Case Series
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Isolated Bilateral Macrostomia: Case Series and Review of Literature

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Abstract

Background: Macrostomia is a rare congenital anomaly of the face, especially the isolated type. This report aims to document our experience in managing this rare pathology.

Patients and Methods: All consecutive cases of children managed for isolated bilateral macrostomia at the Jos University Teaching Hospital were retrospectively reviewed.
Results: Five patients, aged between 10 weeks and 30 months were managed. They were all females and presented with bilateral symmetrical transverse lateral facial cleft (macrostomia). There were no associated anomalies, and no family history of facial clefts or any other congenital anomaly. All the patients had repair of the defect under a general anaesthesia with satisfactory outcome.

Conclusion: Macrostomia can present as an isolated entity. The final outcome of the repair depends on the technique of repair, function of the orbicularis oris muscle and the quality of scar.

Keywords: Macrostomia; Facial cleft; Neocommissure.

Introduction

Congenital macrostomia (lateral transverse cleft) is a rare facial developmental anomaly. It is believed to be caused when there is failure of mesodermal migration or merging to obliterate the groove between the maxillary and mandibular prominences[1-5]. The cleft may be unilateral or bilateral, partial or rarely complete extending from the angle of the mouth to the ear[6,7]. It is often associated with syndromes[4,8], the commonest types are the first and second brachial arch syndromes[1,2]. Macrostomia may also present as an isolated pathology[2,3,7].

A number of techniques have been used to correct this defect[2,9-12]. Generally the aims of surgical repair are to have accurate positioning of the tissues without contractile scar of the oral commissure, to have a functional oral musculature and a minimal cheek scar. In this report we present our experience in the management of a series of five cases of isolated bilateral macrostomia.

CASE REPORTS

Case 1

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Fig. 1: Photograph of case 1 showing bilateral symmetrical macrostomia

A 30months old girl presented to us with an unusually wide mouth since birth. There had been occasional excessive drooling of saliva as well as occasional leaking of food particles from the angles of the mouth. She had no difficulty with speech and there were no respiratory symptoms. There is no family history of similar problem or any other congenital anomaly.

Examination showed a healthy looking child, with a symmetrical bilateral transverse facial cleft (Figure 1). The junction between the normal and abnormal vermillion was well delineated. No other anomaly was

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found in other systems. She had a repair of the defect in three layers; a linear closure of the mucosa and orbicularis oris; the skin of the cheek was closed by Z-plasty.

Case 2

A 12 months old female was delivered with a wide mouth. At presentation she was drooling saliva from the angles of the mouth. There was no history of such abnormality in the family.

On examination, she was healthy looking. The main finding was a bilateral transverse facial cleft with no associated congenital malformation. The clefts were repaired in layers (mucosa, muscle and skin) with a straight line repair of the skin of the cheek.

Case 3

A 9 month old female was brought by her parents to the hospital with the complaint of wide mouth since birth. There was no family history of a similar condition or any congenital anomaly.

The finding on examination was a bilateral lateral facial cleft in a healthy looking child with no other abnormal findings. The defect was repaired in layers (mucosa, muscle and skin) but the skin of the cheek was closed by a straight line repair.

Case 4

A 2years old female presented with abnormally wide mouth since birth. She was drooling saliva and occasionally leaked food particles from the angles of the mouth. There was no family history of a similar condition or any congenital anomaly.

The finding on examination was a bilateral transverse facial cleft with no other abnormal findings. She had the defect repaired in layers with a straight line repair for the cheek skin.

Case 5

A 10 weeks old female presented with abnormally wide mouth since birth. The anomally was not present in the other siblings. There was no associated congenital anomaly and no family history of such anomaly or any other type of anomaly.

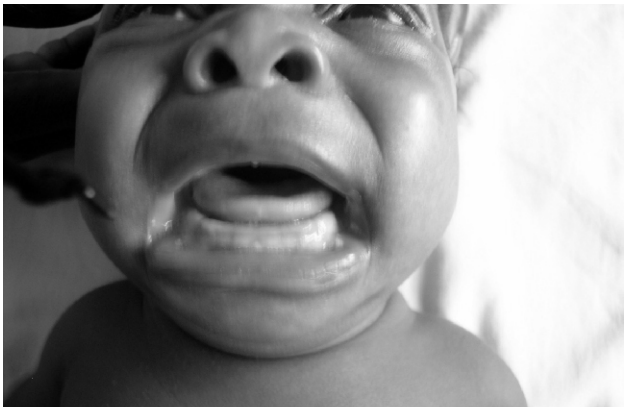


Fig. 2: Photograph of case 5 showing bilateral symmetrical macrostomia

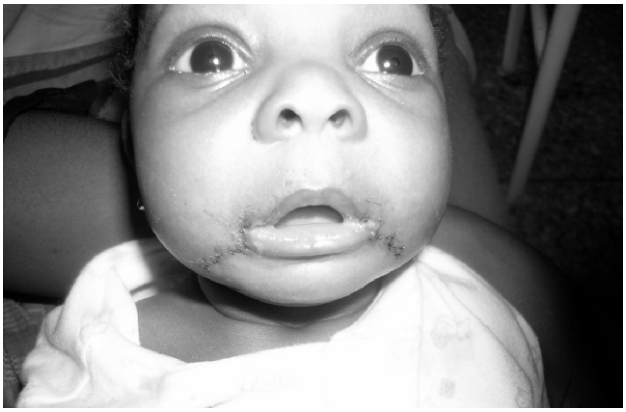


Fig. 3: Photograph of case 5 showing lazy W-plasty.

Finding on examination was bilateral symmetrical transverse facial cleft with the junction of the normal and abnormal vermillion clearly demarcated (figure 2). She had a repair of the defect in three layers; mucosal repair, myoplasty of the orbicularis oris and a lazy W-pasty of the skin (figure 3).

Discussion

Isolated bilateral macrostomia (lateral transverse facial cleft) is very rare and are usually symmetrical[3,4,13,14]. There is no distinct aetiological factor for macrostomia, although the condition is believed to represent a multifactor congenital anomaly. Stelnicki et al[15], in an animal model, demonstrated that lateral facial clefting can occur from constriction, restricting craniofacial growth from

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external forces such as a swallowed amniotic band. The cause of the constriction ring in man is yet unknown[16]. In a case report by Presti et al[7] a prenatal ultrasound diagnosis of a bilateral lateral facial cleft did not show clear evidence of an associated amniotic band. Although in almost all the cases reported, macrostomia was usually associated with other anomalies, none of the patients in the present report presented with other associated anomalies. The defect is believed to be due to a combination of direct tethering of normal tissue migration and an increase in local pressure, which produces cellular ischemia and apoptosis. The relationship between amniotic band and facial cleft has been reported by a number of authors[17-19].

Macrostomia is probably due to a "post zygotic mutation" with no factor to suggest inheritance. Hartsfield and colleague[14] reported a set of twin discordant for bilateral symmetrical; this suggests that macrostomia is probably due to a "post zygotic mutation" with no factor to suggest inheritance. None of the patients in the present report had any family history of congenital macostomia or any congenital anomalies.

Males are more frequently affected in most reports[2,6,13], although a report from France[4] showed equal gender affectation. These reports contrast with our series in which all the patients were females. The reason for these differences is unclear.

Macrostomia has been classified by a number of authors[4] based on different parameters which include clinical, embryological, state of the temperomandibular joint and the surrounding structures. One classification that has some therapeutic implication divides macrostomia into four types namely[4]:

Type I or Minor, in which the cleft terminate at the anterior border of the masseter.

In this type the repair include mucosal repair, repair of the orbicularis oris with the fibres in the vertical direction and the skin closed either by straight line or Z-plasty.

In type II or Major, the cleft extends beyond the anterior border of the masseter.

The muscles of the region (masseter and temporalis) are dissected with the fibres in the vertical direction in other to optimise their function for mastication. The orbicularis oris is reconstructed and a straight line repair of the skin.

Type III or Bilateral Minor is characterised by bilateral clefts that do not go beyond the anterior border of the masseter. The repair involves creation for a neocommissure using standard measurements.

In type IV or Bilateral Major, the clefts are bilateral and extend beyond the anterior borders of the masseter. The repair requires dissection of the regional muscle as in Type II, creation of neocommisures and linear repair of the skin.

Early repair is advocated in early childhood[9,13]. This is to allay the parent's anxiety and to prevent the problems of sialorrhoea, leaking of food/drink during and distortion of speech by maintaining the oral sphincter function. In patients who are treated at a later stage (school age children) there is the greater chance of developing psychological and social problem which in the long run will affect their performance in school. In our series all the patients had their repair in the preschool-age which will allow them develop like any normal child.

The aims of repairing a macrostomia are to restore the function of the orbicularis oris muscle, to maintain symmetry of the lips and to reconstruct the commisure so as to have a natural looking mouth with minimal scar formation[10,12,20]. The reconstruction of the oral commissure is an important part of the repair[9]. Mucosal flaps like triangular flap[6,11,12,20], square flap[9,21] and in some cases modified Estlander-type flap[22] have been used to reconstruct the oral commissure. These flaps placed at the angle of the mouth are used to prevent subsequent contracture and shortening of the oral fissure that will lead to the deviation of the commissure. The flaps are designed to cross the commissure which will prevent the lateral migration of

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the commissure from a linear scar contracture [11].

All these methods of reconstruction of the commissure have their shortcomings either alone or in combination with any form of skin closure. Determining the position of the neocommissure is very important for the purpose of creating commissures that are symmetrical. In unilateral lateral cleft one can rely on the contralateral normal side as a reference point, where measurements are taken and transposed on the cleft side before incisions are made. On the other hand in bilateral macrostomia there is no normal commissure to be used and so the surgeon must define the position without one. The repair therefore requires the aid of extra oral landmarks as well as normal measurements[3]. Standard measurements for different sexes and races are documented[4]. Accurate positioning can be determined by a careful inspection of the muscle and vermillion at the abnormal commissure[10,23]. The point of the neocommissure is at the junction of the normal and abnormal vermillion where the muscles (orbicularis oris) end and the white line thins out[9]. This landmark can be seen clearly in case 1 and 5 (fig, 1 and 2) and it is believed to be more reliable especially when using endotracheal rather than nasotracheal intubation for anaesthesia[2].

In macrostomia orbicularis oris muscle ring is discontinued and this has to be reconstructed[4,21,24] to produce the usual sphincter-like function, a function that is very important in feeding and speech[25]. Myoplasty, involving accurate approximation of the muscle during reconstruction is the key point in the repair of transverse facial cleft[5,12,21,24].

Various methods of closing the skin are described including the straight line repair[6,26], Z-plasty[5,23,27], W-plasty[27,28], lazy W-plasty[9] and the modified Estlander-type repair[22,29]. The main factor that influences the choice of a repair is the outcome of the scar. Eguchi et al[9] as well as Fukuda and Takeda[27] condemned Z-plasty for the cheek closure on the ground of poor cosmetic outcome of the scar despite meticulous suturing. Consequently, Eguchi et al[9] advocated the lazy W-

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Résumé

Arrière-plan: La macrostomie est une anomalie congénitale rare du visage, surtout le type isolé. Ce rapport se propose de rendre compte de notre expérience dans la gestion de cette pathologie rare. ***Malades et méthodes:*** Nous avons fait un examen rétrospectif de tous les cas consécutifs des enfants que nous avons soignés pour la macrostomie bilatérale isolée au centre hospitalo-universitaire de Jos. ***Résultats:*** Cinq malades, âgés de 10 semaines à 30 mois, ont été soignés. Les malades étaient toutes des fillettes atteintes d'une fissure faciale, bilatérale et symétriquement transversale et latérale (la macrostomie). Il n' y avait pas d'anomalies connexes, et aucun antécédent familial de fissures faciales ou de toute autre anomalie congénitale. Pour les guérir du défaut, nous avons opéré toutes les malades sous une anesthésie générale avec des résultats satisfaisants. ***Conclusion:*** La macrostomie peut se présenter comme une entité isolée. Le résultat final de la réparation dépend de la technique appliquée, du fonctionnement du muscle orbiculaire et de la qualité de la cicatrice. ***Mots-clés:*** Macrostomie; fissure faciale; néo commissure.

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