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Tessier 3 and 4 Clefts and Choanal Atresia: An Unusual Association?

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Abstract

Introduction: Craniofacial clefts are rare congenital anomalies that might involve both soft tissue and skeletal components. The association of Tessier cleft number 3 and 4 with choanal atresia appears to be unusual and only few clinical cases have been reported in published literature.

Objectives: Report a series of 13 cases of choanal atresia in patients with Tessier numbers 3 or 4 clefts and the literature review on this topic.

Methods: A literature review was undertaken via PUBMED database before April 2020 addressing the association between Tessier numbers 3 or 4 clefts and choanal atresia. Retrospective chart review of patients diagnosed with both comorbidities at a tertiary hospital expertised in craniofacial anomalies.

Results: Literature review yielded 10 studies describing the relationship between choanal atresia and Tessier 3 and 4 facial clefts. We identified 98 patients diagnosed with medial oro-ocular facial clefts (Tessier 3 and 4) and 119 with choanal atresia at our institution over a 20 years time period. Altogether, 13 individuals were diagnosed with both malformations, 3 patients with number 3 cleft, and 10 patients with number 4 cleft. It represents 13.26% of the cases.

Conclusion: This study highlights the features of Tessier 3 and 4 facial clefts associated with choanal atresia. Although the publications regarding this association are very scarce, the authors present the largest series of cases of Tessier number 3 and 4 clefts with choanal atresia showing that association between these conditions could be not so unusual.

Keywords

Choanal atresia, nonsydromic clefting, nose

Introduction

Craniofacial clefts, classified in detail by Paul Tessier, are rare congenital deformities that compromise beyond the soft tissue, the bony facial skeleton. Choanal atresia, on the other hand, is a rare condition, although it is considered the most frequent congenital nasal anomaly. Like rare clefts, choanal atresia is characterized by the bone involvement in most of the cases.

Tessier number 3 and 4 clefts are considered oblique forms of rare facial clefts. Also known as a naso-ocular or nasomaxillary cleft, Tessier number 3 cleft starts from the inferior eyelid medial to the inferior lacrimal punctum, extends over the lacrimal groove, descends to the area of the lateral wall of the nose, passes around the nasal ala in the nasolabial groove, and ends at the philtrum of the superior lip, like a standard cleft lip (Sesenna et al., 2012). Tessier number 4 cleft, also known as an oro-ocular cleft, spares the nose and extends toward a more centric orbital position. At a point between the philtrum and commissure, the cleft extends superiorly through the nasolabial fold and passes into the orbit just medial to the infraorbital foramen. The main difference on the skeleton is the preservation of septation between the nasal cavity and maxillary sinus (Tessier, 1976).

Choanal atresia may be another anatomical and functional finding present in the rare facial clefts. Anatomical closure of the distal portion of the nasal fossa results from the

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involvement of the following choanal boundaries: the undersurface of the body of the sphenoid bones superiorly, the medial pterygoid lamina laterally, the vomer medially, and the horizontal portion of the palatal bone inferiorly (Ramsden et al., 2009).

The description of the presence of choanal atresia in rare facial clefts is scarce in the literature and the association between Tessier numbers 3 and 4 clefts with choanal atresia is not clear. The objective was to review the literature and present herein the largest series of cases that show this unusual association, reporting 13 cases of choanal atresia in patients with Tessier numbers 3 and 4.

Methods

An online search of PubMed database from inception to April 2020 was performed to search related articles using the keywords "choanal atresia", "cleft", "rare cleft", "facial cleft", and "Tessier cleft", limited to the English language. Two of the authors independently reviewed the titles and abstracts and all outcomes were considered if meeting the following inclusion criteria: Tessier 3 and 4 facial clefts associated with choanal atresia. A retrospective analysis of all patient records diagnosed with Tessier numbers 3 or 4 clefts and with choanal atresia in our department within the last 20 years was also performed (Figure 1). Regarding the ethical criteria, this study was approved by Human Research Ethics Committee of the Hospital for Rehabilitation of Craniofacial Anomalies (HRAC/USP) and informed consents were assigned by parents.

Results

Pubmed yielded 126 publications relevant by examination of titles, abstracts, and full papers, according to inclusion criteria defined a priori. The articles that preceded or did not use the scheme proposed by Tessier in 1976 were reclassified and adapted according to photos and descriptive data of each study (Boo-Chai, 1970, 1990; Dey, 1973). Two case reports were excluded: one reclassified as a paramedian cleft (Wiltshire et al., 2003) and another described by the author as choanal stenosis (Patel et al., 2015). There were only 8 reported cases of choanal atresia associated with Tessier numbers 3 or 4 clefts (Dey, 1973; Tessier, 1976; Thatte et al., 1987; Ragavan et al., 2012; Allam et al., 2014; Ueda et al., 2015; Sung et al., 2020).

The number of patients enrolled in our institution with oblique facial cleft was 98, 76.53% are unilateral forms ($N = 38$ right and $N = 37$ left) and only 23.47% ($N = 23$) are bilateral. In the same period, there were 119 patients with choanal atresia. All individuals were previously evaluated by the Craniofacial and Genetics teams from Hospital for Rehabilitation of Craniofacial Anomalies-University of São Paulo and 13 cases were identified with both entities (Table 1).

Discussion

Facial clefts are rare congenital deformities and the incidence is estimated at 1.4-4.9 per 100 000 live births and 9.5-34 per 1000 common clefts (Fogh-Anderson, 1965; Kawamoto, 1976; Resnick and Kawamoto, 1990). Consequently, Tessier numbers 3 and 4 form a very rare group and due to incomplete case descriptions in the literature, the actual incidence of these clefts as a group and individually is not known (Eppley et al., 2005). Choanal atresia, although it is the most common congenital nasal anomaly, is also a rare condition with an estimated incidence of 1:5000-7000 live births (Kwong, 2015).

The embryologic origin of these 2 malformations, as well as their embryologic relation, is still being discussed. The migration anomalies of the neural crest cells were thought to be a common explanation for these associated craniofacial anomalies (Hengerer and Strome, 1982; Bonafos et al., 2004; Allam et al., 2014).

Boo-Chai in 1970 performed an extensive review of publications dealing with rare craniofacial clefts and identified 12 patients with naso-ocular cleft and 23 with oro-ocular cleft type I, equivalent to Tessier numbers 3 and 4 clefts, respectively (Boo-Chai, 1990). Alonso et al. (2008) presented a large series of 21 patients with Tessier number 4 cleft. Versnel et al. (2011) reported 22 oblique facial clefts (Tessier 3, 4, and 5). Recently, 2 of the largest series of patients with Tessier number 3 cleft were reported by Silva et al. in 2010 and Allam et al. in 2014 with, respectively, 21 and 10 cases. Interesting is the fact that among all these studies, only 1 choanal atresia was cited, by Allam. However, there was no mention of specific details about the nasal cavity of most patients. Curiously, in the definition of number 4 cleft by Paul Tessier, in his remarkable classification in the year 1976, he illustrated with a case report of bilateral cleft and left choanal atresia (Figure 2).

Although this association seems unusual, we observed a relevant incidence in our institution: 3 patients with number 3 cleft and 10 patients with number 4 cleft. It represents a significant portion of 13.26% of our cases. Among the 7 patients with unilateral facial clefts, only 1 had choanal atresia on the same side affected by the cleft and 3 of them had bilateral choanal atresia. Thus, apparently, there is no relationship between the side of the facial cleft and the side affected by choanal atresia in our sample.

To date, no study has been found to indicate a possible relationship between choanal atresia and oblique facial clefts 3 and 4. The following 2 studies are the only ones that describe several cases of the rare craniofacial clefts associated with choanal atresia. Garabedian et al. (1999) ($N = 5$), based on anatomical and pathogenic embryological considerations, proposed a new classification concerning the Tessier numbers 12-2 clefts, including hypogenesis of the nasal fossa and choanal imperforation in the clinical presentation of these malformations. Bonafos et al. in 2004 ($N = 3$), emphasizes the need to search for evidence of choanal atresia as soon as the diagnosis of rare craniofacial cleft is made, particularly when

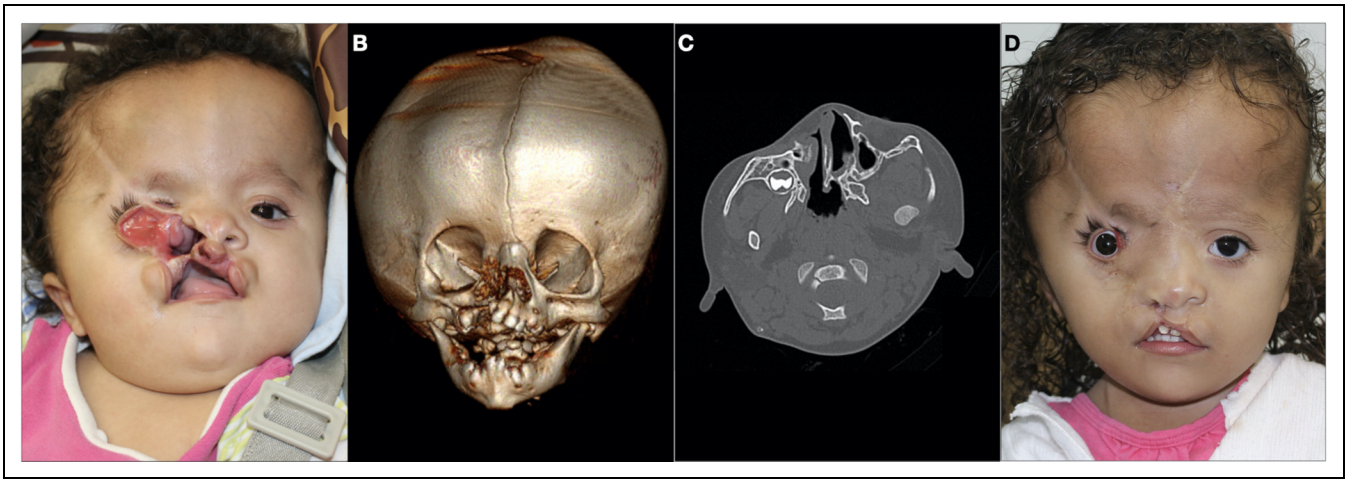

Figure 1. (A) Right Tessier number 3 cleft in a child at 11 months of age, with amniotic band sequence, right anophthalmia, complete right cleft lip, and incomplete left cleft lip; (B) 3-dimensional reconstruction computed tomography (CT) scan shows the oral–nasal–ocular cleft; (C) axial preoperative CT scan illustrates the left choanal atresia. (D) Two years follow-up surgical cleft correction.

Table 1. Eight Reported Cases of Choanal Atresia Associated with Tessier Numbers 3 or 4 Clefts, Published in the English Language and Data From 13 Patients of the Author's Hospital with Choanal Atresia and Tessier Numbers 3 or 4 Clefts.

		Sex	Choanal atresia	Tessier number	Ocular involvement	Amniotic band sequence
Dey (1973)	1	F	Bilateral	4 bilateral	–	–
	2	M	Bilateral	3 left and 4 right	–	–
Tessier (1976)	3	M	Left	4 bilateral	–	–
Thatte et al. (1987)	4	F	Right	3 right	–	–
Ragavan et al. (2012)	5	M	Bilateral	4[a] right	–	–
Allam et al. (2014)	6	M	Left	3 and 7 left	Bilateral microphthalmia	–
Ueda et al. (2015)	7	F	Right	3-11, 5 left and 2-12 right	Right microphthalmia	Present
Sung et al. (2020)	8	F	Bilateral	3 bilateral	–	–
Present study	9	M	Right	3 left	Left anophthalmia	–
	10	F	Left	3 right	Right anophthalmia	Present
	11	F	Right	4 bilateral	–	–
	12	F	Right	4 bilateral	–	–
	13	F	Bilateral	4 right	–	–
	14	M	Bilateral	4 left	–	–
	15	M	Bilateral	4 right	Right microphthalmia	–
	16	F	Left	4 right	–	–
	17	F	Bilateral	4 bilateral	–	–
	18	M	Bilateral	4 bilateral	Right microphthalmia	–
	19	M	Bilateral	3-11 right and 3 left	Left microphthalmia	Present
	20	F	Bilateral	4 bilateral	–	–
	21	F	Left	4 left	–	–

[a] Was reclassified based on the image provided by the authors.
F, female; M, male.

it corresponds to numbers 12-2 or 13-1, paramedian forms. Despite little data in the literature, we routinely carry out an assessment in all cases of rare craniofacial clefts for the detection of choanal atresia through clinical examination, upper airway endoscopy (UAE), and computed tomography (CT).

Craniofacial clefts have long been reported in association with limb and body wall constriction bands, with an incidence between 8.4% and 63% of cases. In 1998, Coady et al. confirmed an association between rare craniofacial cleft and limb ring constrictions, concluding that the 2 conditions may therefore possess common etiology. In their study, the coexistence of these anomalies was largely restricted to the 2-12, 3-11, and 4-10 cleft types (Coady et al., 1998). Even though amniotic band sequence may be implicated in the facial cleft process,

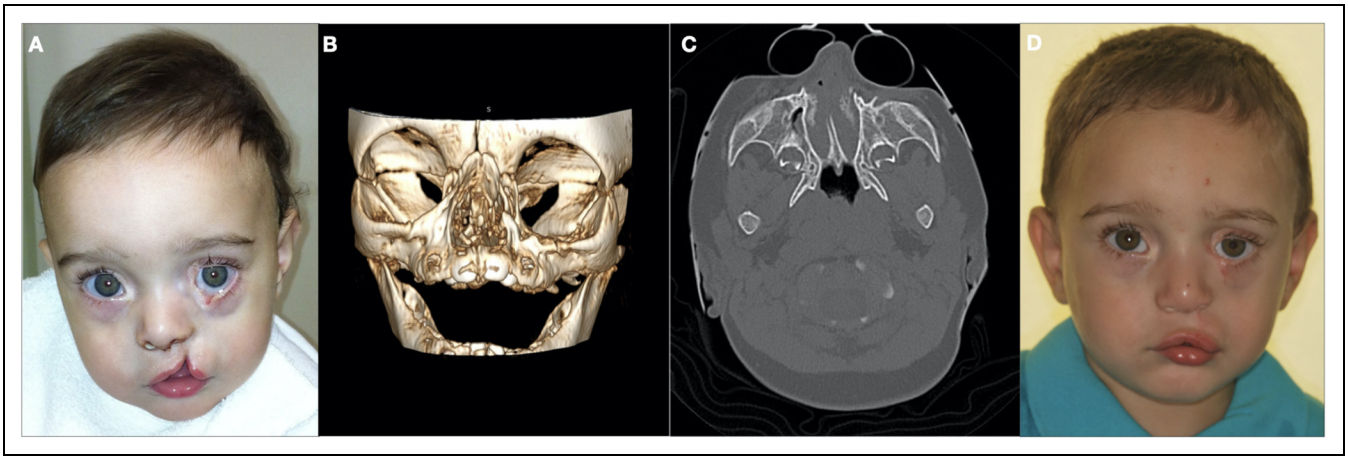

Figure 2. (A) Left Tessier number 4 cleft affecting the upper lip and coloboma of the lower eyelid; (B) preoperative 3-dimensional reconstructed computed tomography (CT) showing the bone gap in the left inferior orbital margin, medial to the infraorbital foramen; (C) axial image preoperative CT scan illustrates the bilateral choanal atresia; (D) 1-year follow-up surgical cleft correction.

we did not find a common explanation for all 3 the concomitant anomalies present in 2 reported patients: choanal atresia, Tessier cleft, and constriction rings.

Approximately 50% of patients with choanal atresia have other associated congenital abnormalities. Considering the bilateral forms, it is even more frequently associated, about 75% of patients (Altuntas et al., 2004). CHARGE syndrome is the most commonly reported disorder related to choanal atresia in the literature. Trisomy 18, Trisomy 21 and Treacher Collins, Pfeiffer, Antley-Bixler, Apert, Crouzon, di-George, Marshall–Smith, Schinzel–Giedion, Kabuki syndromes have also been reported in patients with choanal atresia (Burrow et al., 2009).

Regarding composition, Brown et al. (1996) in a retrospective study reviewing the CT and histologic specimens in 63 patients showed a 29% pure bone atresia, 71% mixed membranous and bone atresia with no pure membranous atresia present. In our study, based on CT, UAE, and intraoperative aspect, all 4 operated cases of choanal atresia associated with Tessier number 3 or 4 clefts were of the mixed bony-membranous type. In the 4 patients not yet operated, they were documented tomographically as mixed type. In 5 cases, it was not possible to adequately define the type of choanal atresia.

The authors are aware of the study limitations since it is a retrospective sample and data regarding clinical evolution and treatment methods were not its aim. Although the publications regarding this association are very scarce, we present the largest series of cases of Tessier number 3 and 4 clefts with choanal atresia. It is important that craniofacial team are conscious of this condition, since early detection and intervention may help to prevent harmful complications.

Conclusion

This study highlights the features of rare facial cleft associated with choanal atresia. The considerable high prevalence observed in our study suggests that association between these clinical conditions could be more frequent that observed in the literature. More research considering rare facial clefts are needed to determine the association.

Declaration of Conflicting Interests

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References

Allam KA, Lim AA, Elsherbiny A, Kawamoto HK. The Tessier number 3 cleft: a report of 10 cases and review of literature. *J Plast Reconstr Aesthet Surg*. 2014;67(8):10551062.

Alonso N, Freitas Rda S, de Oliveira e Cruz GA, Goldenberg D, Dall'oglio Tolazzi AR. Tessier no. 4 facial cleft: evolution of surgical treatment in a large series of patients. *Plast Reconstr Surg*. 2008;122(5):15051513.

Altuntas A, Yilmaz MD, Kahveci OK, Dereköy S, Yücel A. Coexistence of choanal atresia and Tessier's facial cleft number 2. *Int J Pediatr Otorhinolaryngol*. 2004;68(8):10811085.

Bonafos G, Capon-Degardin N, Fayoux P, Pellerin P. Choanal atresia and rare craniofacial clefts: report of three cases with a review of the literature. *Cleft Palate Craniofac J*. 2004;41(1):7883.

Boo-Chai K. The oblique facial cleft. A report of 2 cases and a review of 41 cases. *Br J Plast Surg*. 1970;23(4):352359.

Boo-Chai K. The oblique facial cleft: a 20-year follow-up. *Br J Plast Surg*. 1990;43(3):355358.

Brown OE, Pownell P, Manning SC. Choanal atresia: a new anatomic classification and clinical management applications. *Laryngoscope*. 1996;106(1Pt):97-101.

Burrow TA, Saal HM, de Alarcon A, Martin LJ, Cotton RT, Hopkin RJ. Characterization of congenital anomalies in individuals with choanal atresia. *Arch Otolaryngol Head Neck Surg*. 2009;135(6):543547.

Coady MS, Moore MH, Wallis K. Amniotic band syndrome: the association between rare facial clefts and limb ring constrictions. *Plast Reconstr Surg*. 1998;101(3):640649.

da Silva Freitas R, Alonso N, Busato L, Ueda WK, Hota T, Medeiros SH, Kunz RT. Oral-nasal-ocular cleft: the greatest challenge among the rare clefts. *J Craniofac Surg*. 2010;21(2):390395.

Dey DL. Oblique facial clefts. *Plast Reconstr Surg*. 1973;52(3):258263.

Eppley BL, van Aalst JA, Robey A, Havlik RJ, Sadove AM. The spectrum of orofacial clefting. *Plast Reconstr Surg*. 2005;115(7):101e114e.

Fogh-Anderson P. Rare clefts of the face. *Acta Chil Scand*. 1965;129:275-281.

Garabedian EN, Ducroz V, Roger G, Denoyelle F, Catala M. Nasal fossa malformations and paramedian facial cleft: new perspectives. *J Craniofac Genet Dev Biol*. 1999;19(1):1219.

Hengerer AS, Strome M. Choanal atresia: a new embryologic theory and its influence on surgical management. *Laryngoscope*. 1982;92(8 Pt 1):913921.

Kawamoto HK Jr. The kaleidoscopic world of rare craniofacial clefts: order out of chaos (Tessier classification). *Clin Plast Surg*. 1976;3(4):529572.

Kwong KM. Current updates on choanal atresia. *Front Pediatr*. 2015;9(3):52.

Patel SD, Porras S, Lypka M. Journey to chew: a case of maxillary duplication and bony syngnathia. *J Craniomaxillofac Surg*. 2015;43(1):5761.

Ragavan M, ArunKumar S, Balaji NS. Bilateral choanal atresia with tessier type 3 facial cleft: a rare association. *J Neonatal Surg*. 2012;1(3):48.

Ramsden JD, Campisi P, Forte V. Choanal atresia and choanal stenosis. *Otolaryngol Clin North Am*. 2009;42(2):339352.

Resnick JI, Kawamoto HK Jr. Rare craniofacial clefts: Tessier no. 4 clefts. *Plast Reconstr Surg*. 1990;85(6):843852.

Sesenna E, Anghinoni ML, Modugno AC, Magri AS. Tessier 3 cleft with bilateral anophthalmia: case report and surgical treatment. *J Craniomaxillofac Surg*. 2012;40(8):690693.

Sung JY, Cho KS, Bae YC, Bae SH. Image-guided navigation surgery for bilateral choanal atresia with a Tessier number 3 facial cleft in an adult. *Arch Craniofac Surg*. 2020;21(1):6468.

Tessier P. Anatomical classification of facial, cranio-facial and latero-facial clefts. *J Maxillofacial Surg*. 1976;4(2):69-92.

Thatte RL, Sood MK, Uppal PK. A combination of unusual abnormalities of the lachrymal apparatus and nostrils in a case of bilateral cleft with choanal atresia. *Br J Plast Surg*. 1987;40(6):645646.

Ueda K, Shigemura Y, Nuri T, Iwanaga H, Seno T. A case of complex facial clefts treated with staged-tissue expansion. *Plast Reconstr Surg Glob Open*. 2015;2(12):e264.

Versnel SL, van den Elzen ME, Wolvius EB, van Veelen ML, Vaandrager JM, van der Meulen JC, Mathijssen IM. 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):13341343.

Wiltshire E, Moore M, Casey T, Smith G, Smith S, Thompson E. Fryns "Anophthalmia-Plus" syndrome associated with developmental regression. *Clin Dysmorphol*. 2003;12(1):4143.