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CLINICAL APPEARNCE OF PATHOLOGICAL ENTITIES IN OROFACIAL REGION AMONG PEDIATRIC POPULATION

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The lesions presenting in the oral cavity of children are considerably significant in paediatric dentistry. Although they pertain to the oral cavity, they may reflect an underlying systemic condition. These may be clinically misdiagnosed or left untreated owing to lack of parental education, awareness and resources. Their management requires thorough knowledge of the various lesions and accurate clinical assessment for diagnosis, prognosis, treatment and parental counselling. Majority lesions are asymptomatic and benign hence resolve without any intervention. This review article is an overview for recognition of oral lesions most prevalent in children.

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INTRODUCTION

Children are the future of the nation, the flag bearer of society. Children are one third of our population but still our future. Children undergo important changes of life that includes "physical growth and development, emotional changes, search for one's own identity and an evolving role in the society ". Most of the changes will have an impact on health, also with respect of the oral cavity. Abnormalities in the oral cavity can affect the systemic health, growth, and development of children. Likewise, systemic conditions or their treatments can affect the oral cavity or the feasibility of delivering dental care. Paediatric patients can present with various intraoral lesions that require accurate diagnosis, treatment or reassurance, and possible referral for a dental evaluation. Periodic review of oral soft-tissue pathology can help the medical team to easily recognize common and rare abnormalities affecting children. Early detection of these oral conditions may be lifesaving.¹

Problems associated with teeth eruption

Teething- The term refers to eruption of primary dentition. Primary teeth erupt in the 4th to 6th months of a child's life. In most cases eruption of teeth causes no distress to the child or parents but sometimes the process causes local irritation. Genetic factors, gender and gestational age of the child, low birth weight, growth parameters and nutritional status can influence upon the eruption time of the primary teeth. The various clinical features of teething are: Pain, increased salivation, hyperaemia or swelling of the overlying mucosa,

Corresponding author:* **Taniya Thakur Municipality Complex, Flat no-B4, Kanimore, Jhaljhalia patches of erythema on cheeks, flushing of the skin of the adjacent cheeks, general irritability/malaise, bowel upset, loss of appetite.²

Natal and Neonatal teeth- eruption of teeth at or immediately after birth is a relatively rare phenomenon. These have been defined by Massler and Savara³. "Natal" teeth if present at birth and "neonatal" teeth if they erupt during the first 30 days of life.Several terms have been used in the literature to designate teeth that erupts before the normal time, such as congenital teeth, foetal teeth, predecidual teeth, dentitia praecox. 85% of the natal and neonatal teeth are mandibular incisors, 11% are maxillary incisors, 3% are mandibular cuspids or molars and 1% are maxillary cuspids or molars.

Riga fede disease- Early eruption of natal or neonatal teeth causes ulceration on the ventral surface of the tongue, caused by sharp edges of the teeth. Riga Fede disease presents in early infancy and is characterized by firm, verrucous plaques arising on the oral mucous surfaces. These histologically benign lesions occur as a result of repetitive trauma of the oral mucosal surfaces by the teeth. Early recognition of this entity is important, because it may be the presenting signs of underlying neurological disorders.²

Eruption hematoma- A bluish, purple, elevated area of the tissue which occasionally develops a few weeks before eruption of primary or permanent tooth. The eruption cyst (EC) occurs within the mucosa overlying a tooth that is about to erupt. Usually within a few days the tooth breaks and hematoma subside. Because the condition is almost always self-limited, treatment is rarely necessary.²

Eruption sequestrum- is a small spicule of calcified tissue, white in colour that is extruded through the alveolar mucosa that overlies an erupting molar or any other tooth in children. The child may complain of a slight soreness in the area, probably produced by compression of the soft tissue over the spicule during eating and just prior to its breaking through the mucosa.⁴

Epstein's pearls, bohns nodules and dental lamina cyst

Epstein's pearls are non-odontogenic cyst results from epithelial remnants entombed along the fusion line of the palatal halves.^{5,6} They aresmooth, whitish, keratin filled 1-4 mm papules. They resolve in the first 3 months hence treatment is not needed.^{7,8}Dental lamina cysts arise from remnants of dental lamina. These asymptomatic, multiple, 1-3 mm, nodular, creamy white lesions present bilaterally on the anterior aspect of dental ridges. These lesions are present on birth and are self-resolving hence do not require any intervention.^{9,10}Bohn's nodules are remnants of minor salivary gland epithelium. These asymptomatic, smooth, whitish keratins filled nodules or papules ranging from 1-3 millimetre arise on the buccal and lingual aspects of the ridge away from the midline. They resolve in the first 3 months of life and have an incidence of 47.4%.^{5,7}

Developmental disturbances commonly seen in paediatric population

Cleft lip and cleft palate - are common congenital malformations. Failure in the fusion of the nasal and maxillary processes leads to the cleft of the primary palate (unilateral or bilateral). The degree of cleft can vary from a slight notch on the lip to complete cleft of the primary palate. Cleft of the secondary palate is medial. It varies from bifid uvula to complete cleft palate up to the incisive foramen. When it is associated with the primary palate, a complete uni or bilateral cleft lip and palate is formed. Heredity is one of the most important factors to be considered in the etiology. Cleft lip with or without palate is more common in males than in females whereas the isolated cleft palate is more common in females. Cleft lip may occur with a wide range of severity, from a notch located on the left or right side of the lip to the most severe form, bilateral cleft lip and alveolus that separates the philtrum of the upper lip and premaxilla from the rest of the maxillary arch. When cleft lip continues from the incisive foramen further through the palatal suture in the middle of the palate, a cleft lip with palate (either unilateral or bilateral) is present.11

Microglossia- is characterized by an abnormally small tongue. Microglossia may be due to lack of muscular stimulus between alveolar arches, which results in retardation of growth of the mandible in an anterior direction. Microglossia is frequently associated with hypoplasia of the mandible and lower incisors may be missing.¹²

Macroglossia- meaning large tongue. Macroglossia are of two types true macroglossia and pseudo macroglossia. Physical examination of the oral cavity is helpful to deduce true macroglossia from pseudo macroglossia. Commonly occurs in children. Ranges from a mild to severe degree. In infants, it may be manifested by noisy breathing, drooling and difficulty in eating. Pressure on tongue against the teeth produce crenated lateral borders to the tongue, open bite and mandibular prognathism. Tongue may ulcerate, get infected and finally necrosed. Pseudomacroglossia is the term applied to a condition which forces the tongue to sit in an abnormal position: habitual posturing of the tongue, enlarged tonsils or adenoids, low palate, anterior/posterior deficiency in the maxillary ormandibular arches, severe mandibular retrognathism, neoplasm displacing the tongue, hypotonia of the tongue.¹²

Ankyloglossia- It is commonly known as tongue tie. Ankyloglossia results from fusion between the tongue and the floor of the mouth. It is a congenital anomaly that influences the mobility of the tongue as well as the patient's oral hygiene status is also affected.¹³ Ankyloglossia restricts free movement of the tongue leading to cause speech problems. Ankyloglossia results from failing cellular degradation of the tongue frenum between the 8th to 11th prenatal week¹¹. More common in males.¹⁴

Benign migratory glossitis (Geographic tongue)- is a psoriasiform mucositis of the dorsum of the tongue. Its dominant characteristics are a constantly changing pattern of serpiginous white lines surrounding areas of smooth, depapillated mucosa. The central portion of the lesion is inflamed and the border is outlined by a thin yellowish white line or band. The fungi form papilla are retained in the involved areas as small elevated dots.¹⁵The areas of desquamation remain for a short time in one location and then heal and appear in another location. The changing appearance has led some to call this the wandering rash of the tongue.

Cleft tongue- A completely cleft or bifid tongue is a rare condition that is apparently due to lack of merging of the lateral lingual swellings of this organ. A partially cleft tongue is considerably more common and is manifested simply as a deep groove in the midline of the dorsal surface. The partial cleft results because of incomplete merging and failure of groove obliteration by underlying mesenchymal proliferation.⁴

Median rhomboid glossitis- is a condition characterized by a shiny oval or diamond-shaped elevation, invariably situated on the dorsum of the tongue in the midline immediately in front of the circumvallate papillae. Median rhomboid glossitis presents in the posterior midline of the dorsum of the tongue, just anterior to the V-shaped grouping of the circumvalate papillae. The erythematous clinical appearance is due primarily to the absence of filiform papillae, rather than to local inflammatory changes.^{4,11}

Fissured tongue (scrotal or lingua plicata)- is a textured variation of the surface of the tongue that exhibits a prevalence of approximately 2% to 5%. Classically, a groove is present along the midline of the dorsal surface, with an arborisation of smaller groove from this main fissure. Patients are usually asymptomatic, unless debris is entrapped within the fissure or when it occurs in association with geographic tongue (a common finding. On clinical examination, fissured tongue affects the dorsum and often extends to the lateral borders of the tongue. The depth of the fissures varies but has been noted to be up to 6 mm in diameter.¹¹

Hairy tongue (lingua villosa)- is a commonly observed condition of defective desquamation of the filiform papillae that results from a variety of precipitating factors. The condition is most frequently referred to as black hairy tongue (lingua villosanigra). The basic defect in hairy tongue is the hypertrophy of filiform papillae on the dorsal surface of the

tongue, usually due to a lack of mechanical stimulation and debridement. Normal filiform papillae are approximately 1 mm in length, whereas filiform papillae in hairy tongue are more than 15 mm in length. This condition often occurs in individuals with poor oral hygiene.^{4,11}

Lingual thyroid-is an anomalous condition in which follicles of thyroid tissue are found in the substance of the tongue between the circumvallate papillae and epiglottis., possibly arising from a thyroid anlage that failed to 'migrate' to its predestined position or from anlage remnants that became detached and were left behind. A female predominance of 7:1 is seen. The chief symptoms of the condition may vary, but the presenting complaint is often dysphagia, dysphonia, dyspnoea, haemorrhage with pain, or a feeling of tightness or fullness in the throat.^{4,11}

Microdontia- One or more teeth that are smaller than normal. In generalized microdontia, all teeth in both arches are smaller than the normal. The term relative generalized microdontia is used when mandible and maxilla are somewhat larger than normal but teeth are of normal size, giving the illusion of generalized microdontia. When one or two teeth in the jaw are measurably smaller in size while rest of the teeth are normal, the condition is called focal microdontia. Individual teeth frequently affected by microdontia are the maxillary lateral incisors (peg laterals) and the maxillary third molars.¹¹

Macrodontia - When all teeth in both arches are measurably larger than normal, the condition is termed true generalized macrodontia (seen in rare conditions such as pituitary gigantism). The relative generalized macrodontia is used to describe a condition in which mandible or maxilla, or both are somewhat smaller than normal in size. In this condition the arches exhibit crowding of teeth. Regional or localized macrodontia is occasionally seen on the affected side of the mouth in patients with hemifacial hypertrophy and in segmental odontomaxillary dysplasia. Isolated macrodontia is reported to occur most frequently in incisors or canines but also has been seen in second premolars and third molars. In such situations, the alteration often occurs bilaterally.¹¹

Anodontia- It can be defined as a condition in which there is congenital abscence of teeth in the oral cavity. Total anodontia is the congenital absence of all teeth. It is a rare condition in which patient has no deciduous and no permanent teeth usually occurs in association with a genetic disorder such as hereditary ectodermal dysplasia. The more common form of anodontia is partial anodontia, also termed hypodontia or oligodontia, and involves one or more teeth. The most common congenitally absent teeth are the third molars, followed by the maxillary lateral incisors and second premolars.¹¹

Supernumerary teeth- A supernumerary tooth may closely resemble the teeth of the group to which it belongs, i.e. molars, premolars, or anterior possibly from splitting of the permanent bud itself. Brook found that supernumerary teeth were present in 0.8% of primary dentition and in 2.1% of permanent dentition. Occurrence may be single or multiple, unilateral or bilateral, erupted or impacted, and in one or both jaws. Classification: Supernumerary teeth are classified according to morphology and location. In the primary dentition, morphology is usually normal or conical. There is a greater variety of forms presenting in the permanent dentition. Four different morphological types of supernumerary teeth are-conical, tuberculate, supplemental, odontoma.¹¹

Gemination- Geminated teeth are anomalies which arise from an attempt at division of a single tooth germ by an invagination, with resultant incomplete formation of two teeth. The structure is usually one with two completely or incompletely separated crowns that have a single root and root canal. It is seen in deciduous as well as permanent dentition, and in some reported cases, appears to exhibit a hereditary tendency. Prevalence is 0.5 percent in deciduous teeth and 0.1 percent in permanent dentition. Most commonly seen in maxillary anterior region. Geminated teeth show the presence of a single root canal.¹⁶

Fusion- Fused teeth arise through union of two normally separated tooth germs. Depending upon the stage of development of the teeth at the time of the union, fusion may be either complete or incomplete. If this contact occurs early, at least before calcification begins, the two teeth may be completely united to form a single large tooth. If the contact of teeth occurs later, when a portion of the tooth crown has completed its formation, there may be union of the roots only.¹¹

Dilaceration-is an abnormal angulation or bend in the root or crown of a tooth. It may arise from an injury that displaces the calcified portion of the tooth germ and remainder of the tooth is formed at an abnormal angle. The bend may also develop due to presence of an adjacent cyst, tumour or odontogenic hamartoma. It occurs most commonly in permanent maxillary incisors followed by mandibular anterior teeth. Deciduous teeth may be involved due to injury during neonatal laryngoscopy and endotracheal intubation.^{4,11}

Talon's cusp- talon's cusp is an extra cusp present on the lingual surface of the anterior teeth, extending from the cementoenamel junction to the incisal edge. Mostly affects permanent maxillary lateral incisors, followed by maxillary central incisors, mandibular incisors and maxillary canines. It is rarely seen in children and mostly occurs on maxillary central incisors.⁴

Taurodontism- The term 'taurodontism' was originated by Sir Arthur Keith in 1913 to describe a peculiar dental anomaly in which the body of the tooth is enlarged at the expense of the roots. Shaw classified into hypotaurodont, mesotaurodont, and hypertaurodont forms, with hypertaurodontism being the extreme form in which the bifurcation or trifurcation occurs near the apices of the roots and hypotaurodontism being the mildest form. Taurodontism may affect either the deciduous or permanent dentition, although permanent tooth involvement is more common.⁴

Dens in dente (Dens invaginatus)- is a malformation of teeth probably resulting from an infolding of the dental papilla during tooth development. Affected teeth show a deep infolding of enamel and dentine starting from the foramen coecum or even the tip of the cusps and which may extent deep into root. Teeth most affected are maxillary lateral incisors and bilateral occurrence is not uncommon.⁴Clinically, dens invaginatus appears in the tooth crown at the site of an anatomical lingual pit susceptible to caries.¹⁹

Dens evaginatus- is a developmental condition that appears clinically as an accessory cusp or a globule of enamel on the occlusal surface between the buccal and lingual cusps of premolars, unilaterally or bilaterally, although it has been reported to occur rarely on molars, cuspids, and incisors. It is

usually bilateral and shows mandibular predominance. The elevation in the form of cusp consists of normal enamel, dentin and pulp.^{4,11}

Amelogenesis imperfecta- A complex inheritance pattern gives rise to amelogenesis imperfecta (AI), a structural defect of the tooth enamel. It may be differentiated into three main groups: hypoplastic (HP), hypocalcified (HC), and hypomature (HM), depending on the clinical presentation of the defects and the likely stage of enamel formation that is primarily affected.⁴

Dentinogenesis imperfecta- is a hereditary developmental disturbance of dentin in the absence of any systemic disorder. This condition causes the teeth to be discoloured (most often a blue-gray or yellow-brown color) and translucent. Teeth are also weaker than normal, making them prone to rapid wear, breakage and loss. These problems can affect both primary teeth and permanent teeth. Clinically the affected teeth have a reddish brown to grey opalescent colour. Enamel breaks away from the incisal edge of the anterior teeth and the occlusal surface of the posterior teeth.⁴

Dentin dysplasia- is a genetic disorder of teeth, characterized by presence of normal enamel but atypical dentin with abnormal pulpal morphology. Dentin dysplasia type I (radicular type) is characterized by the presence of primary and permanent teeth with normal appearance of the crown but no or only rudimentary root development, incomplete or total obliteration of the pulp chamber and periapical radiolucent areas or cysts. The pulp chamber is sometimes described as having a "crescent shaped" appearance. Dentin dysplasia type II (coronal type) is characterized by primary teeth with complete pulpal obliteration and brown or amber bluish coloration similar to that seen in hereditary opalescent dentin. The permanent teeth have a normal appearance or a slight amber coloration; the roots are normal in size and shape with a "thistle-tube" shaped pulp chamber with pulp stones. In the deciduous dentition, coronal dentin dysplasia bears a resemblance to Dentinogenesis Imperfecta type II.¹¹

Enamel hypoplasia- Local or systemic factors that interfere with normal matrix formation cause enamel surface defect and irregularities called enamel hypoplasia. Enamel hypoplasia may be mild and may result in a pitting of the enamel surface or in development of a horizontal line across the enamel of the crown. Enamel hypoplasia can occur on any tooth or on multiple teeth. It can appear white, yellow or brownish in colour with rough or pitted surface. In most cases, the quality of the enamel is affected as well as the quantity. Environmental and genetic factors that interfere with tooth formation are thought to be responsible for enamel hypoplasia.¹¹

Pulp pathologies in children

Chronic hyperplastic pulpitis- Also called as "pulp polyp" or "pulpitis aperta". Productive pulpal inflammation due to an extensive carious exposure of young pulp. Characterized by the development of granulation tissue, covered at times by epithelium and resulting from longstanding low-grade infection. Most commonly involved are deciduous molars and first permanent molars as they have an excellent blood supply because of large root opening, coupled with high tissue resistance and reactivity in young persons. Seen in children and young adults.⁴

Osteomyelitis- is an inflammation of all aspects of bone, particularly involving the marrow spaces. It is considered as an inflammatory condition of bone that usually begins as an infection of the medullary cavity, rapidly involves the haversian system and quickly extends to the periosteum. Acute suppurative osteomyelitis, focal sclerosing osteomyelitis and osteomyelitis with proliferative periostitis are commonly seen in the pediatric population (first and second decades of life). Acute suppurative osteomyelitis can occur as a result of periapical infection. Dental infection is the etiologic factor in majority of cases of osteomyelitis of mandible and maxilla. More common in posterior mandible in children and anterior maxilla in infants. Neonatal maxillitis occurs in infants and young children. Osteomyelitis is occasionally caused by bacteremia, local oral infection following minor trauma. There is severe pain and trismus. Paraesthesia of lower lip occasionally may be present. Involved teeth become loose and exudation of pus may be seen usually near the lower border of mandible in children.^{4,18}

Cellulitis (phlegmon)- Cellulitis is a diffuse inflammation of soft tissues which tends to spread through tissue spaces and along fascial planes. Facial cellulitis is classified as nonodontogenic and odontogenic, depending on the source of the infection, and asupper or lower face, depending on the anatomical location. Odontogenic facial cellulitis refers to infections arising from the dentition and its adjacent structure. Upper-face infections supporting periodontal involved swelling located above the lip line, including the maxillary dentition and periorbital, maxillary, frontal, nasal, and upper-buccal regions. Lower-face infections were defined as those below the lip line, and included the mandibular dentition and the mandible, floor of the mouth, and cervical regions. An infectious primary lesion was identified when the infection originated from an infected tooth, due to either dental decay or trauma. In contrast, a secondary infectious lesion was attributed to treatments such as restorations or pulptherapies on the infected tooth prior to the occurrence of swelling. Painful swelling of soft tissues that is firm and brawny. Oral examination reveals a dental abscess. Possible complications of facial cellulitis include dehydration, central nervous system impairment, airway obstruction, and even systemic sepsis if treatment is delayed.^{4,11} The other clinical manifestations include abscess formation, blindness, cavernous sinus thrombosis, pulmonary embolism, and death.¹⁹

Gingival diseases in children:

Eruption gingivitis is a temporary type of gingivitis observed in children when teeth are erupting. Usually subsides after the teeth emerge into the oral cavity. Maximum incidence in the 6 to 7-year age group when the permanent teeth begin to erupt. The gingivitis apparently occurs because the gingival margin receives no protection from the coronal contour of the tooth during the early stage of active eruption, and the continual impingement of food on the gingivae causes the inflammatory process.⁴

Recurrent aphthous ulcer- Also referred to as Recurrent aphthous stomatitis (RAS) or canker sore is an unfortunately common condition characterized by painful ulceration on the mucous membrane. Clinical manifestations include minor (ulcers 10 mm in diameter), major (ulcers >10mm in diameter) and herpetiform (showing multiple small pinpoint ulcers) recurrent aphthous ulcers. Minor recurrent aphthous ulcers are

the most prevalent form (80% of all recurrent aphthous ulcers) and their clinical features include round and oval swallow ulcers, with a greyish white psedomembrane in the centre, enveloped by a thin erythematous halo. These ulcers usually heal within 10-14 days without scarring. These ulcers generally cause considerable pain and discomfort, and can interfere with many oral functions such as speaking, eating and swallowing.⁴

Acute necrotizing ulcerative gingivitis (ANUG), also called as "trench mouth". In India, 54-68% of the cases occurred in children below 10 years of age.²⁰Clinical Characteristics: punched out appearance due to ulcerated and necrotic papillae and gingival margins, ulcers are covered by a yellowish-white or greyish slough termed pseudo membranous, removal of the slough results in bleeding and underlying tissue becomes exposed, a foetor exore is often associated, but can vary in intensity. Seldom associated with deep pocket formation as extensive gingival necrosis often coincides with loss of crestal alveolar bone. The involved papillae are separated into facial and lingual portion with an interposed necrotic depression. Swelling of lymph nodes and increased bleeding tendency are of tenpresent. Fever and malaise are not a consistent. The oral hygiene in these patients is usually poor.²¹

Primary herpetic gingivostomatitis- represents the main pattern of primary infection with herpes simplex viruses. More than 90% of PHG cases are caused by the herpes simplex virus type I (HSV-I) and occasionally by herpes simplex virus type II. Occurs in infants and children younger than 6 years of age. The chief complaints are more often dysphagia. After short period, a clustered, short-lived, vesicular eruption appears followed by painful superficial ulcers circumscribed by a red halo. Lesions can occur in any area of oral mucosa and occasionally on perioral skin. Dehydration is common. Other symptoms are halitosis, excessive drooling, and hypersalivation.⁴

Puberty gingivitis- Gingivitis is common in children and adolescents, especially around puberty. It is believed to be related to an increase in steroidal hormone. Granulomatous changes of the gingivae. Peak prevalence is 10 years in girls and 13 years in boys. Crowded teeth and orthodontic appliances may be important contributors as they render difficulty in practicing oral hygiene measures. Mouth breathing could lead to chronically dehydrated gingivae in the maxillary labial area leading to localized gingivitis. Marginal gingival enlargement is seen characterized by prominent bulbous interproximal papillae far greater than gingival enlargements associated with local factors.Usually only the anterior segment of one arch affected. The lingual gingival tissue generally remains unaffected.⁴

Scorbutic Gingivitis- Vitamin C deficiency causes haemorrhage, collagen degeneration and edema of the gingival connective tissue. The involvement is usually limited to the marginal tissues and papillae.²² Gingiva is bluish, soft, and friable and has a smooth shiny surface. Haemorrhage occurring either spontaneously or slight provocation. Surface necrosis with pseudo membrane formation and necrosis occur as a result of infarcts created in the capillaries supplying the gingiva.²³

Periodontal diseases in children

Localized aggressive periodontitis (lap)- Onset around the time of puberty in an otherwise healthy individual. Aggressive periodontal destruction localized almost exclusively to the incisors and first molars. Familial pattern of occurrence. Low incidence from 0.1 to 2.3 percent. It is more common in girls and it is usually characterized by a lack of the common clinical signs of periodontal disease (inflammation, bleeding, heavy plaque).⁴

Papillon-lefèvre syndrome-It is an inherited disease usually manifested in childhood. Males and females are equally affected. Characteristic features include hyperkeratotic skin lesion, severe destruction of periodontium and in some cases, calcification of the dura. Early inflammatory changes followed by rapid bone loss results in exfoliation of teeth. All the primary teeth may be lost by 5 to 6 years of age. Permanent dentition erupts normally, but they too are lost within a few years due to destructive periodontal disease. There is premature loss of primary teeth, no gingival inflammation, loss of alveolar bone and absence of cementum.⁴

Cysts in the paediatric population:

Eruption cyst-This soft tissue benign cyst arises around an erupting tooth when the dental follicle separates from the crown of tooth and results in fluid collection within this space.^{24,25}It is dome shaped and may appear normal to blueblack, purple or brown in colour subject to the amount of blood in the cystic fluid after trauma (hematomas). It may also be transparent. Since the tooth erupts through the lesion it resolves. The prevalence of eruption cysts of is 22%.^{26,27}

Dentigenous cyst can be defined as an odontogenic cyst that surrounds the crown of an impacted toothcaused by fluid accumulation between the reduced enamel epithelium and the enamel surface, resulting in a cyst in which the crown is located within the lumen. This is one of the most common types of developmental odontogenic cyst, estimated to be about 20% of all jaw cysts (2nd highest in occurrence). Occurrence in mandible is more than in maxilla with the ratio of 2:1. The most common sites of this cyst are the mandibular and maxillary third molar and maxillary cuspid areas, since these are the most commonly impacted. Expansion of bone with subsequent facial asymmetry, extreme displacement of teeth, severe root resorption of adjacent teeth and pain are all possible sequelae brought about by continued enlargement of the cyst.^{4,11}

Odontogenic keratocyst- It is named odontogenic keratocyst because the epithelial lining of cyst produces keratin that gets filled within the cystic lumen. Seen in 1st to 9th decade of life. Seen more often in males as compared to females. Incidence is 12 to 13 percent of total jaw cysts. The mandible is involved twice as frequently as maxilla, with the most common site of origin being the mandibular third molar region and ramus of the mandible, followed by the maxillary third molar region, mandibular first and second molar area, maxillary canine area and mandibular premolar region. Signs and symptoms associated with the cyst often present as pain, swelling, discharge and paresthesia of lower lip.^{4,11}

Nasopalatine duct cyst (NPDC)- is a developmental, nonneoplastic cyst that is considered to be the most common of the nonodontogenic cysts. NPDC is one of many pathologic

processes that may occur within the jawbones, but it is unique in that it develops in only a single location, which is the midline anterior maxilla. Males are affected 1.8 to 20 times more often than females.⁴

Mucocele- This is a bluish, well-circumscribed, translucent, fluctuant swelling. It arises on the lower lip lateral to the midline when the excretory duct of a minor salivary gland ruptures due to mechanical trauma and mucin leaks into the surrounding connective tissues with in a fibrous capsule. It may be normal or whitish and keratinized.²⁸ It may also appear on retromolar region, buccal mucosa, ventral tongue surface and floor of the mouth as a ranula. Superficial mucocele resolves on bursting spontaneously with a shallow ulcer. Treatment minimizes the risk of recurrence.^{29,30}

Ranula- The term ranula is used to describe mucoceles occurring on the floor of mouth. They are mostly found associated with submandibular or sublingual glands. They are usually unilateral. Incidence in children and young adults is more common. Occurrence in the floor of the mouth is common, where they appear translucent blue in colour, that resembles a frog's belly, hence the name. They are small fluctuant swellings. Larger lesions may lift the tongue and cause difficulty in swallowing and speech. The swelling increases in size during meals due to increased secretory activity as a part of gustatory stimulation. Herniated projections of sublingual gland as a result of hiatus or deficiency between anterior and posterior parts of mylohyoid muscle had been reported as etiological factors for plunging ranulas.⁴

Benign tumors of the oral cavity

Congenital epulis of newborn- Congenital Epulis of newborn is a rare gingival tumour that occurs along the alveolar ridge. Clinically it presents as a smooth well defined erythmatous masses arising from gum pad. Size may be large enough to lift the upper lip. The lesion is most common in females, with a female-to male ratio of 8:1, and is three times more common in the maxilla than the mandible.⁷⁶CE clinically appears as a pedunculated, which may interfere with respiration or feeding. Appears as smooth-surfaced, pedunculated, protuberant and sometimes lobulated tumor arising from the alveolar crest of newborn infants.⁷⁷

Hemangioma- This is a benign vascular neoplasm emerging as a macule on birth but may appear a few weeks after and regresses into spotted pigments. The course of disease follows a rapid proliferating phase (0-1 yr), involuting phase (1-5 yr) and involuted phase (5-10 yr). It appears on neck and head, trunk, extremities, lips, tongue, buccal mucosa, palate and uvula [3]. Predisposing factors include infantile age, infant birth weight, childbearing age, gestational hypertension, Kasabach-Merritt syndrome.³⁴

Melanotic neuroectodermal tumor of infancy- This rare pigmented benign neoplasm appears in the first 6 months with a male predilection. It arises from the neural crest cells and may be located on the tongue, buccal mucosa, palate or floor of the mouth. It may be present in the craniofacial region, brain, skull, maxilla, mandible and the genitals. It is a painless, expansile, non-ulcerative rapidly growing, pigmented, lesion with a locally aggressive behaviour. Recurrence is high alongside metastasis and malignant transformation rate.^{35,36} Pvogenic granuloma- Benign vascular lesions that occur most commonly on oral mucous membranes of children caused by bacterial infection and exuberant tissue response to local irritation or trauma. Incidence of occurrence in children and young adults with male predilection in children and female predilection in young adults. Usually occurs as a pedunculated or sessile, smooth and lobulated mass. Surface is characteristically ulcerated and colour ranges from pink to red to purple. Young lesions appear reddish due to high vascularity as compared to older lesions that appear pinker due to presence of collagenized tissue. Bleeds easily due to extreme vascularity. In the oral cavity, it occurs most commonly on the gingiva, where gingival inflammation and irritation resulting from poor oral hygiene act as precipitating factors. Other common sites in the oral cavity are lip, tongue and buccal mucosa.11

Odontogenic tumor in children:

Odontoma- Odontomas are benign tumors of odontogenic origin. The cause of odontoma is unknown, but it is believed to be hereditary or due to a disturbance in tooth development triggered by trauma or infection. Odontomas may be either compound or complex. Budnick³⁸reported that compound odontomas have a propensity for occurance in canine and incisor region, being found more often in the maxillary that in mandible whereas complex odontomas show a predilection for occurance in the posterior jaws. Phlipsen, Reichart and Praetorius³⁹ also found that posterior mandible to be the most frequent site of occurance followed by the anterior maxilla. Although odontomas are usually asymptomatic, they may be the cause of noneruption or impaction of teeth and retained primary teeth.

Infectious diseases in children:

Candida albicans (moniliais, thrush) is caused by an overgrowth of candida albicans. Acute pseudomembranous candidiasis: This is the form most often seen in children. It occurs mainly in debilitated or chronically ill children. Oral lesions include soft, white, slightly elevated plaque occurring on buccal mucosa, tongue, gingiva and floor of the mouth called as thrush. The plaques consist of masses of fungal hyphae with intermingled desquamated epithelium, keratin, fibrin, necrotic debris and leukocytes.⁴

Scarlet fever- is a systemic infection caused by group A, β -*H. streptococci.* The disease is the most prevalent in children between 3 and 12 years old.First, the disease begins as a streptococcal tonsillitis with pharyngitis. During the first 2 days, a white coat covers the dorsum of tongue (white strawberry tongue). After desquamating of white coat, erythematous dorsal surface of tongue appears (red strawberry tongue). Within the first 2 days, the disease is associated with elaboration of an erythrogenic toxin which attacks the blood vessels and develops the cutaneous rash.⁴

Diphtheria- It is an acute, life-threatening, contagious bacterial infection caused by gram-positive bacillus, Coryne bacterium diphtheria. The infection mainly spreads via droplet inhalation. Incubation period ranges from two to five days. Occurrence in children is most common, especially during winter season. A patchy, yellowish-white thin film covered by greyish adherent membrane is seen known as "diphtheritic membrane". Raw bleeding surface is seen left when this membrane is stripped off. Due to involvement of soft palate,

uvula, larynx and trachea, there occurs sore throat, stridor and respiratory difficulties. In severe cases, paralysis of soft palate can be seen.⁴

Syphilis- Children encounter two forms of syphilis: acquired syphilis, which is almost exclusively transmitted by sexual contact and congenital syphilis, which results from transplacental transmission of spirochetes. Oral manifestations show presence of 'mulberry molars' with constricted and atropic cusps, 'screwdriver shaped' incisors. Rhagades, i.e. fissuring and scarring of corners of mouth may be seen. Frontal bossae and saddle nose are the characteristic features in infants suffering from syphilis. Hutchinson's triad of hypoplasia of incisors and molar teeth, eight nerve deafness and interstitial keratitis in eyes is seen.^{4,11}

Aids- Caused by human immune deficiency virus. Virus transmission may occur to the foetus in the pregnancy as early as the first trimester but infection is common perinatally. Children may also get the infection from blood transfusion or blood products. Pseudo membranous and erythematous candidiasis are the most common variants. The pseudo membranous variant may be located anywhere in the mouth. The erythematous variant is usually located on the dorsum of the tongue and the palate. Both types are almost equally likely to manifest. Angular cheilitis is often associated with Candida albicans, and may be seen in HIV-infected patients. Intraoral and lip herpes simplex is a relatively frequent oral viral infection. Oral herpes zoster is a rare occurrence in HIVinfected patients. Hairy leucoplakia is a common oral mucosal feature that has been described among all high-risk groups for HIV infection. Clinically, hairy leucoplakia presents as a whitish, slightly elevated, non-removable lesion of the tongue, often bilaterally. Lesions may extend onto the ventral surface and the dorsum of the tongue. Clinically, HIV-associated gingivitis (HIV-G) is characterized by a fiery red band along the margin of the gingival. Clinically, HIV-associated periodontitis (HIVP) is characterized by soft tissue ulceration and necrosis, and rapid destruction of the periodontal attachment apparatus. Spontaneous bleeding and severe deep pain are common.^{5,6}

Diseases of bones and joints:

Cherubism- is a rare benign hereditary conditionwhich affects only the jaw bones. The disease is characterized by "bilaterally symmetrical enlargement" of mandible or sometimes the maxilla. Cherubism is a disease of childhood that usually presents before the age of five, most often between 12 and 36 months. Males are affected more commonly than females. Wide rim of exposed sclera is noted below the iris, therefore called 'eve to heaven' appearance. Mandibular lesions appear as painless, bilateral expansion of the posterior mandible that tends to involve angles and ascending rami. These along with excessive cheek fullness give rise to typical appearance of "chubby face" to the child. Maxillary involvement is rare but if it occurs, it mainly affects the tuberosity area. Patients with cherubism commonly exhibit increased cheek fullness, expansion and widening of the alveolar ridge, flattening of the palatal vault. Displacement or failure of eruption of tooth, speech difficulty, loss of normal vision or hearing, breathing difficulties.11

Fibrous dysplasia- is a developmental tumour like condition that is characterized by replacement of normal bone by an excessive proliferation of cellular fibrous connective tissue

intermixed with irregular bony trabeculae. It results from a post zygotic mutation in the GNAS I (guanine nucleotidebinding protein, a-stimulating activity polypeptide I) gene. Clinically, fibrous dysplasia may manifest as a localized process involving only one bone, as a condition involving multiple bones, or as multiple bone lesions in conjunction with cutaneous and endocrine abnormalities. When the disease is limited to a single bone- monostotic fibrous dysplasia. This type accounts for about 80% to 85% of all cases, with the jaws being among the most commonly affected sites. Involvement of two or more bones is termed polyostotic fibrous dysplasia. The number of involved bones varies from a few to 75% of the entire skeleton. When seen with cafe au lait (coffee with milk) pigmentation, the process is termed Jaffe-Lichtenstein syndrome. Polyostotic fibrous dysplasia also may be combined with cafe au lait pigmentation and multiple endocrinopathies, such asexual precocity, pituitary adenoma or hyperthyroidism. This pattern is known as the McCune-Albright syndrome. The skull and jaws may be affected with resultant facial asymmetry. Pathologic fracture with resulting pain and deformities frequently noted.¹¹

Osteogenesis imperfecta- is a genetically transmitted disease of bone characterized by defective matrix formation and lack of mineralization, which results in an increased bone fragility. Bowing deformity of the bone with multiple fractures due to increased fragility. Blue sclera with defective teeth in the form of 'bulbous crowns, dentinogenesis imperfecta and blue or brown translucency (opalescent teeth)'. Large head size, frontal bossing, maxillary hypoplasia. Class III malocclusion with anterior and posterior crossbite.Severe attrition of deciduous teeth. Multiple impacted permanent teeth. Excessive bruishing tendency.Increased incidence of development of osteitis and osteomyelitis following extraction of teeth.¹¹

Oral manifestations of haematological disorders in children:

Thalassemia- Clinical manifestations are seen within the first year of life. The most common orofacial manifestations are due to intense compensatory hyperplasia of the marrow and expansion of the marrow cavity and a facial appearance known as "chipmunk" face: enlargement of the maxilla, bossing of the skull and prominent molar eminences.^{39,40}Overdevelopment of the maxilla frequently results in an increased over jet and spacing of maxillary teeth and other degrees of malocclusion.^{41,42,43,44}Tightness of the upper lip are evident. The maxillary alveolar bone and the palate show a retruded position. Mandible becomes less enlarged compared to the maxilla, possibly because the dense cortical plates of the mandible prevent the expansion. Pain and swelling of the parotid glands and atrophic candidiasis have also been reported in these patients.

Sickle cell anaemia- usually presents in childhood. Robinson and Sarnat, 1952, stated that 79.2 percent patients show jaw abnormalities.⁴⁵Orofacial pain, paraesthesia of the mental nerve, mandibular osteomyelitis, prominent maxilla with severe malocclusion, acute facial swelling, gingival enlargement and buccal mucosal pallor. Hypo mineralization of dentin is seen. Interglobular dentin in the periapical area is evident with inclusions in the peri pulpal dentinal tubules. Abrupt alteration of dentin genesis with denticle like calcified bodies in the pulp chamber may be seen. Proliferation of cementum in the apical and mid root areas may be evident.

Haemophilia- Haemophilia may be a challenge to the dental specialists due to the induction of bleeding during treatment, which may be fatal in certain cases. Tooth displacement may be present. Haemorrhage from many sites in the oral cavity is a common finding in haemophilia, and gingival haemorrhage may be massive and prolonged. Petechiae, ecchymoses are very common.

Thrombocytopenic purpura- One of the prominent manifestations of thrombocytopenic purpura is the severe and often profuse gingival hemorrhage which occurs in the majority of cases. This hemorrhage may be spontaneous. Petechiae occur on the oral mucosa, commonly on the palate, and appear as numeroustiny, grouped clusters of reddish spots only a millimeter or less in diameter. Actual ecchymoses do occur occasionally. The tendency for excessive bleeding contraindicates any oral surgical procedure, particularly tooth extraction, until the deficiency has been compensated.^{4,11}

Leukaemia- Acute lymphoblastic leukaemia mainly occurs in children under 10 years. Factors that have been implicated to be of etiologic significance are radiation injury, chemical injury, genetic factors – Down's syndrome, immune deficiency and viral infections. Gingiva appears as swollen, glazed, and spongy tissue which is red-deep purple in appearance with gingival bleeding. Enlargement may appear as a diffuse enlargement of the gingival mucosa, an oversized extension of the marginal gingiva, or a discrete tumour like interproximal mass. It is moderately firm in consistency, but there is a tendency toward friability and haemorrhage, occurring either spontaneously or on slight irritation.⁴⁶

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