Surgical Strategies
Transmandibular K-Wire in the Management of Airway Obstruction in Pierre Robin Sequence

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The Pierre Robin sequence was first described by Pierre Robin in 1923 as a triad of micrognathia, U-shaped cleft palate, and glossoptosis. Although the problems associated with Pierre Robin sequence may be numerous, the most acute problem in affected newborns is upper airway obstruction. The causes of upper airway obstruction are heterogeneous, and treatment is controversial and may include a long stay in an intensive care setting. Most patients could be treated conservatively with prone/lateral positioning and close clinical observation. A more aggressive approach is to hold the tongue forward surgically by a lip-tongue adhesion (glossopexy) technique, and if all else fails, it might be necessary to perform a tracheostomy. The purpose of this article is to report an unconventional technique for the management of airway obstruction in Pierre Robin sequence. A transmandibular K-wire was used in two patients with Pierre Robin sequence to prevent airway obstruction. The author was not the surgeon who placed the K-wire, and in one of the patients, the K-wire was retained for 4 years before being removed by the author. Both patients had surgical intervention to manage the airway problem in the form of lip-tongue adhesion in addition to the transmandibular K-wire. In conclusion, the value of using a transmandibular K-wire in the two cases presented here could not be determined and was questionable.

Key Words: Pierre Robin sequence, K-wire

The Pierre Robin sequence (PRS) is a pathogenetically and etiologically heterogeneous condition that can be a nonsyndromic anomaly or one feature of many syndromes. It was first described by Pierre Robin in 1923 as a triad of micrognathia, U-shaped cleft palate, and glossoptosis commonly complicated by upper airway obstruction. Because the criteria for defining PRS are variable, the reported prevalence also varies, estimated between 1 in 2,000 to 1 in 30,000. Various hypotheses on the cause of PRS have been proposed, but no conclusive evidence is available. Although the problems associated with PRS may be numerous, especially if the primary cause is a multiple anomaly syndrome, the most acute problem in affected newborns is upper airway obstruction. Infants are at increased risk of hypoxic complications such as cor pulmonale, failure to thrive, and cerebral impairment. The causes of upper airway obstruction are heterogeneous, among which are the posterior displacement of a normal-sized tongue secondary to micrognathia/retrognathia, the loss of support of the genioglossus muscle, and negative pressure in the pharynx during swallowing and inspiration. Upper airway obstruction in association with PRS presents at or shortly after birth; however, late presentation of upper airway obstruction has also been reported. Treatment of this obstruction is controversial and may include a long stay in an intensive care setting. Because of the variation in the severity of upper airway obstruction, airway management should be tailored to the individual needs of each patient. The choice of treatment is determined by the severity of the upper airway obstruction. Most patients could be treated conservatively with prone/lateral positioning and close clinical observation. If this conservative approach does not solve the problem, nasopharyngeal or nasotracheal intubation may be necessary. A more aggressive approach is to hold the tongue forward surgically by a lip-tongue adhesion (glossopexy) technique. Finally, if all else fails, it might be necessary to perform a tracheostomy.
Recently, it has been postulated that the musculature of the floor of the mouth is under increased tension, pushing the tongue upward and backward, which causes the respiratory obstruction, and that subperiosteal release of these muscles would alleviate the tension and hence clear the obstruction.7,22

The purpose of this article is to report an unconventional technique for the management of airway obstruction in PRS. Two patients with PRS had a transmandibular K-wire placed to prevent airway obstruction. The option of transmandibular K-wire placement in the management of airway obstruction in PRS patients is discussed.

Case 1

A male infant was born by cesarean section at a private hospital to a healthy mother at 38 weeks of gestation with a birth weight of 3 kg. The infant was noted to have a “small” jaw, was experiencing recurrent episodes of respiratory difficulty, and was kept in the intensive care unit. Shortly after arrival in the intensive care unit, a cleft palate was noted. Because of the deterioration of the patient’s respiratory status, it was decided by the attending surgeon to intervene surgically. According to the mother, surgery was performed when the infant was 4 days old and included tongue-lip adhesion and passing a wire through the infant’s lower jaw (transmandibular K-wire).

The patient was seen at the Cleft Lip and Palate Center at the Jordan University of Science and Technology and King Abdullah University Hospital at the age of 4 years. The mother’s main concerns were the cleft palate and the fact that her baby could not talk because of the wire in his lower jaw and tongue. There was no family history of PRS, cleft lip/palate, or features of an obvious related genetic condition. Genetic counseling did not show any underlying genetic abnormality. The patient had cerebral palsy, which was likely caused by hypoxic injury. Hematological and biochemical investigations revealed that patient had glucose-6-phosphate dehydrogenase deficiency. Lateral and posterior-anterior radiographic views of the skull showed a transmandibular K-wire extending from the region of the right first mandibular molar to the left first mandibular molar (Fig 1). The patient was admitted to hospital, the K-wire was removed at same time as the cleft palate repair, and the patient had an uneventful recovery.

Case 2

This male infant was born at a private hospital by normal vaginal delivery at 36 weeks of gestation. The
birth weight was 2,550 g. Micrognathia and cleft palate were apparent, and the infant was experiencing significant respiratory difficulties and cyanosis soon after birth. The patient was admitted to the intensive care unit, and on day 4 of life, lip-tongue adhesion was performed; however, this was unsuccessful. On day 10 of life, a wire was passed through the mandible (transmandibular K-wire) and a second lip-tongue adhesion was performed. The lip-tongue adhesion failed for the second time, and 2 weeks later, the patient developed a huge facial swelling that was caused by infection related to the K-wire. The K-wire was removed, and a third lip-tongue adhesion was attempted. Feeding was managed using a nasogastric tube, and respiratory problems continued but were less severe than before.

The patient was first seen at the Cleft Lip and Palate Center at the Jordan University of Science and Technology and King Abdullah University Hospital at the age of 2 months after referral by his pediatrician. The parents were second-degree relatives, and the father indicated that his nephew was born with a cleft palate; facial, ocular, and finger abnormalities; and heart problems. These features were suggestive of Stickler syndrome. The parents have another older healthy girl (3 years old), and genetic counseling failed to show any underlying genetic abnormality. The parents did not have a clear idea about the condition of their child and were extremely concerned. The condition and its management were explained to the parents, and they were given advice on feeding and conservative management of the airway obstruction. The patient successfully underwent palatal repair and release of lip-tongue adhesion at the age of 14 months.

**DISCUSSION**

Airway obstruction is the most common presenting feature of PRS, and different theories have been suggested to explain the possible mechanism of this obstruction. One of the widely accepted theories is posterior displacement of the tongue secondary to micrognathia/retrognathia. Negative intrathoracic pressure generated during inspiration and swallowing pulls the tongue back, which may be aggravated by the lack of voluntary control of the tongue musculature. Pharyngeal abnormalities have also been suggested as a possible cause of airway obstruction. Correct identification of the mechanism of obstruction provides guidance for management, and treatment must be individualized to optimize the outcome. The medical literature describes a variety of methods of intervention, including positioning, nasopharyngeal or endotracheal intubation, lip-tongue adhesion (glossopexy), and tracheostomy. More recently, a surgical procedure was introduced for subperiosteal release of the musculature of the floor of the mouth. All interventions are long term until mandibular growth and neuromuscular maturation of the tongue muscles remove the possibility of obstruction.

Reviewing the English literature did not reveal any report of using a transmandibular K-wire in the management of airway obstruction in PRS patients. It seems that the rationale behind its use in the two cases presented in this article was to stabilize the base of the tongue, preventing its backward displacement. The insertion and removal of a transmandibular K-wire can be relatively simple, and compared with the lip-tongue adhesion technique, a transmandibular K-wire may be considered less invasive. The technique is not without complications, however, because it may be associated with damage to the developing tooth germs, inferior dental neurovascular bundle, and other adjacent structures.

In the two cases presented in this report, it was not possible to determine the precise cause of airway obstruction, because one patient was seen at the age of 4 years and the other was seen at the age of 2 months. In addition, both patients had surgical intervention to manage the airway problem in the form of lip-tongue adhesion in addition to placement of a transmandibular K-wire. In conclusion, the value of using a transmandibular K-wire in the two cases presented here could not be determined and was questionable. Comments by interested parties on the management of PRS are invited as to whether the technique merits consideration and further exploration.

**REFERENCES**