DOI: 10.33470/2379-9536.1368

CASE REPORT Volume 8 Issue 2

Use of Mandibular Distraction Osteogenesis to Correct Micrognathia and Airway Obstruction in Newborn Female

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ABSTRACT

We present a case of Pierre Robin sequence and neonatal abstinence syndrome (NAS) in a newborn female patient to highlight the surgical technique of mandibular distraction osteogenesis to correct airway obstruction due to micrognathia. The patient presented as a transport after delivery due to respiratory distress. She had a cleft palate and micrognathia. The absence of other dysmorphic features diagnosed her with non-syndromic Pierre Robin sequence. Mandibular distraction osteogenesis was performed to solve her upper airway obstruction. This procedure allowed the patient to be weaned from all respiratory support and nasogastric tube feeds by the end of her hospitalization. She was able to be discharged home weeks before her internal hardware was surgically removed. Mandibular distraction osteogenesis was previously unavailable in rural Appalachia, making this case novel to the area. WV is disproportionately affected by the ongoing opioid epidemic in the United States. There is currently little literature exploring a potential association between substance abuse during pregnancy and PRS. Our case presents a possible avenue of new research into substance-induced birth anomalies.

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KEYWORDS
Pierre Robin sequence, micrognathia, glossoptosis, airway obstruction, mandibular distraction osteogenesis, neonatal abstinence syndrome

INTRODUCTION

Pierre Robin sequence (PRS) presents with micrognathia, glossoptosis, and upper airway obstruction at birth.[1] Due to the upper airway obstruction, infants born with PRS commonly experience respiratory distress and feeding difficulties in the immediate postnatal period. Up to 25% will need advanced airway placement such as endotracheal intubation or tracheostomy.[17] PRS is a relatively rare condition, occurring in 1/8,500 to 1/14,000 births.[1] This range is due to differing diagnostic criteria and the presence or absence of associated syndromes and anomalies.[2] The exact etiology of PRS remains unknown; however, several studies have pointed to a genetic basis for the condition. Up to 28% of patients with PRS have another family member with cleft lip or palate.[4]

PRS can also present along with several other genetic syndromes, such as Stickler syndrome, Velocardiofacial syndrome, Treacher Collins syndrome, and more. Prenatal diagnosis is possible through ultrasound with inferior facial angle and jaw index measurements, but these are not universally obtained.[5,6]

Presenting signs of airway obstruction in PRS are due to micrognathia. The mandible is hypoplastic in all dimensions, leading to a decreased retrolingual airspace as the tongue is displaced superiorly and posteriorly. The severity of airway obstruction is generally related to the size of the mandible; however, synchronous airway abnormalities, comorbid neurologic, and cardiopulmonary conditions may also affect the infant's respiration. Being able to assess the infant's respiratory status is crucial to preventing

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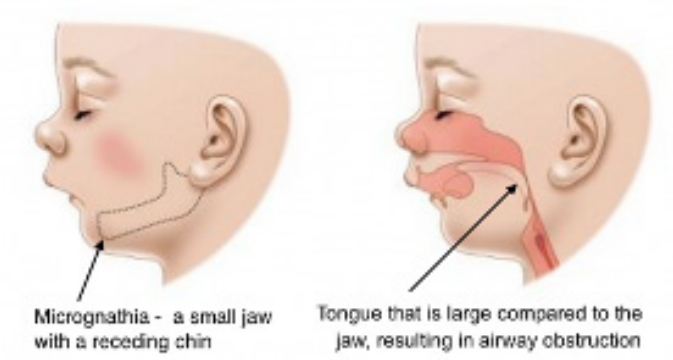

FIGURE 1. Physical manifestations of airway obstruction in Pierre Robin sequence.[20]

hypoxic injury.[7]

Cleft palate is seen in most patients with PRS.[1,7] Palatine development occurs between six and nine-weeks gestation with the lateral palatine shelves fusing in the midline.[6] In children with PRS, it is generally accepted that the superior positioning of the tongue can interfere with the fusion of the lateral palatine shelves.[3] The presence of cleft palate in PRS infants further complicates feeding issues impeded by micrognathia and glossoptosis. Lack of negative pressure in the mouth due to cleft palate, poor suck-swallow-breath reflexes, and respiratory distress can make feeding very difficult for these patients to the degree of aspiration, difficulty gaining weight, and failure to thrive.[7]

There are non-surgical and surgical approaches to treating PRS complications. Placing the infant on its side or prone relieves the airway obstruction in up to 70% of cases.[1] Proper positioning allows the infant to feed and sleep without as many obstructive episodes.[7] If the infant still has difficulty maintaining their airway, a nasopharyngeal (NP) tube can be placed to bypass the obstruction.[1] A newer technique creates an NP airway using an endotracheal tube individualized according to the patient's weight.[1] These airways can be maintained at home with proper cleaning and replacement to avoid surgery.[1,7] If conservative measures are not sufficient in preventing airway obstruction, surgical measures are considered.

Currently, there are several surgical procedures performed to correct the airway obstruction in PRS, including tracheostomy, tongue-lip adhesion, and mandibular distraction osteogenesis (MDO). Of these, MDO has gained popularity as a first-line procedure for a definitive solution to PRS while avoiding tracheostomy.[1] MDO is reserved for infants who have failed conservative treatment and have proven tongue-based airway obstruction.[1] This procedure lengthens the mandible via osteogenesis in the anterior direction, pulling the tongue with it.[8] As the tongue projects forward, the cause of airway obstruction is removed.

CASE PRESENTATION

A newborn female presented as a transfer to the neonatal intensive care unit (NICU) after spontaneous vaginal delivery at 38 weeks gestation to a 24-year-old G3P0211 mother. Pregnancy was complicated by IUGR, tobacco use, heroin and amphetamine use, Hepatitis C infection, HSV-2 infection, and limited prenatal care. The mother tested positive on urine drug screens during the pregnancy for TCAs, THC, opioids, methamphetamines, and amphetamines. The mother stated she stopped using illicit drugs two months before delivery. A urine drug screen of the mother on delivery was negative.

APGAR scores were nine and nine at one and five minutes, respectively. Shortly after delivery, the patient experienced respiratory distress and oxygen desaturations that were only improved by prone positioning and 100% oxygen on CPAP. When the patient's micrognathia and cleft palate were discovered, she was transferred to Cabell Huntington Hospital to be admitted to the NICU for further monitoring and management. A nasopharyngeal airway was placed using a modified 3.0 endotracheal tube, and the infant's work of breathing improved greatly. Due to her micrognathia, glossoptosis, and airway obstruction without any other dysmorphic features, she was diagnosed with non-syndromic PRS with cleft palate. On day of life two, flexible laryngoscopy was performed to visualize the upper aerodigestive tract. During laryngoscopy, glossoptosis was noted without any other masses or lesions. The patient's airway was maintained with a nasopharyngeal tube, and oxygen was slowly weaned as tolerated to room air. Multiple

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feeding trials were performed. The patient showed dyscoordination of her swallow and developed increased work of breathing with feeds. She remained dependent on nasogastric tube feeds. This patient was determined to be a good candidate for MDO. As she was otherwise stable, the decision was made to operate once she weighed over three kilograms.

The patient also began to show signs and symptoms of neonatal abstinence syndrome (NAS) during her first weeks of life. Her NAS scores were consistently above eight for a few days, which is diagnostic for the condition.[15] The patient exhibited withdrawal symptoms such as irritability, tremulousness, and gastrointestinal upset. Due to the mother's history of substance abuse, umbilical cord blood was sent for toxicology after delivery, which came back negative. In response to her withdrawal symptoms, the patient was started on methadone maintenance therapy of 0.22mg as needed. She only required maintenance therapy for a few days.

Direct laryngoscopy yielded a grade 2b view of the larynx on the day of operation. Bronchoscopy was unremarkable for distal airway pathology. Bilateral submandibular incisions were made to approach the inferior border of the mandible, taking care to protect the marginal mandibular branch of the facial nerve. The mandible was exposed from the external angle to the internal angle. KLS Martin (Tuttlingen, Germany) 20 mm end-driven Micro Zurich internal mandibular distractors were positioned and fixated to the mandible. An inverted-L osteotomy was made between the external and internal angles of the mandible bilaterally using a 1 mm diamond burr with identification and preservation of the inferior alveolar nerves. The patient underwent distraction for eight days post-operatively with a total distraction of 13.5 mm (Figure 2). This improved the facial profile and micrognathic appearance, and, more importantly, the airway obstruction was resolved. The patient was weaned to room air. Oral feedings were fully advanced with a specialty feeder given her cleft palate. She was discharged home with plans for internal hardware removal after eight weeks for adequate bone consolidation.

DISCUSSION

Our presented case of a non-syndromic PRS infant highlights the use of MDO to correct micrognathia and upper airway obstruction. Prior to MDO, tracheostomy and tongue-lip adhesion (TLA) were the main procedures offered to address airway obstruction in PRS patients.[9,11,18] TLA is a relatively simple procedure that can offer high rates of airway obstruction correction in PRS. It is associated with its own list of complications, including wound dehiscence, desquamation and ulceration of the exposed tongue, and a need for eventual adhesion release. Rates of airway obstruction resolution are comparable for MDO and TLA. TLA may have a higher rate of gastrostomy tube dependence and need for subsequent intervention.[19] Tracheostomy offers complete bypass of the airway obstruction. However, tracheostomies are associated with up to a 43%

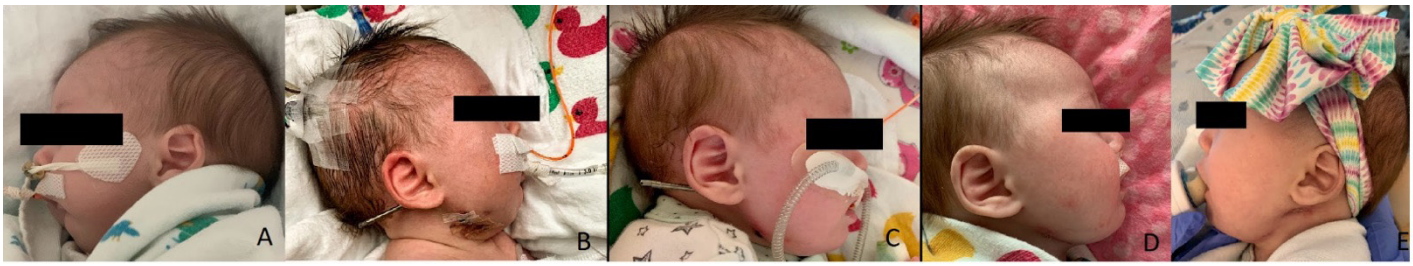

FIGURE 2. Patient with Pierre Robin sequence before and after mandibular distraction osteogenesis. (A) Preoperative, note nasopharyngeal airway and orogastric tube. (B) Postoperative day three with distractor arm exiting posteriorly. (C) Postoperative day seven, weaning O2 supplementation. (D) Postoperative day 18, note director arms have been removed, patient tolerating near full oral feeds. (E) Patient demonstrating good tolerance of bottle feeding within 21 days after surgery with specialty bottle feeder, nasogastric tube removed.

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complication rate and a 0.7%-7% mortality rate.[9,11] Not only can they lead to medical complications, but they also require full-time maintenance from the patient's caregivers. PRS patients have their tracheostomy in place for an average of three years before decannulation, which can lead to speech and other language delays.[9] As an alternative to TLA and to avoid the medical and social burden of a tracheostomy, MDO has gained popularity among surgeons as a first-line procedure for PRS. Unlike tracheostomy, MDO corrects the upper airway obstruction rather than bypassing it. MDO success is predicated on the avoidance of tracheostomy and gastrostomy tube placement. Zhang et al. conducted a study that showed a 95% avoidance of tracheostomy in PRS patients who underwent MDO, while Tahiri et al. had an 88.9%-92.6% avoidance of tracheostomy.[9,12] Although MDO is an alternative to tracheostomy, some patients may not be good candidates. A tracheostomy may be the best option if the patient has a comorbid neurologic or cardiopulmonary disease.

Another unique feature of this case is the patient's neonatal abstinence syndrome (NAS) diagnosis. Managing withdrawal symptoms and weaning these infants off maintenance therapy can prolong their hospitalization considerably. The patient's mother had positive urine drug screens throughout her pregnancy that enforced the diagnosis of NAS. Although the patient's cord toxicology was negative, she still exhibited significant withdrawal symptoms. There is a potential correlation between substance exposure in utero and the patient's PRS diagnosis. There have been documented associations between NAS and craniofacial abnormalities, including orofacial clefting. West Virginia University School of Medicine conducted a study that found a prevalence of orofacial clefting in 6.79 per 1000 births in the hospital's NAS population compared to 1.63 per 1000 births in the general population.[13] Danis et al. reported a prevalence of 3.13 per 1000 births in the NAS population compared to 1.35 per 1000 births in the general population.[14] Both studies concluded that opioid use during pregnancy could be a potential environmental factor for orofacial clefting in infants.[13,14] Orofacial clefting is considered a multifactorial process, so these significant differences in prevalence point to opioid use contributing to the pathology greatly.

The studies mentioned above only included patients with isolated cleft lip, isolated cleft palate, or combined cleft lip and palate. No patients with diagnosed PRS were included in these studies. There is currently little literature exploring a potential association between substance abuse during pregnancy and PRS. Our case presents a possible avenue of new research into substance-induced birth anomalies. West Virginia (WV) would be an appropriate starting point for further research due to its unique patient population. WV is disproportionately affected by the ongoing opioid epidemic in the United States. A study conducted in 2019 showed that the NAS rate in WV is seven times higher than that of other rural states.[16] Such high NAS rates would increase the data pool to analyze for a possible PRS association in the future.

CONCLUSION

Upper airway obstruction in infants with Pierre Robin sequence can lead to significant morbidity and mortality. To permanently relieve this obstruction, surgeons can employ mandibular distraction osteogenesis. This surgical technique can prevent the need for a tracheostomy. The physician should use his or her best judgment to determine if the patient is a good candidate for the operation. Other comorbid conditions should be carefully managed before and after surgery.

CONSENT

The patient's primary caregiver provided consent to use images of the patient and write about her case.

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