A classification and construction of congenital lateral facial clefts

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SUMMARY. Background: The repair of the lateral or transverse facial cleft is a surgical challenge on the account of the abnormal positioning and appearance of the cleft. Materials and methods: Over a twenty-seven year period, 22 lateral facial cleft cases were evaluated at a cleft lip (CL) and palate clinic and seven children underwent reconstruction of the lateral CL. Results: Twenty-two of 3187 (0.69%) cases presented with a lateral CL. Five of these 22 cases (23%) had a bilateral, eight (36%) had a right-sided and nine (41%) had a left-sided cleft. The evaluation of these cases resulted in a new classification (namely an extension of the Tessier 7 cleft) classification for the cutaneous and muscle involvement: a superior (T7.1), middle (T7.2), inferior (T7.3) and agenetic (T7.4) lateral CL. The altered surgical construction: an internal mucosal straight-line closure, a curved cutaneous—musosal red-lip/vermilion-lined flap for the lip commissure, muscle reconstruction at the modiolus and a positional cutaneous z-plasty for the rare lateral cutaneous cleft. Conclusion: The paper introduced a new classification for the lateral CL, as well as an altered surgical reconstructive technique for the most natural functioning of the lateral part of the face. © 2010 European Association for Cranio-Maxillo-Facial Surgery

Keywords: lateral facial cleft, transverse facial cleft, Tessier 7 cleft, classification, surgical reconstruction, repair

INTRODUCTION

Cleft repair is an integral part of the modern cranio-maxillo-facial surgical spectrum and remains a challenge because of insufficient and malformed tissue (minor to major) at the site of the deformity (Mommaerts and Nagy, 2008; Nagasao et al., 2008; Schwenzer-Zimmerer et al., 2008). This in itself will influence the short and long-term aesthetic (soft tissue and facial skeletal appearance) (Nagy and Mommaerts, 2007; Nollet et al., 2007) and functional (occlusal and speech) outcomes (Joos et al., 2006). In addition, the type of surgical repair and at what age it is performed also has a significant influence on the aesthetic (Carlino, 2008) and functional (Li et al., 2006) outcomes. The major challenge is not only understanding the genetics involved (Dostal et al., 2009), but also the design of the surgical procedure required in the uncommon types of clefts (Bütow, 2007).

Lateral or transverse clefts of the lip are quite rare and have generally documented to have an incidence of 0.3—1.0% of the total facial cleft deformity spectrum (Boo-Chai, 1969; Hawkins et al., 1973; Bauer et al., 1982; Verheyden, 1988; Gleizal et al., 2007), or of 0.02% of live births (Kuriyama et al., 2008). The cleft is usually obvious, but it might be difficult to see when combined with another congenital facial deformity (Hawkins et al., 1973), such as hemifacial (lateral facial) microsomia or oto-mandibular dysostosis, bilateral facial microsomia, occulo-auriculo-vertebral spectrum or Goldenhar and Treacher—Collins (Franceschetti) syndrome (Tessier, 1969; David et al., 1987). This specific malformation was classified by Tessier (1969), as a number 7 cleft (Tessier 7), which presents with both a soft tissue (cleft lip (CL) and pre-auricular tags) and a bony component (temporozygomatic cleft). Van der Meulen et al. (1983) classified this cleft as a maxillo-mandibular dysplasia. However, other authors have described the soft tissue involvement as a lateral CL (Van der Meulen et al., 1983; David et al., 1989), with or without pre-auricular appendages or fistulas (Van der Meulen et al., 1983) and bony involvement in the pterygomaxillary junction, or hypoplasia of the alveolar process (molar region), maxilla, zygomatic body, mandibular condyle or coronoid, which might also involve the sphenoid bone, especially the pterygoid plates (David et al., 1989).

The lateral CL may therefore occur in isolation, or in combination with other facial clefts (Tessier 2, 3, 4 or 5). Macrostomia, a lateral facial cleft with normal red-lip mucosal lining or vermilion, especially a short one, might not present as an obvious lateral cleft, and may even go unnoticed. Where there are no normal red-lip structures or vermilion in the cleft area, the defect is usually noticed immediately.

Numerous surgical reconstructive techniques have been developed and used to normalise the appearance of this unsightly defect. Examples are the straight-line closure, without supra-inferior rotation or positioning of the scar (Kaplan, 1981; Ono and Tateshita, 2000; Bauer and Margulis, 2002; Gleizal et al., 2007; Kuriyama et al., 2008), or W-plasty (Bauer et al., 1982; Habal and Scheuerle, 1983; Eguchi et al., 2001). Some authors prefer a type of z-plasty for the closure, which may be positioned horizontally (Boo-Chai, 1969) or inferiorly (Verheyden, 1988; Yoshimura et al., 1992; Torkut and Coğunfırat, 1997).
This report describes a new classification of the lateral cleft, based on the anatomical appearance and a surgical procedure in the form of a four-layered approach with a modified z-plasty for the cutaneous part.

MATERIALS AND METHODS

Database

The database of the CL and palate clinic was analysed for the incidence of lateral facial clefts, as well as its various associated presentations: CL — unilateral left-sided, unilateral right-sided cleft or bilateral — as well as the superiorly rotated, middle-positioned, inferiorly rotated and agenetic lateral CL.

Surgical technique

The surgical technique consists of the creation of a straight-line closure of the mucosa (Fig. 1a), a red-lip commissural flap rotated from the superior part of the lip (which, as a rule, is an extended part of the natural red-lip component) (Fig. 1b), a modiolus muscle reconstruction at the confluence of the orbicularis oris, zyomatic major, risorius and depressor anguli oris muscles (Fig. 1c) and a (mostly superiorly rotated) modified z-plasty flap (Fig. 1d) for cutaneous closure (Fig. 1e, f).

RESULTS

Database

An analysis of the 3187 CL and palate patients revealed that 184 presented with only a CL, 411 with a cleft lip and alveolus (CLA), 1267 with a cleft lip, alveolus and palate (CLAP), and 143 with a combination, such cleft as CL with cleft soft palate or other cleft combination (COMBI). A total of 22 of 2005 (1.1%) (the various groups with CL involvement) patients presented with a lateral or transverse cleft deformity. The subdivisions...
of this lateral cleft accounted for: CL (11), CLA (1), CLAP (0), and combination cleft (COMBI) (10) (Table 1). Fifteen (or 68% of 22) cases presented with a syndrome (Goldenhar: ten; oto-mandibular dysostosis: three; Treacher–Collins one; amniotic band: one). However, there are major variations in the appearance of the transverse facial cleft, and it may also present as: unilateral — left sided (9) (Fig. 2a), unilateral — right sided (8) (Fig. 2b), and bilateral (5) (Fig. 2c) (Table 2).

There are superiorly rotated (Fig. 3a), middle-positioned (Fig. 3b), inferiorly rotated (Fig. 3c) and agenetic lateral CLs (Fig. 3d). It appears that the classically described Tessier 7 cleft, mostly seen in oto-mandibular dysostosis and Goldenhar syndrome patients, is a superiorly rotated or middle-positioned lateral CL. This may involve only the lip, or may present with additional ear tags (very mild form), or rudimentary or displaced ears (severe form), or may even involve the maxillary dento-alveolar ridge, zygomatic or temporal bone. These four different lateral clefts require a classification, and the following is suggested (Fig. 4a, b):

- T for Tessier and (7) seven for his original description, such as T7.1 for the superiorly rotated cleft, subdivided into categories without bone involvement (T7.1a), and with bone involvement (T7.1b); T7.2 for the middle-positioned, subdivided into categories with soft tissue involvement only (T7.2a), and with soft tissue and bone involvement (T7.2b); T7.3 for the inferiorly rotated and T7.4 for the agenetic lateral facial cleft.

The last-mentioned two clefts were not seen with bone involvement, but a subcategory may be used, whenever such a case is described (Table 3). The underlying muscles which are involved: T7.1 — separation of the zygomatic major and risorius muscles (Kaplan, 1981); T7.2 — clefting of the risorius muscle (Verheyden, 1988; Gleizal et al., 2007); T7.3 — separation of the risorius and depressor anguli oris muscles and T7.4 — agenesis, or an anterior partial agenesis of the risorius muscle, with or without partial agenesis of the orbicularis oris muscle.

### Surgical technique

In a lateral cleft, the skin, muscles and mucosa may be reconstructed in such a way as to create a scar resulting in a good facial appearance. This can only be done, if a modified cutaneous z-plasty, rotated superiorly, with modiolus reconstruction is performed.

The repair of the lateral CL in this modified surgical design entails the following: a straight-line mucosal (intra-oral) closure; a rotated red-lip commissural (vermilion-lined) flap, derived from the superior part of the lip, for the creation of a scarless commissure and also

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**Table 1** — Total versus lateral CL (total cleft cases = 3187)

<table>
<thead>
<tr>
<th>All types of CL</th>
<th>CL</th>
<th>CLA</th>
<th>CLAP</th>
<th>COMBI</th>
</tr>
</thead>
<tbody>
<tr>
<td>2005</td>
<td>184</td>
<td>411</td>
<td>1267</td>
<td>143</td>
</tr>
<tr>
<td>Lateral CL</td>
<td>22</td>
<td>11</td>
<td>1</td>
<td>0</td>
</tr>
</tbody>
</table>

**Table 2** — Right, left-side or bilateral positioning of the lateral cleft for CL/CLA/COMBI, sex and race groups

<table>
<thead>
<tr>
<th>Lateral CL</th>
<th>CL</th>
<th>CLA</th>
<th>COMBI</th>
<th>Male</th>
<th>Female</th>
<th>White</th>
<th>Black</th>
<th>East Asian</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bilateral cleft</td>
<td>4</td>
<td>0</td>
<td>1</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>Right side</td>
<td>2</td>
<td>1</td>
<td>5</td>
<td>4</td>
<td>4</td>
<td>8</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Left-side</td>
<td>5</td>
<td>0</td>
<td>4</td>
<td>3</td>
<td>6</td>
<td>5</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>11</td>
<td>1</td>
<td>10</td>
<td>10</td>
<td>12</td>
<td>14</td>
<td>7</td>
<td>1</td>
</tr>
</tbody>
</table>

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Fig. 3 — Rotation of the lateral CL: (a) superiorly rotated (T7.1); (b) middle-positioned (T7.2); (c) inferiorly rotated (T7.3); (d) agenetic lateral CL (T7.4).

Table 3 — Classification of lateral or transverse CL.

<table>
<thead>
<tr>
<th>Main division of lateral CL</th>
<th>Subdivision</th>
<th>Description of lateral cleft</th>
<th>Associated syndromes</th>
<th>Appearance</th>
</tr>
</thead>
<tbody>
<tr>
<td>T7.1</td>
<td>Superiorly rotated (s-r)</td>
<td>32%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>T7.1a</td>
<td>s-r with soft tissue involvement only</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>T7.1b</td>
<td>s-r with additional bone involvement</td>
<td>Oto-mandibular dysostosis, Treacher–Collins Goldenhar</td>
<td></td>
<td></td>
</tr>
<tr>
<td>T7.2</td>
<td>Middle-positioned (m-p)</td>
<td>45%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>T7.2a</td>
<td>m-p with soft tissue involvement only</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>T7.2b</td>
<td>m-p with additional bone involvement</td>
<td>Oto-mandibular dysostosis, Treacher–Collins Goldenhar</td>
<td></td>
<td></td>
</tr>
<tr>
<td>T7.3</td>
<td>Inferiorly rotated (i-r)</td>
<td>14%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>T7.4</td>
<td>Agenetic type (a-t)</td>
<td>9%</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Fig. 4 — Drawing of the four different lateral clefts (T7.1–T7.4): (a) cutaneous involvement; (b) muscle involvement.

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to avoid the collapsed appearance (scar contraction and creeping obliteration) at the red-lip junction (Boo-Chai, 1969) and/or recurrent chelitis; a modiolus muscle reconstruction in the confluence of the orbicularis oris, thereby bringing about muscle continuity at the commissure, with involvement of the zygomatic major, risorius and depressor anguli oris. The distance from the opposite commissure to the centre of the cupid’s bow of the normal side in a unilateral lateral cleft case must be measured and the same distance is then applied to the cleft side. In a bilateral cleft, the vermilion usually shows a small defect, but occasionally there is no red lip or vermilion at the commencement of the cleft. The point where the cleft begins, up to the centre of the cupid’s bow (and an exact mirror image distance on the other side) (Kaplan, 1981) is the length of the new commissure, created by the rotated red-lip commissural flap. The cutaneous part of the lateral CL is closed by means of a special z-plasty, which is more straight for the superiorly rotated CL (or alternatively a straight-line closure may be performed) (T7.1), and the superiorly rotated z-plasty is used for the middle-positioned (T7.2), inferiorly rotated (T7.3) or agenetic lateral cleft (T7.4). The most superior point of the vertical leg of the z-plasty should be positioned more or less at or parallel to the naso-labial fold. The number of z-incisions used, depends entirely on the length of the cleft defect: with less than 1 cm, one might approach the skin reconstruction with a single z-incision (Fig. 5a–d), and with more than 1 cm, two or more z-incisions may be indicated.

Fig. 5 — Patient with a cutaneous z-plasty rotated superiorly with a single z-incision: (a) drawing; (a) neonatal appearance; (b) without smiling; (b) smiling.
In bilateral clefts, both sides are reconstructed at the same time (Fig. 6a–e), for optimal symmetry. Should the modiolus be absent or atrophic (T7.4) the zygomatic major and depressor anguli oris muscles should be surgically connected. However, lateral movement of the lip’s commissure is compromised where there is a weak or agenetic risorius muscle (T7.2 and T7.4).

In cases where the lateral cleft is quite short and lined with red-lip mucosa or vermilion, the reconstruction need only be performed after orthodontic, or after combined orthodontic—orthognathic treatment. This is because a macrostomia allows easier access to the oral cavity for any intra-oral treatment, particularly for the use of a distraction apparatus (Fig. 7a, b).

Only cases with obvious or severe clefts are reconstructed as neonates. Seven (32%), with or without bone involvement, underwent a lateral CL reconstruction while creating a functional commissure at the same time.

**DISCUSSION**

The lateral or transverse facial cleft may be obvious, especially where it is of substantial length and without red-lip mucosa. It is also more obvious when it is rotated inferiorly, or when it is of the agenetic type. Should the cleft be short, superiorly rotated or middle-positioned, lined with normal red-lip mucosa (also described as a macrostomia) and associated with a syndrome, it might remain undiagnosed until a comparison is made between the lengths of the lip on either side. It is therefore not surprising that only a few cases have been documented.
usually reported on an individual basis and when obviously present as a lateral cleft (Hawkins et al., 1973; Verheyden, 1988; Torkut and Coşkunfirat, 1997; Witters et al., 2001; Akinmoladun et al., 2007; Kuriyama et al., 2008).

Before this report describing 22 cases of congenital lateral facial cleft, the largest number of cases reported in a single paper, has been 10 (Gleizal et al., 2007). The incidence of lateral CL in the database, on which this publication is based, is 0.69% in 3178 facial cleft deformities. This is similar to previously reported incidences (Boo-Chai, 1969; Hawkins et al., 1973; Bauer et al., 1982; Verheyden, 1988; Gleizal et al., 2007). The cases reported here appeared with lateral clefts on the left side (41%), right side (36%) and on both sides (23%). Using Tessier’s original classification (Tessier, 1969) and extending the number of these four newer, lateral facial clefts may be categorized as superiorly rotated (T7.1 = 32%), middle-positioned (T7.2 = 45%), inferiorly rotated (T7.3 = 14%), or agenetic (T7.4 = 9%). Fifteen patients were also diagnosed with a syndrome (otomandibular dysostosis, Goldenhar, Treacher–Collins and amniotic band syndrome).

The reconstruction of a lateral CL is a surgical challenge (Ono and Tateshita, 2000). The surgeon must reconstruct the lateral part of the lip and face so that it is at its most natural-looking and functions most normally in relation to the naso-labial fold and he/she must also position the modified z-plasty so that it is neutral when smiling. If this is not done, the face may appear too serious or even angry-looking. The literature does not reveal a particular preference for the procedure of choice in this type of reconstruction (Ono and Tateshita, 2000). In addition it does not describe a technique for the creation of a naturally neutral, or even smiling, appearance. Many elaborate surgical techniques have been used to reconstruct the lateral CL (Verheyden, 1988). The combination of lateral CL (the cutaneous part, which could involve distant soft tissues too, for example ear deformities), with bone deformities (example: mandibular ramus and condyle, maxillary dento-alveolar ridge, maxilla, zygoma, etc.) has resulted in new non-Tessier classifications (David et al., 1987; Gleizal et al., 2007). Unlike the classification described in this paper, these classifications refer neither to the position of the cleft in the lateral part of the face, nor to its muscular involvement.

This new surgical approach, with its four-layered approach and special z-plasty, enhances the natural-looking appearance and the function of the lateral CL, especially for the T7.2–T7.4 patients, and avoids the inferiorly rotated or “goldfish mouth” appearance (Boo-Chai, 1969), which has been reported in many of the published surgical procedures for this cleft type (Kaplan, 1981; Bauer et al., 1982; Habal and Scheuerle, 1983; Verheyden, 1988; Yoshimura et al., 1992; Torkut and Coşkunfirat, 1997; Ono and Tateshita, 2000; Eguchi et al., 2001).

In cases where a lateral cleft is not obvious, especially in cases in syndromic cases, the reconstruction will usually be carried out in the adolescent years. At this age the macrostomia allows good access to the oral cavity for any extensive dental, orthodontic treatment or surgical intervention.

The disadvantage of the surgical design described in this paper is that the z-plasty, which may lead to a slightly longer scar, would be more noticeable if hyperplastic or keloid scar tissue forms. Additional surgical revision with adjunctive treatments may then be required.

CONCLUSION

The incidence of the lateral or transverse facial cleft in our series is low, namely 0.69%. A new classification, based on Tessier’s cleft classification, has followed on the identification of four different lateral cleft types: T7.1–T7.4. The rotational aspect of the appearances of the cleft and the muscles involved are particularly important. The construction or repair, adjusted using classification, utilizes a z-plasty to naturalise the smiling face. This is done by means of a four-layered closure involving the lip’s commissure and the adjacent cheek’s structures.

References


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