Tessier No. 3 and No. 4 clefts: Sequential treatment in infancy by pre-surgical orthopedic skeletal contraction, comprehensive reconstruction, and novel surgical lengthening of the ala base-canthal distance

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ABSTRACT

Background: Repair of facial clefts implies wide tissue mobilization with multi-stage surgical treatment. Authors propose pre-surgical orthopedic correction for naso-oroorcular clefts and a novel surgical option for Tessier No. 3 cleft.

Methods: Two male infants, a Tessier No. 3 cleft (age 7 months) and another Tessier No. 4 (age 3 months), were treated with a modified orthopedic Latham device with additional septo-premaxillary molding and observed to age four years. Tessier No. 3 orthopedic measurements were obtained by image corrected cephalometric analysis. Subsequent repair included tissue expansion on Tessier No. 4 and naso-frontal Rieger flap combined with myocutaneous upper lid flap on Tessier No. 3.

Results: Orthopedic movements ranged from 18.5 mm in bi-planar to 33 mm in oblique analyses. Tissue margins became aligned with platform normalization. Tissue expansion on Tessier No. 4 improved distances from ala base-lower lid and subalar base-lip. The naso-frontal flap combined with myocutaneous upper lid flap on Tessier No. 3 had similar achievement, but also sufficiently lengthened ala base-canthal distance.

Conclusions: Repairs were facilitated by pre-surgical orthopedic correction. The naso-frontal flap combined with an upper lid myocutaneous flap seems viable as a single-stage option to lengthen ala base-canthal distance to advance repair achievement in unilateral Tessier No. 3.

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1. Introduction

Facial, cranio-facial and latero-facial clefts (Tessier, 1976a), also known as rare clefts (Fogh-Andersen, 1965), have been systematically described and classified by Paul Tessier (1976a & 1976b). The Tessier classification, adopted worldwide, numbers clefts from 0 to 14 in a clockwise (right side) or counterclockwise (left side) progression in reference to the orbit. Nevertheless, Tessier’s classification does not meet embryological criteria (Pfeifer, 1991; Fearon, 2008). A more comprehensive classification for craniofacial anomalies, based on embryological and teratological studies, was introduced by Gerhard Pfeifer (Pfeifer, 1974, 1991; Gundlach, 1981). In this paper, the terminology of Tessier (1976a) is used with anatomical description (David et al., 1989; David, 2006). Clefts No. 3 to 5, like the cases presented, are considered the most challenging clefts in which the lip, the cheek and the orbit are compromised (Alonso et al., 2008). There is no consensus in the therapy for the naso-oro-ocular clefts (David, 2006), nevertheless four main approaches for those clefts have been described (Alonso et al., 2008; Ortiz Monasterio and Taylor, 2008) as a Z-plasty proposed by Tessier in which upper lid and lower lid tissue is transposed (David, 2006) a cheek rotation and advancement flap according to van der Meulen (1985) (Alonso et al., 2008; Giglio et al., 2008; Chen et al., 2012; Sesenna et al., 2012; Wu et al., 2013) and soft tissue expansion (Moore et al., 1992; Ortiz Monasterio and Taylor, 2008) or forehead flap (van der Meulen, 1985; Ortiz Monasterio and Taylor, 2008) for major soft tissue deficiency. Other reported therapy to fill the wide cleft gaps employ rib bone grafts (Sari et al., 2003; Eppley, 2003). All those techniques provide, in a different extension, a limited effect on the lengthening of the nose and mid-facial height, which is evident through the commonly observed postoperative lid retraction and malposed ala in an upward direction.

Furthermore, primary surgical repairs were generally isolated from infancy and routinely without Pre-surgical Infant Orthopedics (PSIO) (Resnick and Kawamoto, 1990; Toth et al., 1990; Longaker and Siebert, 1996; Longaker et al., 1997), save for molding plate “passive” PSIO facilitating lip closure in a Tessier No. 3 (Stellzig et al., 1997), and recently pin fixation “active” PSIO for bilateral rare No. 3-4 cleft (Maeda et al., 2014). One article reported a mean age of 5.4 years for 21 cases treated over a period of 20 years (Alonso et al., 2008).

1.1. Case studies

The cases herein presented in infancy are consistent with Tessier No. 4 (Fig. 1) and No. 3 (Fig. 2) clefts.

1.1.1. Case 1

A male infant with Tessier No. 4 cleft (central orbital) included the type 8 variant (quasi-Goldenhar sequence) with ophthalmic malformation (Fig. 1). He presented for consultation at neonatal intensive care unit, William Beaumont Hospital, Royal Oak, MI, USA. Associated deformities included bilateral sensorineural deafness, right coloboma through lower lid, iris, upper lid and brow, bilateral branchial arch remnants, right microtia, right microphthalmia, right ocular dermoid, a patent ductus arteriosus and patent foramen ovale. His lip and alveolar cleft gaps were 25-mm. Oculoplastic examination reported severe defects of right eye unreactive to light, the left eye was able to fix and follow, a large coloboma involving most of the right upper eyelid with a small intact lid margin laterally, and right lower eyelid coloboma was present medially with a clinically absent lacrimal system.

2. Methods (Case 1)

Both cases were treated with the same PSIO protocol. The appliances used were like that reported by Latham (1980) (Fig. 3). The two-armed stainless steel hinge joints were positioned over each maxillary tuberosity. To prevent rotation of the lesser segment (LS) hinge, it was parted to afford LS antero-medial displacement while causing the greater segment (GS) “anchor” to rotate to medial during activation. Delivered under general anesthesia devices were retained by pins and screws (Fig. 3). Parents activated devices daily by clockwise screw rotation for 8 weeks, usually with weekly monitoring. To aid correction, a novel nasal septum-button-head pin (NSBP) (Fig. 1, right & Fig. 2, right) was placed through the GS nostril with the oral-side pin-end chained to the LS for septo-premaxillary molding (Spolyar and Roldan, 2015). Chain placed across appliance gap provided a third corrective force.

2.1. Procedural tactic

Active PSIO and septo-premaxillary molding begun at 3 months of age. At device removal, first stage cleft repair with lip adhesion performed at age 5 months Palatoplasty according to Furlow (1986) combined with bilateral buccal flaps (Mann and Fisher, 1997) performed at age 10 months. Cheek, lid and final lip repair performed at age 3½ years after tissue expanding the cervico-facial flap.

3. Results (Case 1)

Despite successful presurgical orthopodics, soft tissue repair was very challenging. Two expanders were explanted because of patient trauma and dehiscence. The third was placed in the forehead but not used in orbital repair. The cervico-facial rotation and advancement flap lengthened the nose ala-lower lid distance; the lower lid became less retracted and the ala was slightly retracted in an upward direction and the lip length improved (Fig. 4). The dental occlusion became remarkably normal (Fig. 4).

3.1. Case 2

A male new born with Tessier No. 3 cleft (medial orbital) included type 7 variant with affected zygoma, temporal, mandibular, and coronoid components (Fig. 2). He presented for consultation at Craniofacial and Cleft Palate Diagnostic Clinic, St John/ Providence Hospital, Novi, MI, USA. Anthropometry (Table 1A) revealed very wide left-side 25-mm gaps. The defects traversed the philtrum column to nasal floor with alveolus-palate extension. The left alar base was severely pulled up with severe vertical shortening between ala and lower lid. Left medial lower lid harbored a coloboma (medial to lacrimal punctum). There was disruption of lacrimal apparatus and inferior displacement of medial canthus. Right upper lid coloboma presented with lagophthalmos. Both nostrils presented with bilateral nasal septum cysts. The left ear was rated with grade 3 microtia with maximal conductive hearing loss; the right side had a preauricular appendage. The bony cleft extended through alveolus, to piriform aperture, through medial maxilla to medial inferior orbit. Bilateral mandibular hypoplasia with right grade I, left grade II was appreciated. The LS had severe dorsal dystopy and associated malar hypoplasia, short vertically and transversely. Frontally, GS was rotated laterally and elevated (cephalad) with severe nasal septum deviation.
4. Methods (Case 2)

PSIO protocol was the same as for Case 1 with additional objective cephalometric documentation of the active orthopedic phase repositioning the diastatic skeletal components.

Cephalometry employed a portable cephalostat (Spolyar, 1988) (Porta Stat, Inc., Clinton Twp, MI, USA) intra-operatively to secure frontal and lateral cephalometric surveys taken at device delivery (age 7 months) and removal (age 11 months). Lateral surveys secured at 165 cm source to image-receptor distance (SID) and 15 cm midsagittal-plane object to image-receptor distance (OID) and exposed at 5 mAs and 65 kVp, and frontal A–P views exposed at 112 cm SID and 1 cm squama-occipitalis-plane OID with 5 mAs and 65 kVp technique.

Study landmarks included the ends of pins and screws, the button of nasal septum button-head pin, and the device GS and LS button-hooks (Fig. 5). All bilateral landmarks were matched in the lateral and frontal surveys at each time point along with the customary cephalometric landmarks (Table 2 and Fig. 5). Landmark points were digitized with backlighted Numonics AccuGrid digitizing tablet with 0.0127 mm resolution and 0.127 mm accuracy (Numonics Corporation, Montgomeryville, PA, USA).

Three-dimensional measurement (3D) of intra-component landmark distances at each time point was compared to indicate intra-component landmark stability. Orthogonal sagittal plane measurement of parasagittal landmarks was derived from image corrected cephalometric analysis (ICCA) (Spolyar et al., 1993). Based on an earlier study (Spolyar, 1987), the bilateral superior orbital rims were used for reference body required by ICCA method reformat image artifact. The ICCA method accuracy reported at 0.2 mm for bone marker studies (Spolyar et al., 1993). Orthogonal transverse plane measurement derived directly from each frontal survey was assumed reliable. Non-orthogonal measurement of oblique inter-component parameters was derived from the 3D procedure.

4.1. Procedural tactic

Active PSIO and septo-premaxillary molding started at month age 7 and ended at 9; cleft repaired occurred at 11 after 6 weeks. To lengthen ala base-canthal distance, the repair was done and designed to include a fronto-nasal flap according to Rieger (1967), extended with a transposition and advancement upper eyelid myocutaneous flap, (Fig. 6 top-left and middle-row). The exposition of the medial canthal area through the frontonasal flap allowed an exact eyelid reposition. The medial canthal ligament on the affected side was fixed by using resorbable ultrasound driven pins (Sonicweld®, KLS- Martin, Tuttingen, Germany). The repair included a lip rotation and advancement flap according to Millard.
The rational for Rieger flap modification approach was the following:

- The lengthening the ala base-canthal distance is addressed by a frontonasal advancement flap as described by Rieger to repair skin defects of the nose tip (Rieger, 1967). The lengthening problem is further improved by incorporating an upper eyelid myocutaneous flap (Tripier flap) (Elliot and Britto, 2004) transposed and advanced with the Rieger flap to the lower lid.

- This eyelid transposition has again, as an advantage, the reduction of the upper eyelid with enlarged distance in reference to the brow. This flap is expected to expand with dissipation of tension in the midface to further increasing ala base-medial canthal distance as observed when the lateral based...
upper eyelid myocutaneous flap is used for the correction of lower lid retraction (Jackson, 1985).

- The access into the canthal region through the frontonasal flap provides a wide exposition to perform a medial canthopexy, since the medial canthal ligament is typically displaced caudally.

5. Results (Case 2)

5.1. Cephalometric analysis

All Intra-component landmark pairs were stable within an absolute range of (0.07) to (0.87) mm, and for LS study pairs #127–128 (0.38) mm and GS, #132–133 (0.07) mm (Table 1B, 3-D).

Mean horizontal displacement of the LS landmarks #127–128 (pin-ends) was (+)11.3 mm and GS landmarks #132–133 (pin and screw-ends, respectively) (–)2.8 mm in Sagittal Plane (Table 1C). Net Sagittal Plane contracting displacement between LS and GS was 14.1 mm along SN (coordinate horizontal axis) from Sella (constructed coordinate vertical axis through Sella). A 12 mm Transverse Plane contraction occurred in frontal view. Vertical plane relocations of the bilateral landmarks were symmetrical in displacement.

The derived hypotenuse of translation in the horizontal plane of 18.5 mm measured closure or contraction between LS and GS at the locus of pin and screw-ends (Table 1D).

Non-orthogonal Oblique translations measured movement of anatomy peripheral to pin and screw-ends and represented oblique inter-component translations during the pre-surgical period (Table 1E). Those peripheral landmarks relocated with spatial contraction of 33.0 mm from LS posterior screw-tip to button of the NSBP (#129-LS to #92-GS, respectively) and 32.1 mm from device LS “chaining” button to GS posterior retaining pin-end (#93-LS to #131-GS, respectively).

5.2. Surgical assessment

Immediately after surgery, the upper eyelid myocutaneous flap expanded increasingly the ala base-canthal distance (Fig. 5). The eyelid competence could be restored at one stage. The result remained stable over an observational period of two years. At six months, 12 months and 24 months postoperatively the ala base-canthal distance increased progressively, and due to the nature of the eyelid myocutaneous flap, the transposed tissue matches to the lower eyelid in color, texture and thickness providing a pleasant scar (Fig. 6).

6. Discussion

6.1. Pre-surgical orthopedic treatment

Surgical repairs for Tessier type large facial clefts are generally not done in infancy (Alonso et al., 2008). The unique feature of our PSIO device is the marriage of “anchorage” (GS) and “target” (LS) (elements) with orthopedic translations complementarily corrective. With only one reported case utilizing pin retained PSIO

Table 2

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<thead>
<tr>
<th>Landmark type</th>
<th>Location</th>
<th>Lateral # ID</th>
<th>Frontal # ID</th>
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<tr>
<td>Retaining Pin-end:</td>
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<tr>
<td>Retaining Screw-tip:</td>
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<tr>
<td>Device button:(Nasal septum buttonhead pin)</td>
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<tr>
<td>Anatomic:</td>
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<td>Derived: (124–121 relative shifting between surveys)</td>
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LS – Lesser segment of the cleft components, Case left side.
GS – Greater segment of the cleft components, Case right side.
Rt – Right.
Lt – Left.

Fig. 5. Case 2, infant affected by Tessier No 3 cleft. Pre-treatment cephalometric views with the pinned directional orthopedic traction device in place: (left) lateral cephalometric view with numbered landmarks, (right) antero-posterior cephalometric frontal view with numbered landmarks. See Table 2 for definition of numbered landmarks seen in these two views. Anatomic landmarks used for reference between views are Sella and Nasion. Supraorbital was used to define image distortion to perform image corrected cephalometric analysis. Notably displaced is the nasal septum located by landmark #92 in the initial frontal view at the button-head.
(Maeda et al., 2014), the nose and septo-premaxilla became clinically centered without objective documentation and no need for complimentary orthopedic correction as was essential for our cases. Alternatively, compliance sensitive passively retained nasoalveolar molding (NAM) necessarily applied before age 12 weeks (Barillas et al., 2009) is tactically limited as compared to our protocol.

The compliant circum-maxillary suture system and stimulated adjustment response (Latham, 1974) provides the possibility for sufficient corrective displacements done with impunity. Infancy confers the highest growth rate essential for “closed” distraction. The youngest treatment age recommended is after infants achieved 4 kg weight, and about 8 weeks age in deference to drug clearance (Björkman, 2005, 2006) and anesthesia technicalities. Perhaps, the procedure could be applied in children beyond age 2 years following surgical release of resistant buttresses.

Fig. 6. Top row shows intraoperative frontal views of infant affected by a Tessier No. 3 (case 2); (top, left) patient age 11 months at time of surgery after presurgical assistance, (top, center) flaps shown elevated and reflected for repositioning, (top, right) view after closure of flaps. Middle row shows schematic of proposed flaps in three steps: (middle, left) flap design for modified Rieger flap with extended transposition and advancement of upper eyelid myocutaneous flap, (middle, center) dissected flap elevated position of modified Rieger flap, (middle, right) closed design of modified Rieger flap. The bottom row shows postoperative view of patient: (bottom, left) at age 12 months soon after single stage comprehensive. Note the midfacial tissue deficiency replaced by the modified Rieger flap extending the nose ala-canthal distance, (bottom, right) patient at age four years.
Regarding our cephalometric methods for the Sagittal Plane and 3-D measurements, error artifact unlikely exceeds 0.2 mm per non-anatomic landmark studied. The superior orbital rims used for reformatting have less precision and accuracy (Baumrind and Frantz, 1971) compared to bone marker landmarks (Spolyar et al., 1992). Yet, in Table 1C, the bilateral vertical translations were, as expected, equal (0.8/0.9 mm) indicating a valid reformatting process.

The GS retracted 2.8 mm and LS advanced 11.3 mm effectively achieving ala base and piriform rim symmetry (Fig. 1C) and gaining a 4.2 mm [(11.3−2.8)/2] net mean advancement for the centroid of the entire maxilla while restoring a symmetrical maxillary platform. Perhaps, the Class III left-side dental occlusal loss (Fig. 4) reflects a cost related to the GS “anchorage-retraction” supporting the larger LS “target-distraction”. Nevertheless, corrective movements followed rotational paths, although analyzed as linear translations, the complimentary rotations totaled about 45° with segmental translations about 30 mm at the clinical anatomical level.

Growth effects of (PSIO) and surgical repairs provide controversy in common cleft lip/palate treatment. Generally, in oblique facial clefts with diagnostic relevance, midface hypoplasia is likely due, in part, to chronic growth retardation (Versnel et al., 2011) and intrinsic unilateral maxillary hypoplasia (Versnel et al., 2009) in which cases medial canthus and alar base repositioning remained problematic (Chen et al., 2012). Growth injury increases with more interventions (Anastasov and Joos, 2001). Yet multiple surgeries are common for severe facial clefts (Versnel et al., 2011).

Latham PSIO, it alone is faulted for impeding maxillary growth gleaned from occlusal relationship studies (Berkowitz et al., 2004; Henkel and Gundlach, 1997), they too are confounded by gingivoalveoleoplasty, while it alone is faulted for compromised cephalometric values without active PSIO (Meazzini et al., 2010). Notwithstanding, other UCLP occlusion studies with either passive PSIO and sequential primary bone grafting (Hathaway et al., 1999) or active Latham PSIO without primary alveolar surgery (Chan et al., 2003) showed both protocols achieved dental arch relationships in preadolescence like children without PSIO intervention nor alveolar grafting. Finally, active Latham PSIO treatment categorically shifts the native growth-compensated developmental derangement while normalizing cleft component positions (Spolyar et al., 1992). In a UCLP cast study, larger cleft width (derangement) in infancy between cuspid points was significantly associated with less anterior and posterior dental crossbite at age 5 years (Reiser et al., 2010); would that the native growth-compensated developmental derangement having been normalized by PSIO, then lead to more crossbites? Hence, maxillary growth effects are multifactorial reflecting protocol tactics, treatment procedures and the individual treatment response related to sustaining critical life functions, hopefully achieved early on.

6.2. Surgical repair of oro-orbito-facial clefts

Lengthening ala base-canthal distance is the main goal when repairing orbital-facial clefts. A cheek rotation flap addressed this issue on a Tessier No. 4 cleft (case1) with little improvement. However, in the unilateral Tessier cleft No.3, the proposed frontonasal Rieger flap extended with a myocutaneous upper lid flap proved value as a one-stage procedure for the management of soft tissue infra-canthal deficiency. The advantage of the myocutaneous upper lid flap is clearly demonstrated in the management of lower lid retraction in congenital cases like Treacher Collins syndrome (Jackson, 1981) or in acquired cases (Jackson, 1985). The progressive lengthening of the mid-face is remarkable compared to the traditional techniques including the rotational advancement cheek flap, as reported by Chen et al. (2012) in order to avoid Z-plasties in bilateral Tessier Clefts No. 3 and 4; in their presented cases the lower lid retraction was not resolved uniformly and the upper eyelid distance to the brow was not corrected. Many authors performed a V-Y glabellar flap in different extension for the lengthening of the nose (Jackson, 1981; van der Meulen, 1885; da Silva Freitas et al., 2009; Mathijssen and van der Meulen, 2010; da Silva Freitas et al., 2010; Sesenna et al., 2012). The paraglabellar flap (Jackson, 1981) provides a very limited advancement. The Rieger flap is, in contrast, dissected from the epicantal ligament down to the nasolabial sulcus. This flap extension gives more freedom in covering defects by advancement than the classical glabellar flap limited to the upper half of the nose (Rieger, 1967). The Rieger flap was mention by Kawamoto’s group in 1992 in the management of Tessier clefts (Shewmake and Kawamoto, 1992) nevertheless, the value of this flap was not reported again. The upper eyelid myocutaneous flap for the correction of Tessier Cleft No. 3 was reported (Alonso et al., 2008; da Silva Freitas et al., 2010) yet, as a trans-position flap, the rotation is limited as the base of the flap remains almost fixed, unlike our case with myocutaneous flap incorporated with the Rieger flap which is also advanced providing enhanced lengthening of the ala base-canthal distance.

6.3. Pre-surgical and surgical therapy for oro-orbito facial clefts, thoughts and achievements

In the presented cases, independent of cleft width, active PSIO and septo-premaxillary molding, as described, introduces the concept of skeletal cleft contraction; instead of extensive tissue mobilization or expansion, the cleft should be orthopedically narrowed. Having the embryological development of the head in mind (Pfeifer, 1991) the proposed concept is to move the diastatic cleft components forward and medially, conceptually different to the conventional and not embryological concept of “filling the gap”. Mühling and co-workers gave advise about orbito-maxillary osteotomy and medial skeletal advancement before the soft tissue repair (Mühling et al., 1989). Further underscored by Millard, “... as in most architectural constructions, the platform deserves priority” (Millard, 1990). To the best of our knowledge, the Rieger frontonasal flap extended with an upper eyelid myocutaneous flap has not previously been reported as a viable option in the repair of unilateral Tessier Cleft No.3.

7. Conclusions

Presurgical assistance facilitates comprehensive repair of the severe facial clefts, even with single-stage primary defect repair during infancy. Lengthening of the ala base-canthal distance is a key achievement, it can be achieved by performing a frontonasal flap extended with a myocutaneous upper lid flap.

References
