DENTO-MAXILLARY RADIOLOGY AND IMAGING

Course 3

RADIOIMAGING DIAGNOSTIC OF MAXILLO-FACIAL MALFORMATIONS

3.1. MALFORMATIVE PATHOLOGY - TERMINOLOGY

Maxillo-facial malformations are those morphological abnormalities caused by an embryonic development alteration.

From the definition results the character always congenital of the malformations so, the statement congenital malformationis is redundant.

Malformations have a less knowledgeable ethiopathogeny being taken into account:

- **chromosomal abnormalities,**
- **embryopathy,**
- **heredopathy.**

3.2. ONTOGENESIS

Maxillo-facial structures begin to develop from the fourth's week of gestation when the branchial arches develop.

These are embryonic structures which lead to the development of gills in fish.

In humans according to the principle that ontogenesis follows phylogenesis some of the branchial arches will undergo deep changes in order to adapt to a new function and others will disappear.

THE FIRST BRANCHIAL ARCH will lead to the development of:

- **mandible,**
- **maxillary,**
- **palatine bone,**
- **zygomatic bone,**
- **and from the corresponding mesoderm will develop:**
- **mastication muscles.**

THE SECOND AND THE THIRD BRANCHIAL ARCHES lead to the development of - hyoid bone.

From the second branchial arch corresponding mesoderm will develop the mimic muscles and from the third branchial arch corresponding mesoderm will develop superior and middle constrictors muscles of the pharynx and palatoglos muscle.

THE FOURTH AND THE FIFTH BRANCHIAL ARCHES

The structures of the fourth and the fifth branchial arches lead to the development of inferior constrictor muscle of the pharynx and the larynx muscles.

From the corresponding mesoderm will develop the cervical visceras together with thyroid gland, parathyroid glands and the root of the tongue and also the heart, pericardium and the thymus gland which all will leave the cervical region and permanently descend in the thorax.

3.3. MALFORMATIVE PATHOLOGY ETHIOPATHOGENY

The most congenital abnormalities appear during the profound changing processes of branchial arches system into human structures of the face and neck.

CHROMOSOMAL ABERRATIONS

They are genetic diseases appeared as the result of genetic material alteration by mutations which affect the number or the structure of the

cromosoms, autosomals or sexuals, with different phenotypic expressions evident at birth or respectively, secondary syndromes detected after puberty.

EMBRYOPATHIES

They are determinated by exogenous factors which act on a normal primary embryo through toxic noxious.

The malformative result has a great range of variability depending of the precise acting moment of the teratogenic factor during embryogenesis.

Ethyopathogenic, there are recognized many exogenous teratogenic factors:

- **drugs,**
- **ionizing radiations,**
- **consumption of alcohol,**
- **infections,**
- **nutritional and vitamins deficiencies,**
- **morpho-physiological and endocrino-metabolical maternal endogenous factors.**

HEREDOPATHIES

Are hereditary disorders with genetic transmition through autosomal or sexual chromosomes.

Genetic characters are transmitted dominant or recessive with homo or heterozygote phenotypic expressions.

DOMINANT TRANSMITION

Make possible the transmition of a genetic character no matter if the character is present on one or on both chromosomes, namely both in state of homo and in heterozygote state.

So, the dominant character is manifested even when it is present on only one of the two chromosomes.

RECESSIVE TRANSMITION

Recessive character is manifested only if it is present on both of the pair chromosomes, namely in homozygote state (or if it is present on X male chromosome).

So, the recessive character is manifested only in homozygote state, in heterozygote state is not manifested, being masked by the dominant correspondent character.

3.4. MALFORMATIVE PATHOLOGY DETECTION

The phenotypic expressions of different maxillo-facial malformations are manifested as alterations of

- **skeletal structures or**
- **correspondent soft tissues,**

due to the genetic transmition or caused by teratogen factors action.

Any deviation from the normality may be early detected even *in utero* by an ultrasound exam which is part from the follow-up screening of the pregnant woman or by

- **X-ray exam,**
- **CT - scan or**
- **MRI,**

after birth or in puberty, depending on the manifestation period of different malformations.

3.5. CHROMOSOMAL ABERRATIONS

There were described with maxillo - facial involvement the following malformative entities:

- **DOWN SYNDROME,**
- **MARFAN'S SYNDROME,**
- **VAN DER WOUDE SYNDROME,**
- **PROGERIA,**
- **FRAGILE X SYNDROME.**

DOWN SYNDROME (TRISOMY 21)

It's characterized by the presence of an extra 21 chromosome, the incidence being associated with increasing maternal age.

Parents of any age who have had a child with trisomy 21 have a significant risk - about 1% - of having a similarly affected child, equivalent to that affecting births to a mother over 45 years of age.

Even after birth have some characteristics as a mongoloid type facies with: brachycephaly, oblique eyes with epicanthic folds, mandibular prognathism,

small nose and ear pavilions, hypotelorism, absence of the frontal and sphenoid sinuses with hypoplastic maxillary sinuses.

Oral cavity has: ogive palate vault, fissured macroglossia, narrow nasopharynx with hypertrophied tonsils and adenoids.

The dentition exhibits characteristic abnormalities as: hypodontia, enamel hypocalciphication, radicular and crowns malformation with occlusal disharmonies as mesioclusion, posterior crossbites, apertognathia and severe crowding of the anterior teeth.

They also have mental retardation, cardiac malformations, cleft lip and palate with detection after 35 years of age of some dementia type of neuropathologic disorders analogous to those found in Alzheimer disease.

Also an increased risk for Down syndrome has been found in families with a predilection for Alzheimer disease.

MARFAN'S SYNDROME

On chromosome 15 is located Marfan gene (a mutation gene) which produce a change in one of the proteins that produce strength to a component of connective tissue, probably collagen.

It's associated with increasing paternal age being notable due to a number of catastrophic death that have occurred in affected undiagnosed athletes.

Characterized by a plurimalformation syndrome clearly clinic and by imaging detected: long legs and arms, chest deformities, prolapsed mitral valve, aortic dissection and aneurysms, myopia, retinal detachment. Facial massive appears long and narrow, with ogive palate vault.

VAN DER WOUDE SYNDROME

Genetic disorder very closely correlated with the genes that determin the sanguine groups.

It's characterized by the presence of cleft superior lip and palate associated with cystic masses and fistulas of inferior lip.

Even in minor parental forms the descendents have a manifested labio-maxillo-palatine cleft.

Minor forms of the syndrome appear with lateral incisors and superior premolars anodontia, bifid uvula and palatoschizis.

PROGERIA

Chromosomal aberrations which determine pituitary gland dysfunction with a precocious aging, the patients becoming rapidly old, even from childhood.

It's characterized by senile nanism, genital glands insufficiency and precocious aging (hair loosing, weight loosing, skin atrophy and pigmentation, arterial atheroma) with the exit at 16 years of age.

The skull appears with increased volume, small mandible and dentition eruption disorders with dentine development alterations.

FRAGILE X SYNDROME

The most frequently cause of mental retardation with sexual hereditary transmission being affected the long arm of the X chromosome with a fragile site.

Mental retardation is more frequent and severe in males, in females appearing rarely and as only a slightly mental retardation.

Classic clinical triad includes: mental retardation, post pubescent macro-orhidism and characteristic facies (high forehead, big mandible, long ears, ogive palate vault and palatin cleft).

3.6. PLURIMALFORMATIVE SYNDROMES

There are heredopathies with sporadic chromosome aberrations, the phenotypic expressions being usually characterized by multiple abnormalities coexistence which affect many organs and systems.

These multiple abnormalities heredopathies may be classified in General and Regional.

3.6.1. GENERAL HEREDOPATHIES

The heredopathy totally affects a system or a defined specialized tissue. There are described:

- **OSTEOPETROSIS,**
- **OSTEOGENESIS IMPERFECTA,**
- **EHLERS-DANLOS SYNDROME (CUTIS HIPERELASTICA),**
- **CRANIOSTENOSIS,**
- **PHAKOMATOSES.**

OSTEOPETROSIS

It's a recessive transmitted heredopathy characterized by generalized symmetric increase in skeletal density due to abnormalities of bone resorbtion remodeling.

Appear a characteristic generalized dense sclerosis as the marble of the skeleton, being also named "marble bones disease" or Albers-Schönberg disease after German radiologist doctor that first described it.

Radiographic characteristic finding is the skeletal dense aspect as "bone within bone".

The heredopathy is generally divided into two main types:

- the infantil (malignant, congenital),

- the adult (benign, tarda).

also existing a mild intermediate form which appears to be more common.

THE INFANTILE TYPE (malignant, congenital)

It's the severe form characterized by:

- skeletal abnormalities (increase in skeletal density with decrease of resistance and multiple fractures due to secondary fragility),

- neurologic abnormalities (due to thickening of the skull foramina with secondary compression on optic and facial nerve and reactive blindness and deafness),

- hematologic abnormalities (the decrease in the marrow compartment cause: anemia, thrombocytopenia and pancytopenia; extramedullary compensatory hematopoiesis determine secondary hepatosplenomegaly).

THE ADULT TYPE (benign, tarda)

This form has a favorable prognosis, only the skeletal abnormalities being present.

Dento - maxillary findings include: delayed eruption, congenitally absent teeth, unerupted and malformed teeth, enamel hypoplasia and marked mandibular prognathism.

The elevated dental caries index secondary to enamel hypoplasia determine regional infectious spread with the appearing possibility of osteomyelitis, one of the serious complication due to inadequate host response because of the diminished vascular component of osteopetrotic bone.

OSTEOGENESIS IMPERFECTA

It's a periosteal hereditary osteodysplasia due to a heritable defects of connective tissue characterized by an insufficient development of cortical bone which is very thin and also of the cancellous bone which is lax or absent resulting a generalized osteoporosis with multiple spontaneous fractures and secondary skeletal deformities.

It is also named "glass bone disease", due to the fact that the bones are hypostosic, thin and fragile, appearing radiographic as tubular glass structures.

The heredopathy classic clinic triad include: multiple fractures of the long bones, blue sclerae şi otosclerosis with hearing loss, association known as van der Hoeve syndrome or triad.

Dental abnormalities as dentinogenesis imperfect are present in all four types of disease, the temporal dentition being most affected with deformities of the crown (shortened and bell-shaped) and of the root (narrow and short) and discoloration: blue, brown and amber opalescent.

EHLERS-DANLOS SYNDROME (CUTIS HIPERELASTICA)

It's a hereditary mesenchymal disorder of connective tissue, anatomic characterized by a collagen fibers alteration resulting eight variants of expression.

The heredopathy is characterized by joint hypermobility and skin hyperextensibility, severe cardio-vascular disorders as aneurismal aortic dissection, gastrointestinal and urologic with ruptures of bowel and urinary bladder.

At the thoracic level may appear spontaneous pneumothorax and respiratory impairment secondary to chest wall deformities.

Oro-facial features include a narrow maxilla with fragility of gingival and mucosal tissue and also a temporo-mandibular joint dysfunction due to laxity resulting hypermobility and dislocation.

The tongue has a marked extensibility enabling contact with the tip of the nose, as has been described.

Dental abnormalities findings include deep anatomic grooves and excessive cuspal height of the molars and premolars, dental denticles and enamel hypoplasia.

CRANIOSTENOSIS

Cranial malformation group determine by premature closure of sutures characterized by cranio-facial dysmorphism and increased intracranial pressure.

May appear isolated or may be part of multiple malformations depending upon the affected sutures. There are described the following forms:

- **Trigonocephaly - premature fusion of metopic suture, trigon/triangular shaped forehead,**
- **Brahycephaly - premature fusion of coronal suture - short/flat head syndrome,**

- **Dolichocephaly - premature fusion of sagittal suture - long narrow head syndrome**
- **Scaphocephaly - a form of dolichocephaly (like an inverted boat)**
- **Oxycephaly or turricephaly, acrocephaly - fusion of lambdoid and coronal sutures bilaterally, the most severe, tower/high - head syndrome,**
- **Plagiocephaly or unilateral flat head syndrome - anterior: premature unilateral fusion of coronal suture or posterior: premature unilateral fusion of lambdoid suture.**

CRANIOSTENOSIS with FACIAL MASSIVE ABNORMALITIES

From this group there were described:

- **ACROCEPHALOSYNDACTYLY - APERT'S SYNDROME,**
- **CRANIO - ORBITO - FACIAL DYSOSTOSIS - CROUZON SYNDROME,**
- **MICRORINIA - BINDER SYNDROME.**

ACROCEPHALOSYNDACTYLY - APERT'S SYNDROME

It's a cranio-facial malformation determined by the insufficient ventrally growth of the skull characterized by a complex manifestations inherited in an autosomal dominant mode.

Cranio-cerebral disorders: craniostenosis with tilted the anterior region of the skull base and increasing the cranio-basal angle, bulging of the occipital bone with parietal elongation, corpus callosum agenesia, encephalocel.

Facial disorders: exophthalmos, hypertelorism, maxillary hypoplasia with secondary retrognathism and mandibular pseudoprognathism, open occlusion, "saddle nose".

Syndactyly also appearing: absence of the interdigital spaces from hands and feet.

It may be also associated with labio-maxilo-palatin clefts and vertebro-medullary malformations as spina bifida and cifoscoliosis.

CRANIO - ORBITO - FACIAL DYSOSTOSIS - CROUZON SYNDROME

It's a cranio - orbito - facial dysostosis that is inherited in an autosomal dominant mode, like Apert syndrome, without extremities malformations, but with orbits abnormalities, characterized by:

- **acrocephaly caused by premature fusion of the cranial sutures;**

- **maxillary hypoplasia, mandibular prognathism and open occlusion;**
- **“parrot beak nose“;**
- **cataract, exophthalmos and divergent strabismus.**

MICRORINIA - BINDER SYNDROME

It’s a cranio - facial malformation characterized by a stop in maxillary development with craniostenosis and acrocephaly also associated with:

- **abnormally thin nose with narrow narines;**
- **maxillary retrognathism with mandibular pseudoprognathism and open occlusion.**

PHAKOMATOSES

These heredopathies characterize a group of congenital diseases which associate:

- neuraxis different malformations

- small benign ectodermal tumors: vascular or fibromas - generic named “facoma“ due to lens shape - localized on: skin, mucosal level, eyes and central nervous system.

PHAKOMATOSES WITH FACIAL STRUCTURES INVOLVEMENT

There were described:

- **TUBEROUS SCLEROSIS BOURNEVIL,**
- **ENCEPHALO - TRIGEMINAL ANGIOMATOSIS – STURGE - WEBER SYNDROME,**
- **CEREBELLO - RETINAL HEMANGIOBLASTOMATOSIS - VON HIPPEL-LINDAU SYNDROME,**
- **CUTANEOUS NEUROFIBROMATOSIS VON RECKLINGHAUSEN**

TUBEROUS SCLEROSIS BOURNEVIL

It’s a complex congenital disorder that is inherited in an autosomal dominant mode characterized by multiple hamartomas localized in different organs with tuberosity aspect.

Clinic it is characterized by Bourneviel triad which include: skin lesions, epilepsy and mental retardation.

Skin lesions appear as facial milimetric angiofibromas colored rose - yellow - brown as a firm papulas, periunghial fibromas, café - au - lait spots and hypopigmentated maculas.

Cerebral lesions are the most characteristic as solid lesions with tuberosity aspect situated periventricular and different neuronal migration abnormalities, all generating: epilepsy, autism and mental retardation.

Also appear enamel defects, dental or gingival fibromas.

ENCEPHALO - TRIGEMINAL ANGIOMATOSIS - STURGE - WEBER SYNDROME

It's a polimalformative association characterized by classic clinic tetrad which includes:

- **cutaneo - mucosal signs,**
- **eye disorders,**
- **neuro - psychiatric disorders,**
- **cerebral calcification detected at X-ray exam.**

Typical appear: cutaneous angiomas (flat in sensitive territory of trigemenal nerve), congenital glaucoma and calcified vascular malformations often parieto-occipital which generate epilepsy crises.

CEREBELLO - RETINAL HEMANGIOBLASTOMATOSIS - VON HIPPEL-LINDAU SYNDROME

It's characterized by the retinal, cerebella and visceral presence of hemangioblastomas producing different clinical manifestations with ocular, cerebella and visceral disorders.

The presence of oral hemangioblastomas may be confused with submucosal abscesses and any incision attempt can be lethal.

CUTANEOUS NEUROFIBROMATOSIS VON RECKLINGHAUSEN

Type I is characterized by the presence of cutaneous pigmented spots together with cutaneous and perineural neurofibromas.

Neurofibromas, by intra osseous development or indirectly by initiation of vasomotor disorders determine cyst masses to appear on their development route or hyperplasia and osteosclerotic processes.

The most often are the lesions on the trigeminal nerve paths with secondary multiple mandibular cyst masses apparition.

3.6.2. REGIONAL HEREDOPATHIES

These are heredopathies which predominantly affect dento-maxillary region with bilaterally symmetric or unilateral asymmetric developments.

There were described:

- **CHERUBISM,**
- **CLEIDOCRANIAL DYSPLASIA,**
- **ECTODERMAL DYSPLASIA,**
- **GARDNER SYNDROME,**
- **GORLIN-GOLTZ SYNDROME,**
- **PROGRESSIVE HEMIFACIAL ATROPHY - ROMBERG SYNDROME,**
- **HEMIFACIAL HYPERTROPHY.**

CHERUBISM

It's a benign maxillary and mandible malformation characterized by a bilaterally regional thickening which give the face a typical aspect of "cherubin" = chubby.

Radiographic appear the bilaterally increased mandible volume especially at the angles level with multiple polycystic - like geodes which thinning and "blow" the regional cortical bones.

CLEIDOCRANIAL DYSPLASIA

Inherited in an autosomal dominant mode malformation is characterized by the association of:

- clavicles hypoplasia
- brahicephaly,
- facial masive hypoplasia,
- supernumerary teeth,
- prognathism,
- spine, pelvis and long bones abnormalities.

ECTODERMAL DYSPLASIA

Gruoping multiple malformative syndromes characterized by ectodermal development abnormalities is inherited in a recesive or dominant autosomal modde with various cutaneous and dento-maxillary disorders.

Cutaneous disorders are caused by sebaceous or sweat glands deficiency with secondary dryness and dermatitis at the level of irritation - prone regions.

Dento - maxillary disorders also appears as: adontia, dysplasia, hypodontia, dysgnathism, maxillo-palatine clefts.

GARDNER SYNDROME

It is an autosomal dominant mode inherited malformation characterized by the simultaneous presence of ecto-, endo- and mezodermal abnormalities with secondary disorders of:

- **cutaneous tissues (fibromas, sebaceous and dermoid cysts);**
- **skeletal (multiple osteomas or osteofibromas at the level of: maxilla, ethmoid and occiput bones);**
- **digestive tract (intestinal polyposis with malignization tendency).**

GORLIN-GOLTZ SYNDROME

It's characterized by the presence of:

- **multiple basal cell epitheliomas,**
- **maxillary kerato-cysts**
- **ribs abnormalities**

The most of the cysts are localized at the level of mandible angles and the rest appear in lateral maxillary regions being associated with roots resorbtion of the implied teeth.

PROGRESSIVE HEMIFACIAL ATROPHY
- ROMBERG SYNDROME

It is characterized by the presence of ecto - and mezodermale development abnormalities localized at a hemiface level with secondary atrophy of:

- **bones (malar, maxillar, mandible);**
- **subcutaneous cell tissue;**
- **skin (pigmentated and adhesive cutaneous atrophy);**
- **homolateral half of tongue.**

HEMIFACIAL HYPERTROPHY

It is a congenital hemihypertrophy which asymmetrical affects segmental or regional half or even all body by unilateral increase of cells number.

Usually appears as an unilateral hemifacial hypertrophy with excessive development of skeletal tissues, subcutaneous cell tissue and regional tongue half together with various dental abnormalities as macrodontia with malocclusion and root, crown or morphology abnormalities.

3.7. BRANHIAL ARCH SYNDROMES

Multiple congenital abnormalities grouped in different syndromes appearing during the profound changing processes of branhial arches system into human structures of the face and neck.

There are described:

- first branhial arch syndromes

- second branhial arch syndromes

3.7.1. FIRST BRANHIAL ARCH SYNDROMES

There were described:

- **FRANCESCHETTI SYNDROME (TREACHER-COLLINS SYNDROME, MANDIBULOFACIAL DYSOSTOSIS);**
- **PIERRE ROBIN SYNDROME;**
- **GOLDENHAR SYNDROME (OCULO-AURICULAR DYSPLASIA);**
- **MANDIBLE CLEFTS.**

FRANCESCHETTI SYNDROME
(TREACHER-COLLINS SYNDROME, MANDIBULOFACIAL DYSOSTOSIS)

`Imply bilaterally first branhial arch dynamics modifications with secondary maxillary and mandible development stop.

Results a clinic characteristic aspect:

- facial aspect modification with "fish mouth" or "bird neck" due to maxillary, mandible and malar bones hypoplasia;

- ear pavilion and eyelid slot malformation;

- reduction volume of maxillary sinuses;

- ogive vault with mandible hypoplasia, palato-maxillary clefts and dental dysplasia with malocclusion.

PIERRE ROBIN SYNDROME

Characterized by a clinic triad which include: micrognathism, glosoptosis and palatoschisis.

Due to mandible retrognathism and palatoschisis tongue remains placed between maxillary processes keeping a vertical position without horizontal development and anterior growing - it may get fixed in cleft at birth with secondary asphyxia accidents and alimentation disruptions with severe malnutrition.

GOLDENHAR SYNDROME (OCULO-AURICULAR DYSPLASIA)

Imply unilateral disorders resulted from development abnormalities of first and second branhial aches.

Characteristic clinic manifestation include unilateral facial hypoplasia due to:

- auricular pavilion hypoplasia ,

- mandible hypoplasia ,

- temporo-mandibular joint hypoplasia

- dental abnormalities as anodontia, supernumerary teeth.

MANDIBLE CLEFTS

Caused by fusion abnormalities of the two mandible rams resulted from the first branhial arch malformation.

They may be associated with absence of: mandible segments, cervical soft tissue or dental segments.

Due to the absence of anterior fixation of the tongue it may glide posterior and lead to secondary asphyxia accidents.

3.7.2. SECOND BRANHIAL ARCH SYNDROMES

There were described:

- **LATERAL FISTULAS OF THE NECK;**
- **LATERO - CERVICAL CYSTS;**
- **TYROGLOSSAL TRACT FISTULAS AND CYSTS.**

LATERAL FISTULAS OF THE NECK

Appear secondary to the absence of second branhial arch closure with resulting fistulas paths:

- blind with a single extern or intern orifice,

- true with a skin extern orifice along the anterior margin of the sternocleidomastoid muscle and a mucosal intern orifice in tonsil fossa and palatogros arch.

Fistulography - radiological exam which helps totally detect the fistulas trajectory in order to complete resection therapy.

LATERO - CERVICAL CYSTS

Restant rudiments of the second branhial slot with mucous fluid accumulation surrounded by a peripheral epithelial shell mostly localized close to mandible angle or in soft parts of the neck.

May be detected by:

- ultrasound - exam as a transonic structure with posterior strengthening;

- CT - scan as sharp contoured fluid densities without intravenously injected contrast substance uptake;

- MRI - as fluid structures in hyperT2, hypoT1, without intravenous injected contrast substance uptake.

THYROGLOSSAL TRACT FISTULAS AND CYSTS

Appear by totally or rudimentary remanence of glandular tissue in the caudally thyroid gland migration trajectory from foramen cecum (base of tongue region) till in pretraheal final region.

May result cysts or fistulas tongue related or at the neck level, hyoid bone related.

All have the same imaging characteristics but with mediosagital specific localization.

3.8. LABIO – MAXILLO - PALATIN CLEFTS

They are malformations of the face characterized by facial continuity interruption by sulcus which separate independent facial structures in embryonic life and do not follow the normal fusion final process.

After Valerian Popescu these are grouped in the following clinical forms:

- **PARTIAL CLEFTS,**
- **TOTAL CLEFTS,**
- **ASSOCIATED CLEFTS.**

ANTERIOR PARTIAL CLEFTS

UNILATERAL

- **incomplete - imply partial or totally lip and narinar threshold;**
- **complete - imply lip, narinar threshold and alveolar crest.**

BILATERALLY

- **incomplete - imply partial lip structures and eventually alveolar crest;**
- **complete - imply bilaterally lip, narinar threshold and alveolar crest.**

POSTERIOR PARTIAL CLEFTS

- **incomplete - imply partial on the middle line lueta, soft palat and a small portion of hard palat;**
- **complete - imply lueta, soft palat and all hard palate till primary palate.**

TOTAL CLEFTS

- **UNILATERAL - CLEFT LIP "rabbit lip"- imply lateral superior lip, narinar threshold, alveolar crest and on the middle line lueta, soft palat and hard palate.**
- **BILATERALLY - CLEFT PALATE "wolf mouth"- imply totally superior lip, nasal floor, alveolar crest bilaterally and on the middle line lueta, soft palat and hard palate.**

ASSOCIATED CLEFTS

- **an uni or bilaterally anterior cleft coexist with a posterior cleft;**
- **separated by a single intermediary intact osseous segment: alveolar crest secondary palate…**

3.9. MONOMALFORMATIVE SYNDROMES

They are characterized by apparition of isolated malformation in only one precise structure or organ without affecting other structures or organs.

There were described:

- **BELLY OF DIGASTRIC MUSCLE ABNORMALITIES**
- **TORUS**
- **PAROTID CCESSORY GLANDS**
- **HEMANGIOMAS**
- **LYMPHANGIOMAS**
- **DERMOID CYSTS**
- **LINGUAL THYROID GLAND**

BELLY OF DIGASTRIC MUSCLE ABNORMALITIES

Appearing as an unilateral hypoplasia or even aplasia or as an excessive development of anterior accessory digastric uni - or bilaterally muscular fascicle, these last entities must be differentiated from tumors or lymphadenopathies.

TORUS

Mandibulars are internal bilaterally developments on the lingual faces containing normal bone structures.

Palatins are palatinal mediosagitally nodular or extended appearing as regional radioopacity at X- ray exam.

PAROTID ACCESSORY GLANDS

May be unilateral or bilaterally present having a proper excretory tracts system which separately drains in Stenon's tract.

Have the same pathology being the site of morbid entities with high range of malignity: more than 50% from all the tumoral types.

HEMANGIOMAS

Are malformations of vascular endothelium, being present as:

- capillary - capillary hemangioma or

- cavernous - cavernous hemangioma.

With facial localization are detected in regional soft tissue and also in maxillary bones and must be differentiated from other cyst - like structures as cysts and abscesses because their incision may have fatal consequences.

They are easily detected at ultrasound exam, cervical CT - scan or MRI exam native and after intravenous contrast substance injection.

LYMPHANGIOMAS

Appear due to persistent embryonic lymph sacs as tree - like cystic masses with partial communications localized mediosagitally in submental region or laterally in latero -cervical region.

They are easily detected in superficial planes by ultrasound exam, cervical CT- scan or MRI exam.

DERMOID CYSTS

They may contain squamous epithelium, hair follicles and sebaceous gland being localized latero-cervical or medio-sagittal.

Easily imaging detected due to adipose tissue contain with specific negative CT- scan densities and T1, T2 MRI hypersignal.

LINGUAL THYROID GLAND

It’s ectopic development of thyroid tissue caused by stopping from the normal descent process in the pretracheal region.

Appear mostly at the dorsal median lingual level, the whole tongue being rarely affected.

Imaging diagnostic assessed by characteristic hyperdensity on CT- scan images or specific MRI hypersignal, with intense intravenous contrast substance uptake in both imaging methods.