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ORIGINAL ARTICLE

Complete bilateral Tessier's facial cleft number 5: surgical strategy for a rare case report

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Abstract
The oro-ocular cleft number 5 according to the Tessier classification is one of the rarest facial clefts and few cases have been reported in the literature. Although the detailed structure of rare craniofacial clefts is well established, the cause of these pathological conditions is not. There are no existing guidelines for the management of this particular kind of cleft. We describe the case of a 19-month-old girl with a complete bilateral facial cleft. We describe the surgical steps taken to achieve the primary correction of the soft tissue deformation. Embryologic development and radiological approach are discussed, as are also the psychological and social aspects of severe facial deformities.

Keywords Orofacial cleft 5 · Cleft palate · Oblique facial cleft · Craniofacial abnormalities

Introduction

Oblique facial clefts (meloschisis) are the most uncommon of facial clefts [5]. Craniofacial clefts are atypical congenital malformations occurring in less than 5 per 100,000 live births [10, 13, 31, 35]. First reported by Von Kulmus in 1732, the craniofacial cleft was well-described by Paul Tessier [2, 5]. He proposed in 1973 a new classification based on a numbering system which combines the findings of the clinical examination with the underlying facial bone deformity seen at the time of reconstructive repair [30]. It ranges from 0 to 14, according to constant lines through the eyebrows, eyelids, maxilla, nose, and lip.

He presented his classification of facial clefts centered around the orbital cavity as time zones [30]. The most commonly encountered oro-ocular clefts are number 3 and number 4, which begin either through, or slightly lateral to, the philtral column, and then pass through the medial portion of the lower eyelid, either without a bony septation between the nasal cavity and the maxillary sinus (the more difficult number 3), or with a bony septation (number 4) [24]. The number 5 cleft is much rarer, accounting for only 0.3% of atypical facial clefts [12].

Controversy still exists concerning treatment options and timing, but the initial priorities in cases of congenital facial deformities include airway patency, feeding, and protection of the cornea. Restoration of a normal appearance by closure of the soft tissues should be considered as soon as possible to facilitate social integration [31].

We present the case of a complete bilateral orofacial cleft (Tessier number 5), one of the rarest types. Our patient was referred to us by another hospital and was, therefore, already 2 years old when she was operated. We describe the primary surgical steps performed to restore an esthetic appearance and to promote language acquisition.

Case report

We present the history of a 19-month-old girl, born in Switzerland of a Swiss mother and a father originating from Togo. There were no apparent unusual prenatal, consanguinity, or hereditary factors. She was referred to our team at 19 months for a complete bilateral Tessier number 5 cleft. The cleft passed laterally to the cupid's bow (double-curved bow at the top edge of upper lip). and extended bilaterally through the cheeks into the

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inferio-lateral canthal region. The typically decreased oculo-alar and oculo-oral distance for this type of cleft was also present. The bony alveolar cleft was lateral to the incisors on both sides, and lateral to the infraorbital foramen. The lower eyelids on both sides were open in prolongation of the cleft and the left orbit was more severely dystopic, without marked corneal exposure and with eyelashes all along the cleft (Fig. 1). The child also presented multiple bone defects of the maxilla and an almost total absence of hard palate. The hard palate had an apparently intact mucous membrane with several depressions. The soft palate did not have a middle cleft as usual. It had a right lateral cleft separating laterally the palatoglossal and the pharyngoglossal muscles, without insertion abnormalities in the posterior palatal edge. The soft palate was short and largely distant from the pharyngeal wall.

The CT-Scan reconstruction revealed that the alveolar cleft ascended through to the anterior maxilla on both sides, with involvement of the orbital floor. The medial portion of the orbital rim was hypoplastic bilaterally (Fig. 2). The CT-scan showed no evidence of malformation of the brain structures, and the child appeared normally developed for her age.

The first step in the repair of this complex and rare facial cleft was the surgical repair of the soft tissue. This involved closure of the left and right oro-ocular cleft with Z-plasties to increase the skin length. Closure of the lid was achieved by means of cheek advancement flaps and multiple Z-plasties (Fig. 3). The medially retracted pedicled fatty flap is individualized and laterally advanced allowing a regular cheek surface and providing support under the scar.

Multiple cutaneous Z-plasties lengthened the deficient oro-ocular distance, and additional Z-plasties reconstructed the vestibular fold and vermillion contour repairing the large mouth appearance.

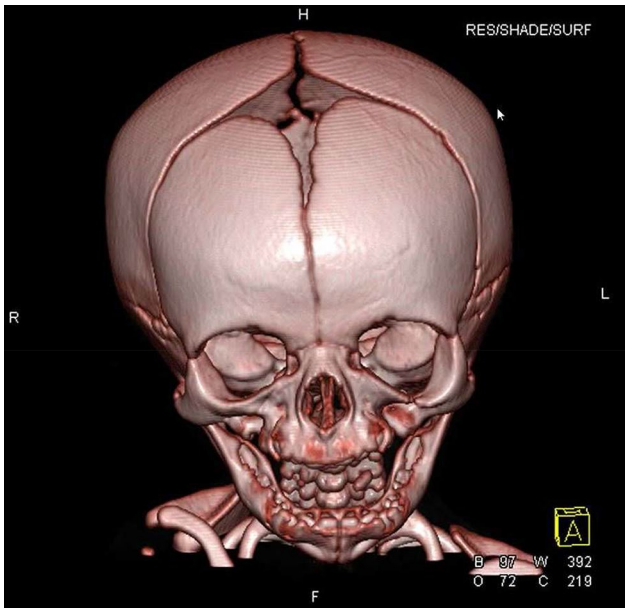

Fig. 2 3D CT-SCAN reconstruction showing the bilateral cleft sparing the orbital floors. The alveolar cleft ascended through to the anterior maxilla on both sides, with involvement of the orbital floor. The medial portion of the orbital rim was hypoplastic bilaterally

Six days after the surgery, the child fell from her bike causing a suture tear on the left side. Several surgical Interventions were necessary until complete healing was achieved.

Concerning the cleft palate, we performed a few months later a modified Von Langenbeck procedure to close the palate in one single operation, creating bipedicled flaps of mucoperiosteum, preserving the greater palatine neuro-vascular bundles, and dissecting and repositioning the palatal muscles to create a normalized muscular sling to promote speech acquisition (Fig. 4a–c). The first incision was made along the free margin of the cleft anterior to posterior to the

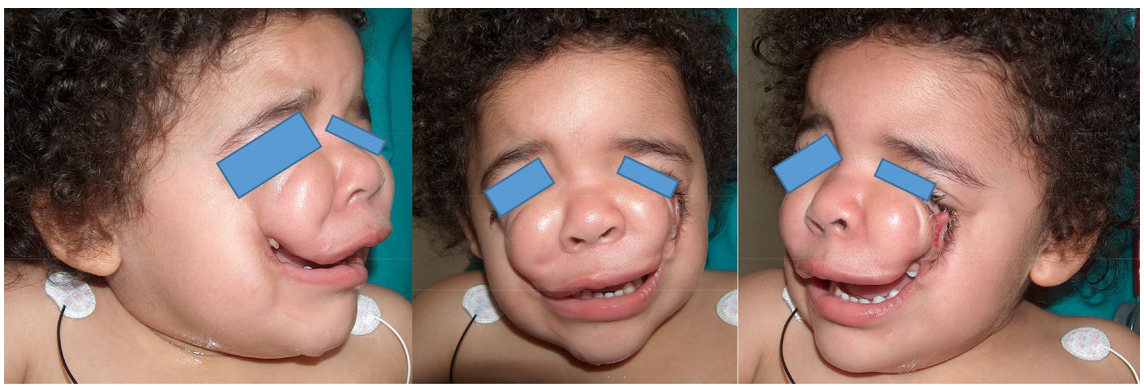

Fig. 1 Preoperative clinical view of the patient. A complete bilateral Tessier number 5 cleft. The lower eyelids on both sides were open in prolongation of the cleft and the left orbit was more severely dystopic, without marked corneal exposure and with eyelashes all along the cleft

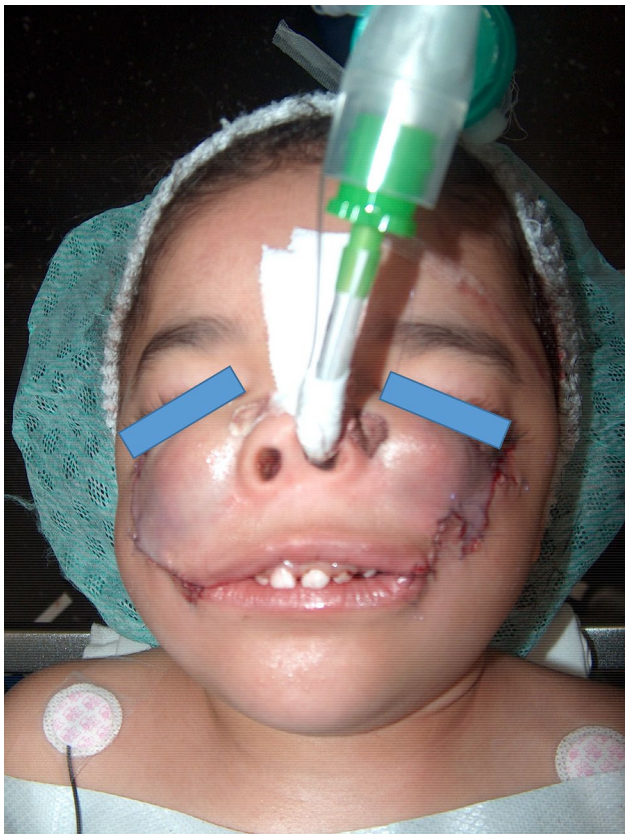

Fig. 3 Postoperative result after surgical correction of left oro-ocular cleft

uvula. The second lateral incision was made from the region of the pterygoid hamulus, transversely along the palatine shelf, parallel and anterior to the cleft. Our surgical technique required that we dislocate the pterygoid hamulus. The width of the two flaps of palatine mucosa and periosteum was measured so as to allow the minimum necessary lift to rotate and close the cleft. The anterior tips of the flaps were left attached to improve vascularisation of the flaps. A two-layer suture was performed. Examination with a microscope revealed no fluid in the middle ear (no secretary otitis media) and grommets were, therefore, not inserted.

Bone reconstruction will be performed later, when the child is 5–6 years old and after previous imagery (CT-Scan). Bone graft will be performed for maxillary cleft, and orbital floor reconstruction will be realized also.

Discussion

A number 5 cleft of the Tessier classification system is an extraordinarily rare facial cleft, representing on average 0.25% of all facial clefts [8, 11, 26]. One-fourth of the cases are unilateral, one-fourth are bilateral, and the remainder are combined with other clefts [6]. We present here the first reported case of a Tessier Number 5 bilateral cleft.

The human face is formed between the 4th and 6th weeks of embryonic development. Facial swellings arise on the frontonasal process (2 medial nasal and 2 lateral nasal processes) and the first pharyngeal arch (2 mandibular and 2 maxillary processes). These processes come together and fuse to form the continuous surfaces of the face. The primary palate is also formed during this period by fusion of the medial nasal and maxillary processes. Subsequently, between the 6th and 12th fetal weeks, the secondary palate is formed as the result of fusion of the palatal processes growing from the oral surfaces of the maxillary processes. All fusion abnormalities could result in a potential facial or palatal cleft.

Number 5 Tessier clefts generally start medial to the commissure of the mouth and lateral to the philtral column (Fig. 1). The cleft continues laterally to the anatomically normal nasal ala into the cheek, and extends through the lower eyelid. This last has a coloboma in the middle third, with a shortening of the distance between the lower eyelid and the mouth. The medial canthus is usually in a normal position, and the lateral canthus is deformed and

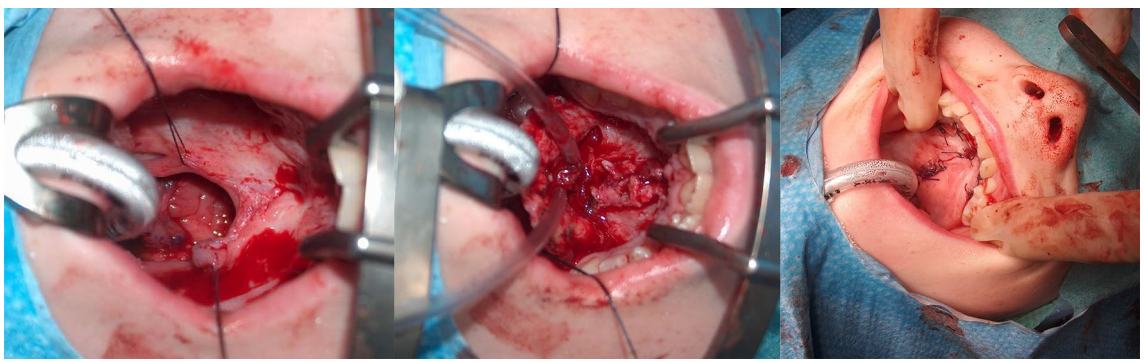

Fig. 4 **a** Bilateral total cleft: preoperative status; **b** nasal plane using vomerian and lateral mucosae; **c** buccal plane after reconstruction

malpositioned. The nasal ala are normal in structure but may be rotated toward the ipsilateral medial canthus. There is significant deficiency in the midface soft tissue. On the bony level, the cleft starts in the alveolus in the premolar region behind the canine. It extends upward, with the infraorbital nerve generally lying medial to the cleft. The cleft finally ends in the lateral portion of the inferior orbital floor. The bony deficiency of the inferior orbital floor and rim results in an inferior displacement of the globe [1, 13, 30].

Although the detailed structure of rare craniofacial clefts is well established, the cause of these pathological conditions is not [9, 30]. The first theory sustains a lack of fusion between the facial processes [11]. This hypothesis is the most popular and explains common clefting deformities. However, rare clefts, such as Tessier number 4 or Tessier number 5, cannot only be explained by this phenomenon. There are additional causes that also play a key role in these deformities [21]. The differentiation of the mesoderm into bone centres and musculature could be involved in Tessier's clefts number 4 and 5 [30, 34, 38].

Several others theories governing the pathogenesis of facial cleft have been proposed, such as malformation of the cranial base [30], a lack of mesenchymal migration and/or penetration [11], insufficient mesodermal penetration [23], error in differentiation of neuroectoderm and mesenchymal precursors of both skeletal and connective tissues of the face [39] and amniotic bands [15].

Many attempts have been made since the nineteenth century to establish a classification of craniofacial defects. Morian was the first to describe, in 1887, three types of facial coloboma [19], depending on the location of the maxillary cleft (between the central incisors and the canina) in relation to the ocular region. Our present case corresponds to a "Morian III" [19]. In 1976, Tessier introduced a classification using the orbit as a central landmark and following southbound (facial) and northbound (cranial) lines through the lips, maxilla, nose, eyelid or eyebrows [30]. As Tessier first stated, the severity of soft tissue and bone deficit is very variable, and the true occurrence of the different facial cleft types is difficult to evaluate because of the coexistence of different clefts. This was further confirmed by Stretch and Poole in 1990 in a review of 170 patients with craniofacial clefts [28]. He showed that midfacial and laterofacial clefts are rarely isolated, but often seen to overlap the adjacent cleft area (according to Tessier's classification) or to extend downward along their axis [18]. More recently, and on the basis of his studies of cleft embryology and anatomy, Van der Meulen renamed this cleft "medial maxillary dysplasia" [36]. Finally, the latest review, by Zhou et al. [40], proposed a supplement to the Tessier classification which conferred greater importance to the severity and localization of the tissue deformity. This STO subclassification (S for skin, T for soft tissue, and O for os or craniofacial bone) was proposed to better describe the clinical aspects, to facilitate pre-surgical planning and to provide standardization of postoperative evaluation [40]. However, the Tessier classification remains the most widely used among craniofacial surgeons and will, therefore, be used in the remainder of this article.

The Tessier cleft number 5 remains one of the rarest malformations, with only one case described in the literature over the past 130 years [23].

Facial bone, muscular atrophy or retraction, and soft tissue abnormalities justify radiological investigations to rule out encephalocele and define orbit and skull base anatomy [8]. The complete number 5 oro-oculofacial cleft includes bone and soft tissue defects to a variable degree. It extends vertically from the inferior eyelid and laterally to the infraorbital foramen on the orbital floor, continuing down through the maxillary sinus and the cheek to penetrate the maxillary arch in the non-classical location of the labiomaxillary cleft (lateral) [13, 31]. Exstrophy of the maxillary sinus is frequently associated, and there is usually a marked decrease in the oculo-oral distance, as was observed in our case. The eye can be functional, microphthalmic or anophthalmic [9].

Surgical management of this exceptional cleft is a real challenge and clear guidelines adapted to its epidemiology are not yet well defined. The sequence followed for a common facial cleft [3, 14, 17, 22, 29, 37] can be adopted. The repair of this kind of complex cleft requires a multidisciplinary surgical approach and multiple staged procedures. The surgical schedule for cleft reconstructions varies from one specialized centre to another according to their respective philosophical and practical priorities. In our center, a simple unilateral complete cleft lip and palate is closed in one procedure that includes a veloplasty to reconstruct the muscular part of the palate (at 4 months of age), a labioplasty and, if possible, a palatoplasty with two small flaps preserving the whole palate. For very wide clefts, closure of the hard palate is done a few months later, especially since over the period of time between these two operations the palatal plates acquire a more horizontal position and the width of the cleft diminishes. This allows one-layer closure of the cleft without lifting the periosteal flaps, which we know has a negative impact on subsequent palatal growth. But since our patient was already 2 years old, we did not follow our usual surgical schedule.

The urgent primary concerns in our case were the closure of the palatal cleft to allow proper feeding and a better chance of normal language acquisition, and the repair of the left lower eyelid to protect the remaining, dystopic eye. But severe midface hypoplasia is to be expected in the long term in any case of bilateral Tessier number 5 cleft, which means that secondary bone grafting and possibly osteotomy may later prove necessary.

There are some case reports of facial clefts associated with cleft palates, but in most of them, there is a lack of

detailed anatomy of the palate. Furthermore, few articles mention the surgical procedure. Sano et al. reported a bilateral facial cleft managed by the Veau–Wardill–Kilner procedure [27]. Uchikawa et al. described their management [33] as a palatoplasty to reconstruct the muscle bands and the velum cleft. We, therefore, chose to follow the one-step modified von Langenbeck procedure to close the palate and normalize feeding and language acquisition [4, 25]. This technique, described 120 years ago, is still one of the most commonly used surgical techniques for palate closure. It involves a straight-line closure of the palatal cleft with bilateral relaxing incisions, which create bipedicled flaps of hard palate mucoperiosteum. The greater palatine neurovascular bundles are kept intact and mobilized. The palatal muscles are dissected through the cleft margins and repositioned for end-to-end suturing. Surgeons attempting to improve velopharyngeal function for speech have widely adopted intravelar veloplasty as a modification of the traditional von Langenbeck procedure [32].

The Tessier number 5 cleft presents with inferior lid coloboma and variable degrees of damage to the eyeball. The lower lid does not close completely, and the exposure of the cornea, combined with an abnormal nasolacrymal duct, usually sets the stage for chronic conjunctivitis and ulcerations, or permanent scarring of the cornea [7]. We decided that a prompt repair of the coloboma was indicated to preserve the cornea as much as possible.

The surgical technique used was multiple Z according to Mustardé, which includes cheek advancement, lengthening of the oro-ocular distance and closure of the cleft soft tissues without tension, thus providing a good primary esthetic result [20]. Recently, Menard has proposed the use of tissue expansion to provide good texture and color as a technique for cases where retractile scar tissue is observed [16].

Conclusions

Severe facial malformations require careful planning of their staged reconstruction. We describe here a rare case of complete bilateral Tessier craniofacial cleft number 5, offer its explanation and report our surgical management of it.

The taking into account of the social and psychological aspects of therapy, as well as a regular multidisciplinary follow-up during the growing-up years, are necessary for a successful result, i.e., a happy, integrated individual.

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Compliance with ethical standards

Conflict of interest All the authors declare that they have no conflict of interest.

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