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PEDIATRIC TUMORS AND CONGENITAL ANOMALIES IN ORAL & MAXILLO-FACIAL SURGERY

PRACTICAL COURSE

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CONTENT

PREFACE

THE IMPORTANCE OF THIS COURSE FOR STUDENTS AND RESIDENTS

We dedicate this book for medical and dentist students, undergraduate dental students and postdoctoral pediatric dental students provide efficient and superior comprehensive care to infants, children, teenagers, and medically compromised patients. It also provides experienced dentists with reference information regarding new developments and techniquesmay be involved in the primary assessment of congenital malformations and tumors patients.The book is written within the project: State Program „Modern personalized surgery in the diagnosis and complex treatment of tumors in children" with the number: 20.80009.8007.06, who's leader was the late honorary member of the SAM (State Academic of Moldova), university professor, illustrious surgeon Gh. TIBIRNA. Medical and dentist studentsacquire a keen eye for details through continued training and practice and due to their knowledge of facial anatomy, they are ideally suited to the diagnosis and treatment of facial malformations. Although dentists may not be involved in all aspects of craniofacial soft tissue treatment, they form an important part of the management team. Dental professionals may be one of the key health care providers who assess the patient postsurgical and provide some aspect of orofacial reconstruction. To be an effective member of the management team, dentists require a thorough knowledge of the diagnosis and treatment of soft tissue craniofacial malformations and tumors. There has been a gradual rise in the incidence of congenital malformations and tumors patients in children, probably due to increased risk-taking and aggressive behavior in children, who are more commonly left to their own devices without close supervision.

1. Congenital disorders of head and neck development in children population

1.1 Disorder of the embryonic development of the face and jaws. The etiology. Pathogenesis.

Congenital malformations refers to morphological and functional disorders condition of the body. Malformation lead to varying degrees of clinical manifestations (cosmetical and functional) and can appear immediately after birth or may have late manifestations. Orofacial clefting is one of the most frequent congenital anomalies. Birth prevalenceof cleft palate and cleft lip with or without cleft palate is approximately one in 700 with characteristic regional variations. Congenital malformations of the head and neck regions have an increased incidence due to local embryological complexity and have a multifactorial etiology.

Etiology. The origin of congenital malformations in most cases are unknown but is considering to be complex and heterogenous with increasing evidence that these anomalies are multifactorial in nature. Most congenital malformations are causes linked to environment, genetic and gene-environment interaction.

The genetic component in the etiology of congenital malformations is considered key to the appearance of malformations (25-30 %) and are determinant. It has been found that responsible for these malformations are not a single gene, but a number of genes. Genetic or inherited causes include: chromosomal defects, single gene defects, dominant inheritance, recessive inheritance. Scientific progress in field of molecular biology and genetics have discovered 70 phenotypes of genes responsible for the abnormal development of the craniofacial region. The large spectrum of anomalies is due interection of teratogenic agents that arise from environmental and genetical conditions, depending on the time of action, intensity and duration.

Four categories of genes suggestive of a genetic susceptibility to orofacial cleftings are: 1) genes expressed in a particular area of the embryo or in a particular period of the palatine arch development, such as the transforming growth factors alpha and beta (TGF alpha, TGF beta 2,

TGF beta 3); 2) genes having biological activities linked to the orofacial cleftings pathogenesis without direct involvement (e.g. the retinoic acid receptor (RAR), the methylenetetrahydrofolate reductase receptor (MTHFR) and the folic acid receptor (FOLR1); 3) genes or locus identified in experimental animals as the homeotic genes MSX-1 and MSX-2; 4) genes involved in the interaction with the xenobiotics metabolism as those in P-450 cytochrome system.

It is estimated that approximately 10–15 % of congenital structural anomalies are the result of the adverse effect of environmental factors on prenatal development. This means that approximately 1 in 250 newborn infants have structural defects caused by an environmental exposure and, presumably, a larger number of children have growth retardation or functional abnormalities resulting from nongenetic causes, in other words, from the effects of teratogens. An agent that can produce an abnormality in structure or function, restriction of growth, or death of the embryo or fetus is known as a teratogen. Teratogenic factors can become any of environmental factors that act directly or indirectly on the embryological schedule, interupting embryological development throuout the pregnancy. Environmental teraatogenic factors comprise exogenous agents (chemical, physical, medications, drugs, biological agents) and endogenous agents or maternal conditions or diseases.

Exogenous teratogenic factors include:

- physical agents (radiation, hyperthemia, hypothermia);
- biological agents and infections (cytomegalovirus, toxoplasmosis, syphilis, influienze virus, mumps virus, varicella etc);
- drugs, toxic metal and chemical agents (polychlorinated and polybrominated biphenyls, toluene, lead, mercury);

Endogenous teratogenic factors or maternal conditions include:

- hormonal disorders (diabetes mellitus. phenylketonuria, obesity, hyperparathyroidism orhypothyroidism),
- nutritional deficiencies (folic acid deficiency and folic acid antagonists. hypervitaminosis A),
- ethanol, smoking and various drugs (marijuana**.** sedatives, metronidazole)**.**

Ionizing radiation can injure the developing embryo due to cell death or chromosome injury. The sources of contamination of the human body with ionizing radiation are classified into two types: natural radiation - contamination with radioactive elements from soil and water; artificial radiation - professional systematic irradiation, medical irradiation. The malformations induced are similar to those noted following exposure to ethanol, retinoic acid or hyperthermia. They vary from sublethal injuries affecting differentiation and cellular interactions, to effects on rates of proliferation and cell death of the cells response to the radiation is depended on cell cycle

*Hyperthermia*is defined as a body temperature of at least 38.9 °C and is an antimitotic teratogen after exposure between weeks 4 and 14. Teratogenic effects depend on the duration of temperature elevation and the extend of elevation. Elevations of 1.5-2.5 degrees of Celsius above normal body temperatures represent the threshold for teratogenesis in human. Mothers who had been exposed during pregnancy to hyperthermia caused by infections or by sauna bathing. or with excessive exercise, febrile episodes at the critical embryonic stage may produce cell death that plays a major role, with the mitotic cells being the most susceptible. It has been suggested though that hyperthermia could result in intra and extracellular leakage of lysosomal enzymes which could lead in type-2 cell death. Infants exposed to maternal hyperthermia are associated with severe mental deficiency, seizures in infancy, microphthalmia, midface hypoplasia, and mild distal limb abnormalities

Hypothermia is defined as a core body temperature of less than 35 °C. Cardiopulmonary bypass in a pregnant patient is associated with a fetal mortality rate of 16 % to 33 %, multiple congenital defects, severe disruptive defects of the brain and distal spinal cord

Infectious has a greate teratogenic impact on the embryo or fetus. The lethal or developmental effects of infectious agents are the result of mitotic inhibition, direct cytotoxic effects, or a vascular disruptive event. However, a repair process may result in scarring or calcification, which causes further damage by interfering with histogenesis. Infections that result in congenital malformations of fetal or neonatal include enteroviruses

(coxsackievirus, poliovirus and echovirus), varicella (or chickenpox), mumps viruses amd other virusis. Microbial infections during pregnancy or fever more than 40 °C during 1-8 weeks of gestation has high-risk of craniofacial cleft. Syphilis and Toxoplasmosis have not resulted in congenital anomalies but have caused significant fetal pathology.

Drugs play an important role in etiology of craniofacial malformations. Teratogenic effects of substancesinclud the type of the drug administered, kind, dose and duration of the medication, impaired metabolism of drugs owing to diseases of the mother, placenta transfer, time of the exposition during pregnancy, genetic disposition. Following drugs are among these pharmacons: Benzodiazepines, Neuroleptics, tricyclic antidepressants, Lithium, Beta-receptor-blocking agents, Beta-sympathomimetics using as tocolytics, Captopril A deleterious effect on the fetus in the first trimester results in abortions and malformations, in the second and third trimester in general retardations of the fetal development, postnatal impairments of the functional development, increasing perinatal mortality, premature deliveries, stillbirths and transplacental carcinogesis. A long-term exposition or the administration of high doses of different substances at the end of pregnancy may produce syndromes which may possibly endanger the newborn.

Lead, mercury and lithium are toxic metal. A woman who has had lead poisoning can pass lead on to her fetus, even if she no longer is exposed to. The adverse effects of lead include spontaneous abortion, stillbirth, increase in minor malformations, including hemangiomas, lymphangiomas, hydroceles, skin tags, skin papillae, and undescended testes. Organic forms of mercury are more toxic than the inorganic forms. Methylmercury, the most toxic organic form, causes severe brain damage, lithium defects in infants exposed to lithium in utero include malformations of the CNS, ear, and ureter, altered thyroid and cardiac function, and congenital goite.

Chemical exposure. It is quite important to notice the prevalence of malformations and its relation with geographic pozition, climate, and socio-epidemiological factors. Greater probabilities of developing malformations were observed on geographic location with risk areas. It`s

means environmental contamination (air, biota, oil, surface water, water springs, and specific chemical contaminants), with a special focus on chemical exposure such as Polychlorinated and polybrominated biphenyls. They have been used for more than 40 years as insulating fluids, heat exchangers, plasticizers, and chemical additives, and they are known to be worldwide pollutants. Women suffering from polychlorinated and polybrominated biphenyls poisoning have infants with parchment-like skin with desquamation and brown discoloration („cola baby"), dark colored nails, conjunctivitis, low birthweight, exophthalmos, and natal teeth. Reduced birthweight and small head size with hypotonicity and hyporeflexia are associated with higher levels of exposure.

The associations may have been influenced by other, uncontrolled chemical sources like occupation, industrial emissions, pesticides, and water contamination. Craniofacial malformations can be caused by the anatomical obstruction (toungue obstruction) during embryological development that breakes fiziological fusion between embryological layers and lead to disorders in the development of the fetus. Early amniotic rupture can severely disrupt the intrauterine development of the fetus leading to different fetal anomalies.

Occupational factors related with job have increase risk craniofacial malformation. Maternal occupations, such as hairdressing, agriculture, dry-cleaning activities and leather or shoe manufacturing as well as exposure to pesticides, and aliphatic acids can make a grate impact in development of the fetus. Such associations were not altered when stratified by maternal age, area of residence, and socioeconomic status.

Maternal characteristics. Anatomical and physiological dizorders, such insuficence of blood supply (hypoxia), or changes in uterine pressure. Hypoxia, mother's anemia, placenta pathologies, repeated vomiting during the first period of pregnancy may hurt the developing fetus.

Obesity.During pregnancy, obesity is associated with adverse outcomes that include macrosomia, hypertension, pre-eclampsia, gestational diabetes mellitus and fetal death. and increased risk of varying birth defects.

Hypoxia producing in the mother body during pregnancy cancause

disorders in the activity of some ferment. It can lead toaresting the synthesis of amino acids, proteins, which cause changes in the structure of the embryological tissures, size reduction and abnormal apposition of the facial prominences.

Hypoxia may result from cigarette smoking, reduced atmospheric oxygen levels and also placental insufficiency.

Healthy and balanced diet have an essential importance for providing the developing fetus with nutriens for proper growth and development. Impact of these agents, as teratogenic factors, is not fully known. It has been proven that some vitamins have teratogenic impact (Vitamins A, B).

Folic acid is responsible for different malformations. Deficiency of folic acid during pregnancy interferes with the embryonic development through a common mechanism that depending on the maternal or embryo genotype. Teratogenic effect on embryonic development of several drugs is known - pseudoephedrine, aspirin, ibuprofen, amphetamine, cocaine, as well as antiepileptic drugs (phenobarbitone, phenytoin and carbamazepine) ethanol. Smoking, radiation, infections, metabolic imbalance, are at higher risk of developing craniofacial cleft.

Metabolic disorders have significant teratogenic potential. A number of hormones and metabolic degradation products can cross the placenta. Diabetes is a disease with an increased risk for disorders of embryological development during pregnancy. Mother suffering from diabetes mellitus or phenylketonuria, has high-risk of craniofacial cleft. Thyroxine deficiency is another experimentally proven factor that causes craniofacial clefts. The action of the hormonal disharmony of the mother in the development of the embryo is not studied in detail, but it is known that the number of mothers with endocrine disorders that give birth to children with malformations is 6 times higher. These abnormalities in mitochondrial size, location, cristae and number suggest a form of metabolic defect that could underline malformations. The suggested explanation is that a defect in energy production could result on in insufficient cellular migration and proliferation and thus be the pathophysiologic basis for craniofacial cleft.

Cretinism and iodine deficiency. In early pregnancy, iodine deficiency induces a critical decrease of T_4 levels with consequent TSH increase responsible for hypothyroidism in about 50 % of iodine-deficient pregnant women. Congenital hypothryoidism associated with deafness and mental retardation, developmental changes in the brain and cerebellum is found in the offspring of hypothyroid mothers. Deafness persists in spite of thyroid replacement therapy. have been described

Ethanol, smoking and drugs. Babies born to alcoholic mothers develop Fetal Alcohol Syndrome that have three characteristics: prenatal and postnatal growth retardation, facial anomalies, and CNS dysfunction. Nicotine is a vasoconstrictor that results in uterine vascular constriction and intrauterine growth retardation through decreased perfusion of fetal tissues. It is a cholinergic agonist and a constituent of tobacco. Cigarette smoking during pregnancy raises the risk of perinatal mortality and morbidity. The active ingredient of marijuana is 8,9-tetrahydrocannabinol, which is fat soluble, crosses the placenta easily, and may persist in the fetus for as long as 30 days. Marijuana use during pregnancy, especially in the first trimester. result in growth retardation, malformations and increased risk of nonlymphoblastic leukemia.

1.2. Embryology of the head and face

Due to the difficulties of studying the embryonic development, a unitary interpretation of the pathogenesis of congenital malformations has not been reached.

Three stages of human ontogenesis development are recognized:

I. The organogenesis stage.
II. Morphogenesis stage, which takes place in two stages (prenatal and postnatal development).
III. Modeling stage.

The organogenesis is a phase of embryonic development that starts at the end of gastrulation and continues until birth. During organogenesis cells of each of the three gems layers undego differentiation by cell signaling cascades that allows cells rearrangement, and ensures that organs forms at specific sites within the organism. Organogenesis for most organ systemss lasts from 0 to 4 months of gestation. There is multiplication of cells that

determine the size and weight of the embryo. Cell multiplication accomplishes through an induction process.

The morphogenesis stage refers to organized spatial distribution of cells during the embryonic development into tissues, organs, organ systems, and organism as a hole and represents a progress towards maturation. Preparing the cells to perform certain functions morphofunctional growth and perfection of organs and sistems (histodifferentiation), known as development. The morphogenesis stage runs from 4 to 9 months (fetus) and continues throughout life, through childhood and adolescence into adulthood. The birth is one of the events produced during the morphogenesis period.

Tissue remodelingstage is the last ontogenetic stage of human development, and refers to reorganization or renovation of existing tissue without essentially altering their size, coordination of growth and differentiation which occurs throughout life, being characterized by restructuring, repairing and remodeling the organ systems.

Embryology of the head and face.

Week 3. Prenatal period of development include three stages: germinal (extending from the time of fertilization to the end of the second week of gestation), embryonic (from the beginning of the third week to the end of the eighth week) and fetal (from the ninth week until birth). The structural development of the head and face occur between the third and eight week of gestation (embryonic period).

The embryonic stage begins at three week of gestation, as two-dimentional planar structure. At this stage central nervous system is set in the area of ectoderm thickening known as neural plate. Neural tube appears as a result of invagination of the ectoderm centreally and simultaneous elevation of ectodermic tissue along side the groove to form the neural folds. These folds fuse with each other in the midline to form the neural tube. The beginning of the fusion stat at the junction of the future brain and spinal cord. As the folds fuse there is a separation of neural tube from the overlying ectoderm and give rise to the formation of neuroectodermal crest cells. They are believed to migrate widely throughout the developing embryo in a relatively cell-free enricherd

extracellular matrix and differentiate into a wide array of cell and tissue types. The majority of connective skeletal tissues of the cranium and face are ultimately derived from derivatives of neural crest cells that are located between the optic vesicle and the overlying ectoderm. The first morphologic evidence of the optic primordia is seen as a thickened area with a shallow sulcus on the lateral forebrain of the neural tube and subsequently optic vesicle that arise from the evagination of surrounding walls. During the third week the major divisions of the brain can be distinguished (prosencephalon, mesencephalon and rhombencephalon). The first stages of recognizable face start at the end of the third week, by futher growth of the cephalic portion of the neral tube that gives rise to the frontonasal process and the branchial arches.

Week 4. The face begins to form during the four weeks. The face is development from five primordial that surround a central area of depression, the oral pit or stomadeum. These primortdia are single cranially located frontonasal process, and two bilaterally located maxillary and mandibular processes. The last tow arise from the the first branchial arch. These processes grow differentially and by obliterating the ectodermal plates or grooves between them give rise to the features of the face. At this time the primitive trisegmented brain (prosencephalon or forebrain) divides into telencephalon (endobrain) with prominent lateral domes (cerebral hemispheres)and the diencephalon which gives rise to the optic vesicles. The mesencephalon part is undevided. The rombocephalon into metencephalon (cerebellum and pons) and myelencephalon (medulla).

At this stage the ectoderm overlying the optic vesicle of the developing eye thichens to form the lens placode. In the cause of a week the optic vesicle deepens to form a optic cup The frontonasal process intervens between the two lateral optic cups.

Week 5. Nasal placodes develop at the inferolateral corners of the frontonasal process. As the region surrounding the the nasal placodes the deepening nasal placodes are transformed into nasal pits, forming in time the anterior nares. In continuity with a developing oral cavity nasal placodes become olfactory grooves and maxillary process continuie to enlage encroaching on the stamadeum to form a primitive oral cavity. As

the optic stalk narrows further to define the primitive optic nerve, retinal pigment appears in the external layers of the optic cup and the inner layer begins to differentiate into the neural elements of the retina. The primitive vitreus body develops in the interventing space as the lens placode invaginates toward the optic cup to form a lense vesicle deteching itself from the overlying ectoderm.

As the features of the face form, thr development of the embryonic skull begins. The mesemchyme, surrounding the developing primitive brain condenses to form a mesenchyme capsule, the desmocranium. At about the fifth week the base thickens forming the cartilaginous cranial base through neuroectoderma; condensation of the mesenchyme.

Week 6. The medial nasal processes approach each other to form a single globular process that in time will give rise to the nasal tip, columella, prolabium, frenulum and primary palate. As this occur the frontonasal process collapses inward to form the nasal septum. Continued growth of the maxillary mass located below the optic vesicles allows fusion with lateral nasal process. During the fusion, nasolacrimal duct start to form. Toward the end of week 6, the maxillary process fuses with the medial nasal fold the globular process, forming a true nostril as it gives rise to the lateral lip element. Posterior to this anterior fusion of the maxillary process to the nasal processes, the developing nasal floor is open to the oral cavity. Within the primitive stomadeum, lateral palatine processes developed from the medial edges from the maxillary process to give rise to the secondary palate. At this time the developing tonger nearly completely fills the oronasal cavity and reaches the nasal septum.

At this stage the eye continues its development. Neurocrest mezoectodermal cells at the lateral aspect of the lens differentiate into the early cornea and sclera. The cavity of optic stalk is obliterated as nerve fibers from the retina grow back to reach the brain to form the optic nerv. The overlying surface ectoderm develops subtle horizontal folds with mezechymal core above and below the developing cornea that represent the beginning of the upper and lower eyelids. During the 6 week period, the external ear develops from the six mesenchymal swelling (hillocks) that surround the first branchial cleft. The first three hillocks arise from the first branchial arch and

the second three from the second branchial arch. From the first branchial arch (three hillocks) become the tragus, the helix, and the cymba concha, the hillocks on the second arch become the antitragus, antihelix, and concha. The branchial cleft lengthens to form the primordium of the external auditory canal. As the face develops the auricle is gradually repositioned to a more lateral cephalic position from the original location low on the side of the neck.

Week 8. By the eight week the face completes closure of the lower facial grooves. The upper and lower jaw is formed by complete fusion. The upper and lower lips are better defined. The frontonasal process continues to collapse, forming a transverse furrow at the nasal bridge and the eyes begin to take a more medial position. The anterior rotation of the eyes continues throughout the remainder of the gestational period and well into childhood. The upper and lower lids continuie to develop and assume a more almond shape as they begin to fuse with each other. The lined ectoderm-derived epithelium between the fused eyelids and the cornea develops into the conjunctival sac, and the lacrimal glands develop from the superolateral angles.

The development of the of the skull continuies with ossification centers arising within the mesenchyme adjacent to the developing brain to form the bones of the calvarial vault the paired frontal and parietal bomnes. The primordial elements of the chondrocranium begin to undergo endochondral ossification to form bones of the basicranium – sphenoid, petrous part of the temporal and occipital bones. Because of the persistent growth of the sphenoethmoidal and spheno-occipital synchondroses and the septal cartilage cranial base grow forward and simultaneously orbital complex allow to inward rotation of the developing orbital complex.

In the face, small nasal bones appear above the nasal capsule.

Ossification center appear on the lateral surface of the nasal capsule, premaxillary and maxillary suface. Ossification center appear within the zigomatic and squamosal portion of the temporal region. The onset of bone formation marks the end the embryonic period and the developing fetus is much more resistant to teratogenic insult. Pathological changes during the skul and face formation may be associated with the absence

or supranumeric occurrence of some organs (anodonts, supranumerary teeth), large deviations from the conformation of the organs (congenital malformations), some incompatible with the viability of the fetus and child.

The morphogenesis of these deformations is not known, but there are two classic theories that explain the appearance of malformation of the head and neck. The theory of fusion by Dursy and the theory of mesodermal migration by Veau, Warbrick and Stark. Later Johnston and Vermeij-Keers theories proposed by them provide possible explanation of some of the rarer defects.

According to the Dursy and his theory free merging facial processes establish ectodermal contact than mesodermal penetration occurs to complete the union. Disruption of the fusion followed by mesodermal penetration leads to various type of clefts.

The supporters of mesodermal migration belive that the free ends of facial processes do not exit and the developing embryonic is composed of a continuous bilamellar ectodermal membrane with epithelial seems demarcating the various processes of the face. Within the bilamellar mesenchyme migrates and penetrates to smooth out the seams. Failure of mesenchymal migration cause a different type of clefts.

Johnston and Weston in their work identified a specialized group of cells, the neuroectodermal cells arising from the dorsal lateral ectoderm of the neural fold. The cells play a significant role in contributing the mesenchyme responsible for scheletal and connective tissue of the face. The cells from differing levels of the neural fold go on to populate specific regions (frontonasal prominence, primitive maxilla, developing mandibula) Malformations result when migration of the cells is incomplete or insufficient in number, or undergoes abnormal differentiation. Vermiej-Keers have shown that migration of the neural crest cells are part of an integrated coordinated set of tissue displacement. According to this theory facial clefting are related not necessarily to failure of migration but failure of local differentiation of the neuroectoderm and mesenchyme.

Vermiej-Keers identified true clefts and secondary clefts or dysplasia. True clefts are associated with the normal planes of embryologic process and are well defined. There are four primary sites in early embriologic

development that could lead to cleft formation (between the junction of the medial and lateral nasal processes and maxillary processes, between the lateral nasal and maxillary processes, between the maxillary and mandibular processes and between the palatine processes). The second type of cleft are happened in later embryonic stage of development depend not only on the well-defined set of membranous bones, but also on the number and location of the ossification center within each of the facial elements. The atypical clefts are believed to be accounted for by the variability in the number of ossification centers within each of the elements. The boundaries between the growths centers can be correlated with the potential sites of atypical facial clefts.

1.3. Developmental disorders

Developmental disorders, which occur in the first three months of intrauterine life, manifest through craniofacial anomalies, consequences of induction, hereditary and coalescence disorders.

Induction disorder refers to disorders or arrest cells proliferation of the facial buds (frontal, maxillary, mandibulary) in the early embryological development of the craniofacial region. These types of malformations are manifested by brain agenesis. partial or total lack of frontal, maxillary, mandibular bone formation and may cause lack of union between left and right orbits create only one in the midline of the face (cyclopia). Non-development of the upper or lower jaw (upper or lower agnathia); hypoplasia or complete lack of tongue.

Hereditary disorders is inherited genetically and passed on from one generation to another through defective genes. There are three types of genetic disorders: single gene or multiple disorders, chromosomal disorders and complex disorders. Most genetic disorders a rare in themselve. Genetic diseases ussualy affects less than 1in 2000 people are classified as rare malformations. Genetic abnormalities can be characterized by a statural hypotrophy associated with a higher microgradation (trisomy 21 or Down syndrome) or a lower microgradation (Edward syndrome, trisomy 17, 18), or with lack of union of palatinal shelves (Patau syndrome or trisomy 13, 15). Many other autosomal abnormalities, also characterized by statural hypotrophy, are associated with microcephaly and hyperthyroidism

(syndrome of chromosome deletion 5 and 21 „in ring l2"), which are sometimes associated with laryngeal development disorders, determining the characteristic „cat voice", the syndrome of „cry du chat".

Fusion disorders. The face forms by integrate difusion of the five facial primordia. Corect fusion of the facial prominences require either cell death or epithelial-mesenchymal transformation of the epithelial cells at the adjacent process. Craniofacial malformations (including facial cleft, craniosynostosis and facial dysostoses), occur because of abnormal development, differentiation and growth of facial prominences.

In the early embryonic development when the brain and facial form are intimately connected deficiency of anterior neural tube plate lead to a malformation named holoprosencephaly. It is refer to complete or incomplete failure of the anterior portion of the neural tube to form the cerebral hemispheres, resulting in a single forebrain. Such early failure before the third gestational week of embryogenesis are associated with monstrous facial dysmorphisms. If the prosencephalon and frontonasal process fail to develop cyclopia, erhmocephaly and cebocephaly can occur.

Median facial defects characterized by orbital hypertelorism arebelieved to occur in the late stage of embryonic development when ocular complex remains in the lateral position because development of the frontonasal process are arrested.

The incidence of congenital malformations of the face is approximately 1 in 700 births. This number includes major defects incompatible with life and minor defects that are surgically correctable. The most severe congenital problems are those that develop early in facial development (4th-8th weeks); relatively minor problems develop later (8th-12th weeks).

Classification of rare craniofacial clefts (median and oblique). The great variety of craniofacial malformations are very diverse, which is why attempts to classify rare malformations were very numerous. The malformations encountered on the midline of the face and the oblique ones were highlighted from all the craniocerebral and craniovisceral malformations.

Median malformations (holoprosencephaly) affect brain structures in 80 %. In relation to facial and brain malformations, they are classified into two groups: 1) with tissue deficiency and 2) with tissue excess. Median malformations with tissue deficiency fall into 5 categories:

Cyclopia (monophthalmia, sinophthalmia, anophthalmia, proboscis (single or double), arrhinia associated with anancephaly); Ethmocephaly (extreme hypothalorism, proboscis (single or double), arrhinia associated with anancephaly); Cebocephaly (hypoteleorbitism, proboscis nose (blind channel or single nostril) associated with anancephaly); Hypotheliorbitism (twisted nose, median cleft, usually associated with ancephaly). Hypotheliorbitism (twisted nose, bilateral cleft lip associated with semi-ancephaly or normal). Embryologically, there is an intimate correlation between the development of the structures of the medial face and the frontal brain. Thus, the degree of deformity of the face represents the severity of brain abnormalities. The first three categories are incompatible with life.

Dysmorphogenesis with excess tissue is also called the median cleft syndrome of the face. The specific features of this syndrome are: 1) orbital hypertelorism; 2) the hair line in the V-shaped frontal region; 3) bifid cranium; 4) the median cleft of the upper lip; 5) the median cleft of the maxilla; 6) the median palatine cleft; 7) primary telecantus. Mental retardation in these defects occurs when severe hipertelorism or extraencephalic deformity is present. (*Fig.1*, *Fig. 2*)

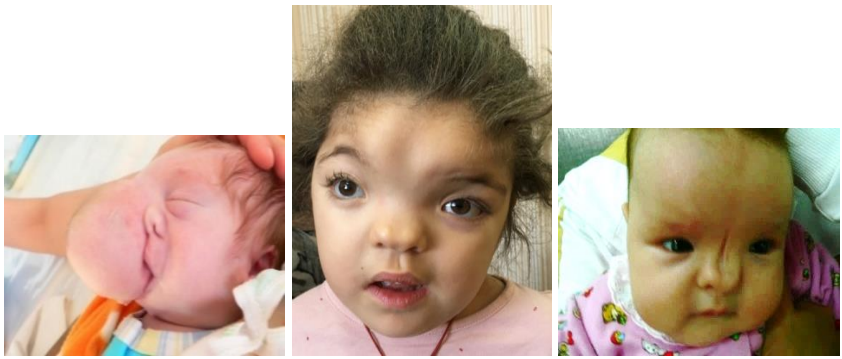


Fig. 1. **Confenital dysmorphogenesis of the craniofacial region (median cleft)**

Classification of oblique clefts. In 1962, the American Rehabilitation Association of Palate Clefts (AACPR) proposed the classification of oblique clefts into 4 groups: 1) mandibular; 2) nasal-ocular; 3) oro-ocular; 4) oro-auricular.

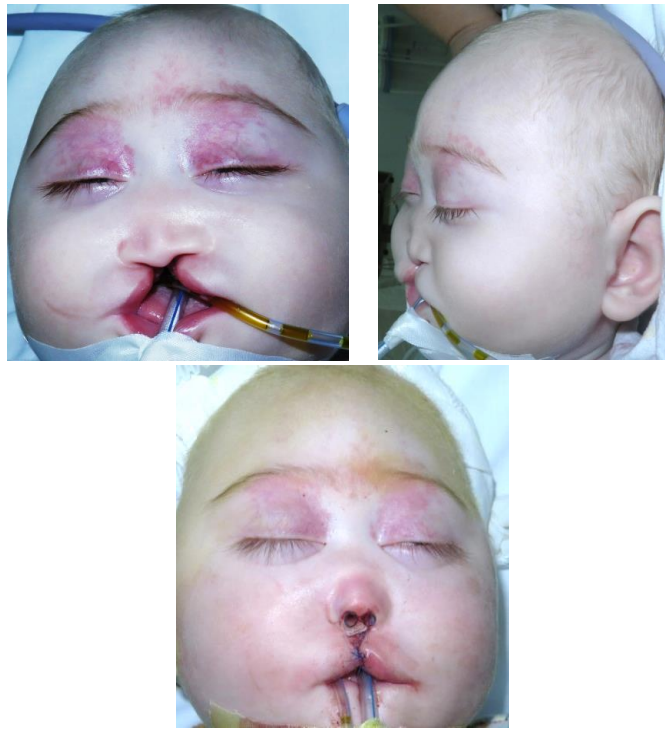


Fig.2. **Confenital dysmorphogenesis of the craniofacial region (median cleft) Cebocephaly.**

The oblique clefts also have another classification proposed by Tessier. In relation to Tessier's classification, 15 groups can be differentiated including the oral cavity, nose, orbit, and cranium. The numbering starts from 0 to 14, against the clock, around the orbit. Orbit is the reference point because it belongs to both the visceral and cerebral skull. In relation to the orbit the clefts are superior and inferior. Clefts 0, 1, 2 are located between the midline and the line projected from the medial edge to the upper lip, but do not pass through the orbit. Clefts 1, 2, 3 start from the Kupidon line, similar to lip clefts. Cleft 0 is characterized by a doubled frenum, diastema between the upper central incisors, the nose often bifid, wide columnar. The cleft 1 passes through the dome of the nose, and 2 reaches the base of the nostril. Cleft 1 and 2 reach the frontonasal line, but

it does not affect the orbit. Cleft 4 begins immediately lateral to the filtrum, and 5 medially to the commissure. Both 4 and 5 also involve the periform opening. Cleft 3 and 4 affect the orbit in the region of the medial canthus. In the third form, the nasolacrimal canal is affected, and the medial canthus is displaced inferiorly and with hypoplasia. Cleft 3 and 4 pass between the lateral and canine incisors, the jaw is with hypoplasia, manifesting tissue deficiency. The cleft 4 the defect passes into the lower eyelid forming colobomas. The cleft 3, 4, 5 associate with dystopia and micro-ophthalmia. Cleft 6 purely orbital, and 7 (*figure 3, 4, 5*) contains only the labial component. Cleft 8 is transverse, starting from the lateral cleft. Starting with cleft 9, they become cranial clefts. Clefts 8, 9, 10 disrupt the integrity of the upper eyelid and eyebrow. Cleft 10 and 11 (*figure 6*) may include orbital dystopia. Clefts 12, 13 and 14 are located medially in orbit and result in hyperthyroidism.

Coloboma is an oblique cleft, caused by the lack of fusion of the frontal and maxillary buds. The cleft extends from the upper lip to the lower eyelid, and the nasolacrimal canal appears as an excavation.

The transverse clefts of the face clinically manifest by macrostomy, due to the lack of fusion of the maxillary and mandibular proemenence.

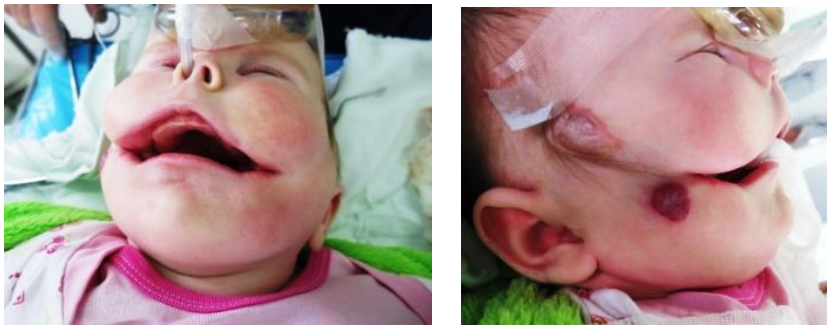


Fig. 3. **Congenital malformation of the face. Acording Tessier classification N 7 and N 8.**

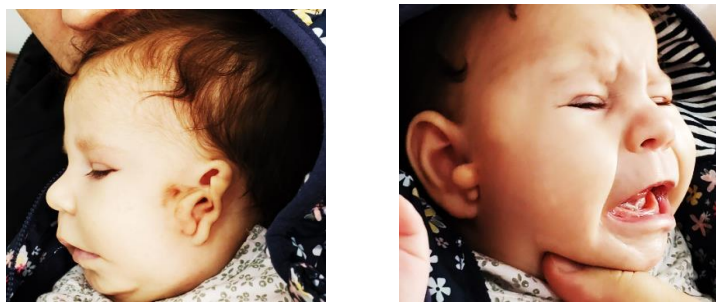


Fig. 4. **Congenital malformation of the face. Acording Tessier classification N 7 and N 8.**

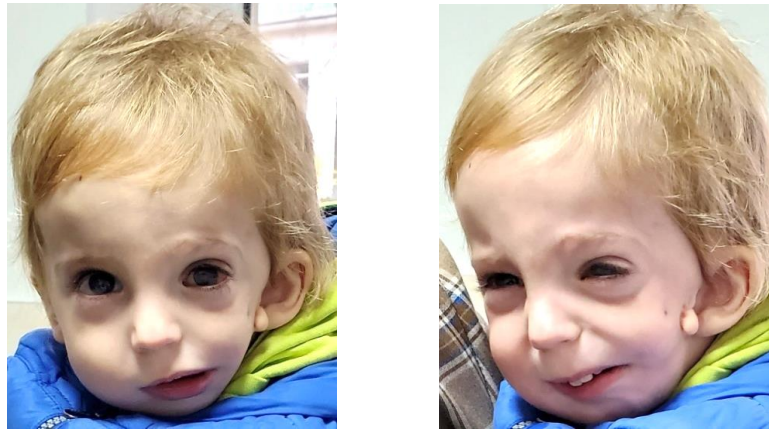


Fig. 5. **Congenital malformation of the face. Acording Tessier classification N 7 and N 8.**

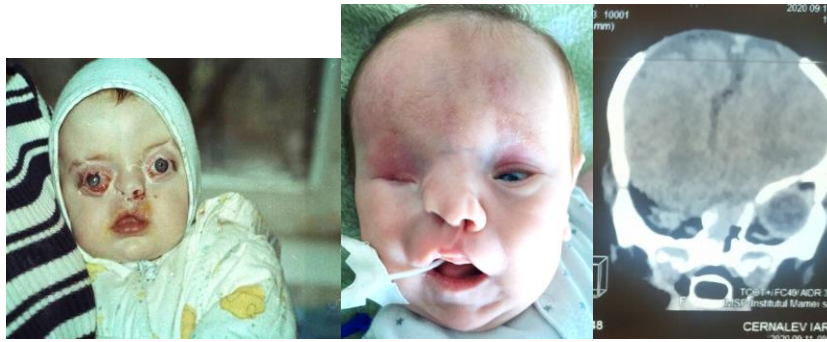


Fig. 6. **Bilateral oblique cleft. (Tessier 5, 6, 10, 11)**

The median clefts of the face are characterized by median sac bottoms on the lower lip, on the nose lobe (dog nose), due to the lack of coalescence of the right mandibular budswith the left mandibular buds, or the internal nasal buds (right and left). rhomboid glossitis or bifid tongue.

Fusion disorders also occur on the tongue, manifested by median The lack of union of the frontal embryological buds of the face with the lateral ones of the tongue determines the appearance of the reddish rhomboid formation, which extends from the blind orifice to the tip of the tongue (median rhomboid gloss).

Cervical cysts and fistulas, located along the anterior margin of the sternocleidomastoid, from the tonsil to the clavicle, are due to the abnormal melting of the ecto-endodermal membranes between the branchial arches.

Persistence and evolution of epithelial remnants. There are a number of conditions (cysts, fistulas, malignancies), which develop from unresorbed epithelial residues in the mesenchyme of the dento-maxillary apparatus, immediately after birth or long afterwards.

Median cervical cysts develop from the unresorbed embriomic glosso-thyroid chanal.

Nasal-palatal cysts are due to the persistence of the Rozemuller organ.

Epithelioid cysts often appear in the spongiosis of the jaws, through the secretory activity of the epithelium of the completely unobstructed dental blade.

Epithelial residues can regain their old powers of intense multiplication, producing adenolymphomas or epitheliomas of the jaws.

Treacher Collins syndrome (called Treacher Collins-Franceschetti syndrome, mandibulofacial dysostosis, Franceschetti-Kleine or Franceschetti-Zwahlen syndrome) comes from the ecto - and mesoderma of the 1st and 2nd branchial arches. It characterizes by bilateral maxilofacial bones hypoplasias - of the jaws, of the orbits with the deformation of the shape in the lower lateral part of the orbits, by deferent stage of the internal and external ear defects, hypoplasia of the zygomatic regions that cause depression of the skull bone located in the temporal area. Macrostomies may also occur, with palatal clefts (30 %), deferent type of

malocclusions. Pre-auricular chondromas. coloboma of the eyelids, abnormalities of development of the ear, micrognathia and preauricular skin tags and fistulas between the ear and mouth, palpebral antimongoloid fissure due to the downward movement of the lateral edge, as a result of a schisis of the lateral orbital margin, the presence of coloboma in the external third of the lower eyelid, in 50 % a deficiency of the eyelid, medial of coloboma, occasional microphthalmia. In more than half of the patients there are cutaneous appendages, blind fistulas or preauricular depressions located between the tragus and the buccal commissure or atrophic skin areas. The ears are deformed, presenting defects of the external auditory chanal: deformed external ears, microtia or flaccidity; in other cases, large, wrinkled and anteriorlylocated ears. In 30 % of cases, the external auditory meatus is stenotic or lacking (atresia of the canal), the conduction deafness or the hearing loss is present, sometimes associated with abnormalities of other organs and systems: cardiovascular, genitourinary.

Goldenhar Syndrome - a complex of craniofacial and neck defects. Clinical picture includes ocular, auricular, unilateral and possibly spinal abnormalities, in which no symptoms are compulsory. The main clinical signs and symptoms evident at birth are: a) facial asymmetry by more or less pronounced unilateral hypoplasia; b) epibulbar dermoids, cystodermoids or lipodermoids located at the tongue or on the corneal border of the inferior quadrant of the eyeball; c) single or multiple pre-auricular appendages located between the tragus and the buccal commissure, sometimes blind fistulas, dysplasia of the external ear; d) unilateral macrostomy;e) dysplasia of the axial skeleton (spine), especially in the cervical region, most often evidenced only radiologically (partial or complete cervical and thoracic vertebral fusions, cuneiform vertebrae, hemivertebrae, supernumerary vertebrae, spina bifida, anomalies of the ribs, possibly scoliosis). Complementary manifestations: dental abnormalities, transmission hypoacusis, occasional mental retardation, cardiac malformations, in approximately 50 % of cases, and pulmonary abnormalities.

Van der Woude syndrome is an autosomal dominant malformation.

Clinically, it has as depressions (pits) near the center of the lower lip, which may appear moist due to the presence of salivary and mucous glands in the pits. The mucus leaks from pitts can be „distressing" (painful). Half of these patients have lip and palate cleft. If a parent has such clinical manifestation (pitts), the child's risk of having depression raise to 50 %, and the cleft of the lip and / or palate – raise about 25 %.

Pierre Robin syndrome is a facial malformation, characterizes by abnormally small mandible hypoplasia (micrognathia), glossoptosis (downwardly displaced tounger) and wide U-shaped cleft palate (palatoschisis). Clinical manifestations are present immidiatly after the birth of the child and are expressed by micrognathia, which causes glosoptosis with narrowing of the upper airways and the medial palatoschisis. The newborn may have severe breaphing and feeding problems due to obstruction of the upper airway. There are also signs of chronic hypoxia, sometimes accompanied with mental retardation. Often it is noted stridor. Due to difficult breathing, the upper part of the chest may be bulged.

Crouzon syndrome or craniofacial dysostosis (figure 7) is a genetic disorders characterized by the premature fusion of skuul bones (craniosynosrosis). Clinical features result from premature fusion of the skull bones

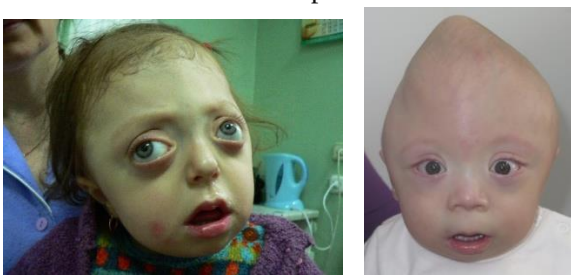


Fig. 7. **Crouzon syndrome or craniofacial dysostosis**

that lead to abnormal growth of facial area (fasciosthenosis). The main clinical signs are turiform shape of the skull with high forehead, bulging eyes and vision problems caused by shallow sockets, with strabismus (eyes that do not point in the same direction), palpebral fissures with slight obliquity (antimongoloid), difficulty or impossibility of convergent rotation of the eyeballs, convergent strabismus and ptosis. Facial deformities are specific

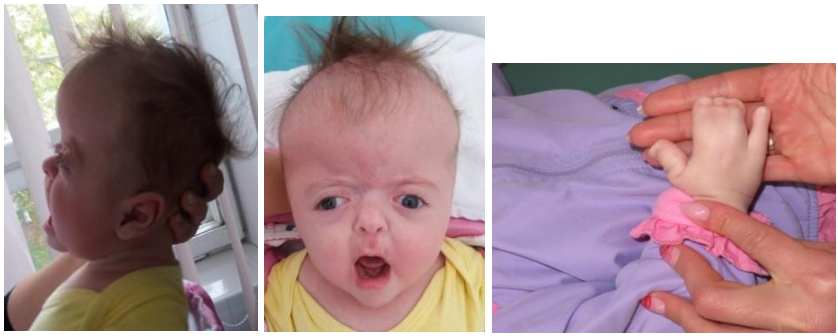


Fig. 8 **Apert syndrom. a. Lateral view. B. Anterior view. C. Hand disorders**

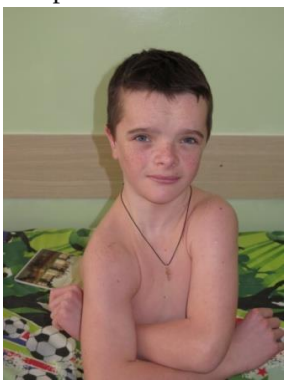
caused by undeveloped upper jaw (hypoplasia) braked nose (parrot's nose), narrow ear canal which is accompanied by hearing loss thin upper lip, a tall and narrow hard palate, malocclusion with crowded teeth.

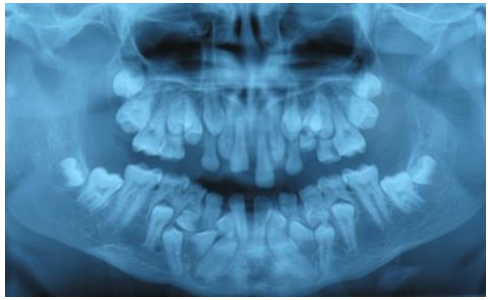
Complementary, it is noted mental retardation from mild to moderate, sometimes progressive visual involvement (up to optic nerve atrophy).

Apert syndrome (figure 8) is a relatively rare genetic disoeders with abnormal skull and facial growth. The shape of the skull is turibrahicephalic due to coronary suture premature fusion (craniosynostoses). A head is long with a high forehead, wide-set and bulging eyes with pooly closing eyelids, hyperthyroidism. Other clinical manifestations results from premature skull sutures fusion that cause false mandibular prognathism because of undeveloped apper jaw, palatal cleft, dentoalveolar disorders. Common syndrome symptoms are abnormal fusion of the bones of the hands and feet (syndactyly) with webbed or mitten-like hands or feet. Abnormal growrh of the skull lead to functional disturbance often with hearing loss, obstructive sleep apnea, repeated ear infection mental retardation.

*Cleido-cranial dysostosis (figure 9a)*is rare genetic condition. The main clinical picture of cleido-cranial dysostosis is abnormalities of the skull, and collarbone along with dental abnormalities. The bones such as spine, collarbones and legs might be more fragile. Undeveloped or absant of the collarbone (clavicles - cleido generates the anterior mobility of the shoulders to the midline. Osteoporosis with coxofemoral and vertebra-medullary deformations, short stature, short finghers, narrow chestscoliosis

may also be encountered. The skull is wide and forehead is protruded with a vertical groove down the center. Nasal bridge is low and wide. Upper and lower jaws are small resulting in small midface. Hard palate is high arched or can be affected by malformation such as palatal cleft. Because of the small upper jaw the eyes are set wide, sinusis are small or absent and ear problems might be. Dental anomalies such as supernumerary teeth, crowded teeth (*figure 9b*), malocclusion is present. Primary teeth do not come out and permanent do not erupt on their own. Thin teeth enamel make them more prone to caries.





a b.

***Fig. 9* Cledo-cranial syndrom: a. clinical picture b. dental radiological image**

Craniofacial microsomia, also called *Parry Romberg's syndrome*, is a rare congenital condition in wich part of one side of the face and skull is undeveloped. In some cases of craniofacial microsomia symptoms are mild: skin tags in front of the ear on only one side of the face, associated with ear deformation and slight jaws undevelopment.

In severe case the disease involves a slow progressive loss of the skin, subcuraneus fat, muscles and bone, pigment change on the skin. The facial characteristics typically include undevelopment of one side of zigomatic bone, eye socket, outer and middle ear, upper and lower jaws, wich cause slant up of the facial asymmetry and displacement of the mouth toword the affected side, deformation or missing external ear, dental problems and difficulties with feeding and speech. The disease settles in childhood has

a slow evolution. It can include a part of the face or progressive involvement of some regeons, called *„en coup de sabre”.* The sensitivity in the affected part is preserved, the function is kept, although the muscles become progressive atrophy on the affected side.

Dermoid and epidermoid cysts are inclusion cyts caused by implantation of epithelial tissue into another structure. They expand very slowly, remain unobserved for a long time and do not cause functional changes. They are discovered accidentally or due to infection complications. Reaching excessive sizes they may cause some functional disorders. Most frequently are located on the head, face, neck and oral cavity.

Dermoid cysts are filled with developmentally mature skin complete with sweat glands, hair follicles, sometimes with luxuriant clumps of long hair, pockets of sebum, blood, fat, bone and cartilage, nails and even teeth.

Epidermoid cysts are lined with stratified squamous epithelium but do not contain the additional skin appendages. Slowly expanding unilocular cysts may produce only mild symptoms.

Located on floor of the mouth result in hinders the movements of the tongue, the phonation and the swallowing, and the clinical examination shows prominences on the midline line, below the tongue, raising the mucosa, exposing the frenulum and movement the tongue up and back. When pressed, the finger leaves a footprint in the form of a goblet, which, on palpation, is well delimited, movable, pasty. Those located under the mylohyoid muscle are prominent in the submental region, with the unchanged skin.

The differential diagnosis is made: with the ranula, systemic adenopathies, cronic and acute types of lymphadenitis.

Thyroglossal cyst or sinus is of congenital neck mass originated from the thyroglossus duct, which is normally atrophies and closed before birth. Thyroglossus duct is an embtiological anatomical structure, located mid-line and forming an open conection between the initial and final position of the thyroid gland.

The thyroglossal cystis lined by pseudostratified, ciliated columnar epithelium. Neck mass presents a sweeling located in the region of the hyoid bone, measuring 2 - 6 cm, Clinical examination show ovoidal soft sweeling,

painless, smooth, fluctuant and relatively mobile on superficial neck structure. The tyroglossus cyst will move upwards on swallowing due to the conection of the duct with foramen ceacum and adherent to the hyoid bone.

Differential diagnosis makes in relation to dermoid cysts, ectopic thyroid, enlarged lymph nodes.

A thyroglossal cyst may be infected and rupture unexpectedly producing thyrodlossal fistula or can appier after incomplete removal of a thyroglossus cyst. Fistulas may be complete and incomplete. The complete ones have two orifices: one - outside (between and the hyroid boneor just above the hyoid bone), another intraoraly (at the foramen ceacum of the tongue). The skin around the fistulous orifice is slightly depigmented, due to saliva that leaks from the orifice.

Surgical treatment consists of removal with resection of the hyoid bone according to radical Sistrunk procedure is an effective option.

Lateral cervical cyst and fistula arise from developmental abnormality arising from failure of obliteration of the second, third and fourth branchial clefts. The cyst is located along anterior to the sterno-cleido-mastoid muscle. They grow slowly, do not cause functional disorders and has a tendency to become infected. Lateral cervical cyst can be detected like a solitary, soft, painless, ovoidal, fluctuant and relatively mobile mass on the deep structure of the neck. The wall of the branchial cyst is cpmposed of squamous epithelium, columnar cells with or without cilia, or a mixture of both, with lymphoid infiltrate. In the fluid of the branchial cyst can be found a cholesterol crystalis

They can become infected, becoming painful, with the tension and congestion of the skin, with the alteration of the general condition.

The differential diagnosis is made with cystic lymphangiomas, cervical deep and superficial limphadenopathies (acute, cronic or specific cronic one); and with tumor mass

The cervical lateral fistulas, sometimes bilateral, may be complete, with an opening to the skin and to the internal opening to the lateral wall of the pharynx or in the supratonsillar fossa. Incomplete, blind fistula with the external orifice is located on the anterior edge of the sterno-cleido-mastoid muscle, below the angle of the mandible, up to sternal fork. There are data

on blind fistulas with the unique internal operning on the lateral wall of the pharynx. A clear, filamentous, slightly viscous liquid, similar to saliva, flows through the fistula. The treatment is surgical, with complete removal of the branchial cyst or fistula.

Pre-auricular sinus is congenital origin, caused by abnormality development of the first branchial arch from which the ear tubercles develop in the third month of gestation. Pre-auricular sinus occurs in front of the ear, with an opening located anteriorly to the external ear. Internal opening usually is located into the ear cartilage. They may become over infected.

Frenulum on the mouth is piece of soft tissue like little strings located in various part of the mouth. There are two most common types of frenulum in the mouth: lip (upper and lower) and tongue. The purpose of a frenum is to give rhe upper and the lower lips and tounger more stability in the mouth. Lingual frenum connects the base of the tongue to the floor of the mouth, Lip type of frenum connect the upper lip/lower lip to the gums and is located between the lower lip and lower gum and between the upper lip and upper gum.

Tongue-tie (ankyloglossia) and lip-tie are a condition where movment is restricted due to a short frenum. These anomalies are the most common developmental defects. Abnormal shape, size, position of the frenum may result in cascading developmental abnormalities and functional disorders in the mouth: normal eruption of the upper two front teeth; speech problems, pronunciation of certain letters of the alphabet. snoring and mouth breathing; exposing of the root of the teeth; dificalty in brestfeeding; struggling to latch on the brest; difficulty breathing during feeding; clicking sound during feeding; slow weight gain; malnutrition; fatigue and colic from breastfeeding.

Treatment for tongur-tie and lip-tie can be done by several methods: frenotomy, frenectomy, frenoplasty.

The frenotomy is indicated for babies less than 12 month when frenulum is thin. The surgical technique may be done with topical anaesthesia. The baby's head is held firmly and the transparnt part of the frenum is simply snipped (divided) with surgical scissors. These results in the immediate release of the tongue or lip and babies can be breastfed immediately after the procedure.

For older children and adults frenoplasty and frenectomy is indicates. Frenoplasty and frenectomy a general or local anaesthetic may be given beforehand. Stitches are needed when the tongue-tie or lip tie have been surgically divided. It may take a few weeks for the mouth to heal. Older children and adults may need speech therapy after the surgery.

Surgical procedures of *frenectomy* consist of their excision, which is done by incision around the frenum (on one side and the other), the thickened part inscribes and the mucosa along with the underlying fibrous tissue is removed.

In severe cases of lip-tie and tongue-tie (in cases of cleft lip and palate) the frenotomy and frenectomy does not achieve sufficient release of the lip or tongue. For thesereason are indicated several type of frenuloplasty: a) the horizontal incision of the frenulum that is followed by creating rhomboid wound that is sutured on the midline; c) „V"-„Y", type of surgery that allowed the lengthening of the frenulum; d) „Z" plastic surgery obtaining the elongation of the lip and tongue bycreating and replacement of the triangular flaps.

Learning objectives

1. Demonstrate on a frontal image of a human face those parts of the face that are formed by contributions from the frontonasal process and those that are formed by contributions from the first pharyngeal arch.
2. Describe the following as to site, composition, time of appearance, and fate: oropharyngeal membrane, oronasal membrane.
3. List the derivatives of the four pairs of facial processes and the pair of palatal processes.
4. Describe the development of the nose and primary palate.
5. Explain the differences between the processes of merging and fusion.
6. List all possible sites where facial or palatal clefts may develop, and for each indicate what facial or palatal processes are involved.
7. Describe the following and list which facial/palatal processes participate in their development: primary palate, secondary palate, and definitive palate.

8. Explain the extrinsic and intrinsic factors that may affect the normal development of the secondary palate.
9. Stages of human ontogenetic development.
10. Classification of rare congenital malformation of the cranio-facial area.
11. Congenital malformation of position of oral cavity lip and toun-gerfrenulum in children.
12. The role of genetic factors in the development of congenital malformations.
13. The most commonly syndromes encountered in the craniofacial region (clinical manifestations).

Tests

1. MC. List the main cinical manidasteation of Pierre Roben syndrome:
   A. deformation of the shape of the skull;
   B. palatal cleft;
   C. muscular ptosis of the tongue and muscles that participate in swallowing;
   D. insufficient development of the mandible in the sagittal plane;
   E. false exophthalmia.
   (A, C, D)
2. SC. From which embryological layer thyroglossal duct cyst arise:
   A. endodermal fold of the second branchial fissures;
   B. embriologicaș thyroglossal duct;
   C. cervical lymphatic bags;
   D. primary tooth buds;
   E. remains of the thymo-pharyngeal tract.
   (B)
3. SC. The chanal of the complete lateral cervical fistulas is shaped like:
   A. uninterrupted canal, with the external orifice on the cervical side, and with the internal one on the palatal amygdala;
   B. blind channel with the external orifice located on the edge of the sterno-cleido-mastoid muscle;
   C. blind channel terminated in soft tissue;
   D. blind channel with internal orifice on the lateral wall of the

pharynx;

E. blind channel with exit into the blind orifice of the tongue.

(A)

4. MC. Muscle ptosis of the tongue is a sign of the syndrome:

A. Pierre Robin;

B. Olbbrait;

C. Van der Woude;

D. Palatal cleft;

E. Patau Syndrom.

(A)

5. MC. The persistence of the thyroglossal duct during the postnatal development period of the child can be the cause of the following diseases:

A. cervical median cysts;

B. epidermoid cysts;

C. cervical lateral fistulas;

D. dermoid cysts;

E. cervical median fistulas.

(A, E)

6. SC. The uninterrupted chanal, with the external orifice on the cervical side, and the internal one on the lateral surface of the faringies is the characteristic for following fistulas:

A. complete lateral cervical;

B. pre-auricular congenital;

C. incomplete lateral;

D. complete medial;

E. incomplete medial.

(A)

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2. Congenital malformation of the face

2.1 General characteristic of malformations of the face

The first information about lip clefts comes from ancient times. In the 1st century AD, Celsius proposed the method of obturation in case of anomalies of the face. There are facts that prove that Galileo (2nd century AD) was aware of such malformation. In 1717, Peter I bought from Russian Federation (Dutch anatomist) an anatomical collection of preparations with different congenital malformations. By his order, this collection was completed for a century. In those times, very little was known about facial clefts. Only in the nineteenth century works about the frequency, etiology and treatment of these malformations appear. In literature, they have different names. In 1956, Zablosky-Deseatovsky described the cleft of the lip as „three lips”. In the people and in practical medicine the splits have long been called „rabbit lip” and „wolf mouth”.

Congenital clefts of the face are malformations characterized by interruption of facial continuity; they have the form of dehiscences that separate the facial structures, which during the embryonic development

were independent. Patients with cleft of the lip and palate have problems associated with anatomical deformities, developmental disorders, dental problems (supernumerary teeth, malocclusions), phonation problems (velopharyngeal insufficiency, secondary joint disorders), otolaryngological problems (Eustachian tube dysfunction), psychological and congenital anomalies associated.

Epidemiology. The lip/palate clefts are the most common congenital malformations of the face. The incidence and morphology of the clefts varies considerably according to race, to the geographical location of the country, to the sex. The incidence in white race groups is 1: 1000. The highest frequency is found in Native Americans (3.6: 1000), Japanese (2.1: 1000) and Chinese (1.7: 1000). The incidence is lower (0.3: 1000) in the Negroid race groups. In Republic Moldova the incidencewas found of 1.1: 1000.

Males are more affected (60-80 %) yjan females. The unilateral lip/palate cleft are found in 80 % of the patients with facial clefts, and the bilateral ones - in 2 0 %, the left side being more interested (60 %). The frequency of clinical forms is 20-30 % in lip clefts, 35-55 % in lip and palate clefts, 30-40 % in palate clefts. In Moldova, the ratio of boys to girls is 1.28: 1. Lip clefts are found in 28.75 % of cases, labio-palatal clefts - in 36.89 %, cleft palates - in 34.36 %.

2.2. Embryological development of the face

Embriological development of the face start during the third week with oral membrane that first is seen at the site of future face, between the primordium of the heart and brain. It is lies at the beginning of the digestive tract and is composed of ectoderm externally and endoderm internally. The opening between the future oral cavity and the forgut appear during the fourth week.

The oropharyngeal membrane breaks down when it stops growing. While tissues around it expand very rapidly, the oropharyngeal membrane's non-proliferating cells are gradually pulled apart because they cannot fill the expanding area.

The external face arice from two sources that surround the oropharyngeal membrane: a) the tissues of the frontonasal process that cover the forebrain, predominantly of neural crest origin; and b) the tissues of the first (or mandibular) pharyngeal arch, of mixed mesoderm and neural crest origin. A series of individualized tissue swellings gives rise to the different parts of the face. These are known as facial processes (prominences). The following facial processes may be recognized: a) the frontonasal process gives rise to: • a pair of medial nasal processes (that later contribute to a single globular [intermaxillary] process), and • a pair of lateral nasal processes. b) the first pharyngeal arch gives rise to: • a pair of mandibular processes (actually the pharyngeal arch itself), and • a pair of outgrowths of the arch: the maxillary processes (that later give rise to a pair of palatal processes).

At the end of the fourth week, two ectodermal thickenings: nasal placodes, appear on the frontonasal process. They are the precursors of the olfactory epithelium, responsible for the sense of smell. During the fifth week, lateral nasal and medial nasal swellings that surround the nasal placodes appear on the frontonasal process. These four nasal processes grow forward, while the nasal placodes remain relatively stationary. This gives the impression that the nasal placodes „invaginate". They stay behind and come to lie in blind nasal pits, surrounded by the nasal processes. This is the first step in the development of the nasal cavities.

Maxillary processes develop near the base of the first pharyngeal arch (mandibular arch). They enlarge and grow ventrally and medially, surrounding the future oral cavity. The maxillary processes grow rapidly, first meeting the lateral nasal processes, and then the lower extension of the medial nasal processes. This lower extension is known as the globular or intermaxillary process and will give rise to the midstructure (philtrum) of the upper lip.

Most facial processes begin as two separate swellings separated by a groove. Merging is the process by which the groove between two facial processes is eliminated. The tissues in the groove „catch up" by proliferating more rapidly than the surrounding tissues, causing the groove to become progressively shallower until it smoothes out. Merging is

critical. Without it, a deep depression (a facial cleft) remains between what used to be the facial processes.

Fusion is the process by which two facial processes, that were initially separated by a space, grow together. An example of fusion is the formation of the secondary palate. where two facial processes grow toward each other, touch each other and then fuse in the midline.

Development of palate By the 6th week the primary palate, formed by the two maxillary and two medial nasal processes, separates the developing oral and nasal cavities. Subsequently, between 6th and 8th weeks, the secondary palate is formed from two palatal processes (outgrowths of the maxillary processes). Primary and secondary palates together form the definitive palate.

Development of the primary (primitive) palate The primary palate develops at the same time as the external face (fifth and sixth weeks). and include pre-maxilla - the area of separation between the anterior regions of the face and oral cavities It consists of a part of the upper lip, the gums containing the four incisors and the anterior part of the hard palate, from the incisal canal to the anterior.

The upper lip arize from three facial processes, which appear in the 4^{th} week of gestation. The first is the frontal-nasal processe, which develop through mesenchymal proliferation. In week 4, there is an active proliferation of the ecto-mesenchyme in the frontal-nasal bud, after which the medial and lateral nasal processes are formed. The external nasal processes remain in a superior position, and the internal nasal one descend and extend, joining the medial line and forming the interma-xillary segment of the upper lip. The lateral part of the lip derives from the two maxillary processes. In the 5^{th} and 6^{th} weeks of gestation, the maxillary processes undergo extensive growth The maxillary first coming into contact with the lateral nasal processes and secondly with the globular process of the merged medial nasal processes (philtrum). Initially the medial nasal and lateral nasal processes come into contact, and secondarily, the medial nasal and maxillary processes come together (just below and in front of the contact site between the medial and lateral nasal processes) and pinch some epithelium between them. This sheet of

epithelium is composed of future nasal epithelium superiorly, and future oral epithelium inferiorly. The two layers of epithelium are then pulled apart, making the mesenchyme between medial nasal and maxillary processes continuous: the core of the primary palate. Posteriorly, behind the primary palate, the nasal epithelium continues to touch the oral epithelium. This patch of epithelium is called the oronasal membrane. Around the 6th week of development this membrane is ripped open in the same manner as the oropharyngeal membrane (cells stop undergoing mitosis). The resulting opening is called the primitive choana, and it connects the nasal cavity to the oral cavity.

Development of the secondary palate. The secondary palate refers to the hard and soft palate and is located posterior to the site of the future incisive foramen of the hard palate. The secondary palate includes the palatine processes of the maxillary bones and the soft palatal muscle (the soft palate and the hard palate) and is located posteriorly the primary palate. The pathogenesis of the secondary palate begins at week 5 of gestation and ends completely at week 12 of gestation.

As the face grows in an antero-posterior dimension, the primary palate soon is too short to provide adequate separation between the nasal cavities (respiratory function) and the oral cavity (digestive function). A new structure: the secondary palate develops to further separate these cavities.

Formation of the secondary palate and nasal septum arise from palatine shelves that grow medially from the maxillary swellings. During the same period, growth of the nasal septum separates the left and right nasal passages. The palatine shelves at first grow inferiorly on either side of the tongue but then rapidly rotate upward to meet in the midline, where they fuse with each other and with the inferior edge of the nasal septum. During the seventh and eighth weeks, the medial walls (the oral surfaces) of the maxillary processes produce a pair of thin medial extensions, called the palatal processes (shelves). Initially these grow predominantly vertically: downward and parallel to the lateral surfaces of the tongue. By the beginning of the eighth week the tongue begins to move and the lower jaw grows downward and forward. Thus, by the end of the eighth week, the

palatal processes rotate rapidly upward to a horizontal position and fuse with each other and with the primary palate. Palatal shelf-elevation is the result of an intrinsic shelf elevating force, chiefly generated by the progressive accumulation and hydration of hyaluronic acid. The fused palatal processes form the secondary palate - together with the primary palate they form the definitive palate.

The successful development of the secondary palate depends on many factors: *Extrinsic factors* • swallowing movements of the tongue, moving the tongue out of the way from in between the two palatal processes and allowing them to move upward • downward and forward growth of the lower jaw and tongue complex, providing more space above the tongue for the palatal processes • straightening of the cranial base as the result of growth of the neural mass, establishing the mechanical environment for the palatal processes to swing upwar a face, the broken lines indicate the possible locations of facial clefts. The areas between the broken lines correspond with the areas formed by the original facial processes. *Intrinsic factors* in palatal processes • mesenchyme cell proliferation - increasing volume- ceases hours before palatal processes swing upward extracellular matrix production - increasing volume hydration of extracellular matrix - major increase in volume and turgor of palatal processes just before they swing upward • medial edge epithelium (MEE) - covering the free edges of the palatal processes apoptosis of MEE surface cells immediately prior to fusion development of a temporary glycoprotein coat, enabling adhesion between MEE cells of the two opposing palatal processes.

2.3 Facial malformations. Classification. Clinical manifestation

Various types of cleft lip and cleft palate may be encountered clinically. Complete clefts indicate the maximum degree of clefting of any particular type (e. g. a complete cleft of the secondary palate, a complete cleft of lip, alveolar process and primary palate, or a combination of these two). Incomplete clefts are found when some merging or fusion has taken place during development. Clefts may be unilateral or bilateral. The important thing to remember clinically is: each site where

merging or fusion occurs during development of face and palate is a potential site for a facial/palatal cleft.

Classification. The upper lip, premaxilla, and primary palate are formed by the merging of three structures: the frontonasal process and the right and left processes of the maxilla. Any disturbance in the merging of the above processes results in the formation of the clefts. The incisive foramen is a basic anatomic landmark for classification of cleft lip and palate. There are about one hundred combinations of the cleft lip and cleft palate. Proper diagnosis of this cleft formation and its severity assessment helps in planning and execution of the appropriate treatment. Various classifications of facial clefts cleft lip and cleft palate have been reported.

First person to classify malformations of the face was Forster3 in 1861, a Pathologist from Wurzburg (Germany). Davis and Ritchie1 in 1922 classified the congenital clefts into three groups according to the position of the cleft in relation to the alveolar process. Group I: Pre-alveolar clefts, unilateral, median, or bilateral; Group II: Post-alveolar clefts involving the soft palate only, the soft and hard palates, or a submucous cleft; Group III: Alveolar clefts, unilateral, bilateral, or median. Their classification had many shortcomings such as, insufficient descriptions of cleft lip, cleft of the primary palate with intact secondary palate and presence or absence of alveolar involvement, and the incisive foramen.

Veau's classification (1931) comprises 4 groups. Group I – defects of the soft palate only; II – defects involving soft and hard palate extending not further than the incisive foramen; III – complete extending from the soft palate to the alveolus, usually involving the lip; IV – complete bilateral cleft, resembles Group III but is bilateral. When cleft is bilateral, pre maxilla is suspended from the nasal septum.

In 1942 Fogh Anderson gave a very similar classification based on embryological development, which is as follows: Group 1 – clefts of the lip- unilateral or bilateral; Group 2 – clefts of the lip and cleft palate (single or double); Group 3 – clefts of the lip and palate upto the incisive foramina.

The Spanish surgeon Vilar Sancho classifies the clefts into complete and incomplete. The location of each cleft is indicated by its first

letter or the first letter of the Greek word: K - for the lip (*kilos*), G - for the jaw (*gnato*), U - for the palatal vault (*uranium*), S - for the soft palate (*stafilos*), SK for cleft (*skisis*).

In 1958 Kernahan and Stark recognized the need for a classifycation based on embryology rather than morphology. Primary palate comprised of premaxilla, anterior septum, and lip. The roof of the mouth - from the incisive foramen or its vestige, the incisive papilla, to the uvula - is termed the secondary palate. The incisive foramen is the dividing line between the primary and secondary palates. Their classification was as follows: Clefts of primary palate: Unilateral, Bilateral and median Clefts of Secondary palate: Unilateral, Bilateral and median Clefts of primary and secondary palate: Unilateral, Bilateral and median based on embryological principles used by Kernahan and Stark, Harkins and associates (1962), presented a modified version of classification of facial clefts.

Cleft of Primary Palate

A. Cleft Lip

(1) Unilateral: right, left (a) Extent: one-third, two-thirds, complete

(2) Bilateral: (a) Extent: one-third, two-thirds, complete

(3) Median (a) Extent: one-third, two-thirds, complete

(4) Prolabium: small, medium, large

(5) Congenital scar: right, left, median (a) Extent: onethird, two-thirds, complete

B. Cleft of Alveolar Process

(1) Unilateral: right, left (a) Extent: one-third, two-thirds, complete

(2) Bilateral: (a) Extent: one-third, two-thirds, complete

(3) Median (a) Extent: one-third, two-thirds, complete (4) Submucous: right, left, median

(5) Absent incisor tooth

II. Cleft of Palate

A. Soft Palate

(1) Posteroanterior: one-third, two-thirds, complete

(2) Width - maximum (mm)

(3) Palatal shortness: none, slight, moderate, marked

(4) Submucous cleft (a) Extent: one-third, two-thirds, complete

B. Hard Palate

(1) Posteroanterior (a) Extent: one-third, two-thirds, complete

(2) Width - maximum (mm)

(3) Vomer attachment: right, left, absent

(4) Submucous cleft (a) Extent: one-third, two-thirds,

Mandibular Process Clefts

A. Lip (a) Extent: one-third, two-thirds, complete

B. Mandible (a) Extent: one-third, two-thirds, complete

C. Lip Pits: Congenital lip sinuses

Naso-ocular: Extending from the narial region toward the medial canthal region.

Oro-ocular: Extending from the angle of the mouth toward the palpebral fissure.

Oro-aural: Extending from the angle of the mouth toward the auricle.

Schuchardt and Preifer's (1963) simvolic classification, proposed simvolic indexing cases of lip and palate cleft through a graphical representation (pentagon). Its sides indicate the severity of the defect, the areas involved, the width of the affected area and the degree of premaxillary protrusion.

In 1979, the embryological classification was integrated into the International Classification of Diseases (ICD) by the World Health Organization16 in 1979. The sequence though, was not absolutely correct, it is as follows: 749.0 cleft palate; 749.1 cleft lip; 749.2 cleft lip and palate. In Chapter XVII of WHO ICD Version 200717, discusses about the congenital malformations, deformations and chromosomal abnormalities (Q00-Q99) and Cleft lip and cleft palate (Q35-Q37). Q 35 Cleft palate includes fissure of palate, Palatoschisis and excludes cleft palate with cleft lip. Q35.1- cleft palate, Q35.3- cleft soft palate, Q35.5- cleft hard palate with soft palate, Q35.7- cleft Uvula, Q35.9- cleft palate unspecified. Q 36 Cleft lip includes Cheiloschisis, congenital fissure of lip, hare lip, labium leporinum and excludes cleft lip with cleft palate. Q 36.0 Cleft lip, bilateral, Q 36.0 Cleft lip, bilateral, Q 36.1 Cleft lip, median, Q 36.0 Cleft lip, unilateral. Q37 includes cleft palate with cleft

lip. Q 37.0 Cleft hard palate with bilateral cleft lip, Q 37.1 Cleft hard palate with unilateral cleft lip, Q 37.2 Cleft soft palate with bilateral cleft lip, Q 37.3 Cleft soft palate with unilateral cleft lip, Q 37.4 Cleft hard and soft palate with bilateral cleft lip, Q 37.5 Cleft hard and soft palate with unilateral cleft lip, Q 37.8 Unspecified cleft palate with bilateral cleft lip and Q 37.9 Unspecified cleft palate with unilateral cleft lip.

Changes in classifications from ancient times to the present continue to be proposed.

Spina in 1974 proposed a modification of classification presented by the nomenclature committee of the American Cleft Palate association.

Smith et al (1998) modified the Kernahan Y classification further, in an attempt to make up for the shortcomings.

The LAPAL system 2007 consists of only five Arabic numerals that describe accurate anatomic components and the extent of any cleft.

Using the Kernahan concept with modification, Schwartz et al (1993) developed a three-digit numerical system RPL system to record the location and number of anatomic components involved in cleft deformities

Koch and Koch in 1995 proposed a new extended classification, LAHSN of cleft deformities. In addition to the lip, alveolus, hard palate, soft palate, they also considered the Vomer and the micro forms in three dimensions

Mortier et al (1997) developed a dual scale, which included two indicators: one corresponding to the severity of the cleft (ISS, or initial severity score) and another related to the surgical result (PRS, or postoperative results score). This indicator considered seven features to describe the patient. A comparison of the ISS and PRS allows for more objective judgment of the surgical result. However, it has been applied only to unilateral incomplete clefts of the primary palate.

Tessier (1976) formulated a classification system based upon his extensive personal experience.

Percy Rossell-Perry gave the Lima clock diagram 2009, is the design of a new diagram for cleft lip and palate, based on the degree of severity of the four basic cleft components: nose, lip, primary palate, and secondary palate.

Guțan A.M. is one of the scientists, from Republic Moldova, who tried to change the classification according to embryological development and clinical manifestations.

Guțan A. (1982) classifies the clefts into two genetic groups. The first genetic group includes unilateral and bilateral clefts. 1. Lip clefts: a) marginal (includes lip red); b) incomplete (includes the lip socket); c) complete non-penetrating (it comprises the lip in full, up to the nostril); d) complete penetration (it comprises the lip in its entirety, passing through the nostril). 2. Clefts of the lip and alveolar apophysis: a) marginal (recess in the alveolar ridge); b) incomplete (extends within the body of the alveolar ridge); c) complete non-penetrating (reaches the incisor foramen); d) complete penetration (passes through the incisive foramen). 3. The cleft of lip and palate. Clefts of the lip, alveolar apophysis and palate: a) total, b) incomplete.

The second genetic group - median palatal cleft involving the secondary palate: a) of the uvula (involving only the uvula); b) of the soft palate (within the limits of the soft palate); c) incomplete; d) complete non-penetrating (extending not futher than the incisive forament); d) complete penetrated cleft (extending from the uvula to the incisive foramen in the midline, then deviating to one side and usually extending through the alveolus at the position of the future lateral incisor tooth).

The International Confederation of Plastic Surgery officially adopted the classification of cleft lip and palate proposed by Kenahan and Stark, 2002. These authors considered embryological aspects to classify clefts into three groups: Group 1, anterior, primary cleft palate, involving right, left, or both lip and alveolus; Group 2, anterior and posterior clefts of the palate, primary or secondary, involving lip, alveolus and hard right, left or both palate; Group 3, fissures of the right, left or both posterior palate, hard palate and soft palate. Some facial clefts are rare and may be oblique, transverse, of the lower lip, nose and upper lip median cleft, with or without hypoplasia

The clinical manifestations of the clefts have there own characteristics of anatomical and functional disorders. According to the anatomical disorders of the lip clefts clinical manifestation are bazed on the

extending area (lip, alveolus, nasal floor), laterality (unilateral left, unilateral right, bilateral) and severity (width and structures involved). Thus, all lip clefts according to the anatomical disorders of the scheletal, mucosal and skin interruption, have general clinical features a) defect on the upper lip due to the interruption of the continuity of the orbicular muscle, skin snd mucosa; b) shortening of the upper lip along the cleft due to the insertion of the muscle fibers at the base of the aperture pyriformis; c) asymmetry of the nose structures. Thus, anatomical characteristics of the unilateral cleft lip include nasal deformities of the tip, columella, nostril, alar base, septum and skeletion. The lower lateral cartilages on the cleft side have short medial crus, lateral crus that is lomger and draw into a S-shaped fold. The caudal septum is deviated toward the noncleft side. Columella is shorter on the cleft side and together with the nasal tip is directed toward the noncleft side. due to muscles imbalance. The alar base on the cleft side is positioned more horizontal, laterally, inferiorly and posteriorly.

2.4 Cleft lip. Clinical picture. Diagnosis. Treatment approach

The **submucous lip** *cleft s(microform)* is a very mild form of incomplete and has no visual clinical manifestation. Only concern the interruption of muscular continuity, leaving the entire mucosa and the upper lip skin whole. Diagnosis can be established by clinical inspection or by magnetic resonance imagining (MRI). It's appears in the form of a groove on the skin, whose feature can variably include intended mucosa, notched vermillion, disrupted white roll, furrowed philtral skin, flattened nasal sill and nasal ala.

The unilateral incomplete lip cleft (*figure 10*) involves the partial vertical height of the lip and appear as a small notch in the inferior part of the upper lip. On the superior area of the lip there is a Simonart band that consists of the skin with variable amounts of orbicularis oris muscle fibers. The nasal floor and alveolar processes remain unaffected; Mild facial asymmetries arising from the orbicularis oris muscle fibers that are asymmetrically oriented along the cleft margins and continuous across Simonart's band, loss of Cupid's bow, and nasal deformity (displacement

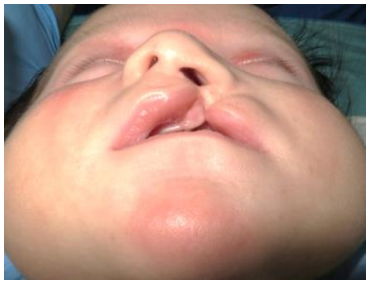
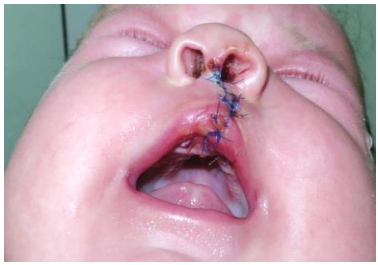
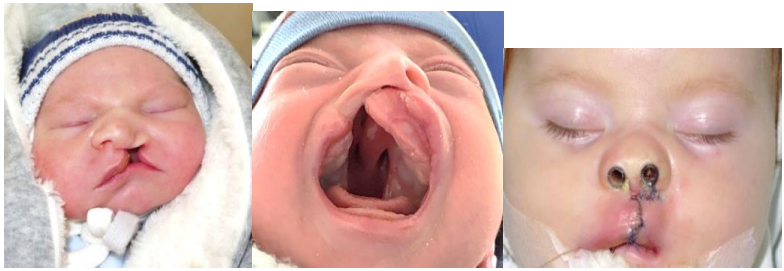



Fig. 10. **Congenital malformation of the face. a. Uncomplete left cleft lip. b. Uncomplete left cleft lip and gum.**

of the ipsilateral lateral crus of the alar cartilage laterally, inferiorly, and posterior, the tip is flattened and deflected to the non-cleft side, the ipsilateral nostril is oriented horizontally rather than vertically). Functional changes are of minor importance, breastfeeding is possible.

The total unilateral cleft lip/palate (*figure 11*) concern primary palate



a b c

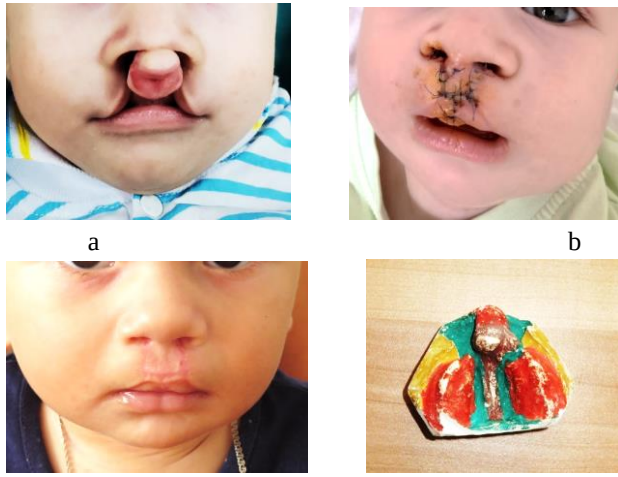
Fig. 8. **Congenital malformation of the face. a. unilateral left/right cleft lip/palate before surgery, c. after surgery.**

which is separated into two fragments (big and small). This separation extends beyond the upper lip, base of the nose and includes the bones of the upper jaw, the nostil, the alveolar ridge and the hard palate up to the incisive orifice (primary palate). The sagittal gap of the cleft pass between the lateral insisor and canine on the left or/and right side of the gum. The intermaxillary relations near the cleft is disordered. The anterior part of the biggest one often is displaced anteriorly and tilted superiorly. Sometimes the big fragment is displaced anteriorly up to 1 cm. The small one is hypoplastic and has palatal position.

There is rotation and distortion of the vermillion with loss of Cupid's bow and philtral landmarks on the cleft side. Continuity of the orbicularis oris muscles is compromised with abnormal insertion thus being asymmetrically oriented along the cleft margins. In the medial lip element, the orbicularis inserts into the anterior nasal spine and columella and in the lateral lip element, the orbicularis inserts in the lateral aspect of the alar base, nasolabial fold and into the nostril base periosteum of the pyriform rim.

Dental position on the anterior part of the fragments is displaced. Lateral incisor, may or may not be present. Occasionally, supernumerary teeth appear, form disorders, hypoplasia, or lack thereof. Anatomical changes in some forms of upper lip cleft disrupt the breastfeeding function of the mew-born, and they have to be artificially breastfed in an early period.

The total bilateral cleftslip (figure 12) affect the primary palate bilateral which is separated into three fragments (two maxillary and



a b c d

Fig. 12. **Congenital malformation of the face. Bilateral cleft lip and palate a. b. c. Clinical case: the total bilateral lip cleft. d. the cast of the child with the total bilateral lip clefts.**

premaxilla). The cleft extend to upper lip, the nasal floor and the alveolar processes. Oral cavity communicates with nasal cavity on either side of the nasal septum. In bilateral cleft lip the premaxilla is fused with the vomer bone, may be undeveloped or obviously developed and displaced on the left or right side, The prolabium, consist of soft tissues of the premaxilla without muscle fibers, also lacks Cupid's bow and philtral columns bilaterally. The columella is severely shortened or absent while the lateral crura are displaced laterally, producing a broad, flat nasal tip.

The treatment of children with cleft lip deformity has long challenged surgeons. Numerous surgical techniques have been developed to restore function, symmetry and aesthetics. The treatment of lip clefts vary depending upon the severity of the cleft and include preoperative treatment, operative primary closure of the upper lip (*Fig. 13*) and postsurgical rehabilitation.

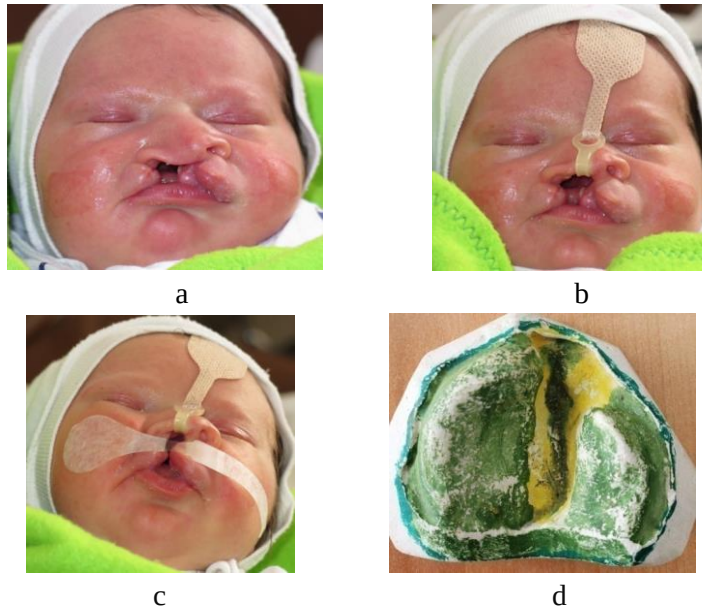


Fig. 13. **Right cleft lip/palate. a. b, c, preoperative adesive band, b. intraoral model of the hard palate in children with cleft lip/palate.**

Preoperative orthopedic treatment is a procedures performed by an orthodontist who specialize in treating craniofacial deformities. Presur-

gical orthopedic management is performed to reduce the width of the bony cleft and to align the maxillary arch prior to definitive lip repair Naso-alveolar molding and different type of applies gradually brings the palate and lip together and provides symmetry of the nose, preparing the patient for surgical outcomes. Presurgical molding start in the first few weeks after the birth and continuing until the patient is ready for primary closure. For the repositioning of the alveolar segments, active or passive palatal mouthguards can be used too. Sometimes, surgical lip adhesion is practiced. Adhesive tape for imobilization prior to surgical repair of unilateral and bilateral clefts consists of long strips was applied from cheek to cheek for 6 week. In this way reduction of the alveolar gap and lip narrowing from partial to complete apposition is obtained. The surgical reconstruction of the lip and nose must keep the following instruction: upper lip of the affected side to be elongated and to create an equal to that of the unaffected side, and in the bilateral clefts to correspond to the equivalent length. The groove of the lip in the unilateral clefts (filtrum) must be preserved, and the edges reconstructed symmetrically. The cutaneous mucosal margins and cupid's bow should be symmetrically restored; a groove with a satisfactory depth should be preserved or formed on the midline of the lip; symmetrical restoration of the nasal floor, columella, nostrils.

The surgical techniques in the lip primary closure passed several steps. First successful closing of a cleft lip was reported in 390 BC in China. Since that time, numerous techniques have emerged for the treatment of the cleft lip.

Early surgical techniques in treatment of lip deformity involved straight line repairs. Ambroise Paré (1575) first described a technique in which the labial aspects, previously incised with a long needle, were transfixed and brought together with wires anchored to this needle. William Rose (1879) and James Thompson (1912), Ladd, Braun and Veau (1938), described a similar technique, now known as the Rose-Thompson, consisting of cuerved incisions across the labial edge parts, so that during the suturing, there is an extension of the lip to prevent the „notches" in the vermilion. The priority of these methods is the coincidence

of the incision line with the upper lip filter, simplicity and speed but in wide clefts this method do not give satisfactory extention of the upper lip, asymmetric cupid's bow, a prominent scar and retrusion of the maxilla, Complications of these techniques consist of the linear scars after surgery, limitations of the straight closing and notch in the vermilion. Though the straight line repair techniques are less used,

The next step came with techniques using flaps. The goal of these procedures were lengthening the lip and preventing scar retraction, trying to keep the lip symmetry and „cupid's bow" more natural. LeMesurier (1945), May and Trauner proposed a reconstruction with quadrangular flaps. Charles Tennison (1952), Obuhova (1955), Randall, Lemos and Spina (1963) disclosed a method with triangular flaps and Z-plasty. The technique proposed by these authors, allows the extension of the upper lip, which is directly proportional to the size of the triangular flap create on the big fragment of the upper lip. Instead there is no ideal Cupid's bow, because of the incision of the upper lip filter.

The third step was the technique of advancement and rotation, presented by Ralph Millard Jr at the First International Congress of Plastic Surgery, in Stockholm. This technique was first published in 1957, and spread quickly due to the simplicity of its preparation and the good results.

The goal of primary closure of the cleft lip is to create the upper lip with appropriate vertical length and symmetry, repair of underlying structures with normal muscle function and primary treatment of associated nasal deformities. Cleft lip is usually is repaired between 3 and 6 months of age. During those first few months, new-born is monitored for adequate weight gain and nutrition, and to make sure that there are no issues relative to breathing while eating. One rule, which successfully passed the test of time, is the one in „10", applied for the first surgery (the child must be over 10 weeks, over 10 % hemoglobin and weight over 10 kg).

Labiaplasty technique developed by Millardis the most widely used by cleft surgeons. Its principles serve as the foundation of many unilateral repairs today. Millard preserved anatomical landmarks: the cupid's bow and the philtral column. Downward rotation of the medial

lip element restores vertical lip height and advancement of the lateral lip element repositions the alar base.

Key points:

1) The position of point 3 may be determined by transposing the distance between 1 and 2, such that the distance between 1 and 2 is equal to the distance between 1 and 3.

2) The distance between the alar base and Cupid's bow peak on the noncleft side should equal that on the cleft side, ie, the distance from 2-5 is equal to the distance from 7-6.

3) The difference between the distance from the columellar base to points 2 and 3 represents the deficiency in vertical length that must be gained to level the Cupid's bow. Although the rotation incision allows point 3 to drop inferiorly, some vertical deficiency of the cleft side may remain. The added length may be gained by making a small backcut medial to the philtral column on the noncleft side. The advancement flap derived from the lateral element fills the opening created by the rotation incision and any backcut in the medial element; hence, the distance from 3-5 plus the added length gained by the backcut equals the distance from 6-7. Introduction of a small triangular flap from the lateral element into a small transverse incision in the lower part of the lip may also serve to lengthen the cleft side of the medial element and to improve the contour of the lip. The base width of this flap is equal to the height of the vermilion-cutaneous roll.

4) The rotation incision curves gently from point 3 to the columellar base, hugging the columellar-lip junction, and stops medial to the philtral column on the noncleft side. Crossing the normal philtral column results in an undesirable elongation of the lip on the noncleft side. In the infant with a rectangular philtrum, this incision may be modified as described by Mohler.

5) Place point 7 on the lateral element at a point level with the Cupid’s bow peak on the noncleft side (point 2) and where the white roll remains well developed. Placing this point too far laterally produces an unnatural shortening of the lateral lip element, which results in a noticeable imbalance. Placing this point too far medially, where the white roll

is poorly developed, results in a noticeable irregularity of the white roll. To gain some extra vertical height, if needed, point 7 may be moved 1 mm laterally and point 3 moved 1 mm medially.

6) The advancement incision curves from point 7 to point 8, then a variable distance to point 9, depending on the amount of rotation needed to correct the flare of the displaced alar base.

7) When possible, line up the point at the junction of the wet and dry vermilion; this point is also called the red line.

Repair of the orbicularis oris. Reorientation and repair of the orbicularis oris muscle bundles are essential for normal lip function and eversion of the lip border. Failure to adequately address the muscle at the time of lip repair results in abnormal motion or contour when pursing the lips and in a characteristic bulge in the lateral lip element. A variety of techniques for reorienting the orbicularis oris muscle fibers in unilateral clefts have been described, although the optimal method of muscle repair remains to be determined. Park advocates careful identification and precise reapproximation of the superficial and deep components of the muscle. Primary nasal correction. In every case, reconstruct both the lip and the nose at the primary operation. Repair of the lip in infancy while delaying nasal repair until later in childhood is no longer appropriate. Reconstruction of the cleft nasal deformity remains the most challenging aspect of cleft surgery. Principles of primary nasal correction include the following: 1. Wide undermining of the nasal skin on the cleft side, freeing the skin from the underlying nasal skeleton. 2. Elevation of the slumped alar cartilage on the cleft side to the normal level using internal or external suspension sutures. 3. Medial advancement of the lateral crus and alar base on the cleft side.

Bilateral cleft lip repair. Prior to surgical repair, the use of presurgical orthopedic appliances can reduce significant premaxillary protrusion. The premaxillary segment varies considerably in size and in the extent of its protrusion. In incomplete clefts, attachment of the premaxilla to one or both lateral maxillary segments limits premaxillary protrusion. In complete clefts, retroposition of the premaxilla prior to definitive lip repair often is necessary. This procedure may be

accomplished through presurgical orthopedics, using external traction devices or passive orthodontic plates. Retroposition of the premaxilla may also be accomplished through surgical lip adhesion. Surgical setback of the premaxilla, a technique popular in the 19th century, is associated with subsequent midfacial growth impairment and should be avoided. Modifications of the Millard straight-line, banked, forked-flap technique currently are the most widely used methods for repair of bilateral cleft lip. These techniques work well for the repair of complete bilateral clefts and may be modified for the repair of incomplete and/or asymmetrical clefts.

Key points.

1.The prolabium is always used in reconstruction of the philtrum, even if severely deficient.

2. The prolabial vermilion nearly always is deficient, and the prolabial white roll usually is indistinct. Therefore, in most cases, the prolabial mucosa is turned down to line the buccal alveolar sulcus; the tubercle and white roll are reconstructed using paired white roll-vermilion-orbicularis marginalis flaps from the lateral lip elements brought beneath the prolabium.

3. The prolabium must not be left too wide and should rarely exceed 5-6 mm in width.

4. The orbicularis peripheralis muscle is freed from its abnormal attachments at the alar bases and from the overlying dermis. This allows the muscle to be mobilized and reconstructed over the premaxilla.

5. Anatomic positioning of the alar cartilages is performed at the time of primary lip repair.

2.5. Postoperative details. Complications

Postoperative details. 1**.** After surgery, feeding is resumed using a soft crosscut nipple. 2. Infants remain hospitalized for intravenous hydration until oral intake is sufficient (usually 24 h). 3. The suture lines are kept clean by gentle application of a dilute hydrogen peroxide solution, and a small amount of antibiotic ointment is applied to the repair 3 times daily and after feeding. 4. If nonresorbing suture material is used, the sutures are removed by the fifth postoperative day. 5. Soft elbow restraints

are used for 2-3 weeks to keep the infant from manually disrupting the repair. *Follow-up.* Arrangements for suture removal are made prior to or immediately after discharge. All patients require long-term follow-up care. A dedicated multidisciplinary team approach and evaluation in different stages of the patient's life is important. Assess speech, language, hearing, somatic growth, and development regularly. Appropriately assess general dental health. Orthodontic management and seconddary surgical procedures, such as bone grafting, are carried out during the school years. Patients with significant midface retrusion may require treatment. Secondary procedures to correct the tip in nasal asymmetry may be performed at school age; however, if the reconstruction involves osteotomy, delay the procedure until the completion of nasal growth (age 16-17 y). Emotional difficulties may emerge because of poor self-esteem during adolescence and should be recognized and addressed early.

Complications. Complications following cleft lip repair are unusual. Wound infections following surgery are uncommon and are treated with appropriate antibiotic therapy. Although immediate wound dehiscence is best repaired prior to discharge, treat delayed dehiscence after the scar has settled. Achieving good consistent results in primary repair of cleft patients depends on in-depth understanding of the cleft lip-nose deformity. Secondary deformities may result from poor operative planning, operative error, or postoperative scar contraction. Most secondary deformities that are encountered by the senior author are from inadequate understanding of the biology of the cleft deformity or of the described technique. This probably results from inadequate technical appreciation of how to release, reshape, and reconstruct the lip and nose adequately while minimizing a detrimental scar.

Learning objectives

1. In which period of intrauterine development the formation of the upper lip takes place?
2. Ethiopathogenesis of lip clefts.
3. Morbidity of upper lip clefts compared to other forms of clefts. The spread of lip clefts in children in Moldova.

4. Anatomical deformities of the upper lip clefts.
5. Classification of upper lip clefts.
6. Functional disorders in upper lip clefts.
7. Characterization of cheiloplasty methods (Limberg, Tenieson-Obuhov, Milard).
8. Basic purpose in the surgical treatment of upper lip plasties.
9. Optimal age of upper lip plastic surgery.
10. Preparation of the child for cheiloplasty surgery.

Tests

1. CM. The surgical treatment of anatomical recovery of the upper lip aims at:
   A. elongation of upper lip;
   B. restoration of continuity;
   C. harmonious restoration of nostril relief;
   D. restoration of suckling reflex;
   E. restoration of the connection between the upper and lower alveolar apophyses.

   (A, B, C)
2. CS. The Tenieson-Obuhov surgical procedure to remove the upper lip cleft is based on:
   A. advancing the quadrilateral flap;
   B. linear incisions;
   C. sliding the triangular flap into the inside of the lip;
   D. edge stitching and suturing;
   E. no answer is correct.

   (C)
3. CM. In the cleft lip the orbicular muscle at defect level is inserted at:
   A. top of the cleft;
   B. pyriform opening;
   C. alveolar apophysis;
   D. center of cleft;
   E. vomer.

   (A, B)
4. CM. For all lip clefts, anatomical disorders are characteristic for:

A. shortening of upper lip;
B. elongation of alveolar apophysis;
C. exposed nostril;
D. the nasal lobe deflected to the healthy side;
E. deviation of nasal septum.

(A, C, D)

5. CS. Cleft of the orbicular muscle with preservation of the integrity of the skin and mucosa of the upper lip characterizes anatomically the clefts:

A. completely unilateral;
B. completely bilateral;
C. median;
D. incomplete;
E. camouflaged (skisis).

(E)

6. CS. Clefts of the lip that partially interest the lip (the cleft of the lip) or the lip and the threshold of the nose, leaving the alveolar ridge integral are characteristic for the clefts:

A. camouflaged;
B. incomplete;
C. complete;
D. totally unilateral;
E. totally bielateral.

(B)

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3. Congenital cleft palate. Clinical manifestation. Methods of diagnosis and treatment

3.1 Anatomy of the normal and cleft human palate

Normal palate development is a complex process that involves numerous growth factors, their receptors and intracellular signaling pathways. Clefting of the palate can result from failed midline fusion of the paired palatine shelves due to embryologic errors of formation including inadequate growth of the palatine shelves, failed shelf elevation and fusion, and secondary degradation after fusion. The normal palate is composed of the hard and soft palates. The greater palatine neurovascular bundles emerge bilaterally from the palatine canals via the greater palatine foramina. The hard palate is responsible for continues the width and anterior projection of the maxillofacial framework, the soft palate raise the sphincter toward the posterior pharyngeal wall, thereby separating the nose from the mouth and are responsible for phonation, breathing, swallowing, and blowing. The muscular soft palate, or velopharyngeal sphincter, is composed of five pairs of muscles: the levator veli palatini, tensor veli palatini, uvulae, palatopharyngeus muscles, and palatoglossus muscles.

Embryologically, the muscles of the palate are all derived from the fourth pharyngeal arch and therefore innervated by vagus nerve (pharyngeal branch). The sole exception to this is the tensor veli palatini, which is derived from the first pharyngeal arch, and therefore innervated by the mandibular division of the trigeminal nerve. The tensor and levator veli

palatini muscles both arise from the Eustachian tube and are important anatomic components of cleft palate repair. The tensor veli palatini muscles arise from the medial pterygoid plate and spine of the sphenoid bone. They travel inferiorly as a tendon, coursing around the pterygoid hamulus, and insert on the soft palatal aponeurosis near the soft-hard palate junction. They function to control opening of the Eustachian tube, permitting aeration of the middle ear. The levator veli palatini muscles arise from the petrous portion of the temporal bone and from the cartilage of the Eustachian tube. They travel inferomedially and insert at the midline on the palatine aponeurosis. This interdigitating insertion forms the bulk of the anterior „levator sling" and therefore, the anterior portion of the velopharyngeal sphincter. The uvula originates from the nasal spine and palatine aponeurosis and insert on the mucosa of the uvula, assisting in elevation of the uvula. The palatoglossus and palatopharyngeus muscles originate from palatine aponeurosis and course inferolaterally to insert on the lateral tongue and pharyngeal wall, respectively; both assist in narrowing the orophangeal aperture. An additional pair of muscles is the superior pharyngeal constrictors; these originate from the medial pterygoid plate and Hamulus, pterygomandibular raphé, and mylohyoid line of the mandible, and course posteriorly to insert on the midline pharyngeal raphe.

The role of the palate is to provide a barrier between the nasal and oral portions of the respiratory tract. Velar actions with deglutition, respiration, and phonation are similar to those of a sphincter; hence, the velopharyngeal mechanism is often termed the velopharyngeal sphincter.

Familiarity with the anatomy of the palate is essential in understanding functional and surgical repair. Blood is supplied to the hard palate by the greater palatine artery, which enters via the greater palatine foramen. Nerve supply originates from the maxillary branches from the trigeminal nerve, which forms a plexus that innervates the palatal muscles. Contributions from cranial nerves VII and XI enter posterior to the plexus.

The palatine aponeurosis is the principal structural element within the velopharynx. In provides an anchoring point for muscles, adding a degree of stiffness, and is continuous laterally around to the hamulus with the tensor

veli palatine muscle. The aponeurosis is diamond shaped. More posterolaterally, the salpingopalatine ligament, the fascia of Tröltsch, and the internal fascia of the phanrynx (which all form the membranous portion of the Eustachian tube) contribute to the velopharynx.

The normal structure and function of the soft palate is dependent on the levator sling. This structure comprises portions of the tensor veli palatine, palatoglossal, palatopharyngeal, and uvular muscles. Functionally, the levator veli palatine, palatoglossus, and musculus uvulae muscles either elevate the soft palate or alter its shape. Other muscles, such as the superior constrictor, palatopharyngeus, palatothyroideus, and salpingipharybgeus muscles, are involved with movements of the lateral and posterior pharyngeal walls. The tensor veli palatine is involved mainly with middle ear aeration. In patiens with cleft palate, the muscle attachments are directed anteriorly and attach onto the posterior portion of the bony palate. These fibres must be surgically reoriented to achieve proper palatal function.

During velopharyngeal closure, contraction of the posterior velum touches the posterior pharynx.

3.2 Frequency. The etiology. The clinical picture cleft palate

Cleft palate refers to a gap in the upper lip or palate or in combination. Cleft palate can occur in isolation when the palatal shelves fail to fuse in the medial line or in combination with cleft lip.

Pare was the first to describe the use of the obturator for the perforation of the palate (1564). In 1552, Jacques Houllues proposed suturing the edges of the cleft together. In 1764, Le Monnier, a French dentist, made the first successful plastic surgery of the velum cleft. Dieffenbach closed the cleft on the soft and the hard palate in one session in 1834. Von Langenbeck first described the palate's mucoperiosteal flaps in 1861. In 1868, Billrot proposed to fracture the pterygoid apophysis in the plastic surgery of defects. Von Langenbeck’s procedure continued to be modified by Gillies, Fry, Wardill, Veau, Dorrance. In 1944, Schweckendiek proposed the plastic surgery of cleft in young children.

Frequency. Clefts palatal are the most common forms of congenital malformations.

The diagnosis of palatal clefts may be ascertained at the 17th Week of intrauterine development, by ultrasonographic methods. The incidence of palatal clefts, with or without lip clefts, is 1:1000 newborns. The incidence of cases varies by race. Among the children of American Indians, the number of children with clefts increases considerably (3.6: 1000 newborns). In Africa, their number decreases to 0.3: 1000. The labial clefts constitute 20 % (18 % unilateral, 2 % bilateral), those of the palate and upper lip – 50 %, (38 % - unilateral, 12 % - bilateral), palatal ones 3 0% of the total number of clefts. The incidence of palatal clefts is 1: 2000 newborns. The frequency is higher in girls, compared to boys (4: 1). In approximately 20-30 % of cases, palatal clefts are clinical signs in groups of children with different syndromes. The incidence of submucosal cleft is 1: 1200, 1: 2000 newborns, and the ratio girls: boys - 1:1.

In Moldova, palatal clefts occur in 34.4 % of cases, in all forms of lip-maxilla-palatal clefts. The number of children with palatal clefts increased with 0.16 per 1000 newborns (I. Lupan 2005).

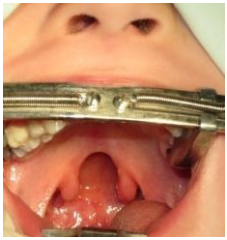
The etiology. The genetic origin of the clefts is heterogeneous and multifactorial. For all parents the risk of having children with clefts is 1 in 700. If the parents do not have first degree relatives affected, the rate of birth of children with clefts is 2.5 %, and if one of the first degree relatives has such defect, the risk of having children with clefts increases up to 10 %. If the parents are affected, the risk eise to 10-12 %. If the malformation is part of the autosomal dominant syndrome, the risk rise to 50 %. In 30 % of cases, the clefts can be associated with syndromes. More than 400 syndromes associated with clefts are described.

In general, the etiology of the clefts is considered unknown. At the same time, there are mentioned some factors with external ones, which can influence the development of the child during the gestation period of development. Alcohol and narcotic intake during the embryological period can serve like a teratogenic action. Mechanical action in embryological development can also influence the development of the child.

The clinical picture. Palatal clefts are observed immediately after the birth of the child, through functional disorders and aesthetic defects. Anatomy of the cleft palate involves disruption of the levator sling and palatal aponeurosis, and normal muscular insertions thereon. Muscles

such as the tensor and levator veli palatini, which would normally insert at the midline in a cleft, run longitudinally along the cleft margin before inserting into the posterior border of the hard palate. As a result, the normal sphincteric contraction of the soft palate and posterior pharynx is compromised. Functional disorders of the patients with the palatal cleft are very severe. They manifest themselves by passing the food from the oral cavity into the nasal cavity, causing asphyxiation, breathing disorder, difficulty in feeding. Incomplete closure of the soft palate results in air escape through the nose during the pronunciation produce hypernasal speech. Additionally, as the tensor veli palatini assists with Eustachian tube opening, aberrant function likely contributes to the otopathology seen in cleft patients, including recurrent otitis media. Because of the interruption of the anatomical continuity of the jaw bone and correspondingly the destruction of the growth areas development of the jaws is compromised. Over 275 syndromes have been identified in which clefting is a primary clinical feature. Palatal cleft in 30 % of cases may be associated with syndromes (Pierre-Robin syndrome). Incomplete clefts of the soft or submucosal palate may remain unobserved for a long time. Nasal reflux, nasal speech, are the clinical signs of these clefts.

Palatine submucosal clefts. In 1825, Roux described the submucous cleft palate. He presented three factors that characterize this cleft: the presence of a bluish line in the the soft palate, the shortening of the soft palate and the abnormal expansion of the nasopharyngeal region. Later, in 1956, Calnan added to these deformations the bifid uvula, the diastasis of the palatine muscles and the presence of depression on the hard palate. Bone defect may be detected by palpating the posterior edge of the hard palate. (*Fig. 14*)



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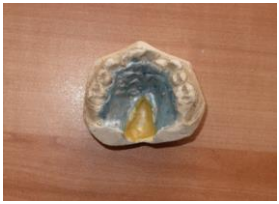

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Fig.14. **Palatal cleft. A. Intraoral view, b, c. cast model**

This type of cleft often is difficult to diagnose because the palatal mucosa maintains visible continuity, the underlying muscular anatomy of the submucous cleft is similar to that of the class. The diagnosis is made occasionally, during the prophylactic examination, sometimes after the adenotomies, once the nasal speech appears. A submucous cleft palate is a form of cleft palate that may appear to be structurally intact, but in fact there are both muscular and bony deficits. The defects include a bony notch in the posterior part of the hard palate, a bluish line at the midline of the soft palate (zona pellucida) indicating a separation of the submucosal palatal musculature, and a bifid uvula. The bony notch can be seen or felt where normally the posterior nasal spine is found along the posterior border of the hard palate. The submucous cleft of the hard palate is not functionally significant. However, the muscle disorder in the soft palate often is functionally significant. The levator muscles in these cases are often found to be inserted into the hard palate instead of midline to form the normal levator sling. This condition creates further complexity, which may contribute to velopharyngeal incomepetence and in consequences nasonate speech.

Cleft of the secondary palate. There is several type of the cleft of the secondary palate based on anatomical position (cleft of the soft palate, cleft of the soft and hard palate) and usually they are located on the midline and simmatrically. Those clefts extend up to the incisive foramen are incomple, and those extend futher than incisive foramen are complete. Thus in clefts of secondary palate, oral cavity communicates with the nasal cavity. The vomer, usually small, suspends in the oral cavity on the midline, between the free palatal shelves, which are hypoplastic and verticalized. The soft palate is short and the nasal and oral pharynx wide. The soft palate muscles are inserted at the posterior edge of the palatal plates, acting as antagonists, which is why, during the speech and swallowing, the defect increases. In these clefts the integrity of the alveolar processess is preserved. As the child grows up, growth disorders of the jaws may occur. Anatomical disorders result in

functional dificalty associated with feeding difficulties, speech and language development, increased risk of middle ear effusions and subsequent infections. grat risk for developing malocclusion.

Unilateral complete cleft lip and palate affect the structures of the soft and bony parts of the primary and secondary palate, manifested by interrupting the continuity of the upper lip, alveolar processes, hard palate and soft palate. The cleft is asymmetrical divided into two pieces: the greater alveolar segment (including the premaxilla) and the lesser alveolar segmen (on the cleft side).

The typical unilateral complete cleft lip deformity results from both a deficiency and a displacement of the soft tissues, the underlying bony structures, and cartilaginous structures. An imbalance of the normal muscular forces acting upon the maxilla results in an outward rotation of the premaxillary-bearing medial segment and posterolateral displacement of the smaller lateral segment. The jaw on the affected side is hypoplastic. At the level of the cleft, there are often supernumerary incisives. The inferior edge of the anterior nasal septum is displaced out of the vomerine groove into the noncleft nostril, and the anterior septum leans laterally over the cleft. The overlying columella invariably is short on the cleft side and distorted by the displaced caudal septum. In the nasal tip, the alar cartilage is characteristically deformed, and the medial crus are displaced posteriorly. The dome is separated from that of the noncleft side, and the lateral crus is flattened and stretched across the cleft. The axis of the nostril on the cleft side is characteristically oriented in the horizontal plane. This position is in contrast to the normal vertical axis of the nostril on the opposite side.

The muscular fibers of the orbicularis oris do not decussate transversely as in the normal lip; rather, they course obliquely upward, paralleling the cleft margin toward the alar base on the lateral side of the cleft and toward the base of the columella medially. The philtrum on the cleft side is short, and the presumptive Cupid's bow peak is displaced superiorly. The vermilion is deficient on the cleft side of the medial element.

The orbicularis oris muscle is the primary muscle of the lip and can be divided functionally and anatomically into 2 parts. The deep compo-

nent, in concert with other oropharyngeal muscles, works in swallowing and serves as a sphincter. The superficial component is a muscle of facial expression and inserts into the anterior nasal spine, sill, alar base, and skin to form the philtral ridges. In a complete cleft, the deep fibers of the orbicularis oris muscle are interrupted by the cleft and end on either side of the defect instead of making their way around the mouth. In addition, the superficial component of the orbicularis oris turns upward, along the margins of the cleft and ends beneath the ala or columella. Incomplete cleft lip behaves in a similar manner, except when the cleft is less than two thirds of the height of the lip. In this case, the fibers of the muscle run along the margins of the cleft, then change direction and run horizontally over the top of the cleft. These muscle fibers are interspersed with connective tissue.

In the intermaxillary relationships, laterognathia is determined on the affected side, and anteriorly, on the large fragment - a deep occlusion. The vomer merges with the palatine processes of the unaffected side, it bends towards the healthy side being skewed and forming the vomero-palatine suture. The soft palate is shortened, the pharynx enlarged. The velum muscles insert on the posterior edge of the palatal processes and sometimes extend to the middle edge, having an oblique direction of the fibers. The mucosa of the nasal cavity is swallowed, with hyperemia and hypertrophied nasal cornices. The pharynx is filled with 3rd degree adenoid vegetation, the hypertrophied tonsils.

Complete bilateral clefts result from failure of the premaxillary segment to fuse with the lateral maxillary segment on the right and left side. The defect is located on the upper lip, alveolar processess, soft and hard palate, bilaterally of the vomer. This cleft distinguishes three fragments - premaxilla and two maxillary. Acording to the anatomic characteristicsthe cleft is symmetrical, altghou they are determined by the degree of completeness of the cleft and its symmetry.

The size and position of the premaxilla vary and effectively can be excluded with a collapse of the alveolar arch. Subsequent forward growth of the premaxilla, attached only to the vomer above, leads to its projection beyond the lateral segments. Within the isolated prolabium,

the skin is foreshortened vertically, the white roll is underdeveloped, and the vermilion is deficient. The prolabium lacks muscle fibers, and the philtral ridges, the central philtral dimple, and Cupid's bow are absent. Although the prolabium varies in size, it is usually retracted and lacks muscle fibers. The bilateral cleft nasal deformity is characterized by flaring of the alar bases and wide separation of the domal segments of the alar cartilages. The columella is markedly shortened, causing the nasal tip to be depressed. In addition, the columella is absent and the prolabium appears attached to the top of the nose in some cases. The extent of nasal deformity associated with cleft lip varies from patient to patient. The jaws in relation to the mandible are hypoplastic. The premaxilla contains the upper incisors The dental arch is narrow, of conical shape, and because of this, there is a bilateral laterognathia in the intermaxillary relations, accentuated in the canines, with the deep occlusion (sometimes open) at the premaxilla level. The palatine processes of the jaws do not merges with the vomer, leaving it suspended freely in the oral cavity. Most often, it is thickened and projects below the palatal plates, sometimes with hypoplasia. Oral cavity communicates with nasal cavity through bilaterally gaps between vomer and palatal shelves. The intermaxillary relations in the lateral parts are reversed, and the occlusion is deep in the anterior part.

The blood vessels parallel the course of the muscle fibers and run along the margins of the cleft toward the columella or alar base, where they form anastomoses with nearby vessels.

3.3 Functional disorders in complete uni and bilaterally cleft

Palatal clefts manifest functional disorders, including feeding problems, airway obstruction, and otitis media, thus children with palatal cleft require medical management prior to repair.

Feeding problems. Feeding a baby with a cleft lip and palate is a challenge for parants. A team of professionals is available to help the family meet this challenge by providing information regarding feeding and nutritional needs.

A speech/language pathologist who specializes in feeding and swallowing disorders can provide the family with information regarding the most appropriate feeding position and equipment to use to make the

feeding as normal as possible. An evaluation of the patient's swallowing skills for signs of aspiration or dysphagia is an important part of this assessment. A nutritionist can help establish a feeding regimen that provides appropriate energy intake for optimal nutrition and growth. In general, a newborn needs 100-150 mL of breast milk or formula per kilogram of body weight per day.

Craniofacial orthodontist who specializes in anatomical and neuro-reflection theory of development of children with facial clefts, can construct an appliance to assist with feeding for infants who cannot tolerate prefabricated feeders. Since no separation exists between the oral and nasal cavities, children with a cleft palate (with or without a cleft lip) have difficulty obtaining adequate intraoral pressure for sucking and extracting liquid from the nipple. This can cause the baby to tire easily and to be unwilling or unable to suck long enough to obtain enough milk. In addition, food or liquid may back up and run out of the baby's nose and cause choking, coughing, or spitting up.

Most babies with a *cleft palate are fed with a bottle*, although breastfeeding is not possible. Only in some cases (lip and alveolar ridge and not the palate), breastfeeding may be attempted only. A baby with a cleft palate is not likely to obtain adequate suction to extract the milk because there is no achievable lip seal. There are several modifications to help improve oral feeding. These modifications include special cleft feeding equipment, properly positioning the infant during feeding, adjusting the placement of the nipple, supporting the cheek, and altering
the rate of feeding.

With bottle-feeding, the baby with a cleft palate typically feeds slower and needs help regulating the flow of liquid. Various bottles and nipples can be used to assist with feeding. A cleft feeding nipple must have a large enough opening to allow the formula to flow easily to prevent sucking fatigue, but it must not be so large as to cause choking. Nipples should be soft and compressible, allowing liquid to flow easily. Soft nipples designed for premature infants used with a regular bottle often work well. Occasionally, the hole in the nipple designed for a premature neonate may need to be enlarged to increase the flow of milk. This is best performed by creating an X-shaped opening to help regulate

the flow of formula. Enlarging the opening too much may result in the free flow of milk, which can cause the baby to choke.

With regard to positioning, a semiupright position, as upright as possible, is best. This position helps prevent food and liquid from entering the nasal cavity. Upright positioning may also decrease eustachian tube reflux, which may lead to middle ear inflammation (otitis media). Eustachian tube reflux can also cause otorrhea in children with ear grommets.

Placement of the nipple within the oral cavity is important. Optimally, the nipple should be positioned to the back and along the side of the mouth on the noncleft side (in the case of unilateral cleft palate). Supporting the cheek by gently squeezing the cheeks together around the nipple may also improve oral suction.

Mothers who attempt breastfeeding must be aware of the signs of dehydration and the need to seek medical attention at the first sign of problems. Signs of dehydration in the baby include sleepiness and listlessness, urination fewer than 10 times per 24 hours, and urine that is strong smelling and/or dark and concentrated. In addition, the mother should carefully monitor the baby's weight and take into account the baby's frustration with feeding, sucking fatigue, and signs of hunger when deciding to continue or discontinue breastfeeding. Frequent burping is important because babies with clefts tend to take in a lot of air with sucking. The gastroesophageal reflux may also be increased because of excessive air intake. Regular burping during feeding may help minimize spitting up. It may be beneficial to feed the baby smaller meals and to increase the number of feedings throughout the day, especially if the baby fatigues quickly with sucking. A nutritionist should carefully monitor these changes to ensure adequate energy intake for optimal growth. Ensuring that the mouth and lip area are clean after feeding and prior to placing the baby in a reclined position is important to prevent choking.

Spoon-feeding and feeding of textured and table foods usually evolves in the same quantities and developmental sequence as with any baby or child, even if the cleft palate remains open. Things to consider

with spoon-feeding include presenting the food slowly, allowing the baby to remove food from the spoon by using its lips, and allowing the baby to regulate the timing of the next mouthful. For textured and table foods, the feeder should continue with the slow rate of presentation and provide sauce or gravy with small textured foods, such as rice, that tend to spread throughout the mouth. Adding sauce or gravy helps the pieces of food stick together as they are transported through the oral cavity. Most babies and children with a cleft lip or palate learn to eat orally, with modifications. They become adept at moving the bolus through the oral cavity around the cleft. If food falls from the nose or gets stuck in the palate, the feeder should not become alarmed because the food does not interfere with breathing or cause harm. Occasionally, the child may sneeze when food enters the nasal cavity. Food can be removed with a finger or a cotton swab without frightening the child. If the patient with cleft palate continues to have feeding difficulties even with the appropriate modifications, further consultation may be necessary to rule out such problems as dysphagia or sensory integration difficulties. If the lack of weight gain due to feeding difficulties is a problem, use of a feeding tube should be considered. If problems with weight gain do not respond to feeding therapy, a gastrostomy may be necessary.

Airway obstruction. Airway obstruction may present in children with a cleft palate, especially those with mandibular hypoplasia (ie, a Pierre Robin sequence). Upper airway obstruction results from posterior positioning of the tongue, which is prone to prolapse into the pharynx with inspiration. Nasal obstruction can also result from the tongue protruding into the nasal cavity. Airway obstruction is usually managed by placing the child in a prone position to prevent prolapse of the tongue. In severe cases in which the obstructed airway is not relieved with conservative measures, a tracheotomy may be necessary. In these instances, such measures as a lip-tongue adhesion are generally not as effective and not as well tolerated as a tracheotomy.

Otitis media. Otitis media is a common complication of a cleft palate and is present in nearly all children with unrepaired clefts. Although recurrent suppurative disease can be a problem, the primary complication is that of persistent middle ear effusion with resultant

hearing loss. Medical management for this problem typically involves careful observation, which must be performed in light of the potential complications of prolonged hearing loss, especially in a child at risk for speech problems due to a cleft palate. In most instances, grommet insertion for middle ear ventilation is the preferred treatment to avert potential speech problems due to conductive hearing loss.

3.4 Treatment and profilaxy of complications in the complete cleft lip and palate

Children with palatal clefts require *complex, multidisciplinary treatment* in all periods of development - from newborn to adolescent. The objectives of the treatment are the restoration of the physiognomic-aesthetic aspect, the harmonious development of the teeth and jaws, the achievement of a satisfactory phonation, the harmonious development of the organs and systems. For this purpose, the children need a special monitoring, in special centers, by the pediatrician, psychiatrist, plastic surgeon, otolaryngologist, craniofacial orthodontist, speech therapist. The treatment of clefts starts right from the first days after the child is born. An important role in the rehabilitation of these children is the family, overwhelmed by negative emotions, anxiety, disappointment and feelings of guilt. Individual professional medical care is performed by a nurse or a team of doctors (pediatric speech therapist, craniofacial orthodontist), which will contribute to the psychological recovery of the family and to the difficulties of caring for the child in the first days.

The complex rehabilitation comprises two stages: early and late. Early stage: pre-surgical orthopedic treatment; the primary plastic surgery of the upper lip and the soft palate; speech-language rehabilitation; early orthopedic treatment; alveolar bone grafts; orthodontic treatment in permanent dentition. Late stage: secondary plastic surgery of the nose, lip and septum (cheilorinoseptoplasty); the treatment of vleopharyngeal incompetence; orthognathic surgery; prosthetic therapy. (*Fig.15*)

The preoperative orthopedic treatment covers the age period of 1-4 weeks and aims at maintaining and/or repositioning preoperatively the alveolar fragments, separating the oral cavity from the nasal one.
General agreement exists that surgical correction of a cleft palate should be accomplished when patients are younger than 1 year, before

Fig. 15. **Rehabilitation part of treatment. (orthopedic, dentistry, speech therapeutic)**

significant speech development occurs. The potential benefits of an intact velum as a child begins to speak are believed to outweigh the possible complications of early closure, namely later collapse of the maxillary arch with a resultant crossbite.

How closure is accomplished is subject to some variation. Generally, 1-stage closure of the soft palate and/or the hard palate can be accomplished when the patient is aged 11-12 months. However, some advocate a 2-stage closure, with repair of the velum (soft palate) when the patient is aged 3-4 months. This procedure results in narrowing of the hard palate cleft, facilitating closure at a later date, usually when the patient is aged 18 months. Similar to a lip adhesion for a wide cleft lip, a 2-stage approach may be useful when the cleft palate is particularly wide.

When cleft palate repair is deferred to later childhood or adulthood, repair often involves a pharyngeal flap. Incorporating a pharyngeal flap

into the repair can help close a large defect and compensate for velopharyngeal dysfunction and speech problems.

The optimal age for a palatorrhaphy in one session is considered to be 18-20 months. At the same time, there are persons advocating for the primary palateclosure in two stages. The age for the primary surgical closure of the defect on the soft palate is about 12 months. The first stage creates favorable conditions for the hard palate surgery closure, conditioned by its gradual narrowing. Uranoplasty is performed at age of 18-20 months.

If cleft lip is present, its repair can precede palatoplasty. Although early repair seems to have an advantage in decreasing the chances of speech delays, the risk for facial growth abnormalities and other midface-related problems may be increased. In the past, several criteria have been listed for patients undergoing any of the procedures. Some of these criteria include a hemoglobin level higher than 10 g/dL, weight gain, the absence of infection, and a full preoperative evaluation by a pediatrician.

Preoperative details. Repair of the hard palate is not always possible when the soft palate is repaired, especially with wide bilateral clefts. The cleft size can decrease as much as 7 % with growth in patients aged 3-17 months. The size can be further reduced with early repair of the soft palate (in patients aged 3-4 months) followed by closure of the hard palate in patients aged nearly 18 months. This fact should be taken into consideration in planning the time and the type of the repair. The defect is usually smaller than it was originally when closure is performed after the soft palate defect has completely healed. The procedure can be performed in patients aged as young as 3 months, with a second procedure for closure of the defect when they are 6-12 months.

A great deal of debate exists regarding the timing of the repair. In the past, many surgeons believed that hard palate repair should be delayed until after eruption of the molar teeth. Currently, most centers focus on completion of the cleft palate before the patient is 12 months. Debate had existed over whether or not delayed closure of the hard palate was beneficial or harmful to facial growth, but the evidence for either side has not been conclusive.

The purpose of the surgical treatment in patients with cleft palate is to separate the nasal cavity from the oral cavity, to stop the air and liquid reflux from the oral cavity into the nasal cavity, to create the conditions necessary for the development of an adequate speech. Removing the defect also contributes to the harmonious growth of the facial skeleton and dentition, and conformity of the dorsal surface to the pharyngeal wall. Three important factors are required for the function of the soft palate in restoring speech: mobility, length, pharyngoconstriction. For this purpose, it is determined: the degree of shortening of the soft palate and the index of the cleft.

The degree of shortening of the soft palate is a metric method of determining the velar insufficiency. It is determined on the basis of distance measurements: 1) from the alveolar apophysis to the tip of the uvula; 2) from the alveolar processes to the posterior wall of the pharynx. The difference between these variables determines the degree of shortening of the soft palate.

The cleft index determines the degree of defect in the horizontal plane and the complication of the surgery, based on measurements according to the formula A1 + A2: A = P. (A1 - the width of the palatine flap on the left, at the level of line A; A - the border between the soft and the hard palate; A2 - the width of the palatine flap on the right, at the border between the soft palate and the hard palate). The palatal cleft is considered uncomplicated in the surgical plane, when P = 1.6 mm.

Intraoperative details. Investigators in a multicenter study involving surveys of more than 300 surgical teams attempted to establish the common ground for repairs of cleft defects. Although no single technique was used universally, a trend has been established toward the use of earlier palate closure over the last several years. Of the closure techniques surveyed, the Furlow procedure was the most common technique for cleft palate closure. The basic surgical techniques included the following: von Langenbeck, Schweckendiek, 2-flap, 3-flap (V-to-Y), and double reverse z-plasty (Furlow) palatoplasties. Although most of the repairs do not involve repairing the muscular sling, doing so allows

better palatal and eustachian tube functions. Descriptions of the major techniques used for palatoplasty are outlined below.

The von Langenbeck technique. First described in 1861, the von Langenbeck technique underscores the importance of separating the oral and nasal cavities. Virtually every repair performed today incorporates principles initially included in this technique. Bipedicle mucoperiosteal flaps of both the hard palate and the soft palate are used to repair the defect. After their elevation, the flaps are advanced medially to close the palatal cleft. Advantages of this technique include less dissection and its simplicity. A disadvantage of the von Langenbeck repair is that it does not increase the length of the palate, which results in an inability to close primary and secondary clefts. Other criticisms of this technique include the occurrence of anterior fistulas and the resultant inferior speech due to the short soft palate. Airway obstruction during sleep seems to be an insignificant problem with this repair. Because of the physical limitations in lengthening the palate with this technique, many modifications have been made over the years.

Schweckendiek technique. In the 1950, Schweckendiek began to repair clefts in a staged fashion. In this technique, the soft palate is first repaired when the patient is young (typically 3-4 months), and this is followed with hard palate closure when they are nearly 18 months. During the first and second closure of palate, an obturator is used to allow swallowing and speech. This technique has the advantages of achieving closure when the patients are young and causing minimal disturbance of facial growth. However, the disadvantages include the need for additional operations; the need for frequent changing of the dental prosthesis.

The initial repair is usually performed in a patient aged 3-12 months. The second stage is usually performed when the patient is 18 months, but it may be delayed until the patient is 4-5 years. Longer delays (ie, until primary dentition is established) were believed to be advantageous in that they prevented lateral contraction of the palatal arch. Currently, speech and feeding difficulties with delayed closure are thought to outweigh the dental alignment problems, and the current trend is to use earlier closure. Collapse of the maxillary arch is now dealt with

by means of palatal expansion when the patient is young. The initial repair is accomplished by making incisions in the soft palate along the margins of the cleft. The levator muscle, which is abnormally attached to the posterior free edge of the bony palate, is dissected free and reoriented. A 3-layer closure of the nasal mucosa, the levator muscles, and the oral mucosa is then performed. The resultant hard palate fistula is closed at a later date.

Although many methods to close the hard palate exist, one technique is the use of the vomer flap. The mucoperiosteum of the vomer bone is elevated in an inferior-to-superior direction. This flap is then rotated laterally for attachment to a small palatal mucoperiosteal flap. This procedure can provide a watertight closure with minimal elevation of the palatal mucoperiosteum. The preferred method involves raising the mucoperiosteal flaps on the oral and nasal surfaces of the hard palate and closing them in 2 layers across the defect. The vomer flap is primarily useful with wide or bilateral clefts. Vomer flaps have the disadvantage of requiring closure of 2 suture lines on the nasal surface. When used with oral mucoperiosteal flaps, the vomer flaps are attached to the flaps raised from the nasal surface of the cleft.

Two-flap technique (Wardill-Kilner-Veau). This method is the most common technique used for closing complete clefts. Limberg A.A. (1926), Kilner and Wardill (1937), independently described the V-Y repositioning technique. This technique is primarily used for repair of incomplete clefts or clefts of the secondary palate. The incisive foramen is the anterior border of the repair, and the uvula is completely divided posteriorly. The theoretical advantage of this technique is that pushing back the flaps adds length to the palate and reduces closure tension at the junction of the hard and soft palates, helping to prevent fistula formation.

The Limberg A.A. procedure (1926) includes 5 stages of palace plastic surgery.

1. Incisions are made along the free margins of the cleft and extended anteriorly from the apex of the cleft to where the canine teeth erupt. Dissection is then continued posteriorly along the oral side of the alveolar ridge to the retromolar trigone. Mucoperiosteal

flaps are elevated from the nasal and oral surfaces of the bony palate. The soft palate muscles are released from the posterior regions of the palatal processess and move in the medial and posterior direction.

2. Dissection of the greater palatine vessels from the foramen is performed by Interlaminar of the sphenoid bone osteotomy in the posterior margin of the hard palatine. The tensor veli palatini muscle is elevated off from the hamulus. In this way the sufficient mobility of the flaps in the sagittal and horizontal plane is obtained.
3. Infracture of the hamulus or stripping the levator veli palatini muscle from the hamulus can be made; these changes greatly improve medial rotation of the mucoperiosteal flaps. Once the nasal mucosa is freed, the palate can be closed in layers: the nasal and oral layers anteriorly and the nasal, muscular, and oral layers posteriorly.
4. Mesopharyngoconstriction is achieved by releasing the lifting muscles of the palatine veil from the pterigomandibular lining.
5. Staphylography. Once the nasal mucosa is released from the nasal surface of the hard palate, the suture is placed anteriorly in two planes and posteriorly in three planes. Advantages of this procedure: reduction of posterior oro-nasal fistulas, sufficient elongation of the palatine veil, reduction of tension at

the junction of the palate.

Double reverse z-plasty. In 1986, Furlow described a technique to lengthen the velum and to create a functioning levator muscle sling. This method is difficult to perform in wide clefts. However, it is considered a good method when the cleft is narrow or if a submucous cleft exists. The technique involves opposing z-plasties of the mucosa and the musculature of the soft palate. The goal is to separate the nonfunctioning attachments to the posterior border of the hard palate and to displace the mucosa and the musculature posteriorly.

The first z-plasty is created on the oral mucosa side, while the second z-plasty is inverted on the nasal mucosa side. The incisions are

made, and the oral mucosa is dissected free from the underlying muscle. On the left side of the patient, the oral mucosa flap also contains the muscle. On the patient's right side, the muscle is kept with the underlying nasal mucosa. The 2 muscle-bearing flaps transpose posteriorly, while the thin nonmuscular flaps are placed anteriorly. This technique has the effect of rotating the muscular sling posteriorly and lengthening the soft palate. One potential problem with this technique is the formation of a fistula at the junction of the hard and soft palates.

Alveolar bone grafting. Alveolar bone grafting is an integral part of repairing clefts that involve the anterior maxilla. Establishing a bony union can help to prevent maxillary segmental collapse, to close oronasal fistulas, and to encourage eruption of teeth. Regardless of whether the repair is early or late, the neonate should be fitted with an obturator within the first month after birth. Bone grafting in patients younger than 2 years is considered primary, and secondary grafting occurs afterward. Graft material can be obtained from the hip, the ribs, the extremities, or the outer table of the skull. Although morbidity can exist at the various donor sites, the benefit of closing the maxillary gap outweighs the potential risk.

The surgical procedure involves raising mucosal pedicles on either side of the maxillary defect. With the use of any of the described donor sites, the graft of cancellous bone is placed into the pocket. The mucosal flaps are closed in a simple fashion. Many times, the depression of the alar base is immediately corrected on completion of the procedure.

Postoperative details. Immediate postoperative concerns in cleft palate repair include airway management and analgesia. Repairing the palate changes the nasal/oral airway dynamics and may present problems in the immediate postoperative period, especially in children with a Pierre Robin sequence. The lasting effect of narcotics used for anesthesia may also alter upper airway dynamics. Since placement of an oral airway may disrupt the palate repair, a ligature of 2-0 chromic (or silk) suture is placed through the anterior tongue to allow forward traction on the tongue while the patient is in the postanesthesia area. This suture is removed once the child is fully alert and able to maintain the upper airway. Adequate analgesia is important in the postoperative period to allow patients to return to their activities as

quickly as possible. However, the use of analgesics must be balanced with the risks of oversedation and subsequent airway compromise. Generally, acetaminophen with codeine is sufficient for this purpose. Analgesics may be continued as needed for as long as 7-10 days postoperatively with few problems; the most common adverse effect is constipation. In infants and younger children, arm restraints or „no-no's" are used when the child is unattended to prevent the placement of fingers in the mouth because this may disrupt the repair.

Diet in the postoperative period is generally limited to liquids and soft foods that do not require chewing. The use of bottles is avoided because the nipples may interfere with the repair. The use of spoons is also avoided for similar reasons. Feeding is accomplished by using either a cup (not a sipping cup) or a Breck feeder (a red rubber catheter attached to a syringe). Normal diet and feeding may be resumed after 10-14 days, depending on the type of repair. At 3 weeks, all dietary and feeding restrictions are removed.

Oral hygiene is best performed by rinsing with clean water, with the patient taking care to remove all collected food particles. The use of hydrogen peroxide should be avoided because it may inhibit healing. After 5-7 days, careful toothbrushing may be resumed.

Follow-up. Once discharged from the hospital, the patient should have follow-up visits at 7-10 days and at 3 weeks. If a small fistula or a wound breakdown is noted in this period, waiting at least 6 months prior to attempting closure is advised. This delay allows for maximal wound contracture and for reestablishment of the blood supply to the tissues.

3.5. Complications

Airway obstruction. As mentioned previously, postoperative airway obstruction is the most important complication in the immediate postoperative period. This situation commonly results from prolapse of the tongue into the oropharynx while the patient remains sedated from anesthetics. Intraoperative placement of a tongue traction suture helps in the management of this situation. Airway obstruction can also be a protracted problem because of changes in airway dynamics, especially those in

children with a small mandible. In some instances, placement and maintenance of a tracheotomy is necessary until palate repair is complete.

Bleeding. Intraoperative hemorrhage is a potential complication. Because of the rich blood supply to the palate, significant bleeding requiring transfusion can occur. This can be dangerous in infants, in whom total blood volume is low. Preoperative assessment of the hemoglobin level and the platelet count is important. Injection of epinephrine prior to palate incision and intraoperative use of oxymetazoline hydrochloridesoaked packing material can reduce blood loss. To prevent postoperative blood loss, demucosalized areas of the palate should be packed with Avitene or a similar hemostatic agent.

Oro-nasal (*palatal) fistula.* Wound dehiscence (palatal fistula) can occur as a complication in the immediate postoperative period, or it can be a delayed problem. A palatal fistula can occur anywhere along the original cleft site. The incidence has been reported to be as high as 34 %, and the severity of the original cleft has been shown to correlate with the risk of fistula occurrence. Complete dehiscence is uncommon, but immediate reclosure should be attempted if it does occur. Small fistulas that occur at areas of maximal wound tension are more common. These typically occur at the junction of the primary and secondary palates anteriorly or at the junction of the hard and soft palates posteriorly.

Postoperative cleft palate fistulas can be managed in 2 ways. In a patient without any symptoms, a dental prosthesis can be used to close the defect with good results. A patient with symptoms may require surgery. Poor blood supply, especially the anterior supply, is the major reason for failure of fistula closure. Therefore, closure of persistent anterior or posterior fistulas should be attempted no sooner than 6-12 months after surgery, when the blood supply has had an opportunity to reestablish itself. Currently, many centers wait until the patient is older (at least 10 y) before attempting fistula repair. If simple closure methods fail, vascularized tissue flaps, such as an anterior tongue flap, may be required for closure.

Speech therapy. Nasonate phonation is the most common complication, which occurs after primary surgery. This aspect occurs when the

soft palate cannot contact the posterior wall of the pharynx, because of the motility restrictions, and in the presence of oro-nasal fistulas. To improve and eliminate the nasonate speech respiratory gymnastics, massage of mucseles, myotherapy, speech-language exercises are indicated constantly before school age

Midface abnormalities. Maxillary growth restriction in patients with cleft palate is a common complication. Early primary closure may lead to midface hypoplasia and deformity, malocclusion, decreased anterior or posterior dimension of the upper jaw, a narrower dental arch, or an abnormal height. LeFort I osteotomies is indicated for reconstruction of midface hypoplasia. Local factors specific to patients with clefts, which limit the orthodontic possibilities, are: 1) previous surgeries, which produce retractable palatal scars; 2) there is no palatal suture on the midline, but a fibrous scar that greatly reduces the possibilities of orthodontic expansion; 3) for the correction of a posterior crossbite occlusion, the lateral expansion of the dento-alveolar is limited by the postoperative scars.

Secondary plastic surgery procedures.

A. Secondary plasticity of the nose and lip. Even if the most rigorous technical procedures will be followed, after the restoration of the lip and the palate on the part with the cleft, there is a distortion of the nose, the tip of the nose and the nasal septum. Around the age of 5-6 years it is possible to intervene at the level of the vicious scars of the lip. Cheilorinoseptoplasty, usually, is performed at age of 15-18 years, when the growth period ends.

B. Velo-pharyngeal incompetence, manifested at shortening of the palatine veil, which is the cause of the loss of air column through the nose. 3 degrees of vein-pharyngeal incompetence are known:

Degree I. Horizontally, during the phonation, the right and left sides of the soft palate merge, and the width of the split at the border level between the soft palate and the hard palate does not exceed 5 mm. In the sagittal plane, during the phonation, the posterior edge of the soft palate joins the posterior wall of the pharynx, and at rest it does not exceed 5 mm.

Degree II. Horizontally, the width of the cleft at the border between the soft and hard palate is 5–10 mm. At the moment of phonation, the right and

left sides of the soft palate do not join, and at rest, the distance between the posterior edge of the soft palate and the wall of the pharynx is up to 5 mm. At the time of the phonation, the distance narrows but does not disappear.

Degree III. Horizontally, the cleft width exceeds 10 mm. At the moment of phonation, the distance between the left and the right side of the soft palate is more than 5 mm. In the sagittal plane, the fissure between the posterior edge of the soft palate and the wall of the pharynx exceeds 10 mm, and during the phonation - 5 mm.

The surgical techniques used to improve the velo-pharyngeal incompetence are: velopharyngoplasty (medial pharyngeal flap with inferior or superior pedicle), pharyngoplasty and implants in the pharyngeal posterior wall.

C. Orthopedic surgery is usually performed during the age period when the jaw growth is completed (17-19 years). It is indicated, in the case of the large discrepancies in the mandible-maxillary bone relations, which cannot be corrected orthodontically.

Surgical displacements of the jaws can be performed sagittally, horizontally or vertically. But a number of factors may be restrictive for orthognathic surgery. First, the postoperative scars prevent the jaws from advancing and reduce post-surgical vascularization and bone healing. Secondly, the phonetic adaptation disorders of patients with clefts greatly limit the degree of maxillary advancement.

D. Prosthetic treatment aims to correct some dental abnormalities or edentation therapy after the growth period. In cases with bilateral clefts, the premaxilla remains mobile; this can be immobilized by fixed prosthetic works.

Patients with hypoplastic jaw, which need to improve the occlusion and the physiognomic appearance, are applied prosthetic devices, over existing teeth.

3.6. Follow-up and principles of rehabilitation of children with cleft lip and palate

Rehabilitation of children with labial-palate clefts is a long-term process, which requires the surveillance and treatment of several specialists. The dispensary center is endowed with a technical-material base and

includes complex rehabilitation of patients with cleft labial-palate, socio-pedagogical rehabilitation and medical-genetic consultation.

The technical-material base has a specialized department of oral and maxillofacial surgery, a dental clinic with profiles of therapy, orthodontics and orthopedics, an advisory department in the field of ophthalmology, speech therapy, otolaryngology, audiology, cardiology, psychology and general therapy (pediatrician); specialized preschool and school institutions.

The *pediatrician's* surveillance begins in the newborn period and continues until adolescence. Dystrophies, bronchopneumonia, heart disease often affect children with cleft palates, which is why they require surveillance and treatment. The role of the pediatrician in the complex treatment of children with clefts is to provide a complex nutrition, diet, physical and psychological development of the child, the condition of the respiratory and cardiac systems. The active period of surveillance includes the postnatal, preoperative and postoperative periods.

Otorhinolaryngologist. 60 % of the children with lip and palate cleft concomitantly suffer from rhinitis, otitis, tonsillitis, adenoid vegetation, hearing loss. The otolaryngologist provides conservative and surgical treatment of these organs, in order to improve the conditions of the plastic surgery of the basic disease. The active observation periods are pre-school and pre-operative.

Speech patology. Children with lips and palates cleft have defective, intelligible speech, they do not pronounce the sounds F, V, L, T, D, C, H, Ţ. Active periods of surveillance: a) 1–2 years, when practicing respiratory gymnastics, myomasages and myogymnastics of the veil muscles, spelling sounds and words; b) 4-5 years: articulation and speech exercises; c) 7-13 years - the most active period of myogymnastics and myomasages on soft cheek, speech-language exercises.

Psychoneurologist. 5 % of children with cleft lip and palate suffer from psychological retardation. Medical-pedagogical rehabilitation actions, medication correction therapy, psychotherapy are required. These patients need to be follow up for a long time by a neurologist.

In order to recover development of the craniofacial and oral maxillo area in patients with cleft lip and palate follow up require by the *oral and maxillofacial surgeon.* A special examination need to be done at each periods of development: newborn, 6 months –1,5 years, 5-7 years, 9-12 years, 16-18 years.

The craniofacial orthodontist. 85 % of lip and palate clefts are accompanied by tooth-maxillary deformations. The goal of orthopedic recover before and after the surgery to ensure the growth and development of the jaw according to the occlusion standarts. The aims of treatment during the preoperative period is to contribute for isolating the oral cavity from the nasal one and adjusting the fragments of the fragments. Postoperatively, to create conditions for development of dentoalveolar system and make a propfilaxy of facial deformities. Follow up period include: newborn, preoperative and postoperative, until adolescence.

The peadiatric dentistry. Damage of the teeth and periodontal tissues are present in all children with congenital malformations. Children with cleft lip and palate require dental prophilaxy and treatment during the preoperative and postoperative periods and prophylaxis of periodontal disease. According to the degree of healthy group each patient need to be examed not less than twice a year.

3.7 The complex rehabilitation of children with clefts lip/palate

In the first week of life of the newborn, the rehabilitation center is informed by the maternity ward and the surveillance begins. The problem of the newborn care is solved together with the pediatrician, craniofacial orthodontist, surgeon, psychiatrist etc. Separation of the oral cavity from the nasal cavity is performed in the first days of life by adapting the palatine mouthguards. The parents are trained to feed the child, its position, special feeding equipment (Haberman nipples). Initially the syringe or special sounds may be used for feeding.

The period of the newborn (the first month). The child is examined by the pediatrician, the oral and maxillofacial surgeon, pediatric dentistry. The degree of damage, the localization of the malformation and the presence of other defects of the organs and systems are determined. The cleaved stumps of the jaws are prepared by aligning them with palatal

devices (especially in cases with bilateral clefts), in order to improve the surgical conditions. Nasal retainers are indicated, in order to form the nasal shape during the preoperative period.

Age of 1–3 months: consultation of children by pediatrician, otolaryngologist, dental surgeon, pediatric dentistry; assessment of the general condition, the thymus, the otolaryngological organs, the effectiveness of the obturator, the situation of the bilateral premaxilla. The treatment plan includes cheiloplasty in unilateral clefts; unilateral or bilateral cheiloplasty of clefts; modeling of nasal retainers, in order to maintain the nasal relief and to maintain the alveolar apophysis stumps.

Age 3-6 months: daily consultation of the specialists of the medical consulting center; assessment of the general condition and physical development of the child by the medical committee; determining the effectiveness of nasal retainers, palatine mouthguard and correcting or performing a new obturator.

Age 6-12 months: daily consultation of specialists and anesthesiologist; assessment of the general condition of the child, the ORL organs, the thymus; examining the function of the velo-pharyngeal ring; examining the relations of the stumps of the palatal cleft and the premaxilla. At 12 months, velopharyngoplasty is performed (removal of the cleft in the soft palate and the uvula).

Age 1,5–2 years: repeated consultations of ORL specialists (chronic tonsillitis, adenoid vegetation), dentist, orthodontist, speech therapist. Attention is paid to the dental eruption, the position of the teeth, the degree of affectation of the dental caries, the character of the breathing and the speech, the degree of velar insufficiency. The surgical treatment during this period will be reduced to the second stage of plastic surgery of the hard palate. Orthodontic and speech therapy treatment is prolonged.

Age of 1,5–7 years. The children are examined by the center's specialists once a year. If necessary, they are sent to special pre-school institutions. Attention is paid to the dental status, the affectation degree of tooth caries, the harmonious development of the jaws, the formation of the occlusion, the character of the breath, the speech, the relief of the nasal wings, the degree of velar insufficiency. Correction operations are

performed – rhinoseptocheiloplasty, healing of the oral cavity, correction of orthodontic devices, respiratory gymnastics, myogymnastics and myomasages.

Age 8–16 years: consulting specialists once in 1-2 years or when needed; sending children to special school institutions; appreciation of the anatomical relief of the nose, lip and the character of the breath. Attention is paid to post-operative defects, secondary deformities of the palate, jaw development, intermaxillary relations, occlusion, eruption of dental teeth, condition of periodontal tissues, degree of velar insufficiency. It is performed surgeriesfor correcting the lip, nose, soft palate, hard palate, removing deformations and secondary defects; oral cavity healing, orthodontic treatment and speech therapy.

Age 17-18 years: consultation of the maxillofacial surgeon and orthodontist in dynamics; appreciation of the shape of the nose, lip, septum, remaining defects of the palate, condition of the dental-maxillary device. Surgical interventions are indicated, in order to remove secondary defects and deformities, the plastic surgery of the nose and lip relief.

Genetic medical consultation is a process in which patients and relatives of patients suspected of congenital malformations obtain information about the causes and consequences of congenital malformations, probability of transmission and methods of prophylaxis. The consultations can be organized in the prospective and retrospective period. In the first stage, the consultations have a prophylactic objective (in case there is a probability that a child with congenital malformations will be born). The consultations may be organized up to the stage of pregnancy being indicated in families with an unfavorable genetic history, in the case of kinship between partners; also, the action of the external and internal factors unfavorable for the mother and the fetus during the period of ovulation and active development of the facial skeleton.

Retrospective consultations are performed after the birth of the child with congenital malformations and with the objective of detecting the causes of this disease, i. e. it is a primary genetic mutation or it is caused by a camouflaged carrier. The consultation is done in three stages. The first stage starts with establishing the diagnosis. This process

is performed together with the pediatrician, dentist, neuropathologist, psychoneurologist, otolaryngologist, based on clinical, anthropometric, radiological, functional data, etc. of family and first degree relatives. The information is recorded in special files. In the second stage, the genetic analysis of the malformation is performed, the mode of transmission and the risk of its repetition. The forms of transmission of congenital malformations are: monogenic pathology - the analysis of the frequency of malformation among the relatives allows to determine the type of heredity; polygenic pathology - hereditary signs are the summary cause of several gene mutations; autosomal abnormalities; spora-dic cases of congenital anomalies, in which it is not possible to identify the relative cases. In the third stage, the risk of malformation in the family members is predicted. From 0 % to 10 % the risk of repetition is low; from 11 % to 20 % - medium; more 21 % - high.

Socio-pedagogical rehabilitation is necessary for children parallel to the aesthetic relief of the face, which has a great significance in integrating children with congenital malformations in society. At the initial stage, priority is given to preschool and speech therapy education. The socio-pedagogical rehabilitation is considered effective under the conditions of the specialized pre-school and school institutions, where the education is done in psychological conditions suitable for children with malformations.

Learning objectives

1. The stages and origin of the secondary palate during the intrauterine development period.
2. Classification of palatine clefts.
3. Incidence of palatine clefts.
4. Anatomical disorders in different forms of palatine clefts.
5. Functional disorders of the palatine clefts.
6. Diagnosis methods of palatine clefts.
7. Feeding of children with palatine clefts.
8. Determination of optimal indications for surgical restoration of soft and hard palates.
9. The operation of velouranoplasty. The objectives of the surgery.

10. The role of the pediatrician, ORL, dentist, orthodontist, psychologist in the complex treatment of these patients.
11. Monitoring of children with palatine clefts.

Tests

1. CS. Complex treatment of labial-maxilla-palatine clefts includes:
   A. Orthodontic treatment;
   B. Speech therapy;
   C. Genetic consultation;
   D. Surgical treatment;
   E. All answers are correct.

   (E)
2. CM. Functional disorders of the palatine clefts are:
   A. impossibility to breastfeed;
   B. shortening of palatine veil;
   C. nasal speech;
   D. open rhinolalia;
   E. frequent superficial breathing.

   (A, C, D, E)
3. MC. The purpose of the surgical treatment in patients with cleft palate is:
   A. to determine the degree of shortening of the soft palate;
   B. to separate the nasal cavity from the orqal cavity;
   C. to stop the air and liquid reflux from the oral cavity into the nasal cavity;
   D. to create the conditions necessary for the development of an adequate;
   E. to detrmine the index of the cleft.

   (B, C, D)
4. SC. The degree of shortening of the soft palate is:
   A. a method of detremining the mobility of the soft palate
   B. a metric method of determining the velar insufficiency
   C. a method to determine the length of the soft palate
   D. a method to determine the degree of defect in the horizontal plane

E. a method to determine the degree of the complication of the surgery

(B)

5. MC. The basic surgical techniques included the following:
   A. von Langenbeck technique
   B. flap technique
   C. 2-flap technique
   D. Double z-plasty
   E. Z – plasty

   (A, C)

6. SC. How many degrees of velo-pharyngeal incompetence are known:
   A. 3 degrees
   B. 2 degrees
   C. 4 degrees
   D. 5 degrees
   E. 6 degrees

   (A)

7. MC. The basic surgical techniques included the following:
   A. von Langenbeck technique
   B. flap technique
   C. 2-flap technique
   D. double z-plasty
   E. Z – plasty

   (A, C)

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4. General peculiarities of tumor lesions in children. Principles of classification, diagnosis and treatment. Soft tissure tumors in head and neck area

4.1 General characteristic. Etiology. Pecularities in children

A tumor is a lump or mass of tissue that forms when cells divide uncontrollably. A growing tumor may replace healthy tissue with abnormal tissue. Tumors and like tumor lesions of the maxillofacial region in children are subject to the same common laws characteristic for all tumors. At the same time, tumors in children are characterized by pecularities of etiology, morphological structure and clinical manifestation.

Sometimes tumors in children population can become aggressive and can lead to disability or death, particularly if signs and symptoms are ignored.

Tumors in children are different from adult in their ethiology, pathomorpholocical strcture and clinical appearance. 12,4 % of oral and maxillofacial tumors are occur in children, however, 95 % are benign, and 5 % malignant.

Tumors in children are the result of deviations from genetic and embryonic shedule disorders during the period of organogenesis and morphogenesis and during intracellular multiplication, growth and differentiation of the embryo. Tumors are likely to develop from immature or ectopic tissue and may appear in conjunction with obvious developmental anomalies such as an unerupted displaced tooth. They may arise in areas of rapid growth.

That is demonstrated by the examinations showing that most of the tumors in children are occur before 5 years, they have mesenchimal origin (no epithelial), very othen tumors in the oral and maxillofacial region occur toghether with others malformations. Most of the soft tissue tumors in maxillofacial region in children arise from conjunctivetissue (vessels tumors). Oral tumors a connecting with odontogenesis and arise from odontogenic epithelium. The benign tumors of the soft parts, originating from the connective tissue (the vascular tumors) predominate, followed by the tumor of the facial bones. Among the tumor formations of the tissues and organs of the oral cavity, tumors originating from the covering and dental epithelium, from the epithelium of the large and small salivary glands; the rare ones come from connective, vascular, lymphatic, and very rarely - neurogenic tumors.

Tumors and like tumor lesions with localization in the bone tissues of the jaws affect most often children aged of 7-12 years, less often those of 12-16 years and very rarely - children up to one year. There is an opposide picture in the tumors of the oral cavity: the most affected are the children up to one year and those of 12-16 years.

The disontogenetic tumors can be observed immediatly after the birth of the child or in the first year of life. The increasing number of tumors in children of 7-11 years corresponds to the active growth of the facial bones, and their increased frequency at 12-16 years - with the intensive activity of the endocrine system (increased body growth, sexual development).

The frequency of some tumor forms is directly proportional to the age of the child. For example, hemangiomas, lymphoangiomas, dermoid cyst, myoblastoma, mesenchinoma, melanoma neuroectodermal tumor of the baby are characteristic for children under 5 years, reticular sarcoma - for those of 2-5 years. In older children, usually, benign tumors, Ewing's sarcoma, are encountered.

Some tumor processes and dysplasias are dependent on hormonal activity. In boys, osteoblastoclastoma, lymphangioma, angiofibroma, malignant tumors of the lymphatic system are more frequently encountered; in girls - hemangiomas, teratomas, papillomas of the mucosa of the oral cavity, Olbrait syndrome.

Tumors in children population (even benign ones) grow much faster than adults. Because of this, benign tumors are often confused with malignant ones. One of the main features of benign tumors, in childhood, is the family predisposition and it can be tumors of heredity origin, such as: gingival fibromatosis, neurofibromatosis, cherubism, hemangioma. Their presence in parents facilitates their diagnosis in children.

A specific peculiarity of some tumors in children is their ability of involution over time (hemangioma, lymphangioma, papillomatosis of the mucosa of the oral cavity). Tumors are likely to develop from immature or ectopic tissue and may appear in conjunction with obvious developmental anomalies such as an unerupted displaced tooth. They may arise in areas of rapid growth.

The hemangioma, which is considered a disorder during the embriogenesis development of blood vessels, has an infiltrative growth, it infiltrates the healthy neighboring regions, it can damage the organs, destroying their anatomical and morphological structure, and their surgical removal is very difficult. The rhabdomyoma and hamartoma evolve identically. Ossification of pathological foci in the bones of the jaws during adolescence manifests in some patients with cherubism.

Etiology. There are many factors that can cause dysregulations within the cell-multiplication, making the development of tumors. In many cases, its origins may lie in changes in the Genetic conditions: 1. Genetic disorders and gene mutations are predisposing factors for

some benign and malignant soft tissue tumors. 2. Radiation can produce disorders and cause postirradiation bone tumors, postirradiation fibrosarcomas. The pathogenetic mechanism is the emergence of radiation-induced genetic mutations that encourage neoplastic transformation. 3. Chronic lymphedema may predispose individuals to the development of lymphangiosarcoma. 4. Some environmental factors can be carcinogens. The occurrence of hepatic angiosarcoma, for example, has been linked to arsenic, thorium dioxide, and vinyl chloride exposure. 5. Infection can be considered an etiological factors. A classic example of an infection-induced soft tissue tumor is Kaposi sarcoma resulting from human herpesvirus type 8 in patients with human immunedeficiency virus (HIV). 6. Trauma of soft tissue can lead to development of tumors. The relationship between trauma and soft tissue tumors appears to be coincidental. Trauma probably draws medical attention to a pre-existing lesion. Exposure to dietary, lifestyle and environmental factors play a major role in the development of cancers in adults, but only a few risk factors, such as ionising radiation and a mother's exposure to X-rays during pregnancy have been identified as risk factors in children.

There are differences between age group in terms of etiology, pathophysiology and treatment of the pediatric neck mass. In adult neck mass is to be considered maligmant until proven otherwise. The etiology of the pediatric neck mass is more often infectious or congenital.

Environmental factors must be considered in the ethiology of the pediatric neck mass. Radiation exposure is associated with an increased risc of thyroid cancer and salivary tumors. Dilantin and other drugs may lead to cervical adenopathy. Tropical diseases may appear in those immigrants from endemic areas, or in those with recent travel histoties to those areas. Breakage of skin by a cat’s claws may lead to cat screach fever. Cockroaches and cat have been known to carry toxoplasmosis. Fungal diseases have been associated with swimming in contaminated waters.

Pecularities. Children's cancer is not only less frequent than adult cancer, it is also different. None of the most common adult cancers - prostate, breast, lung, colon - are found in children. Instead, children tend to get leukemias, brain tumors and other cancers of the blood and connective

tissues. Adult cancers are thought to occur because of years of cumulative damage to the cells. In children, this kind of long-term damage has not had a chance to take place. Their cancers are thought to occur, instead, during periods of vulnerability during early development, when the organs are still forming. Tumors at the pediatric skull baze can be broadly categorized into congenital (nazal glioma, encephalocee, and dermoid cyst as well as teratoma, hamartoma and choristoma) and true neoplasma (olfactory neuroblastoma, angiofibroma, nazopharyngeal carcinoma and rhabdomyosarcoma). Clinical evaluation often gives significant clues as to the nature of skull base pathology. The time of onset (at birth or postnatal), localization (near the lines of fusion or not), and tumer progression (static or enlarging) all contribute to the accurancy of diagnosis. In children and adolescents, neoplastic lesions are often benign and are of mesenchymal origin. Choung and Kaban were of the opinion that tumor histology in this age group did not correspond to their clinical behaviour.

There are significant differences that must be considered in the surgical management of skull base lesions in the pediatric patient. Neoplastic lesions as well as surgically manipulated tissue dynamically interact with growth potential of the regional anatomy before and after treatment. This impact on growth is an iportant distinction between pediatric and adult skull base surgery that has to be considered during oncologic surgery as well as reconstruction.

There are differences between age group in terms of etiology, pathophysiology, and treatment of the pediatric neck mass. In adult neck mass is to be considered maligmant until proven otherwise. The etiology of the pediatric neck mass is more often infectious or congenital.

Proper diagnosis of tumors in children begins with history andphysical examination especialy in the cases of pediatric neck masses. In younger patients, the parents will need to provide the history. Whereas congenital lesions frequently present at birth, some not be noticed until later infancy (e. g. vascular malformations), early childhood (e.g. vascular malformations) early childhood (e.g. thyroglossal duct cysts), or young adulthood (e.g. branchial cleft cysts). Granulematous diseases such as scrofula usualy follow a chronic course, acompanied by fever or

low-grade temperature. Acute bacterial infection such as supperative sialoadenits usually presents suddenly with pain and discomfort, inflammatory signs, an fever. Neoplasms typically have either an asymptomatic presentation or nonspecific constitutional symptom complex (e. g. Hodgkin's lymphoma). Few malignat lesions are present at birth, with the exeption of rhabdomyosarcoma, or an occasional neuroblastoma presenting in the cervical region. Malignant lesions tend to present in the teenage years. The spectrum of malignancies in children is age related and different from that of adult. Pain associated with these lesions usually is seen in late stages. The masses may result in airway compromise because of direct compression. Increased tenderness and swelling during meals may be associated with obstruction of a salivary gland duct by stricture or stone. Enlagment of the neck mass during crying mai be assotiated with filling of vascular channels as a result of decreased venous return.

The physical examination of a pediatric patient may pose unique challenges. Compliance may be difficult or impossibile to obtain. In any case, one must make the best attempt, so as to acquire the most information about the patient's condition. Although the area of concern is typically smaller than in the adult, one should keep in mind that the neck of a child not a smaller version of the adult. The cartilaginous infant trachea is less prominent and found more cranially than in the adult. Pediatric pathology is more apt to show metabolic or embriologic abnomalities than the adult population. There are also marked differences between subgroups of the pediatric population. Minimal changes in the shape or volume of underlying structures can produce masses that serve as the primary motivator for parents to seek medical advic. The pediatric neck has an abundane of adipose tissue until the age of 9 months when the amount of baby fat startsto decrease, continuing throughout the second year of live. This may lead to a delay in seeking medical advice and more a challenge in diagnosis and treatment.

Sebaceous cysts are usually firmly attached to the epidermis, and therefore one is not able to roll the skin over the surface. Benign masses usually are not attached to skin or deep structurie and thus are usually

mobile. Malignizations, both primary and metastatic, can be attached to a skin and adjacent organs. Cancer are usually localized lesions in the pediatric population. Benign lymph nodes feel soft, well defined, and mobile. Advanced malignancies typicaly feel woody and hard. Limphomas tend to feel rubbery. Cysts feel as through there is an interface between solid and liquid. Cavernous hemangiomas may feel like a ‚bag of worms' with occational fiemness from particles of calcium (phleboliths). Abscess are tender and feel fluctuant. Air in the neck is perceived as crepitance. Pulsation may be derived from an aneurysm or carotid body tumor.

The diagnosis of benign and malignant tumors in children is more difficult to establish than in adults. The late diagnosis is due to the lack of pathomorphological and radiological signs in most bone tumors. Some tumors cannot be identified morphologically in the first years of life (neurofibromatosis and lymphogranulomatosis).

The early diagnosis in oncostomatology in children is a fundamental question. Doctors should know the early clinical signs of benign and malignant formations most commonly encountered in children. Prophylactic examination of children with tumor in the maxillofacial area is performed together with the dentist, pediatrician and oncologist. Children up to one year old must be examined once in a month, and then 3-4 times per year until they rich 5 years old and once a year until adolescence.

The prophylaxis of tumor includes the detection of family predispositions, the elimination of the harmful factors that can affect the fetus during the prenatal period.

4.2 Principles of classification of tumors in oro-maxillo-facial region

The World Health Organization (WHO 2002), classification of soft tissue tumors proposes the morphological classification of tumors, based on the location of the tumor, its morphological structures and their evolution. So far, there are 6 type of tumors classification in the oral and maxillofacial region in children. 1) tumors of soft parts; 2) tumors with localization in the oral cavity and in the pharynx; 3) odontogenic bone tumors; 4) maxillary

cysts; 5) bone tumors; 6) tumors with localization in the salivary glands; 6) tumors with localization on skin.

Soft tissue tumors are a large and heterogeneous group of neoplasms. Traditionally, tumors have been classified according to histogenetic features. (Fibrosarcoma, for example, is categorized as a tumor arising from fibroblasts.) However, histomorphologic, immunohistochemical, and experimental data suggest that most, if not all, sarcomas arise from primitive, multipotential mesenchymal cells, which in the course of neoplastic transformation differentiate along one or more lines. According to the tissue from which they derive, they are classified as follows: 1) epithelial tumors; 2) tumors of the soft parts; 3) bone and cartilaginous tumors; 4) lymphoid and hemoblastic tumors; 5) tumors with multiple genesis; 6) secondary tumors; 7) unclassified tumors; 8) tumor-like raised lesions.

As part of this 2002 WHO classification, soft tissue tumors are divided into the following 4 categories.

- *Benign* - The benign tumors are characterized by slow growth,
- lack of metastases and recurrences after their surgical removal. The cells of these tumors have a high differentiation and are not sensitive to chemical therapy. These usually do not recur locally, and if they do, the recurrence is nondestructive and almost always readily curable by complete local excision.
- *Intermediate* (locally aggressive) - These tumors show an infiltrative and locally destructive growth pattern. However, although they may recur locally but they do not give metastasize. They usually require excision with a wide margin of normal tissue for better local control. The example in this category is desmoid (fibromatosis).
- *Intermediate* (rarely metastasizing) - These tumors are often locally aggressive, but in some cases, they also have a tendency to produce distant metastases (usually in a lymph node or lung). This risk is low (<2 %), but histomorphologically, it is not reproducibly predictable. The classic examples in this group are

plexiform fibrohistiocytic tumor and angiomatoid fibrous histiocytoma.

Malignant - Soft tissue sarcomas are locally destructive with the potential to recur. The risk of distant metastasis is significant. (Depending on histologic type and grade, the potential ranges from 20 % to almost 100 %). Histologically low-grade sarcomas have a lower chance of metastasis (only 2-10 %).[18]However, the recurrences of such tumors may advance in grade and attain a higher risk of metastatic potential similar to that associated with myxofibrosarcoma and leiomyosarcoma.

4.3 General principles of diagnosis and treatment. Complaints and anamnesis

The diagnosis of tumors in children usually is delayed. They are observed by parents during their exteriorization period. In particular, the tumor of the jaws remain for a long time without clinical signs, because they first extend to the maxillary sinus, and then they cause asymmetric deformations of the anatomical shape of the face. The subjective changes, during the intra-bone growth of the tumor, the children cannot relate it, treating them as normal. Usually, they are observed by the parents accidentally or during the prophylactic examination. Sometimes the affected region (sublingual, submental, submandibular or volume increase of the tongue - macroglosia), lead to functional disorders: blurred speech, disturbirns of dental eruption, occlusion, dental position, etc. The causes and the period of occurrence cannot be explained by the parents, often considering them as consequences of the traumas.

Swallowing and difficult breathing, bleeding caused by hard foods can be caused by hemangioma, dystonia of the thyroid gland, adenoma at the root of the tongue, tumors located in the pharynx.

Some tumors in children are associated with developmental defects, such as congenital lymphangioma, tongue cleft, tongue-tie with camouflaged clefts of the mandibular alveolar processes, congenital pathology of the lower extremities and cardiac defects, and others.

Some tumors are congenital (vascular tumors, dermoid, myblastoma). The congenital tumors in children is characterized by a short and latent period from the occurrence of the tumor to the first clinical signs.

The diagnosis of tumors, the time elapsed from the first symptoms, the diseases of the organs and systems supported by the child until the moment of addressing are recorded in the history of the disease. Unjustified indications in the treatment of formations of unclear origin may change the character, evolution and clinical signs, making the diagnosis much more difficult.

The inspection of the maxillofacial region determines the changes of the anatomical shape, the degree of asymmetry of the face, neck and oral cavity; it establishes the extension of the tumors in the neighboring anatomical areas, their relation to the deep layers, the color changes of the covering skin. For example, clinical manifestation hemangioma refers to color changes from cyanotic to reddish. The tumor located in the supraorbital region, lateral cantus of the eye or base of the external nose are characteristic for dermoid cysts. Inspection in the pediatric neck exam, includes the identification of normal landmarks. Assymmetry, deformities, masses, scars, pulsations, discolorations, sinuses, or fistular are noted. The oral cavity and oropharynx must be visually inspected. Havimg the patient swallow wil also help identify abnomalities. Masses attached to hyoid bone and move with deglutination or tongue protrusion, is a clinical manifestation of tyreoglossal duct cysts, Dermoid cysts are usually attached to the skin. Cavernous hemangiomas tend to increase in size when the head is lowered as a result of venous congestion.

The severe deformities of the anatomical shape of the face are caused by vascular tumors, large salivary glands tumors, embryonic cysts, neurofibromatosis, pseudotumors etc.

The tumors located in the oral cavity have their specificities in the diagnosis. In infants, myoblastoma is localized under the gingival mucosa of the alveolar prosses. At the root of the tongue can be detected the fibroma, the fibroadenoma, which requires differentiation with the embryonic aberrations of or with the total dystopia of the thyroid gland. Rhabdomyoma is often present in the lateral regeon of the tongue. Malignant tumors are localized in the tongue: fibrosarcoma, angiosarcoma, rhabdomyosarcoma.

Tumors of the small salivary glands are often located on the border between the soft and the hard palate, or inside of the big salivary glands like parotid gland. Hemangiomas, lymphoangiomas, retention cysts of the small salivary glands, papillomas, etc. are located on the mucosa of the lip. On the cheek mucosa, at the level of the dental occlusion line, the fibroma, the papilloma is often located.

The oral cavity inspection includes the dental examination of the interdental papillae, the mucosa. Lingual or vestibular dental displacements, deformations of the dental arches, changes in the intermaxillary relations are caused by the compressive action of the soft parts tumors on the dento-alveolar complex or the benign and malignant tumors of the maxillae. The most often, these deformities are caused by hemangioma, fibromatosis, reticular sarcoma, eosinophilic granuloma. The functional disorders of the temporo-mandibular joint for example trismus or spontaneous pain are cause by the spread of the tumor tissues in the pterigo-mandibular and parotid-maseteric spaces. Usually this clinicil signs of tumors appear in the late stage of development.

Palpation may prove to be a challenge in young children, particularly those with tender lesions. One should attempt to to control head motion with the opposite hand while palpating. Location of a mass, may point to the diagnosis. The sternocleidomastoid muscle serves as a landmark that divide the neck into the anterior and posterior triangular spaces. Cervical masses are more commonly found in the anterior triangule. Approximately two yhird of all neck masses in children are inflammatory. Of the remaning lesions, the most common mass of the anterior triangle is the branchial cleft cyst. Midline lesions are commonly thyroglossal duct cysts. The bimanual palpation of the affectted region determines the consistency, the shape of the covering surface, the extantion and the relations of the tumor with the deep planes, and the mobility and the pain. The soft or elastic consistency of the tumor, the warty surface, without limited border, confirms the origin of the soft parts or the inflammatory origin. The pulsations in the projection regions of the tumor formations determine the presence of blood vessels.

The palpation of the lymph nodes allows determining the degree of enlargement and their shape, mobility, consistency, presence of pain and relations with deep planes. Lymph nodes enlarged in volume and painful, indicate the malignant character of the tumors or the presence of metastases in the lymphatic system.

4.4 Morphological and imaging diagnosis

Early tissue diagnosis is the most important component of multimodality treatment for soft tissue tumor. *The morphological examination* (cytogenetic or biopsy) is the final step in diagnosis and there is no other specific laboratory tests for diagnosing soft tissue tumors. The biopsy or cytogenetic examination must be done taking into account the local and general clinical signs. Proper and timely biopsy is critical. An inadequately performed biopsy may complicate patient care and result in loss of limb or life. Biopsy usually is indicated for a soft tissue mass arising in a patient without a history of trauma or for a mass that persists for more than 6 weeks following local trauma. All soft tissue masses larger than 5 cm, as well as any enlarging or symptomatic lesions, also should be biopsied. Small, subcutaneous lesions that persist unchanged for years may be considered for observation rather than biopsy. A high level of suspicion is necessary to ensure early treatment.

Several biopsy techniques are available, including FNAB, core needle biopsy, incisional biopsy, and excisional biopsy. The choice of biopsy is based on the size and location of the mass and the experience of the surgeon. Excisional biopsy is indicated only for small, superficial masses (<3-5 cm in greatest dimension), in which the probability of malignancy is low. Effective reexcision is more likely for smaller malignant lesions that initially are unintentionally treated as benign.

Fine-needle aspiration biopsy - This is a cytologic technique involving the use of a fine-gauge (usually 21- to 25-gauge) needle to aspirate individual tumor cells and microfragments from the mass. The aspirated material can be examined as a cytology smear, with immediate evaluation of specimen adequacy.

Some peculiarities growth and development of the child’s body, related to its growth and development, make the morphological examination

difficult. These particularities refer to the predisposition of the organism to pseudotumor hyperplasias, the identity of the embryonic and blastic tissues.

The final diagnosis is made on the basis of clinical and paraclinical examinations: accusations, anamnesis, inspection, radiographic examination, computed tomography, magnetic resonance imaging examination, cytological or biopsy results.

Imaging diagnosis. In the past 2 decades, imaging studies have contributed greatly to the management of soft tissue tumors. Imaging studies should be obtained before biopsy to ensure that a biopsy of a potentially malignant lesion is taken in a manner that will not preclude limb-salvage surgery. Imaging should also be performed before biopsy, to prevent the biopsy tract from adversely affecting the capture of anatomic detail by magnetic resonance imaging (MRI). The relationship of the tumor and surrounding normal structures to the planned biopsy site should be evaluated, as should the functional status of the involved limb, signs of lymph node involvement, and any other factors that could compromise optimal surgical or radiation therapy.

Because prognosis is primarily dependent on the disease stage rather than the histologic tumor type, evaluation of local and distant extent is pivotal in the ultimate management of soft tissue sarcoma. Imaging methods commonly used for such evaluation include plain radiographs, computed tomography (CT) scanning, MRI, and bone scintigraphy (bone scan). Positron emission tomography (PET) scanning is being used more frequently to assess the metabolic activity and, presumably, the biologic aggressiveness of a lesion. Angiography to evaluate any vascular involvement by soft tissue tumors has essentially been replaced by MRI. CT scanning

- Check for presence and number of pulmonary metastases.
- Consider performing a CT scan of the liver in cases of intra-abdominal or retroperitoneal tumors.

Magnetic resonance images (MRI)

- In contrast to CT scanning, MRI is not limited to the transverse (axial) plane. Coronal, sagittal, and oblique planes may be imaged.

- MRI best defines the relationship between a tumor and adjacent anatomic structures, such as compartment boundaries, nerves, vessels, and muscle.[7,8]
- Although for most patients MRI alone suffices, the information obtained from CT scanning and MRI of the primary tumor occasionally may be complementary. Bony involvement may be better assessed with a CT scan, as may the boundary between normal muscle and fibrous lesions

4.5. Principles of treatment

Surgery is the most important component of any treatment plan for a clinically localized primary or recurrent soft tissue sarcoma. On the basis of the achievable margin, 4 types of excisions may be performed.

The fundamental principles of treatment of tumor formations are surgical removal, chemotherapy and radiotherapy. The choice of the environment (outpatient, inpatient) in which the treatment will be carried out is determined by the age of the child, the character, extent and location of the pathological process, the clinical evolution and the individual particularities of the child.

The limited tumor formations of the organs or soft tissues of the oral cavity (papillomas, retention cysts, fibroids), during the preschool and school age, are treated in the outpatient surgical wards. During the young age, the surgical treatment is performed under inpatient conditions. Malignant tumors will be treated in the oncology inpatient wards.

The basic method in the treatment of neoplasms of the soft parts is surgery. Satisfactory results are obtained, if two important conditions are met: 1) complete removal, according to the determined protocol; 2) obligatory morphological examination of the removed tissues.

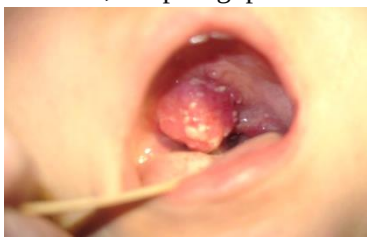
On the basis of the achievable margin, 4 types of excisions may be performed.

- Intracapsular excisions and amputation - The excision or amputation passes within the tumor itself. The tumor inside the pseudocapsule is removed (often piecemeal). Incidence of local

recurrence with these types of excisions is virtually 100 %; these procedures are performed only in unusual circumstances.

- Marginal excisions and amputation - The excision is performed through the pseudocapsule surrounding the tumor. Shelling-out procedures and most excisional biopsies belong to this category. The chance of local recurrence is 20-75 %, depending on the nature of the tumor and whether or not radiotherapy is used.
- Wide excisions and amputation - The tumor is excised with a wide margin of surrounding normal tissue but within the muscular compartment. Without adjuvant therapy, the incidence of local recurrence following wide excision varies but may reach 30 %; the rate of recurrence depends on the selection criteria used and the adequacy of the histologically assessed surgical margin. A wide amputation is performed through the normal tissue proximal to the reactive zone around the tumor but remains within the involved compartment. Limb-sparing procedures belong to this category.
- Radical excisions and amputation - These are en bloc excisions of the tumor along with the entire muscle compartment. Amputation with disarticulation of the joint proximal to the involved compartment is called radical amputation. The risk of local recurrence is lowest with this procedure.

The papilloma is a benign tumor, originating from the striated flat epithelium. Causal factors: small and repeated trauma, viral origin, teratogenic factors, chronic and acute inflammatory processes. It is found at all ages, even in the newborn, but most often at 7-12 years. It is located on the tongue, on the cheek mucosa, the palate, the gums, the short type frenulum, the pterigopalatine flaps. (*Fig.16*)



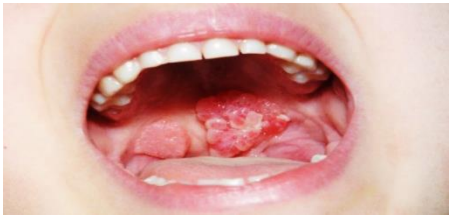


Fig.16. **Papiloma in the oral cavity (soft palate)**

It develops slowly, having a narrow or wide implant pedicle. It has variable dimensions, but does not exceed 7-8 mm in diameter. It is simple or multiple; forms the so-called oral papillomatosis; has a light pink color; it does not cause functional disorders and is not painful. Differential diagnosis is not difficult. The color changes and their erosion are problematic. Large tumors with a broad pedicle resemble fibroids. Their location on the alveolar apophysis is different from the Serra cysts. The treatment includes surgical removal, along with the implant pedicle, and suture. Sometimes electrical coagulation is used.

The fibroma is formed of collagenous tissue. It is found in children aged 7-15 years, rarely in newborns. Causal factors - chemical, thermal and mechanical excitants. Localization - tongue, genial mucosa, alveolar apophyses, mucosa of the palate and lips. The fibroma is round, bordering; may have a wide pedicle; the covering mucosa is unchanged. It grows slowly. The differential diagnosis is made with Serra cysts. Treatment - surgical, by removal within the boundaries of healthy tissues.

The rhabdomyoma is a benign tumor, originating from the striated muscles; it is very rare. It has disontogenic congenital origin. It locates on the tongue, in the pterigomandibular and retroauricular muscles, with a slow, painless growth. It is present in the newborn, but the diagnosis is made up to 3 years.

The lipoma - a benign tumor, with a slow growth, rarely encountered in children. It is located in the thickness of the cheek, near the Bichat bubble, evolving towards the skin. The dimensions can reach 5-6 cm. It has a round, curved shape, soft, pseudofluctuent consistency. It consists of adipose tissue, but histologically fibrous, vascular or nerve interstitial proliferations can be detected. It is usually superficial, limited, but it can also be observed into the infratemporal fossa. The treatment is surgical.

The hemangioma *(figure 17)* is a very common benign tumor in children: 1.1-2.6 % in newborns; 08-1.4 % in children up to one year of all tumors of the body. It is 3-5 times more common in girls than in boys. 30 % are present in newborns, and 70 % occur in the first months of life

of all tumors with localization in the oral and maxillofacial region. Multiple lesions are present in 20 %. The head and neck regions are affected in 50 % of cases, the body – 25 % and the extremities – 15 %.

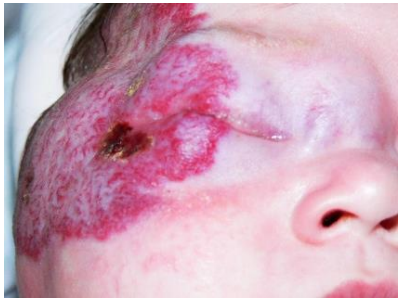


Fig. 17. **Hemangiomas on the lateral part of the face**

Depending on the sizes, the hemangiomas can be: small (up to 1 cm^2); medium, (from 1 cm^2 to 10 cm^2); large, (from 10 cm^2 to 100 cm^2); massive, (larger than 100 cm^2).

According to the classification of G.A. Fiodorova, hemangiomas are divided into two groups: true (malignant) and false (congenital malformations). True, the hemangiomas are capillary, cavernous, racemic; false - flat angiomas (stains of wine, stellates, botriomicoma, medial spots).

The malignant hemangiomas have a tumorous appearance, are present in the newborn or arise at an early age. Their evolution comprises three stages: up to one year - the accelerated growth stage followed by the second stabilization stage and the third involution phase. It can stay for a period, after which they begin to increase progressively in volume through the proliferation of the endothelium and the dilation of the vessels that compose it, sometimes becoming monstrous (*figure18,19*). Others are stationary, with the ability to regres in time. Malignant hemangioomas have a reddish-purple coloration, bulge at the level of the skin or mucosa, have a soft, elastic consistency, reducing by pressure in size, and then returning to the initial volume. The size of the vascular tumors growths when the blood pressure in the head and neck region raised (doenward position of the head, when crying, or in periods of effort).

a b c

Fig. 18. **Hemangiomas. a. before, b. during, c. after treatment.**

The malignant hemangioma may be accompanied by anemia, thrombocytopenia, ulceration, hemorrhage, inflammation.

The capillary hemangiomas are characterized by dilated vessels, which form limited tumors, with endophytic or exophytic growth.

The cavernous angioomas have dilated vessels realize large sinuses, that communicate with each other, forming diffuse and bloody cavities, without precise boundaries, with rapid evolution. They invade

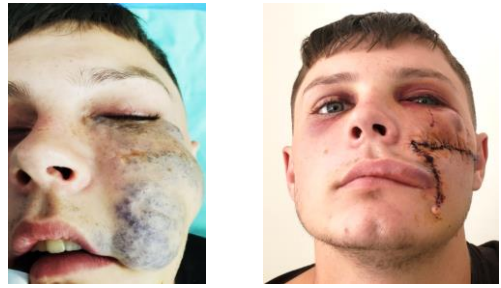


Fig.19. **Limphangiomas. a. before, b. after surgery**

neighboring tissues, which they dissociate or even destroy.

The vascular malformations are errors of morphogenesis; they represent a collection of blood vessels deviated from the norm, with normal endothelium, non-invasive, present in the newborn, growing in proportion to the body of the child, without the ability to return in time.

Flat hemangioomas, called *port-wine stains*, located on the skin or oral mucosa, have different size, invoving a smaller or bigger surface. They are flat, superficial lesions and usually preserve the characteristic of the normal tissue Some of them may infiltrate the derma and appears

like a raised lesion. The coloration differs: light-pink, deep red, bluish-red to bluish. When vitreous pressure on such hemangiomais extend, the coloration disappears. A particular form is Sturge-Weber Disease where the plane hemangioma involves a trigemen nerve branch territory and also meanings and ocular lesions.

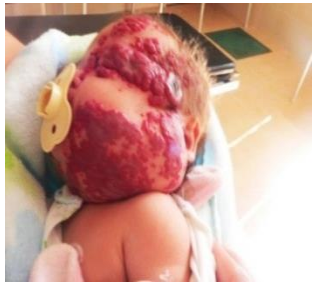
The star-like hemangioomas is a little radial dilatation of the capillaries having a central red point. It is considered as a little arterio-venous anastamosis.

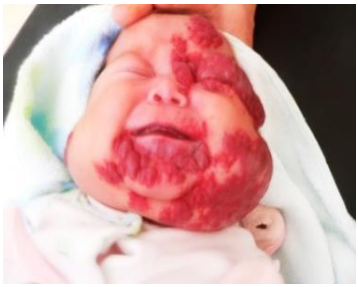
The median stains are present in newborns; it is located in the medial regions of the face (nasal crest, frontal, occipital, upper eyelid). They are visible in physical exertion or during crying. Usually, it does not require treatment and disappears at the end of the first year of life.

The botriomicoma - hyperplastic and infectious tumor; it can be located both on the lip and on the jugal or lingual mucosa. It seems that the causative agent is botriococcus, a germ similar to staphylococcus. It occurs due to local traumatic factors.

Anatomically-pathologically, the tumor is formed of inflammatory connective tissue with numerous vessels. Usually, it occurs at different time intervals, after minor traumas (bites or stings). The tumor is small, pediculate, with dimensions not exceeding 1 cm, red in color, with a finely granulated, muriform or ex-ulcerated; it has elastic consistency and bleeds to the slightest touch. When it reaches a certain volume, it is stationary. In some cases, it is ulcerated, infected or necrotized, accompanied by inflammatory adenopathies. The differential diagnosis is made with papilloma, angioma or carcinoma. The treatment is surgical - excision. It relapses in cases where it has not been radically removed.

The treatment of hemangioomas varies from one form to another, as well as the age of the child. The treatment methods used are: cryotherapy, electrocoagulation, sclerosing therapy, intratumoral hormone therapy, laser therapy, radiotherapy, surgical treatment, combined treatment.





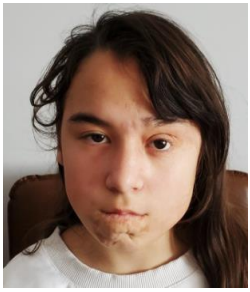


Fig 20. **Malignant hemangioma. a, b, before, c. after treatment**

The ma lignant hemangioomas are treated by the combined application of the methods. The purpose of the treatment is primarily to arrest growth. For this purpose, sclerosing therapy, intratumoral hormone therapy or the action of laser waves are indicated. The action of laser waves is based on the selective absorption capacity of the vascular tumor, depending on the type of the hemangioma, after which the fibrous transformation of the tumor walls takes place. The surgical removal is performed after tumor remission.

The stellate hemangioomas can be electrocoagulated with a very small needle inserted into the central vessel of the tumor. In *flat angiomas (stains of wine)*, contact radiation therapy (buckyterapia), laser wave action, or surgical removal are practiced. The harmful action of the roentgen rays on the growth and development of the child, determines the reduced indications of these method in the treatment of hemangiomas, replacing it more and more with other methods. Good results are obtained by applying carbon snow (cryotherapy), remaining supple whitish scar. Superficial skin abrasion can be used in superficial plane hemangioomas and in delimited ones, with deeper interest,the surgical excision can be used.

The angioendothelioma is a variety of vascular tumor, rarely found and localized in the muscles of the tongue. Clinically, it manifests by one or a few limphnods of cyanotic color, soft consistency, with the covering mucosa without modifications. The vascular drawing instead of tumor projection is intensified.

The angioendothelioma has a tendency to ulceration, due to its location in the superficial layers. It is found in children aged 11-15 years. Treatment - surgical removal. (*Fig. 21*)

Kasabach-Merritt syndrome is characterised by the combination of rapidly growing vascular tumour, thrombocytopenia, microangiopathic haemolytic anaemia and consumptive coagulopathy. The blood clotting disorder results from platelets and other clotting factors of the blood being used up within the tumor.

Venous malformation or venous hemangioma is acongenital tumor with abnormal vascular development that result in aberrant and ectatic venous connections. Venous malformation is anautosomal dominant disorders and rare slow growing benign tumor. Being aberrant in venous connections, venous hemangioma leads to venous congestion, thrombosis, phlebolith formation and gradual expansion of these lesions.

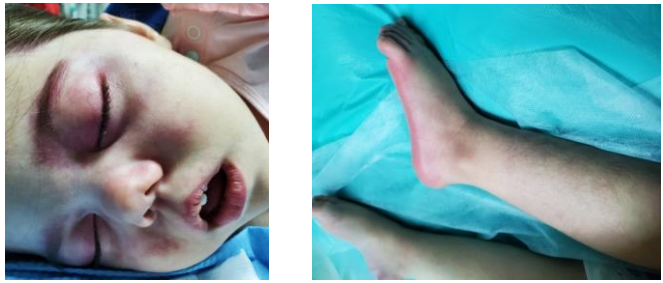


Fig. 21. **Kasabach-Merritt syndrome**

Venous and lymphatic malformations form during fetal development and are present at birth, but they may or may not be noticeable at birth.

Lymphatic malformation (lymphangiomas) are formed by abnormally enlarged lymphatic vessels. Normal lymphatic vessels drain excess fluid from tissues and eventually recycle this fluid back into your veins.

In lymphatic malformations, fluid drains more slowly and pools in the enlarged area. Lymphatic malformations can become infected and cause pain, leak out of the skin, or bleed.

The pathogenesis theory of development of the lymphatic malformation, recognized by most scientists, start in the early pregnacy period. The mesenchymal primordium grows branches and forms the future lymphatic channels. The branches are located along the blood vessels. They grow, develop and form six lymphatic pouches, two of which are located around the jugular vein. Gradually, the lymphatic pouches merge between them to form the lumphatic system. Under the action of teratogenous factors, disorders of the lymphatic system developed result.

Microscopically, typically consists of closely packed thick walled vessels, which are variably dilated with multiple cavities, lined with endothelium and filled with clear, pale yellow fluid. Sometimes display thrombosis with occasionally formation of phleboliths (calcified thrombi).

Lymphatic malformation can form anywhere in the body but most common in the head and neck area. Affected areas can becheek, buccal floor and nasopharyngs and oropharyngs space, along the vessels of the neck, tongue. Clinical fetures ussualy appears as a growing, spongy-feeling lump that is present at birth. In deep mass overlying skin may appear normal or bluish discoloration, If it's close to the skin surface, it can look bruised. The skin over lymphatic malformations often has small bubbles, called **vesicles** (VESS-ih-kuls). These look like tiny blisters. The fluid in them starts out clear and colorless, but will turn dark red if blood leaks into it. With more cutaneous involvement, the lesions appear darker blue or purple. Enlarged part of the neck and face areas, where lymphatic malformation is located, decreased motor control and sensation can occur. At the palpation, a soft tissue is not reducible, there is no fluctuation.

Mostlymphatic malformationsthat appear suddenly will decrease in size and pain withouttreatment, but very rarely go away ontheirown. Incision (cutting into) and drainingthe lymph from a lymphatic malformation can temporarily reduce its size. This is usually only done for diagnosis or to treatm an infection.

The lymphatic malformation can be: capillary, cavernous and cystic. The clinical manifestation depends on age, location, size, histological structure and can be well limited or diffuse. At the same time, lymphatic malformation has common clinical signs. As a lymphatic malformation grows they may put pressure on nearby area such as gums, teeth, upper or lower jaws that lead to disorders in growth and cause anatomical deformities of the shape. The diffuse type of lymphangioma of the tongue is the cause of inferior progeny and excess development of the chin, occlusion disorders, breaphing, speaach and swallowing dysfunction. The well defined hemangiomas located on the mucosa of the oral cavity or on the tongue are manifested by small cysts, which appear clinically in the form of „dew drops", with translucent white or bloody content.

All lymphatic malformation have the capacity to increase in volume during periods of acute infections (tonsillitis, stomatitis, etc.) and come back after the infection go back.

The cystic type of lymphatic malformation consists of one or several cavities filled with serous citrine fluid. It is located in the regions of the neck and and submandibulary regeon, being well defined. It is located in the cervical, submandibular lateral regions. The skin is not modified or gets cyanotic. It is painless, fluctuating. The cystic type of lymphatic malformation are not prone to inflammation.

The differential diagnosis is made with hemangiomas, neurofibromatosis, medial and lateral cervical cysts, dermoid cysts. The lymphangiomas located on the upper lip are different from Milkenton-Rozental syndrome. In the period of inflammation, the differentiation of lymphangiomas is made with cellulites.

The treatment is made according to the shape, volume and location of the tumor, the age of the child. Surgical excision is employed for the more superficial lesions, while deeper lesions are typically treated with sclerotherapy, the injection of a chemical agent that causes the lesion to shrink. In most cases, a multidisciplinary treatment is reqiued, which includes the treatment of chronic outbreaks, including the dentistry one, the therapy with antibiotics, surgical elimination of tumor, orthodontic and speech therapeutic treatments.

The mioblastomyoma is a rare tumor of indeterminate origin. The lesion is thought to derive from the striated musculature, but some opinions support the neuronal theory (originating from the connective tissue of the nerves). Two varieties of this tumor are known: granular cell myoblastoma and congenital myoblastoma, found only in newborns.

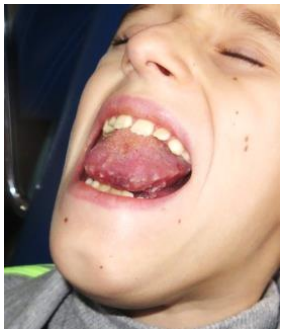
The granular cell myoblastomyoma is most often located on the tongue, in the form of single or multiple nodules, and sometimes in slices; it can grow considerably. The covering tissues look normal. Clinically, it may resemble to leukoplasty. It is determined in newborns and children of 3-12 years.

The congenital myoblastomyoma is a very rare tumor, found in newborns. Clinically, it is located on the alveolar apophysis, in the form of a fistula, located on the foot, the size of a grain. It comes from the alveolar apophysis and contains portions of the odontogenic epithelium. It differs from fibroma. The treatment is surgical.

The tumors of the tongue (figure 22) are related to the embryogenesis of the child. In the anterior part of the tongue are located papillomas, neuromas, myoblastomas, as they are associated with some pathologies of language development.

In the posterior part of the tongue, the present formations are due to the embryogenesis disorders of the thyroid duct (congenital cysts and fistulas) and the thyroid gland. In some cases, at the root of the tongue, a portion of the thyroid gland is located, and in others at the root of the tongue, the base portion of the thyroid gland is located. In all cases of presence of tumor formations in the posterior third of the tongue, the endocrinologist investigation is required.

The tumor-like lesions of the soft parts. *The papillomatosis* is located on the mucosa of the oral cavity, in the form of multiple papillary hyperplasia, identical to the papilloma. The elements have a large red implant base. It is located on the mucosa of the buccal cavity, at the level of the lip, cheeks and commissures and most often caused by trauma. Treatment – removal of traumatic factor and surgical removal.



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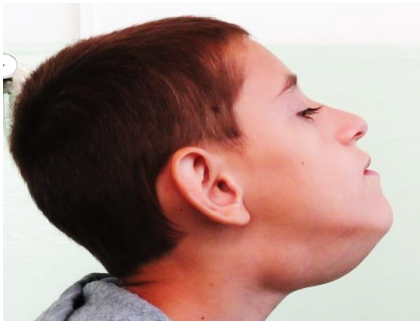

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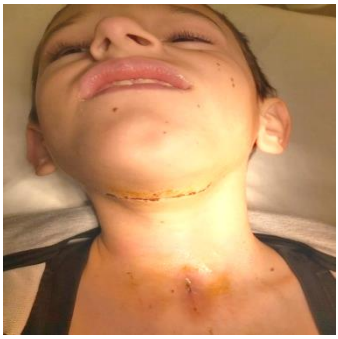

d

Fig. 22. **Limphangiomas on the tounge and submental area. (a, b, before, c, d, after treatment)**

The fibrous inflammatory hyperplasia can be localized in any region of the mucosa of the oral cavity. It is a consequence of the child's ability to bite his lips, tongue, cheek. They are also caused by the chronic trauma of the obturations, the sharp edges of the teeth. Clinically, it appears in the form of swelling, hard, sore, with unchanged covering mucosa. The treatment consists in removing the irritating factors.

The gingival fibromatosis (juvenile gingival hyperplasia- *figure 23*) is a pseudotumoral lesion, which manifests by the massive growth of the gum, with very slow evolution and family character. The condition is rare and is determined by the autosomal dominant gene. The associations of gingival fibromatosis have been found with other abnormalities: enlarged nose, loose and elongated ears, splenomegaly, bone abnormalities, hypertrichosis.The

histopathological appearance is characterized by intense fibroblastic proliferation of the chorion and perivascular lymphoplasmocyte infiltrates, while the malpighian epithelium of the gum is normal or slightly modified. The dense masses of fibrous connective tissue predominate.

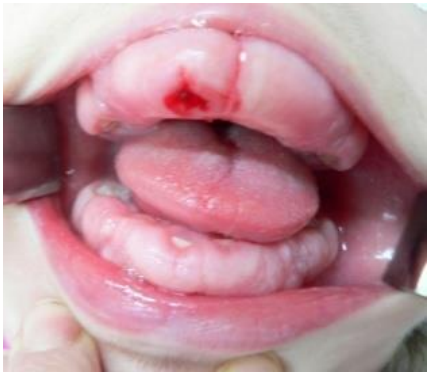

Fig. 23 **The gingival fibromatosis**

Installed in childhood, often during the eruption of permanent teeth, the disease manifests through hypertrophied gum, with dense, diffuse, smooth or nodular growth of the gingival margin along the arches (vestibular and oral), gradually covering the teeth. It may comprise one or both arches. The mucosa has a normal appearance, the gingival hypertrophy is firm or hard, painless, preventing the normal eruption of teeth through various abnormalities of dental position. The gingival tissue, by increasing its volume, can cover the teeth, causing chewing, phonation disorders. The jaws are often deformed. Sometimes, generalized gingival hyperplasia is associated with hypertrichosis, mental impairment, epilepsy, bone or soft tissue abnormalities. The radiological examination reveals partial anodonts of primary and permanent teeth.

The predisposing factors of fibromatosis are: 1) low reactivity of the organism; 2) hereditary factors; 3) endocrine disorders in the transient period.

The differential diagnosis must be made with gingival hyperplasia, manifestation of various general conditions (hemopathies, dysendocrinopathies, hypovitaminosis, etc.), with consecutive gingival hyperplasia.

The surgical treatment consists of gingivoectomy, followed by the orthodontic monitoring.

The giant cell epulis with a structure identical to that of the central granuloma, which contains giant cells. It occurs more often at the age of 8-14. Location of the epulis - alveolar ridge, more commonly on the vestibular slope (*figure24*).

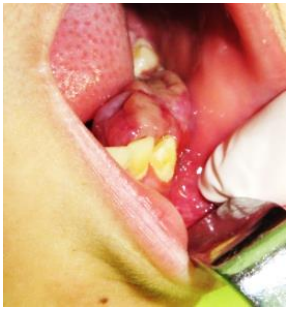

Fig. 24. **Epulis.**

The epulis begins with a gingival hypertrophy located at an inter-dental papilla or at the free edge of the gum, very rarely intra-alveolar, causing odontalgy and dental mobility, dental displacements. It grows slowly, insidiously, without pain, developing freely in the oral cavity. The epulis may have a broad implant base or pedicle, growing on the vestibular, oral or both sides of the alveolar apophysis. The granulomatous epulis has a nipple, reddish-purple surface, elastic consistency; it ulcerates, bleeds slightly. The differential diagnosis is made with fibromatous and angiomatous epulis. The prophylactic treatment: oral cavity remediation, rigorous oral hygiene. The treatment is surgical.

4.6 The tumors and cysts of the salivary glands are divided into epithelial, non-epithelial and unclassified tumors. There are cysts of the large and small salivary glands. The epithelial tumors of the large and small salivary glands are rarely encountered. (*Fig. 25*).

Adenoma. Pleomorphic adenoma (mixed tumor) is most commonly located in the parotid (*figure25*), submandibular, and small salivary glands of the soft palate and hard palate. It is found at the ages of 7 to 11 years. The onset is nodular, with a slow evolution (a few years), painless spontaneously and under pressure, well delimited, of reluctant consistency, it does not

infiltrate the surrounding tissues, it is not accompanied by adenopathies and relapses. It has a nodular shape, a bumpy surface, surrounded by a connective capsule. Sometimes tumors can be found outside the capsule and this is why it is considered multifocal (pleumorphic). The tumor is gray-ashen, tiger color; in the translucent section, sometimes cystic, hemorrhagic or mucoid areas can be observed, which alter with areas of hard, cartilaginous, and sometimes bony consistency. Microscopically, the tumor is characterized by the simultaneous presence of glandular epithelial cells and mesenchymal cells. The epithelial cells have various forms (cubic, round, fusiform), and stroma - various aspects (mucoid, myxomatous, pseudocartilaginous, hyaline, rarely osteoid). It differs from hemangiomas, lymphangiomas, cylindroma. Total surgical removal of the tumor is the basic treatment.

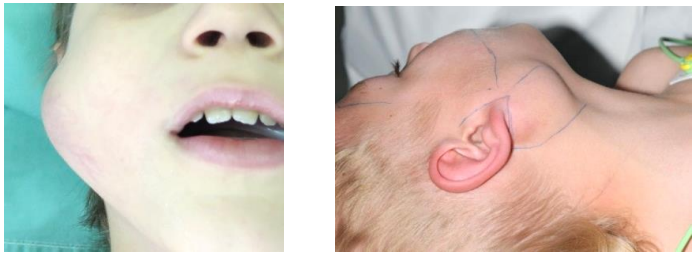

Fig. 25. **Tumor of the parotid salivary gland.**

The monomorphic adenoma or adenolymphoma (Warthin tumor) is a tumor lesion of salivary glands of disontogenic origin, located in the parotid gland. The tumor is variable in size and represents a yellowish, rather thick and creamy capsule. Microscopically, it has an epithelial composition, with cylindrical and cuboid cells, and a lymphatic composition, with numerous granular centers.

The clinical symptomatology is very similar to that of the polymorphic adenoma (slow evolution, painless, without adenopathies and paralysis of the facial nerve). It is well encapsulated. Having an elastic consistency, it can be confused with the lipoma and pleomorphic adenoma, where the lipomatous tissue predominates. The treatment is surgical.

The mucoepidermoid is a variety of salivary gland tumor lesions; it is characterized by the ability of the epithelium to differentiate, through

intermediate forms, into mucosal cells or cells of epidermoid character. It has an increased potential for malignancy, due to the epithelial-glandular structure, in which malpighian, muciparous and intermediate cells are present. Clinically, it is identical to the polymerphic adenoma, but it has a rapid growth and a higher malignant capacity, especially when the histopathological intermediate cells predominate. It is very rare and affects children over 12 years old. The surgical treatment consists of the removal along with the entire salivary gland.

The cylindroma is mainly located in the mucous secretion glands - the soft or hard palate, at the root of the tongue and the small salivary glands. It is very well delimited; sometimes extremely infiltrating forms can be encountered. It consists of two types of cells: canalicular and myoepithelial, oriented around cystic spaces. This tumor is characterized by the growth along the nerve trunks with peri- and intranural invasion, which are aborous the appearance of early, irradiating spontaneous algae - the clinical signs of this lesion. The cylindroma is prone to hematogenous relapses and metastases. Treatment of mucoepidermoid and cylindroma consists of the combination of surgical and radiotherapeutic treatments, especially for low-differentiation epidermoids. Surgical treatment: removal with the salivary gland. Differentiated mucoepidermal lesions will be treated only surgically.

The cysts of the small salivary glands and rarely congenital. Depending on the location, there are two types of cysts –of large salivary glands (sublingual and submandibular) and of small salivary glands (lips, cheeks, palatal, lingual).

The sublingual ranula starts insidiously, often being accidentally discovered; exobuccal, without changes. Endobuccal, the ranula is highlighted when the tongue is raised, having the appearance of a „frog goiter". The tumor is round or oval, covered by smooth, glossy mucosa, blue in color, with mucoid fluid content, of soft, fluctuating consistency, adherent to the deep soft planes, where its limits cannot be specified. As the tongue grows, it can move up and down, causing functional disorders (left tongue movements, mastication and phonation). The causes of this tumor are the inclusion and the cystic transformation of embryonic remnants of the second

branchial arch or of the thyroid canal (Neuman), or the cystic transformation of the acini of salivary glands from the buccal floor, resulting from theobstruction of the excretion channels or infection.

The tumor is located on the left or right sides from the tounger, above the milohioidian muscle, bulging under the mucosa of the buccal floor. Occasionally, it develops back and down, passing into the submandibular lobe, either through the space between the milohioid and hioglossus muscles, or by dissociating the fibers of the milohioid muscle, giving the tumor a cystic appearance in bilobate form, in bi-sac. Very rarely, usually in cases of incomplete extirpation, the cyst develops only under the milohioid muscle.

The cystic membrane is composed of three layers: 1) the peripheral layer, composed of fibers and adult conjunctival cells (fibroblasts) and extremely adherent to the surrounding tissues (mucosa, salivary gland, muscles); 2) the middle layer with a young conjunctival structure, with an embryonic character, very well vascularized; 3) the inner layer (sometimes missing) is composed of a discontinuous epithelium. The content is a clear, viscous liquid, similar to egg white, rich in albumin and mucin, with suspension of epithelial cells.

The Blandin-Nunov cysts are a variety of salivary cysts, located in the anterior region and on the lower surface of the tongue.

The differential diagnosis is made with the cystic dilatations of the Warthon canal, where the swelling increases during meals; with the median localized dermoid cyst, with the covering mucosa of normal appearance and pasty consistency; with salivary cyst, usually smaller, located much more superficially; with the angioma, which has the property to change its volume in the declining position of the head.

The suprahydroid ranula is located paramedian, protruding between the basilar margin of the mandible and the hyoid bone. Covering skin looks normal, being mobile, non-sticky. It is of soft, fluctuating consistency. By endobuccal and exobuccal palpation, the contents can be pushed from one compartment to the other. Functional disorders are not caused.

The differential diagnosis is made with the branchial cyst, which cannot be detected by the endobuccal route.

The treatment is surgical - removal of the cystic membrane or marsupialization. Often the cyst is removed along with the gland.

The Recklinghausen's disease (neurofibromatosis -*figure 26*) (is a serious hereditary disease characterized by the presence of multiple subcutaneous neurofibromas. Usually, neurofibromatosis is accompanied by disorders of the endocrine and vegetative systems.

***Fig. 26*. Neurofibromatosis.**

The etiology is not definitively known. It is considered that itstarts in the period of embryonic development. At the base of the etiology are the disorders of ectoderm and mesoderm formation. The participation of the ectoderm is based on the clinical signs of impairment of the nervous system, and of the mesoderm - on changes in the bone system. In children with neurofibromatosis, embryonic disorders in the development of the central nervous system and retardation in intellectual and physical development are often present.

The clinical signs of neurofibromatosis in children may occur in the first days or in the first year of life, by unilateral increasing in volume of the soft parts of the face region (auricle, nose, macrodentia and bulky dental buds). In young children, the covering skin is not modified, and the elasticity - high, the turgor preserved. In the first 4-5 years of life, the tumor evolves clinically and morphologically, identical to myoma, lipoma, fibrolipoma, lymphangioma. Specific clinical signs appear at 4-5 years, more obvious at 12-15 years - brown spots on the skin of the abdomen, back, chest; low turgor, early skin wrinkles. The affected mucosa loses its normal appearance, becoming smooth, with a yellowish hue, without gloss. In the thickness of the tumor, hard, painless nodules can be palpated. There are signs of facial nerve damage, with paresis of mimic muscles. Radiography shows outbreaks of osteoporosis, giant teeth.

The differential diagnosis until the installation of the clinical signs is made with lymphangioma, which has signs of local inflammation at the time of acute viral infections, etc. Lipoma macroscopically characterizes the tumor in the early period of the child. Sometimes, both the lipoma and the hemangioma cannot be differentiated morphologically from neurofibromatosis and only with age the diagnosis materializes, when the clinical signs characteristic to the tumor appear. The treatment is surgical and is made in the form of palliative removal.

The nevus is a congenital formation of the skin, which appears after birth. It is characterized by the modification of the skin (hyperpigmentation or depigmentation), the character of the surface (with hairy, verrucous covering) and the surface it occupies. The nevus is located at the level of the epidermis and dermis, the dermis. Depending on the location, the nevus may be epidermal, intradermal or combined. Clinically, the nevus has a flat spot or brownish-dark node, slightly protruding above the skin, sometimes the surface is smooth (*figure 27*). The intradermal nevus appears clinically in the form of delimited flat spots, of various shapes and sizes, with brownish hues. The treatment is surgical and consists of its removal being performed plastic manipulations of the postoperative defect. When the nevus encompasses important surfaces, then the unaffected skin of the child is collected,

***Fig. 27*. Nevus on the skin of the face**

grown in vitro, then applied to the postoperative surfaces, gradually removing the nevus.

Learning objectives

1. Peculiarities of the evolution of soft parts tumors in children.

2. Characteristic of benign tumors of soft parts in children.
3. Methods for diagnosing pathological formations.
4. Lymphatic malformation and venous malformation (etiology, clinic maniferstation).
5. Hemangioma (etiology, classification, clinical picture, principles of treatment).
6. Methods of treatment of hemangioma and lymphemangioma.
7. Tumors of soft tissues; methods of diagnosis.
8. Tumors-like lesions of the soft tissue.
9. Methods of treating the tumor processes of soft parts.
10. Indications for admission.

Tests

1. CM. Treatment of the cysts of retention of the sublingual gland will be performed by:
   A. Curettage;
   B. Surgical removal along with the sublingual gland;
   C. Marsupialization;
   D. Conservative treatment with iodine-containing solutions;
   E. All answers are correct.
   (B, C)
2. CS. Clinical examination of the sublingual ranula will reveal a tumor:
   A. Round or oval;
   B. Smooth;
   C. Blue colored;
   D. Translucent;
   E. All answers are correct.
   (E)
3. CM. The content of the sublingual ranula is a liquid:
   A. thick/cloudy;
   B. clear;
   C. viscous;
   D. red-brown;
   E. colloidal.
   (B, C)

4. CS. Localization of retention cysts of small salivary glands of Blandin-Nunov type:

A. in buccal floor;

B. paramedian, above the milohioid muscle;

C. on the inferior surface of the tongue;

D. in small salivary glands (on the inferior lip);

E. in small salivary glands (on the superior lip).

(C)

5. CM. The differential diagnosis of sublingual ranula is made with:

A. hemangioma;

B. myxoma;

C. lymphangioma;

D. botriomicon;

E. dermoid cyst.

(C, E)

6. CS. The papilloma develops from:

A. small salivary glands;

B. fibrocytic cells;

C. planar multilayered epithelium;

D. giant cells;

E. connective tissue.

(C)

7. CM. Stellate angiomas can be treated by:

A. sclerosis;

B. electrocoagulation;

C. cryotherapy;

D. contact radiation therapy;

E. surgical excision.

(A, E)

8. CM. Cavernous hemangiomas are treated by applying methods as:

A. cryodestruction;

B. electrocoagulation;

C. radiotherapy;

D. sclerosis;

E. surgical excision.

(D, E)

1. CS. Treatment of cavernous hemangiomas includes:

A. sclerosis;

B. surgical removal;

C. cryotherapy;

D. thermal coagulation;

E. all answers are correct.

(E)

10. CS. Hemangiomas with capillary dilation, with a red spot in the center, are called:

A. flat;

B. capillary;

C. stellate;

D. cavernous;

E. median spots.

(C)

11. CM. Name the true hemangiomas:

A. stellate;

B. flat;

C. cavernous;

D. capillary;

E. branchy.

(C, D, E)

12. CS. Medial hemangiomas are characteristic for children:

A. newborns;

B. 2–3 years;

C. 3–7 years;

D. 7–12 years;

E. 12–16 years.

(A)

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5. Benign tumors in children. Odontogenic and non-dontogenic tumors in children

5.1 General characteristic tumors in head and neck area. Etiology

Tumors arising in the pediatric facial skeleton represent a formidable challenge in clinical practice because of the complex anatomy of the head and neck and because of facial development during this time span. Although rare, these conditions include a broad spectrum of lesions with a varying degree of malignant potentia. Their true incidence is difficult to establish due to the paucity of reports in the literature and because most published articles are either isolated case reports or series dealing with a specific histological diagnosis. Depending on lesion origin, tumors can appear in different period of development of the children and adolescence. Evolution is slowly growing tumor and does not change the general condition of the patient. For this reseon they are diagnosed accidentally, late or on the occasion of complications. Some children are not diagnosed correctly at the initial stages as having a neoplasm and are wrongly treated for infections by antibiotic administration. Subsequent to an unresponsive antibiotic therapy radiographs are taken to reveal a radiolucent or radiodense lesion in the jaws. Finally a tissue diagnosis becomes necessary in order to diagnose and initiate proper therapy. The rapid growrh and development process in childhood

and adolescence affects the growth potential of the tumors and tumor like lesions and can result in considerable morbidity.

Most tumors of the jaw in children are odontogenic and constitute a wide range and diverse kind of lesions derived from tooth forming apparatus and its remnants. Odontogenic tumors originate from epithelium or ecthomesenchyme or from both, showing varying degrees of inductive interaction between these embryonic components of the developing tooth gem. Most commonly they are located within the bones of the jaw (central odontogenic tumor), while extraosseous odontogenic tumors occur nearly always in the tooth-bearing mucosa (peripheral odontogenic tumor).

Nonodontogenic tumors encompass a wide range of pathologic conditions and may arise from mesenchymal tissue within the jaw or from the osseous tissue of the jaw.

According to the current literatures the etiology of tumors remains unknown. However, genetic background, environmental factors such as viral infection, chronic malnutrition, trauma, alcohol and tobacco intake has been suggested in the aetiopathogenesis of maxillofacial tumours. Pathologic disorders include various histological types and different clinical behavior. However, benign tumor has their own characteristic: mature and well differentiated cells structure, which replaces the bone elements, but do not destroy them and they vary widely in their level of aggression. They do not relapse and do not give metastases. The potential sources for development of an odontogenic tumor include: a) the prefuncional dental lamina (odontogenic epithetlium with ability to produce a tooth, which is more abundant distal to the lower third molars; b) the post-functional dental lamina, a concept that covers, those epithelial remnants such as Serre's epithelial rests, located within the fibrous gingival tissue, the epithelial cell rests of Malassez in the periodontal ligament and the reduces enamel organ epithelium, which covers the enamel surface until tooth eruption; c) the dental papilla, origin of the dental pulp, which has the potential to be induced to produce odontoblasts and synthesize dentin (dentinoid); d) the dental follicle; e) the periodontal ligament, which has the potential to induce the production of fibrous and cement-osseous mineralized material.

5.2. Peculiarities in clinical signs and symptoms of pediatric jaw tumors

Pediatric tumors of the jaw can present a diverse clinical pattens and can vary from odontogenic to non-odontogenic lesions. Pediatric patients encompass the varied range of conditions connecting with long term changes that take place in the maxillofacial area. It's refers to the specific characteristic of the region: continuous development of the region with permanent growth of the primary dentition, mixed and permanent dentition. During the mixed dentition period odontogenic cyts or odontogenic tumors have been considered. Because of their slow-groing tendency, usually they do not cause pain. At this period, odontogenic tumors are observed causually or after appearance of nonspecific symptoms. In the majority of cases tumors in the head and neck area are first seen by general practitioners or pediatricians with subsequent delays in inverstigations and diagnosis. A physical sighns and symptoms of the jaw tumors depend on to certain on the dimentions, A small lesion is unlikely to be diagnosed on a routine examination. Exceptions are some early lesions that may present in conjunction eith a devitalized tooth, which is detectable on clinical examination. Some cyts may be secondary infected, leading to their diagnosis. Clinical absence of one or more teeth without the history of extraction may indicate odontogenic tumors. Many of these lesions are associated with impacted teeth or congenitally missing teeth. Enlarging tumors between two teeth can cause the crowns to converge and the roots to diverge. Growth that is nearly undetectable visually may lead to difficulty with denture retention. As the tumor enlarges expansion of the bone may be seen directly. This is usually toward the buccal surface of the alveolar bone because this is the thinnest area and expansion occurs here most easily. Clinically evident expansion is often a late finding, especially in the lesions developing within the ramus or angle of the mandible or within the maxillary sinus. Long time evolution of the benighn neoplasmsmay cause destruction of the bone that can result to mechanical airway obstruction and compromise digestive system and adjacent structures.

Tumors grow in the jaw, through the haversian system, without metastases but with a high probability of relapse. Some of these tumors may disappear spontaneously without any treatment.

The development of the tumors in children is more accelerated than in adults because of the growth potenthial of the child. Peculiarities of the evolution of the neoplams are related to development of the dental system (physiological change), the presence of growth areas in the maxillofacial bones, the presence of periods of growth and intensive development, the continuation of the differentiation processes of the tissure, the accelerated metabolism. The pains caused by tumors are often confused with the dental ones. The dental extraction, can accelerates tumor growth.

Sometimes neoplasms (osteoma, fibrous dysplasia, cementoma) evolve slowly, quietly, insidiously and without pain. Other neoplasms (osteogenic sarcoma) evolve very rapidly, infiltrating the neighboring tissues, ulcerating the mucosa and skin, with metastases and deadly end. Initially the clinical manifestation of the chondroma, the ameloblastic fibroid is poor (slow growth, painless) and in advanced stages they have a malignant evolution. In a short time, they reach large size, they invade the soft and metastatic parts in the organs.

In children, benign and malignant tumors have different aspects. Thus, desmoplastic fibroma, myxoma, lithic form of myeloplax tumor, fibrous dysplasia with proliferation are not included in the notion of benign tumors neither clinically, radiologically nor morphologically, especially in infants. K. A. Moskaciov (1961) wrote: „The clinical and histological criteria of malignant processes in children are relative. Often tumors confirmed histologically as benign develop clinically malignant". In this connection, the notion of intermediate tumors, characteristic only for children, has emerged.

5.3. Classification of the tumors of the maxillary bones

Jaw tumors in childhood show considerable differences than in adult. These differences refer to the spectrum of diseases. But when the

diseases are similar, there are sometimes differences in their clinical behavior.

Tumors of the head and neck represent only 2 % to 5 % of all pediatric tumors. Odontogenic tumors in children constitute approximately 3 % of all tumors like growths in the oral cavity, jaw and salivary glands in all age groups. But now day characteristics and epidemiology of jaw tumors have been described mostly in adults. Compared with their adult counterparts, jaw tumors in childhood show considerable differences. In the same time the incidence and prevalence of the tumors has been increasing in recent years, and it remains a significant cause of morbidity and mortality in this population.

Classification of jaw and facilal tumors in children will necessitate further modification and subsequent changes in the classification system according with new findings of new genetic and molecular changes. However, tumors can be classified as primarily originating in the facial bones or as metastases to the facial skeleton. Tumors arising within the jaws can be further subdivided into odontogenic and non-odontogenic.

There are almost 50 classifications of tumor processes and bone dysplasia. It is recognized that the classification of tumors after WHO (1972, series N 6) is based only on morphological principles (cell differrentiation, character of intercellular substances). Primary jaw tumors are broadly classified into odontogenic and nonodontogenic geoups. The World Health Oganization (WHO) classified this group of lesions in 1971 and 1992. IN 2005, the WHO published the latest updated edition of classification of jaw tumors. Classification of the pediatric jaw tumors include various classification system proposed by authors. And are enumerated below:

Jaw Tumors in Children

1. Classification of non-odontogenic jaw tumors in children:

I. Benign mesenchimal tumors: a) Giant cell lesions; b) Fibro-osseous lesions; c) Myxoma

II. Hematopoietic and reticuloendothelial tumors: a) Langerhans cell histiocytosis; b) Burkitt's lymphoma; c) Lymphoma

III. *Neurogenic tumors: a) Neurofibroma; b) Neurilemmoma; c) Neuroma; d) Ganglioneuroma; e) Neuroblastoma; f) Melanotic neuroectodermal tumor*

IV. *Vascular lesions: a) Vascular malformation (capillary, lymphatic venous, arterial, combined); b) Hemangioma; c) Aneurysmal bone cyst*

V. *Malignant mesenchymal tumors: a) Osteogenic sarcoma; b) Chondrosarcoma; c) Fibrosarcoma; d) Ewing's sarcoma*

VI. *Malignant epithelial tumors: a) Squamous cell carcinoma; b) Mucoepidermoid carcinoma; c) Adenoid cystic carcinoma; d) Adenocarcinoma*

2. ***Classification of odontogenic jaw tumors in children***

I. *Epithelial tumors: a) Ameloblastoma (Peripheral, Unicystic, Solid, Multicystic); b) Adenomatoid odontogenic tumor; c) Calcifying epithelial odontogenic tumor*

II. *Mesodermal tumors: a) Cementoma; b) Periapical cemental dysplasia; c) Cementifying fibroma; d) Cementoblastoma; e) Odontogenic fibroma*

III. *Mixed tumors: a) Ameloblastic fibroma; b) Odontoma*

3.Classification of Small Round Cell Tumors in children

a. *Soft tissue Rhabdomyosarcoma: a) Soft tissue (extraosseous) Ewing's sarcoma; b) Hemangiopericytoma*

b. *Osseous Ewing's sarcoma; Small cell osteosarcoma; Mesenchymal chondrosarcoma; Hemangiopericytoma of bone*

c. *Neural: Neuroblastoma; Peripheral neuroectodermal tumors (Askin's tumor, Neuroepithelioma); Pheochromocytoma*

4.International classification of childhood cancer

Malignant Bone tumors: a) *Osteosarcomas; b) Chondrosarcomas; c) Ewing tumor and related sarcomas of bone; d) Other specified malignant bone tumors; e) Unspecified malignant bone tumors*

Soft tissue and other extraosseous sarcomas*:*

a) Rhabdomyosarcomas; b) Fibrosarcomas, peripheral nerve sheath tumors and other fibrous neoplasms; c) Kaposi's sarcoma;

d) Other specified soft tissue sarcomas; e) Unspecified soft tissue sarcomas.

5.Classification of the Latest Benign Fibro-Osseus Lesions of the Craniofacial Complex

a. Bone dysplasias

Fibrous dyspla: A) Monostotic; (Polyostotic; Polyostotic with endocrinopathy (McCune-Albright); Osteofibrous dysplasia; b) Osteitis deformans; C) Pagetoid heritable bone dysplasias of childhood; D) Segmental odontomaxillary dysplasia

b. Cemento-osseous dysplasia: *A) Focal cemento-osseous dysplasia; B) Florid cemento-osseous dysplasia*

c. Inflammatory/reactive processes: *A) Focal sclerosing osteomyelitis; B) Diffuse sclerosing osteomyelitis; C) Proliferative periostitis*

d. Metabolic Disease: hyperparathyroidism

e. Neoplastic lesions (Ossifying fibromas): *A) Ossifying fibroma; B) Hyperparathyroidism jaw lesion syndrome; C) Juvenile ossifying fibroma*

In the practice of oncostomatology, this classification does not include tumors that are found only in the bones of the jaws,for example: true myxoma, fibroosteoma, cherubism, etc.

Ideal would be the classification according to the etiological principle. However, at the contemporary stage of science development, very little is known about the etiopathogenesis of tumor processes in children. Currently the WHO classification includes two MGKO series: - „Histological classification of odontogenic tumors, maxillary cysts" (series 5) and „Histological classification of primary bone tumors and pseudotumoral processes" (series 6).

5.4. Clinical diagnosis

Diagnosis of pediatric tumor is a big challenge because of peculiarities of behavior and physiological changes that take place in the maxillofacial area. Pediatric tumors in maxillofacial bone can be observed casually or after the appearance of nonspecific symptoms. Pathological changes in the jaws and facial bones are perceived by children as

normal and they can't appreciate their subjective state, making them to be diagnosed in late period. Often pediatric tumors are usually observed by parents when facial asymmetry appears. Facial deformity and functional impairment may be the result of asymmetric facial growth related to the lesion itself or to its treatment. In order to make an early diagnosis of the tumors, physical examination must be done, thorough medical hystory, inspection, palpation, percussion, etc.

Medical history. The first signs of pediatric tumors often are missed. Anamnesis plays an important role in the early diagnosis of jaw and facial bone tumors. Parents usually are first who can detect the tumor but in a very late period when facial asymmetry, deformation of the bone, dental mobility and trismus in temporo mandibular joint, of different degrees, with or without pain. Often, the swellimg are connecting with traumas by parents.

Pediatric tumors have poor clinical symptoms. The pain is earliest clinical sighn of malignant tumors of the jaw and facial bone, leaving the shape of the bone unchanged. Pain can be spontaneous or may be caused by some factors. Pain is irradiated or localized, diurnal or nocturnal etc.

Clinical manifestation of benghin tumor is non-specific and include swelling, facial asymmetry, induration, pain, paresthesia, bleeding, or increased tooth mobility. Due to the slow growth rate, most lesions do not cause pain.In the later stage of development the benign tumors lead to maxillary deformities, changes in dental occlusion, abcent of the teeth, disorders of nasal breathing, pathological fractures (the lithic form of osteoblastoclastoma). Exception is the osteoma-osteoid, in which the pains are neurally torturous, in the absence of external changes.

For the child's body, primary tumors in the maxillofacial area and, in very rare cases, metastases, as opposed to adults, are present, in which metastases of the maxillofacial bones are present in 20 % of cases.

The age of the patient is of utmost importance. Bone tumors have a predilection for specific age groups, and while some are more often seen in younger individuals, others are more commonly encountered in older patients. However, ages should be regarded as approximate, as there can be exceptions Pediatric tumors and like tumor lesions of the maxillofacial bone can appear in each period of development but some of them are

characteristic for some age. For example, eosinophilic granuloma occurs usually during the preschool period, and fibrous dysplasia - at the age of 10-12 years. Hemangioma of the maxillary bones and odontoma were detected at the age of 7-12 years. Ewing sarcoma, osteosarcoma more often is seen in younger individuals 10 - 20 - year-old. Osteochondroma, osteoid-osteoma affect periods of 10-30 years.

Inspection. The general condition of the child in benign tumors and like tumors lesions is not oltered, even in the advanced development stages. The clinical picture in malignancies (reticular sarcoma, Ewing's sarcoma), especially in infants, often develops acute infection (febrile condition, leukocytosis, increased erythrocyte sedimentation rate (ESR), similar to those of inflammatory origin (osteomyelitis). Anemia and cachexia characterize jaw bone sarcomas in advanced stages. In malignant lesions, gradually progressingindurated swelling occur. The vascular drawing becomes accentuated, the veins dilated, the cyanotic skin, slightly edematous.

Any swelling of the maxillary bones indicates the presence of a tumor. Some lesions can appear on the gingiva or alveolar process and presents itself as a pediculated or sessile lesion. During the mixt dentition some kind of lesion is like a marginal periodontitis which involve the gingival papilla and its look like reddish, blooming tumor, or blackberry appearance, soft and bleeds. Sometimes is smooth swelling covered by a pink mucosa, and has firm consistency.

Deciduous dentition may be spontaneously shed prematurely, absence of one or numorous teeth, displacement and lack of eruption. from the periosteum or from the superficial bone layers are observed in the early development stages. Later in the deep regions, especially those located at the maxilla, are detected. Hard consistency, round and well msrginated verrucous surface and surrounding borders are signs of benign tumor lesions. The cystic lesions of the jaws in children, unlike adults, do not manifest by thinning of the cortex plate (Dupuytren symptom) and depressibility, like a celluloid ball, as a result of bone elasticity.

Cortical plates of the bone and overlying mucosa or skin are almost invariable intact in the benign tumors, in the early stage Binghn tumors

of the jaws can be associated with the skin pigmentation over the trunk, vary in size and shape, and are of brownish coffe color.

The intensive growth of benign tumors, after a period of stability, warns their malignancy. Some benign tumors (myeloid neuroectodermal tumor of the suckling, desmoplastic, ossifying and ameloblastic fibroma, myxoma, lithic form of osteoblastoclastoma) are rapidly evolving, injuring the bone and invading the soft limb, such as malignant tumors. In infants, shortly (up to 2 months), malignancies exceed the size of the jaw, have an explosive evolution, invading the soft parts.

At *the palpation* of the neoplastic lesion, the local temperature, the skin hyperesthesia, the increased sensitivity, in comparison with the unaffected part are determined, the location, the extent, the shape, the appearance of the surface, the consistency, the mobility, the relations with the neighboring planes and the bone plane. The hard consistency, the tuberous surface and the clearly limited borders characterize a benign tumor process, as opposed to inflammatory processes, in which inflammation is lost in the unaffected soft parts.

The buccle and dentistry examination, by inspection and palpation of the mucosa of the oral cavity, is performed not only in the affected areas, but also throughout its surface - the general examination of the dental system. If changes are observed, the degree of extantion and their clinical feature are determined. Thus, the unique erosions present on the mucosa of the oral cavity may reveal a tumor process, a chronic trauma or a disease caused by specific infection (tuberculosis, syphilis). The tongue may have foul deposits, which need to be removed, to observe the pathological changes. The palpation of the tongue is done by grasping between the fingers and highlighting the tumors located in the depth. Then, the number of anomalies, shape, structure and positions of the teeth, dental malpositions, deviations, inclinations, dental mobilities, sensitivity to percussion and to thermal and physical agents are noted. Tooth position abnormalities are characteristic for both benign and malignant tumors. Most often, position abnormalities are present in sarcoma, osteoblastoclastoma, chondroma, fibrous dysplasia, eosinophilic granuloma, ameloblastic fibroid.

Dental mobility occurs in late periods of tumor development, with changes in bone structure, determined clinically and radiologically. Sometimes, dental mobility is a symptom of benign tumors. The changes in the periodontal disease and in the tumor lesions are differentiated.

High-frequency, clean percussion tonality characterizes healthy bone tissue, and low and deaf frequency - bone pathological changes.

The mandibular movements in full volume in the three planes show that the temporal-mandibular joint is not affected. The trismus of the temporo-mandible joint of the mandible, accompanied by pain or lateral-deviations, highlights the changes caused by the extension of the tumor from the bones of the jaws in the temporal-mandibular joint and the late periods of tumor development. The limited mandibular movements and accompanied by pain involve the localization of the tumor near the joint or masticatory muscles. Tumors located in the coronoid procces generate trismus during the early stage of development of the tumors. Functional changes of the temporal-mandibular joint are more pronounced in primary malignancies of the jaws. They are evident when the tumor invades the pterigo-mandibular space. Benign tumors, even if they are located near the temporal-mandibular joint, do not alter the function of the mandible.

5.5 Radiographic and morphological study

The primary goal of radiographic assessment is to more precisely define the primary lesion and to detect metastatic disease for clinical staging. Radiologically, benign tumors have specific general criteria: well-defined contour, blowing and thinning of the cortical bone, but not interrupting it, growing without periostal reaction, changes of structure (lithic, osteosclerotic, mixed), the tumor pushes the soft adjacent structures, but does not them infiltrate, the structure of the surrounding bone tissue is normal.

Chest radiographs are useful screens for mediastinal limphadenopaphy. Ultrasound is able to differentiate a solid from a cystic mass and give general relationships of the mass to adjacent structures. Axial and coronal computerized tomography allows documentation of bone erosion and invasion of adjacent structures and completes the information regarding the benignity criteria of the tumor. Magnetic resonance

imaging detects the belonging of the lesions, determines the degree of vascular implication, the benignity and malignancy of the lesions and offers improved tissue contrast and definition. Angiography delineates the blood supply to a lesion and offers the ability to embolize specific factors to decrease blood loss associated with excision of vascular lesions. Bone scans and liver spleen scans offer modalities to detect systemic disease.

Symptoms of progression of malignant neoformations. As the tumor progresses, general and local clinical symptoms appear. Tumors located in the maxilla are the causes of unilateral exophthalmia; intracranial and intraorbicular compression conditions the edema of the orbital nerve. Invasion of the malignant tumor into the median wall of the nose and nasal cavity causes difficult nasal breathing and serous elimination. The tumor invasion into the parotid gland is manifested by paralysis of the facial nerve. Tumors that have extended from the bone to the soft tissue (skin/oral mucosa) usually are extremely large in size and result in erosions on the skin or oral mucosa.

Dental pain become more pronounced, dental mobility occurs, pathological root dental resorption is manifested, and an important radiological sghins is the location of the dental roots in the tumor mass. In the malignanr tumor the general condition of children is altered. They complain fever of 38-39 ºC, hypochromic anemia, cachexia, metabolism disorders, sometimes psychological disorders.

Radiological diagnosis. Most diagnostic information as to tumors type comes from clinical assessment plus the plain films. Radiological examination is the most informative method of diagnostic of bone tumor. The radiological changes that are required to be analyzed on the radiography of a skeletal part are those of shape, size, contour, position, structure.

The tumor lesion of the bone are the consequence of a disorder in the normal physiology of the bone tissue being the results of breaking the balance between osteoforming and osteodestructive process. Osteclasts resorbtive activity on both cortical and cancellous bone surfaces lead to bone destruction.

Knowing the type of bone destruction is helpful for determining tumor aggressiveness. Other findings such as age, tumor location, tumor matrix, periosteal reaction, and soft tissue involvement are important aspect to establish diagnosis of bone tumor.

Most notable are the margins of the lesion. The relative biological aggressiveness and the rate of tumor growth can be appreciated by the presence of schlerotic margins (white rim) around of the destruction. Well defined, sclerotic margin refers to benign, slow-growing disorders (bone cyst, non-ossifying fibroma, enchondroma). Lesions without sclerotic margins refer to sharply defined edges with normal trabeculae up to the edge of the lesion but totally removed along along a plane of contact between the tumor and normal bone (giant cell tumor). Ill-defined margins is a focall destructive but locally infiltrative, creates a wider zone of transition at the margin of the lesion less well-defined with a complete cortical penetration (fibrosarcoma, chondrosarcoma).

The most aggressive lesions have a characteristic appearance called „moth eaten" or „permeative" – Ewing's sarcoma, osteosarcoma, angiosarcoma, high grade chondrosarcoma). Cortical destruction is a frequent finding in bone lesions. However, it is not very useful in differentiating benign versus malignant lesions. Complete cortical destruction may be present in high-grade malignant lesions like osteosarcoma, Ewing sarcoma and also in locally aggressive benign lesions. Ballooning is a particular type of cortical destruction, involves the destruction of inner cortex and new bone formation outside cortex at the same time. Seen in giant cell tumor (locally aggressive expansile lesion with cortical destruction, a wide zone of transition, interrupted new bone formation peripherally) and in case of chondromyxoid fibroma (well defined, expansile lesion with regular destruction of the cortex and uninterrupted new bone formation). A group of small cell tumors involving marrow like Ewing's sarcoma, lymphoma, small cell osteosarcoma can spread along entire Haversian canals without cortical destruction.

Periosteal reaction is one of the tools for cathegorizing tumor aggressiveness. Formation of bone tissue from the periosteum (solid layered or single-layered) demonstrate benign bone growth. Depending

on the aggressiveness and the time when pathological process start, the periosteal reactionscan be multiple-lamellated appearance, also known as „onionskin appearance," suggests an intermediate aggressiveness or transformation benign to malignant tumor. The most aggressive periosteal appearance is spiculated „hair on end" or buttress that means malignant tumor.

There is several type of bone destruction in scheletal tumor: osteoporosis, osteolysis, pressure atrophy and osteonecrosis.

Osteosclerotic lesions is a pathological modification of the normal turnover with excessive osseous deposition. May be found around the roots of the tooth like painless process, radiopaque area around a tooth, usually molar or premolar, is found during routine radiographs.

Osteoporosis (demineralization) consists in decreasing the amount of phospho-calcium salts in the skeletal part, which means low bone mass with alteration of the microarchitectural deterioration of the bone tissuer. Usually appear in systemic skeletal (Paget's osteodystrophy)

Osteolysis – is a progressive destruction of the mineral and protein composition in a certain bone area. This phenomenon is the result of a rapid demineralization process, which favors osteoclastic resorbtion, followed by the development of granulation tissue. The radiological aspect is characterized by areas of bone resorption of variable dimensions, with a well-defined or ill-defited contour. Occasionally, this condition is called „soap balloons" or „honeycomb". Osteolysis occurs in bone cysts, benign lesions (adamantinoma), where the bone is replaced with pathological tissue (pus, granulations, tumor tissue).

The atrophy is the consequence of a great pressure exerted on the bone. High pressure causes at the site of action an edema with the installation of demineralization. As a result of this phenomenon, the bone becomes plastic, incurving. At the same time, at the periphery, the premises of a compensatory osteosclerosis process appear. Under the action of tumor formations (hemangioma, fibroma, sarcoma), pressure atrophy of hard tissues occurs.

Hyperostasis, exostosis, oedostosis show changes in the shape of the bone. Hyperostasis defines a segmental or total thickening of a bone.

Exostoses defines as the benign growths of bone extending outwards from the surface of bone, having the same morphological elements as the normal bone.

Oedostosis represents the deformation of the bone by pushing the compact and the cortex of the bone by a structure modification with slow development „blown bone". The appearance of „blown bone" is found in the cystic tumor and benign tumors with mandibular localization (mandibular adamantinoma, myeloplax tumor, mandibular fibroma, etc.). At the level of the mandibular branch, the appearance of „blown bone" is manifested by increasing the diameters, by reversing the curvature of the sigmoid incision, by rounding the tip of the coronoid process and the gonion, by thinning the cortex in the vicinity of the area. At the level of the body of the mandible, the „blown bone" is characterized by thinning and swelling of the inferior margin of the bone and disorientation of the tooth axis.

Calcification or matrix mineralization are essential features to differentiate bone tumors. There are two types of matrix mineralization. The chondroid matrix presents in cases of cartilaginous tumors like enchondroma, chondroblastoma, chondrosarcoma and presents as a ring and arc, floccular, stippled, or popcorn-like. Osteoid tumors demonstrate the osteoid matrix. Trabecular ossification pattern in case of an osteoid matrix and cloud-like bone formation in case of osteosarcoma. Chondral calcification manifests as „rings-and-arcs", flocculent or „popcorn-like" calcifications. Osseous matrix has a „fluffy" or „cloud-like" aspect and is poorly defined. Fibrous tumors have a characteristic „ground glass appearance"

Changes in size are usually the consequences of developmental changes and are classified into: aplasia, hypoplasia, hyperplasia, dysplasia. Aplasia is the absence of one or more skeletal parts. Hypoplasia - the existence of a bone with small diameters, but with harmonious development. Hyperplasia appears normal in form and structure, but with all diametersincreased. Dysplasia is a group of changes in the size of the bones, the consequence of disorders of the enchondral or periosteal ossification.

Large bone volumes create osteogenic osteosarcoma. Tumor proliferation may be caused by endoosteum, odontoma, included tooth. Reactive condensation also occurs in dystrophic and tumor processes. Homogeneous radiotransparency, constantly bordered by a calcareous border, which clearly delimits the formation of the uninjured sponge, are found especially in cystic pathology (root cysts, pericoronary cysts, follicular cysts or fissure cysts). The intra-cystic inflammatory processes give the cystic image less clear boundaries.

A Codman triangle refers to an elevation of the periosteum away from the cortex. This term is classically associated with an osteosarcoma, but is not limited to this as any number of malignant or benign processes can cause this feature.

The tumor lesion may have a homogeneous or non-homogeneous structure. The homogeneous structure characterizes by a single intensity and it is found in benign bone tumors (fibroma, myxoma, cysts), and in some malignant bone tumors (osteolytic osteosarcoma, epithelioma, Ewing's sarcoma).

The radiographic images with a non-homogeneous structure have several shades of different hues. Homogeneous osteosclerosis is characteristic for osteoma, odontoma, cementoma, and inhomogeneous osteoporosis is found in cementoma, Paget osteodystrophy, osteogenetic osteosarcoma.

The appearance of *cumulus cloud* is found in the mandibular adamantinoma of a unilocular form. Radiologically, we find a homogeneous radiotransparency, with the outer limits consisting of united circle arches, which give them gibbous, convex, round marginal images, between which sharp incisions with the tips oriented towards the inferior of the radiotransparency are required.

The appearance of the *heart* or *apple* is characteristic for the incisive cysts. The radiotransparency is located on the midline, under the nostrils, and represents a medial upper groove, determined by the anterior nasal spine and an inter-incisive extension.

Morphological examination. The biopsy is the only method that can specify the diagnosis of tumor. Biopsy of jaw tumors allows histologic evaluation of the mass. Excisional biopsy is often therapeutic as well as

diagnosric. Incisional biopsy is required in cases where the lesion is large, or the lesion is relatively inaccessible. Fine needle aspiration for cyrologic study is useful in salivary gland and thyroid gland lesions.

5.6. The odontogenic cysts of the jaws in the primary and permanent dentition

The pecularities of onset, evolution and treatment in relation to age. Odontogenic cysts are pathological cavities or sac, with epithelial-lined wall and surrounded by fibrous connective tissue, filled with fluid or semi-fluid material. Originate from odontogenic tissues they occur in tooth-bearing regions of maxilla and mandible. Odontogenic cysts have developmental or inflammatory origins.

Classification of odontogenic cysts of the jaws in children and adolescents (WHO, 1974). Maxillary cysts are classified into three types: 1) a) odontogenic developmental cysts (dentigerious, primordial, eruption, gingival, odontogenic keratocyrs, calcifyied epithelial odontogenic cyts) and odontogenic inflammatory cyts (radicular, residual, inflammatory periodontal cyts); 2) non-odontogenic cysts a) nasopalatine, nasolabial, median palatine, globulomaxillary, median mandibular; 3) pseudocysts (solitary bone cyts, aneurismal and hemorrhagic cyts).

Depending on the size cyts are devided in three types: small cysts - up to 1.5 cm in diameter, inscribed within the limits of the alveolar process; medium cysts - up to 2.5 cm in diameter, extended into the mandibular body; large cysts - over 2.5 cm in diameter, deformed.

The physiological peculiarities of development of the dentigerous cyst *system in children*. In children 3-4 years of age, the dental buds are located in the region of the root bifurcation of the primary teeth.

At 6-7 years, during the period of intensive growth of the child, the dental buds migrate to the root apex of the primary teeth. The degree of displacement may be different, in relation to the presence of chronic apical infection of the primary teeth. The apical root cysts develop an internal pressure, which can moves the dental buds, lateral and inferior, towards the inferior margin of the mandible.

At 8-12 years, refer to the period of development of permanent dental buds. The growth of the jaws becomes slow; the period of tissue

differentiation begins. With the development of the dental root, the dental buds move towards the alveolar process, realizing the root resorption of the primary teeth. Correspondingly, they migrate to areas of the rarefied alveolar process, causing bone formation on the lateral side of the alveolar process. The formation of the root cyst at the apex of the primary teeth during this period leads to the inclusion of the dental bud in the cystic cavity. The development of the dental buds does not allow the growth of the cystic cavity in the body of the mandible. Predominantly, it develops in the vestibular, displacing the lateral cortex.

In pediatric dental practice, most often the inflammatory cysts from primary and permanent teeth are encounteredof all the odontogenic cysts. They are due to the pulp necrosis, which extends into the periapical tissues, causing the formation of the apical infection. Normally, around the roots of the teeth are present the Mallasez epithelial (embryonic epithelium). These cells remain inactive for a long time. Because of the chronic apical infection, their potential of multiplication retains. Under the harmful factors (contamination, the action of chemical agents in cases of conservative treatment, dental trauma, etc.) they begin to proliferate.

Inflammatory cysts from primary teeth are found in 73 % of cases. 90 % of them manifest themselves after repeatedly endodontic treatment. Cysts are located 3-4 times more often in the lower jaw than in the upper jaw.

At 12-16 years, the frequency of odontogenic cysts from permanent teeth reaches 20 %, located more often at the maxilla. The cause of root cysts toan be ophendening tooth and trauma.

7 % of all cysts belong to the dentigerious cysts of the permanent teeth, which are characterized by the presence of the dental crown of the permanent tooth in the cystic cavity. At the same time, the root system may be at different stages of development, sometimes with fully formed roots. Most often, the cysts are located in the region of the upper canines and premolars and the lower molars.

The clinical picture of periapical (radicular) odontogenic cysts are common, inflammatory derived from rests of Malassez, usually is

associated with a partially necrotic or necrotic tooth and involves a portion of the root, usually with loss of continuity of the radiographic lamina dura. Periapical cytsare determined by some peculiarities of the child's development. Fast growth of cysts in children is due to intensive metabolism processes, flexibility of the jaw bones and uneven growth of portions in different age periods.

The clinical picture of cysts of the jaw bone is similar to that of benign tumors and is characterized by the presence or absence of infection. In the first stage of development (intraosseous evolution), the cyst can be accidentally detected on the radiographic image. In the externalization phase, the first clinical sign is the deformation of the jaws. The intraoral examination shows a swelling on the vestibular area of the jaw, well defined, painless with a normal-looking mucosa. In the advanced stage of development, the cyst can lead to facial deformities but with the normal color of the skin. In late stage of development the maxillary cysts can spred into the maxillary sinus or nasal cavity, leading to the functional desorders (hard nasal breathing).

Periapical cysts is lined by stratified squamous epithelium of variable thickness, often edematous or may have scattered ciliated cells,
particularly if inflammatory process erodes bone of maxilla and process perforates to sinonasal cavity.

The X-ray shows well-defined round or pear-shaped, unilocular, lucent lesions in the bearing part of the jaws. In the primary dentition the lessure is located near the root and may displace adjacent teeth, dental buds or cause intensive root resorption of the milk teeth. The associated tooth usually has a deep restoration or large carious lesion. Surgical treatment include extraction of the offendening primary teeth. Permanent teeth shoud be fiiled followed by apical resection.

The dentigerous cystt are slow-growing benign and non-inflammatory cyst (*Fig. 28*). The dentigerous cyst is the most common type of noninflammatory odontogenic cyst Follicular cystis considered to be developmental cyst formed by accumulation of fluid between the reduced enamel epithelium and crown of an unerupted tooth develops at the eruption of permanent teeth, histology shows stratified squamous epithelium.

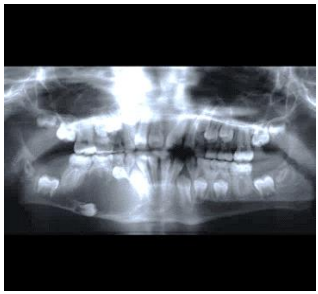


Fig. 28. Radiological picture of the dentigerous cystt.

The cyst wall is fixed to the included tooth package, surrounding its intra-cystic located crown. It is characteristic for children and adolescents. In 57 % of cases, the cysts are located in the mandible, in the premolar or molar area. Patients are typically pain free. The most important features of this cyst are its ability to expand asymptomatically and its potential to displace or resorb adjacent teeth or bone. In the dentigerous cyst, in the area of radiotransparency, the crown of the included tooth is visualized, the roots being initially extra-cystic.

At radiography, dentigerous cysts appear as well-defined, round or ovoid, corticated, lucent lesions around the crowns of unerupted teeth, usually third molars. The roots of the involved tooth are often outside the lesion and in mandibular bone. Dentigerous cysts can vary in size; cysts 2 cm in diameter or larger may cause mandibular expansion. Large jaw cysts expectedly result in pains, swellings, and other difficulties. Extremely large dentigerous cysts often develop ill-defined borders due to unevern rates of expansion through areas of varying bone density; the resulting radiographic appearance is comparable with that of a larger odontogenic keratocyst or ameloblastoma. In endosinus-growing cysts, the sinus appears veiled, as in odontogenic sinusitis. When there is no certainty of a cystic cavity, an x-ray with contrast substances can be made, which specifies very well the limits and the connections with the neighboring structures.

Pathological anatomy. The cyst wall is made up of two layers: external and internal. The external layer is made up of connective tissue cords, represented on the outside by a dense and fibrous, thin but quite

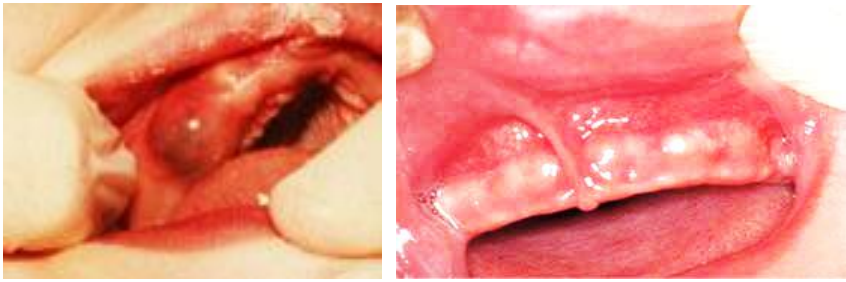
resistant tissue plane. The internal layer is made up of loose connective tissue, representing in its thickness a chronic inflammatory lymphocyte infiltration. The conjunctival wall is lined inside with non-keratinizedstratified pavement epithelium. The overinfection causes thickening of the cystic wall, its adhesion to the bone wall, and hyperplasia or destruction of the epithelium.

The cystic content is characterized by a sterile liquid, with characteristic serous citrine, filamentous appearance, containing cholesterol crystals with an intra-cystic pressure greater than the capillary pressure. The color of the cystic fluid may change: it becomes brown, if there has been an intrachysmal hemorrhage, or yellowish (purulent), if an overinfection has occurred.

The dentigerous cyst wall has the same structure, but is thinner. The best preserved epithelium can be malpighian stratified, with a smaller number of layers, sometimes cylindrical. Epithelial islands may be encountered in the thickness of the connective wall. Dentigerous cyst epithelium may keratinize or undergo ameloblastic transformation.

The differential diagnosis is made with osteoblastoclastoma, ameloblastoma, fibrous dysplasia, intraosseous fibroid, Ewing's sarcoma.

Eruption cysts**.** Associated with erupting primary and permanent teeth. Appear as bluish and translucent dome-shaped soft tissue lesions overlying an erupting tooth. Results from fluid accumulation within the space surrounding the erupting tooth. (*Fig. 29*)



a b

***Fig. 29*. a Eruption cysts. b. Gingival cyst of the newborn**

Gingival cyst of the newborn Gingival cysts of newborns occur as solitary nodules, located on the alveolar ridges Originate from remnants

of the dental lamina Histologically - true cyst with a thin epithetlial lining. The lumen filled with keratin but may contain some inflamematory cells, dystrophic calcifications, and hyaline bodies, No treatment is required for these lesions, (*Fig. 29*)

The principles of treatment of cysts in children is done according to the same principles as in adults. The objectives of the treatment of maxillary cysts are based on four principles: 1) removal or creation of conditions for the complete removal of pathological tissues; 2) preservation of healthy erupted teeth and those that can erupt; 3) the preservation of the neighboring structures, such as the vascular-nerve bundle, the integrity of the maxillary sinus and the nostrils; 4) restoration of the affected area as close to the normal contours.

Treatment consists of enucleation of the cyst lining. There are two type of surgical methods: cystectomy (complete enucleation - Partsch I) and uncomplete or marsupialization (Partsch II). The first one is indicated in small, medium and large uninfected cysts. Alveolotomy and cystectomy using Partsh II method is the most usual method of treating patients who have the diagnosis of follicular jaw cyst.

If the tooth buds are damaged during surgical enucleation, it can be put back immediately into the sochet. Dental buds, displaced by cystic pressure in young children, return to normal position within 3 months. In children aged 5-7, the provisional orthopedic appliance is indicated during the period until the permanent teeth erupt, in order to prevent deformities of the gum and restore the function.

The anevrismal bone cyst according to the World Health Organization defined as expansive osteolytic lesions consisting of blood-filled spaces and channels divided by connective tissue septa that can contain osteoid tissue and osteoclast-like giant cells. It is a rare benign lesion of the bone, which are infrequent in craniofacial skeleton. The anevrismal bone cyst is a highly destructive benign bone tumor, occurring in less than 1 in 100,000 people per year. The cause is unknown and can to grow in response to a disturbance of the blood vessels in the involved bone, or may grow because of a pre-existing tumor.

Anevrismal bone cysts are characterized by rapid growth with resultant bony expansion and facial asymmetry (exteriorization period). It is found in the mandible during periods of pre-puberty and puberty. The main symptoms are painless deformation and depending on its location include dull pain. headache, diplopia, loss of vision, proptosis, tooth mobility amd bleeding, hearing loss, etc (*Fig. 30*).

Radiographically, cystic bone expansion represent an radiotransparent image, multilocular, well-defined with irregular margins resembls honey-comb or soap bubble like inner structure. Sometimes destruction of the bone cortex and periosteal reaction can be observed.

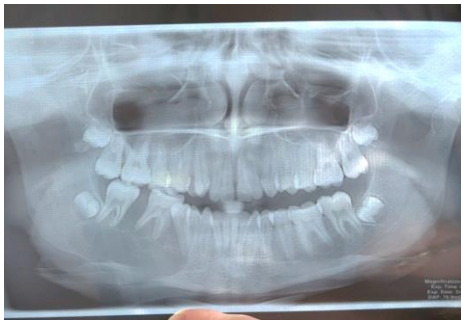


Fig. 30. **Anevrismal bone cysts**

Histologically, it is characterized by numerous spaces, of different sizes, filled with blood. The walls are made up of fibrous connective tissue, multinuclear giant cells and osteoid tissue; does not contain epithelium. The differential diagnosis is made with other unichameral cyst, ossifying fibroma, ameloblastoma, giant cell granuloma and with brown tumours.

It is applied the surgical treatment, consisting of cavities curettage.

Solitary bone cyst (traumatic cyst, hemorrhagic cyst, extravasation cyst, unicameral bone cyst, simple bone cyst, and idiopathic bone cavity) of the maxillofacial region is an uncommon pseudocyst and makes up about 1 % of all jaw cysts. The solitary bone cyst occurs in children especially during the period of intensive growth of the skeleton (12-14 years). The pathogenesis is unknown, but it is considered that during this period the bone marrow fails to grow in relation to the intensive growth of the bones,

consequently, the hemorrhagic cysts are formed. Some authors believe that they are the result of bleeding caused by dental trauma.

Most solitary bone cyst present no clinical symptoms of swelling or other functional signs and are found during panoramic radiographic examination. Pain is the most frequent presenting symptom of jaw lesion, affecting 10 to 30 % of patients. Tooth sensitivity, fistulas, paresthesia, delayed tooth eruption, and pathological fracture of the mandible can occur. In the majority of cases, the pulp of the teeth in the involved area is vital, and swelling or rare pain may be the presenting complaint. When the cystic cavity is opened surgically, it is found to contain either a small amount of serosanguinous fluid, shreds of necrotic blood clot, fragments of fibrous connective tissue or nothing but a naked, raw, and empty bony cavity without an apparent cystic lining

Histologically, solitary bone cyst represent lacks an epithelial lining Radiographically - a unilocular, well-defined radiolucent image, located in the teethbearing part of the mandible, between the canine and molars, above the mandible canal or in the chin region.

The differential diagnosis is made with root cysts, osteoblastoclastoma, ameloblastoma. The treatment consists in the curettage of the lesion.

5.7 Nonodontogenic cysts

The nasopalatine duct cyst (incisive canal cyst) is an extrusive cyst, located in the midline of anterior maxilla near the incisive foramen. It arises from the remnants of epithelial of nasopalatine duct, the communication between the nasal cavity and anterior maxilla in the developing fetus. probably under the action of chronic irritations. It belongs to the category of cleft cysts. It is one of the most common nonodontogenic cysts of the oral cavity occurring in about 1 % of the population

Clinically is asymptomatic lesion being detected on routine radiograph picture. The crown of central incisors can be displaced labially, mesially or oraly. Often, nasopalatine duct cyst is associated with an infection of a previously asymptomatic nasopalatine duct cysts and consist of swelling on vestibular, palatal or concurrent ways on both sides, drainage, and pain. The vitality of nearby teeth should not be affected;

however, it is not uncommon to see evidence of endodontic therapy because the nasopalatine duct cyst was previously clinically misdiag nosed as a periapical cyst or granuloma.

On the maxillary occlusal radiograph showed a well-defined round, oval or heart-shaped radiolucency lesion, in the midline and between the central incisors. The lesion can cause displacement or resorbtion of the roots of the incisors.

Cyst formation is due to spontaneous cystic degeneration of residual ductal epithelium. The vast majority of cases contain non-keratinized stratified squamous epithelium alone or in combination with other epithelia histologically. Approximately 30 % cases contain respiratory epithelium. The content of the cyst is yellow, with no cholesterol.

The differential diagnosis is made with the periapical cyst or odontogenic subperiosteal abscesses, schwannoma in the incisive canal regione, the nasal furuncle, which has a rapid evolution, and at the examination of the nasal vestibule, the characteristic necrotic head are observed; superior salivary labial cyst, which evolves more towards the mucosa. Enucleation The surgical treatment is usually curative, and recurrence is rare.

The globulo-maxillary cyst develops between the maxillary lateral incisors and the canine teeth; it is considered a fissural cysts, originating from the persistent epithelial remains at the level of the junction between the maxillary and globular process. The globulo-maxillary cyst arise from epithelium entrapped during fusion of the globular portion of the medial nasal process with the maxillary process. The globular portion of the medial nasal process is primarily united with the maxillary process and a fusion does not occur, therefore, epithelial entrapment should not occur during embryologic development of this area.

Clinically, it evolves asymptomatically for a long time, being accidentally discovered following a routine X-ray that appear as inverted pear shaped well-defined radiolucency, extending upto the periapical region with displacement of roots associated with left maxillary lateral incisor and canine. In late period, usually swelling extend to the soft-

tissue of the maxillary anterior mucolabial fold, lateral to midline leading to obliteration of the nasolabial fold.

The wall of the cyts has nonkeratinized epithelial lining of cuboidal to stratified squamous cells, the vitality of the neighboring teeth, the lack of the tooth, the dental buds included in the cyst, the root divergence of the neighboring teeth. The cyst contains yellow liquid without cholesterol.

Surgical treatment include enucleation using intraoral approach under local anesthesia and adrenaline.

The differential diagnosis is made with the tumor with myeloplax and sometimes with ameloblastoma. The treatment is surgical enucleation. Before treatment, the vital test for the neighboring teeth is indicated.

The gingival cyst also known as Epstein's pearl *(Serra cyts)* is a dysentogenic type of cystsoriginates from the dental lamina. It is a superficial cyst in the alveolar mucosain newborns and usually are transient in nature. It can be seen inside the mouth as small and whitish bulge. It appears in children in the first year of life and is located on the alveolar process, in the form of small, whitish cystic formations, with exophyte growth, painless, of elastic consistency.

The cyst is lined by odontogenic epithelium which is covered by a thick layer of keratin, which gives the cyst its yellow color. The majority of these cysts break by themselves, a few days after birth, exuding the keratin. In some cases however, they may remain for a period of several months. In somecases surgical opening is indicated.

*The eruption cyst*also known as eruption is a form of soft tissue benign cyst accompanying with an erupting primary or permanent teeth and appears shortly before appearance of these teeth in the oral cavity. It is a soft tissue analogue of the dentigerous cyst, but recognized as a separate clinical entity. Usually appear on the mucosa of a tooth shortly before its eruption.

The exact etiology of occurrence of eruption cyst is not clear but it can be trauma, infection and the deficient space for eruption. The eruption cyst is located superficially around the crown of the erupting tooth. Clinically, the lesion appears as a dome-shaped lesions, fluctuant, often translucent swelling of the alveolar ridge over the site of the

erupting tooth. When the cystic cavity contains blood, the swelling appears purple or deep blue; hence, the term „eruption haematoma".

Histologically, presents the same microscopic characteristics as the dentigerous cyst, with connective fibrous tissue covered with a fine layer of non-keratinized cellular epithelium.

The treatment consists in excising the mucosa, which covers the crown of the tooth, after the tooth spontaneously erupts. Sometimes, only one incision of the mucosa is sufficient to evacuate the fluid.

Odontogenic keratocysts are rare benign cystic lesions involving the mandible or maxilla and arise from dental lamina. **Odontogenic keratocyst is a** developmental lesion but locally aggressive and tend to recur after excision. They originate from epithelial cell rests (stratified squamous keratinizing epithelium) found along the dental lamina and periodontal margin of the alveolus of the mandible Inflammation may impede histologic characterization. It's occurred predominantly in younger patients and is located in either the body or ramus of the mandible or maxilla. The keratocysts have a frequency of 6-8 % of the jaw cysts. The radiographic examination detects expansile well-defined solitary unilocular lesion extending longitudinally in the posterior portions of the mandible without being in contact with an included or erupted tooth. Clinically discovered incidentally but in late period symptoms associated with these tumors can be jaw swelling and pain. Less commonly, trismus and paresthesia may occur. Morphological examination reveals a thin-walled, friable cyst containing fluid and debris. Viscosity of the contents ranges from straw-colored fluid to purulent and cheese-like masses. Histologically, the cystic cavity delimited by a thin conjunctive membrane lined with stratified pavement epithelium is observed. There is an ortho- or para-keratinized layer, and inside the cyst – keratin on the surface of the epithelium.

Learning objectives

1. Classification of cysts in children (WHO).
2. Which cysts are most common in children and adolescents?
3. Etiology of cysts. Which cells are responsible for the formation of cysts in the jaws?

4. Clinical manifestation of eruption cysts in children.
5. Pathogenesisi periapical maxillary cysts in children.
6. Methods of diagnosis of cysts in children.
7. Differential diagnosis of the odontogenic and non-odontogenic cysts.
8. Clinical-radiological feature of follicular cysts in children.
9. Principles of treatment of cysts in children in primary and permanent dentitions.
10. Indications for various forms of cysts treatment.

Tests

1. C.S. Surgical treatment of infected root cysts in the primary dentition:
   A. cystotomy with apical resection of the tooth cause;
   B. antibiotic therapy concomitant with endodontic treatment;
   C. tooth extraction, followed by cystectomy;
   D. periostotomy concomitant with antibiotic therapy;
   E. no answer is correct.
   (C)
2. CS. The Serra glands are located on:
   A. mucosa of the alveolar apophysis;
   B. lower lip mucosa;
   C. upper lip mucosa;
   D. the occlusion line on the genial mucosa;
   E. the hard palate mucosa.
   (A)
3. CS. Clinically, the eruption cyst is located:
   A. superficially, around the crown of a tooth;
   B. at the level of the teeth 38, 48;
   C. superficial, around an erupting tooth;
   D. on the alveolar apophysis, at the level of the erupting tooth;
   E. all answers are correct.
   (D)
4. CM. Clinical signs of a follicular cyst may be:
   A. cyanotic mucosa at the level of the erupting tooth;
   B. jaw deformation in the region of a segment;

C. absence of a permanent tooth;
D. persistence of a primary tooth;
E. frequent inflammation.
(B, C, D)

5. CS. The radiological image of the dental cyst is characterized by:
A. the apex of the tooth is in the radio-clear area;
B. the crown of the permanent tooth is included in the cystic cavity;
C. the radio-clear image replaces a tooth that has not formed;
D. the radio-clear image has neighborhood relations with the teeth;
E. single- or multilocular radiotransparency.
(B)

6. CM. Neodontogenic cysts are:
A. dentigerous cyst;
B. globulo-maxillary cyst;
C. eruption cyst;
D. median palatine cyst;
E. aneurysmal cyst.
(B, D)

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6. Benign odontogenic and non-odontogenic tumors and tumor-like lession of the jaw in children and adolescents

6.1 Benign non-odontogenictumorsin children and adolescents

Tumors arising in the pediatric facial skeleton represent a formidable challenge in clinical practice because of the complex anatomy of the head and neck and because of facial development during this time span. Although rare, these conditions include a broad spectrum of lesions with a varying degree of malignant potential. Their true incidence is difficult to establish due to the paucity of reports in the literature and because most published articles areeither isolated case reports or series dealing with a specific histological diagnosis.

Symptoms of tumors in facial bone in children are often non-specific and include swelling, facial asymmetry, induration, pain, paresthesia, bleeding, or increased tooth mobility. However, due to the slow growth rate, most lesions do not cause pain. Biopsy are crucial for a successful multidisciplinary management. Multidisciplinary treatment requires a close collaboration between pediatricians, radiologists, medical oncologists, maxillofacial surgeons, radiotherapists, and pathologists. In addition, long-term follow-up is necessary in many pediatric cases because of subsequent facial deformity.

The osteoma is a benign osteogenic, slow-growing neoplasm that arises most frequently in the craniofacial skeleton. There is no predilection for age and can appear from 4.8 months to 50 years of age mostly occurring in young adults. In children they are very rare and form 2.4 % of the benign tumors of the facial bones. According to the location osteoma are categorized as central, peripheral, or extraskeletal. Osteomas of jaws may arise on the surface of the bone (peripheral, periosteal, exophytic, or parosteal) or they may be located in the medullary bone (endosteal or central). Generally it is asymptomatic

The pathogenesis of the osteoma is unclear but it has been considered to be a true neoplasm, developmental anomaly, or reactive lesion triggered by trauma, muscle traction, infection or can be caused by endocrine factors mesenchymal tumor, located mainly on the flat bones and at the level of the facial massif. It affects the young age. The radiological aspect: a zone of round or ovoid in shape well-circumscribed radiopaque masses (osteosclerosis),

Histologically, they can be compact osteoma, cancellous osteoma, and mixed osteoma. The bones grow very slowly and remain unobserved for a long time.

The peripheral osteoma (exostosis) is a prominent, rounded, well-shaped tumor, sometimes pediculate, hard, painless, of variable size. The mucosa that covers it is thin, without changes. In most cases, multiple exostoses appear, often symmetrical, especially on the internal face of the mandibular ramus. Large exostoses cause functional disorders. The peripheral osteoma is diagnosed at an early stage of development.

The central osteoma is detected in late developmental periods, usually during a prophylactic radiograph. It is encapsulated and produces displacement of surrounding tissues. Sometimes it reaches large dimensions, causing even pain, tooth displacement, deformations of the region.

The osteoma with localization in the maxillary sinus is a rare tumor form; it can be pediculate, but it is usually a massive, hard formation with a broad base on the sinus wall. The clinical picture in the advanced periods of development stems from facial deformities, exophthalm, neuralgia, diplopia, decreased vision, nasal breathing disorder. The peripheral osteoma is a sign always present in Gardner's syndrome, which is familial in nature.

The differential diagnosis of the osteoma is made with Paget's disease, myeloplax tumor, adamantinoma, odontoma and so on.

Patients with multiple osteomas affecting the mandible should be evaluated for Gardner's syndromeinpatients suffering from this syndrome is characterized by triad symptoms: colorectal polyposis (which may undergo malignant transformations), skeletal abnormalities, that is multiple osteomas, and multiple impacted or supernumerary teeth.

Osteomas are managed by surgical excision but it is indicated only in symptomatic patients.

The osteoid osteoma is a benign, osteogenic tumor, rarely encountered, occurring at an early age (between 5 and 10 years). It has small dimensions (up to 1 cm in diameter). It is located in the jaw. The main symptom is pain.

Osteoid-osteoma is a type of nidus which appeared as a hard osseous core composed of densely set trabeculae of newly atypical formed bone. Initial notable changes in the lesion is as an increased vascularisation and destruction with replacement by new atypical bone following resorption of the destroyed tissue. The stroma consisted of osteogenic connective tissue containing numerous blood channels. A cortical lesion which produced this bony replacement stimulated the overlying periosteum to lay down new bone of fairly normal architecture

Morphologically, it is characterized by a central area, composed of compact tissue, with different degrees of calcification, sprinkled with connective-vascular tissue.

The differential diagnosis is made with the myeloplax tumor, which is characterized by larger dimensions and results clinically without pain.

The treatment of osteoid osteoma consists in the surgical removal of the lesion.

The chondroma is a rare, benign bone tumor. In the facial bones, it involves anterior maxillary region adjacent to the nasal spine and nasal septum and in mandible, it involves symphysis, body, coronoid process and mandibular condyle. This tumor arises from remnants of the embryonal cartilaginous tissue that escapes resorption during endochondral ossificationand composed of mature hyaline cartilage, with areas of calcification or necrosis, containing microcysts with viscous filamentous fluid. It is found in children during preschool period. Clinically, it appears as a spherical nodular tumor, of firm consistency, with a moderate degree of elasticity, painless. It grows slowly, but it can reach giant size. It's a lithic tumor. The lysis area has variable shapes, contours well delimited by compact, which is thinned and blown, resulting in an oedostotic (blown bone) diaphysis. Within the lysis zone, several calcifications are highlighted.

In the mandible, the chondroma is located in the growth areas: the symphyseal region, the coronoid process, and in the maxilla - in the area of the incisor canal, the malarial proccess, in the palace or on the alveolar ridges. It may also develop from the nasal cartilage. Rarely, it is located

in the soft parts, where there is no cartilage: on the pharyngeal tonsils, tongue, parotid gland.

The peripheral location (echondroma), clinically, appears in the shape of a prominent hemisphere in the mouth cavity, well defined, of firm consistency. The skin and the covering mucosa remain unchanged and mobile. Reaching a large volume, it produces deformations and pain.

The applied treatment is surgical, the tumor being resistant to irradiation treatment. *Central Giant Cell Granuloma* (CGCG) first described by Cooper and Travers in 1818. Giant cell lesions of the bone are common in the long bones, and more rare in the facial bone. Central giant cell lesions of the jaw are relatively uncommon lesions that cause thinning and expansion of the cortical bone more destructive than reparative. Central Giant Cell Granuloma is a benign tumor of the jaw accounts for approximately 7 % of all benign tumors of the jaws and 2 - 12 % of all giant cell tumors of the body. Mostly, it have seen in children and young adults. Based on the clinical behavior Central Giant Cell Granuloma can be locally aggressive or non-agg aggressive. After treatment with curettage or surgical giant cell tumor of bone can occasionally recur.

The recent World Health Organization classification in 2013 defines a giant cell-rich tumor with histopathologic characteristics resembling with those of a central giant cell granuloma as a giant cell lesion of the small bones. The histopathologic features are fibrous tissue with hemorrhage, hemosiderin deposits, irregularly distributed giant cells, and reactive bone formation. Despite the similarity of the histopathologic findings, it is considered that this newly defined group does not include central giant cell granuloma of the jaw. In recent years, giant cell reparative granuloma has been classified as an osteoclastic giant cell-rich tumor (namely, giant cell lesion of the small bones), which comprises very rare tumor-like lesions consisting of fibrous tissue with hemorrhage, hemosiderin deposits, irregularly distributed giant cells, and reactive bone formation.

The ethylological factors can be trauma, inflammation, blood disorders. According to the pathogenesis of the tumor, they occur from the

tissue germs left in the embryonic life. The giant cell lesion of the small bones (central osteoblastoclastoma) *(figure 31)* is most commonly located in the mandible - in the region of premolars and molars, and in the maxilla - in the region of premolars. According to the pathomorphological picture, we distinguish two type of lesion: lithic and cystic forms.

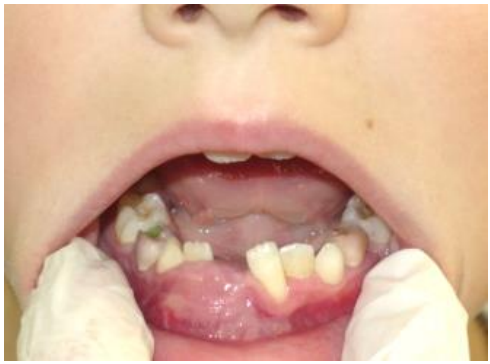

Fig. 31 **Osteoblastoclastom**

The cystic osteoblastoclastoma has a slow growth and it occurs more often at the age of 8-15 years. The cortical bone in the tumor region is blown and has ill-defined border.

The lithic type of giant cell lesion is local aggressive, can cause pain, resorption of the tooth, destruction of the bone, displacement of the erupted tooth, and germs of the unerupted tooth and pathological fractures. The invasion of the soft parts changes the color of the mucosa into purple or brown. The lithic type of giant cell lesion can be confused with osteosarcoma.

On the radiographic image the cystic type appears with polycyclic, well-defined contour the appearance of „honeycomb". Treatment of the giant cell tumors consist in surgical resection of the jaw, including removal of surrounding bone and nerves, followed by reconstructive surgery.

*The peripheral giant cell granuloma (*giant cell epulis, *epulis)* is the most common oral giant cell lesion. This condition is associated with two other diseases, pyogenic granuloma and peripheral ossifying fibroma. The lesion is not a true neoplasm, but rather may be reactive in nature. The development of epulis is favored by some general factors

(trauma, hormonal disorders) and caused by chronic irritation or inflammation caused by the dental deposition, chronic gingivitis.

Peripheral giant cell granulomas are seen exclusively in gingiva appear as a soft tissue extra-osseous purplish-red nodule. They arise from periodontal ligament or periosteum. Usually is located between the first permanent molars and the incisors. It starts with gingival hypertrophy located at an interdental papilla or at the free edge of the gum and very rarely interdental. It grows slowly, insidiously, with a wide base of implantation or pedicle, causing dental displacement and dental mobility, or unerupted tooth and can cause resorption of alveolar bone.

Fibroblasts are the basic element of peripheral giant cell granulomas. Scattered throughout the fibroblasts are abundant multinucleated giant cells believed to be related to osteoclasts.

Differential diagnosis is made from a pyogenic granuloma. Although a peripheral giant cell granuloma is more likely to cause bone resorption than is a pyogenic granuloma. A biopsy provides definitive diagnostic results. Microscopically, a peripheral giant cell granuloma is identical to its central or intraosseous counterpart, the central giant cell granuloma. The differential diagnosis is made with odontogenic cysts, cystic adamantinoma, Ewing's sarcoma, eosinophilic granuloma, fibrous dysplasia, ameloblastoma and so on.

The treatment of tumors can involve radiotherapy, surgical removal or mixed. Surgical excision is the preferred treatment for peripheral giant cell granulomas. Removal of local factors or irritants is also required. Recurrences, which are seen occasionally, are believed to be related to lack of inclusion of periosteum or periodontal ligament in the excised specimen.

Intraosseous hemangiomas are one of the rarest lesion of jaw, comprises 0.5-1 % of all intraosseous tumors. Many authors believe it to be hamartoma which arises from the proliferation of mesoderm that undergoes endothelial differentiation which is further localized and vascularized.

World Health Organization considers it as a true benign neoplasm of vascular origin due to endothelial proliferation which differentiates into

blood vessels. The election localization is at the level of the mandibular angle and at the level of the mandibular body. Intraosseous hemangiomas is asymptomatic, sometimes may present discomfort, pulsation, bluish slow growing mass, compression of surrounding structures, mobile teeth, and hemorrhage. The correct diagnosis of cavernous hemangioma is relatively challenging due to similar radiologic features of ameloblastoma, odontogenic myxoma, fibrous dysplasia, and aneurysmal bone cyst. However, orthopantomogram, computed tomography and magnetic resonance imaging is available for a diagnosis purpose. Radiographic features of cavernous hemangioma has variable appearance and can present tube-like arrangement of radio-opaque striae, periphery shows well-defined or ill-defined corticated area with scalloped margin, or can reveals an area of altered radiodensity usually osteolytic with occasionally central radio-opaque areas and altered trabecular pattern similar to spokes of wheel, radiating from center to periphery, or osteolytic areas like a soap bubble or honeycomb-like appearance. Due to the presence of a variable degree of radiolucency cavernous hemangioma may be multilocular or unilocular and may give sunburst and tennis racket appearance also

A biopsy is not done due to a higher risk of hemorrhage. Histopathological examination revealed endothelial cells proliferate to form a plexiform pattern of vascular space. The thin-walled cavernous spaces are lined by a single layer of endothelial cells interspersed among bony trabeculae.

Treatment modalities for cavernous hemangioma include hemorrhage control, complete eradication of the lesion, and prevent recurrence. These include: noninvasive radiotherapy, intralesional injection of sclerosing agents and embolization, curettage and radiation, and resection followed by osseous reconstruction.

The fibroma of the bone is a benign tumor. There are *non-ossifyingand ossifying fibroma. Non-ossifying fibroma* occurs commonly in the long bones of young people (in children and adolescents) and rarely occur in the mandible or even more rarely in the maxilla. Most common localizations of nonossifying fibromas are situated in growth areas some authors regard them as localized growth-related disorders. That's why

there is controversial whether it represents a true neoplasm or rather a developmental disorder of growing bone. Nonossifying fibromas are not characterized by any specific symptoms, a fact which places special constraints on diagnosis and therapy. Ossifying fibroma is common in the maxillofacial bones.

Clinically, there are peripheral and central ossifying fibroma. The peripheral ossifying fibroma is located only on the alveolar ridge between the teeth (interdental papilla). The most common site is related to an upper incisor or upper cuspid tooth. Clinical features can be associated with red or pink in colour, a smooth or pebbled irregular surface, ulcerated or not, a broad base limpet-like attachment or less often on a short stalk (pedunculated), small, but very large lesions have been reported. The most common presentation of a peripheral ossifying fibroma is as a slow growing, ulcerated, red, outward growing lump less than 2cm in diameter in an adolesecent. It can reach a considerable size quite quickly, but there is often a delay of months or years before presenting for treatment, depending on the degree of discomfort, aesthetic appearance and development of ulceration. Peripheral ossifying fibroma is distinguished from other common lumps in the mouth such as pyogenic granuloma, oral irritation fibroma, giant cell fibroma or peripheral giant cell granuloma. The histology is characteristic with bone, cement or calcium deposits in cellular connective tissue. X-rays rarely show involvement of the underlying jaw bone.

The peripheral ossifying fibroma should be completely excised. Extraction of teeth is rarely required. In addition, any predisposing causes should be treated such as plaque or irritation from a dental prosthesis. Unusually for benign lesions, the recurrence rate is high, up to 20 %, occurring on average 12 months following initial excision. Therefore regular follow-up is required.

Central ossifying fibrom is the most common benign fibroosseous neoplasm of the oral and maxillofacial region. Often is located in mandibular premolar and molar areas. It grows slowly, expansively and concentric. The growth of the tumor may be stimulated by some favoring factors (trauma, infections).

Central ossifying fibroma usually presents clinically as a painless and expansive spherical or ovoid jawbone mass that may displace the roots of adjacent teeth and cause root resorption. Radiological picture of central ossifying fibrom demonstrates well defyned either completely radiolucent or a mixture of radiolucent and radiopaque appearance (depending on the amount of internal calcification). Histologically composed of proliferating fibroblasts and osseous products that include bone and cementum-like material.

At the upper jaw, (nasopharyngeal fibroma) histologically occur with relative frequency in the nasal passages and paranasal sinuses, and these are believed to be reactive rather than neoplastic. On occasions, the fibroma grows first in the nostrils, in the sinuses of the jaws, in orbit, and then can invade the soft tissue however, they may rich a considerable size, and even expand bone accompanied by mobility and dental displacements.

The differential diagnosis must be done with such as focal cementum-osseous dysplasia, osteoid osteoma, and fibrous dysplasia, ameloblastoma, fibrous dysplasia, chondroma. The treatment is surgical: the excision of the tumor within the healthy tissues. Odontogenic fibroma is located in the tooth-bearing areas of the jaws, and which sometimes contains epithelium of apparent odontogenic type, thus implying a possible odontogenic origin. Desmoplastic fibroma is a rare neoplasm of fibroblasts first described by Jaffe (1958) in which the predominant feature is the production of collagen. It does not metastasise, but may cause considerable local destruction and occasionally extend into soft tissues. Almost any bone, including the mandible and maxilla, may be involved, but it is most common in the long bones. Histologically, many collagen fibres are seen interspersed with small indolent fibroblasts and abundant hyaline intercellular substance. Giant cells are not a feature, and osteogenic activity is absent. Mitotic figures are rare. The lesion is overall much less cellular than the non-osteogenic fibroma.

The odontogenic myxoma is a rare tumor that occurs in the mesenchymal tissue of the mandible and maxilla and represents 3-6 % of all odontogenic tumors.In children, it can occur even less frequently than in

adults but tends to have the same pathologic and clinical features. Most patients are asymptomatic at diagnosis and typically only present with image findings of the tumor. However, the tumor can also present as a facial deformity with no associated pain or inflammation

The tumor is not delimited and has a tendency to invade the neighboring bone, and histologically it is formed of thin collagen fibers, round or polyhedral and rare cells, fragmented by odontogenic epithelium, located in an abundant mucoid stroma.

Clinically, myxoma is a destructive, invasive, fast-growing lesion, causing painless bone deformities, displacement or dental mobility. It has no preferential location on the jaw, may be associated with an included tooth It can reach large size and has a recurring tendency. Extensive-invasive growth along the spongy tissue can result in Vensan symptom.

The radiographic image has the appearance of unilocular with trabeculae and that is sometimes associated with tooth displacement. diffuse and irregular contour.

Macroscopically, myxomas are spherical or nodular tumors, gelatinous, transparent, of gray-yellow color, of varying sizes.

The differential diagnosis is made with the chondroma, cysts, ameloblastoma, etc. The histological examination definitively determines the diagnosis. There is no evidence-based consensus in terms of treatment of the odontogenic myxoma. Usually a large spectrum of surgical treatments is used, varying from conservative enucleation of the tumor to aggressive resections such as mandibulectomy and maxillectomy. *The melanotic neuroectodermal tumor of infancy* is a rare, local aggressive tumor of neural crest origin that usually develops during the first period of life. In 90 % of cases, it is located in the head region. It is a rare lesion, with localization, in 2/3 of cases, in the frontal region of the maxilla.

The histological profile demonstrates a biphasic cell population within a stroma of moderately vascularized fibrous tissue with large epithelioid, melanin-containing cellsare arranged in alveolar or tubular formations around clusters of small neuroblastic.

Clinically, the tumor appears suddenly, with the intramaxillary, paramedian location. It has a smooth surface, elastic consistency, and the covering mucosa with normal or pigmented appearance. The tumor causes displacement, early eruption and exfoliation of primary teeth. Due to the large size of tumor, it can cause eating, phonation, physiognomic disorders.

Radiographically, it does not have its own aspects. The image is osteolytic, more or less delimited. It evolves relatively quickly, but it is considered benign, with a reduced tendency for relapses. Computed tomography imaging typically reveals a well-demarcated, hyperdense lesion with contrast enhancement and hyperostosis of adjacent bone

A differential diagnosis would include other small round cell tumours occurring in infancy, such as rhabdomyosarcoma, neuroblastoma, melanoma and lymphoma.

The tumor must be removed completely and the patients must be under observation for a long time.

6.2 Benign odontogenic tumors in children and adolescents

The odontoma is a tumor arise from the hard dental tissue, of epithelial origin (enamel) and mesenchymal (dentine and cement). It is the most common lesion in children of odontogenic origin (from 4 % to 67 % of cases). The odontoma is a tumor of the young age, especially during the period of formation of the permanent dentition. The most frequently reported location is the incisor-canine area of the upper jaw (67 %) followed by lower-anterior and lower-posterior areas of the lower jaw (33 %).

Odontoma are classified taking into account organization and degree of alteration of odontogenic cells, there are two classifications: compound and complex. Compound odontogenic tumor exhibits morphological and histological differentiation, while complex odontogenic tumor only presents histological differentiation. In compound odontogenic tumor multiple amorphous dental structures are formed (denticles), while in complex odontogenic tumor a solid mass of dental soft and hard tissues is formed, these tissues are haphazardly arranged and do not resemble the morphology of a tooth.

Etiology of tumors is unknown. Lesion can arise after trauma in primary dentition, Malassez paradental remains, inflammation processses, odontoblastic hyperactivity and hereditary anomalies (Gardner and Herman syndrome). The tumor is considered a disorganoplasty that results from the deviated development of the dentoforming organ. Tumor is composed of mature dental tissues (enamel, dentine, cement), usually arranged irregularly in the tumor mass.

Odontoma are benign, not aggressive tumors which usually do not elicit symptoms; over half the cases are discovered as radiographic findings often associated with impacted teeth. The complex odontoma - a hard tumor, with irregular contour, encapsulated, constituted of masses of dentine, enamel and cement in variable proportions, arranged disorderly. It may be associated with a dentigerous cyst, or it may be the cause of dental retention; it has small dimensions. The evolution is asymptomatic. Radiologically, there are areas of irregular radioopacity, surrounded by a radiotransparent line, often associated with an included tooth.

The compound odontoma is a tumor composed of adult dental structures, organized, in the form of small, rudimentary teeth or dental fragments, surrounded by a cystic wall or fibrous sac; the number of teeth varies between 3 and 4.

From a histological point of view, compound odontoma are characterized by presence of dental tissue, demineralized enamel, dentin, cement and pulp, arranged in an organized manner of dental structures and partially surrounded by a connective tissue capsule. Conversely, complex odontoma exhibit a disorganized mass of hard dental tissue; odontogenic epithelium strands can be found in the periphery,and sometimes, presence of phantom cells, cementicles and ameloblastic epithelium can be detected.

Radiographically, compound odontoma has the appearance of radio-opaque masses with irregular margins adopting a tooth-like configuration; they present radiolucid peripheral borders, whilst complex odontoma exhibit unique radio-opacity.

The odontomas must be differentiated of the odontoameloblastoma, the ameloblastic fibroodontoma and the maxillary cysts; it may be confused

with infected odontoma, with inflammatory appearance and acute or chronic evolution; it also differs from the central ossified fibroma and osteoma.

Treatment of choice consists in conservative surgical enucleation by means of the removal of the conjunctive tissue capsule that surrounds it, suitably preparing the sample for the histo-pathological study which will support an accurate diagnosis.

The ameloblastoma - It is a benign, slow growing, epithelial odontogenic neoplasm but locally invasive neoplasm. Ameloblastoma is one of the most common odontogenic tumors of the maxillofacial region. The lesion occurs in patients between the ages of 30 to 60 years, predominantly in the fourth and fifth decades. The most favored sites of ameloblastoma are ascending ramus the mandibular molar region and proximal body of the mandible (80 %). In childhood is a very rare more frequent in children of over 10 years (10 % -28 % of odontogenic tumors).

The etiological factors are considered disorders related to the development of the embryo and the eruption of the teeth, especially the wisdom tooth. Malassez (1885) established the genesis of this tumor in close connection with the epithelium of the dental germ (*Malassez islands*). The development of ameloblastoma occurs in an early period of the evolution of the dental germ, i. e. until the differentiation of the internal epithelium of enamel into adult cells, adamantoblasts. So, ameloblastoma arises from remnants of the dental lamina, enamel organ, and cell rests of Malassez

Ameloblastomas are divided into two subtypes, based on radiological appearance. Multicystic (solid adamantinoma) ameloblastomas account for approximately 85 % of all ameloblastomas and occur in the third to seventh decades of life. On radiographs, there is marked buccolingual cortical expansion with internal osseous septae, giving rise to a „soap bubble" appearance. Tooth displacement or root resorption may occur.

Unicystic (cystic ameloblastoma) ameloblastomas occur in a younger age group and tend to be non-invasive. They present as a wellcircumscribed, unicystic, radiolucent lesion, mostly in the region of the mandibular third molar. Some unicystic ameloblastomas are associa-

ted with unerupted teeth and, therefore, cannot be differentiated radiographically from dentigerous cysts. Tooth displacement or root resorption may occur. Sometimes the teeth in the tumor area become mobile and can exfoliate on their own.

There are distinguished two stages of evolution: intraosseous and extraosseous. In the intraosseous stage, the patients do not complane pain or other functional symptoms. At this stage, the tumor is painless and remains asymptomatic as it enlarges. Without clinical manifestation; the tumor are discovered usually quite accidentally, on the occasion of a radiographic examination. In the externalization stage, the patient notices a gradual jaw expansion producing facial asymmetry. The cortex is often thinned but is seldom penetrated by the growth. The recurrence rate is high after surgical treatment.

The differential diagnosis is made with the myeloplax tumor, the root and follicular cysts, the dysplasia of the jaws, the sarcoma. The treatment regimen for ameloblastoma can be divided into three modalities: conservative (enucleation and curettage), marsupialization, and radical surgery (resection with or without continuity defect). In case of solid/multicystic ameloblastoma the treatment choice is in general resection

The cementoma are benign jaw tumors that originate from periodontal ligament elements. According to morphological characteristic, four type of cementomas are described: periapical fibrous dysplasia, benign cementoblastoma, cementifying fibroma, and florid osseous dysplasia.

The usual location is around the apex of the premolars, the cementomas is asymptomatic, with slow growth, causing tooth displacement and bone deformation in a late period of evolution; the tooth, in relation to which the tumor developed, is vital. These tumors originating from (i. e. in connection with) the tooth root and are usually slowly enlarging. It, therefore, obliterates the periodontal ligament space.

Radiologically, the appearance of the tumor is typically with radiodense or mixed-density varies depending on the stage of development. Initially, the image in relation to the apex of a tooth is radiolucent, sometimes it is partially radiolucent and partly radiopaque (well-circumscribed lobular radio-opaque mass surrounded by a radiolucent

margin), can represents a slightly denser radiopaque area than the surrounding bone. Loss of the periodontal ligament space along with root resorption/loss of root outline are common.

The macroscopic specimen showed lobular mineralized masses with a „ginger root"-like appearance. Microscopic examination showed a lobular calcified mass with a peripheral zone of fibropsammomatous tissue corresponding with the radiolucent margin.

The differential diagnoses, which include osteoma, ossifying fibroma and cementoblastoma.

6.3 Tumor like lesion of maxillofacial bones

In addition to inflammatory processes and tumors, maxillary bones can be interested by a series of quantitative and qualitative metabolic disorders, which distort the development of bone tissue. Such disorders have been called *systemic dysplasiaor osteodystrophy*. The dysplasias are diseases of the bone, which have as a morphopathological substrate the defective development of the bone growth processes, in which the radiological changes of structure predominate. They determine the changes in shape, contour and size of the bones.

The fibrous dysplasia is a benign bone diseasethat replaces normal bone with fibrous-type tissue was individualized by Lichtenstein and Jaffe in 1942. This tissue is not as hard as normal bone, and because it is soft and stringy, makes the bone more fragile and prone to break. Fibrous dysplasia is a skeletal developmental anomaly of the bone-forming mesenchyme that manifests as a defect in osteoblastic differrentiation and maturation. Any bone in the body can be affected. It is a nonhereditary disorder of unknown cause.

The exact cause of fibrous dysplasia is unknown, recent studies point to a mutation of Gs alpha protein during prenatal development as a contributing factor. Fibrous dysplasia is not hereditary, meaning parents do not pass the condition to their children.

There are two types of fibrous dysplasia, monostotic form, occurring in 70 %, which is more common in the jaws and cranium, and polyostotic, occurring in 30 %, wich is often associated with McCune-Albright's syndrome (cutaneous pigmentation, autonomic hyper-functioning endocri-

ne glands, and precocious puberty). The neck and head are involved in half of these patients. Skull involvement occuers in 27 % of monostotic and up to 50 % of polyostotic patients. The fibrous dysplasia of the jaws is more common at the age of 8-15 years, during the period of fast growth of the head and area, and during tooth eruption. Fibrous dysplasia involving the face and skull is called „Leontiasis ossea"

The evolution is slow, with periods of remission and exacerbationin. The lesion is not encapsulated and continues, without any demarcation line, with the healthy bone. causing a bone deformity, bone fractures or deformities, bone pain, dfficulty walking. Bone lesions may stop growing when the child reaches puberty. If your child's fibrous dysplasia is affecting his face or skull, he may have specific symptoms including: facial asymmetry, shifting facial structure that can affect any bone in the face, nasal airway obstruction, jaw and bite misalignment, in some cases making it difficult to chew and swallow, visual and hearing problems due to compression of the optic nerve or acoustic nerve.

Children with polyostotic fibrous dysplasia may also experience: endocrine gland problems, such as early puberty, thyroid disorders and related issues, unusual skin patches, called café-au-lait pigmentation, ranging from light brown to dark brown in color

When the swelling reaches significant dimensions, it can cause tooth displacement, occlusion disorders, displacement of the eyeball. Also, abnormal protrusion of the eyeball (exophthalmos) may develop and eventually cause complete loss of sight because its presses on the optic nerve. In addition, there may be interference of the nasal passage and with eating. It may also cause facial nerve paralysis or dizziness.

The monostotic type is the most common type seen when the jaw is involved The maxilla is the most common site of involvement. The lession does not cross the midline and tends to be limited to one bone.

The histological structure of fibrous dysplasia varies with the developmental stage. It is characterized by the destruction and metaplasia of normal bone tissue, with the formation of a partially calcified osteoid tissue or even the replacement of the bone by a fibrous cellular tissue. The bone and medullary tissue undergoes a fibrous transformation, in

the diffuse mass appearing outbreaks of osteoid tissue with non-uniform calcification, so that clearer holes remain, with micro-cysts appearance.

The radiological image is extremely varied - from homogeneous, mono- or multilocular radiotherapy to more or less intense radio-opacity, in relation to the fibrous or calcified structure of the lesion. It is considered a finely granulated radio-opaque area, the contour of which exceeds on the one hand the normal limits of the bony plane, and on the other hand is lost on the neighboring bone. The flocculent astructural image, with darker islands, with rounded-oval foci of cystic radiotransparency, without marginal reaction. Sometimes, outbreaks of amorphous hypercalcification appear, which give the bone a stained appearance.

Treatment should be deferred, if possible until skeletal maturity. Children with fibrous dysplasia should be followed quarterly with clinical and radiographic evaluation. Non-aggressive lesions that have been observed to exhibit no growth are treated by contour excision for esthetic and/or functional reasons. When disabling functional impairment or paresthesia occurs, contour or en bloc excision may be performed. Accelerated growth or aggressive lesions require early surgical intervention with en bloc resection and bone graft reconstruction. Malignant Transformation has been reported after radiation therapy, which is contraindicated.

McCune-Albright Syndrome (leonteasis ossea) is a rare fibrosseous lesion, characterized by a classic triad of polyostotic fibrous dysplasia, café macules and underlying endocrinopathies. *McCune-Albright syndrome* involve many bones and often they are confined to one side of the body. The medullary bone is replaced with fibrous tissue developing areas of abnormal scar-like (fibrous) that may lead to fractures, uneven growth, and deformity. Endocrinopathies include early puberty and enlargement of the thyroid gland.

McCune-Albright syndrome is caused by embryonic postzygomatic somatic mutation in the Gsa gene, located on chromosome 20q13.2-13. This mutation causes proliferation of the osteoprogenitor cells, leading to formation of fibrous matrix with woven bone.

Craniofacial fibrous dysplasia may cause asymmetric growth of the face, ocular effects like globe displacement and loss of vision due to the

involvement of sphenoid and ethmoid bones. Dental findings include malocclusion, delayed eruption, tooth displacement and tooth anomalies like tooth rotation, oligodontia and taurodontism. Fibroosseous lesions of craniofacial bones are characterized by Slow growing painless swelling involving both jaws and thickening of the facial bones giving the appearance of *fascias leonteasis*. Gradually enlarged bone cause spacing between teeth and noticeable facial asymmetry. This swelling was not associated with pain, paresthesia or pus discharge.

Radiographically, the lesions might progress from a totally radiolucent multilocularity, mixed radiolucent – radiopaque stage to a totally radiopaque stage, depending upon the degree of maturation. Some of the common radiologic presentations include ground glass appearance, salt and pepper, orange peel or thumb print appearance. The lesion has ill defined borders, with an absence of a capsule or cortical border and gradually merge into the surrounding bone. The mandibular canal may be displaced in any direction depending on the epicenter of the lesion, with an upward displacement, being the most common. Fibrous dysplasia of the maxilla may cause obliteration of the maxillary sinus and displacement of the orbit.

The differential diagnosis is made with osteoblastoclastoma, sarcoma, cyst, chronic osteomyelitis of the jaws.

Bisphosphonates are used to prevent recurrent fractures and they act as antiresorptive agents. Surgical treatment consists of either excision followed by reconstruction or surgical recontouring of the affected bone to improve esthetics and function.

The cherubism is a rare benign, fibro-osseous pathology of the mandible and/or maxilla of unknown origin. Cherubism was first described in 1933 by Jones, and first was found in children between 2 and 5 years of age. It is a genetic, autosomal dominant trait and usually it is a familial disease. The genetic basis for cherubism was identified in 1999, when the gene responsible for it was mapped to chromosome 4p16.3.14. The word „cherub" refers to angels with childish full cheeked face (chubby cheeks) often gazing upward as if eyes to heaven as depicted in the Renaissance era.

The disease is classified as a benign and appear during early childhood, but with age cherubism tends to regress.

In cherubism disease mandible or/and maxilla is replacement of with multulocular cyst-like tissue growths. In spite of being a self-limiting condition that subsides with age, in some cases it can cause serious orbital abnormalities and impairment of hearing On the basis of extent of involvement, some athers proposed a grading system for cherubism: grade 1 (involvement of both mandibular ascending rami; grade 2 (same as grade 1 plus involvement of both maxillary tuberosities); grade 3 (massive involvement of whole maxilla and mandible, except the condylar processes); grade 4 (same as grade 3 with involvement of the floor of the orbits causing orbital compression). Skeletal involvement i. e. ribs, humerus and femur is rare.

The first signs of manifestation of the disease are generally observed at about 2 years of age, followed by accelerated growth from 8 to 9 years and spontaneous interruption after puberty. The disorder is characterized by bilateral, painless, symmetrical swelling, giving rounded „chubby" appearance of the face.

Usually cherubism onset may cost 14-15 months, but the disease becomes clinically evident at 2-4 years, and at 5 years is well expressed by the excess volume of the jaws. Occasionally, swelling of the maxillary alveolar process leads to a decrease in the palatal arch, which is causing chewing, swallowing and breathing difficulties due to important bone changes there are disorders of the temporal and permanent dentition, in the form of dental displacement, dental mobility, eruption disorders.

Radiographically, the ascending branches and the angles of the mandible show expansile remodeling of involved bones. The bone appears as numerous welldefined multilocular radiolucencies of the jaws. The borders are distinct and divided by bony trabeculae. In mandible, it causes thinning and expansion of the cortical plates with occasional perforation. Displacement of the inferior alveolar canal may be noted. Unerupted teeth are often displaced and appear to be floating in the cyst like spaces At the maxilla, the multilocular radiolucent image is projected into the sinus of the maxilla. On the CT there is no demonstrable

sighns due to superimposition and the anatomical complexity of the jaws. The MRI findings of cherubism is as nonspecific, homogeneous, but it's helpful for determining soft tissue involvement in patients with aggressive cherubism and for assessing the vascular structures preoperatively.

Histopathologically, the lesions are composed of a vascularized fibrous stroma containing multinucleated giant cells, resembling giant cell granuloma. From histological point of view the lesion has been characterized into three subtypes: I – predominance of multinuclear cells; II – predominance of inflammatory activity; and III – predominance of fibrosis

Nowadays, genetic tests should be used for the final diagnosis of cherubism. Being a self-regressing condition, generally, minimally invasive treatment is done. Surgical treatment is mainly used for the esthetic needs and for unerupted teeth. Liposuction has been proposed to reduce the mass of the lesion in particular cases. Curettage alone or in combination with surgical contouring for cosmetic purposes has been considered the treatment of choice. Some authors point medical therapy in the form of calcitonin as a possibility to curtail the disease and obviate the need for surgery.

*Paget's disease*of bone was first described by Sir James Paget under as the term „osteitis deformans" in 1877. It is a family disease, autosomal dominant chronic progressive disease involving single bone (monostotic) or many bones (polyostotic) of the body. Paget's disease is a bone lesions, symmetrical, affecting the vertebrae, femur, skull, sternum, bones of the pelvis. The disease is characterized by rapid bone resorption and deposition, resulting in numerous reversal line formations which gives the mosaic pattern to the lamellar bone with profuse local vascularity and fibrous tissue in the marrow.

The etiology of Paget's disease is still unknown, but genetic and environmental factors may play a role. Viruses such as paramyxovirus, canine distember virus, and respiratory syncytial virus are the causative agents

Clinical symptoms include pain, deformity, and may lead to fracture of the affected bone, even though the initial course of the disease may be asymptomatic. Compared to other bones of the body, the jaws are less affected, in about 15 % of cases. In the oral and maxillofacial areas, the

maxilla is more frequently interested than the mandible Clinically, it is manifested by bone thickening, dental displacement, hypercementosis, deformation and thickening of alveolar ridges, slow healing of postextraction wounds. The enlarged bones may compress surrounding nerves and vessels causing neurological symptoms like hearing loss. Facial disfigurement may be consequence of enlargement of the maxilla and/or mandible bones.

Radiographical picture shows characteristic cotton wool appearance, well-circumscribed radiolucency, loss of lamina dura, pulpal radio-opacity, root resorption, and hypercementosis.

Anatomopathologically, the bone structure is irregular, with a mosaic appearance, determined by anarchic intensive resorption and bone formation, resulting in thickening and loss of normal architecture.

Treatment is based on powerful antiresorptive agent. Bisphosphonates form the main stay of medical management of Paget's disease. These agents suppress or reduce bone resorption by osteoclasts. Five bisphosphonates are approved for the treatment of Paget's disease. These include pamidronate, which is given intravenously, etidronate, tiludronate, alendronate, and risedronate, all of which are taken orally.

Langerhans cell histiocytosiss was previously known as Histocytosis X where the term Histocytosis refers to intense inflammatory proliferative reaction of histocytes and X denotes its unknown etiology of the disease. It has now been proposed that loss of heterozygosity on chromosomes 1, 4, 6, 7, 9, 16, 17 and 22, chromosomal instability and elevated expression of oncogene products, such as p53, H-ras and c-myc, causing disrupted cell-cycle regulation are considered to cause *Langerhans cell Histiocytosis*.

Histocytosis X is composed of three clinical variants, classified by Lichtenstein in 1953 depending on the patient's age, onset and distribution of the lesions into *Eosinophilic granuloma* (the most frequent and benign of the clinical forms which appears as a uni- or multifocal lesion in a single or various bones with or without soft tissue involvement, without systemic involvement and presenting at any age), *Hand-Schüller-Christian* chronic diffuse disease (which usually appears in

children or young adults with the characteristic triad of exophthalmos, osteolytic lesions of the cranium and diabetes insipidus and *Letterer-Siwe* disease acute disseminated (affects children under three years old involving multiple organs and systems such as liver, lung, lymph nodes, skin, bone and bone marrow and has a fatal outcome in a short time).

The eosinophilic granuloma is one of the three clinical forms of Langerhans cell histiocytosis being a benign inflammatory reaction to an unknown etiologic agent. It most commonly occurs in children and young adults. Eosinophilic granuloma can manifest in almost any bone. The most frequently involved bones are the skull, ribs and femurs, but the jaw are involvement in 77 % of cases.

Based on the location, jaw lesions are divided into the alveolar and intraosseous types. The alveolar type of lesions are commonly multiple, involve the alveolar process and has the appearance of a periodontal disease. Alveolar type commonly starts in mid-root region of tooth and with progression, a scooped-out appearance of alveolar process. Intraorally *eosinophilic granuloma* manifests withpain in the gums, associated with mobility of the teeth. The gingiva is soft, tender and bleeding, friable and swollen gingiva with severe recession exposing the roots of the teeth, early eruption of primary and permanent teeth.

Intraosseous lesions occur elsewhere in jaw and are manifested by local tension, pain, followed by bone swelling located farther from the alveolar ridge, of variable consistency, between hardness and pseudofluorescence, without changes of the soft parts and without local inflammatory phenomena, simulating an odontogenic cyst. *The diffuse form*, in which a bony segment of the maxilla is diffused in its entirety the eosinophilic granuloma can simulate a process of osteomyelitis, a giant cell tumor, an ameloblastoma or a malignant tumor, due to the more destructive nature of the lesions.

Radiologically, these lesions are totally radiolucent and the margins may be smooth or somewhat irregular. The bone around the teeth, including lamina is destroyed; as a result, the „floating teeth appearance" may be seen. Intraosseous type lesions are usually solitary with mandibular ramus most commonly involved. Intraosseous lesion may be irregular, oval or round and may stimulate periosteal bone formation.

Diverse therapeutic options are available such as surgery, radiotherapy, chemotherapy and steroid injections, alone or in combination. However, in general, no therapy is required for localized osseous eosinophilic granuloma. It is because, in many instances, the biopsy itself is enough to initiate healing and spontaneous resolution. However, steroid injection, curettage, excision or radiation can be reserved for multifocal lesions or disseminated disease.

Learning objectives

1. General characteristic of benign tumors of the bony parts.
2. Frequency of neodontogenic and odontogenic tumors in children.
3. Clinical signs of early maxillary tumors.
4. Methods of examining benign maxillary tumors in children.
5. Methods of treatment of benign tumors in children.

Tests

1. CS. Tumors and pseudotumors of the maxillary bones are commonly encountered during the age periods:

A. up to 5 years;
B. 12–16 years;
C. up to 1 year;
D. 7–12 years;
E. 1-3 years.
(B)

2. CM. Maxillofacial tumors most commonly found in girls are:

A. teratoma;
B. papilloma;
C. lymphangioma;
D. Olbrait syndrome;
E. angiofibroma.
(A, B, D)

3. CM. Indicate which tumors of the maxillofacial region have the specific feature of involution:

A. hemangioma;
B. neuroectodermal myelin tumor of the infant;
C. lymphangioma;

D. myoblastoma;
E. papillomatosis.
(A, C, E)

4. CM. Benign tumors in children are characterized by:
A. slow growth;
B. adjacent metastases;
C. spontaneous regression;
D. lack of relapses;
E. insensitivity to radiology.
(A, D, E)

5. CM. The brown spots on the skin of the face and neck region are satellite signs of the following bone neoformations:
A. cherubism;
B. fibrous dysplasia;
C. Olbrait syndrome;
D. eosinophilic granuloma;
E. neurofibromatosis.
(C, E)

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7. Fetal tumors. Malignant tumors of the soft parts and facial bones in children

7.1 Fetal tumors. Etiology and mechanisms of carcinogenesis. Classification

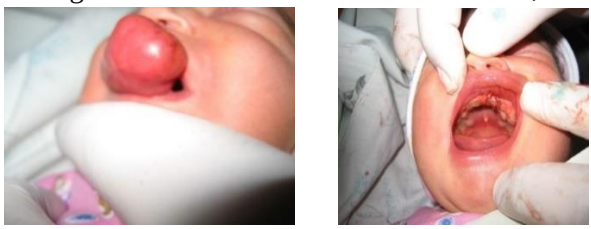
Fetal tumors are rare, but they have important implications for the health of both the fetus and the mother. Once a fetal tumor has been detected, close surve illance by a multidisciplinary team of doctors is mandatory, with anticipation and early recognition of problems during pregnancy, labor and immediate postnatal life. When the sonographic diagnosis is uncertain, fetal tissue biopsy may be necessary to obtain a histological diagnosis. In rare cases, intrauterine treatment may be possible. Some fetal tumors may be malignant and could metastasize to other fetal organs and the placenta; maternal metastases in such cases are unknown. In contrast, on rare occasions, maternal malignancies (melanoma, leukemia and breast cancer) can metastasize to the placenta; in about half of the cases with placental metastases, mostly with malignant melanoma, the tumor can metastasize to fetal viscera.

Etiology and mechanisms of carcinogenesis. Developmental errors during embryonic and fetal maturation may result in embryonic tumors. One hypothesis is that more cells are produced than are required for the formation of an organ or tissue and the origins of embryonic tumors rest in developmental errors in these surplus embryonic rudiments. Embryonic tumors developing after infancy are explained by the persistence of cell rests or developmental vestiges. Developmentally anomalous tissue (such as hamartomas and dysgenic gonads) is a source of neoplasms in older children and adults. When any of this developmentally abnormal tissue is present at birth, it is inferred that the cells failed to mature, migrate or differentiate properly during intrauterine life.

Neoplastic transformation of cells in tissue culture and in vivo carcinogenesis are dynamic, multistep and complex processes that can be sepa-

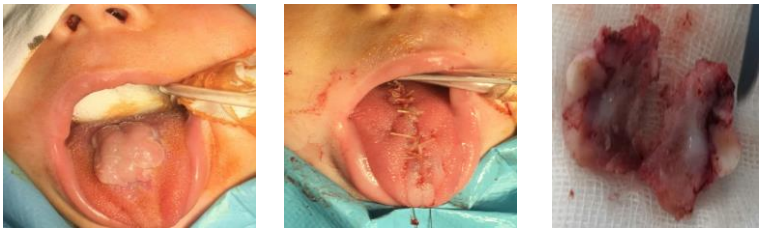
rated artificially into three phases: initiation, promotion and progression. These phases may be applied to the natural history of virtually all human tumors, including embryonic ones. Initiation is the result of exposure of cells or tissues to an appropriate dose of a carcinogen; an initiated cell is permanently damaged and has a malignant potential. The initiated cells can persist for months or years before becoming malignant. During the promotion phase, initiated cells clonally expand. Promotion may be modulated or reversed by a variety of environmental conditions. In the last phase, progression, the transformed cells develop into a tumor, ultimately with metastasis. Embryonic tumors can, therefore, be regarded as defects in the integrated control of cell differentiation and proliferation. A genetic model of carcinogenesis has also been introduced in an attempt to clarify the pathogenesis and behavioral peculiarities of certain embryonic tumors. According to this hypothesis, embryonal neoplasms arise as a result of two mutational events in the genome. The first mutation is prezygotic in familial cases and postzygotic in non-familial; the second mutation is always postzygotic.

Benignity of fetal and infantile neoplasms. (Fig 32,33) Some neonatal and infantile tumors have a benign clinical behavior despite histological evidence of malignancy. Examples include congenital neuroblastomas and hepatoblastomas in the first year of life, and congenital and infantile fibromatosis, and sacrococcygeal teratomas in the first few months of life. The factors responsible for this „oncogenic period of grace", which starts in utero and extends through the first few months of extrauterine life, are uncertain.



a b

Fig. 32 **Fetal and infantile tumors. (a. before, b. after treatment)**



a b c

Fig. 33. **Fetal and infantile tumors (a. before, b. after, c treatment).**

Association of neoplasia and congenital malformations. The concept that teratogenesis and oncogenesis have shared mechanisms is well documented. Probably, there is simultaneous or sequential cellular and tissue reaction to specific injurious agents. The degree of cytodifferentiation, the metabolic or immunological state of the embryo or fetus, and the length of time of exposure to the agent will determine whether the effect is teratogenic, oncogenic, both, or neither. Many biological, chemical and physical agents known to be teratogenic to the fetus or embryo are carcinogenic postnatally. Alternatively, a teratogenic event during intrauterine life may predispose the fetus to an oncogenic event later in life. This would explain neoplastic transformation occurring in hamartomas, developmental vestiges, heterotopias and dysgenetic tissues. It is postulated that the anomalous tissues harbor latent oncogenes which, under certain environmental conditions, are activated, resulting in malignant transformation of a tumor.

Classification. A formal classification of fetaltumors does not exist. Apart from distinguishing solid from cystic lesions, probably the best classification should be by location. The main compartments of fetaltumors are the head and brain, face and neck, thorax (including the heart), abdomen and retroperitoneum, extremities, genitalia, sacrococcygeal region, and skin.

Prenatal diagnosis The approach for prenatal diagnosis of fetal tumors should be based on three sets of ultrasound signs: general signs, organ-specific signs and tumor-specific signs. The general sonographic features, that should raise the suspicion of an underlying fetal tumor, include:

(1) Absence or disruption of contour, shape, location, sonographic texture or size, of a normal anatomic structure;

(2) Presence of an abnormal structure or abnormal biometry;

(3) Abnormality in fetal movement;

(4) Polyhydramnios; and

(5) Hydropsfetalis.

Polyhydramnios is particularly important, because almost 50 % of fetal tumors are accompanied by this finding. The underlying mechanisms include interference with swallowing (such as thyroid goiter or myoblasttoma), mechanical obstruction (such as gastrointestinal tumors), excessive production of amniotic fluid (such as sacrococcygealteratoma), and decreased resorption by lung tissue in lung pathology. Intracranial tumors are also commonly associated with polyhydramnios and the mechanism may be neurogenic lack of swallowing or inappropriate polyuria.

Tumor-specific signs include pathological changes within the tumor mass (calcifications, liquefaction, organ edema, internal bleeding, neovascularization and rapid changes in size and texture). Organ-specific signs are rare, but in some cases they are highly suggestive of the condition (such as cardiomegaly with a huge solid or cystic mass occupying the entire heart, suggesting intrapericardialteratoma).

In some cases, normal and abnormal sonographic findings may mimic fetaltumors. Examples may vary from severe cases of bladder exstrophy (where the protruding bladder mass appears as a solid tumor-like structure), to rare cases of fetal scrotal inguinal hernia (where bowel loops occupy the scrotum, appearing as huge masses).

Prognosis Apart from intracranial tumors (where the prognosis is generally poor), the prognosis for tumors in other locations is variable and depends on the size of the tumor (with resultant compression of adjacent organs), degree of vascularization (with the risk of causing heart failure and hydrops), and associated polyhydramnios (with the risk of preterm delivery).

7.2 Pediatric Head and Neck Tumors

Tumors or growths in the head and neck region may be divided into those that are benign (not cancerous) and malignant (ie., cancer). Fortunately, most growths in the head and neck region in children are considered to be benign. These benign growths can be related to infection, inflammation, fluid collections, swellings, or neoplasms (tumors) that are nonlife-threatening. The malignant growths, on the other hand, may be life-threatening and it causes other problems related to their growth and spread. Even the malignant growths in the head and neck are usually treatable.

Benign Tumors. It is very common for children to have enlarged tonsils and adenoids. These are almost always from an infection or inflammation. It is very rare that children develop a cancer, lymphoma, or sarcoma of these areas. When the tonsils, adenoids, or othe rareas of the mouth or throat remainen larged or are enlarged on only one side, it is important to have an evaluation by a specialist in ear, nose and throat or otolaryngology-head and neck surgery.

The lymph nodes of the neck region may become enlarged during childhood. Most of the time, this is reactive in nature and related to inflammation or infection. However, if the lymph nodes remain enlarged for a period of time without going away, it is important to have an otolaryngologist-head and neck surgeon evaluate the problem.

Other benign growths in the face and neck include cysts (fluid collection) such as branchial cleft cyst, thyroglossal duct cyst, cystichygroma, and dermoid cysts. These often require removal due to the incontinued growth and potential for infection. Growths of blood vessels often are seen in the face and neck and these are often referred to as hemangiomas, vascular malformations, lymph atic and arterio venous malformations. Some of these may require removal or treatment depending upon the type and location.

Sinus and bone growths. Most children have often nose bleeds cause by occasional allergies and sinus infection. Sometimes tumors of the nose and sinus present with similar symptoms with nose bleeds. Generally recommended that a child with continuous sinus problems or nose bleeds be evaluated by an otolaryngologist and head and neck surgeon to be sure it is not a tumor condition.

Non-epithelial neoplasms constitute the majority of sino-nasal tumors in children and adolescents. Among these, rhabdomyosarcoma or undifferentiated sarcoma and non-Hodgkin lymphoma account for the majority of cases. Among head and neck rhabdomyosarcoma 14 % arise from the nasal cavity and 10 % from the paranasal sinuses. Nasopharyngeal carcinoma accounts for one third of the nasopharyngeal neoplasms in children. Less frequently, Ewing's sarcoma can present in this location. These tumors have also been described as secondary malignancies following treatment of retinoblastoma and other neoplasms. Other less common sino-nasal tumors presenting in children include hemangioma and hemagiopericitoma, fibroma and fibrosarcoma, malignant fibroushistiocytoma, and desmoids fibromatosis.

Salivary gland tumors. There are three paired sets of salivary glands in the headand neck region. These include the ones in front of the ears (parotid), below the jaw (submandibular), and underneath the tongue (sublingual). Additionally, there are numerous very small salivary glands through out the mouth and throat. Although tumors can arise in these areas, they are rare. Thus, any child with a growth in the seareass should be seen by an otolaryngologist and head and neck surgeon.

7.3 The peculiarities and diagnosis of malignancies in children

Primary-origin tumors in pediatric patients are very rare but more aggressive and exhibit high levels of recurrence due to the growth potential of developing cells. Evolution of the tumors exhibit rapid local growth, with local invasion and tissue destruction, which often do not match their benign histological appearance. Tissue damage caused by tumor is of greater impact, in children, because they directly alter facial growth and development as well as psycho-social

Practically all basic tissues of the body can develop tumor lesions but the upper and lower jaw in children are afflicted in about 20 %. Nevertheless, jaw tumors possess a unique characteristic, because they have teeth included. Therefore, lesions that can be found in these structures which cannot be found in any other anatomical area of the body.

The diagnosis malignant tumors in children should be made as short as possible. Nevetheless, diagnosis of malignancy in children is difficult.

It must be done based on the clinical examination. Initial assessment of children with tumor lesions must include clinical history and physical exploration. All the following factors must be carefully taken into account: age, gender, familial genetic history, lesion onset, evolution and present state, present signs and symptoms, clinical data such as pain, paresthesia, paralysis, lesion discoloration, lesion bleeding, time of growth and degree of destruction or invasion to adjacent anatomical structures, mobility or fixation of the lesion, consistency, contents and size of tumor.

Other important signs and symptoms that might lead us to consider a possible malignant lesion are considerable weight loss, asthenia, adynamia, high temperature, or tegument pallor. It is important to take into account diagnostic auxiliary tools such as laboratory exams, X-rays computed or three-D tomograms, magnetic resonance or even ultrasound.

Histological origin or malignancy degree are prone to confusions, therefore, harvesting the biopsy is the most important element to achieve accurate diagnosis. Because tumor in children occur infrequently only a few oral pathologists have had the opportunity or experience in diagnosing these lesions and predicting their biological behavior. For this reseon, it is recommended to count with experienced pathologists, well able to recognize tumors in the facial region, since accurate diagnosis will lead to an appropriate surgical behavior. Histologically cells of malignant tumors have young, atypical cells with atypical multiplication that infiltrate the surrounding bone tissure structure and the surrounding soft tissuer. Unlike benign tumors, malignant tumors relapses and metastasizes.

In relation to the histological elements from which they onset, there are several anatomo-pathological forms of sarcomas: fusocellular (they onset from the connective tissue of the support or from the periosteum), with slow evolution; osteogenetics (they onset from osteoformative tissue); sarcomas with round cells (they originate in the differentiated elements of the blood-forming bone marrow); lymphosarcomas, myelosarcomas, reticulosarcomas, rapidly evolving Ewing's sarcoma. There are also associated forms: osteosarcomas, chondrosarcomas, fibrosarcomas, osteochondrosarcomas. Rabdomiosarcom (*Fig. 33*)

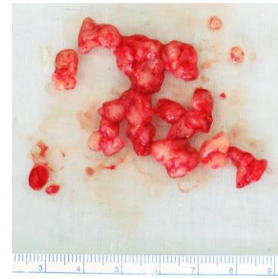



Fig. 33. **Rabdomisarcomas.**

The imaging exploration, in order to establish the diagnosis, includes: 1) radiographic exploration that highlights the tumor and directs the diagnosis to elements of malignancy; 3) computerized tomography - completes the information regarding the malignancy criteria of the tumor, and highlights the invasion of the soft tissues; 4) Bone scintigraphy reveals tumor hypercaptation due to the rich, anarchic tumor vascularization with multiple blood lakes.

Radiologically, the malignant tumors have specific criteria: 1) the tumor has a clear, imprecise contour; 2) the cortex of the bone is interrupted by tumor extension; 3) malignant tumors cause periosteal spiculiform reaction. At the periphery of the tumor, instead of affecting the periosteum, the periosteal spicules of triangular shape appear, with the name of „Codman's triangle"; 4) the structural changes may be lithic, osteosclerotic, mixed; 5) the soft structures are infiltrated, invaded by multiple bone inclusions.

The general clinical signs of sarcomas. From the clinical and the onset point of view, two forms of sarcoma are known: with peripheral onset (periosteal sarcomas) and with endoosseous onset (central sarcomas).

Mandibular sarcoma. In the forms with peripheral onset, bone deformation is observed, more commonly with vestibular localization. At first, the tumor has a hard consistency, then it becomes variable, the hard areas alternating with the depressive or soft consistency areas. Initially, some tumors are similar to epulids. Gradually, the tumor infiltrates the soft parts of the vestibule, cheek or floor, in the form of a fleshy, voluminous, polylobate mass, with red-violet covering mucosa,

stretched or ulcerated. The tumor vegetation covers the mobile and painful teeth. The patients claim spontaneous pain, which is accentuated during the functional actions.

In forms with deep and endoosseous onset, the clinical signs are minimal. The first signs appear in the form of dental or neuralgiform pain, the parastatic disorders in the territory of the chin, dental mobility. Subsequently, the deformity of the bone cortex - irregular surface appears, covering the appearance of a tumor that makes the body common to the bone, which appears enlarged in volume and with a variable consistency with respect to the histological type. After rupture of the cortex, the tumor usually has a rapid evolution, invading the soft perimandibular parts or the buccal cavity, in the form of vegetation that fills the vestibular or sublingual groove with hemorrhages at the slightest trauma. The impressive clinical aspects, due to the volume of the tumor and the rapid evolution, cover the lymphosarcomas, myelosarcomas and reticulosarcomas. In forms with distal localization, the tumors manifest by trismus of varying degrees, embarrassment at chewing and swallowing. The pains are of great intensity, radiating in the affected hemimandibula or in the hemicranium.

Radiographically, it presents different aspects, in relation to the histological form, the osteolytic or osteoblastic character. The characteristic images of sarcomas, such as „hedgehogs", „needle pillows", „combed bone", „onion sheets" are described. Initially, the destructive osteolytic tumor processes highlight the deletion of the trabecular bone pattern and the appearance of diffuse radiotransparency areas, disseminated in the thickness of the bone. Then there are broad, uneven radiotransparent gaps that may interest large portions of bone, where teeth seem to float.

The differential diagnosis is made with the epulis in the forms with peripheral onset, with the myeloplasma tumor, with the mandibular osteodystrophies, with the osteomyelitis, etc.

*The jaw sarcoma.*The sarcomas can start peripherally and deeply. The infarct sarcoma shows clinical signs similar to those of sarcoma with peripheral onset of the mandible. The first clinical signs are: neuralgiform pains, odontalgia, dental mobility, gingivorrhagia. The tumor grows accelerated, generating bone tumor deformation; it has a vegetative,

polylobate character, it covers teeth that can be later expelled. By its growth, the tumor invades the vestibule and the palatine vault, modifying the facial appearance. The tumor ulcerates, and then bleeds. The functional disorders in mastication and phonation are present.

The first clinical signs in tumors with mesostructural onset are sinus: pain, fetid, unclear bloody and sanguinopurulent discharges, through the nostril of the affected part. The anterior rhinoscopy can detect fairly early tumor vegetation in the middle meatus. Parallel to sinus signs, patients claim dental mobility, pain, which often leads to dental extractions. Usually, after the dental extractions, the fleshy clusters appear in the alveolar wound, which grow progressively, are painful and bleed. In the more advanced phases, the tumors evolve into the buccal cavity, where they are externalized in the form of a vegetative tumor or they protrude the
genic region, producing a marked asymmetry.

At first, the skin is only pushed by the tumor, then by rapid infiltration of the tumor, they change their color, into reddish-purple, adhere to the tumor and at very advanced stages can ulcerate. The tumor extends into the neighboring structures: orbit (palpebral edema, chemosis, ophthalmic neuralgia), nasal fossa, producing deformities of the nasal pyramid, obstruction phenomena, epistaxis; towards the zygomatic pit and the base of the skull, producing intense headaches.

The radiographic examination is less conclusive than in the mandibular locations. Initially, the tumor image looks like a banal sinusitis by opacifying the sinus, then by deforming the sinus walls and destroying the bone walls.

The osteosarcoma is one of the most aggressive malignant tumors of bone. Although rare, osteosarcoma is the more common type of all the sarcomas. Osteosarcomas may involve the mandible or maxilla and rarely the ethmoid region. The most common sites of involvement are the body of the mandible and the alveolar ridge or the antral area of the maxilla. The majority of tumors arise within the medullary cavity of the affected bone with rare examples developing on the bony surfaces. The long bones like the femur, tibia and humerus are most frequently affected Osteosarcomas are prone to develop hematogenous metastases, mainly to the lungs.

When the tumor occurs in the jaws, the main complain is the presence of a rapidly growing swelling and pain. The affected teeth may be loosened, displaced and paresthesia may develop. Usually, the parents report the symptoms related to osteosarcoma, as being of a dental nature, which is why the extraction in the tumor area is practiced in 40-50 % of cases. Sometimes, the sarcoma is caused by trauma. Itching sensations, dental mobility, hypoesthesia occur in the innervation regions of the chin or infraorbital nerve. With localization to the upper jaw, the clinical signs appear later. Because of the initial invasion in the paranasal sinus, the tumor appears by the deformation
of the jaws, edema in the soft parts, high temperature, increased ESR.

Radiographically, two types of osteosarcoma were established: osteoblastic and osteolytic. The earliest radiographic sign is a widening of the periodontal ligament space or a radiolucency around one or more teeth. Later on, the lesion assumes an osteolytic radiolucent form, an osteoblastic radiopaque form or a mixed radiolucent image with radiopaque foci. The malignant nature of the disease gives it an irregular, ill-defined borders. There is expansion and destruction of the cortical plates. In one-third of the cases, thin spicules of new bone extend outwards away from the bone cortex, producing the characteristic sunray, sunburst or fan-shaped appearance. On CT, the tumor displays a spectrum of bone changes from well demarcated borders, notably the low grade osteosarcoma (uncommon), to lytic bone destruction with indefinite margin and variable cortical bone erosion, to the ostoeblastic form, where the bone is sclerotic. The majority of osteosarcomas have matrix mineralization, calcifications of the osteoid or osteoid-Iike substance within the tumor and some tumors show a sunburst effect caused by radiating mineralized tumor spiculae. Cortical breakthrough and interruption of alveolar margin is common in advanced cases.

The osteoblastic form is most commonly found in children and young people, and the osteolytic form - in adults with low immunity and minimal bone formation possibilities.

The fibrosarcoma It is a malignant tumor of fibroblasts in which there is no deposition of osteoid or bone. Fibrosarcoma is more uncommon than

osteosarcoma and chondrosarcoma. It is relatively rare (2-10 %) tumor, originating from connective tissue or periosteum. It is seen in the skull base (common), maxilla and orbit (less common), and cartilage of the nasal septum (rarely).

Histologically, chondrosarcoma of the craniofacial region can be divided into subtypes: the conventional subtype with myxoid and/or hyaline components, the aggressive mesenchymal and dedifferentiated subtype and the extremely rare clear cell subtype. The conventional type, which is the most common form, is slow growing, and rarely metastatic. On the other hand, mesenchymal chondrosarcoma is more aggressive and tends to metastasize. They slowly increase in size, and the majority of them are already extensive at the time of diagnosis. The patient experiences pain, swelling, loosening of teeth and paresthesia.

The tumor occurs in older patients and in young children. As with osteosarcoma, fibrosarcoma is occasionally associated with Paget's disease or may result from therapeutic irradiation.

The radiographic appearance may simulate the radiolucent form of osteosarcoma, and have ill-defined borders. The teeth may be displaced. On CT scan, chondrosarcoma shows a soft tissue mass with characteristic multiple stippled and amorphous areas of calcifications that may be associated with bone destruction and an inhomogeneous pattern of contrast enhancement

When it reaches large proportions, the lesion generates functional disorders. Most often, deep-growth fibrosarcoma is observed by parents at an advanced stage of development, because, first, the tumor grows toward the maxillary sinus, and in the second, children cannot express their subjective sensations, considering them normal.

The Ewing's sarcoma (*Fig. 34*) is a malignant tumor of bone derived from mesenchymal connective tissue of the bone marrow. The frequency of Ewing's sarcoma is 88 % between 6-20 years, with a priority at the age of 13 years. The lesion is most common in the femur and tibia. Of all sarcomas in the body, the interest in the facial bones rarely occurs and it is found in only 1 % of cases. It may be seen in the orbital wall, sphenoid and maxilla and mandible, in the angular regions and the mandibular branch.

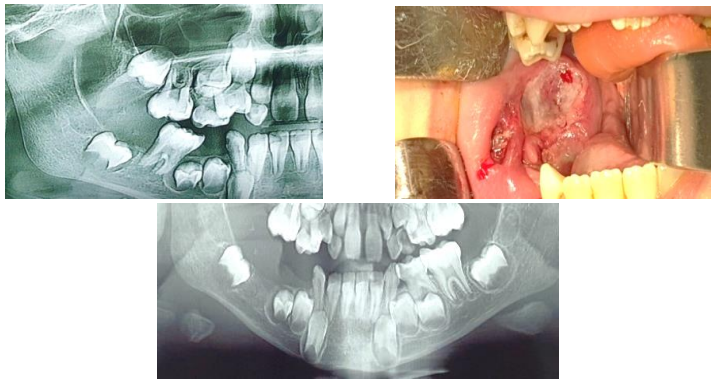


Fig. 34. **Malignant tumor of the mandible. (*Ewing's sarcomas)***

It is a rapidly growing, highly invasive tumor with early and widespread metastasis. Because of the intense vascularity of the tumor, hemorrhage and necrosis are common. Pain and swelling are the most common manifestations often with fever, anemia, leukocytosis, increased ESR. In later stages dental mobility appears. The Ewing's sarcoma is characterized by alternating periods of exacerbation and remission. During the remission period, the swelling is reduced, the pains disappear, the dental mobility reduces.

The radiographic appearance is that of an ill-defined destructive mottled radiolucent lesion which may be unilocular or multilocular. The margin is diffuse with unsharp edges and extensive cortical destruction. It may be associated with perpendicular bony spicules and shows the characteristic onion peel appearance of periosteal reaction, and less often with a sunburst type of periosteal reaction. In the early stages, the mottled rarefaction may resemble an osteomyelitis. In later stages, it may stimulate the periosteum to produce thin layers of bone, resulting in an „onion skin" effect. Advanced cases may exhibit a sunburst appearance. Often Ewing's sarcoma is confused with acute osteomyelitis. The evolution is peculiarly aggressive, with early metastases to other bones or lungs. Over 25 % of patients have metastases at diagnosis. The prognosis is very poor, most cases lead to death within a few years of diagnosis.

The hemoblastoses is a malignant tumor of the blood forming and lymphatic tissue, which includes two groups: 1) leukemia, malignant tumor of bone marrow; 2) lymphomas – malignant tumor of the lymph nodes,

without the primary involvement of the bone marrow. They occur in children with a frequency of 50 % of all malignant tumors. All *hemoblastoses* are characterized by general clinical manifestations: fatigue or pale skin, infection and fever, easy bleeding and or bruising, weakness, unexplained joint pain, anemia, granulocytopenia, thrombocytopenia, changes in the immune system; fever, weight loss, loss of appetite

Leukemia is a malignant neoplastic disease of the bone marrow and peripheral blood. The disease is characterized by overproduction of white blood cells with replacement of the normal bone marrow, circulation of abnormal cells in the blood, and infiltration of other tissues. Leukemia accounts for 5 % of all malignancies. In children, it is the single most common cause of cancer deaths. The acute and chronic forms can occur at any age, however, the acute form generally occurs in children whereas the

chronic form generally occurs in adults. Clinical manifestations include fatigability, anemia, lymphadenopathy, hepatosplenomegaly, bone and abdominal pain, secondary infection, and hemorrhagic lesions secondary to thrombocytopenia. Failure of liver and spleen is due to infiltration by malignant cells. Oral lesions include gingival bleeding, mucosal ulceration, gingival enlargement, pain and periodontitis. Radiographic findings are those of periodontal disease, severe bone loss, loss of lamina dura and loosening of teeth. Local dental cause must be ruled out to make an interpretation of leukemia.

The reticulosarcoma has its origin in the differentiated elements of the blood-forming bone marrow. It is rarely encountered (5 % of the total malignant tumors of the jaws); it is growing relatively slow. The general systemic clinical manifestations - fever, weight loss, indisposition - are minimal, even if local changes occur. This relatively satisfactory general condition, despite widespread local bone destruction, differentiates bone lymphomas from other malignant tumors. In young children it evolves rapidly. The first clinical sign is the deformation. The pains settle in the stages of advanced evolution. With the swelling, the signs of inflammation of the skin and mucosa, such as hyperemia, edema, become evident. Its characteristic the febrile condition, increased ESR. In advanced stages, the functional disorders occur - disorders in the chewing,

swallowing, heavy nasal breathing. The dental mobility in the affected area is a permanent clinical sign. The submandibular and cervical lymph nodes are enlarged.

The radiographic examination of the mandibular locations shows destructive osteolytic lesions, which appear at the onset of the disease. At first, the pseudo-cystic round radiospray, without peripheral condensation contour observes. In more advanced stages, the image highlights gaps of ununiformed radiotransparency, with irregular contour. The interradicular septaare destroyed, and the teeth float in the areas of radiotransparency. The osteolysis may comprise large areas (body, angle and mandibular branch).

Malignant lymphomas are a group of immunologic neoplasms which arise in lymphoid tissue. The various types of lymphomas are Hodgkin' sdisease, lymphosarcoma, reticulum cell sarcoma and giant cell lymphoma. Depending on the type of lymphoma, some or all of the following clinical features may be exhibited: painless enlargement, fever, loss of weight, anemia, generalized weakness, and anorexia. All lymphomas, with the exception of Hodgkin's disease, affect middle and older age groups. On a radiograph, the radiolucencies are similar in appearance to periodontal abscesses. There is loss of lamina dura around the teeth. In advanced stages, large multilocular destructions are seen. To make a complete diagnosis of malignant lymphoma, clinical, radiographic, microscopic and laboratory information must be correlated.

Hodgkin's disease also called lymphogranulomatosis or limphoadenoma is a a form of malignant lymphoma, most commonly seen in children. It is characterized by progressive chronic inflammation and enlargement of the lymph nodes, spleen and occasionally liver and kidneys and by lymphoid infiltration along the blood vessels. Lymph nodes are located in the neck are, armpit, groin and mesentery which at first are painless and of elastic consistency.

There are two types of lymphoma: Hodgkin and non-Hodgkin lymphoma. Hodgkin lymphoma tends to affect the lymph nodes in the head and neck whereas non-Hodgkin lymphoma can affect any lymph node or related tissue in the body. (*Fig.36*)

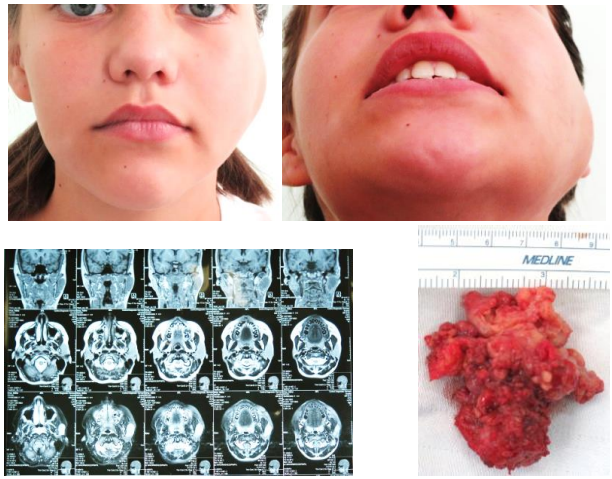


Fig. 26. **Non-Hodjkin limphomas.**

The exact cause of Hodgkin's disease is unknown but there is increasing evidence that infections (such as the virus that causes glandular fever) may play a part in its development by genetic mutation of the lymphocyte. There is an increased risk of the disease among parents, siblings and identical twins of lymphoma patients. Riscs factors can be also young adults or people over 55; male are more likely to develop lymphoma than females; people with immune system diseases or drugs untaken that can suppress their immune system; some infection including the Epstein –Barr rirus and Helicobacter pylon infection. Hodgkin lymphoma occurs more frequently among young children from developing countries than among those from countries of advanced socio-economic status.

The early symptoms of Hodgkin's diseaseare painless swelling of one or more lymph nodes, causing a bump under the skin. Along with the local advancement of lyph nodes pathology, general clinical manifestations include unexplained fever, unexplained weight loss, drenching night sweats

There are 4 stages of tumor development: 1) envolving one are of lymph nodes or neighboring groups; 2) envolving regional of several groups of lymph nodes; 3) generalal envolving of lymph nodes and spleen; 4) envolving lymphoid organs - bones, liver, lung system, digestive tract.

The differential diagnosis of lymphogranulomatosis is made with odontogenic infection, lymphadenitis caused by specific infection like tuberculous lymphadenitis, mononucleosis and from leukemia and sarcomas.

The common clinical signs of swelling lymph node caused by odontogenic infection are: 1) the presence of the offending tooth; 2) the odontogenic infection involves regional lymph node, whereas lymphogranulomatosis affects the cervical and supraclavicular lymph nodes; 3) the swelling lymph nodes are painful; 4) the swelling lymph nodes response to antibiotics.

The swelling lymph node caused by tuberculous is characterized by: 1) the adhesion of the lymph nodes between them and with the deep cervical layies; 2) the tendency of fistularization; 3) samplesto tuberculin are positive.

The mononucleosis (kissing disease), caused by Epstein-Barr virus resemble a virus infection that. is characterized by: 1) enlargement of the lymph nodes in the cervical area; 2) acute onset, which can develop with acute angina, and after tonsillectomies - with pharyngeal mucosal hyperemia; 3) abdominal pain, enlargement of the spleen, liver; 4) lymphocytosis.

Hodgkin lymphoma must be deferentiated from another form of blood cancer - *leukemia.* The difference is that leukaemia develops in the bone marrow and affects normal blood cell production. Lymphogranulomatosis develops in the lymphatic system and does not affect normal blood cell production.

The exact diagnosis of Hodgkin lymphoma can be confirmed based on morphological examination. Hodgkin lymphoma is distinguished from other types of lymphoma by the type of cancer cell formed – the Reed-Sternberg cell – which is not found in any other blood cancer. The main treatment for Hodgkin lymphoma in children is chemotherapy. Sometimes patients requied radiotherapy alone or in combination with surgery.

The lymphosarcoma is one of the most aggressive lymphomas; it occurs in 8–9 % of all malignant lesions, especially in children aged 3–5 years. In children, lymphosarcoma affects the lymph nodes of the mediastinum, the abdominal cavity, the soft tissues, the bone tissue of the

upper and lower limbs, but especially the nodules in the cervical and supraclavicular regions. At the onset of the disease, the lymph nodes increase in volume, become painful at palpation, of elastic consistency, with slight signs of intoxication. At this stage of development, lymphosarcoma resembles reactive adenitis, often being confused with odontogenicreactive adenitis. For this reason, inappropriate treatment (surgical or conservative) is applied, which greatly favors the acceleration of the tumor. Subsequently, there is an intensive growth of lymph nodes, with changes in consistency, firm, adherent to each other and to the deep planes.The tumor has a soft reddish-gray consistency, with invasive and destructive growth around the lymph nodes and lymphatic tissue.

The tumor lesions in children, even benign ones, have the capacity to grow fastn a short period, with invasion of adjacent areas or organs, subsequently with function disorders. Inert surveillance of diseases with localization to the soft or bony parts can lead to deformities and secondary disorders, which require further treatment. The early determination of the diagnosis at the primary stage, then the referenceto the specialized departments will be done shortly.

The treatment of malignant tumor lesions in children is combined. The surgical treatment in combination with the action of the radiological rays on the tumor masses, the chemotherapy remain basic, for Ewing's sarcoma. Depending on the cells detected in the malignant tumor masses, the combinations are practiced: 1) chemotherapy and surgical treatment; 2) surgical treatment and chemotherapy; 3) chemotherapy and surgical and radiological treatments; 4) radiological treatment and chemotherapy.

The surgical treatment of benign and malignant tumor diseases consists in the removal of tumors by partial or total resection of the jaw. Surgery can lead to facial deformities, disorders of growth and developmen of facial bones. Reconstruction options include reconstruction bar with iliac crest bone graft or free flap reconstruction. The decision to use nonvascularized graft vs free tissue is made based on the size of the defect and the location of the defect, as well as the extent of the resection or the need for adjuvant therapy. When the periosteum is resected as a margin, the wound bed becomes more hostile for bone regeneration as

well as for nonvascularized graft healing. For larger defects of this nature, composite free tissue transfer is a good option. Additional procedures were performed for recurrence, dental implants, cosmetic revision, second look, and repeated biopsy.

The rehabilitation is a complex of controlled medical environment to help body heal and to regain strength, relearn skills that have been lost after surgery or trauma or find new ways to do activities that after surgery or trauma may be difficult, social and professional measures to achieve the maximum purpose in the functional activity.

There are three main types of rehabilitation therapy: occupational, physical and speech. Rehabilitation varies from person to person and may include different treatment techniques such as therapeutic exercise, manual therapy, music or recreational, neurological re-education or modalities for pain relief. Each form of rehabilitation serves a unique purpose in helping a person reach full recovery, return him to a healthy and active lifestyle. Each type of rehabilitation therapy can be accessed in various healthcare settings - in-patients and/or out patient conditions.

The occupationl therapies help to restor an individual's ability to perform necessary daily activities. This may mean working to improve fine motor skills, restore balance, or assist patients in learning how to increase their functional ability via use of adaptive equipment, among other potential treatment options. Occupational therapists help by making changes to things that hinder someone's ability to complete tasks such as eating, dressing, brushing one's teeth, completing school activities and working. Modifications may include changing the way the task is approached, changing the environment in which the task is completed or helping a person develop skills necessary to complete certain tasks.

The systematic clinical monitoring aims at detecting the relapses, the metastases, ensuring the harmonious growth of the child through orthodontic, prosthetic and surgical assistance.

Speech therapy or speech-language pathologists provide treatment for those who have speech issues and help to treat a wide variety of issues involving language, communication, voice, swallowing and fluency. *The*

speech therapy rehabilitation begins immediately after suspecting and establishing the tumor diagnosis, and it lasts throughout the life. The goal of speech therapy is to combine the mechanics associated with speech with the use of language. The end result is to help the patient communicate in more useful and functional ways.

Physical type of rehabilitation therapy works to improvemovement dysfunction. provides treatment for those who are experiencing pain or difficulty in functioning, moving or living life normally. Therapists work with patients to restore movement, strength, stability and/or functional ability and reduce pain via targeted exercise and a range of other treatment methods.

Because tumor require continuity resection of the mandible or maxillectomy, an appropriate reconstructive strategy can be planned for while undertaking definitive treatment, whether that involves fabrication of an obturator, precontouring a reconstruction plate, or planning a free tissue transfer. Postsurgical defects of the jaw and face requies surgicall reconstruction or/and orthopedic remodeling splints immediately after the removal of the tumors. Orthopedic splints (movable or fixed) help individuals to protect the postsurgery wounld, to restore the continuity of the dento-alveolar, dento-maxilary or maxillo-facial defects. Tissue damage caused by tumors can modify facial growth and development causing physical, esthetic and psychological alterations. By applying different devices the functional and anatomical disorders is mantaining and contributes to the adequate development of children, to the early return and adaptation of the children in the preschool and school groups.

The orthopedic devices are prepared preoperatively, with 10-15 days. Immediately after surgery, they protect the operative wound; then, after 20 days, they are permanently changed or modified. The child is examined by the craniofacial orthopedic every 3 months, because, with it grows and develops, the need to change the devices appears.

The surgical rehabilitation of children with tumors in the maxillofacial region is performed in four stages: 1) preoperatively (from the moment of diagnosis until surgical removal); 2) postoperatively (within 10 days after

surgical removal); 3) late (from 10 days after surgery until the discharge of the patient); 4) from the day of discharge and throughout life.

The removal of the tumor lesions in the growing body causes essential disorders in the cosmrtic appearance of the face (scars), in the development of the facial skeleton and functional disorders (mastication, phonation, speech). The bone defects can be partial (without interruption of bone continuity) or total (with interruption of bone continuity). They are the causes of secondary deformities of the facial skeleton due to defect at the level of dentoalveolar, dentofacial and craniofacial regeons. Bone defects lead to the muscle dysfunctions (masticators and mimics), atrophy of the soft parts in the facial and cervical areas.

The postoperative bone defects are removed immediately (primary or momentary), in the same session as tumor removal; secondary plastic surgery - one year after tumor removal; repeated plastic surgery - after the failure of the primary plastic surgery. The primary, secondary or repeated bone graft surgeries have the functions: anatomical goal - to restore the integrity of the jaws; mechanical –to support the soft parts; biological –to stimulate the osteogenic processes, to accelerate the consolidation.

The primary reconstruction by bone graft is indicated in children after resection of a bone portion because of a benign tumor or a tumor like lession, when there is no risk of relapse. Indication for primary bone graft is the presence of a sufficient quantity of soft, well-vascularized tissues.

The secondary reconstruction by bone graft is indicated in children after tumor removal, when tissue insufficiency is detected to cover the bone graft or in cases of malignant tumor lesions. The secondary plastic surgery is performed later in a 3-4 months after tumor removal, when the soft covering parts preserve their vascularization and there are no secondary changes caused by yhe scars. After malignant tumors are removed, bone graft surgery is indicated only after 1-1.5 years.

The repeated reconstruction is performed in cases where the primary and secondary surgeries have failed and the bone graft has been expelled or resorbed. After these failures, surgery is indicated only after 6-8 months.

When planning the treatment of children with postoperative defects and deformities of the jaws after the tumor is removed, are divided into three groups. The first group is children with bone defects, in which the plastic surgery is planned in the same session. In the second group, there are children in whom the defect plastic surgery is not planned within the same session. From the moment of discharge, these children are treated with deferent casts and supervised by the craniofacial orthodontist. The third group comprises the children in whom the plastic surgery of the defect and the orthodontic treatment were not performed.

The bone grafting in children are accompanied by peculiarities, which are related to the constant growth and development of the body and jaws, and determine the results of the plastic surgery. One of the peculiarities of the bone plastic surgery is the size of the transplant, which must be larger than the defect. With the growth of the jaws, the harmonious growth of the jaws ensures. The facial symmetry is restored after 3-4 years from the primary bone plastic surgery. Until the definetive growth of the jaws, the bone plastic surgeries in children can be repeated.

The problem of fixation and immobilization of the transplant is the second peculiarity in the bone plastic surgery in children. The anatomical-physiological peculiarities of the jaws in children create unfavorable conditions for fixing the transplant and postoperative immobilization. The immobilization is done with movable or fixed devices.

The surgical results depend on some peculiarities of pre-operative preparation: 1) anamnesis of life, with the purpose of determining the suffered diseases; 2) detection of chronic or specific conditions, which can influence the operative results (active forms of tuberculosis, diabetes, haematological disorders); 3) detection of the diseases that require multidisciplinary treatment both during the preoperative and postoperative period.

Immediate postoperative period requires special care: 1) infection prophylaxis by administration of antibiotics; 2) immobilization of fragments, for a period of 50-60 days, hygiene of the postoperative wound, hygiene of the oral cavity; 3) the clinical-radiological records of

the regeneration processes; 4) remodeling of bone by craniofacial orthodontics with fixed or removable splints; 5) follow up of children until adolescence, The children with bone plastic surgery after removal of the tumor are examined every 3 to 3 months in the first year; once in 6 months the second year; and once a year for 15 years. At each stage, radiologic examinations are made to determine the regeneration status of the bone graft. Occlusion and the degree of secondary dental-alveolar deformations must be evaluated by craniofacial orthodontist. He/she permanently ensures the smooth growth of the jaws, and the systematic correction of the occlusion. Early detection of tumor lesions would be of the utmost importance to avoid severe complications derived from radical surgical treatments.

Learning objectives

1. Osteogenic sarcoma. Diagnosis, clinical picture. Treatment.
2. Lymphogranulematosis. Diagnosis. The clinical picture.
3. Ewing's sarcoma. The clinical picture. Differential diagnosis.
4. Monitoring of children with malignant tumors.
5. Surgical rehabilitation of children with malignancies.

Tests

1. CS. The characteristic radiological image for osteogenic osteosarcoma will be:

A. combed bone;
B. multilocular;
C. unilocular;
D. „honeycombs”;
E. „soap balloons”.
(A)

2. CS. Ewing's sarcoma develops as:

A. phlegmon;
B. abscess;
C. inflammatory infiltrate;
D. periostitis in the intraosseous stage;
E. acute odontogenic osteomyelitis.
(E)

3. CS. The differential diagnosis of lymphosarcoma is made with:
A. lymphangioma;
B. lymphadenitis;
C. lymphogranulomatosis;
D. chondrosarcoma;
E. Olbrait syndrome.
(C)
4. CS. Which sarcomas are characterized by early metastases:
A. lithic osteosarcoma;
B. chondrosarcoma;
C. Ewing's sarcoma;
D. reticular sarcoma;
E. all answers are correct.
(C)
5. CM. Treatment of lymphosarcoma includes:
A. lung therapy;
B. interferon;
C. chemotherapy;
D. radiotherapy;
E. surgical treatment;
(C, D)

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