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Case Report

Cayler Syndrome with Isolated Cleft Palate: Case Report

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ABSTRACT
The present study was focused on assessing the clinical Isolated cleft palate is the term used when a cleft occurs only within the palate. In clinical terms it is differentiated from other clefts, affecting the primary palate because it as such doesn't affect either the alveolar ridge or the perioral musculature. This makes the diagnosis of the cleft, by facial analysis, more challenging, since it is concealed in the palate. Cayler syndrome is an abnormal condition at birth represented by hypoplasia of the depressor angular oris muscle (HDAOM) in an addition to cardiac birth defects. A considerable range of multiple organ system anomalies are also to predicted to be present. Paediatrician must make awareness regarding this minor facial anomaly as it can be related to multiple, cardiac and internal organs abnormalities. I present to you a paediatric rare case of cleft palate which appears to be isolated, along with cayler syndrome. To my knowledge no previous report on situs inverse totalis was reported. Additionally, the patient does not have any major cardiac anomalies associated with the cayler complications.

Keywords: Isolated cleft palate, Cayler syndrome, Palatoplasty, Palatogenesis, Clefting.

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INTRODUCTION

The hallmark sign of the Cayler cardio facial syndrome is marked by the hypoplasia of the depressor angular oris muscle and cardiac birth defects. it may be sporadic or related to 22q11.2 micro deletion. Hypoplasia of the depressor angular oris muscle results in failure of one edge of the mouth to move in a downward and outward manner while crying or while frowning and hence, is described as an "asymmetric crying face.".[1] A range of congenital cardiac defects is described. On an occasional basis, other organ system abnormalities could also be additionally present.[2] DAOM hypoplasia at birth is kind of frequent; the estimated frequency is 6.3% per 1,000 neonates in India.[3] The differential diagnoses to rule out are, Goldenhar syndrome, Moebius syndrome, Velocardiofacial, digeorge syndrome, Opitz GBBB syndrome, etc.[1]

Isolated cleft palate is the term used when a cleft occurs only within the palate.[4] The cleft can involve either the soft or hard palate and will extend as far as the nasopalatine foramen but there›s no accompanying cleft lip. Approximately half of the affected individuals with cleft palate appear to be either a part of a genetic syndrome or any other malformation and the remaining 50% occur as solo defects, which are usually cited as non-syndromic clefts.CPO is one of the rarest types of oral clefting.[1]

Anatomy and Biology of Palatogenesis

The human palate comprises of a palatum durum (hard palate) and palatum velem (soft palate). The hard palate is further separated into primary and secondary portions. The hard palate is located anterior to the anterior palatine foramen, and the soft palate lies posterior dividing the nasal passage from the pharynx. The palatal shelves emerge around week 6 of the gestation period in humans. It consists of cranial neural crest-cells which are mesenchymal in origin and mesoderm-derived which are endothelial in nature, which together is concealed by derived epithelial cell-pharyngeal ectoderm. Typical evolution of the palate depends on various aspects like; migration should be proper, growth, differentiation, and cell apoptosis. The growth occurs in three stages: vertical expansion of the tongue's shelves toward the sides, followed by, as the mandible extends, the palatal shelves are raised to a horizontal posture, and the palatal shelves are fused together, this results in the creation of a temporary epithelial seam in the midline which eventually transitions from epithelial to mesenchymal. A failure in any of the above phases can result in a cleft palate.[1]

Classification of cleft palate

Clefts of the palate are generally divided into three categories: unilateral, bilateral, both of which can be complete or incomplete, and the last one being submucous (Figure 1). Veau divided clefts into four categories in1931: (1) soft palate cleft only; (2) both hard and soft palate cleft (3) unilateral cleft lip and palate and (4) bilateral cleft lip and palate. However, a major drawback of this classification was that, it doesn't address principal palate clefts or differentiate between incomplete, complete lip and palate clefts.

CASE REPORT

A year and a month-old female infant were born to a non-consanguineous couple at our hospital, with a current weight of 9.1 kg, the mother has had a typical vaginal delivery and the mother was healthy during the pregnancy. The mother was breastfeeding him and no difficulties were reported and all vaccination for immunizations was given. the patient was admitted to NICU for 3 days for depression of angular oris muscle hypoplasia along with cleft of both hard and soft palate, was advised no active intervention and was advised surgery at 11 months of age. The patient was a follow-up case for cleft palate with complaints of a hole in the palate region. After performing extra oral and intraoral examination patient was diagnosed with cleft of both hard and soft palate which was isolated, associated with cayler syndrome. On Physical examination, a 2 × 3 cm defect involving both palates along with the angle of mouth deviation to the left side, the facial features were normal when the infant was sleeping. However, asymmetry was noted in the form of drooping of the right angle of the lips when she cries

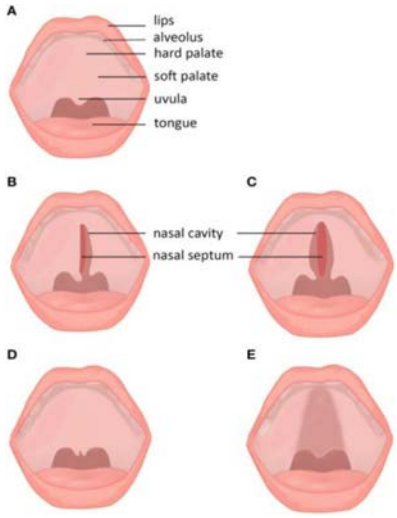

Figure 1: Subtypes and subclinical forms of cleft palate.
(A) Normal lip and palate. (B) Unilateral cleft palate. (C) Bilateral cleft palate. (D) Cleft uvula. (E) Submucous cleft palate.

and deviation of head eye, and ear to the left side. CNS Examination revealed abnormalities in the 5th, 6th, and 7th nerves. A 2-D echo study was performed which showed intact IAS and IVS, No PDA (patent ductus arteriosus)/COA (Coarctation of Aorta)/ TAPVC/ No PAH (pulmonary arterial hypertension), and effusions was noted. palatoplasty was performed for the cleft defect. Post-op analysis revealed intraoral sutures over both of the palates along with facial symmetry. The patient was discharged with oral antibiotics and multivitamins and is on follow-up.

DISCUSSION

The rarest form of oral clefting was found to be, isolated cleft palate. it varies by geography from 1.3 to 25.3 per 10,000live births with the highest rates in British Columbia, Canada and the lowest in Nigeria an Africa. Besides genetic and environmental risk factors, the etiology of isolated cleft palate is multifactorial. As for the genetic risk factors, maternal tobacco smoke has been documented has the highest and strongest risk factor. As for the treatment options, surgery remains as the most viable option. The sole goal of the surgery is the closure of the defect, but more emphasis has to be given on quality of speech.[1] Congenital hypoplasia of DAOM presents with drooping at the corner side of the mouth also known as " asymmetric crying face". Parents are to be counselled for the future complications that is, learning difficulties, and mental retardation.[3]

CONCLUSION

Facial study found that the facial pattern was unaffected, that palatoplasty of the hard and soft palates was successful, and that facial symmetry was addressed.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

ABBREVIATIONS

HDAOM: Hypoplasia of depressor angular oris muscle; **CPO:** Isolated cleft palate; **IAS:** interarterial septum; **IVS:** Interventricular septum; **Pda:** Patent ductus arteriosus; **COA:** Coarctation of aorta; **TAPVC:** Total anomalous pulmonary venous connection; **PAH:** pulmonary arterial hypertension; **PICU:** Paediatric ICU; **NICU:** Neonatal ICU.

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