Oblique facial cleft, from an odontological point of view

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Abstract

Introduction: Craniofacial clefts are the result of disruption of the normal developmental program during embryogenesis affecting the face, skull or both. Objective: To analyze the literature on oblique facial clefting, highlighting the different subtopics such as its etiology and prevalence, clinical manifestations, diagnosis, treatment and dental management. Methodology: Articles published in databases such as PubMed, SCOPUS and Google Scholar were analyzed, with emphasis on the last 5 years. This was done using keywords such as “oblique facial cleft”, with the subtopics “etiology”, “oral management”, “diagnosis”, “epidemiology” and “treatment”. Results: The oblique facial cleft is one of the rarest forms, its incidence is believed to be in the range of 0.24% of all facial clefts. Its etiology is embryologically a failure of the lateral and maxillary processes along the naso-optic sulcus. Its clinical manifestations are very variable, but a cleft can be found originating from the Cupid’s bow and can involve the alveolus between the lateral and canine, passing laterally from the medial piriform opening to the infraorbital foramen and ending in the lower orbit. Diagnosis is made in the 2nd and 3rd trimester, based on sagittal, coronal and axial views of the fetus. Treatment is based on the protection of vision and corneas, in addition to creating an adequate rehabilitation. Conclusion: As it is a manifestation that involves the oral region, a dental team is required to complete its general treatment.

Keywords: Oblique Facial Cleft, “Etiology”, “Oral management”, “Diagnosis”, “Epidemiology” and “Treatment”

1. Introduction

Craniofacial clefts are the result of disruption of the normal developmental program during embryogenesis affecting the face, skull or both [1]. Oblique facial clefts are among the rarest clefts and few cases have been reported in the literature [2]. These are defined as clefts extending between the nasolateral and maxillary prominences combined with the cleft lip include types 2-6 of the Tessier classification [3, 38, 39, 40]. Most of these arise sporadically and are attributed to a failure of fusion between the medial nasal prominence and the maxillary and lateral nasal processes [4]. They are in the paramedial line of the facial structures, especially in the orbit [5], extending from the nasal ala to the medial canthus, variably affecting the skin, soft tissues, cartilage, bone and mucosa [6]. Generally, there is severe asymmetry and multiple facial areas are affected, with the passage of time the deformities become more obvious and a clear three-dimensional underdevelopment of hard and soft tissues results [7]. Clefts vary in severity and have individual differences, this must be measured, as well as their distribution to correct them surgically [8]. There is some controversy in the treatment options and timing, the priorities in these cases include airway patency, feeding and corneal protection [9].

Therefore, we must be very clear about what this malformation consists of and what it entails, as well as its causes and phenotypic characteristics, to be alert and be able to make a timely diagnosis in patients so that they can be treated appropriately. In the dental area, its importance lies in the affection of the alveolar bone, dental treatments should meet the same objectives as in any other patient, but we must have certain considerations and work in a multidisciplinary
manner with a well stipulated treatment plan. The objective of this review was to analyze the literature on oblique facial cleft, highlighting the different subtopics such as etiology, epidemiology, diagnosis, treatment, and dental management.

2. Materials and methods
Information from articles published in PubMed, Science Direct and EBSCO was analyzed with emphasis on the last 5 years. The quality of the articles was analyzed based on the PRISMA guidelines, i.e., identification, review, choice, and inclusion. The quality of the review was assessed using the measurement instrument for evaluating systemic reviews (AMSTAR-2) [10]. The search was performed using Boolean logical operators AND, OR and NOT. It was realized with the words "oblique facial cleft", "etiology", "epidemiology", "diagnosis", "treatment", and "dental management", in conjunction with logical Boolean operators Or y And.

3. Results & Discussion
3.1 Etiology and Prevalence
Craniofacial clefts have a prevalence of 1.43 to 4.85 per 100,000 births [9, 11, 12]. Less frequently occurring in an oblique manner, therefore, it is the rarest form, its incidence is not estimated, but is believed to be in the range of 0.24% of all facial clefts [13, 14]. Its etiologic factors are not clear, it usually has no familial tendency, syndromic association or gender predominance. Embryologically it is a failure of the lateral and maxillary process, along the naso-optic sulcus [15]. The loss of some elements in craniofacial bone formation, by virtue of its downward cascading nature causes failure of bone formation and fusion, creating a large defect [16].

Its cause is mainly attributed to a primary developmental arrest, neurovascular insufficiency or tears in the developing maxillary process, recent studies suggest that they are caused by a combination of amniotic band migration and increased local pressure that produces cellular ischemia [14]. On the other hand, studies also report that injury or rupture of the stapedial artery early in embryogenesis has been postulated as a potential cause [17]. Genetic studies have revealed that disruption in the SPECCIL protein results in oblique facial clefts, demonstrating that these have a genetic basis [18]. This protein plays an important role in adherens junctions involved in cell adhesion, actin cytoskeleton organization, microtubule stabilization, and bone organization and cytokinesis. Thus, it controls the delamination of cranial neural crest cells during facial morphogenesis [19].

Although the prevalence of the oblique facial cleft is not yet estimated, it is believed to be found in 0.24% of all facial clefts. The etiology is clearly attributed to embryologic failure, as there is a failure of closure of the lateral and maxillary processes.

3.2 Clinical Manifestations
It has a wide clinical presentation [15, 20]; in its phenotype the cleft runs vertically through the lacrimal portion of the lower eyelid, lateral to the lacrimal puncta, through the infraorbital rim and orbital floor, medial to the infraorbital nerve, through the maxillary sinus and cheek [21]. It originates from the Cupid’s bow and may involve the alveolus between the lateral and canine, passes laterally from the medial piriform aperture to the infraorbital foramen, the cleft terminates in the lower orbit. Soft tissue and skeletal defects may be observed, in addition the lacrimal apparatus may be involved, the inferior lacrimal canaliculus is usually hypoplastic or absent; the cleft may end in an association of coloboma and the orbital contents may prolapse into the maxilla, in addition to presenting microphthalmos or anophthalmos [20]. In the alveolus, the cleft is in the usual position for a complete cleft palate [31]. Speaking of patients' self-esteem, it has been reported that in addition to facial deformities, patients may present psychological, social and physical stress [17]. Because the face is the primary means by which human beings interact with each other and is the main method of emotional expression and social interaction [22].

Its manifestations are very broad, we can observe a cleft that originates from the Cupid’s bow to end in the lower orbit, involving soft tissue and skeletal defects; psychosocial factors are important in these patients by influencing their self-esteem.

3.3 Diagnosis
It has been reported that clefts are usually diagnosed during the second and third trimester of pregnancy, based on sagittal, coronal and axial views of the fetal head and face [23]. Accurate prenatal diagnosis of these is critical to establish long-term treatment planning, prognostic prediction and parental counseling [24]. Imaging and assessment of the deformity play an important role [25], transabdominal ultrasound represents the first approach [26]. An accurate assessment of these malformations is often seen on ultrasound performed during pregnancy but the accuracy of this depends on the experience of the sonologist, the fetal position, amount of amniotic fluid and the type of cleft we are dealing with [27]. Three-dimensional ultrasound and prenatal MRI improve diagnostic accuracy by providing a more accurate picture of the defect [28]. All this requires a multidisciplinary team involving different areas such as genetics, obstetrics, neonatology, pediatrics, radiology, maxillofacial surgery and medical ethics, in order to proceed with the prognosis [28].

Despite being present before birth, this team continues until the patient reaches skeletal maturity [29]. The diagnosis of these manifestations can be made in the second and third trimester of pregnancy with the help of imaging by the physician, it is important to have a multidisciplinary team from the moment these manifestations are detected in order to issue a prognosis and advise the parents.

3.4 Treatment
Facial clefts are a challenge for plastic surgeons because they can involve soft tissues or facial bones, even both [20]. Their repair is anatomical in all cases [17]; an important step in their treatment would be a better understanding of their genetic pathogenesis [18]. The main objective of its rehabilitation is to protect the vision or corneas; the soft tissue component can be treated in the first month of life; for patients with more accentuated deformities, each facial component should be evaluated in order to restore and give a more natural appearance [13]. Their repair is usually complicated, with more complex surgeries; multiple surgical procedures are required in early infancy, along with lifelong multidisciplinary treatment [11, 18]. Minimally invasive techniques are now included, with minimal incisions to achieve a natural appearance [30]; although this may involve maxillary bone grafting, cheek flaps, rhinoplasty and transcranial correction of orbital dystopia [11], in addition to the use of tissue expanders, along with autologous soft tissue repair and bone grafting [31]. The treatment represents a challenge for surgeons due to the amount of tissue involved; its goal is vision or corneal

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protection. Minimally invasive surgery with the aid of bone grafting is currently used to also give a more natural and esthetic appearance.

3.5 Dental Management

In addition to soft tissue and bone abnormalities, the alveolar and palatal regions may be affected, generating a cleft-related maxillary discontinuity with asymmetry and disproportion [32]. Collapse of these maxillary elements is frequently observed and demonstrates narrowed and recessed dental arches [33]. For improved esthetics and function, cleft management includes maxillary plastic surgery, which can have complications such as marginal bone destruction, root resorption and lesions in dental organs [34]. To treat such root resorption, endodontic therapy followed by internal dressing with calcium hydroxide is recommended to stop resorption [32]. Calcium hydroxide increases dentin pH and inhibits hydrose activity while activating alkaline phosphatases [35]. In addition to the above, these patients face risk factors such as dental caries, enamel hypoplasia and structural abnormalities [36]. Another situation that may be present is a diminution of the oral vestibule leading to altered occlusion and articulation along with difficulty in brushing and flossing [39]. The desire to improve facial esthetics is one of the reasons why dental treatment is sought by an orthodontist and a maxillofacial surgeon, depending on the case [37].

When oral regions are affected, a maxillofacial surgeon is involved in multidisciplinary care for the correction of maxillary collapse, in addition to other specialists such as orthodontists for dental alignment and endodontists in cases where there is root resorption.

4. Conclusions

The oblique facial cleft is a rare manifestation with a percentage of 0.24 of all facial clefts, its etiology is a failure in the closure of the lateral and maxillary processes, it is characterized by a cleft that goes from the cupid's bow to the lower orbit with possible involvement of the palate. It is diagnosed using imaging techniques such as three-dimensional ultrasound and prenatal magnetic resonance imaging. Its treatment is complicated and involves a multidisciplinary team for the multiple surgeries it requires. As it is a manifestation that involves the oral region, a dental team is required to complete its treatment.

5. References

1. Balaji SM. Single stage correction of bilateral tessier 4 cleft. Indian J Dent Res. 2017;28(1):105-108.
2. Binet A, de Buyss Roessingh A, Hamedani M, El Ezzi O. Complete bilateral Tessier’s facial cleft number 5: surgical strategy for a rare case report. Surg Radiol Anat. 2019;41(5):569-574.
3. Corona-Rivera JR, Zapata-Aldana E, Bobadilla-Morales L, Corona-Rivera A, Peña-Padilla C, Solis-Hernández E, et al. Oblique facial clefts in Johanson-Blizzard syndrome. Am J Med Genet A. 2016;170(6):1495-501.
4. Eppley BL, van Aalst JA, Robey A, Havlik RJ, Sadove AM. The spectrum of orofacial clefting. Plast Reconstr Surg. 2005;115:101e-114e.
5. Jin T, Liu J, Gui L, Niu F, Yu B. Computer-assisted design of sequential surgical procedure for oblique facial clefts with mandibular outer cortex autografts. J Craniofac Surg. 2015;26(2):373-7.
6. Morgan AL, Cason R, El Amm CA. Anthropometrically-Based Surgical Technique for Tessier 3 Cleft Reconstruction. J Craniofac Surg. 2016;27(8):e785-e787.
7. Versnel SL, van den Elzen ME, Wolvius EB, Biesmeijer CS, Vaandrager JM, van der Meulen JC, et al. Long-term results after 40 years experience with treatment of rare facial clefts: Part 1–Oblique and paramedian clefts. J Plast Reconstr Aesthet Surg. 2011;64(10):1334-43.
8. Nagashima H, Sakamoto Y, Ogata H, Miyamoto J, Yazuwa M, Kishi K. Evaluation of bone volume after secondary bone grafting in unilateral alveolar cleft using computer-aided engineering. Cleft Palate Craniofac J. 2014;51(6):665-8.
9. Bubanale SC, Kurbet SB, De Piedade Sequeira LMG. A rare case of cleft number nine associated with atypical cleft number two. Indian J Ophthalmol. 2017;65(7):610-612.
10. Shea BJ, Reeves BC, Wells G, Thuku M, Hamel C, Moran J, et al. AMSTAR 2: a critical appraisal tool for systematic reviews that include randomised or non-randomised studies of healthcare interventions, or both. BMJ. 2017;358:j4008.
11. Prasad V, Ahmed R, Singh AK, Kumar V. A rare case of bilateral oblique facial cleft and accessory maxilla with repaired unilateral cleft lip and palate. Nat J Maxillofac Surg. 2019;10(2):241-244.
12. Canoglu E, Turgut MD, Tekcicek M. Healing of External Inflammatory Root Resorptions and Periapical Lesions without Surgical Treatment in an Operated Oblique Facial Cleft Case. Eur J Dent. 2010;4(2):208-14.
13. Balaji SM. Two-stage Corrections of Rare Facial Tessier’s Cleft - 3, 4, 5, 6, 7. Ann Maxillofac Surg. 2017;7(2):287-290.
14. Abdollahafekhshim S, Shahidi N, Bayazian G. A bilateral Tessier number 4 and 5 facial cleft and surgical strategy: a case report. Iran J Otorhinolaryngol. 2013;25(73):259-62.
15. Oh JH, Park YW. Anatomical repair of a bilateral Tessier No. 3 cleft by midfacial advancement. Maxillofac Plast Surg. 2018;40(1):9.
16. Carstens MH. Anatomía patológica del paladar blando, parte 1: Embriología, plataforma de tejido duro y evolución. J Labio leporino Craniofac Anomalía. 2017;4:37-64.
17. Bello SA, Ibikari AB, Oketade I, Balogun SA. Atypical Facial Clefts From Northcentral Nigeria, Review of 36 Cases. Cleft Palate Craniofac J. 2019;56(4):514-520.
18. Gfrerer L, Shubinets V, Hoyos T, Kong Y, Nguyen C, Pietschmann P, Morton CC, Maas RL, Liao EC. Functional analysis of SPECC1L in craniofacial development and oblique facial cleft pathogenesis. Plast Reconstr Surg. 2014;134(4):748-59.
19. Bhoj EJ, Haye D, Toutain A, Bonneau D, Nielsen IK, Lund IB, et al. Phenotypic spectrum associated with SPECC1L, pathogenic variants: new families and critical review of the nosology of Teebi, Opitz GBBB, and Baraitser-Winter syndromes. Eur J Med Genet. 2019;62(12):103588.
20. Ascha M, Harvey D, Becker D, Rowe D, Sarma H, Lakin GE. A Novel Single-Stage Technique for the Treatment of Soft Tissue in an Oblique Facial Cleft. J Craniofac Surg. 2016;27(6):1517-20.
21. Tessier P. Anatomical classification facial, cranio-facial and latero-facial clefts. J Maxillofac Surg. 1976;4(2):69-92.
22. Rifkin WJ, Kantar RS, Ali-Khan S, Plana NM, Diaz-Siso
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JR, Tsakiris M, et al. Facial Disfigurement and Identity: A Review of the Literature and Implications for Facial Transplantation. AMA J Ethics. 2018;20(4):309-323.

Wilhelm L, Borgers H. The “equals sign”: a novel marker in the diagnosis of fetal isolated cleft palate. Ultrasound Obstet Gynecol. 2010;36:439-444.

Jones MC. Prenatal diagnosis of cleft lip and palate: detection rates, accuracy of ultrasonography, associated anomalies, and strategies for counseling. Cleft Palate Craniofac J. 2002;39(2):169-73.

Alpagan Ozdemir S, Esenlik E. Three-Dimensional Soft-Tissue Evaluation in Patients with Cleft Lip and Palate. Med Sci Monit. 2018;24:8608-8620.

Marginean C, Sasanean V, Marginean CO, Melit LE, Marginean MO. Prenatal diagnosis of cleft lip and cleft lip palate - a case series. Med Ultrason. 2018;20(4):531-535.

Evans MI, Hume RF Jr, Johnson MP, Treadwell MC, Krivchenia EL, Zador IE, et al. Integration of genetics and ultrasonography in prenatal diagnosis: just looking is not enough. Am J Obstet Gynecol. 1996;174(6):1925-1931.

Sreejith VP, Arun V, Devarajan AP, Gopinath A, Sunil M. Psychological Effect of Prenatal Diagnosis of Cleft Lip and Palate: A Systematic Review. Contemp Clin Dent. 2018;9(2):304-308.

Lewis CW, Jacob LS, Lehmann CU; Section on Oral Health. The Primary Care Pediatrician and the Care of Children with Cleft Lip and/or Cleft Palate. Pediatrics. 2017;139(5):e20170628.

Luo X, Huang H, Yin X, Shi B, Li J. Functional stability analyses of maxillofacial skeleton bearing cleft deformities. Sci Rep. 2019;9(1):4261.

Chen ZQ, Wu J, Chen RJ. Sagittal maxillary growth pattern in unilateral cleft lip and palate patients with unrepaired cleft palate. J Craniofac Surg. 2012;23(2):491-3.

Kahnberg KE, Vannas-Löfqvist L, Zellin G. Complications associated with segmentation of the maxilla: a retrospective radiographic follow up of 82 patients. Int J Oral Maxillofac Surg. 2005;34(8):840-5.

Sáez MDM, López GL, Atlas D, de la Casa ML. Evaluation of pH and calcium ion diffusion from calcium hydroxide pastes and MTA. Acta Odontol Latinoam. 2017;30(1):26-32.

Zschiesschack B, Grabowski R. The influence of caries of the deciduous teeth upon development of the dentition in patients with cleft lip, jaw and palate. J Orofac Orthop. 1999;60(3):215-24.

Alhayek S, Alsalem M, Alotaibi Y, Omair A. Evaluation of facial appearance in patients with repaired cleft lip and palate: comparing the assessment of laypeople and healthcare professionals. Maxillofac Plast Reconstr Surg. 2019;41(1):5.

Harkins CS, Berlin A, Harding RL, Longacre JJ, Snodgrass RM. A classification of cleft lip and cleft palate. Plast. Reconstr. Surg. 1962;76(2):212-224.

van der Meulen JC, Maddalena P, Mazzola R, Vermey-Keers C, Stricker M, Raphael B. A morphogenetic classification of craniofacial malformations. Plast Reconstr Surg. 1983;71(4):560-72.

van der Meulen JCH. Oblique Facial Clefts. Plastic and Reconstructive Surgery. 1985;76(2):212-224.

van der Meulen JC, Mazzola R, Vermey-Keers C, Stricker M, Raphael B. A morphogenetic classification of craniofacial malformations. Plast Reconstr Surg. 1983;71(4):560-72.

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