CASE REPORT

Bilateral macrostomia

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SUMMARY

Macrostomia as a rare facial deformity is classified among facial clefts. It is a rare congenital anomaly which affects the aesthetics and function of the oral cavity. It is usually associated with deformities of other structures developed from the first and second branchial arches. Bilateral transverse cleft occurring alone is uncommon. Here we report a case of bilateral macrostomia (bilateral lip cleft) in a 5-year-old girl as a sole entity without other skeletal and facial deformities.

BACKGROUND

Macrostomia, commissural or lateral facial clefts (Tessier 7 soft tissue) are unusual lesions that result from failure of the embryonic mandibular and maxillary processes of the first branchial arch to fuse properly and form the corners of the mouth.¹⁻² It may be seen alone or in combination with other anomalies, accompanied by varying degrees of severity and extent. It may range from a simple presentation in the form of a wide oral aperture with loss of commissural anatomy to a full-thickness defect of the face extending from the mouth to the tragus. Associated anomalies may also be present in the form of pretragal skin tags, defective development of the auricle and external ear, absence of the abnormalities of the temporomandibular joint, zygomatic arch, eyelids and polydactyly. There may also be varying degrees of skeletal hypoplasia and discontinuity of bone between maxilla and zygomatic bone, frontal and zygomatic bone and between the temporal and zygomatic bone. These lesions are usually unilateral and rarely bilateral. Lateral clefts are the most common orofacial clefts, second to the more common isolated cleft lip, with an incidence of 1 in 3500–5000 live births.¹⁻⁸ Facial cleft does not affect the patient, but nevertheless causes an abnormal facial appearance and disordered daily life functions. In this case report, we present a 5-year-old girl with bilateral facial clefts.

CASE PRESENTATION

A 5-year-old girl along with her parents reported to our outpatient department with a deformed face and a wide mouth since birth. Her perinatal history was normal and uneventful. Her milestones were recorded to be normal. She showed good psychological development, abilities of comprehension, cognitive skills and was of average intelligence, attending primary school. On examination, there was a wide mouth and bilateral extension of the angles of the mouth to the masseter area (figure 1A,B). The clefts were lined with skin externally and buccal mucosa internally. A clear line of

demarcation was noticeable where the lips ended and the defect began. Further physical examination did not show any other abnormalities. Based on the above finding bilateral macrostomia diagnosis was made.

DIFFERENTIAL DIAGNOSIS

- ▶ Treacher Collins syndrome
- ▶ Hemifacial microsomia

TREATMENT

The patient was referred to the department of oral and maxillofacial surgery for the surgical correction of bilateral clefts.

OUTCOME AND FOLLOW-UP

The prognosis of the current case was good.

DISCUSSION

Macrostomia is most often associated with a transverse facial cleft, classified by Tessier as a number 7 cleft. It is found in 1 out of 80 000 births and occurs more frequently in men. Most cases are unilateral, but bilateral involvement can be seen in 10–20% of cases. More than 50% of bilateral cases are isolated without additional ear or skin deformities. However, the defect is often larger than in unilateral macrostomia.³ In the present case, bilateral facial clefts were present.

Bilateral congenital macrostomia cases are more often isolated, without ear or skin deformities, than are unilateral cases. Although the aetiopathogenesis of macrostomia remains unknown, many theories have been proposed. The mandibular dysplasia and transverse soft tissue deficiency could be explained by a lack of growth of the mandibular process. However, although four cases of bilateral macrostomia did present with mandibular hypoplasia, the majority were isolated. Others have suggested a vascular aetiology, explained by the presence of a haematoma in the territory of the stapedia artery preventing fusion of the maxillary and mandibular processes. In the case of bilateral macrostomia, however, both stapedia arteries need to be pathological, making this explanation less plausible.²⁻⁴ Amniotic bands have been proposed as another alternative. Physical restricting forces from these bands can lead to the formation of unusual clefts such as macrostomia. The embryonic origin of lateral lip cleft may be related to the seventh embryonic week, when the maxillary and mandibular processes of first branchial arch merge laterally to fuse in a posterior to anterior manner and construct the corners of mouth and cheeks. Any disruption in this process may lead to lateral clefts. It is also claimed to be a postmerging anomaly due to

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Figure 1 (A and B) Profile of the patient depicting bilateral cleft of the angles of the mouth.

considerable clinical variability in expression.^{5–8} The manifestations of lateral facial cleft may vary from a slight commissural involvement and macrostomia, which sometimes remain undiagnosed specially when combined with other facial anomalies to a gross combination of soft and hard tissue extended from mouth to tragus.⁸

Although over 50% of the reported cases of bilateral macrostomia are isolated, this condition presents a therapeutic challenge. In the case of unilateral macrostomia, the presence of a normal side can serve as a guide for the surgeon to localise the best starting point for the commissure through transposition to the cleft side. This is quite accurate on the upper lip, but can be somewhat difficult on the lower lip unless a definite midline can be identified by a groove in the vermillion or between the mentalis muscles of the chin. In the case of bilateral macrostomia, the surgeon must define the commissure position without a normal contralateral guide. Repair thus requires the use of extraoral landmarks and normal measurements.^{2 6 8}

Because bilateral macrostomia represents a distinct entity from unilateral macrostomia, a revised classification with therapeutic implications could be made as follows:

Type I or minor unilateral macrostomia: the cleft termination resides medial to the anterior border of the masseter, with a cleft length between 1 and 2 cm. Repair of the cleft requires vertical reorientation of the orbicularis muscle. **Type II or major unilateral macrostomia:** The cleft extends sagittally to the tonsillar pillars (type IIa) or laterally distal to the anterior border of the masseter and extending into the area of the tragus (type IIb). Repair requires a masseter reconstruction associated with orbicularis reorientation with or without a reconstruction of the tonsillar pillar muscles. Linear sutures for skin reconstruction may be preferred as it does not require significant skin dissection. **Type III or bilateral minor macrostomia:** The therapeutic protocol is the same as in unilateral forms, but the surgeon must define the commissural position without the assistance of a normal contralateral side.^{2 9} **Type IV or bilateral major macrostomia with sagittal cleft extension (type IVa), or bilateral major macrostomia with transverse cleft extension (type IV b).** Treatment involves the reconstruction of soft tissue followed by reconstruction of the bony frame. Soft tissue reconstruction should be performed in the preschool period. In this period, excision of skin tags and the correction of macrostomia and prominent ear are appropriate. When unnecessary, the bone frame is not reconstructed in the early ages. Bone grafting and augmentation must be applied at older ages. At 5 years of age, cranio-orbito-zygomatic development is at 85% of adults. For

this reason, skin and bone reconstruction of the middle face have to be performed after age 6, and that of maxilla and mandibular interventions must be performed in the adolescent period.^{1 3 5 7} It is hoped that with public campaign and enlightenment activities, societal attitudes to children with congenital deformities will change. It is also hoped that the authorities will enact and enforce laws to ensure the rights of such babies, especially right to care and life.

Learning points

- ▶ Bilateral macrostomia as a rare soft tissue deformity can be repaired successfully with minimally visible scar by simple linear triangular flap.
- ▶ Z-plasty or W-plasty techniques are unnecessary in repairing transverse cleft lips.
- ▶ Aesthetics as well as function, especially in participants without other anomalies, is a great concern. Surgery should be performed in young age to avoid unwanted anxiety and psychological impacts on child and family as well as correcting sialorrhoea, speech problems and compromised chewing ability.

Competing interests None.

Patient consent Obtained.

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