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Developmental Disturbances Of Lip And Palate – A Brief Review

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Abstract:

The most prevalent birth abnormality affecting the orofacial region that results from abnormal embryonic development is cleft lip and palate. It is a congenital condition that is characterized by the child's facial and oral cavity splits. Van der Woude syndrome (VWS) is an uncommon craniofacial abnormality that exhibits exceptional expression heterogeneity and autosomal dominant inheritance. The small salivary glands of the lips are the primary target of cheilitis glandularis (CG), an inflammatory disease with an unclear etiology. Melkersson-Rosenthal syndrome, which consists of recurrent orofacial edema, recurrent facial nerve palsy, and tongue fissuring, is another name for cheilitis granulomatosa.

Keywords: Cleft lip and palate, Van der Woude syndrome, Cheilitis glandularis, Cheilitis granulomatosa

Introduction:

The intricate process of lip and palate development necessitates precise synchronization of cell migration, proliferation, differentiation, and apoptosis[1]. The first branchial arch is the source of the oral cavity's structures. The two maxillary, two mandibular, and frontonasal processes are apparent by the end of the fourth week of development.The upper lip merges by 6 weeks gestation, while the face and palate finish midline fusion between 6 and 12 weeks gestation[2]. Cleft lip, cleft palate, or cleft lip and palate are the outcomes of midline fusion failure during embryogenesis. In weeks five and six of embryonic development, the medial nasal tissues unite to create the lip; in weeks six through ten, the anterior hard palate, alveolus, and philtrum form[3]. During weeks 10 to 12 of pregnancy, the uvula and soft palate unite to form the posterior hard palate, which is created by the maxillary prominences. The prevalence of cleft lip and palate, a common congenital defect, is 17 per 10,000 live births[4].One of the most prevalent clefting syndromes is van der Woude syndrome (VWS). Congenital lower lip pits along with cleft lip, cleft palate, or both[5,6] are the typical presentation in patients with VWS. Cheilitis glandularis (CG) is an uncommon inflammatory disease that mostly affects the lips' surrounding tissues and small salivary glands. Almost only white people are affected, and adults (over 40) are more affected than young ones. The male to female involvement ratio is 3:1. As of yet, no particular cause or contributing factor has been linked to the start of the condition[7]. Cheilitis granulomatosa is an uncommon and distinct illness. Granulomatous inflammation causing persistent swelling of one or both lips is the hallmark of this illness. The causes of these lip and palate developmental abnormalities could include anticonvulsant medications taken by

the mother during pregnancy, which could produce an orofacial abnormality in the unborn child. Numerous additional elements also play a role in the development of clefts. These include circulating drugs like alcohol, pollutants, and vitamin deficiencies, especially those related to folic acid and vitamin A. Other contributing causes include mechanical abnormalities where the size of the tongue may inhibit the fusion of palatine shelves, lack of genetic developmental force, physical, emotional, and traumatic pressures during pregnancy, and a damaged vascular supply to the area. The precise etiology is still unknown after several clinical and experimental studies, but genetics remains the most significant contributing element[8].

Cleft Lip and Cleft Palate:

Clefts of the palate and upper lip are the most prevalent craniofacial birth abnormalities [Leite and Koifman, 2009]. Although environmental, syndromic, and genetic variables have all been implicated, the exact cause of clefts is still unknown. Although midline lip cleft is uncommon, cleft lips can be unilateral, bilateral, median, or limited to the soft palate (mucous cleft). Females without a racial preference are more likely to have isolated cleft palates[4]. Palatal clefts, lip and palatal clefts, lip clefts, and lip and alveolar clefts are the four primary types of lip and palate clefts. The definition states that the maxillary alveolus is not included in palatal clefts. Another kind of palatal cleft is a submucosal cleft, which occurs when the cleft is concealed by mucosa. Both the soft and hard palates, or just the soft palate, may be affected by palatal clefts. Both unilateral and bilateral lip and palatal clefts can reach the hard and soft palates by passing through the lip and alveolar ridge. Males and people of Asian and Caucasian heritage are more likely to have cleft lip with palate, which is twice as prevalent as solitary cleft palate. A condition is more likely to be linked to cleft palate without cleft lip[3].Organic solvents and agricultural chemicals, particularly pesticides, as well as viral infections, metabolic disorders, and illegal narcotics are teratogen exposures linked to the formation of clefts. Diazepam, phenytoin, phenobarbital, topiramate, and lamotrigine are antiepileptic drugs linked to orofacial defects[9–11]. Numerous genetic diseases, including as trisomy 13, oral-facial-digital syndrome, Treacher Collins syndrome, Van der Woude syndrome[12], and the spectrum of chromosomal 22q11, are linked to cleft lip and/or palate. Another cause of cleft palate is mechanical interference with palatal fusion in the fetal tongue, which typically results from micrognathia with mandibular hypoplasia, sometimes referred to as Pierre Robin syndrome or Robin sequence. In addition to rigorous feeding support and airway screening, the newborn with cleft lip and/or palate needs to have their hearing and genetic makeup evaluated[13]. Primary palatal repair and surgical primary lip repair are frequently performed at 3 and 9 months of age, respectively. After all procedures, residual hypernasal speech and language deficits are typical. A multidisciplinary cleft and craniofacial team consisting of skilled practitioners from the medical, surgical, dental, and allied health fields achieves the best possible care[14].

Van der Wounde Syndrome:

Demarquay published the first account in the literature in 1845 [15], and Van der Woude provided a thorough and in-depth description of it in 1954 [16]. The syndrome has a high and incomplete penetrance (80–100%) and is genetically transmitted in an autosomal dominant form with variable manifestation[17]. Approximately 2% of patients with clefts develop lip pits[18]. A second VWS locus (VWS2-OMIM) has been mapped at 1p34[19], but the majority of VWS cases have been associated with chromosome 1q32-q41, also known as VWS locus 1. The distribution of sexes is equal, and there is no sex-related difference in expression[20]. Three forms of congenital lip pits can be distinguished based on where they are located: commissural, midline upper lip, and lower lip[21]. The elimination of sinuses and cosmetic relief from the deformity are the two objectives of treatment. Complete excision of sinus tracts, correction of aberrant elevations and protrusions, and preservation of the orbicularis muscle ring are all necessary for the best possible lip function when using the suggested surgical procedure. The prevention of subsequent malformations, like whistling abnormalities, is also very important. However, because of the dismal surgical results, other methods have been proposed, including resection with AlloDerm graft implantation, the split lip advancement technique (SLAT), and vertical wedge resections. The inverted-T lip reduction technique was presented as a substitute treatment for lower lip pits more recently[22]. Genetic counseling is recommended since a cleft patient with lip pits has a 10-fold increased chance of having children with cleft lip, with or without cleft palate, compared to

those without lip pits. It should be noted that VWS is an autosomal dominant mode of transmission, meaning that all parents are at 50% risk of having a kid with cleft lip and/or cleft palate [23].

Cheilitis Glandularis:

Despite being an uncommon disease, CG can occasionally be seen by the doctor. The primary cause of the lesion's appearance is a process of renewed yellowish plaque production that baffles medical practitioners. Topical corticosteroids don't seem to work well on their own. Although intralesional steroid injections have gained some popularity, there is disagreement about whether they are always effective. Vermilionectomy has long been the standard of care for CG, but it has side effects include paresthesia and persistent itching. There was ductal ectasia of the small salivary glands, fibrosis inside the glands, and chronic inflammation with varying degrees of nonspecific sialadenitis as the predominant nonspecific histological findings[24]. Contact cheilitis and cheilitis granulomatosa are two potential differential diagnosis for CG[25,26]. Food allergies, primarily to cinnamon and benzoate chemicals, may be linked to the development of cheilitis granulomatosa in a small number of cases[27]. Tacrolimus had a key role in avoiding crust development, presumably as a result of its ability to effectively reduce glandular inflammation by reducing the small salivary glands release of proinflammatory cytokines.

Cheilitis Granulomatosa:

Cheilitis granulomatosa is an uncommon and distinct illness. Granulomatous inflammation causing persistent swelling of one or both lips is the hallmark of this illness. This uncommon inflammatory condition was initially identified by Miescher in 1945[28,29]. It is an incomplete or monosymptomatic type of Melkersson-Rosenthal syndrome (MRS), which is characterized by recurrent facial nerve palsy, fissuring of the tongue, and orofacial edema. Wiesenfeld first described Orofacial Granulomatosis in 1985[30]. CG is also regarded as a subset of this rare disease. The cause of CG has not yet been determined. Some cases may exhibit an autosomal dominant inheritance pattern, according to reports, with the relevant gene mapping to chromosome 9 p11 [31]. Other authors have suggested a variety of explanations, such as autoimmune mechanisms, allergic reactions, persistent infections odontogenic foci, linkages to sarcoidosis and Crohn's disease, or even oral manifestations of systemic disorders[32]. Young adulthood is often the onset age, and there is no racial or sexual preference. The sample showed variable-thickness hyperplastic epithelium coated in fibrinopurulent exudate. The lamina propria on the surface was edematous and loose. The underlying fibrocellular connective tissue showed perivascular lymphocyte aggregations, Langhans type multinucleated giant cells, and many non-caseating granulomas. There are several known therapies for CG, antibiotics such tetracycline and clofazimine[33,34], tranilast[35], oral and intralesional steroids[36,37], surgical resection[38]. Quick enhancement In situations of severe disfigurement, surgical surgery and radiation therapy have been suggested as medical options for CG, and post-operative relapses are frequent[39].

Conclusion:

One of the most common birth abnormalities and a serious health problem is the orofacial cleft. The highest number of dental abnormalities are found in children with the most severe clefts. Tooth developmental problems that manifest as oligodontia, supernumerary teeth, hypoplasia, or hypomineralization are frequently linked to clefts. Both treatment planning and budgetary estimates should take these observations into account. A rare disorder with a notable clinical appearance is VWS. It may be linked to either cleft palate or cleft lip. Lip pits are the only sign of VWS, in which case cosmetic surgery may be explored. With information on the inheritance pattern and the consequences of these traits highlighted, genetic counseling is essential for impacted parents and patients. Chelitis Grandularis may be more severe in albino patients and is closely linked to sun sensitivity. Squamous cell carcinoma may be more likely to develop on the swelling, sun-exposed lip. Treatment for Melkersson-Rosenthal syndrome is challenging because it is an uncommon

condition. It highlights the value of food allergy testing as well as the part food allergens play in the etiopathogenesis of the illness.

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