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Hippocrates provided the first description of craniostenoses in 100 B.C. ✓

He noted the variability in appearance of the calvarial deformities and correlated it with the pattern of cranial sutural involvement. ✓

CLINICAL AND RADIOGRAPHIC FINDINGS

Scaphocephaly-Sagittal Synostosis

- ✓ Premature fusion of the sagittal suture is characterized by a narrow, elongated cranial vault and reduced bitemporal dimension.
- ✓ It occurs predominantly in males .

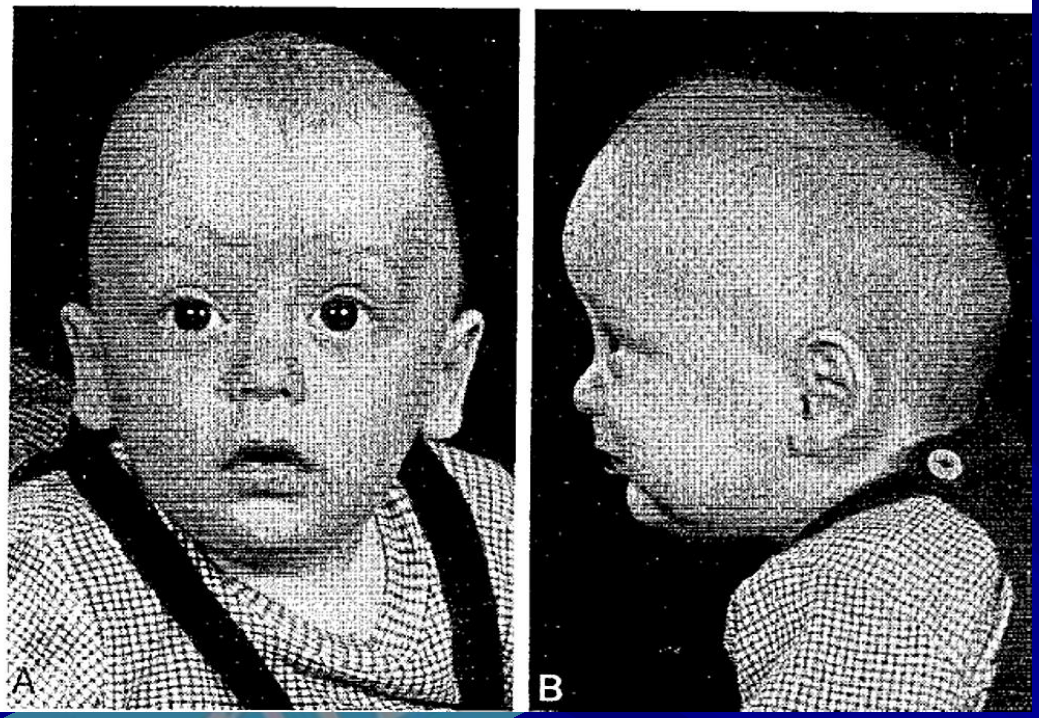

Figure 61-2. Scaphocephaly-sagittal synostosis in an infant., *A,* Frontal view. B, Profile.

Figure 61-3. Scaphocephaly-sagittal synostosis in an adult. *A,* Frontal view. B, Three-quarters view.

Trigonocephaly-Metopic Synostosis

There is a triangular-shaped deformity of the anterior cranial fossa and forehead resembling a midline keel. ✓

✓The orbits are medially displaced, with an associated hypotelorism.

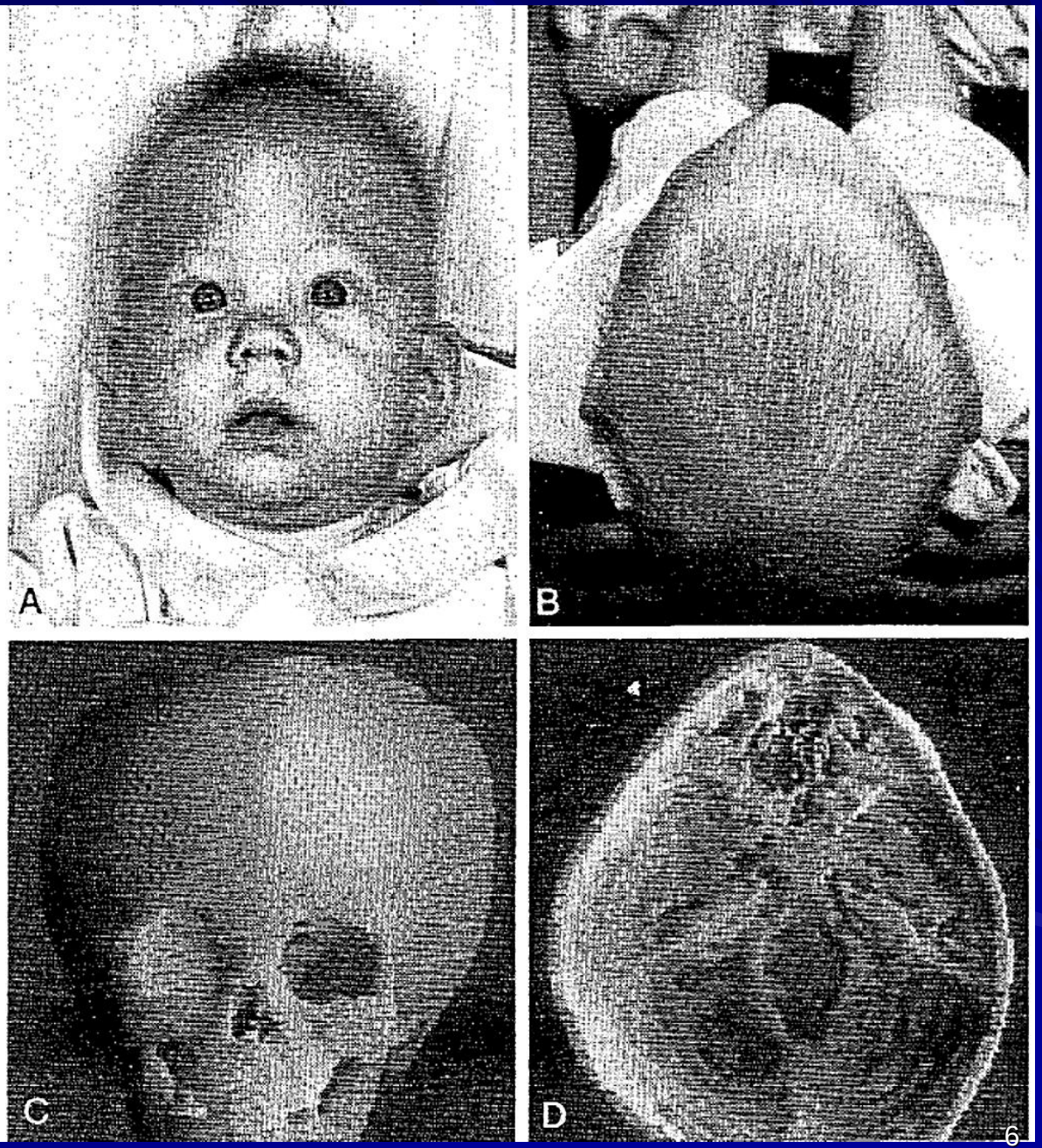

Figure 61-4. Trigonocephaly-metopic synostosis in an infant. *A,* Frontal view. Note the orbital hypertelorism. B, Superior view. C, Three-dimensional CT view of the trigonocephaly forehead deformity and the associated orbital hypotelorism. 0, Three-dimenslonal CT intracranial view.

Brachycephaly-Bilateral Coronal Synostosis

- Fusion of both coronal sutures is associated with a reduction of the anteroposterior dimension of the cranial vault and a compensatory increase in the bitemporal distance. ✓
- A mild degree of exophthalmos can be observed if the supraorbital rim is recessed. ✓

Figure 61-5. Brachycephaly-bilateral coronal synostosis in an infant. *A,* Frontal view, *B,* Profile,

Plagiocephaly-Unilateral Coronal Synostosis

- Flattening of the forehead and recession and elevation of the brow and superolateral aspect of the orbit are observed on the affected side. ✓
- On the contralateral side, persistent growth produces frontal bossing, inferolateral orbital dystopia, and bulging of the occipital prominence. ✓
- The nasal tip is usually deviated to the affected side and the ear on the more affected side can be more superiorly and anteriorly positioned. ✓

These findings can be found in patients without coronal synostosis like Unilateral craniofacial microsomia or muscular torticollis. ✓

Oxycephaly-Multiple Suture Synostoses.

Oxycephaly, literally translated as "***pointed head,"*** is characterized by a retroverted forehead, tilted posteroinferiorly on a plane with the nasal dorsum.

The forehead is usually reduced in the horizontal dimension and capped by an elevation in the region of the anterior fontanel.

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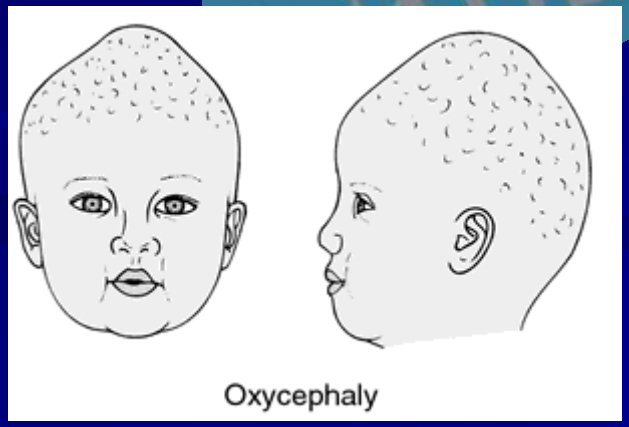


Crouzon's Disease

Described by a French neurologist in 1912, ✓
Crouzon's disease

exorbitism and midface retrusion. ✓

Figure 61-9. Crouzon's disease in an infant.. Note the exorbitism, midface hypoplasia, and turricephaly. *A,* Frontal *view.* B, Profile.

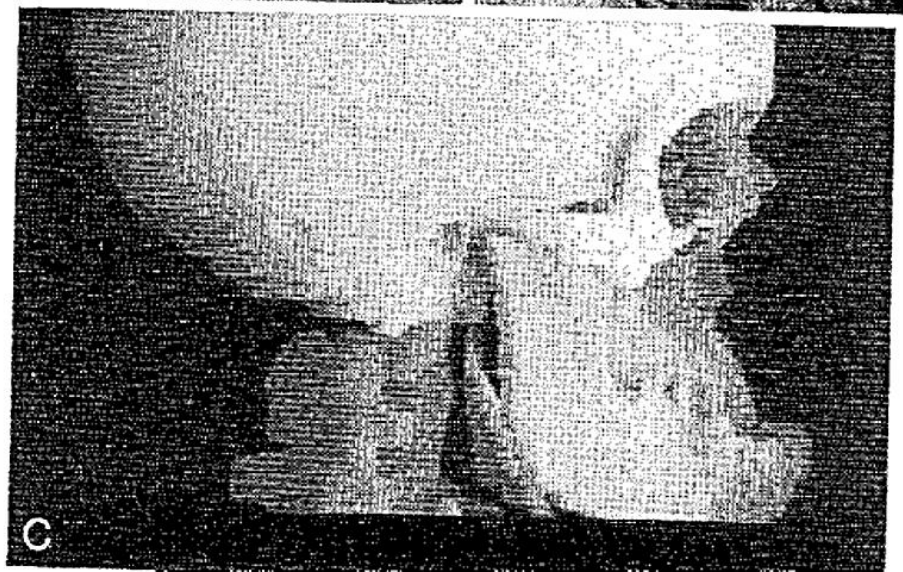
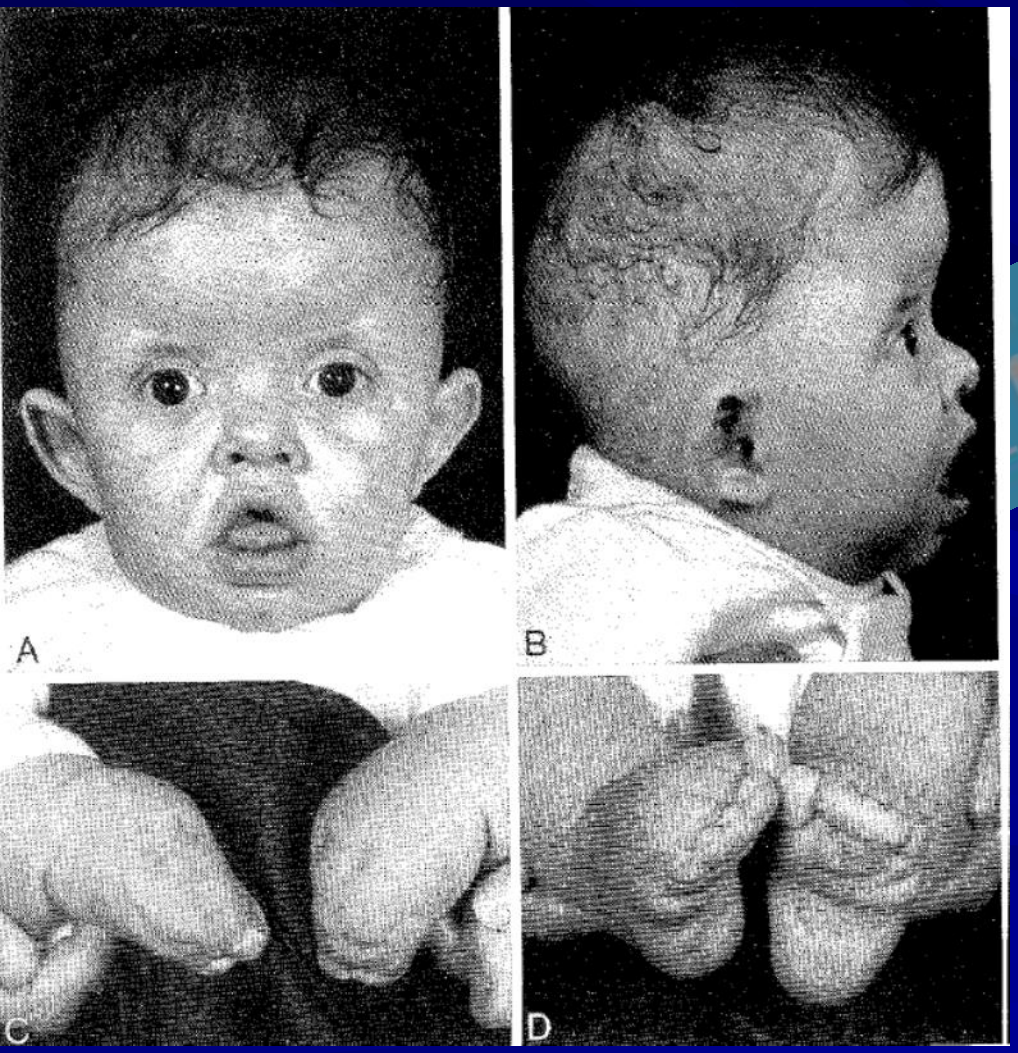


Figure 61-10. Crouzon's disease in a young female. Note the moderate exorbitism and midface hypoplasia. *A,* Frontal *view.* B, Profile. C, Three-dimensional CT scan illustrating the degree of midface hypoplasia and anterior crossbite in a patient with craniofacial synostosis.

Apert's syndrome



- The acrocephaly, ✓ frontal bossing, midface hypoplasia, and open bite.


A
B

Hemifacial microsomia

(Goldenhar syndrome)

_ unlateral or bilateral asymmetrically hypoplastic ears and ramus

_ ear tags and / or pits

_ micrognathia

_ cardiac defects

_ variably cleft lip or palate

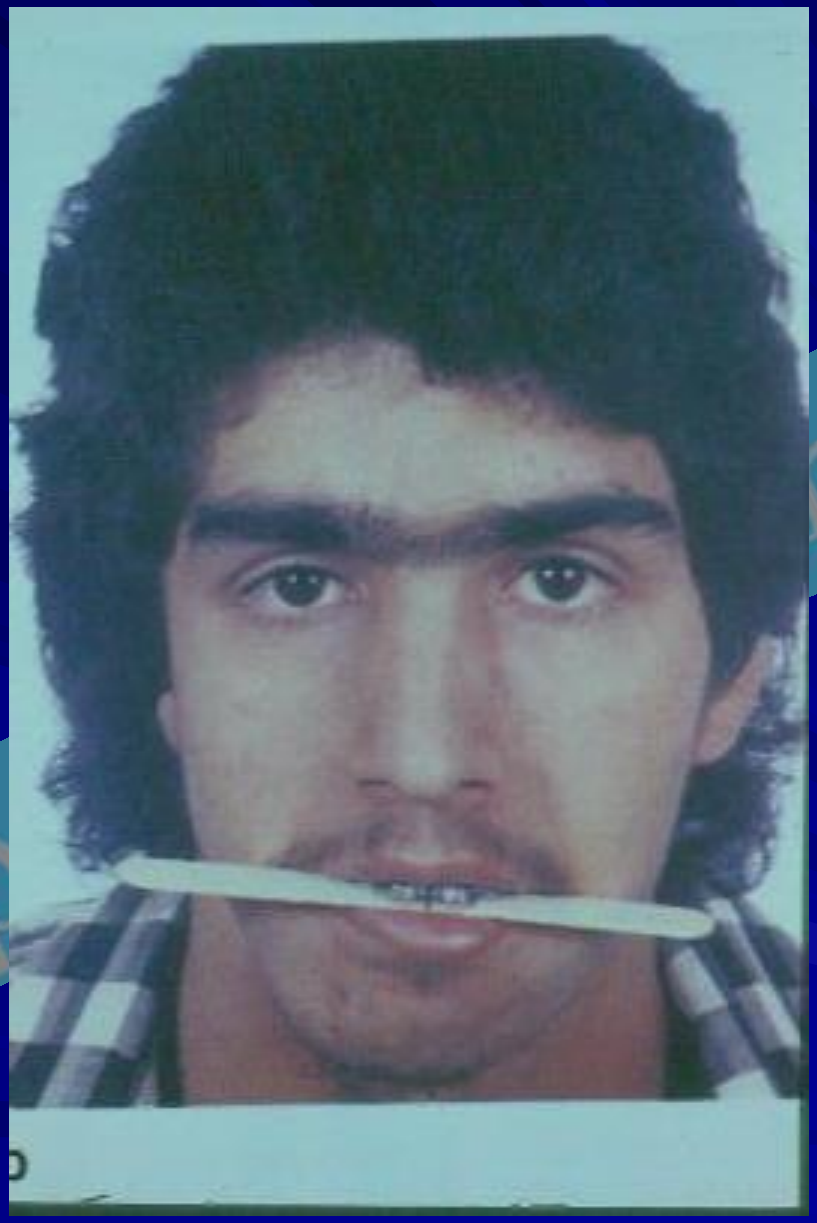
_ vertebral anomalies

Treacher collins syndrome

_ Symmetrically hypoplastic low _ set ears

_ Micrognathia

_ Some times cleft palate



Mandibular deficiency

_ Robin syndrome

_ Treacher collins syndrome

_ Nager acrofacial dysostosis

_Wildervanck _ smith syndrome

_Hemifacial microsomia (Goldehar syndrome)

_ Mobius syndrome

_ Hallermann _ streiffsyndrome

Pierre Robin syndrome

Microganthia , cleft palate , glossoptosis

Robin syndrome is part of stickler syndrome

Anterior open bite

_ Amelogenesis imperfecta

_ Beckwith _ wiedemann syndrom

Amelogenesis imperfecta

_ Discolored teeth : hypomaturation , hypoplasia or hypocalcification of enamel

_ Anterior open bite

Osteogenesis imperfecta

_ fragile bones

_ blue sclerae

_ deafness

_ dentinogenesis imperfecta tooth condition

_ mandibular prognathism

Mandibular prognathism

_ Basal cell nevus syndrome (Gorlin syndrome)

_ Klinefelter syndrome

_ Marfan syndrome

_ Osteogenesis imperfecta

_ Waardenburg syndrome

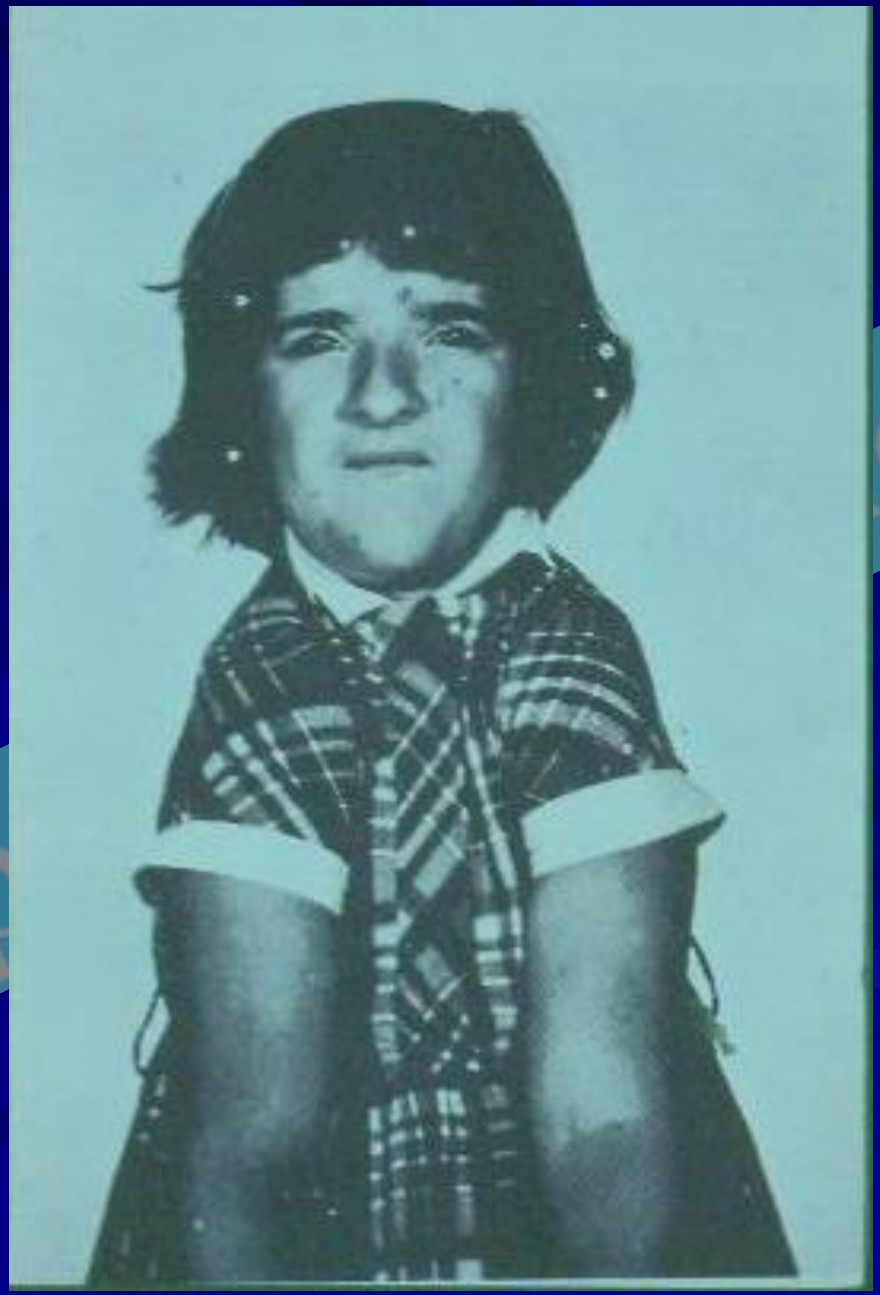
Cleidocranial dysostosis

_ Unilateral , bilateral , partial , absence of clavicle

_ Delayed cranial suture closure

_ Maxillary retrusion

_ Mandibular protusion



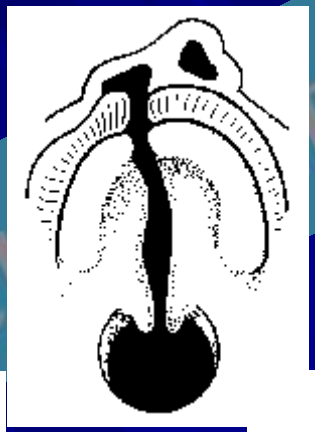
b

Classification of Clefts and Clinical Features

Clefts are classified as

unilateral or bilateral cleft of the lip .1
unilateral or bilateral cleft lip and palate .2
Palatal cleft .3
bifid uvula .4

Types of Clefts



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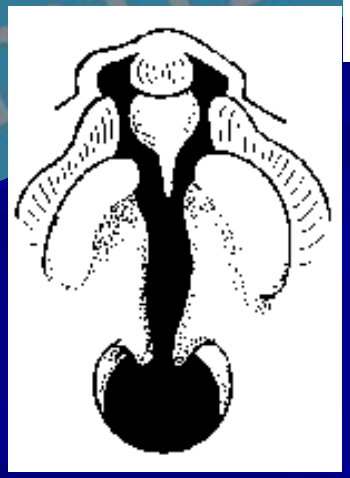

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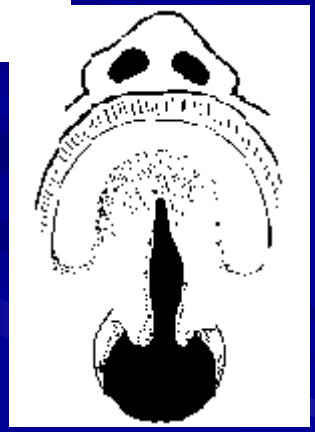

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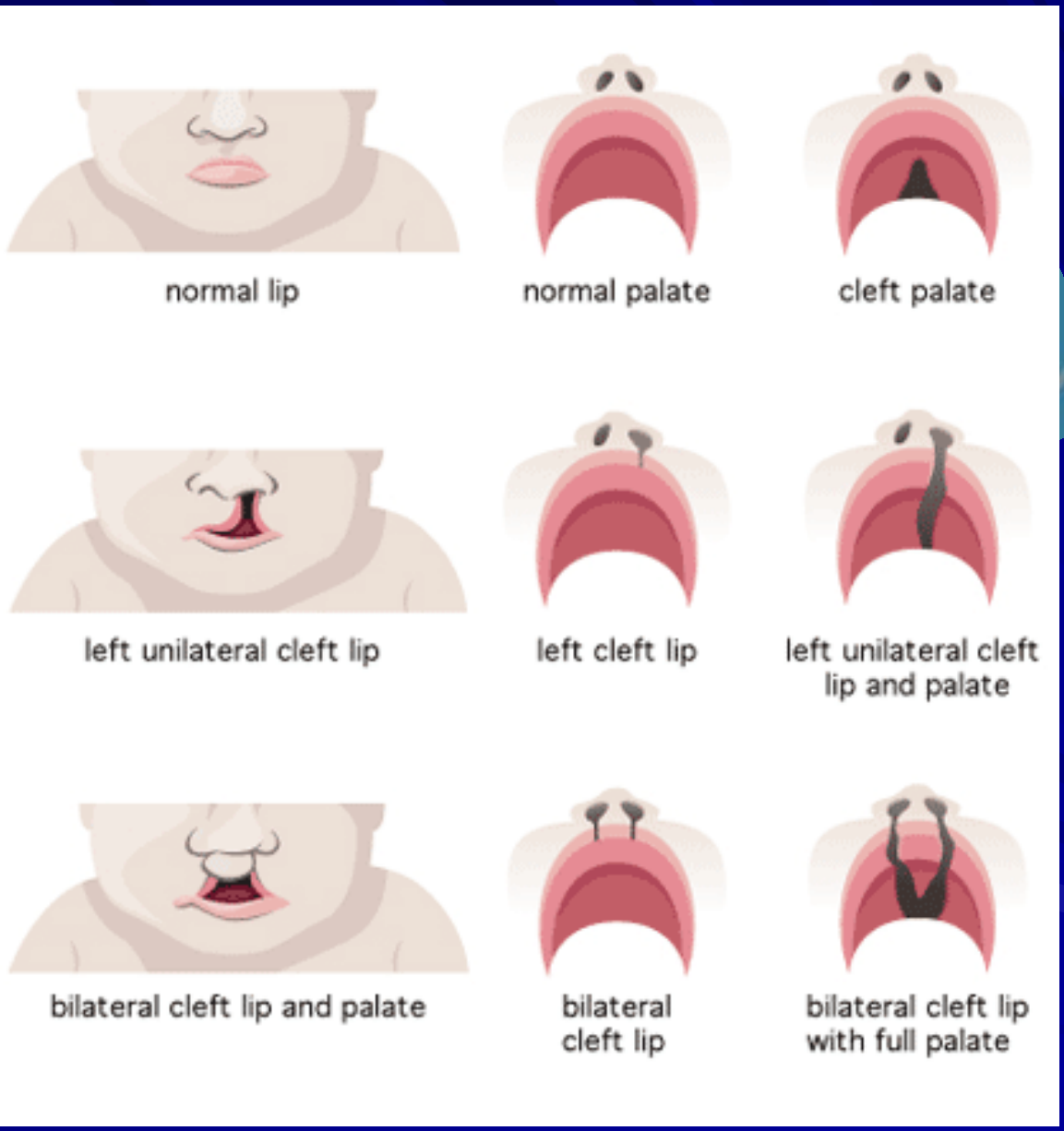

normal lip
normal palate
cleft palate
left unilateral cleft lip
left cleft lip
left unilateral cleft lip and palate
bilateral cleft lip and palate
bilateral cleft lip
bilateral cleft lip with full palate

Cleft Palate : Various theories have been given for its development.

- Failure of tongue to drop down

- Non fusion of palatal shelves

Team Consists Of:

- Pediatrician
- Orthodontist
- Pedodontist
- Oral surgeon
- Plastic surgeon
- Psychologist
- Speech therapist
- Prosthodontist

Genersl surgeon

Gender

.

- Clefts palate alone is found in approx 1 in 1000;
- females are more often affected than males.

ETIOLOGY

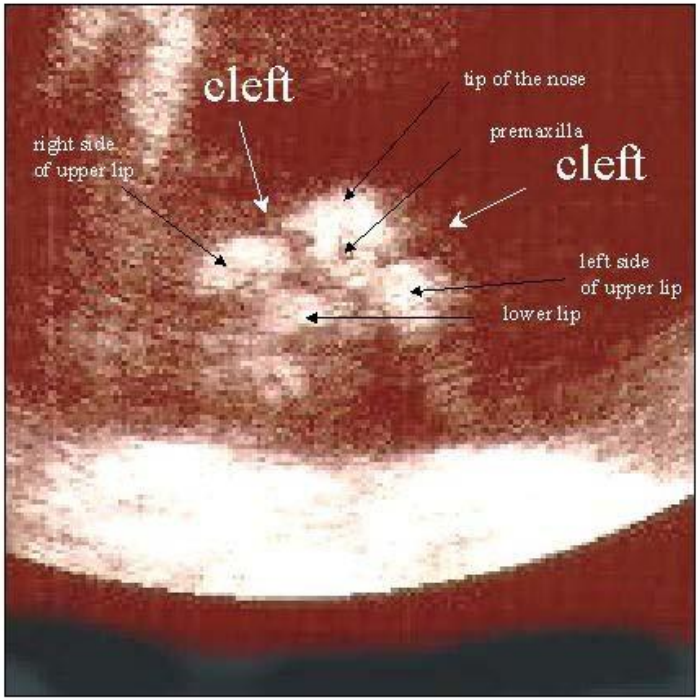
1. Genetic factors
2. Nutritional disturbances during development
3. Physiologic, Emotional or traumatic stresses during developmen
4. A mechanical disturbance where the size of the tongue may prevent the union of parts
5. Various environmental factors like infections (e.g. Rubella), exposure to radiation, drugs like thalidomide, antiepileptic durgs, hormonal pills, etc.
6. Maternal consumption of alcohol and smoking

Diagnosis

- Physical examination at birth
- Ultrasound: not always visible
- Genetic testing for parents to determine risk of having additional children with cleft lip/palate

Ultra Sound at 18th Week of Pregnancy

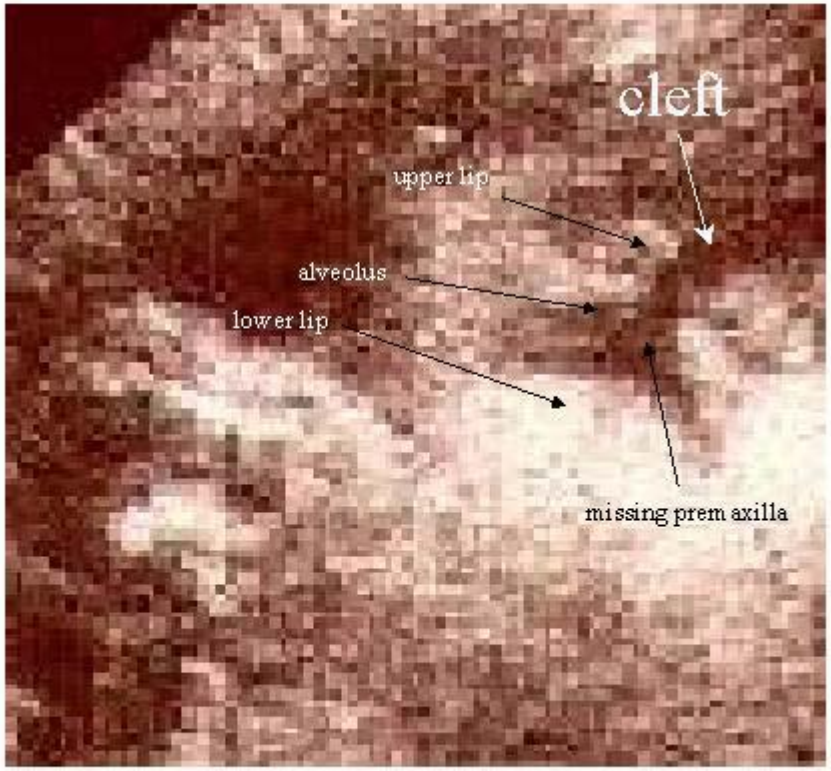
Bilateral cleft lip - *Ultrasound at the18th week of pregnancy*

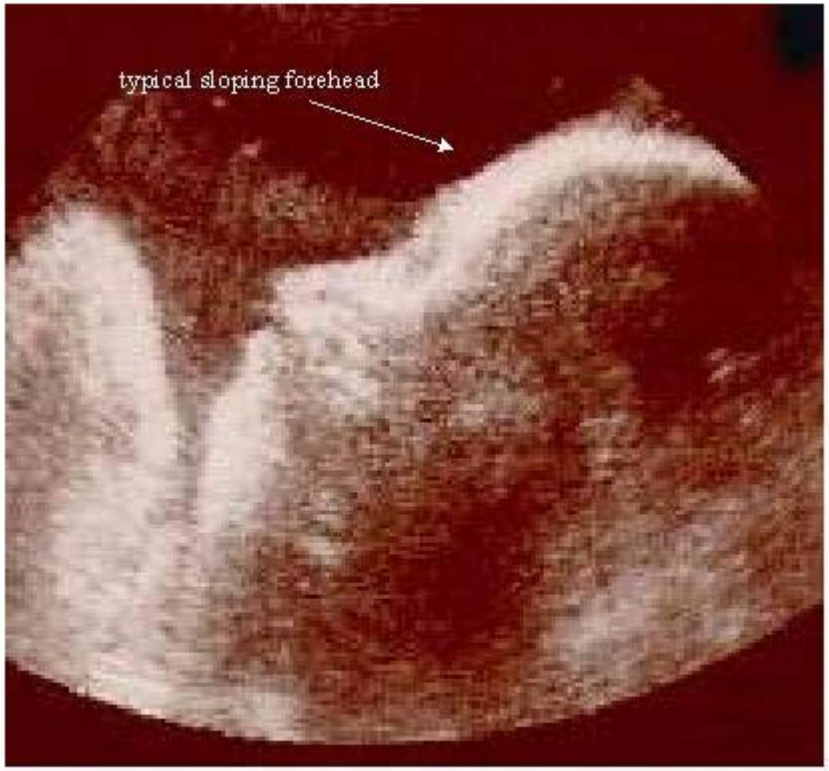


Courtesy of Dr. P. Calda, Prague (calda@obgyn.anet.cz)

Median cleft lip in holoprosencephaly sequence

- *Ultrasound at the 16th week of pregnancy*





Courtesy of Dr. P. Calda, Prague (calda@obgyn.anet.cz)

Stages in cleft lip / palate treatment

1- Presurgical infant orthopedics

2- Lip closure

3- Palate closure

4- Speech therapy

5- Early orthodontics

6- Alveolar grafting

7- Comprehensive orthodontics

8- Pharyngeal flap surgery

9- Orthognathic surgery

10- Fixed prosthodontics

Stage : presurgical infant orthopedics

Age : 1 to 4 weeks

Comment : Repositioning palatal segments can facilitate lip repair , done less frequently now

Lip closure

Age → 10 weeks

Weight → 10pounds

Hemoglobin → 10 grams

Stage : lip closure

Age : 8 to 12 weeks

Comment : may be preceded by preliminary lip adhesion as an alternative to presurgical orthopetics

Soft palate closure → 12 months

Hard palate closure → 1 years

Soft and hard palate closure → 18 ,24 months

Closing only the soft palate initially is an alternative , but one stage closure of the hard and soft palate is the usual procedure

Hard and soft palate closure

Early closure

- Normal speech
- Maxillary underdevelopment

In cleft

. Laterals are missing or undersized

. Supernumerary teeth are common

Stage : speech therapy

Age: 6 to 11 years

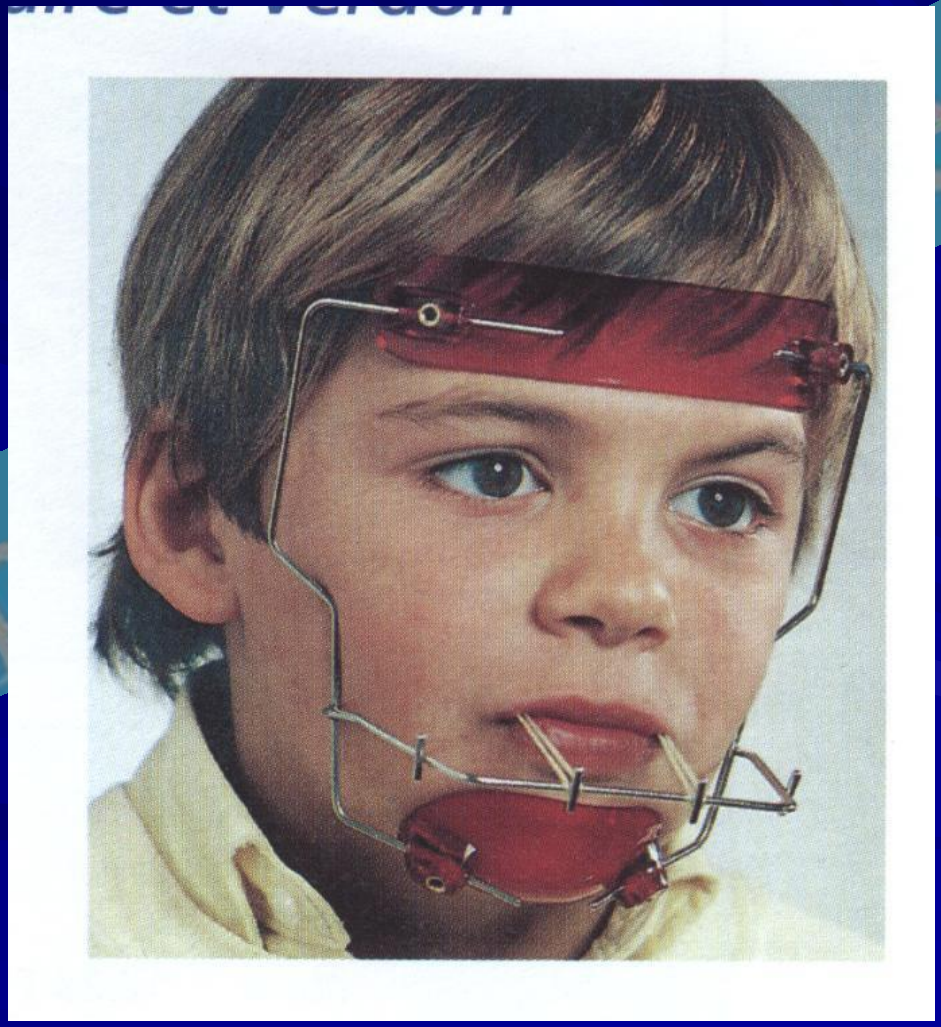
Comment : Articulation errors often develop as child tries compensate for cleft

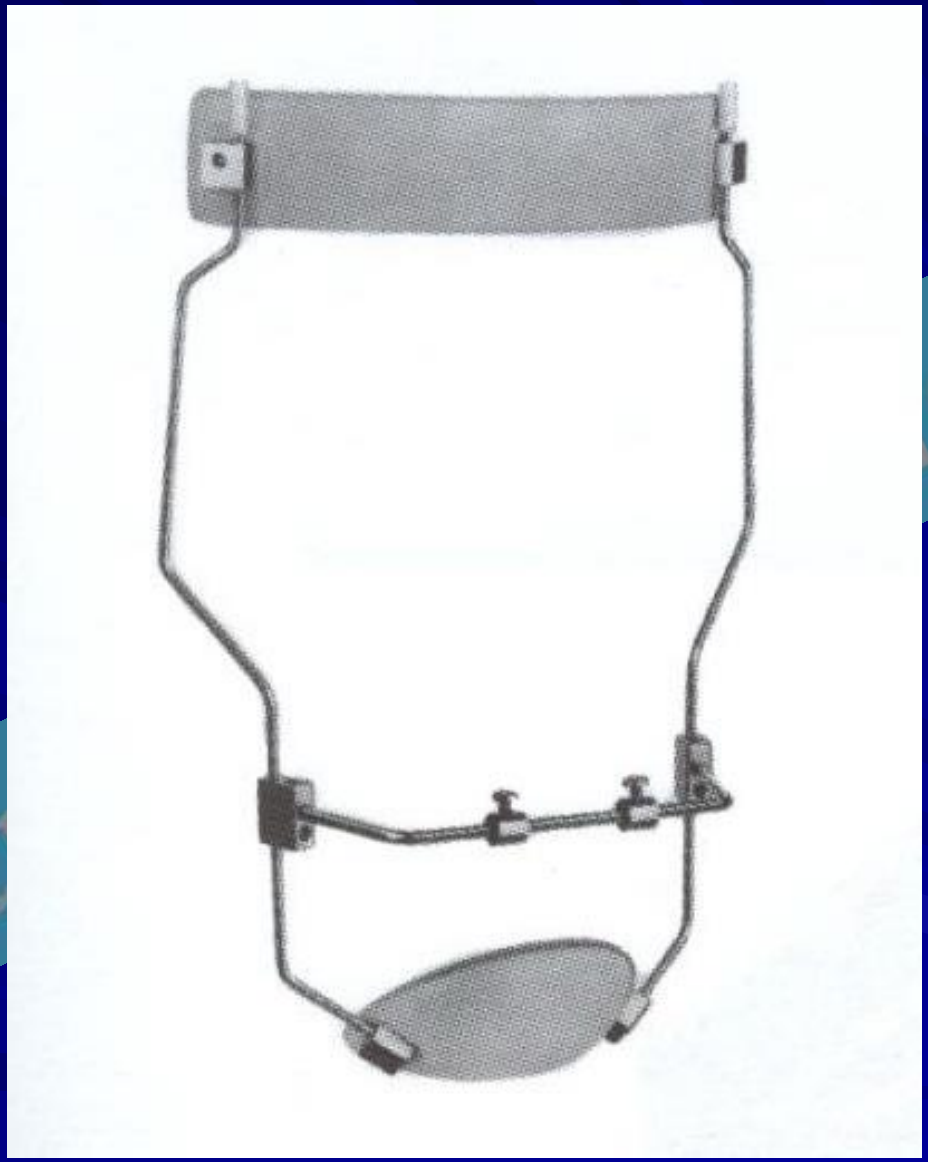
Stage : early orthodontics

Age : 7 to 8 years

Comment : usually incisor alignment and maxillary transverse expansion

Delaire Face Mask





In this girl of 7 years with a Cl III malocclusion and a receding midface , anterior traction was applied to the maxillary structures .

Goals of early orthodontics

. Correct of incisor malalignment

. Correct of incisor rotation

. Correct of anterior cross bite

. Correct of posterior cross bite

Stage : Alveolar grafting

Age : 6 to 10 years

Comment : Needed before permanent canines Erupt : timing determined by stage and sequence of dental development

The ideal time for alveolar graft

. As late as possible in maxillary growth

. Before the eruption of teeth

Stage : comprehensive orthodontics

Age : 11 to 14 years

Comment : Class III elastics and reverse chin cup often very helpful

Stage : orthognathic surgery

Age : 17 to 19 years

Comment : maxillary advancement , mandibular set back

Stage: fixed prosthodontics

Age :17 to 19 years

Comment: replacement of missing lateral , temporary bridge when fixed orthodontic appliance removed , comprehensive treatment after growth completed .

Stages in cleft lip and plate treatment

Presurgical infant orthopedics	1 to 4 weeks
Lip closure	8 to 12 weeks
Palate closure	18 to 24 month
Speech therapy	6 to 11 years
Early orthodontics	7 to 8 years
Alveolar grafting	6 to 10 years
Comprehensive orthodontics	11 to 14 years
Pharyngial flap surgery	9 to 19 years
Orthodontic surgery	17 to 19 years
Fixed prosthodontics	17 to 19 years

Mead Johnson/Enfamil Cleft Feeder

Special Needs Feeder / Haberman Feeder

Pigeon Feeder

Dr. Brown's Natural Flow *to relieve gas*

Scar tissue

- It may take several months to form.
- Maxillary deficiency

Treatment plane

1. Growing patients

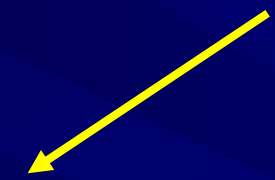
2. Nongrowing patients

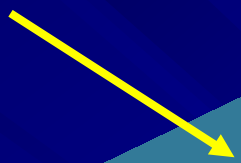
Treatment of Cl III

Growing → Growth modification

Non growing
- Camouflage
- Surgical

Growth Modification





Maxillary Deficiency

Mandibular Excess

Treatment plan in growing patients

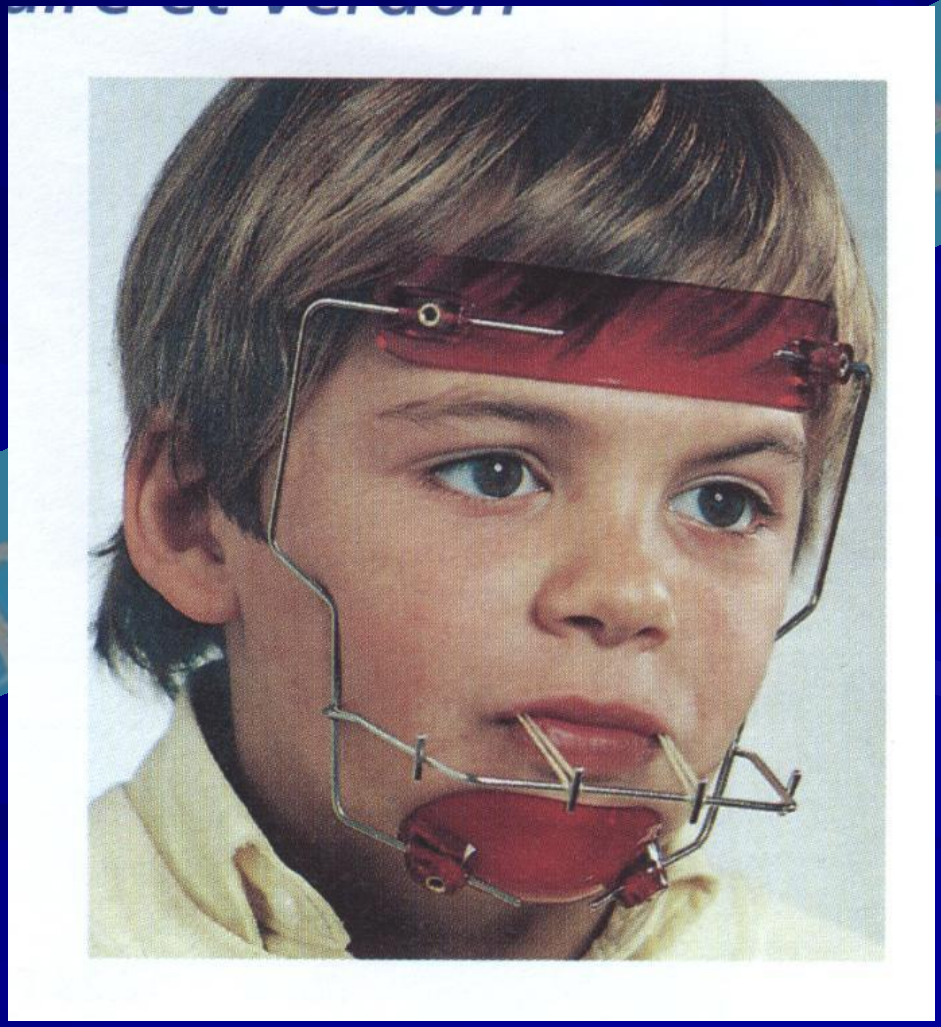
lateral : expansion

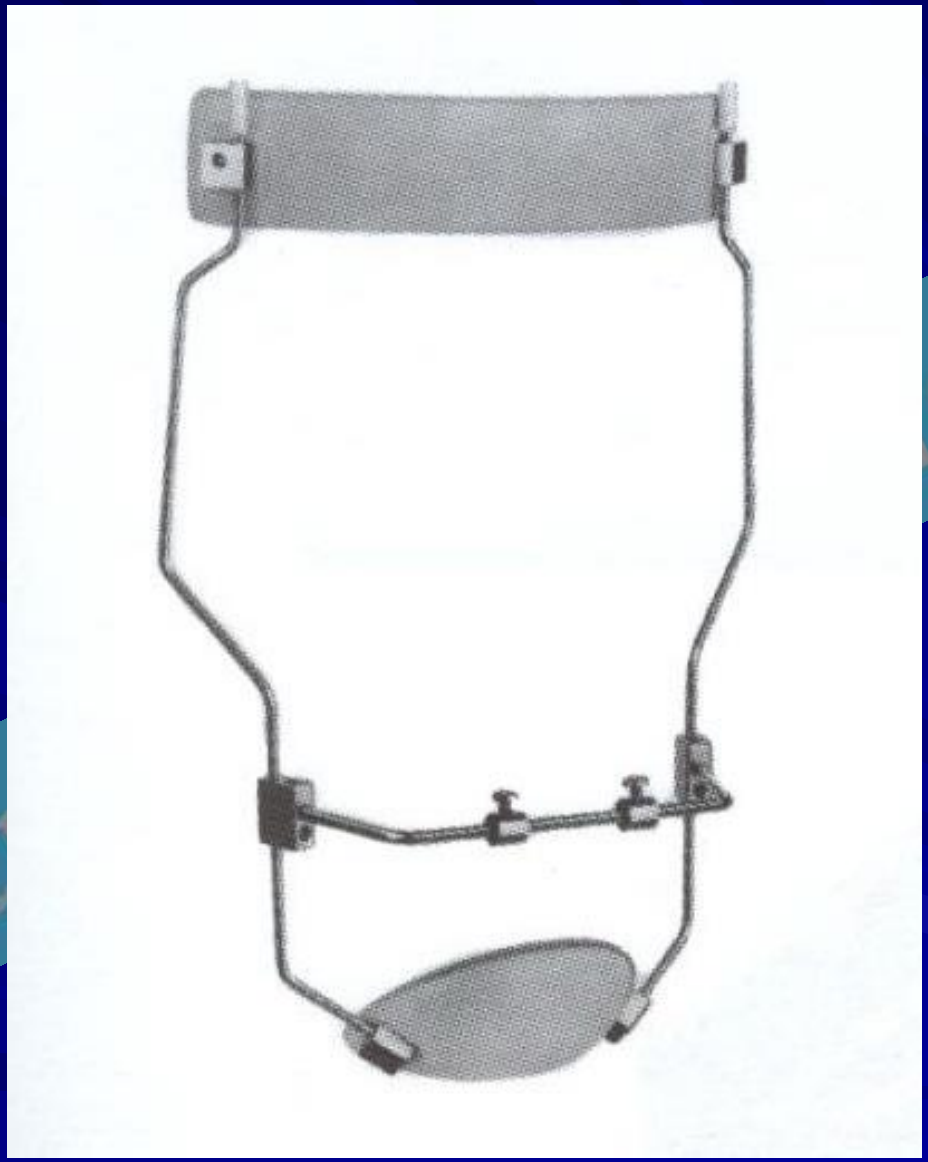
Antero posterior

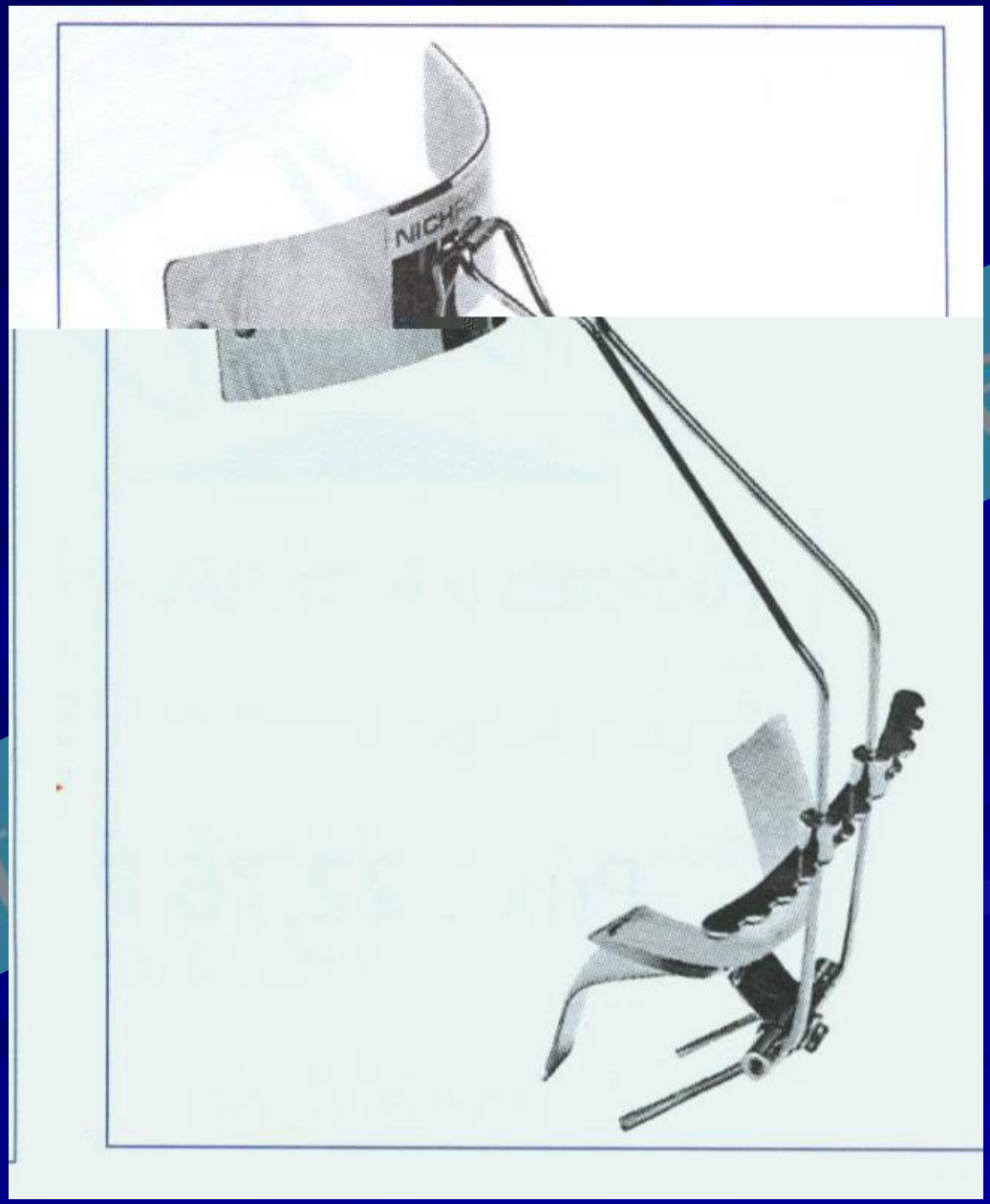
- **Reverse chin cup**
- **Face mask**
- **Cl III elastic**
- **Tongue appliance**
- **Tongue plate**

In this girl of 7 years with a Cl III malocclusion and a receding midface , anterior traction was applied to the maxillary structures .

Delaire Face Mask

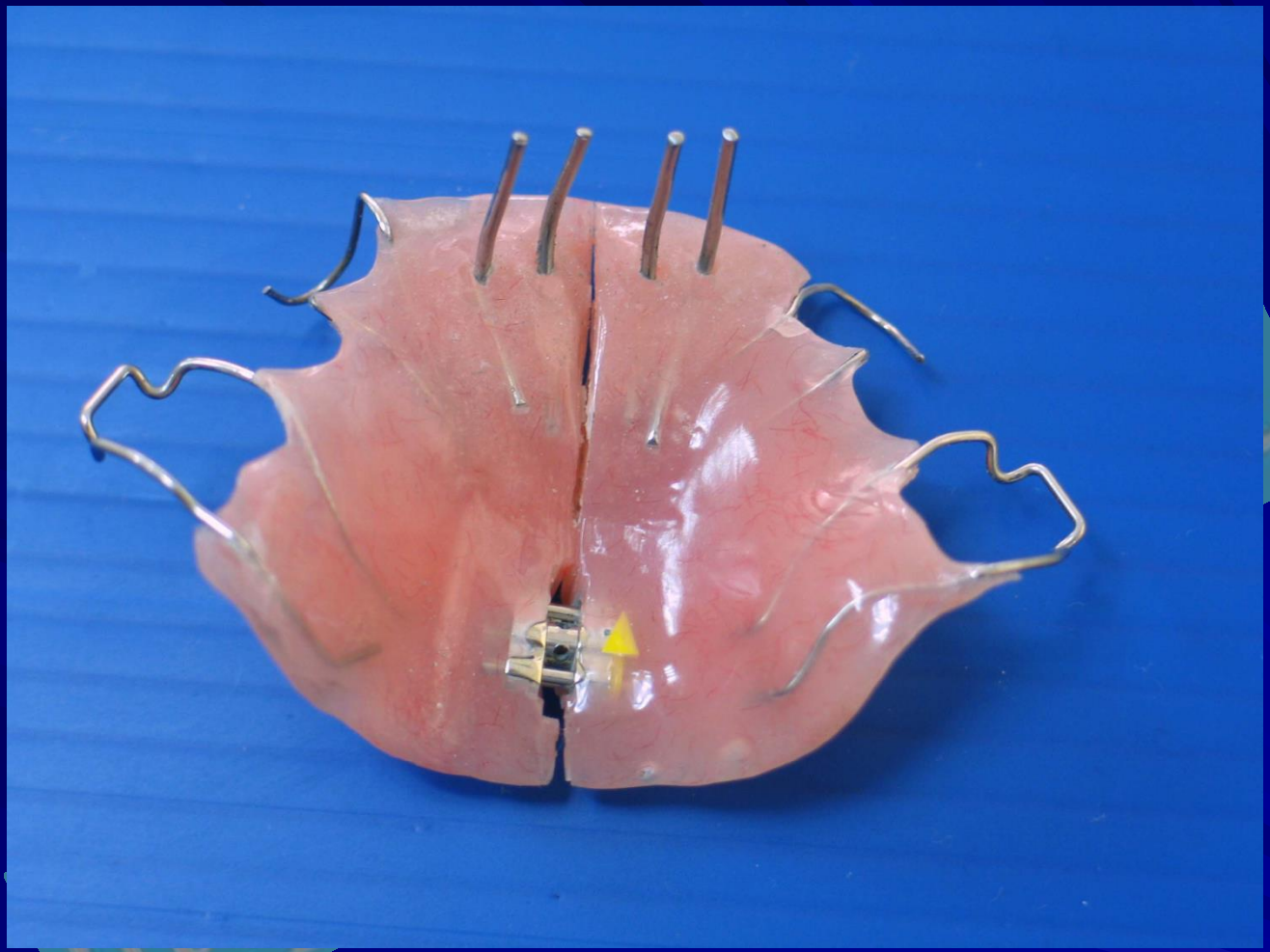






Tongue appliance structure

1. Base plate
2. Posterior bite plate
3. Palatal cribs
4. Clasps



Tongue appliance phylosophy

1. Swallowing → 17.5 minutes

2. Rest
   - Position of the tongue
   - Position of the sprue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Quadhelix for expantion

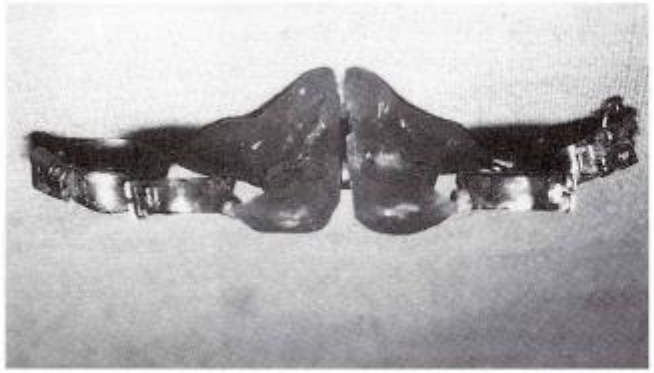
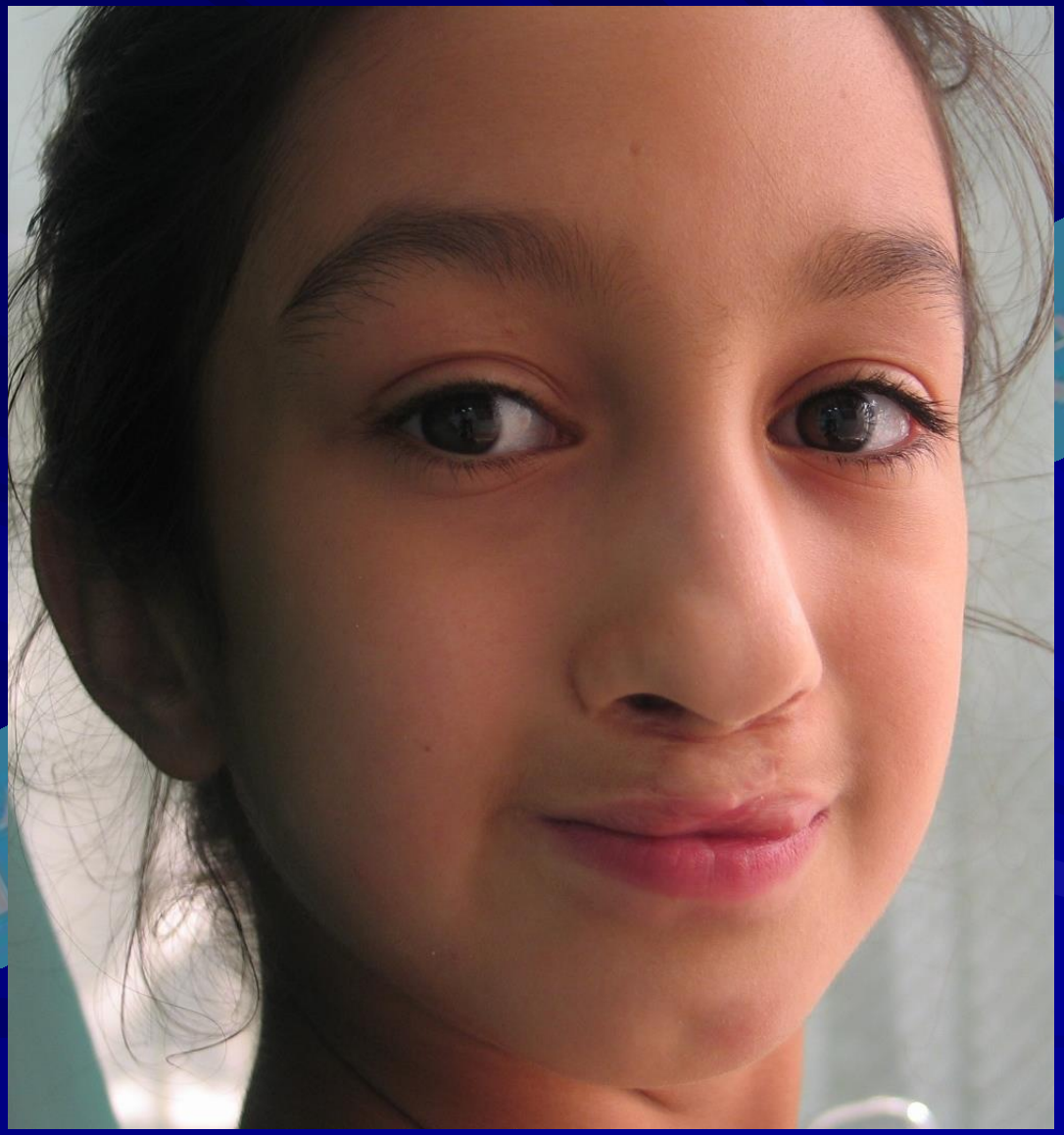
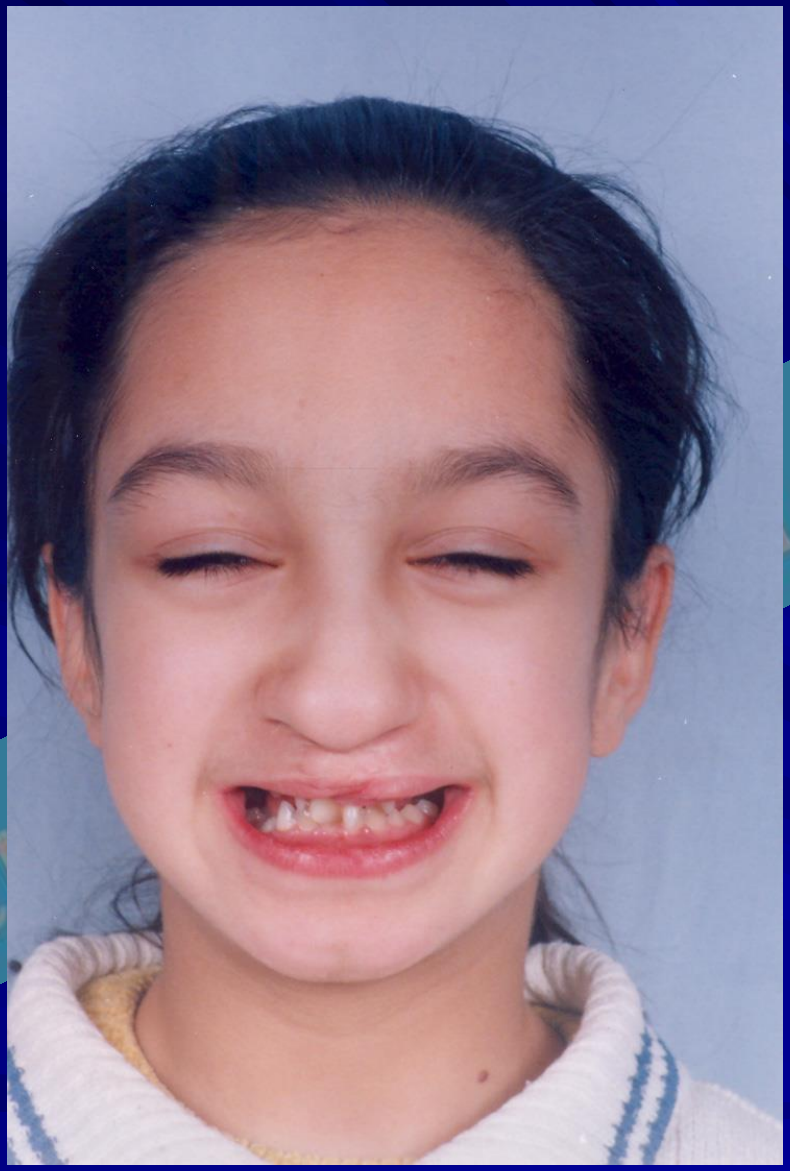
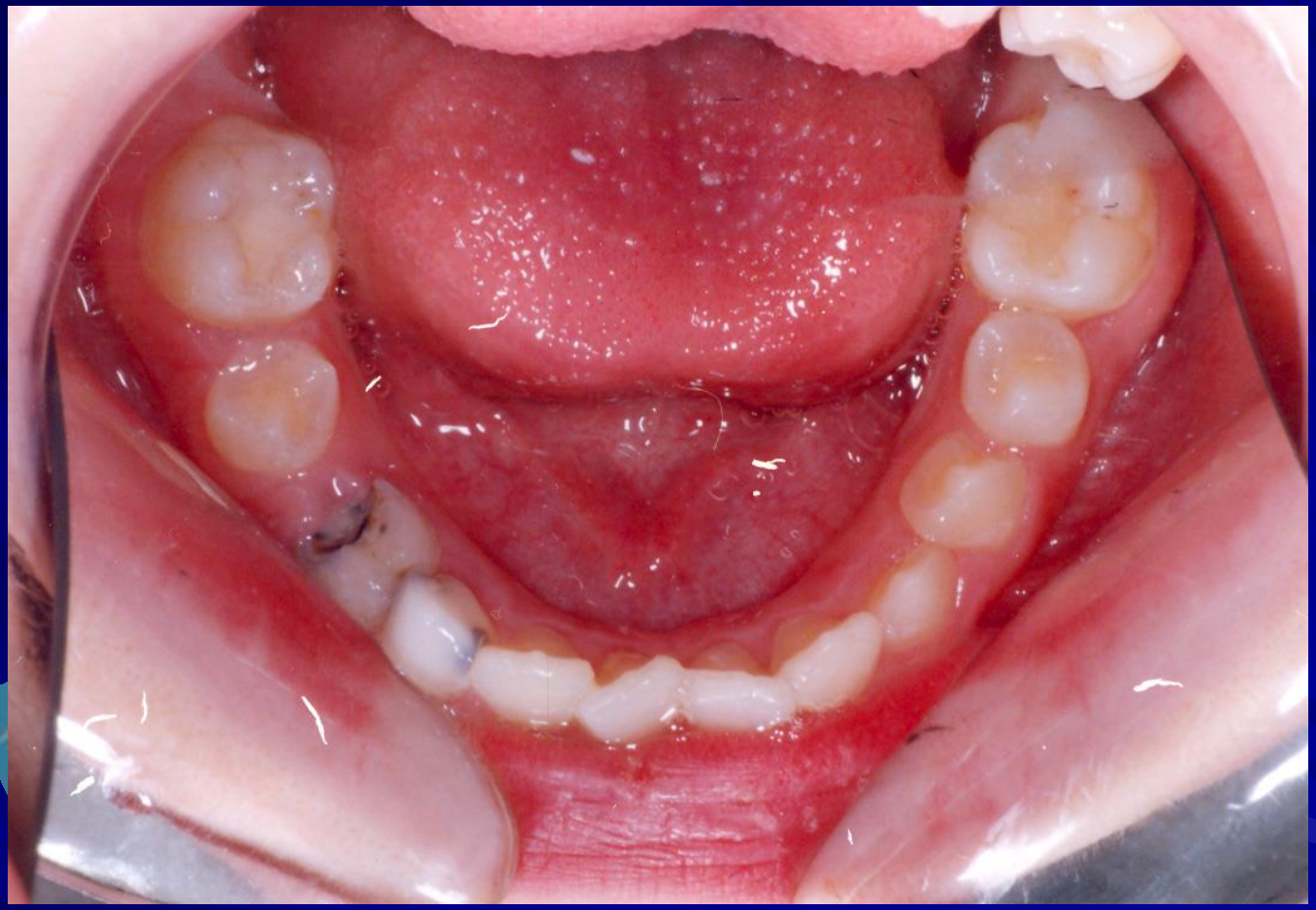
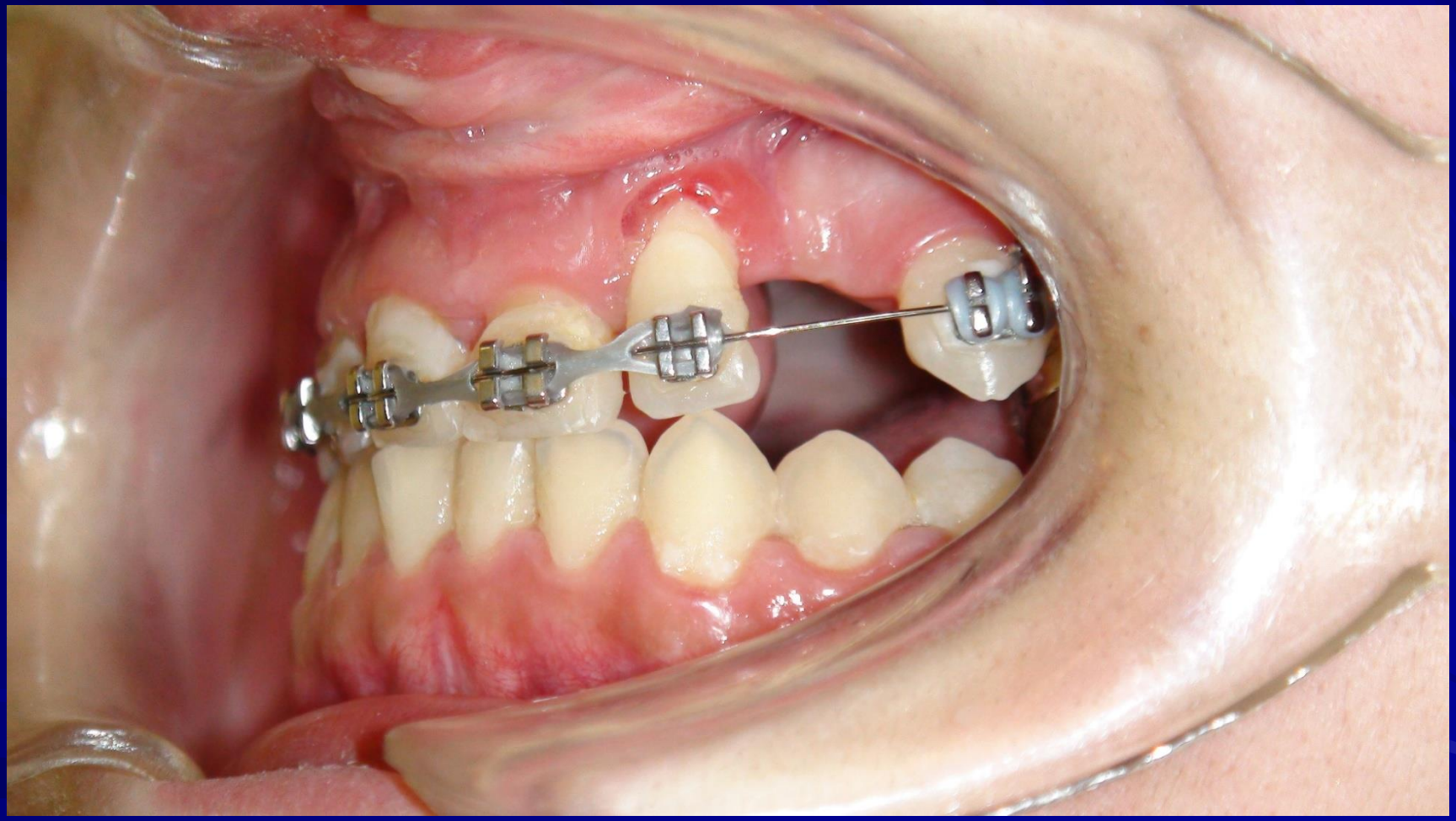


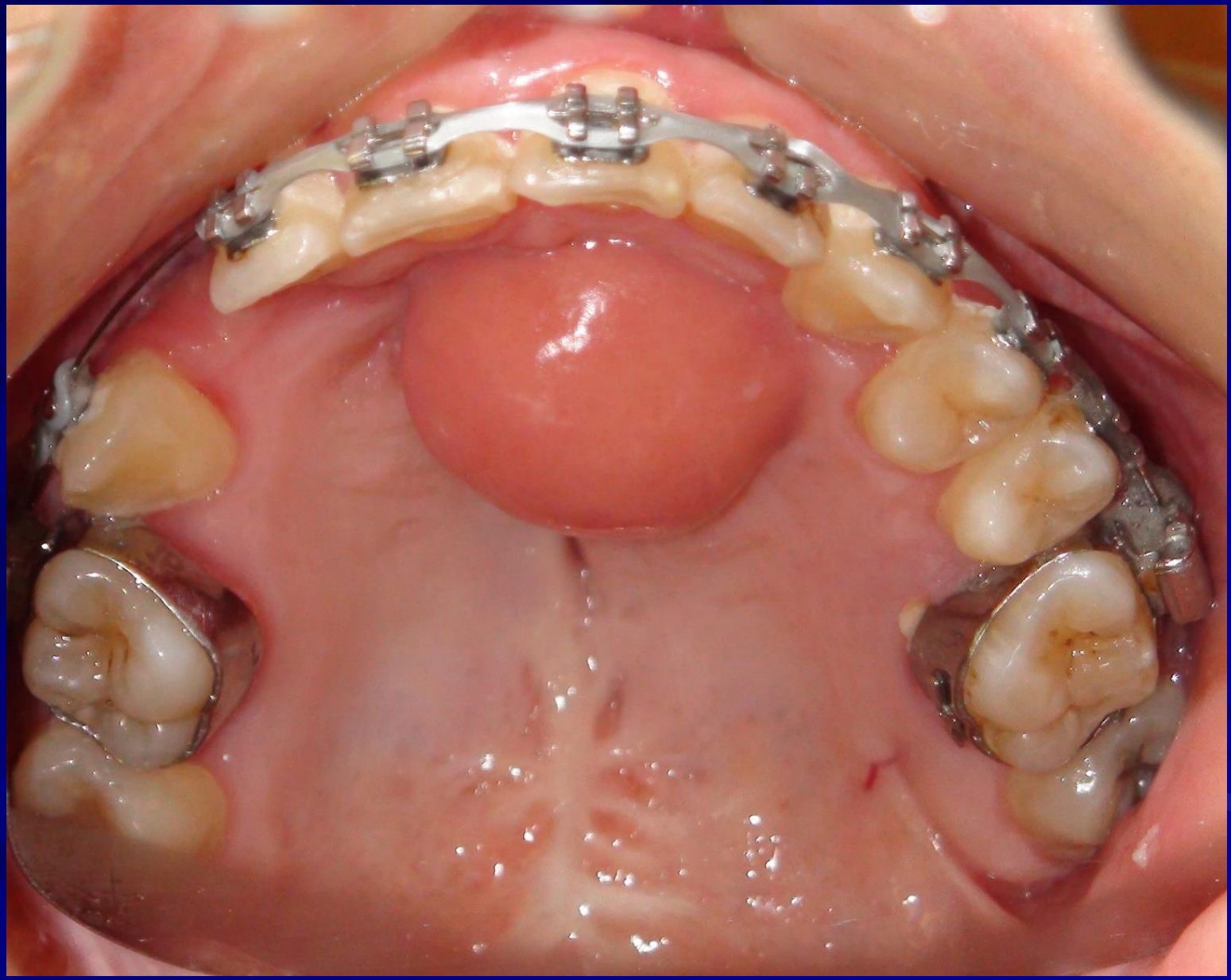
FIG. 19-33

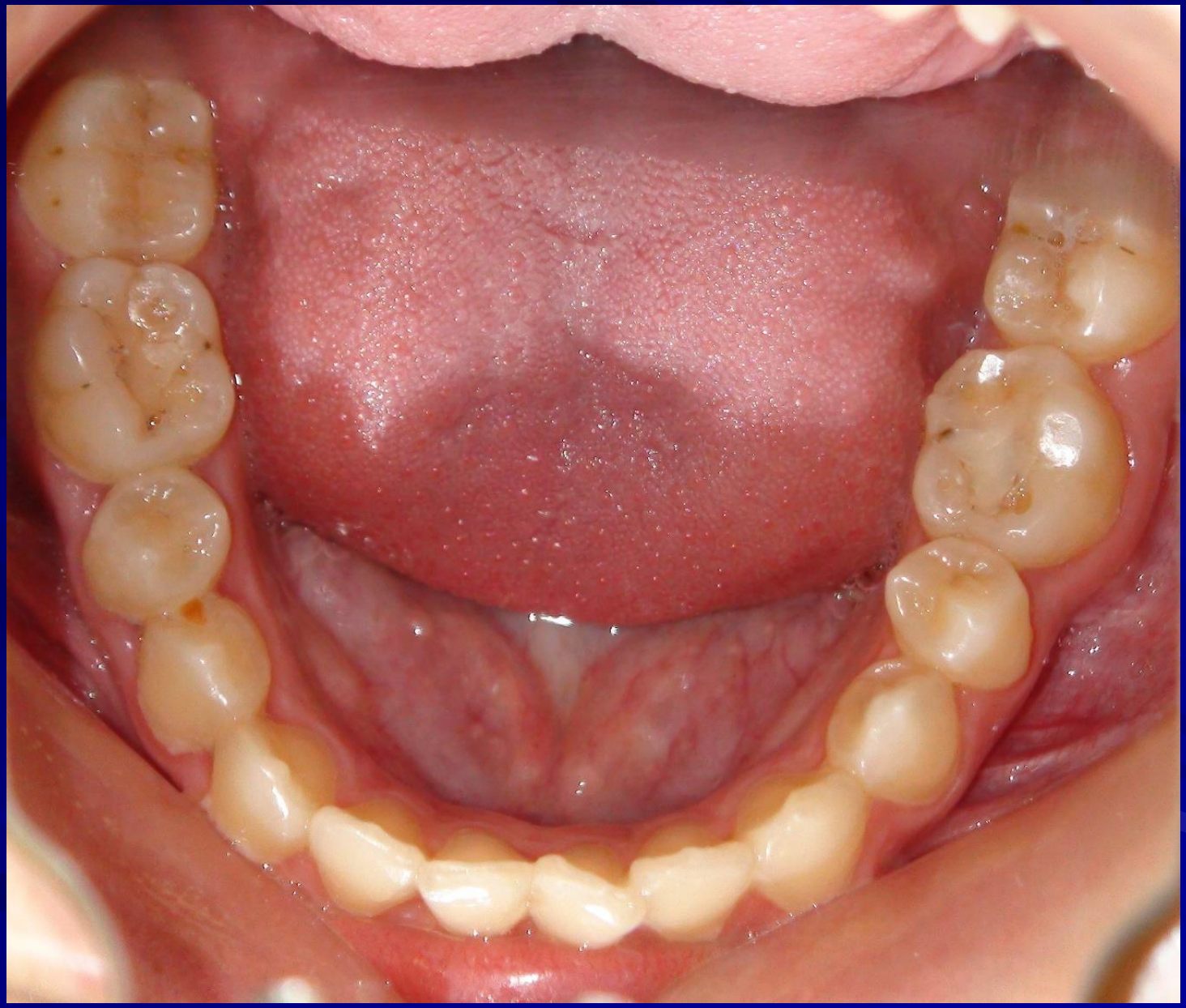












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