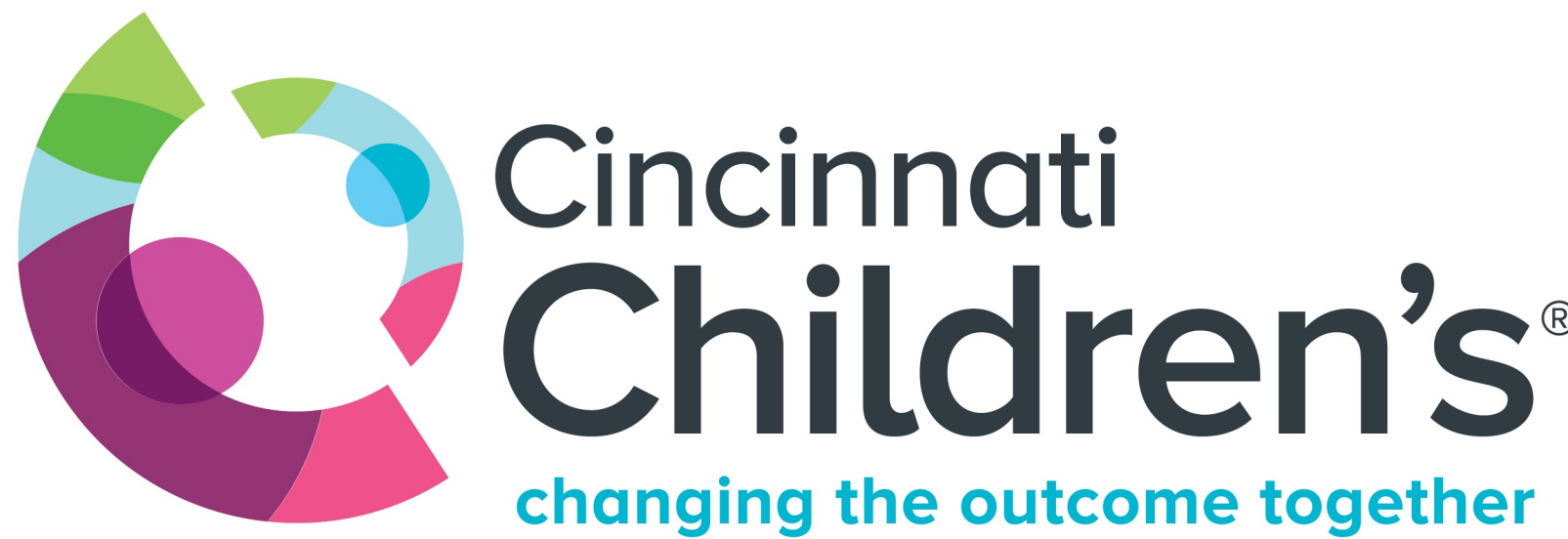


Surgical Correction of Median Cleft of the Mandible: A Case Report and Multidisciplinary Approach to Care

Nina O. Bredemeier^{1,2}, Mohamed Salim³, Omer Abdoun³, Angelo A. Leto Barone², Brian S. Pan^{1,2}, Raquel M. Ulma^{1,2}, Haithem Elhadi Babiker^{1,2}

¹Division of Plastic Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, OH USA, ²University of Cincinnati College of Medicine, Cincinnati, OH USA, ³Khartoum Dental Teaching Hospital, OMFS Department, Khartoum, Sudan



Introduction

Orofacial clefts are the most common congenital craniofacial malformation found in populations globally¹. Median mandibular cleft (also called Tessier 30 facial cleft) is one such anomaly with the first case reported by Couronne in 1819². In the 200 years since then, only 80 cases have been reported, lending to a lack of consensus on the presentation and treatment options for median mandibular cleft. A wide range of variations in the severity of this anomaly have been reported, ranging from simple cleft lip to cleft lip with defects including the mandible, strap muscles, hyoid bone, thyroid cartilage, tongue, and the floor of the mouth. There have also been cases reported with associated anomalies such as lower lip hemangioma, cleft palate, cervical dermoid cyst, cardiac malformations, aplasia of the epiglottis, and malformations of the laryngeal cartilage which can significantly compromise the airway, swallowing, and speech³.

Most cases of median mandibular cleft are discovered early on and require a series of surgeries throughout adolescence, including scar revision and orthognathic surgery following initial repair. These patients also require a long-term multidisciplinary team including, but not limited to orthodontists, speech therapists, ENT specialists, plastic surgeons, and psychiatrists. However, as most affected patients are from underserved populations with limited access to medical care in the Middle East and Asia, lack of resources and specialized professionals often serves as a limitation to care that needs to be addressed.

Case Presentation

Patient Presentation and History

A 30-month-old child presented with multiple facial defects associated with median mandibular cleft.

The chief complaints of the patient's parents were the poor aesthetics regarding her significant drooling and facial cleft.

The patient's past surgical history included ankyloglossia repair performed at 6 months of age by a general pediatric surgeon.

There was no familial history of similar conditions. The patient was delivered at full-term with no history of maternal illness or complications during pregnancy.

Physical Examination



Figure 1: Pre-operative picture showing bifid tongue.

Upon examination, a midline defect in the lower lip and a midline neck scar causing limitation in neck flexion was noted. Intraorally, a midline mandibular defect with completely separated and mobile bony segments was noted, as well as an anteriorly bifid tongue. All teeth were sound and, although malocclusion was present, the patient had competent lips with severe drooling of saliva (Figure 1).

Speech appeared to be grossly normal. After pediatric evaluation, the patient's heart was also assessed and cleared for anesthesia.

Following pre-operative clearance, the patient underwent one-stage surgical correction of the bifid tongue, median mandibular cleft, and hypoplastic strap muscles causing neck contracture.

Surgical Intervention

Following pre-operative clearance, the patient underwent a one-stage surgery with an "inside-out" order of surgical corrections performed, beginning with reconstruction of the floor of the oral cavity, followed by fixation of the mandible, and finally Z-plasty of the neck contracture.

Anesthesia

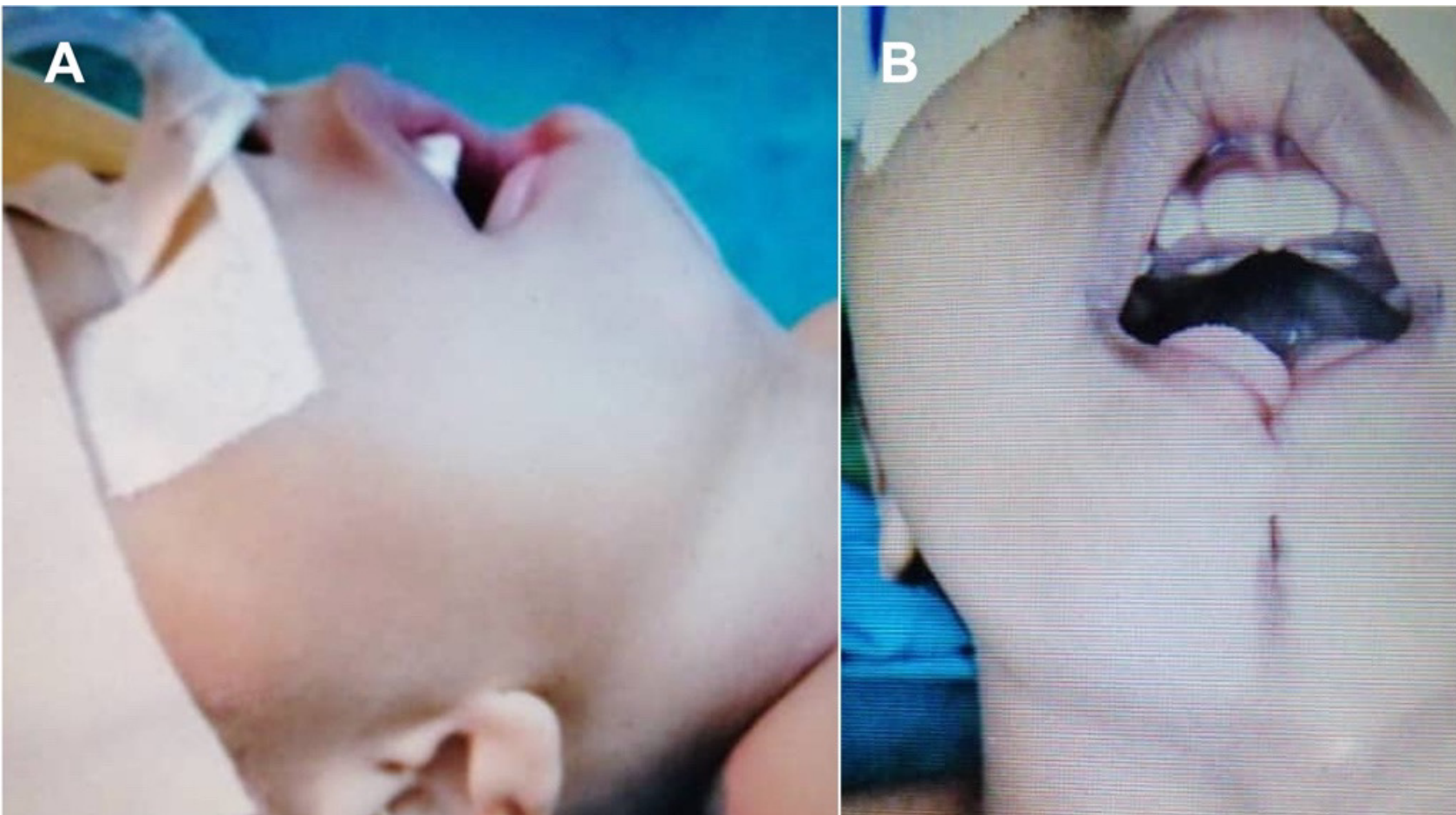


Figure 2 (A, B): Intra-operative pictures showing lateral profile view showing hypoplastic mandible, lip incompetence, and decreased thyromental distance (A), as well as patient's cleft mandible (B).

The patient was evaluated by an anesthesiologist, and despite mobility of the two mandibular segments and short thyromental distance, conventional nasal intubation of the patient was successful (Figure 2A). Nasal intubation of the patient also allowed for the patient's oral cavity to remain open and unobstructed for the surgeon to operate.

1. Reconstruction of Oral Cavity Floor

The midline cleft lip was incised, releasing three layers (orbicularis oris, skin, and mucosa). Subsequently, the bifid tongue was corrected by approximation of the two tongue halves in layers.

2. Semi-Rigid Fixation of the Mandible

The edges of the midline mandibular cleft were freshened and burred down to bleeding bone using surgical burs. A single titanium miniplate was then used for semi-rigid fixation of the split mandible (Figure 3). Due to the absence of proper occlusal guidance for reduction, the primary molars were used as a guide with a slight anterior open bite at the end of fixation.



Figure 3: Intra-operative view showing single miniplate fixating the mobile mandible halves.

3. Z-Plasty of Neck Contracture

The midline of the neck was incised utilizing a double-opposing Z-plasty technique. After careful dissection, fibrous tissue was removed, breaking the linear scar. Complete absence of the hyoid bone was noted. Closure of all incisions was completed in a layered fashion.

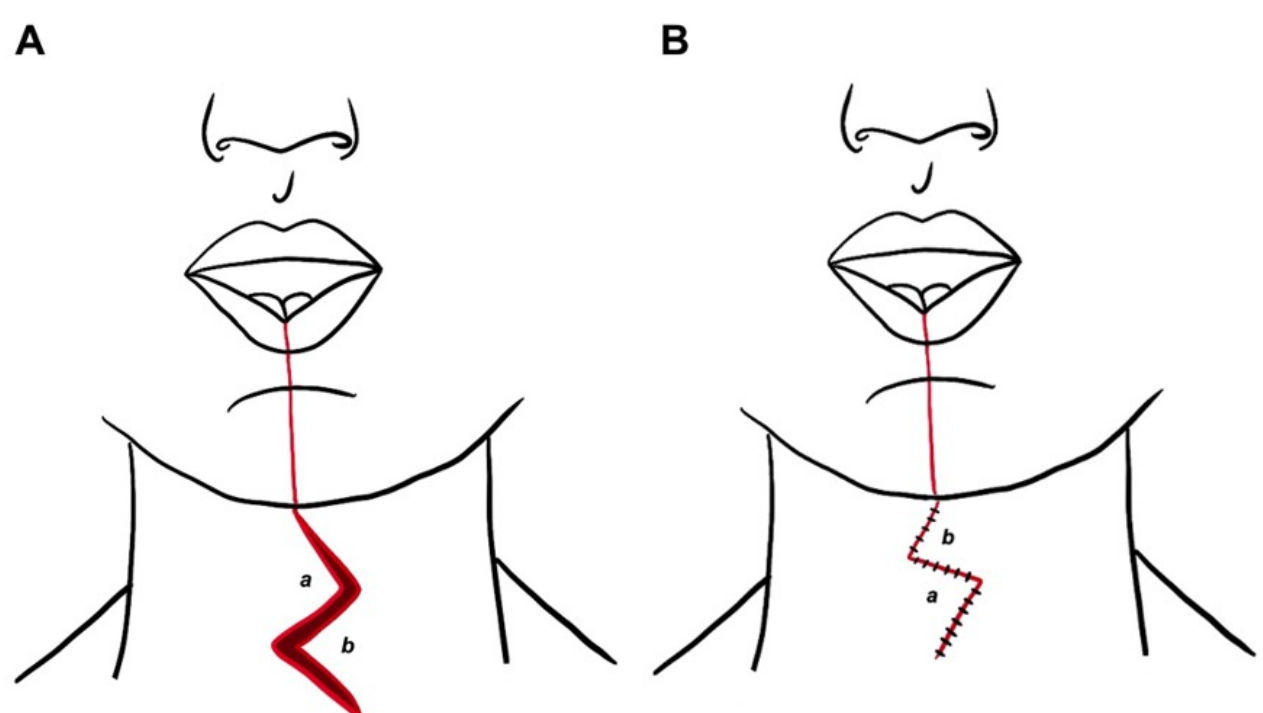


Figure 4 (A,B): Illustration demonstrating basic technique of the double-opposing Z-plasty technique. Red lines indicate sites of incision and creation of skin flaps a and b (A), and the later closure of incisions following the opposition of skin flaps (B).

Surgical Follow-up

The patient was seen again at 15 months after the initial surgery, showing minimal facial scarring, misaligned anterior teeth, and improved neck flexion (Figure 5A, 5B, 5C). The mandibular miniplate was removed 15 months post-operatively (Figure 6A, 6B), and the patient was seen again 2 weeks after miniplate removal, showing minimal scarring with healing of the skin and lower lip (Figure 7).



Figure 5 (A, B, C): 15-month post-operative appearance of the lower lip (A), misaligned anterior mandibular teeth (B), and neck scar after Z-plasty (C).

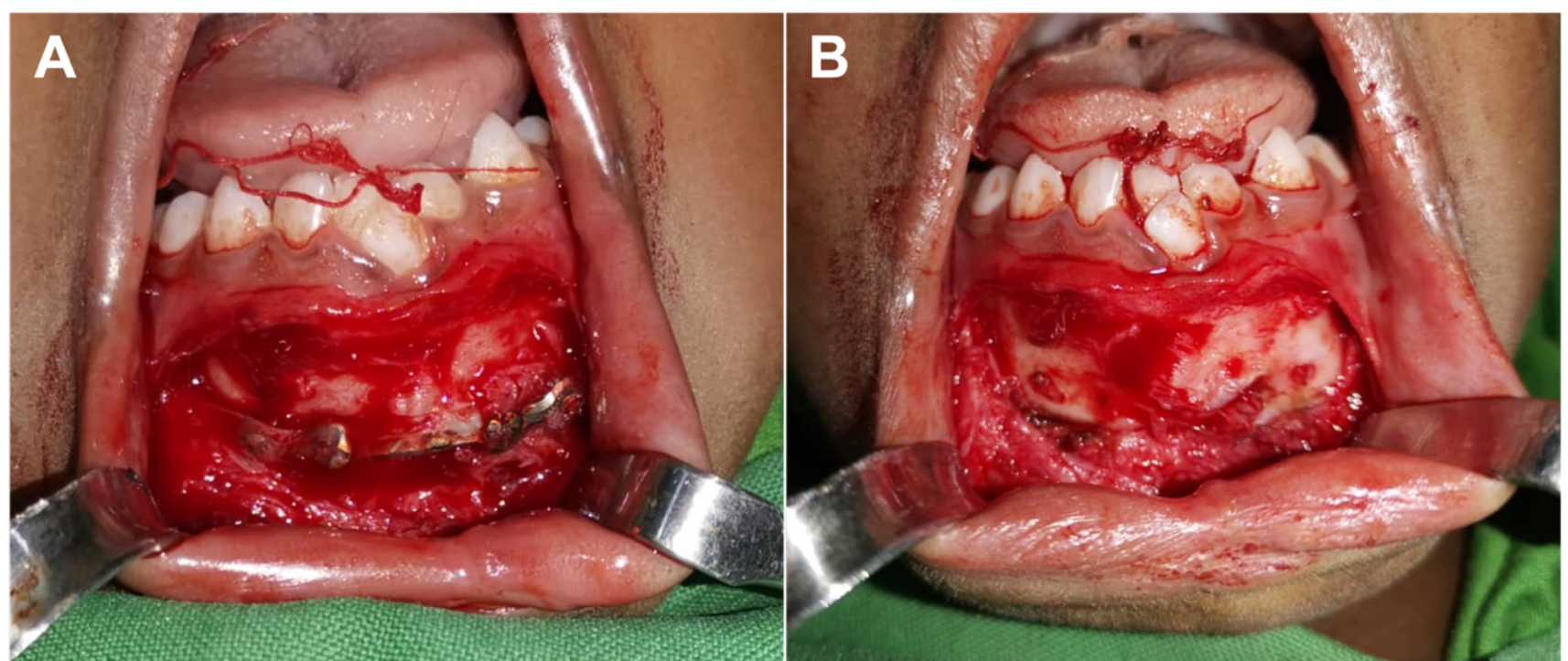


Figure 6 (A, B): At the time of removal of single miniplate, showing bony union of the mandible.

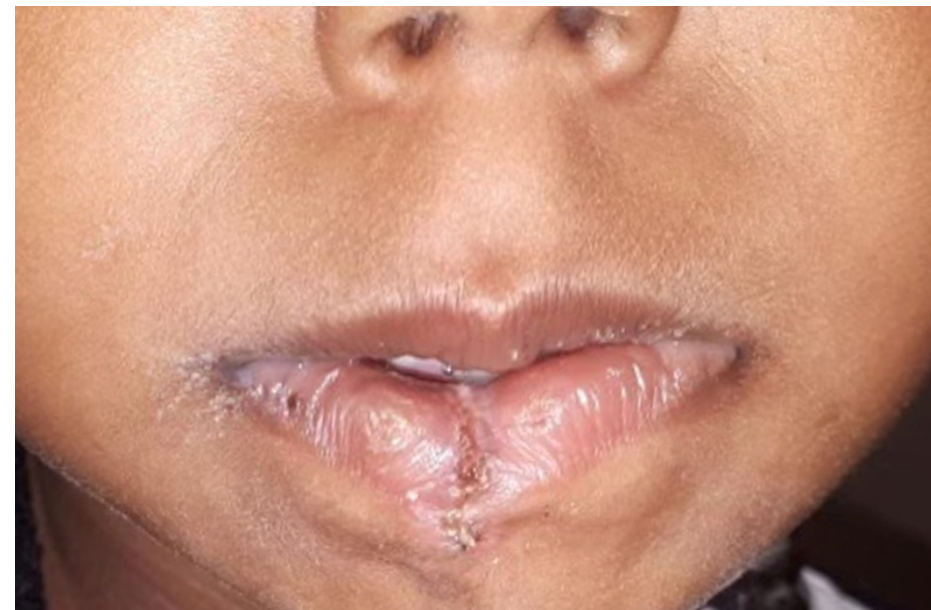


Figure 7: 16-months post-operative appearance of the lower lip.

Multidisciplinary Care

Most patients with median mandibular cleft require a series of surgeries and long-term follow-up with a multi-disciplinary team to ensure holistic care for optimal outcomes.

Plastic Surgery

- Perform necessary variety of surgical interventions to improve patient quality of life

Orthodontics and Dentistry

- Surgical interventions to optimize teeth and jaw alignment, dental health

ENT

- Monitor and perform surgeries pertaining to the ears, nose, and throat

Speech Language Therapy

- Assist in speech by improving articulation and resonance, reducing sound errors

Psychology/Psychiatry

- Support patients and families in dealing with societal stressors due to appearance

Genetics and Obstetrics

- Counsel patient and family on family planning, prenatal screening/care

Pulmonology and Sleep Medicine

- Assist in airway maintenance in patients with breathing issues

Feeding Specialists

- Assist in individualized feeding methods and optimize nutrition for growth

Discussion

One-stage vs. Two Stage Surgical Correction

- One-stage correction was chosen in this patient as she had a complete cleft with mobile mandibular segments⁴.
- Resource limitations in Sudan also indicated for one-stage correction due to lack of surgical specialists permanently practicing in the area. A one-stage surgery also reduced costs and burden of care as there is a high risk of patient loss to follow-up due to transportation barriers of patients.

"Inside-Out" Method of Surgical Correction

- The proposed method of one-stage surgical correction of median mandibular cleft and associated defects is to correct each defect in an "inside-out" order in surgery. First, the tongue must be released in the case of ankyloglossia. Next, the floor of the mouth is reconstructed. Then, the mandible is fixated in a semi-rigid manner. Finally, the neck contracture is corrected via Z-plasty technique.

Impact on Dentition

- The long-term impact of the miniplate hardware on permanent mandibular dentition will only be known with time. This patient demonstrated misaligned anterior mandibular teeth; however, they are not critical as they are of primary dentition.

Follow-Up Care

- Patients require a multidisciplinary team throughout their adolescence to assist in proper development in feeding, breathing, sleep, language, and mental health.
- Follow-up corrective jaw surgery in later adolescence is necessary to realign the jaw and teeth.

Geographic Distribution of Patients and Limitations to Care

- The majority of cleft patients are from low and middle-income global areas due to lack of proper nutrition and prenatal care, such as in parts of the Middle East and Asia⁵. These areas also lack specialists, facilities, and supplies, as well as limited patient transportation.
- This emphasizes the need for more awareness and funding for mission trips to train healthcare providers in under-resourced areas, as well as awareness as to how vital an investment in having a multidisciplinary team would be to patient care in these areas.

Conclusions

Median cleft mandible is a rare condition with a variety of presentations and associated defects ranging from mild to severe. Surgical interventions can be completed in one or two stages according to age, medical condition, specific anatomical findings, and socioeconomic conditions. To ensure best functional outcomes for the patients, they require a multidisciplinary team to provide holistic care in various aspects of their life. These patients will also often require multiple surgeries throughout adolescence as part of their follow-up care to ensure optimal mandibular function and form.

References

- 1-.Guttikonda LK, Nadella KR, Uppaluru V, Kodali RM, Nallamothu R. Nonsyndromic mandibular symphysis cleft. Case reports in dentistry. 2014;2014.
- 2-Monroe CW. Midline cleft of the lower lip, mandible and tongue with flexion contracture of the neck: case report and review of the literature. Plastic and reconstructive surgery. 1966 Oct 1;38(4):312-9.
- 3-Knowles CC, Littlewood AH, Bush PG. Incomplete median cleft of the lower lip and chin with complete cleft of the mandible. A preliminary report. British dental journal. 1969 Oct 7;127(7):337.
- 4-Almeida LE, Ulbrich L, Togni F. Mandible cleft: report of a case and review of the literature. Journal of oral and maxillofacial surgery. 2002 Jun 1;60(6):681-3.
- 5- Stewart, Barclay T., et al. "Estimate of Unmet Need for Cleft Lip and/or Palate Surgery in India." JAMA Facial Plastic Surgery, vol. 18, no. 5, 15 Sept. 2016, pp. 354–361., <https://doi.org/10.1001/jamafacial.2016.0474>.