the Child Dental Patient with

Williams Syndrome

ORares
MALADIES RARES ORALES ET DENTAIRES
STRASBOURG
CRMR COORDINATEUR

Les Hôpitaux Universitaires de STRASBOURG

European Reference Network
for rare or low prevalence complex diseases

Network
Craniofacial anomalies and ear, nose and throat disorders (ERN CRANIO)

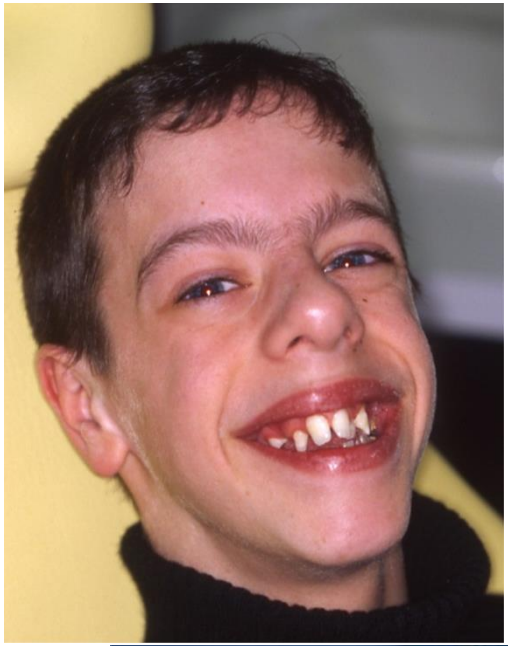
Member
Hôpitaux Universitaires de Strasbourg — France

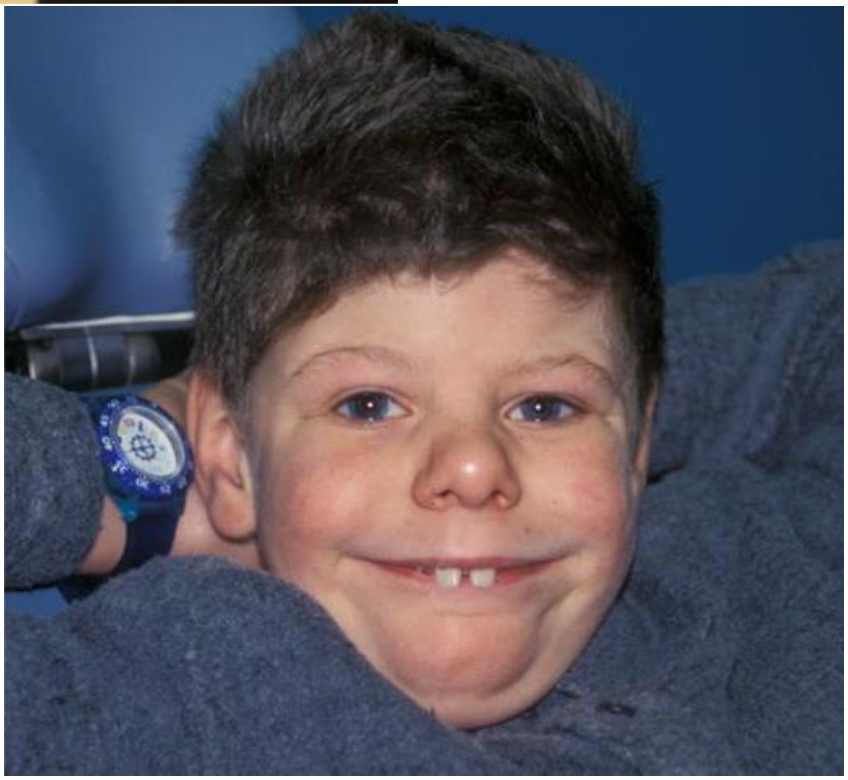
General characteristics of Williams syndrome

- Williams syndrome (= Williams-Beuren syndrome) affects approximately one in 10 000 people and is caused by the deletion of genes on chromosome 7q11.23 which code for elastin.
- Williams syndrome is a multisystem disorder.
- The phenotypic appearance of people with Williams syndrome is well characterized, but there continues to be new genetic and therapeutic discoveries.
- The neurodevelopmental profile of patients with Williams syndrome is characterized by high sociability and a variety of retained and compromised linguistic and cognitive abilities. Also they have an hypersensitivity to sound.
- Feeding and eating problems are common.
- Patients with Williams syndrome have increased morbidity and mortality under sedation and anesthesia, largely as a result of cardiovascular abnormalities (mainly : aortic stenosis, pulmonary valve stenosis, ventricular/auricular septal defects).

Physical features

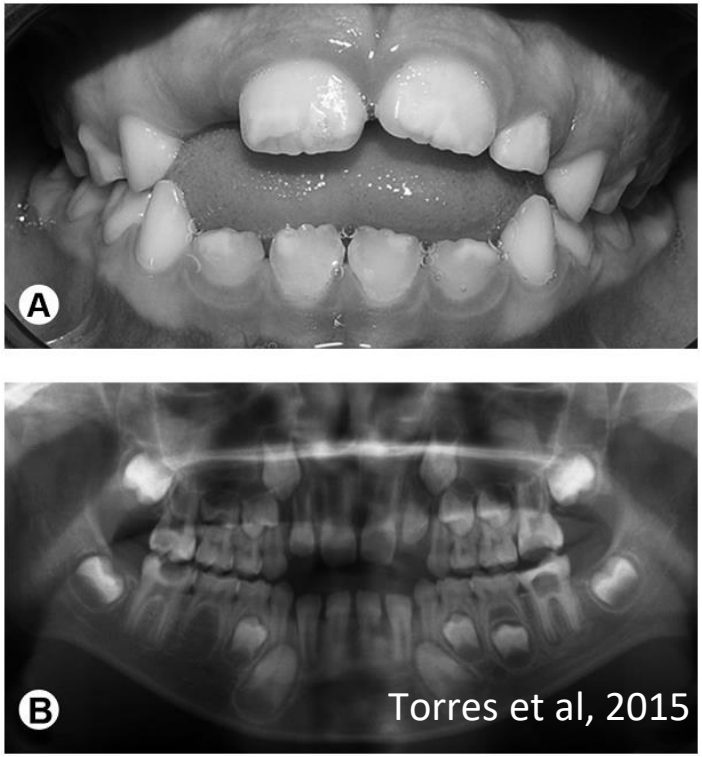
- **Main facial features:**
  - Flat midface
  - Depressed nasal bridge
  - Anterverted nostrils
  - Long philtrum
  - Thick lips
  - Wide open mouth
  - Blue eyes with stellate iris pattern
- **Also found:**
  - Short palpebral fissures
  - Ocular hypertelorism
  - Epicanthal folds
  - Peri-orbital fullness
  - Strabismus, visual impairment, photophobia
  - Sensorineural hearing loss, hyperacusis
  - Hoarse voice



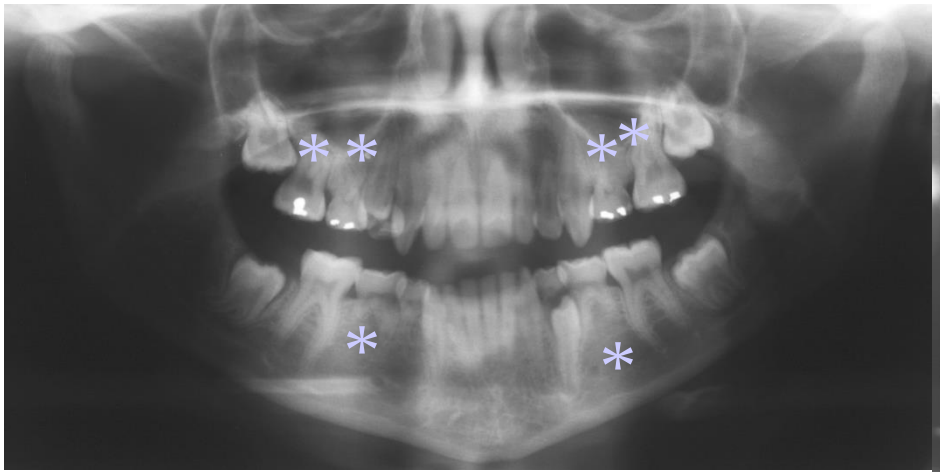


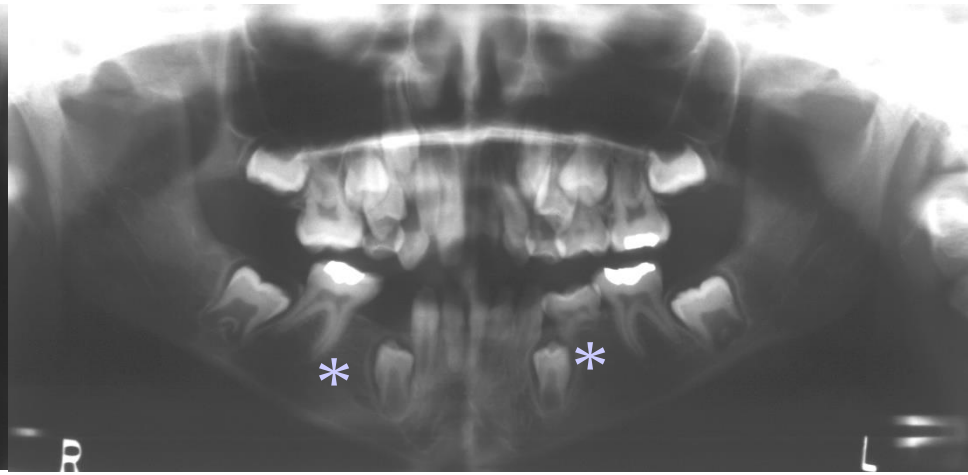
Oral features

- SPECIFIC to the syndrome:
  - Anomalies of dental development = 90% of the patients are concerned
  - Orofacial dysfunction (sucking problems, chewing and swallowing problems, mouth-breathing…)
  - Hypersensivity mouth
  - Eruption disorders
  - Malocclusions
- Non specific:
  - Dental caries
  - Periodontal problems



Anomalies of tooth number

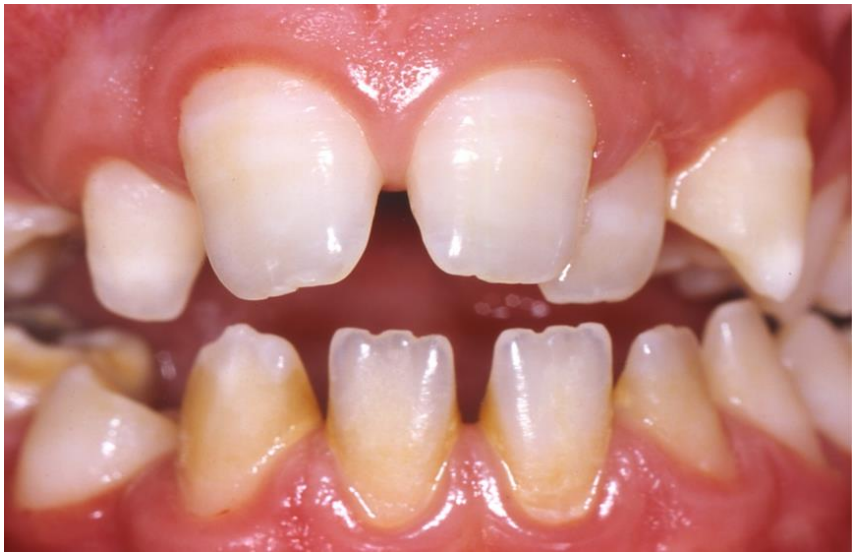


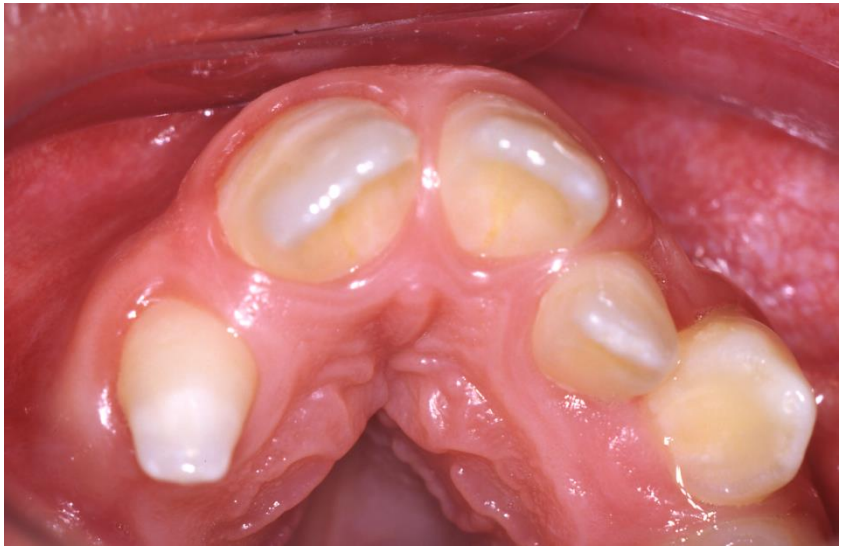


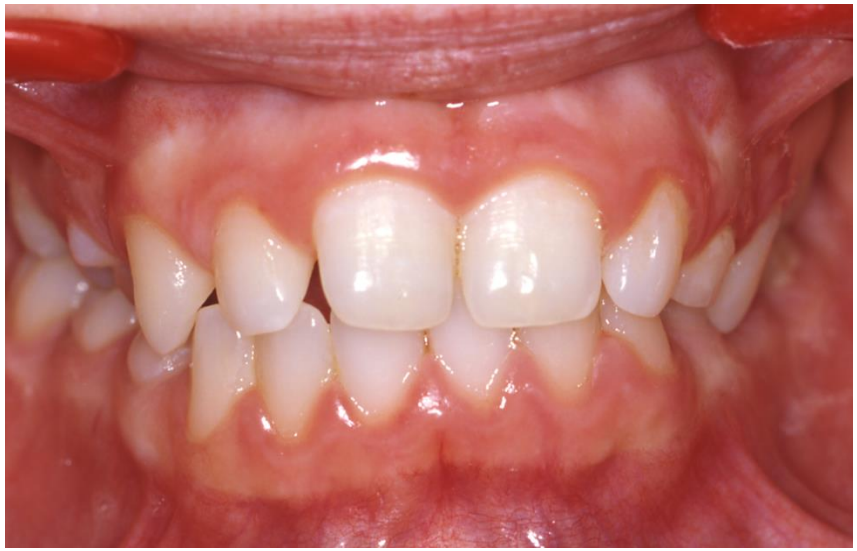
- 40,5% of children have agenesis of one or more permanent teeth = hypodontia (Axelsson et coll, 2003)
- 12% have oligodontia (6 missing teeth or more)
- Premolars are often concerned

Common other dental anomalies: abnormal tooth morphology

- Size and shape anomalies found in:
  - 12,5 % in primary dentition
  - 40,7 % in permanent dentition
- Microdontia:
  - mesiodistal incisor crown dimensions significantly smaller
- Anomalies of shape:
  - Tapered or screwdriver shaped incisors
  - Conical teeth
  - Taurodontism

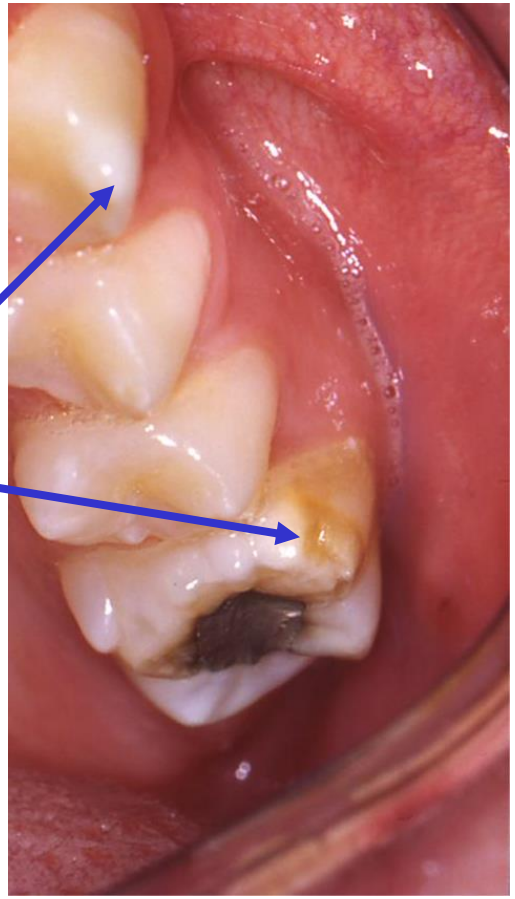


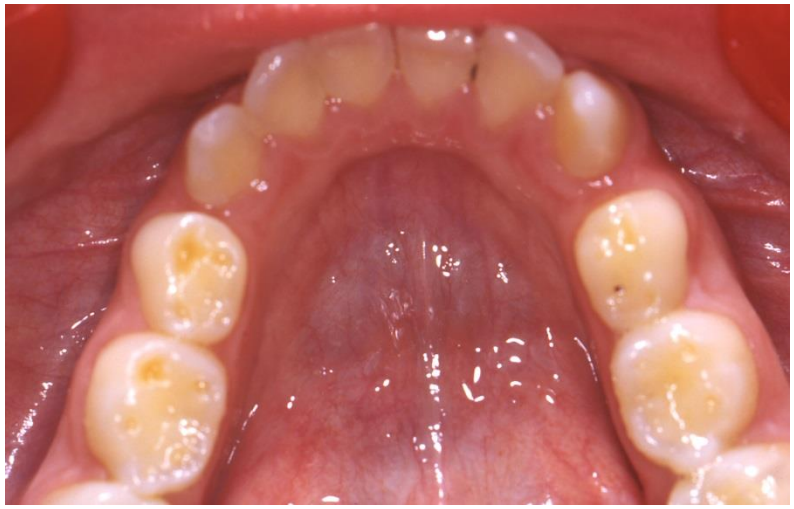




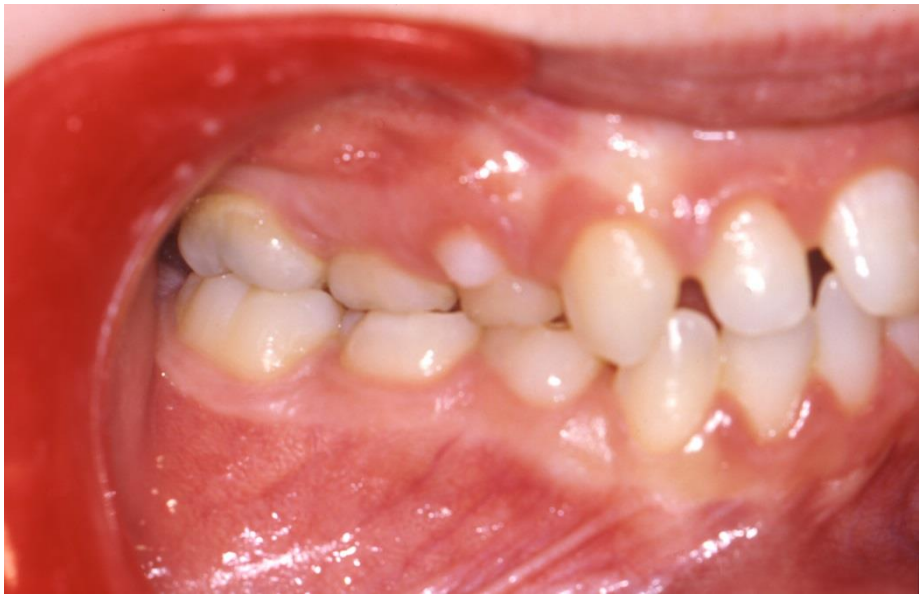
Developmental defects of enamel

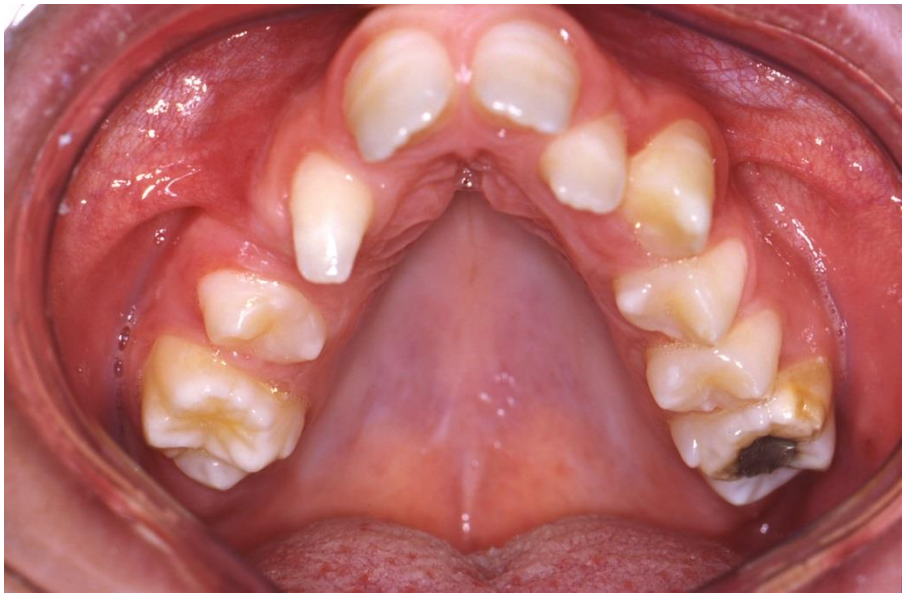
- 9,4% of the patients present at least one defect in primary dentition
- 18,5% of the patients in permanent dentition
- Type of defect :
  - Hypomineralisation = qualitative defect (yellow or white spots - opacities)
  - Enamel hypoplasia = quantitative defect



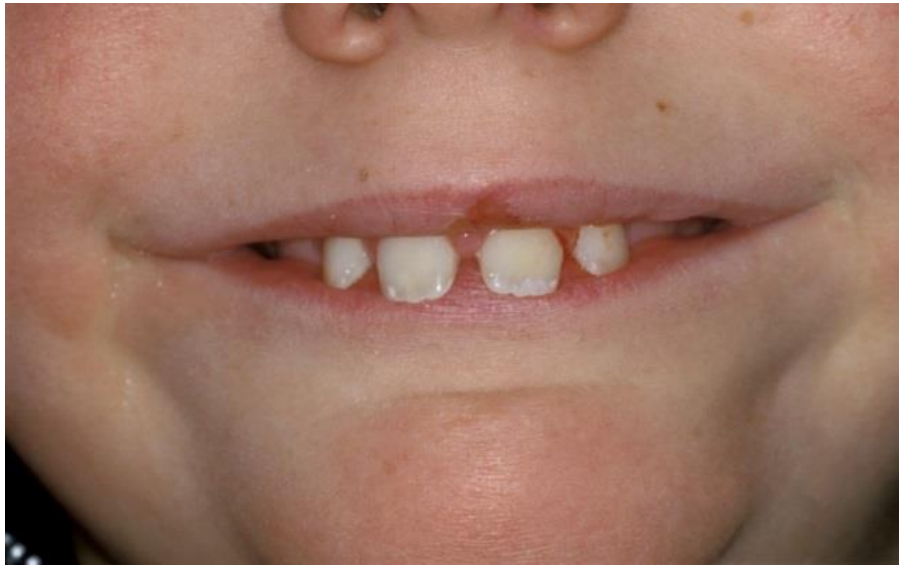


Malocclusions





- Higher prevalence of Class II and III occlusions
- Narrow or large maxilla - Deep palate
- Late exfoliation of primary teeth
- Ectopic eruption
- Crowding or spacing (missing teeth)
- Cross-bites
- Anterior Open-bite (25% of the patients)



Non specific pathologies

- Increased carious risk:
  - Mild intellectual deficiency
  - Behaviour management difficulties
  - Some cardiac medication may contain sucrose.
- At risk of gingivitis and periodontal diseases:
  - Poor oral hygiene practices, linked to the weak cooperation of the young patients, also associated with malocclusion and agenesis that limit the self-cleansing
  - Severe alteration of elastin components of gingival connective fibers, increasing the susceptibility of periodontal tissue to be attacked by plaque bacteria.

Medical and dental management

- The medical management of children with Williams syndrome requires a multidisciplinary team familiar with the organ systems affected as well as the developmental and behavioral problems.
- Guidelines exist for pediatricians and many tertiary centers now offer clinics for patients with Williams syndrome.
- The website of the Williams Syndrome Association (www.williams-syndrome.org) proposes helpful information for parents as well as physicians.
- The dentist should particularly pay attention to:
  - The congenital heart defects
  - The impaired orofacial motor function
  - The oro-facial anomalies
  - The hypersensivity to sound and of hypersensitive mouth
- The dentist has to evaluate the benefice risk of sedation and GA for patients with Williams syndrome. If conscious sedation used, careful monitor of the vital signs and oxygen saturation during the procedure

Dental management

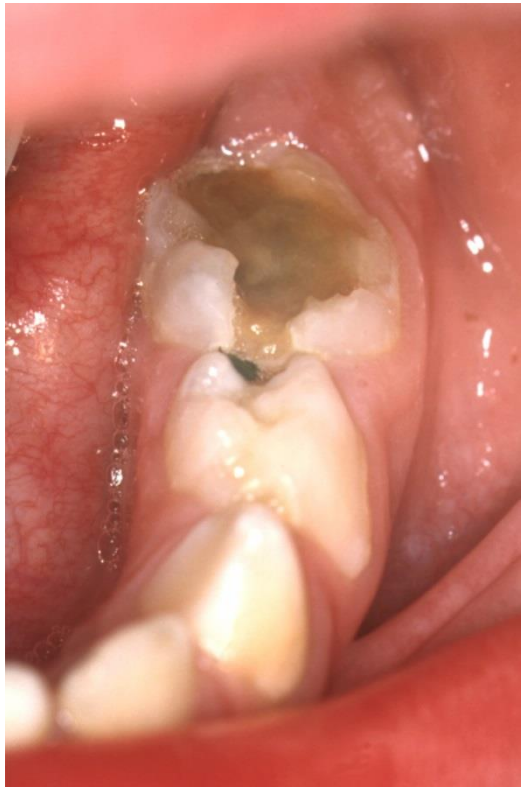
- Dental anxiety and communication disorders are present in almost all the children with Williams syndrome.
- One third of the children present several anomalies and/or pathologies as anomalies of number, size, shape and structure, or severe malocclusions. These malformations, associated with oral motor impairment, may cause oral dysfunction, beginning early in the infancy.
- The limited motor skills shown in patients with WS contributed to a poor oral hygiene, requiring a comprehensive preventive approach to reduce the caries risk and activity.
- A comprehensive radiographic examination may reveal the presence of dental anomalies, which are typical findings of this syndrome, and help planning the restorative and orthodontic rehabilitation.
- Nevertheless, orthodontic management is often limited.
- The use of headphones could be recommanded to minimize the sounds and noises of the dental office, which caused great discomfort to the child due to his hypersensitivity to loud sounds.

Management of infectious risk

More than 80 % of the patients have a cardiac malformation

Reduce the risk of infective endocarditis:

- Antibiotic prophylaxis before certain dental procedures
- Preoperative oral antiseptic mouth wash, such as 0,2% chlorhexidine gluconate
- Eliminate all dental infections
- Pulpotomy or pulpectomy contraindicated
- Evaluate benefice versus risk for orthodontic treatment



Dental care under general anesthesia

Risk factors:

- Congenital heart diseases
  - Exemple : death associated with general anesthesia in a Williams syndrome patient with a aortic stenosis (Burch et al, 2008)
- Complicated intubation linked to difficult airway:
  - Craniofacial/oral malformations: micrognathia, macroglossia
  - Breathing difficulties
- Sleep apneoa
- Drug interactions (with certain cardiac medication)

Recommandation for dental management and follow-up

Work together with other professionals as a member of the rehabilitation team!

1. Primary dentition
2. Mixted dentition
3. Adolescence
4. Adult

" Preventive dental protocols and dietary counseling must be individually designed and implemented "

1. Primary dentition

Age	Objectives	Management
12-24 months	Control of eruption Early diagnosis of dental anomalies Assessment of oral function (sucking, feeding abilities)	Parental counseling about preventive measures: tooth brushing methods, diet, use of fluorides… Prevention of mouth hypersensitivity
30 months	Control of eruption Early diagnosis of dental anomalies Assessment of oral function (eating disorders..) Introduction to the dental environment	Preventive program: oral hygiene, diet… Prevention of mouth hypersensitivity Topical application of fluoride, sealant (if indicated)
3 y	Control of eruption Diagnosis of dental anomalies Assessment of the occlusion and of oral habits Carious risk assessment	Intraoral radiographies if indicated Sealants if enamel defects Conservative treatment (if needed) Physiotherapy/orofacial therapy if indicated
4 y	Follow up	Preventive program Physiotherapy/orofacial therapy if indicated
5 y	Follow up	Preventive program Physiotherapy/orofacial therapy if indicated

2. Mixted dentition

Age	Objectives	Management
6	Control of eruption Diagnosis of dental anomalies Assessment of occlusion and of oral habits, tongue position, speech articulation problems Carious risk assessment	Preventive program Topical application of fluoride Panoramic Xray Sealant of the first permanent molars Orofacial therapy
7	Control of eruption Diagnosis of dental anomalies Assessment of occlusion and of oral habits	Preventive program Topical application of fluoride Sealant of the first permanent molars Orofacial therapy
8	Follow up	Preventive program Topical application of fluoride
9	Follow up Orthodontic examination	Preventive program Topical application of fluoride Panoramic Xray – lateral cephalogram Orthodontic treatment if indicated
10	Follow up	Preventive program

3. Adolescence

Age	Objectifs	Prise en charge
12	Diagnosis of sketetal malocclusion, tooth malpositions, oral soft tissue and dysfunction Evaluation of gingival health Eruption of permanent teeth	Preventive measures Topical application of fluoride Sealant of the second permanent molars Panoramic Xray
13	Follow up (one visit every year if low carious risk)	Preventive measures Orthodontic treatment
14	Follow up	Preventive measures Orthodontic treatment
15	Follow up	Preventive measures Orthodontic treatment
16	Follow up	Preventive measures Orthodontic maintenance

4. Adult

Age	Objectives	Management
16 - 20	Evaluation: occlusion, periodontal diseases, eruption of wisdom teeth Multidisciplinary evaluation if orthognatic surgery needed	Preventive program (gingivitis and periodontal diseases) Scaling Panoramic Xray
> 20	1 visit every year	Preventive program (gingivitis and periodontal diseases) Orthognatic surgery (if indicated)