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Congenital macrostomia management in children in a country with limited resources: A case series

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1. Introduction

Congenital macrostomia is a rare facial malformation characterized by an excessively enlarged mouth at the labial commissures. It corresponds to type 7 of the classification of cranio-facial clefts according to Tessier [1]. In the literature, some previous cases of macrostomia have been reported particularly in America, France, Japan, India and Nigeria. Several surgical procedures have been described. We report here a series of three consecutive cases successfully managed in our center after thirty years of pediatric surgical practice. The authors advocate *Z*-commissuroplasty in a country with limited resources and provide an update on an epidemiological, diagnostic and therapeutical point of view. This case series has been reported in line with the updated PROCESS 2020 Guideline [2].

2. Presentation of cases

This study is a retrospective case series of congenital macrostomia in children. These consecutives cases have been managed by a junior surgeon in our teaching hospital center. Informed consent was always obtained prior to surgery.

2.1. Case 1

A 04-month-old female infant was admitted in consultation for a congenital mouth malformation. In her medical records, a positive HIV serology status was noticed. Clinical examination noted an enlarged mouth at the left labial commissure on two centimetres with a rotation of the lower lip. There were no further clinically visible malformations (Fig. 1A). The diagnosis of left unilateral congenital macrostomia was retained. Under general anaesthesia and orotracheal intubation, patient in supine position, we performed a Z-commissuroplasty. The different stages of the intervention were as follows:

- The outline of the incision has required four landmarks: normal labial commissures, the limits of the malformation, the nasolabial fold and the bitterness fold (Fig. 1B).
- A frank incision along the line involved the three layers of the deformity: cutaneous, muscular and mucous (Fig. 1C).
- The reconstruction of the mucosal plan was performed by resorbable thread knotted in the oral cavity. After dissection of the orbicularis muscle and vertical reorientation of its two edges, it was repaired at the level of the neolabial commissure by a simple suture according to the Kaplan technique. We used a slow resorption wire (Fig. 1D). A cutaneous plasty with *Z*-flap imbrication were done (Fig. 1E).

Analgesia and intravenous antibiotic (ceftriaxone 50 mg/kg per day) were administrated for three days. The postoperative period was uneventful. The infant was discharged from the hospital on the third postoperative day. After twelve months of follow up, we noted a fairly good

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symmetry of lips and labial commissures, slightly visible scars with no flanges (Fig. 1F).

2.2. Case 2

A 02-month-old female infant was seen in outpatient consultation for a congenital malformation of the mouth. Her clinical record showed no peculiarity. Examination of the cephalic extremity revealed an enlarged mouth transversally at the level of each labial commissure on 2.5 cm, without rotation (Fig. 2A). Any further associated anomalies were identified. The diagnosis of bilateral congenital macrostomia was made. A *Z*-commissuroplasty was performed under general anaesthesia and orotracheal intubation (Fig. 2B). Analgesia and an intravenous antibiotic (ceftriaxone 50 mg/kg per day) were administrated for five days. The postoperative course was uneventful. The infant was discharged from the hospital on the fifth post-operative day. Twelve months after surgery, good lips symmetry and labial commissures were observed. Good scars and no flanges were noticed (Fig. 2C) and an acceptable mouth opening (Fig. 2D).

2.3. Case 3

A 30-month-old female infant was admitted in our unit for a congenital malformation of the mouth. Her clinical record showed no peculiarity. On examination, an enlarged mouth was noted at the level of the left labial commissure on 2 cm and a deviation of the jaw to the left. No other clinically visible malformations were observed (Fig. 3A). The diagnosis of left unilateral congenital macrostomia was made. A *Z*-commissuroplasty has been performed under general anaesthesia and nasotracheal intubation. The immediate postoperative appearance with reconstituted left labial commissure and operative Z scar was showed (Fig. 3B). Analgesia and an intravenous antibiotic (ceftriaxone 50 mg/kg per day) were administrated for four days. The postoperative period was uneventful. The infant was discharged from the hospital on the fourth post-operative day. At twelve months of follow up, there has been an acceptable symmetry of the lips and labial commissures with no migration of the neo commissure (Fig. 3C), with a slightly visible scar without flange (Fig. 3D) and an acceptable mouth opening.

3. Discussion

Epidemiologically, congenital macrostomia is one of the so-called rare cranio-facial clefts as opposed to labio-alveolar-palatal clefts [3]. It represents 0.3 to 1% of craniofacial clefts [4]. Its overall incidence was estimated at 1 in 60,000 to 1 in 300,000 live births [5]. In Nigeria, the most important series of literature has been reported with thirteen cases [6]. In Togo, we report the first series of three cases after thirty years of pediatric surgical practice. The diagnosis of congenital macrostomia is clinical. Only 10% to 20% of patients with macrostomia have bilateral involvement [3]. In our series, we recorded two unilateral cases on the left and one bilateral case. Straight unilateral clefts are more frequent with a predominance of lower rotation and the male sex is the most found in the literature [5]. Patients were all female with two cases of unilateral clefts rather left with lower rotation. It can be isolated or syndromic hence requiring a complete malformative assessment [3]. We did not note any syndromic symptoms in our series.

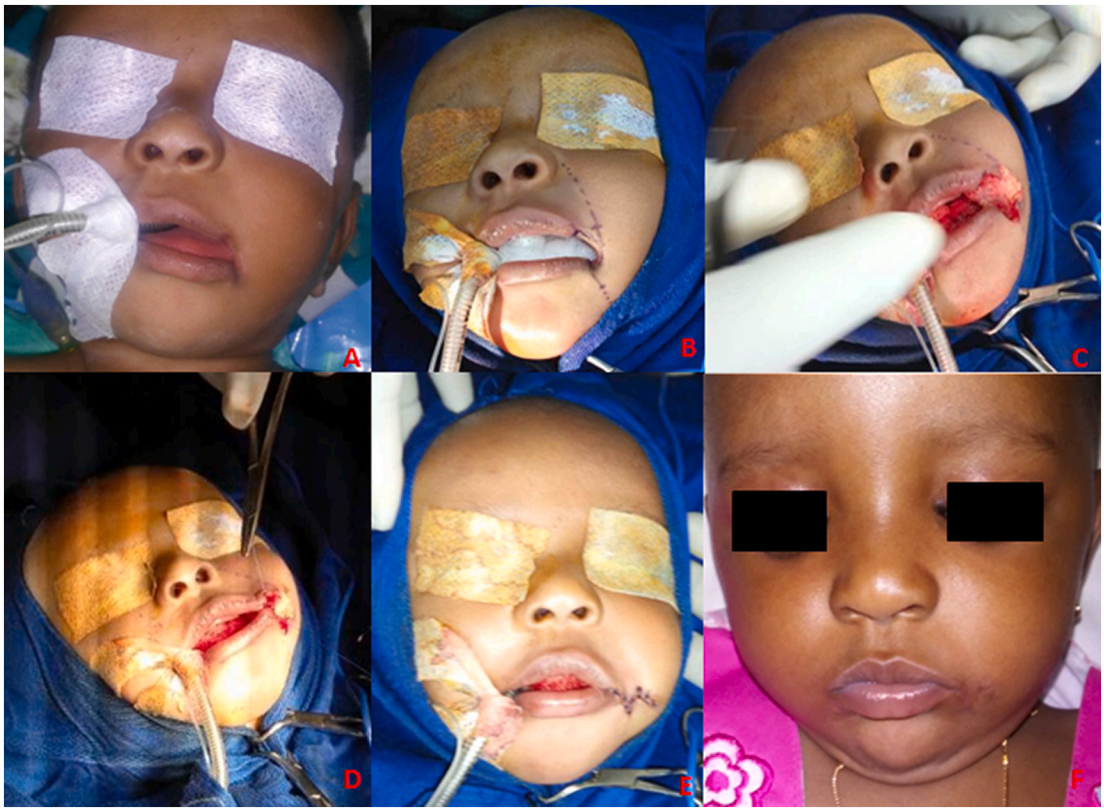

Fig. 1. Left unilateral macrostomia.
A. Preoperative view.
B. Outline of the incision requiring 4 landmarks: normal labial commissures, the limits of the malformation, the nasolabial fold and the bitterness fold.
C. Incision along the line involved the three layers of the deformity: cutaneous, muscular and mucous.
D. Orbicularis muscles repair using simple suture according to the Kaplan technique
E. Cutaneous plasty with *Z*-flap imbrication.
F. Good symmetry of lips and labial commissures, slightly visible scars with no flanges.

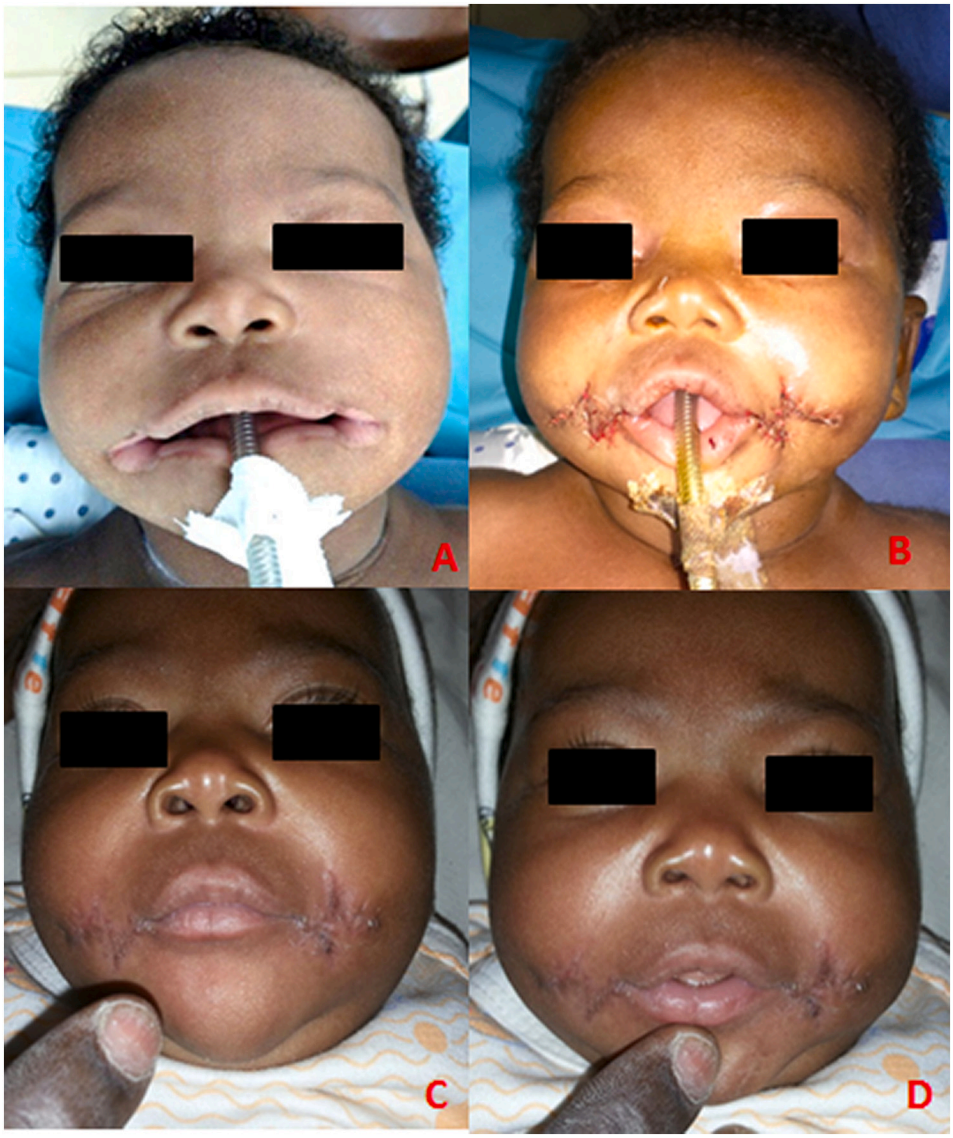

Fig. 2. Bilateral macrostomia.
A. Preoperative view.
B. Z commissuroplasty.
C. Good symmetry of lips and labial commissures.
D. Slightly visible scars.

According to all the techniques described in the literature, the objectives of macrostomia commissuroplasty are: to reconstitute symmetrically the neocommissure, to restore the oral function by repairing the orbicularis muscle, to close the oral mucosa to obtain an outline normal, to prevent the lateral migration of the neocommissure and to obtain a scar not very visible [7]. The objectives of the repair that generated the most debate are the reconstruction of the commissure and the closing of the skin with a scar not very visible. Several commissuroplasty techniques have been developed with various advantages and disadvantages. They concern the cutaneous level: simple linear suture [7], W plasty and Z plasty [8] or the triangular double flap method [9]. On the muscular level, Kaplan offers a simple suture edge to edge of the ends of the orbicularis muscle of the lips; in contrast to Skoog which suggests a simple suture with entanglement of the ends of the orbicularis muscle of the lips with adequate tension at the level of the commissure [3]. On the mucous level, a simple suture with resorbable thread with knots tied in the oral cavity is suggested [3]. In our series, the mucosa was reconstructed by resorbable thread knotted in the oral cavity. The orbicularis muscle was repaired by simple suture at the neocommissure using the Kaplan technique. The skin plan has been reconstructed by Z-plasty. Proponents of Z and W-plasties conclude that skin contracture and lateral commissure migration are minimized with their techniques [8]. On the other hand, proponents of linear suture of the skin claim to obtain a more aesthetic appearance of the postoperative scar without lateral migration of the commissure [7]. A repaired orbicular muscle with adequate tension would provide a medial dynamic counterforce to lateral displacement resulting from contractile force [10]. In the short and middle courses, we obtained a rapid, non-hypertrophic scarring without lateral migration of the commissure. Our aesthetic results appreciated by the symmetry of the commissures and the lips, the size and the visibility of the operative scar seem to be acceptable. The functional results of our series evaluated postoperatively by the degree of mouth opening, the fasciculation of the skin during mimicry, seem satisfactory.

4. Conclusion

Macrostomia is an uncommon birth defect. Various surgical procedures have been proposed for its repair. However, the choice of the surgical procedure should be based on functional, aesthetic results and the surgeon's experience. Even in underdeveloped countries, Z-commissuroplasty could be performed with good results. Further research should focus on long-term follow-up, regardless of the type of surgical procedure, with long series.

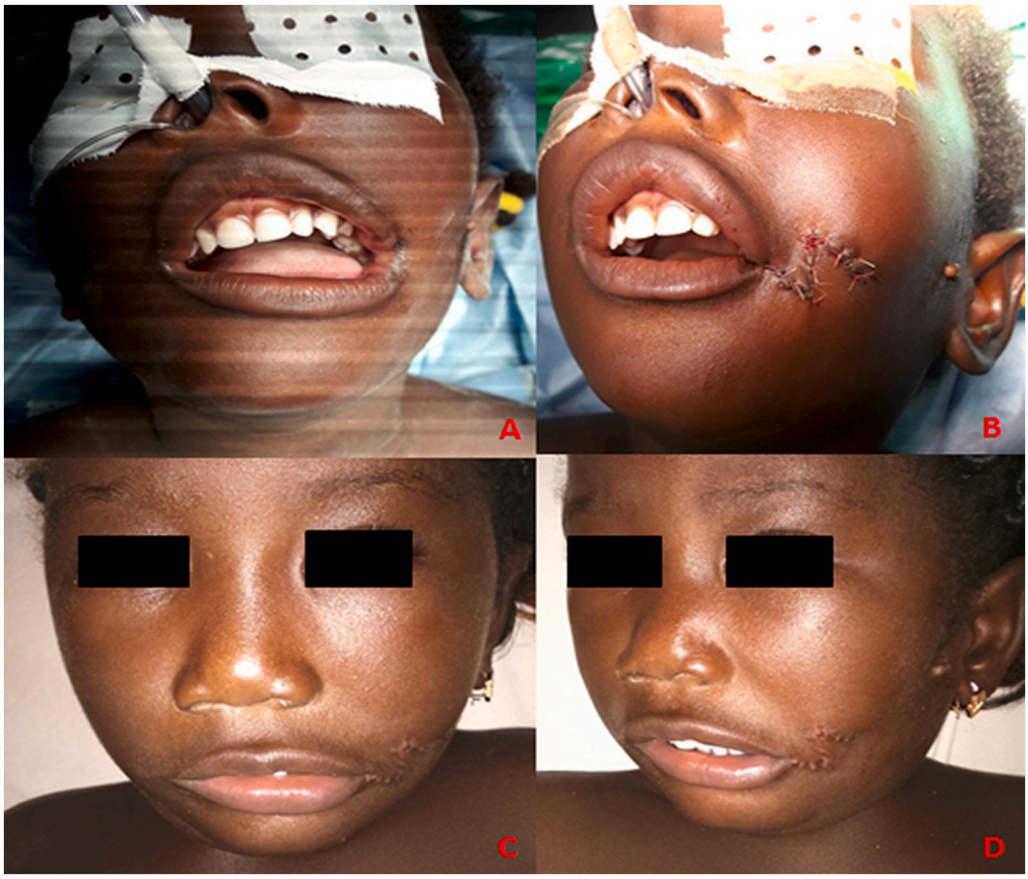

Fig. 3. Left unilateral macrostomia.
A. Preoperative view.
B. Z – commissuroplasty.
C. Good symmetry of lips and labial commissures.
D. Slightly visible scars.

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Ethical approval

Ethical approval has been exempted by my institution for reporting this case.

Consent

Clinical figures were taken in this manuscript after parents informed consent obtained. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor

Dr. Amoussou Sedjro Clotaire Romeo Houegban is the guarantor for the work.

CRediT authorship contribution statement

Eudes Ulrich Elvis Mahougnon Goudjo: Conceptualization, Methodology, Writing – original draft. **Octave Exupère Désiré Dongmo Miaffo:** Conceptualization, Methodology. **Codjo Serge Metchihoungbe:** Methodology, Writing – original draft. **Amoussou Sedjro Clotaire Romeo Houegban:** Methodology, Writing – original draft, Writing – review & editing, Visualization. **Houenoukpo Koco:** Writing – original draft. **Okassate Sibabi Akpo:** Writing – original draft. **Tely Bailo Kante:** Writing – original draft. **Komlan Gnassingbe:** Supervision.

Declaration of competing interest

Authors have no conflicts of interest to declare.

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