Pierre Robin Sequence in a Neonate With Suckling Difficulty and Weight Loss

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Pierre Robin sequence (PRS) is a combination of micrognathia and glossoptosis that leads to airway obstruction, feeding problems, and potentially other sequelae that can complicate early life. Currently, therapy for these newborns and infants is focused on preventing fatality or complications until the jaw can grow sufficiently to correct airway obstruction and associated problems. For patients with mild PRS, treatments include conservative measures such as airway maintenance and a feeding tube. For more severe cases, a surgical procedure is necessary. To the authors’ knowledge, the effectiveness of osteopathic manipulative treatment of patients with PRS has not been reported in the literature. The current case describes the use of osteopathic manipulative treatment to help alleviate difficulties with breathing and latch and sucking in a 15-day-old newborn with PRS.

J Am Osteopath Assoc. 2014;114(9):727-731
doi:10.7556/jaoa.2014.142

In 1923, a French physician described a set of facial abnormalities that led to obstruction of the upper airway. These included micrognathia (an undersized jaw) and glossoptosis (an abnormal posterior positioning of the tongue).1 The physician, Pierre Robin, MD, postulated that the abnormally small mandible led to the malpositioning of the tongue such that it occluded the upper airway, making it difficult to breathe.

Today, we recognize Pierre Robin sequence (PRS), also known as Pierre Robin syndrome, as micrognathia, glossoptosis, and obstruction of the airway, often accompanied by palatal abnormalities.1-7 Because the positioning, size, shape, and orientation of the mandible aids in the positioning of the tongue, the micrognathia seen in PRS leads to a shorter floor of the mouth and is believed to be the cause of the posterior positioning of the tongue and sucking difficulty.2,4-7

With airway obstruction as a key feature of PRS, often a newborn will present in the first few days of life with oxygen desaturation, apnea, and cyanosis.4 Specifically, the airway obstruction hinders inspiration, leading to increased respiratory effort, in which the newborn must use the accessory inspiratory muscles, which can be detected on physical examination.4-7 Simple positional changes may provide some relief. For example, placing the newborn in a prone position may be helpful compared with the supine position, which further aggravates breathing.2,4,7

Another life-threatening characteristic of PRS is that the tongue’s position in the posterior pharynx makes it difficult for the newborn to breathe and eat at the same time.2,6,7 Further complicating the problem is that the characteristic shape of the mandible as well as any palatal abnormalities, such as a cleft or a high-arched palate, makes latching difficult
and inhibits sucking, all of which ultimately leads to poor intake and failure to thrive. Other sequelae include gastroesophageal reflux and aspiration, which can lead to further complications, such as aspiration pneumonia and failure to thrive.

In addition to intrauterine insults in the seventh to eleventh weeks, genetic aberrancies can result in the same mandibular structural abnormalities leading to PRS. Pierre Robin sequence has been associated with other congenital abnormalities, such as Stickler syndrome, velocardiofacial syndrome, craniofacial microsomia, Treacher Collins syndrome, and Down syndrome. With treatment, either through palliative measures, a surgical procedure, or osteopathic manipulative treatment, the jaw can be repositioned, allowing the tongue to move anteriorly, improving the newborn’s or infants’ ability to breathe and eat.

We present the case of a 15-day-old newborn who had difficulty breathing and sucking and latching. His diagnosis of PRS was successfully managed with osteopathic manipulative treatment (OMT).

Report of Case
A 15-day-old male newborn presented with limited sucking ability. The patient was born via spontaneous vaginal delivery at 38 weeks to a gravida 3, para 2 mother (2 term deliveries, 0 preterm deliveries, 0 abortions, 2 living children). The patient was in the breech position until a successful conversion was performed 24 hours before birth. Labor was induced with oxytocin, and the mother received epidural anesthesia during labor. Neither forceps nor a vacuum extractor was used during delivery. At birth, the newborn weighed 7 lb, 5 oz (3321 g), and his Apgar scores were 8 at 1 minute and 9 at 5 minutes.

In the family care unit, the patient became cyanotic; he was noted to have nasal airway noise that indicated difficulty breathing and a small mandible. At that time, 3.5 hours after birth, the patient was transferred to the neonatal intensive care unit (NICU) because of respiratory distress. He spent the next 8 days in the NICU, where he was evaluated and treated for respiratory distress and hyperbilirubinemia. During that 8-day span, he was noted to have episodes of oxygen desaturation during feeding as well as during periods of crying. As a result he required tactile stimulation to recover from his apneic episodes, feeding via nasogastric tube, and a fiber-optic blanket for his hyperbilirubinemia.

The patient’s physical examination results were normal with the exception of a subjectively small and receded mandible, loud upper-airway noises, increased breathing effort, and desaturation episodes while supine, feeding, sucking on a pacifier, or when agitated. Although he showed oral responses when breastfeeding was attempted, with brief latching and oral movement, true transfer and nutritive sucking were lacking. In the NICU, a lactation consultant determined that the mother was producing adequate amounts of milk and that the newborn was better able to feed from a bottle. His discharge diagnosis was mild PRS, with instructions for the mother to breastfeed and supplement bottle feedings as needed. The patient weighed 7 lb, 7 oz (3375 g) at discharge.

Despite the newborn’s ability to feed from the bottle, his mother noted that during feeding, he would become exhausted after 5 to 10 minutes and remain hungry. This new development prompted her to consult her osteopathic physician about OMT for her newborn’s symptoms.

On presentation, the patient’s mother related that her newborn received a diagnosis of mild PRS and that they were awaiting genetic test results for cystic fibrosis. The mother also related that there had been no postdelivery infection, and she reported a family history of type 2 diabetes mellitus. She emphasized the newborn’s limited ability to latch and suckle. Physical examination results were as follows: weight, 7 lb, 8 oz (3402 g); pulse rate, 140 beats per minute; respiration rate, 40 breaths per minute; heart rate and rhythm, normal and without murmurs, rubs, or clicks; lungs, clear to auscultation bilateral.
ally without wheezes, rales, or rhonchi; normocephalic/atraumatic; intact extraocular movements; positive grasp bilaterally; positive rooting bilaterally; positive Moro reflex; and was able to move all 4 limbs equally. Micrognathia and limited sucking (evaluated by inserting a pinky into the patient’s mouth, allowing him to engage in sucking, and evaluating the pressure generated) were also present. On osteopathic examination, the patient had a sphenobasilar synchondrosis compression, left occipitomastoid (OM) suture compression, tight left sternocleidomastoid, sacroiliac joint restricted on the left, and a left in-flare of the innominate. The sacrum, OM suture, and sphenobasilar synchondrosis compression were treated with osteopathic cranial manipulative medicine, and the sternocleidomastoid and pelvis were treated with balanced ligamentous tension. At the end of treatment, the patient’s sucking had improved when retested, in the same manner as above, and his breathing had become less labored on visual examination.

At the follow-up appointment 3 weeks later, the mother reported that he had gained 1 lb, grown 2.5 in, and was eating better with less effort and longer time to fatigue. Review of systems at this time was normal with the exception of the presence of oral thrush. Physical examination results were as follows: weight, 8 lb, 7 oz (3827 g); height, 21.5 inches; pulse rate, 150 beats per minute; respiration rate, 30 breaths per minute; heart rate and rhythm, normal and without murmurs, rubs, or clicks; lungs, clear to auscultation bilaterally without wheezes, rales, or rhonchi; normocephalic/atraumatic; intact extraocular movements; positive rooting bilaterally; positive Moro reflex; and was able to move all 4 limbs equally. The patient’s micrognathia and sucking had improved. Osteopathic examination results showed the left parietal overriding the left temporal bones, a mild compression of the left OM suture, atlantoaxial joint rotated left, a right second sacral vertebrae restriction, and a left hemidiaphragm restriction. The cranial, cervical, and sacral lesions were treated with osteopathic cranial manipulative medicine, and the diaphragm was treated with balanced ligamentous tension. At the end of the treatment, sucking was intact, and the parietal bone was no longer overriding the temporal bone on the left.

The patient was evaluated 3 weeks later for complaints that were unrelated to PRS. At that time the mother reported that her son’s condition had improved. After the initial treatment, she said that he had started to refuse the bottle after breastfeeding, was no longer fatigued after 5 to 10 minutes of breastfeeding, had gained approximately 2.5 pounds in the first week after the initial treatment, and had grown 3 inches in that same period. Physical examination results were as follows: weight, 10 lb (4536 g); pulse, 150 beats per minute; respiration, 38 breaths per minute; heart rate and rhythm, normal without murmurs, rubs, or clicks; lungs, clear to auscultation bilaterally without wheezes, rales, or rhonchi, although stridor was present; positive rooting bilaterally; positive Moro reflex; and was able to move all 4 limbs equally. His micrognathia remained, and his sucking ability was strong. Osteopathic examination results revealed a restricted right OM suture with no overriding of the left parietal on the left temporal bone, tight right sternocleidomastoid, and a right unilateral flexion at first sacral vertebrae. These areas were treated with osteopathic cranial manipulative medicine.

Several weeks after the last follow-up, the mother noted continued improvement in suckling, eating, weight, and development. She also stated her intent to continue with the treatments because they had provided substantial improvement.

**Discussion**

Owing to the complexities of PRS and possible genetic abnormalities, treatment of these patients requires a multidisciplinary approach to investigate the anatomic abnormalities that result in airway obstruction and the degree of airway obstruction. Specialists who should be involved in the management of PRS include those in plastic surgery, otolaryngology, pulmonology, speech...
pathology, neurology, and anesthesia. A genetics consultation should also be considered in PRS cases in which other genetic disorders are suspected.

The goal of management and long-term treatment for the first 6 months to 1 year is to ensure the survival and health of the patient.\(^4\) This time should allow the mandible to grow sufficiently to allow the tongue to move anteriorly, clearing the airway and improving feeding.\(^2\)

**Conservative Management**

Mild cases require a conservative approach, involving positional changes while sleeping and while feeding. Sleeping and eating positions should be changed from supine to prone or lateral to minimize or prevent the desaturation events that can occur during these activities.\(^4\) If the positional change does not sufficiently prevent or minimize the desaturation events, nasopharyngeal airway is placed to ensure that the patient receives sufficient oxygen. Nasogastric tube feeding is also used in mild cases of PRS in which the patient has feeding difficulty without airway obstruction and positional change does not help.\(^5\)

In patients who do not require surgery, OMT can be used. Cranial techniques in combination with other modalities of osteopathic manipulative medicine can help patients with PRS breathe easier and struggle less when feeding without the complications and side effects associated with surgery.

**Surgical Management**

**Tongue-Lip Adhesion**

If the tongue is the sole cause of airway obstruction, and conservative measures have failed, the patient may be a candidate for tongue-lip adhesion (TLA). In this procedure, the tongue is pulled forward, out of its posterior position in the laryngopharynx, and sutured to the bottom lip.\(^4\) Once the mandible has grown sufficiently, at the age of 9 months to 1 year, the TLA can be reversed. As with any surgery, there is a risk of bleeding, infection, and complications associated with anesthesia. Further, if teeth have grown in, they may cut through the suture prematurely.\(^4\)

**Distraction Osteogenesis**

When conservative measures and TLA have failed, and the tongue is the sole cause of the airway obstruction, distraction osteogenesis of the mandible is an option. This procedure begins with fracturing the mandible on the lingual surface and distracting it forward 2 to 3 days later. The distraction process is continued each day for 2 to 3 months moving the jaw 0.5 to 2 mm per day.\(^4\) The result is an increase in the size of the mandible, with the secondary effect of pulling the tongue forward and out of the laryngopharynx. Complications associated with the procedure include those associated with surgery and anesthesia, as well as inferior alveolar nerve neurapraxia, tooth root injuries, severe scarring, reabsorption or ankylosis of the temporomandibular joint, poor union of the fracture, and failure to provide relief of symptoms.\(^4\)

**Conclusion**

Wolff’s law (Julius Wolff, MD) states that “bone under a mechanical load will modify its structure along the axes of force and stress.”\(^9\) Chamay and Tschantz\(^6\) elaborated on that law, stating that intermittent forces placed on a bone activate and sustain osteogenic processes to a greater degree than a sustained force.\(^9\) The mandible is greatly influenced throughout growth and development by Wolff’s law.\(^10,11\) In the current case, the mandible and the muscles that allowed it to function, were manipulated to alter the forces that were placed on it. The third tenet of osteopathic medicine states that structure and function are reciprocally interrelated.\(^12\) The current case depicted altered structure and hence poor function in a newborn with PRS. Osteopathic manipulative treatment of this patient improved structure in conjunction with the natural growth and healing processes of the body and thus improved function.
Acknowledgments

We thank Amber Koon, OMS V, for her contributions to this case and manuscript, and the Rocky Vista University College of Osteopathic Medicine library staff for their help in editing.

Author Contributions

Student Doctor Summers, Student Doctor Ludwig, and Dr Kanze provided substantial contributions to conception and design, acquisition of data, or analysis and interpretation of data; drafted the article or revised it critically for important intellectual content; and gave final approval of the version of the article to be published.

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