

Original Research Article

Repair of Congenital Macrostomia with Inferiorly Based Vermillion Square Flap and Z Plasty – Case Series and Experience from Eastern India**Dr. Biswajit Mishra¹, Dr. N.C. Dash², Dr. Gayatri Mahali³, Dr. S.Roy⁴**¹Associate Professor, Department of Plastic Surgery, SCB Medical College, Cuttack, Odisha, India.²Senior Resident, Department of Plastic Surgery, SCB Medical College, Cuttack, Odisha, India.³MCh Resident, Department of Plastic Surgery, SCB Medical College, Cuttack, Odisha, India.⁴MCh Resident, Department of Plastic Surgery, SCB Medical College, Cuttack, Odisha, India.**Corresponding Author**

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Received: 03-03-2024 / Revised: 09-03-2024 / Accepted: 16-04-2024

ABSTRACT**Background**

Macrostomia is a rare congenital anomaly characterized by enlargement of mouth at oral commissure. It is also known as Tessier cleft 7. There are various techniques described for achieving optimal function and aesthetic results. We describe inferiorly based vermilion square flap with z plasty on the skin for correction of macrostomia.

Methods

A retrospective analysis of 7 patients with macrostomia operated over a period of last 9 years were included in the study. All operations were conducted by senior surgeon. Pre operative and post operative documentation was done by photography. Age at presentation, operative technique and outcome were analyzed. Parameters used for assessing outcome are position of commissure, symmetry, thickness of vermilion, scar result.

Results

In all the patients repaired with myoplasty and modified lower vermilion square flap, lip commissure was formed with satisfactory shape, position, thickness with no commissure contracture. One patient had hypertrophy scar.

Conclusion

Inferiorly based vermilion square flap with z plasty method produces satisfactory aesthetic and functional result in all patients.

Keywords: Congenital macrostomia, Cleft 7, Vermillion square Flap, scar.

INTRODUCTION

Macrostomia correction aims at normal positioning of neocommissure with minimal scar on face. Previously described techniques place the scar at angle of mouth which has theoretical risk of pain during mouth opening and contracture at angle of mouth. This was negated by vermilion square flap technique.

Kaplan^[1] in 1981 described upper lip vermilion technique to create neo commissure in which upper lip vermilion flap was transposed to lower lip. This results in a commissure with no suture line scar. Since the final scar was not at angle of mouth, theoretical chance of lateral migration of neo commissure was minimized. It also decreases the risk of contracture and pain

with movement. In Kaplan's method skin closure was done by Z plasty. Z plasty changes the direction of scar, there is less chance of scar contracture and stabilization of the commissure. However, scar at lower border in Kaplan's method has a tendency to become more noticeable over time because of the tension when mouth is open. So Eguchi^[2] has modified Kaplan's technique in which he described elevation of a vermilion square flap from the lower lip and final scar was at upper lip. Skin closure was done by W plasty. We tried to repair macrostomia by inferiorly based vermilion flap which will place the scar at upper lip and a skin closure was done by Z. Rationale of our technique is that it will prevent the downward pull on the scar and benefits of Z plasty will be utilized.

AIM AND OBJECTIVE

To retrospectively analyse the outcome of macrostomia repair by inferiorly based vermilion square flap technique and Z plasty.

MATERIALS & METHODS

The patients for the present study comprised of 7 patients of congenital macrostomia who were admitted and operated in our Department from July 2016 to March 2023. They were classified according to whether they have isolated macrostomia or macrostomia with other deformities. All the patients were operated by a single surgeon using inferiorly based vermilion square flap technique with Z plasty. Preoperative and post-operative outcomes were analysed by clinical photograph. Written informed consent was obtained from parents/guardians of subjects before enrolment in to the study. The study was approved by the research and ethical committee of the institution.

Inclusion Criteria

1. All patients with congenital macrostomia.
2. No contraindication for general anaesthesia.

Exclusion Criteria

Associated cardiac anomalies, malnutrition, anaemia, or other conditions contraindicating for General Anaesthesia.

Surgical Technique

Patients were positioned in supine position. Surgeries were carried out under general anaesthesia through endotracheal intubation. To obtain symmetry of lip tube was placed in midline of lower lip. Oral commissure on the normal side was marked A. Oral commissure on the cleft side was marked B. Peak of Cupids Bow were marked C1 (normal side) and C2 (contralateral side). Midpoint of lower lip was marked as D. The distance from A to C1 was calculated ($= X$). Same distance was transposed from point C2 towards cleft side. The position of neo commissure was marked as B1. ($C2$ to $B1 = X$). Distance from D to A was calculated ($= Y$). The same distance was transferred to cleft side on lower lip (B2). It corresponds to a point in where there is transition from normal vermilion to abnormal vermilion and where the white line thins. Vermillion square flap three components. Two horizontal limb and one vertical limb. Line joining points B2 and B3 forms lower component horizontal limb, line joining B3 and B4 forms the vertical component of square flap. Points joining B4 and B5 forms horizontal component of the square flap in lower lip. Point B6, B7, B8 were marked on the upper lip so that line joining B7 and B8 forms the lower horizontal limb. Line joining B1 and B6 forms the upper horizontal limb of the square flap. The line joining B1 and B7 forms the vertical component of square flap in upper lip. (Figure 1A, 1B, 1C). In patients with bilateral macrostomia, position of new commissure was determined by asking the patient to look straight

ahead and a vertical line was dropped from medial side of sclera corneal junction (limbus). (Figure 2) A 3 mm × 3 mm mucocutaneous vermilion flap was designed in all the cases. Local infiltration with 2% xylocaine and adrenaline in 1:200000 concentration was given at the incision line in dose of 7 mg per kg body weight. Skin and mucosa were dissected to expose muscle. Excess tissue of buccal mucosa was excised and sutured in linear fashion using absorbable suture. The orbicularis oris upper and lower bundles were identified and sutured in overlapping fashion using absorbable suture (polyglactin 910) 5-0 with superior muscle bundles placed anterior to inferior muscle bundle using horizontal mattress suture as suggested by Eguchi (Figure 3). Skin was closed with z plasty using 6-0 undyed polyglactin 910 suture (Figure 4). After surgery wound was covered using neosporin ointment. Patients was discharged on third post-operative day.

Figure 1

Schematic images for correction of macrostomia

Oral commissure on the normal side was marked A. Oral commissure on the cleft side was marked B. Peak of Cupids Bow were marked C1 (normal side) and C2 (contralateral side). Midpoint of lower lip was marked as D. The distance from A to C1 was calculated (= X). Same distance was transposed from point C2 towards cleft side. The position of neo commissure was marked as B1. (C2 to B1 =X). Distance from D to A was calculated (= Y). The same distance was transferred to cleft side on lower lip (B2). Line joining points B2 and B3 forms lower component horizontal limb, line joining B3 and B4 forms the vertical component of square flap. Line joining B4 and B5 forms horizontal component of the square flap in lower lip. Point B6, B7, B8 were marked on the upper lip so that line joining B7 and B8 forms the lower horizontal limb. Line joining B1 and B6 forms the upper horizontal limb of the square flap. The line joining B1 and B7 forms the vertical component of square flap in upper lip.

RESULT

Total of 7 patients were included in this study with male to female patient ratio of 4:3. Age ranged from 4 months to 14 years. In all patients commisuroplasty was done by vermilion square flap technique and skin closure was done with Z plasty. All patients had satisfactory shape, position and thickness with no commissure contracture. One patient has scar hypertrophy. -This patient had familial history of keloid formation. It was managed by intralesional injection of triamcinolone acetate. Z plasty had produced more favourable scar. None of the patients had suffered from commissure contracture during follow up period. There was no feeding problem. Mouth opening was satisfactory and lip seal was adequate for all patients.

Sl. No	Age	Sex	Unilateral / Bilateral	Associated anomalies	Complications
1	5 yr	M	Bilateral	No	Nil
2	6yr	M	Unilateral	Epibulbar Dermoid	Nil
3	8 yr	F	Bilateral	Preauricular Skin Tag	Nil
4	14 Yr	F	Unilateral	Preauricular Skin Tag	Scar Hypertrophy
5	3 Yr	F	Bilateral	No	Nil
6	6 months	M	Bilateral	No	Nil
7	5 months	F	Unilateral	Skin Tag	Nil

Table 1: Demographic data

Case no. 6 – A 6-month-old male patient presented with enlargement of mouth opening on both the sides (Figure 5a). CT scan with 3 D reconstruction was done to rule out any bony

abnormality, which did not reveal any abnormality. Two week follow up of the patient revealed healed scar without any discharge or wound dehiscence. (Figure 5b)

Case no. 7

A 5-month female presented to OPD with skin tag and abnormal mouth opening. CT scan with 3D reconstruction was done to rule out any bony abnormality. Patient was operated by the same technique .18 month follow up of the patient revealed good outcome without any commissural migration. Commissure contour was maintained. (Figure 6A, Figure 6B)

Case no. 1

5 year old male presented with enlarged mouth opening on both the sides. Fig 7a represents the patient with mouth closed. Figure7 b represents the patient with mouth open. Figure 7c represents immediate post op result.

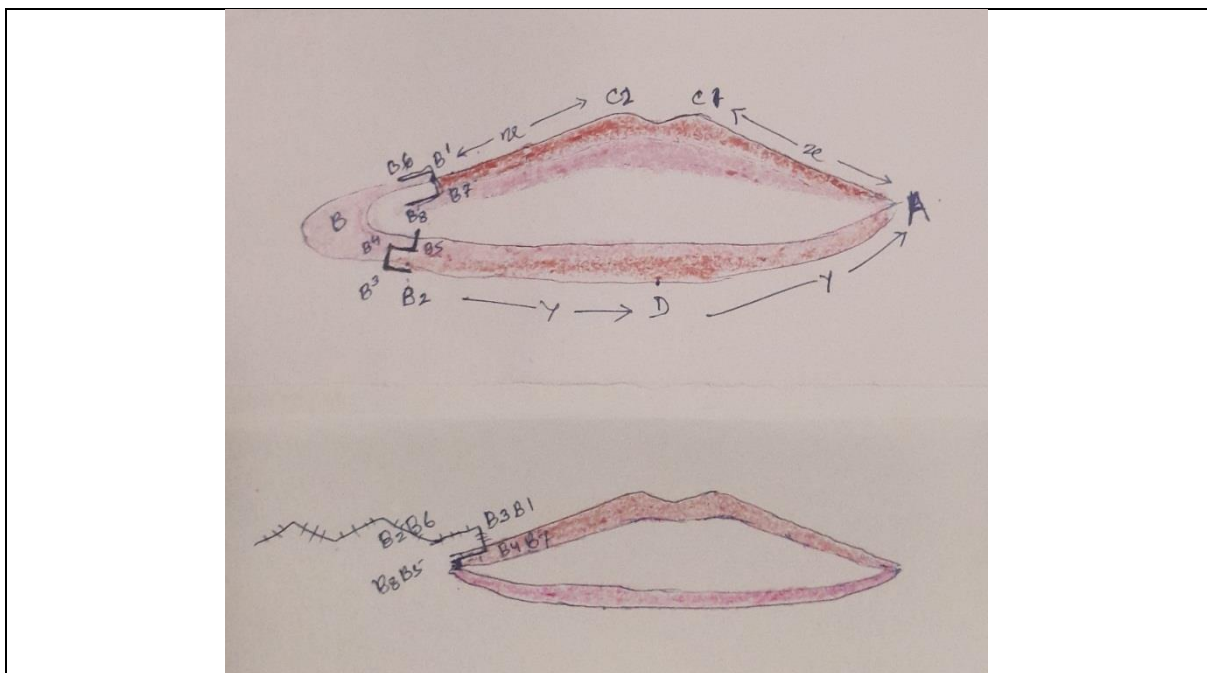


Figure 1 a: schematic diagramme

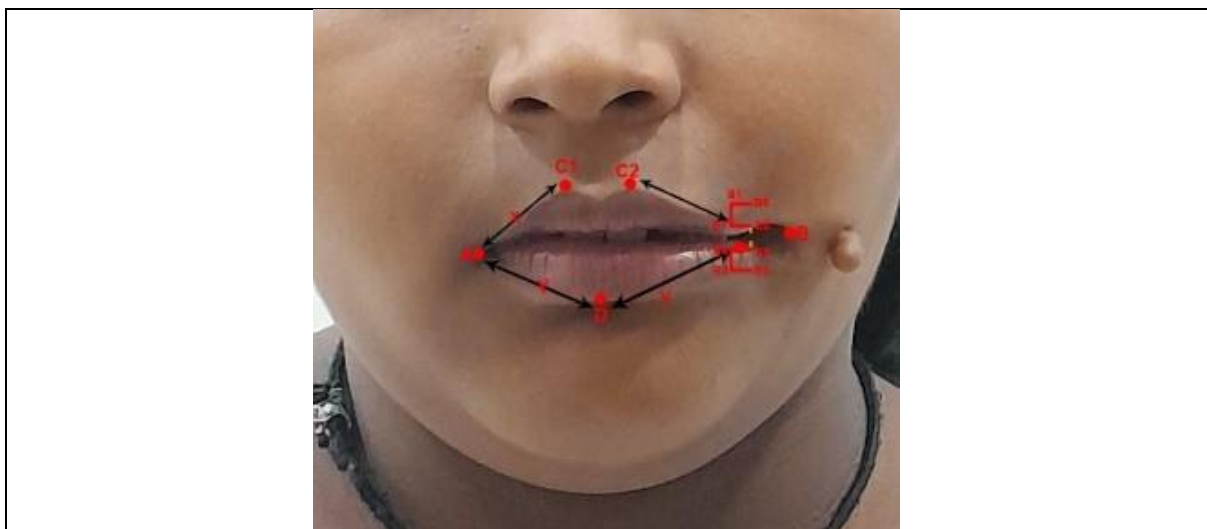


Figure 1b: Markings on a left sided macrostomia



Figure 1c: Proposed final marking



Figure 2: Marking in bilateral macrostomia



Figure 3: Myoplasty



Figure 4: Z Plasty closure



Type 5a: pre op picture of bilateral macrostomia



Figure 5b: post op result of macrostomia



Figure 6a: Isolated unilateral macrostomia

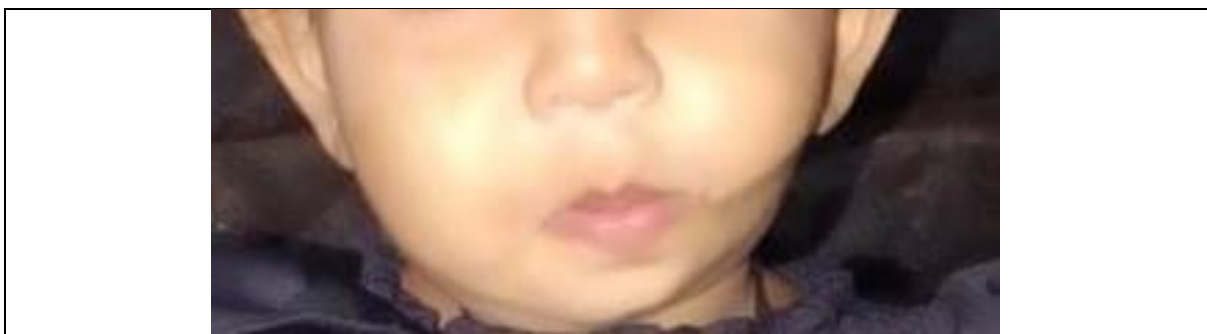


Figure 6b: Post op result macrostomia



Figure 7a: Bilateral macrostomia



Figure 7b: Abnormal vermillion of an isolated bilateral macrostomia



Figure 7c: Immediate post op result

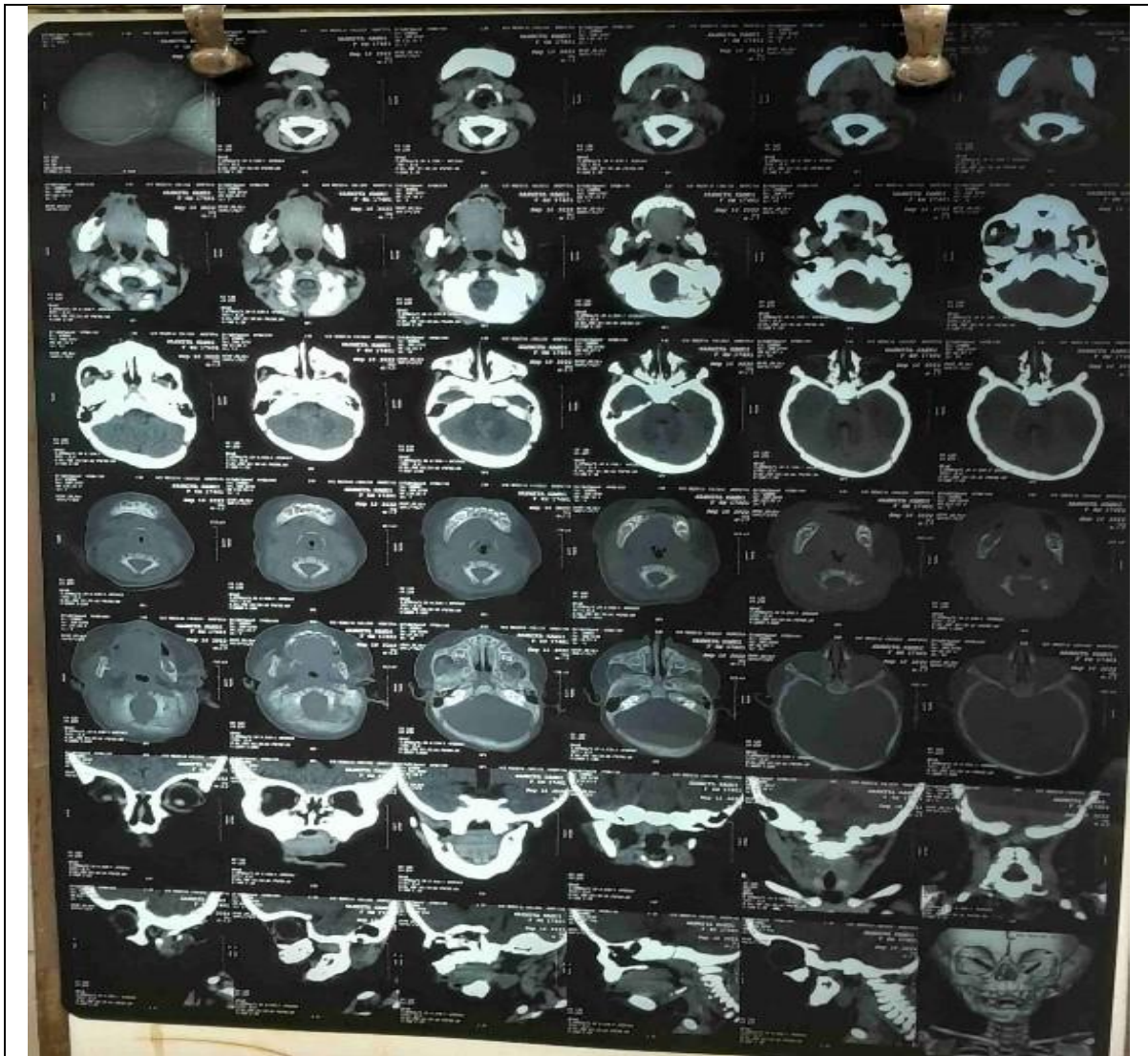


Figure 8: Normal CT scan of isolated bilateral macrostomia

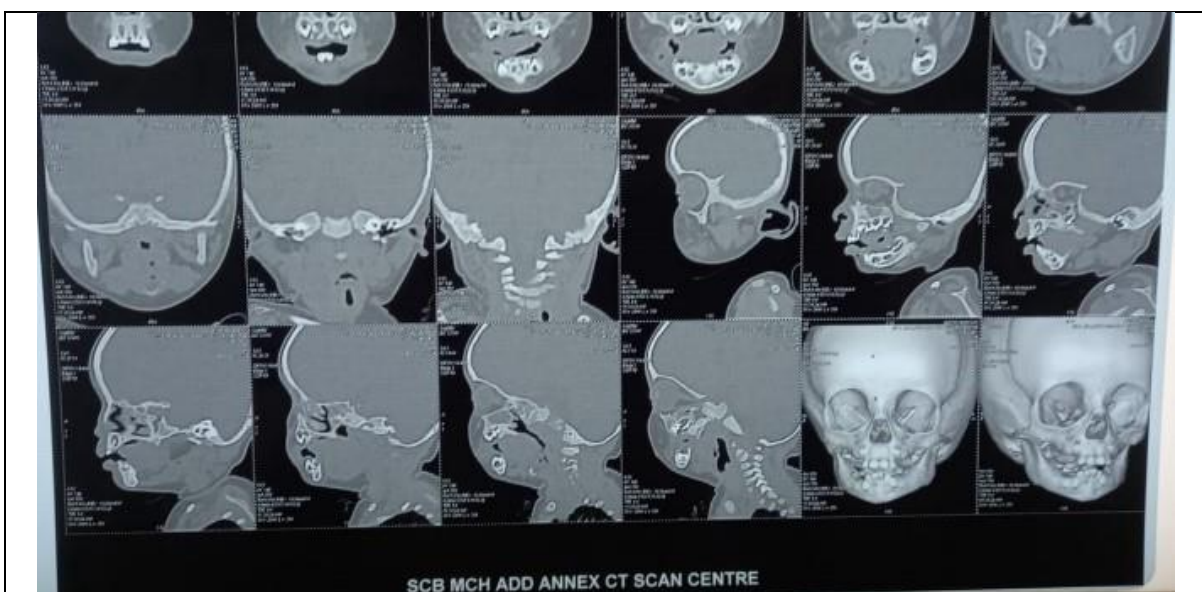
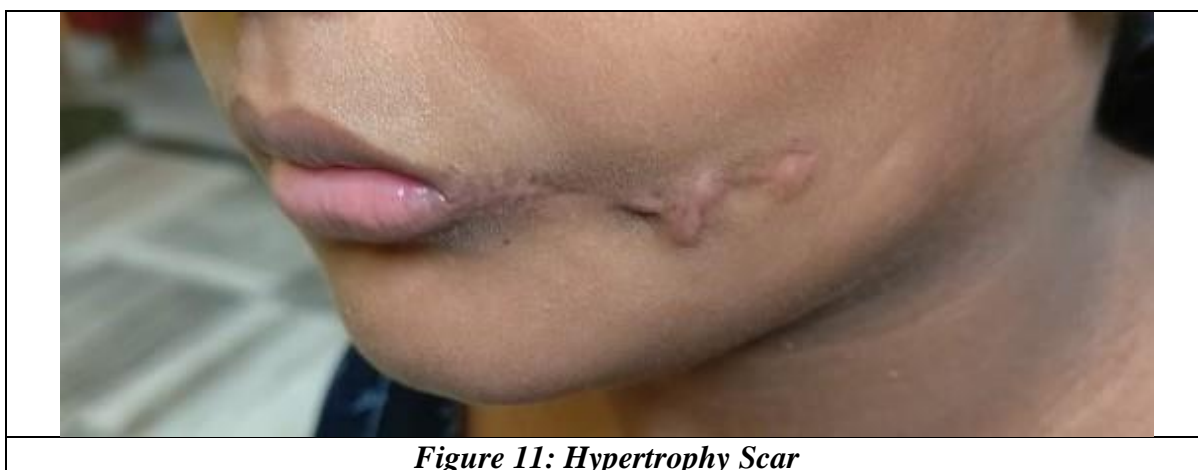


Figure 9: Normal CT Scan of Isolated Macrostomia



DISCUSSION

Macrostomia is a rare congenital anomaly. It is called Tessier cleft 7, transverse facial cleft. It is defined as enlargement of mouth at oral commissure.^[3,4] The incidence is 1 in 60,000 to 1 in 3 lakh live birth.^[5,6] It may occur unilateral or bilateral. Unilateral involvement is more common than bilateral. They are bilateral in 10%–20% of cases.^[5,6] In our series bilateral involvement is found in 57.1 % cases. This can be attributed to the fact that being a referral centre unilateral cases might have been operated by others. Since bilateral cases require more technical expertise more no of such cases are referred here.

Bilateral macrostomia can be syndromic or non syndromic. Syndromic bilateral macrostomia is usually associated with deformities of other structures developed from the first and second branchial arches. It can be associated with maxillary cleft and duplication.^[4] Among bilateral variety, syndromic variety is more common. All over world only 42 cases of nonsyndromic isolated bilateral facial clefts have been reported till date.^[7] There are very few cases of isolated bilateral macrostomia reported from India. Most of the cases are from Caucasian ethnicity.^[8] Most of literature are case reports only.

We have four cases with bilateral involvement. Out of four bilateral cases three had presented as isolated non syndromic variety. One case had bilateral preauricular skin tag. In rest of the three cases there was no preauricular skin tag. There was no asymmetry of face.

Computed tomography of the head and face with three-dimensional (3D) reconstruction was done in two of our cases which revealed normal maxilla and mandible with no bony clefts [Figure 8, 9], with involvement of skin and soft tissue including the orbicularis oris bilaterally. We have observed that degree of involvement was not symmetrical on both the side of the face. None of the previous studies have highlighted regarding this feature.

It is widely accepted that macrostomia should be corrected as early as possible.^[9] Stark and Sanders^[10] reported neonatal repair of macrostomia by direct excision and layered closure. In our series earliest timing of repair was 5-month oldest patient was 14 years.

Numerous techniques have been described for commisuroplasty and skin closure. May,^[11] who was the first to describe macrostomia repair, performed it using the Estlander flap in 1962. In this technique full thickness vermilion was used for commisuroplasty. But this technique did not use layered closure. Later on, layered closure technique was developed for muscle and oral commissure closure.

Reconstruction in macrostomia principally involves closure in three layers. Inner mucosa layer, middle muscle layer and outer skin layer. Inner layer closure is straight forward. Excess of buccal mucosa is excised and closed in straight line. Muscle layer is closed in such a way that upper orbicularis oris muscle crosses over lower orbicularis oris muscle. Improper alignment can lead to formation of web formation.^[11]

In 1969 Boo-Chai^[12] proposed landmark for positioning of new commissure. It corresponds to the point of junction of normal and abnormal vermilion. At this point orbicularis oris muscle border the sides, there is inversion of a triangular portion of skin up to this point. Suturing of free ends of orbicularis oris at commissure was advised to prevent gold fish mouth appearance (Figure 10). Kaplan^[1] in 1981 described upper lip vermilion technique to create neo commissure in which upper lip vermilion flap was transposed to lower lip. This results in a commissure with no suture line scar. Since the final scar was not at angle of mouth, theoretical chance of lateral migration of neo commissure was minimized. It also decreases the risk of contracture and pain with movement. Skin closure was done by Z plasty.^[5,6,12,13,14] However, scar at lower border in Kaplan's method has a tendency to become more noticeable over time because of the tension when mouth is open. So Eguchi^[2] has modified Kaplan's technique in which he described elevation of a vermilion square flap from the lower lip. Skin closure was done by W plasty.

The aim of commisuroplasty is to locate the neo commissure at normal anatomic position and prevent lateral migration of commissure. The resultant scar should be aesthetically pleasing, there should be no scar contracture.

A disadvantage of most of the surgical techniques described for macrostomia repair is that the resultant scar tissue is perpendicular to the tension lines of the face and is therefore pronounced, and the oral commissure moves laterally due to scar contracture.^[15] Even though Z-plasty performed on the edge of the oral commissure allows appropriate localization of the commissure, it may not prevent lateral movement of the commissure.^[16,17] because final suture line was at the angle of mouth .

Lateral movement of the commissure can be prevented when a flap is transposed from the upper and lower lips to the commissure. In these methods final scar is not at the angle but either at upper lip or lower lip. It is also recommended that flaps should not be placed exactly on the commissure to obtain a natural appearance.

Skin can be closed by W plasty, Z plasty or straight line. W-plasty was first used by Bauer^[18] for treatment of macrostomia. He hypothesized that W plasty will provide better cosmetic results than Z-plasty in reconstruction of facial scars.

Initial workers used Z-plasty to close transverse facial cleft.^[18,19,20] Z plasty skin closure is advocated by most authors as this change's direction of scar, there is less chance of scar contracture and stabilization of the commissure. But later on, it was noticed that the Z-plasty

left a more visible scar. This drawback of Z-plasty led to development of straight-line closure.^[21] Straight line closure theoretically is prone to result in contracture and lateral displacement although actual studies had not proven this. Some studies have shown inconspicuous scar is formed after straight line closure.

Kawai et al.^[22] in 1998 described a technique to correct macrostomia with a simple straight-line incision and incorporation of a small triangular flap to achieve proper positioning of the commissure with minimal visible scar.

Yu et al described more favourable result will be obtained if medial limb of z plasty was made perpendicular to relaxed skin tension line.^[23]

We have used vermilion square flap for commisuroplasty. This is based on the observation that corner of the normal mouth is not a corner but rather a smooth and continuous segment of the vermilion.^[24] It is difficult to achieve a natural contour of the corner of the mouth when the scar is located at the angle.^[12] Vermilion flaps do not place the scar at the angle and it maintains the continuity of vermilion. We have used method proposed by Eguchi where an inferiorly based vermilion flap was created to form commissure and final position of scar is on upper lip vermilion instead on lower lip.

Our approach differs from Eguchi in that we have used z plasty for skin closure. Eguchi described W plasty. In our series Z plasty had acceptable result.

One of our patients had developed scar hypertrophy (Figure 11) which was managed by intralesional triamcinolone injection.

Our study highlights the need of awareness for correction of such deformities. Even with the awareness for early cleft surgery by various government and non-government organisation still people hesitate to report early as evident by one of our cases who presented at age of around 14 years.

LIMITATION OF THE STUDY

We have not compared Z plasty with straight line repair or with W plasty. It is a single centre data, multicentric double blinded trail may be done to prove superiority of Z plasty with W plasty or straight-line repair. But since it is a rare congenital anomaly, it may be difficult to do such type of study.

CONCLUSION

Bilateral macrostomia is a rare congenital anomaly which should be corrected early. Inferiorly based vermilion square flap with Z plasty is a valuable alternative for macrostomia repair. Still awareness is required among the public for early referral of the case.

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