the Child Dental Patient with

CLEFT LIP OR/AND PALATE

ORares
MALADIES RARES ORALES ET DENTAIRES
STRASBOURG
CRMR COORDINATEUR

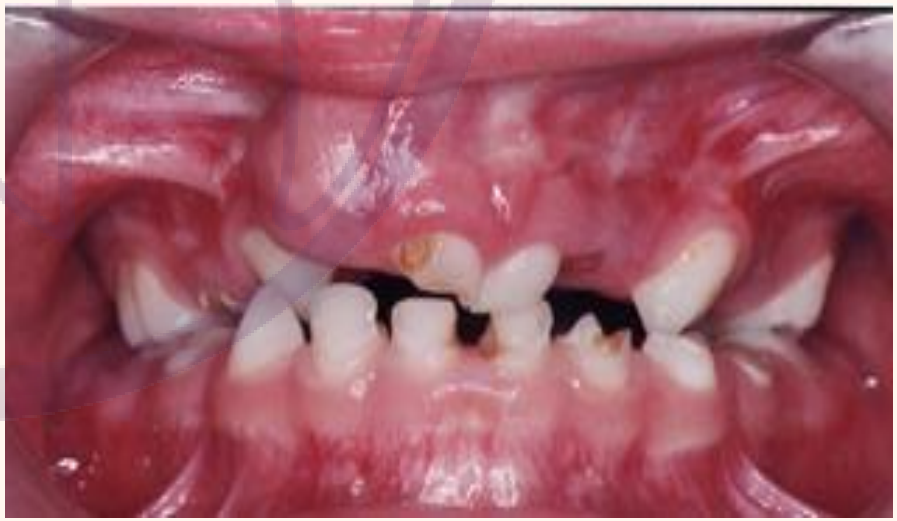
Les Hôpitaux Universitaires de STRASBOURG

ASOCIAȚIA NAȚIONALĂ DE STOMATOLOGIE PEDIATRICĂ
A.N.S.P.R.
DIN ROMÂNIA

UNIVERSITAS MEDICINAE ET PHARMACIAE "CAROLUS DAVILA" - BUCURESTIS
DOCENDO DISCITUR
A.D. 1857

Background

- Cleft lip or/and palate is the most common craniofacial malformation, affecting 1 in 700-1000 new-borns.
- There is a lack of continuity of tissues forming the lip, alveolar bone and soft/hard palate.
- Type of cleft: incomplete/complete; unilateral/bilateral.



CLEFTS

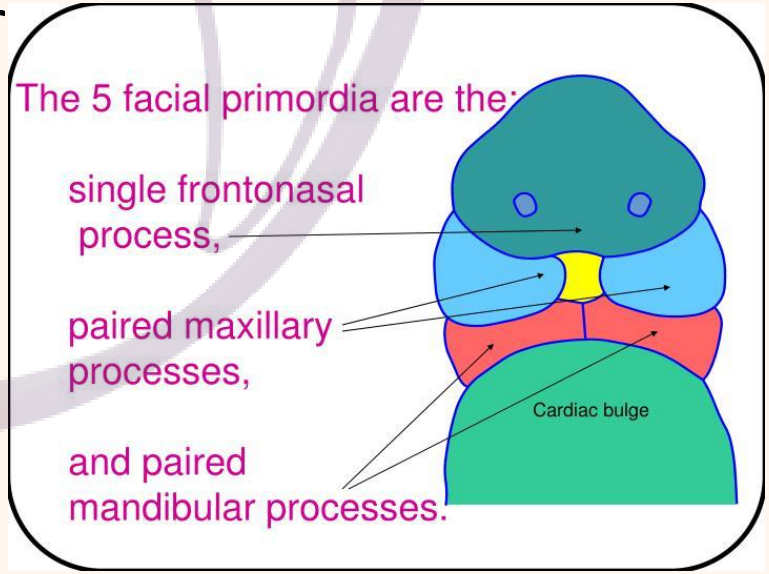
- isolated (non-sindromic)
- Syndromic = associated with other birth disorders or syndromes (Down's, Crouzon, Vander Woude's, Treacher Collins, Klippel-Fiel, Pierre Robin and oro-facial-digital syndrome)

Aetiology

- Multifactorial: genetic, environmental factors and gene-environment interactions.
- Family history in 1/3 cases
- The disrupting factor occurs between 5-8 weeks of intrauterine life.
- Cited factors involved : German measles, X-rays, drugs (Thalidomide), active smoking, alcohol, vitamin B deficiency (including folic acid), intensive exposure to UV light.

Aetiological mechanisms of clefts

- Differentiation and fusion of the different facial primordia : complex mechanisms with molecular regulation begin in the 4th week of embryonic development (ED) : formation of **single frontonasal, maxillary and mandibular processes**
- Spatial and temporal regulation by environmental and genetical factors : **genome-environement interactions**
- Underlying cellular and molecular mechanisms: cellular proliferation, neural crest cells migration, fusion, apoptosis

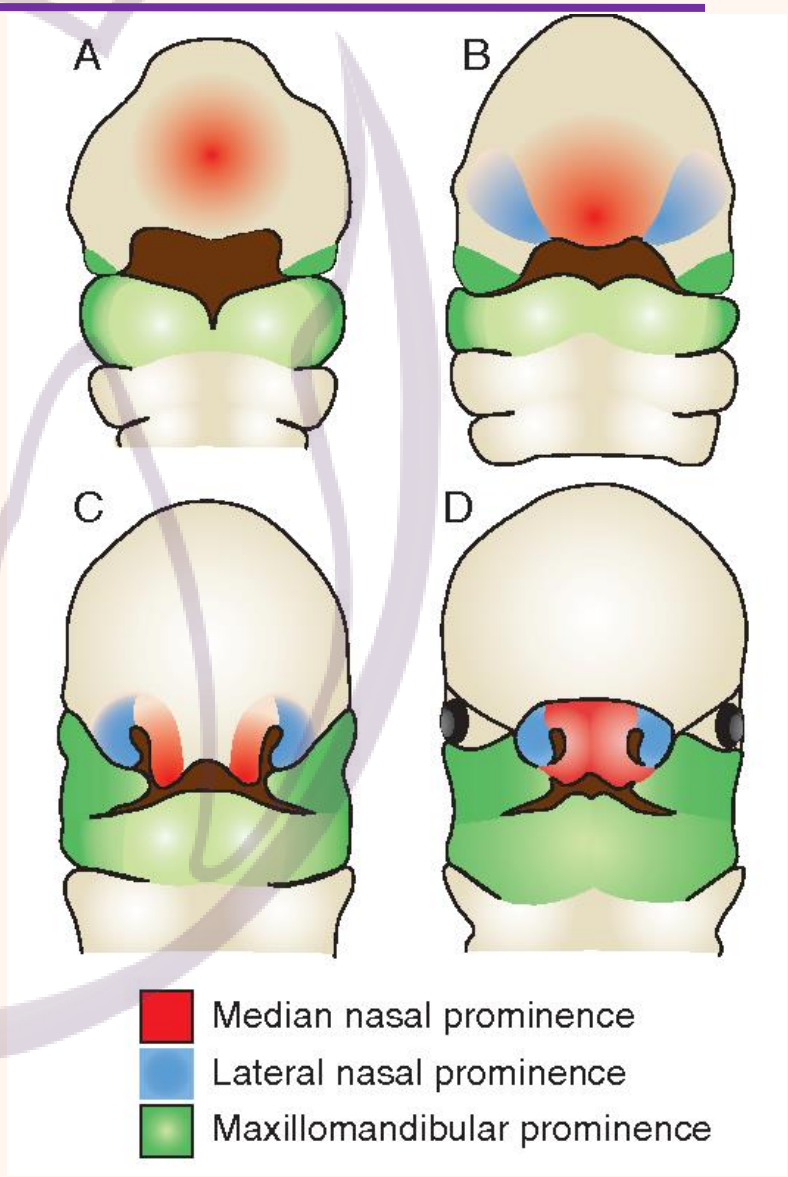


Aetiological mechanisms of clefts

- **6th-7th week of ED**: fusion of maxillary process-lateral and median nasal processes
  Defect at this stage: **labio-alveolar cleft with or without cleft palate**

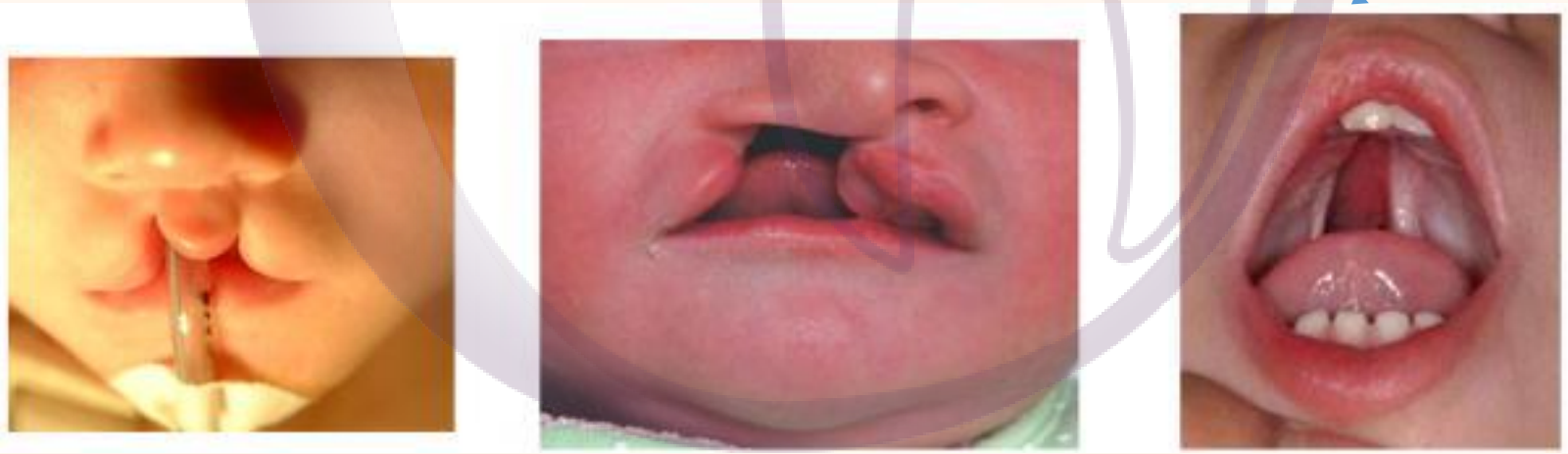
- **7th week of ED:** palatogenesis with development of palatal processes from the maxillary process and fusion on the median line (**MEE**: medial edge epithelia)

- Defects in development or elevation of the palatal processes : **secondary palatal cleft**



Aetiological mechanisms of clefts

- Strong clinical variability and genetic heterogeneity of clefts
- Early defects : lip and primary palate cleft
- Late defect of palatogenesis: secondary palate cleft



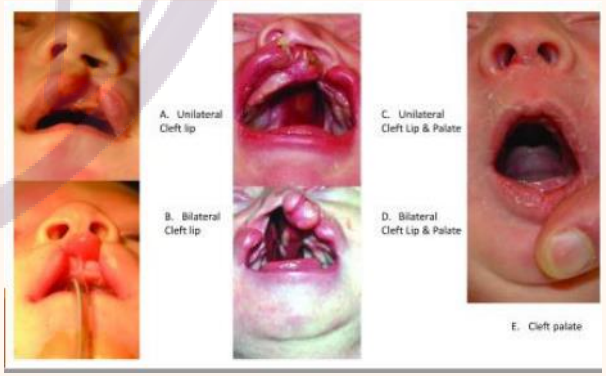
Leslie et al., 2013

Non-syndromic CLP: genetic susceptibility

Different molecular approaches are available:

- linkage analysis: use of polymorphic markers of informative families
- Genome Wide Association Studies (GWAS): SNP (Single Nucleotid Polymorphisms) research distributed along the genome
- direct sequencing, DNA-chip analysis or exome sequencing
- Identification of pathogenic polymorphisms**: IRF6, PAX7, PAX9, MSX1, BMP2, BMP4**

Main difficulty: phenotypic and genetic heterogeneity



Identification of genetic mechanisms

- Familial forms of non-syndromic CLP with different intra-familial transmission modes
- Complex segregation analysis (CSA) and studies on monozygotic twins confirmed the genetic mechanisms involved in CLP

Consistency in monozygotic twins: 40-60%
Dizygotic twins: 3-5%

Pre-natal diagnosis

- Ultrasound examination may detect clefts of the lip and alveolus;
- Cleft palate is difficult to diagnose through routine screening;
- Around the 20th gestational week → examination, cephalic presentation of the child, low body mass index of the mother;
- Genetic counselling of the family and gathering information about family history of exposure to any teratogenic factors;
- Amniocentesis;
- Involvement of the cleft team as soon as possible → counselling and treatment strategies;
- Psychological and emotional support for the family.

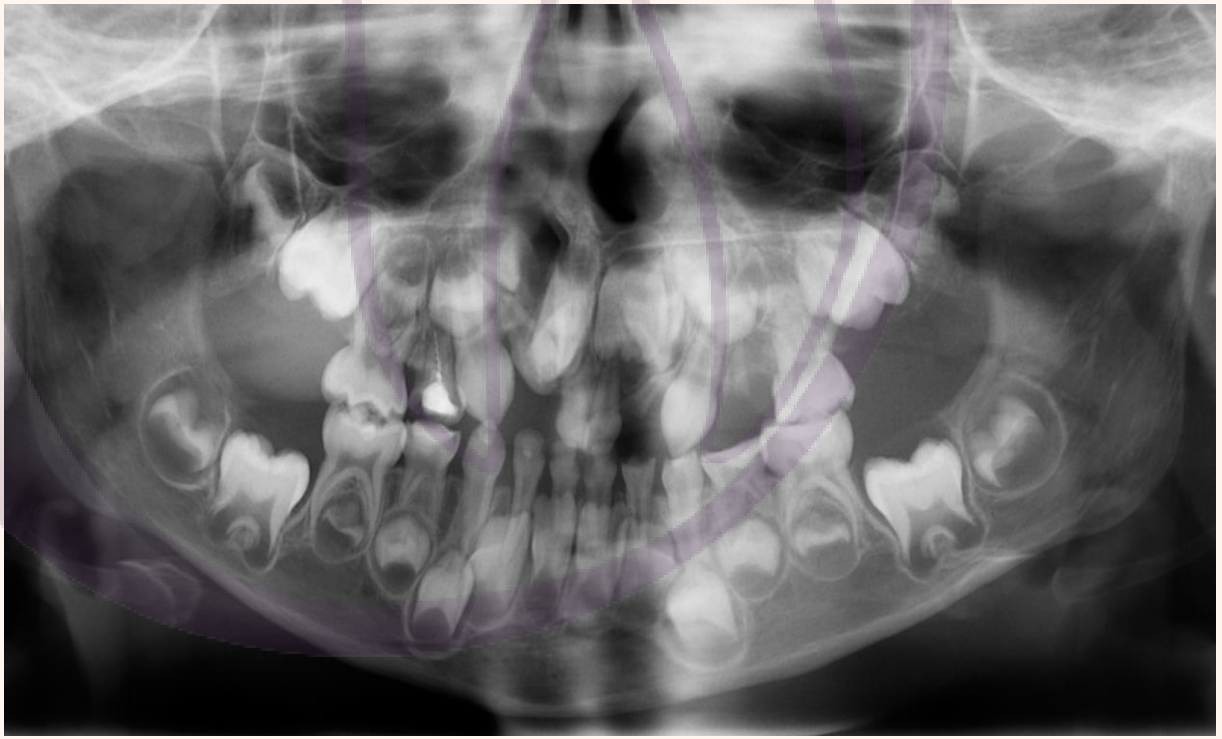
Clinical features

- Facial asymetry → aesthetic problems
- Mid-face retrusion→ profile is generally concave
- Speech problems → nasal twang in the voice and difficulty in articulation, due to velo-pharyngeal insufficiency
- Disorder of the middle ear → hearing difficulties and ear infections
- Inability to feed due to poor oral suction, nasal regurgitation of fluids→ risk of chocking and excessive air intake
- Increased nose width, reduced mouth width, nose asymmetry, increased nose width/mouth width ratio, reduced upper lip length, reduced lip elasticity

- Shy, nervous or uncooperative behaviour

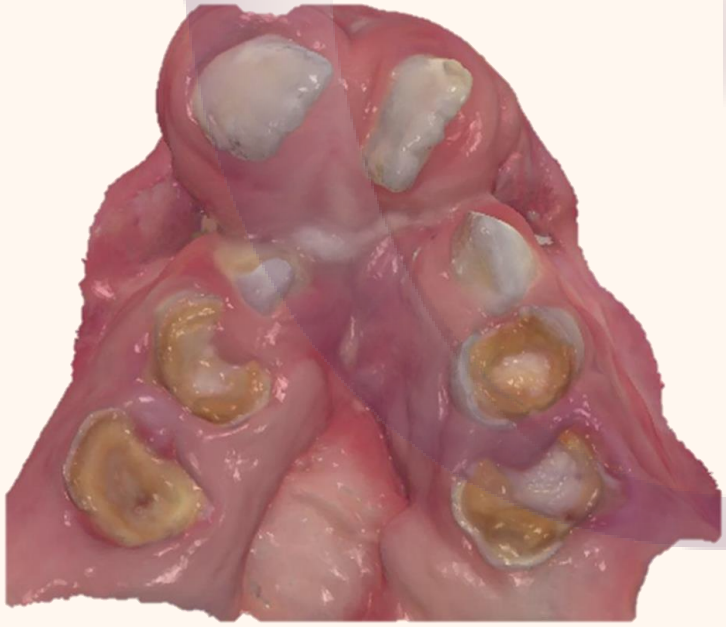
Dental abnormalities

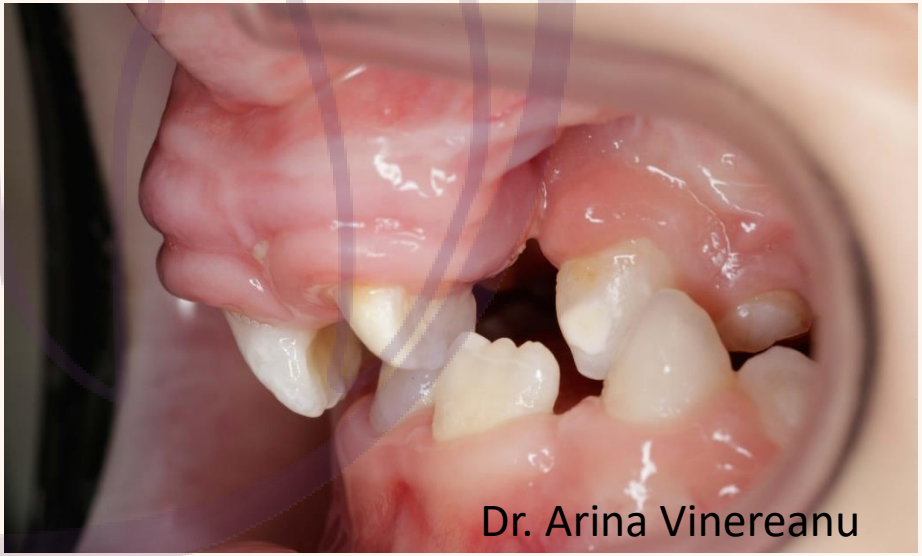
- Anomalies of number→ congenitally missing teeth (especially lateral incisors and second premolars) and supernumerary teeth
- Natal/neonatal teeth
- Ectopic eruption (palatally)
- Impaction
- Transposition



Dental abnormalities

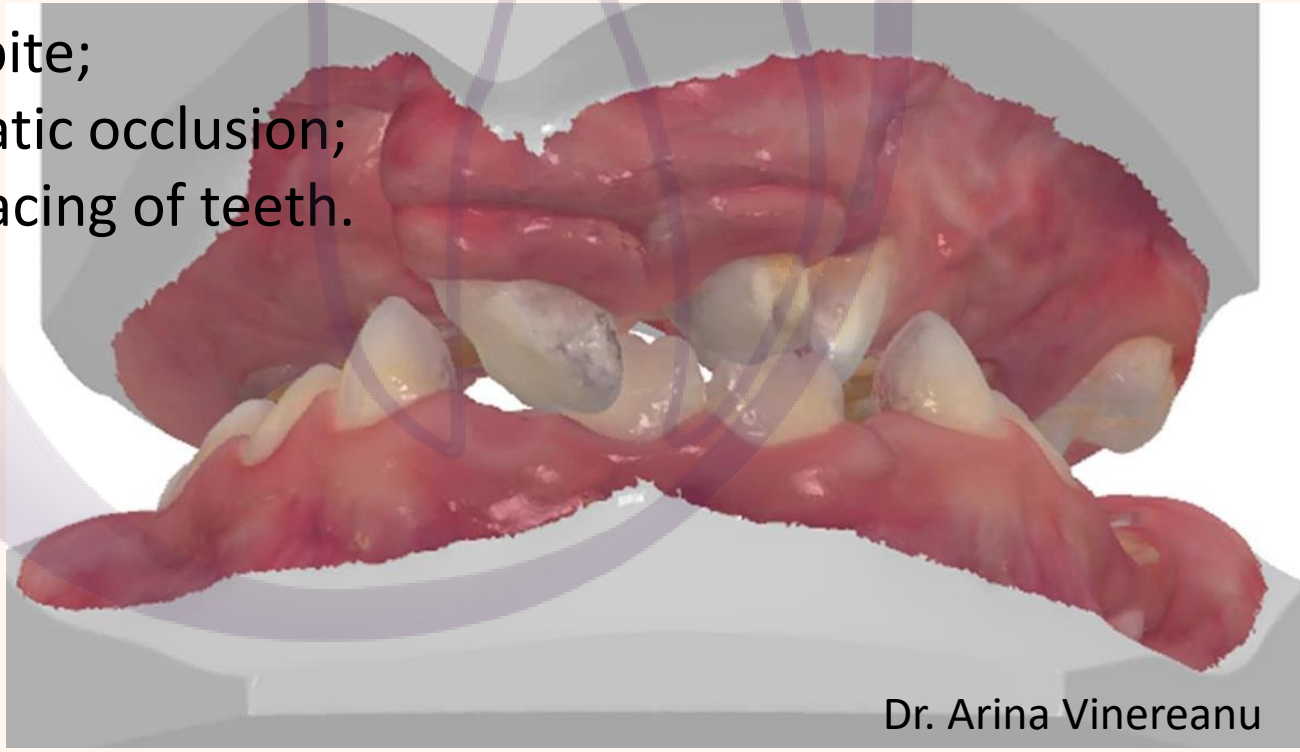
- Tooth rotations and deviations of axial root inclination
- Anomalies of tooth size →micro- and macro-dontia
- Anomalies of shape → fused teeth, conical crowns, aberrations in crown shape
- Anomalies of tooth structure→ enamel hypoplasia/ hypomineralization





Orthodontic problems

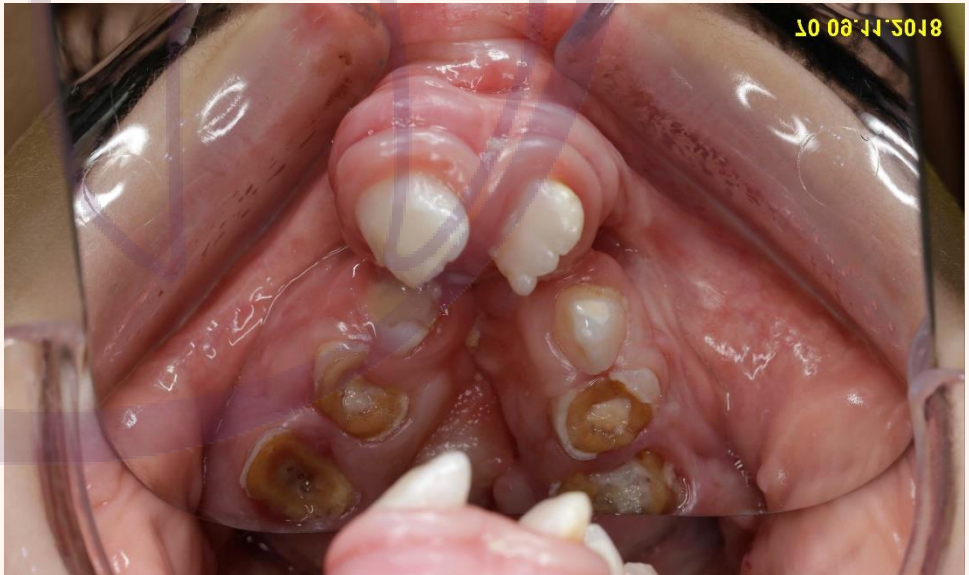
- Maxillary hypoplasia and retrognatia;
- Development of class III occlusion;
- Buccal segment open bite;
- True/pseudo mandibular prognatism;
- Deep bite;
- Posterior crossbite;
- Anterior traumatic occlusion;
- Crowding or spacing of teeth.



Dental caries

- Increased caries prevalence when compared with children without clefts;
  - more prone to caries due to altered tooth structure and malocclusions

- Higher DMF-T values compared to individuals without cleft;
- Early loss of deciduous teeth due to caries → loss of arch length.



Periodontal diseases

- Poor gingival health;
- Tendency to neglect oral care → gingivitis and periodontitis;
- Crowding is a favouring factor;
- Periodontal trauma due to multiple malpositions and maloclusion;
- Permanent teeth adjacent to the cleft have a deficiency of supporting alveolar bone → susceptible to premature loss.

Management

- Interdisciplinary team of specialists:
  - Cleft Nurse Specialist;
  - Cleft Surgeon;
  - Speech and Language Therapist;
  - Orthodontist;
  - Paediatric Dentist;
  - Audiologist and/or Ear, Nose and Throat (ENT) Specialist;
  - Paediatrician;
  - Clinical Psychology, Genetics and Sonography.

- Interventions for cleft patients start as early as intrauterine and continues into late adulthood.

Preventive approach

- Special teat and bottles that favour milk delivery to the back of throat → easier swallowing;
- Cup and spoon method → useful for some babies;
- Palatal prosthesis → seal the cleft;
- Dietary awareness in relation to dental caries;
- Regular dental examinations (every 3- 4months)→ routine prophylaxis and fluoride varnish;
- Modified tooth brushing technique, using a small-headed soft toothbrush and lifting the lip to allow the labial surfaces to be cleaned;
- Toothpaste with high fluoride content from young ages:
    - 1000 ppm from the first tooth
    - 1450 ppm after age 3

Surgical treatment

- Pre-surgical orthopaedics and naso-alveolar moulding → can reduce the defect and tension in soft and cartilaginous tissues → will facilitate surgical soft tissue repair;
- Cleft lip repair → ages of 1 and 6 months;
- Cleft palate repair → ages of 5 and 15 months;
- In mixed dentition (6-12 years): alveolar bone grafting to correct the maxillary defects and to provide maxillary-alveolar ridge continuity for tooth eruption and alignment;
- When growth is complete → Orthognathic surgical correction of maxillary retrognatia;
- Surgical revision of the nose (rhinoplasty) will be the last surgical step.

Dental management - Paediatric dentist

- Monitor and investigate the eruption of permanent teeth (especially front teeth);
- Fissure sealing of permanent molars/ premolars as they erupt;
- Atraumatic restorative technique for initial carious lesions (without risk of pulp contamination);
- Restore (stainless steel crowns), rather than extract deciduous molars →keep arch length;
- Maintain supernumerary and/or malpositioned deciduous teeth adjacent to the cleft → preserve bone tissue that is already defective in this region;
- Rubber dam isolation for dental treatment whenever possible (especially in cases of unrepaired cleft palate);
- Anaesthesia might be more painful and difficult to perform (secondary scar fibrosis);
- Anaesthesia of the palatal region is always necessary.

Orthodontic treatment

- Monitor facial growth → planning and timing interventions;
- Mixed dentition → interceptive orthodontic treatment: reposition the teeth adjacent to the cleft; correct the transverse relationship using palatal expansion appliances;
- Permanent dentition → definitive orthodontic treatment;

- **Objectives**:
  - maintain teeth and supporting structures,
  - correct impacted and transposed teeth,
  - manage congenitally missing teeth;
- Extractions and prosthetic treatment may be taken into consideration (implants and bridges);
- Scar tissue formation might affect the prognosis.