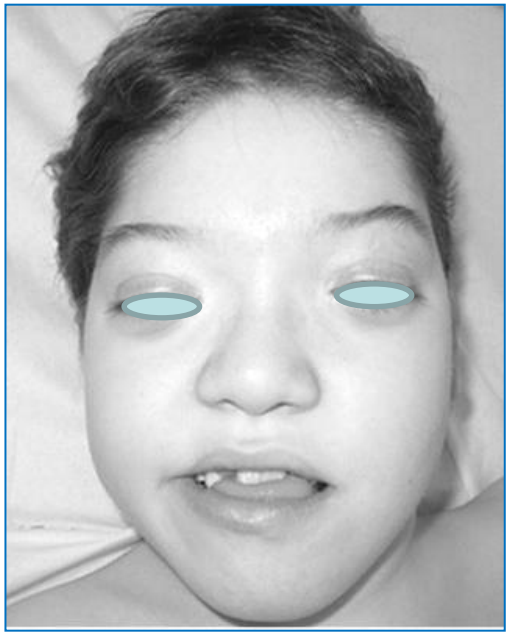
Clinical case

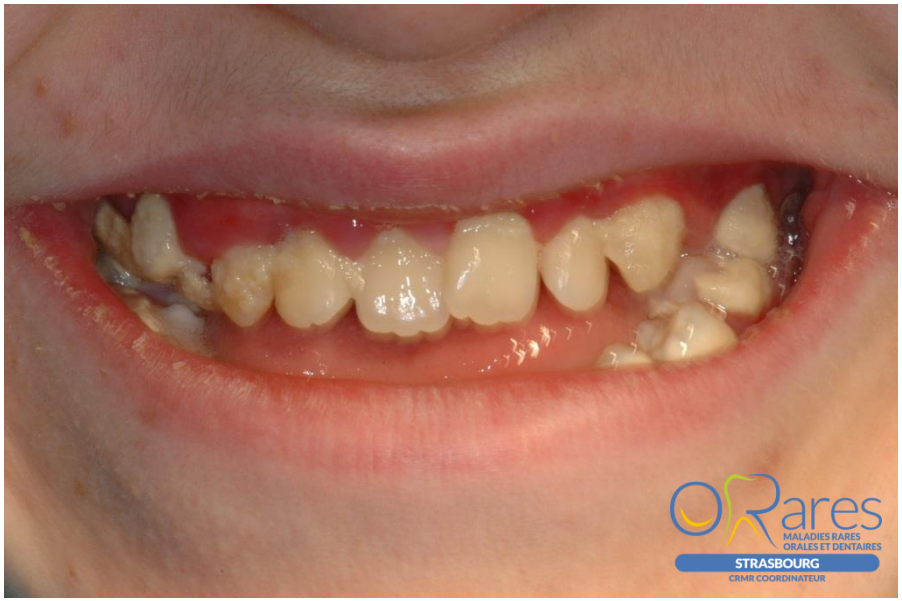
- 14 years-old boy
- Referred for an orthodontic assessment
- Its first visit to a dentist
- Schooled in a specialised institute

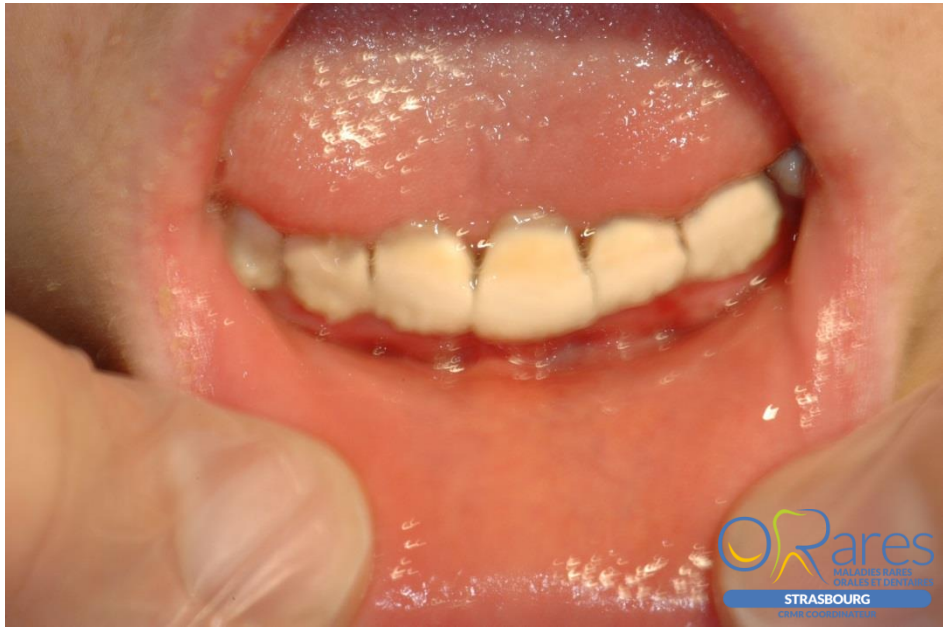


General health assessment:

- He is suffering from a physical impairment (Larsen syndrome):
  - Multiple joint deformation
  - Musculo-skeletal abnormalities
  - Dysfunction of arms and hands
  - Use an electric wheelchair but anable to manage it by himself
- Moderate hearing disability
- Moderate visual disability, glasses
- Medically compromised
- Poor motor control of the soft tissues of the mouth

Intraoral examination





Intraoral examination

Results of the initial intraoral examination

Soft tissues and oral hygiene:

- Dental plaque +++ (index 3, Loe & Silness)
- Significant accumulation of calculus with lower incisors totally covered by a thick layer of calculus
- Severe gingivitis (index 3, Loe & Silness): inflammation in all sectors, spontaneous bleeding
- Halitosis

Dental examination

- Mixed dentition
- Several primary teeth failed to exfoliate, covered with calculus
- Several premolars erupted lingually
- Severe class III malocclusion
- Relative macroglossia
- Dental caries impossible to detect

Poor oral health and severe malocclusion

Assesment of pain and anxiety

- The child reports pain when eating
- Also reports chewing difficulties (obliged to eat only mixed food since at least two years)
- Clinical inspection of the oral cavity was possible but he strongly contracted his lips
- Sometimes, he was seized with tremors in his legs

- He is able to explain clearly what he is feeling
  - Does not like that we spread his lips to examine the teeth
  - Fear of dental instruments
  - Fear from getting water in the mouth, fear of choking
  - Fear of the surgical suction
- Venham scale 3 during examination

How to explain such a poor oral health of this adolescent?

He is affected by a physical impairment but presents no learning difficulty
History of good oral health in primary dentition, no caries

At the special school

- He has his lunch at school but the education team do not help the children with toothbrushing after the meal
- He progressively quit chewing when primary molars failed to exfoliate spontaneously →poor clearance of foodstuff from the mouth
- He had more and more bleeding during meals so the school decided to mix all his food

Decision taken: to send pictures of his mouth taken before and after the first scaling so that the medical and education staff understand the severity of the situation and the obligation to assist him for the toothbrushing as part of daily routine

At home

- The parents are both affected by physical impairment (the father has Larsen syndrome)
- They worried about provoking bleeding and harm him with the toothbrush, so they progressivly stopped the brushing
- He had a lot of surgeries and physiotherapies so the dental follow up has been neglected

Decision taken: to invite both parents at the clinic to explain periodontal disease, to involve them in the preventive measures, and to see what toothbrushing technique would be adequate, especially with regard to posture given their physical impairments

Treatment plan: criteria to be considered

Need for emergency care?

- No acute infection but moderate pain
- Neglected oral health
- Needs scaling and extraction of several primary teeth

Behaviour management?

- Relative ability to cope
- Dental anxiety
- Fatigability
- Cervical pain: importance of confortable body posture during dental care
- Good verbal communication – Good sense of humour

Medical risk assessment ??

Establishing the dental treatment plan

Discussion of the modalities of management:

- Behaviour management in the dental setting
- Pharmacological management:
  - N2O/O2 conscious sedation
  - Intravenous sedation
  - General anaesthesia

Decision needs a benefic/risk assessment

<u>Criteria to take in account:</u>

- Medical aspects (infectious risk, bleeding risk, other risks?)
- Type and complexity of dental treatment
- Expected level of cooperation
- Parental and institution support

Medical condition

Medical history

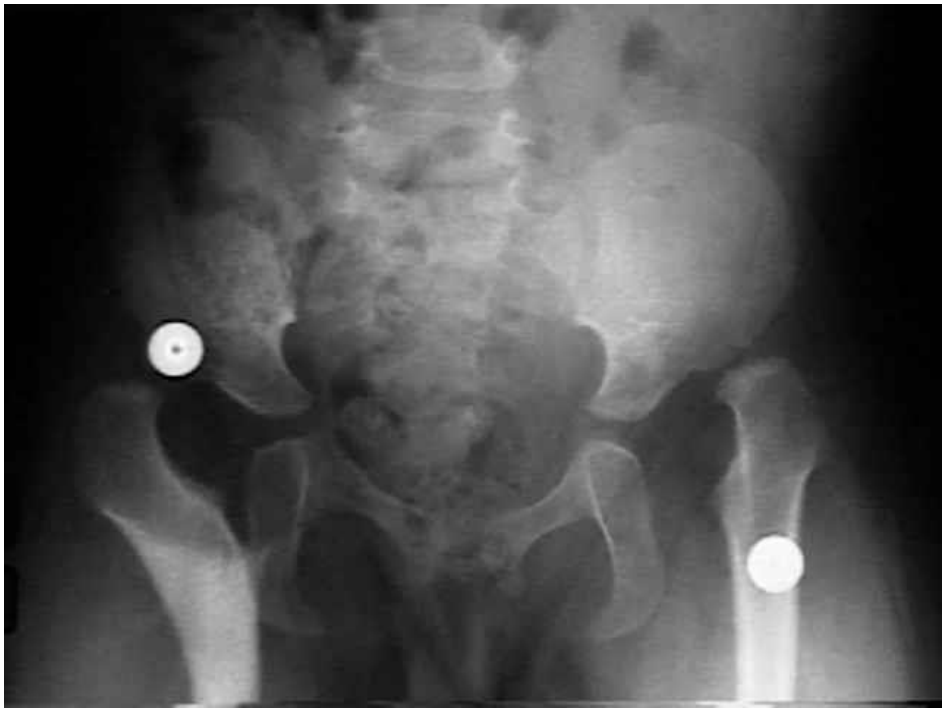
- Severe motor impairment due to Larsen syndrome
- Gastric ulcer (under medication)
- History of surgery: inguinal hernia and spine
- Multiple orthopedic problems

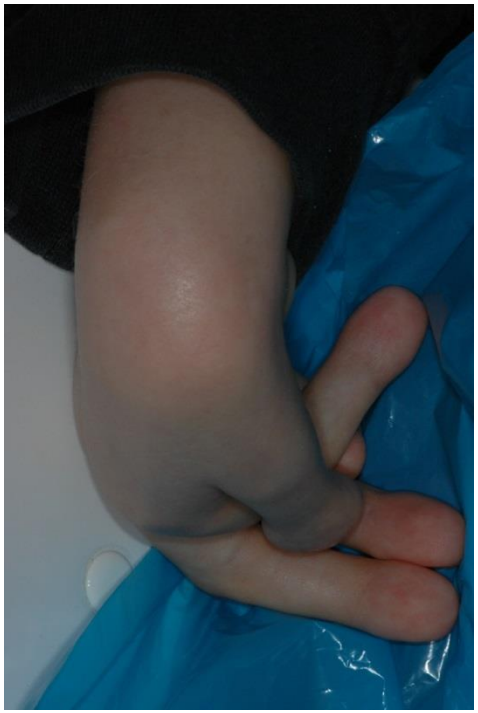
Larsen syndrome

- Syndrome characterized by congenital dislocation of large joints (hip, knee and elbow) with equinovarus or equinovalgus foot deformities, cervical spine dysplasia, scoliosis, spatula-shaped distal phalanges and distinctive craniofacial abnormalities, including cleft palate
- Rare genetic disease (OMIM 150250)
- Birth prevalence is estimated to be less than 1 in 250 000 in Europe
- Autosomic dominant or recessive
- Due to missense mutations or small in frame deletions in the *FLNB* gene (localized to 3p14.3) that encodes cytoskeletal protein filamin B.

https://www.orpha.net

Clinical characteristics of Larsen syndrome

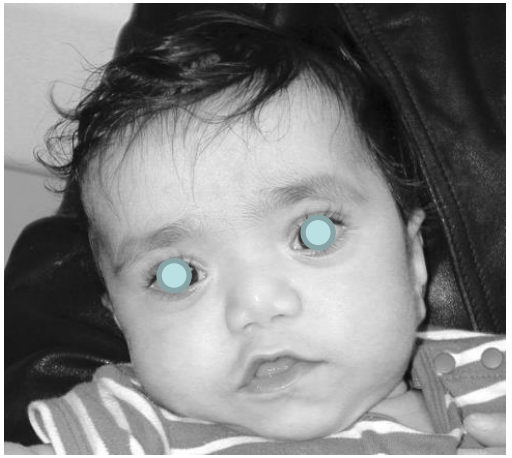


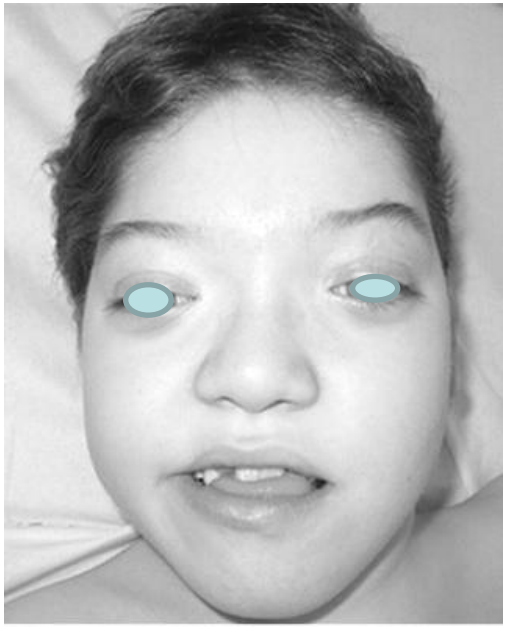


- Luxation of big joints like hips
- Spinal deformities such as scoliosis and cervical spine kyphosis
- Supernumerary carpal and tarsal bone
- Spatula-shaped distal phalanges with the thumb almost always affected

Cranio-facial abnormalities of Larsen syndrome

- Prominent forehead
- Ocular hypertelorism
- Depressed nasal bridge
- Flattened midface
- Cleft palate possible
- Macroglossia
- Midline cleft palate possible
- Conductive hearing loss





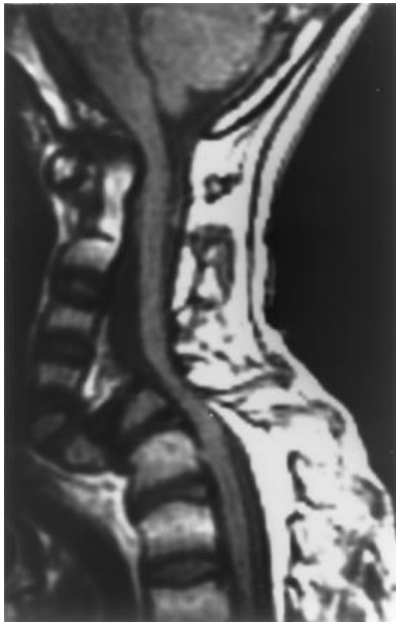
Larsen syndrome and anaesthetic risk?

Respiratory problems

- Thoracic and neck / craniofacial malformations
- Cervical instability
- Laryngomalacia
- Tracheo- et broncho-malacia

Congenital cardiac disease

- Aortic dilatation
- Interventricular or interauricular communication
- Arterial canal
- Mitral valve prolaps



Major anaesthetic risks

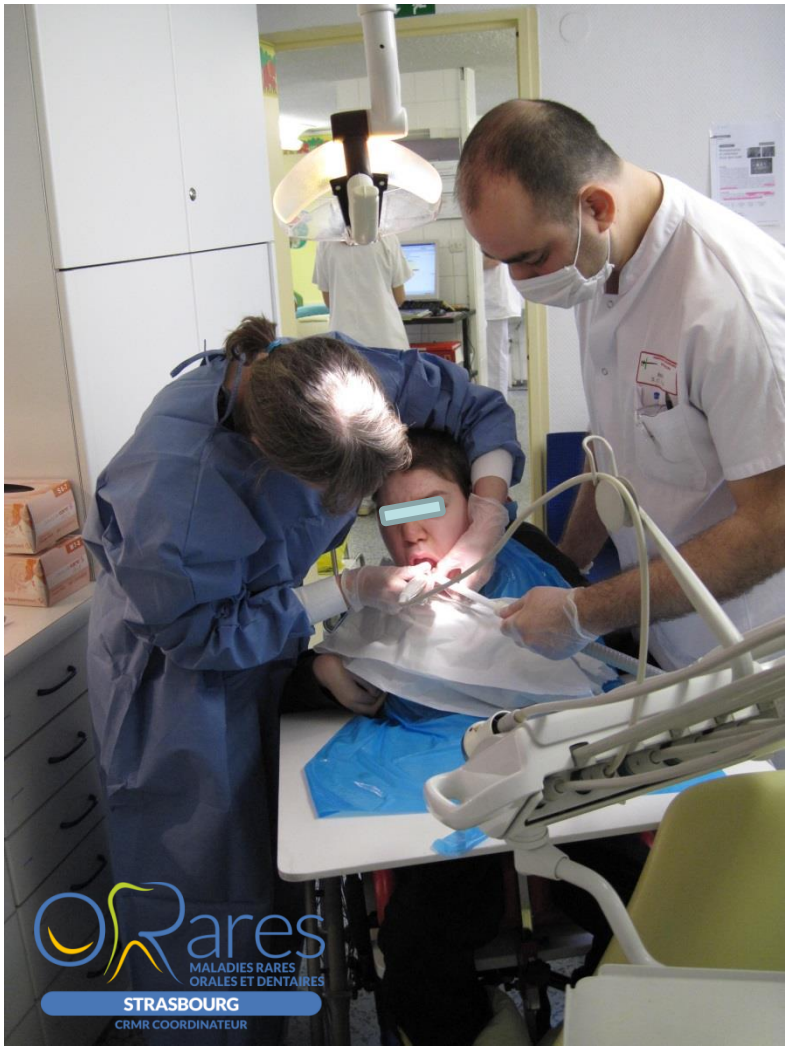
- Intubation and extubation difficulties
- Death by cardiac arrest
- High rate of respiratory post-operative complications
- Cases reported of malignant hyperthermia
- Estimated ASA III with high risk morbidity /mortality

GA and IV Sedation contraindicated for dental care

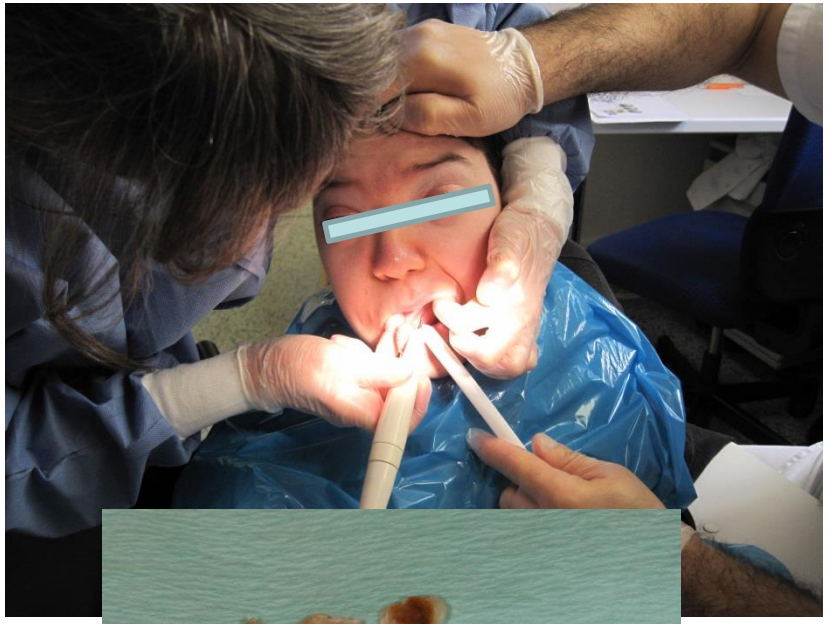
Choice for dental management for this child: vigil care in the dental office

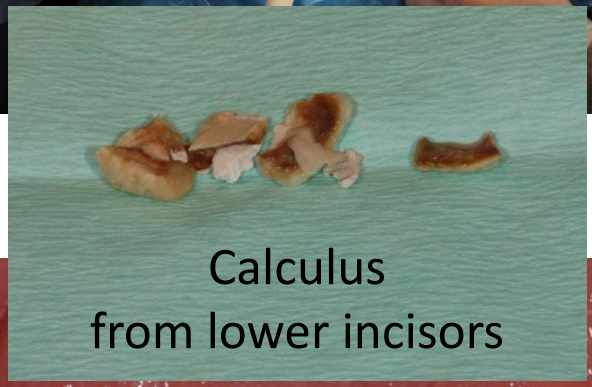
Important criteria for its behaviour management in the dental surgery:

- Body posture:
  - Patient treated in his own wheelchair
  - Careful positionning of his head and stabilization
- Short sessions to avoid any discomfort and tiredness
- Careful surgical sucking to avoid deglutition or inhalation of calculus

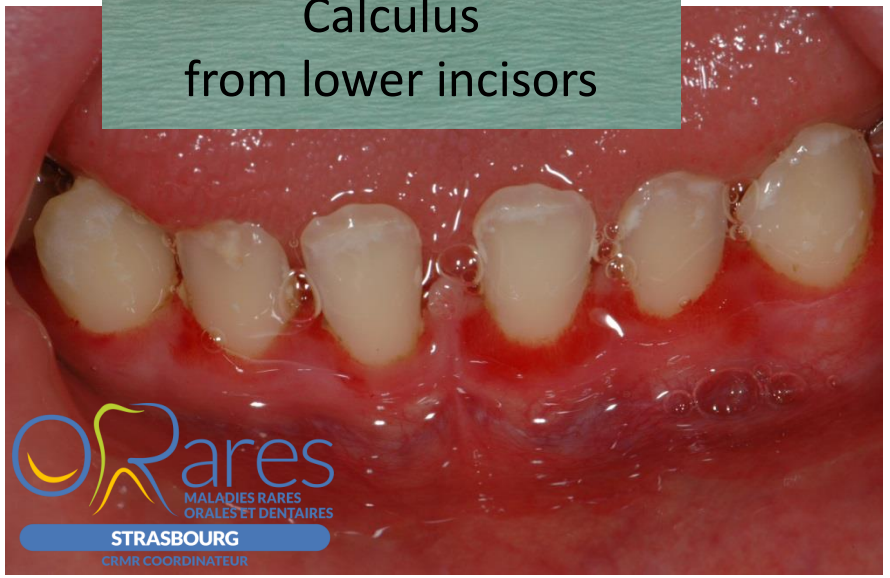


First session

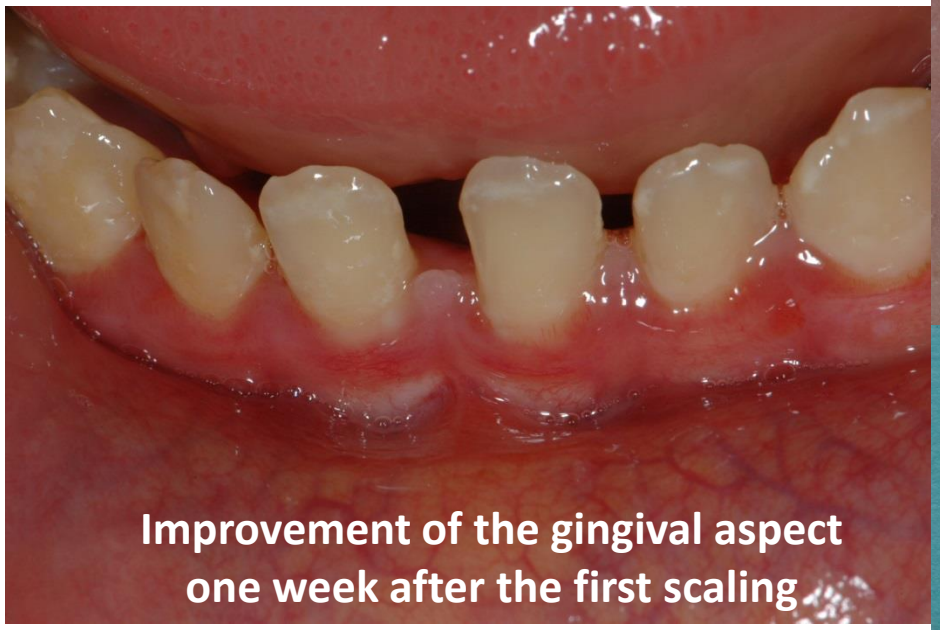




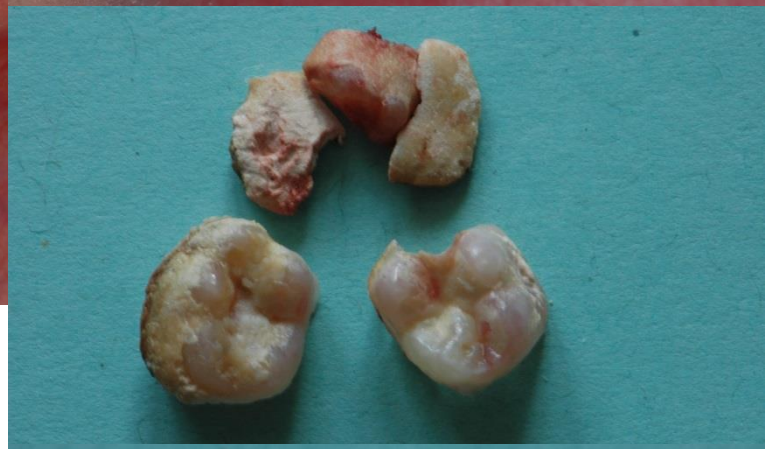
Calculus
from lower incisors

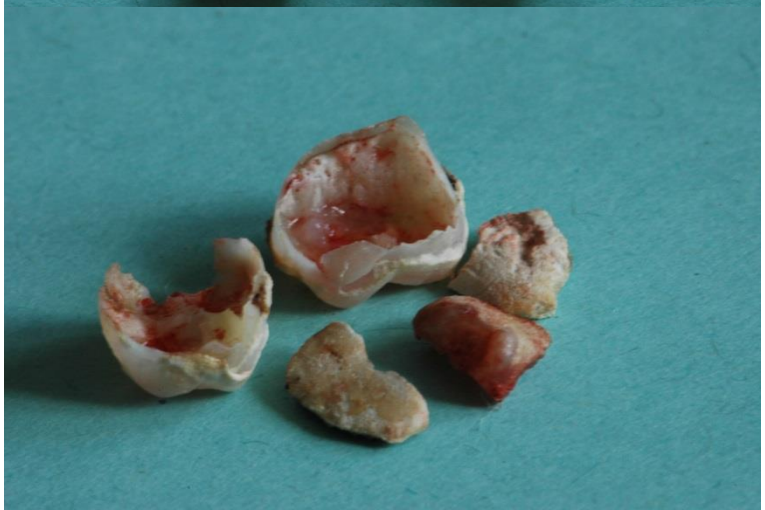


Second session (+ 8 days)



Improvement of the gingival aspect one week after the first scaling





Objectives :

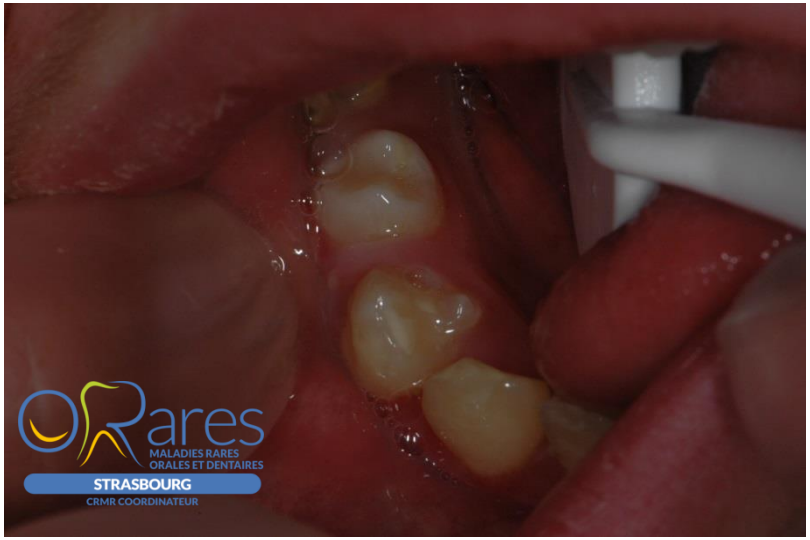
- To perfect the scaling of the anterior teeth
- Extraction of 3 primary molars under local anaesthesia (85, 55, 74) and with antibioprophylaxis

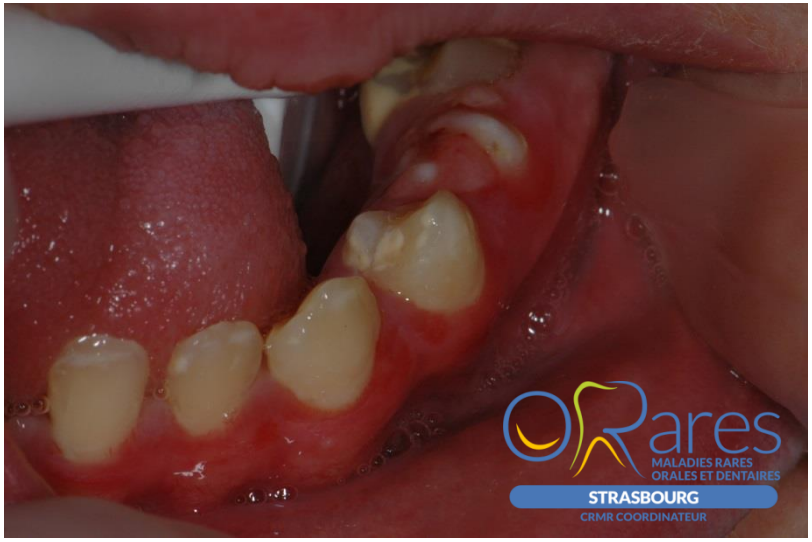
Third session

- 5 days after the 2nd session, under antibioprophylaxis
- The patient is more relaxed, comes to the dental department with pleasure and appreciates having a clean mouth
- Objectives of this session:
  - To evaluate the toothbrushing at school and at home
  - To scale the posterior teeth

Third session - Results

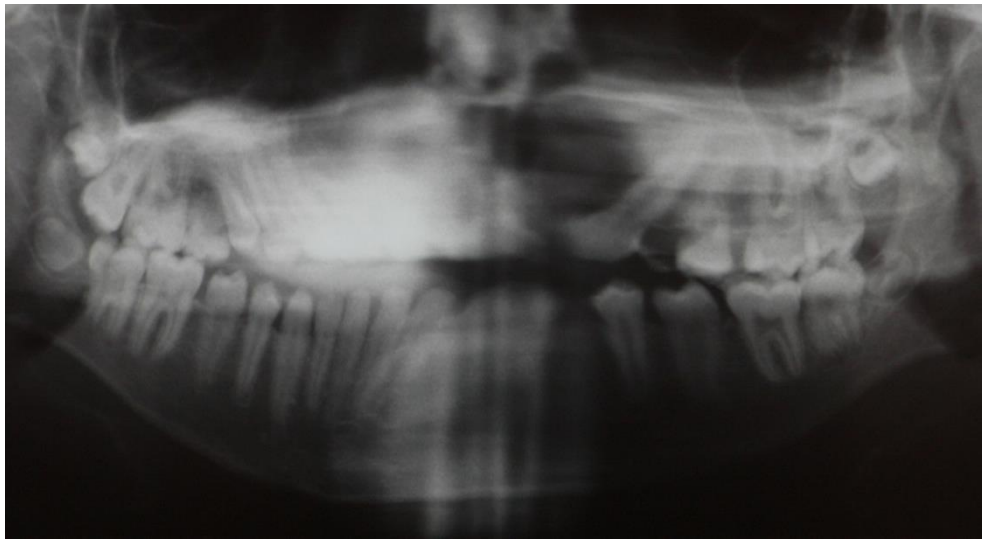
- Toothbrushing is effective but there is still some bleeding
- Access for scaling of the posterior teeth more difficult
- Examination of the upper jaw: persistance of a primary molar (65) in a palatal position, to be extracted in a next session
- Decision taken to try to do a panoramic radiograph at the next visit





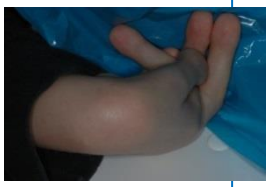
Panoramic radiograph

Difficulty: stabilization of his head and his cervical spine

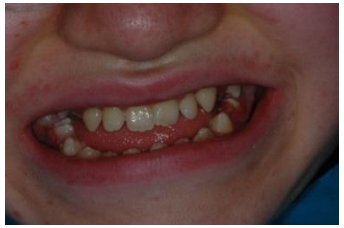


Absence of carious lesions (confirmation of the clinical examination)
Persistance of the right second primary molar
Presence of germs of 18, 28, 38 and 48

Conclusions: Great improvment of his quality of life!



- Adolescent at risk of periodontal disease but with low caries risk
- Dental management was possible without sedation and good cooperation was obtained for short sessions of dental care
- Rapid relief of all oral symptoms, including halitosis
- Improvement of his oral hygiene at school: the education staff are now aware that its toothbrushing has to be totally carried out by a caregiver
- Improvement of the toothbrushing technique by the parents
- Important change in diet: no more mixed food, he enjoys his meals again!
- Treatment of Class III malocclusion - impossible (contraindication of maxillofacial surgery)
- <u>Regular dental follow-up</u>:
  - Control every 4 months for the first year
  - Regular scaling (twice a year and more if needed)
  - Preventive measures (fluoride toothpaste)



This relatively simple dental management, together with the cooperation of the school staff and parents, lead to a better general health and had a great positive influence on the quality of life for this adolescent.

Keeping a clean and healthy mouth is essential for seriously disabled patients

Selected references and further reading

- Disability and Oral Care. Edited by Dr June Nunn, iADH; FDI World Dental Press Ltd, 2000
- Girisha KM et al. Phenotype and genotype in patients with Larsen syndrome: clinical homogeneity and allelic heterogeneity in seven patients; BMC Med Genet. 2016 Apr 6;17:27
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