Polysomnographic Indications for Surgical Intervention in Pierre Robin Sequence: Acute Airway Management and Follow-Up Studies After Repair and Take-Down of Tongue-Lip Adhesion

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The major concern for neonates with Pierre Robin sequence is the stability of the airways. The accepted management has been close clinical observation followed by surgical intervention if the airway was felt to be unstable. Six newborns with this diagnosis were admitted for evaluation in a 7-month period. Each underwent transternal oxygen and carbon dioxide monitoring in a resting state. Each infant then was evaluated with infant polysomnography to ascertain the presence of obstructive apnea. Pulse-oximetry was utilized to document the oxygen saturation of each child during the polysomnography. On the basis of these studies, four of the neonates required a tongue-lip adhesion to stabilize the airway. These four infants were reassessed with polysomnography postoperatively, prior to cleft palate repair, following palatoplasty, and after their tongue-lip adhesion was released. This method of evaluation allows early testing of the stability of the airway in a way that augments and confirms the clinical assessment of the infant, allowing appropriate surgical intervention when necessary. This method of evaluation also allows the safe prediction of airway stability following palatoplasty and release of the tongue-lip adhesion.

KEY WORDS: airway obstruction, Pierre Robin sequence, polysomnography, tongue-lip adhesion

The presence of retrognathia and glossoptosis in a newborn, leading to respiratory problems, with or without a cleft palate, is referred to as the Pierre Robin sequence. Untreated, many of these infants can present with numerous complications ranging from failure to thrive and malnutrition to cor pulmonale, hypoxia, pneumonia, and even death. This has been referred to as a "glossoptosis-apnea syndrome" (Cozzi and Pierro, 1985). Although Robin introduced the term "glossoptosis" in 1934 to describe the tongue falling back and causing pharyngeal obstruction, its treatment still remains controversial.

Four major methods of treatment have evolved. These include: (1) positional; (2) intra-oral or nasopharyngeal endotracheal airway; (3) tongue-lip adhesion; and (4) tracheostomy. What has been lacking is a standardized, predictable approach for establishing when the more invasive treatments are indicated.

To date, most decisions are based on clinical criteria such as: can the child breathe easily while resting or sleeping, does he or she become cyanotic during feeding, or does he or she show a progressive gain in weight and strength over the first 7 days of life? We propose a noninvasive approach that can quickly and reproducibly determine the infant's need for intubation, surgical intervention, or both.
When faced with six newborns admitted to our neonatal nursery meeting the criteria of the Robin sequence, we found nothing in the literature search to provide criteria that would predict those infants who would not need surgical intervention. An article written in 1982 suggested the above-mentioned clinical criteria as the only satisfactory way to recommend the need for surgical intervention (Parsons and Smith, 1982). Our technique allows the decision regarding surgical intervention to be determined using standardized, instrumental recordings, and not having to rely on clinical findings alone.

**METHODS**

Six newborns with the diagnosis of the Robin sequence were admitted for evaluation over a 7-month period. All infants were born at term and had birth weights ranging from 2,395g to 4,820g. All patients had micrognathia, cleft palate, and glossoptosis with airway compromise. All infants were evaluated by a geneticist to rule out other congenital conditions that might lead to airway obstruction (i.e., Treacher-Collins syndrome).

Upon admission, a bedside evaluation of the infant’s average transcutaneous oxygen (T\textsubscript{O\textsubscript{2}}) and transcutaneous carbon dioxide (T\textsubscript{CO\textsubscript{2}}) levels over a period of time were determined (Hansen and Tooley, 1979; Rowe, et al 1980). The transcutaneous monitor used (SensorMedics) has a memory mode that stores average O\textsubscript{2} and CO\textsubscript{2} levels every minute. When placed back on the “mother module,” each bedside unit is capable of plotting histograms based on this stored memory. Each infant was monitored for a minimum of 8 hours (range 8 to 18 hours). Average oxygen levels below 60 mm Hg and carbon dioxide levels above 50 mm Hg were considered by us to be abnormal for the purposes of this study (Brouillette, et al 1982; Talbot and Robertson, 1973).

For confirmation of the reliability of the transcutaneous monitoring a modified polysomnographic (sleep) study was then performed. Each infant was monitored on a Grass Model 78 polygraph. Nine channels were simultaneously utilized including nasal and oral thermister and nasal end-tidal CO\textsubscript{2} to document air flow, thoracic and abdominal strain gauges to monitor mechanical respiratory effort, and diaphragmatic EMG to monitor the phrenic nerve output. Oxygen saturation levels via a Nelcor N-100 pulse oximeter and heart rate with an ECG and tachograph were recorded (Fig. 1). Because we were able to monitor both airflow via use of the end-tidal CO\textsubscript{2} and thermisters while simultaneously recording thoracic and abdominal movements (strain gauges), we could accurately determine the presence of airway obstruction. As O\textsubscript{2} saturations were also being measured, the significance of such obstructive episodes could also be determined. All children were studied in lateral, prone, and supine positions for a minimum total of 45 minutes. The study was analyzed to determine the presence of obstructive apnea and O\textsubscript{2} saturation levels both at rest and during obstructive episodes. Using these data to support our clinical impressions, we made decisions to recommend surgical intervention for airway management. The procedure chosen was a tongue-lip adhesion so that tracheostomy could be avoided (Parsons and Smith, 1980). The infants were again studied as described for the initial evaluations after they were extubated (approximately 3 to 5 days postoperatively). The definitive palatoplasty was scheduled when the children were 1 year of age and had a normal sleep study. Six weeks following palate repair, all children were again evaluated by the same criteria. No obstruction was noted. The release of the tongue-lip adhesion was done without untoward sequelae. Airway patency was again confirmed by our studies.

**CASE REPORTS**

The following two patients represent the two categories that resulted from our assessment. The first child underwent a tongue-lip adhesion having met the criteria established by our assessment while the second child did not require surgical intervention.

**Case 1: Baby DR**

This patient, a white male infant, was the product of an uneventful term pregnancy, labor, and vaginal delivery to a 27-year-old G\textsubscript{1} P\textsubscript{1} A+ mother. The birth weight was 2,395g and the APGAR scores were 8\textsuperscript{1} and 8\textsuperscript{5}. Upon admission to the nursery the child was noted to have a large “U” shaped cleft of the palate, micrognathia, right metatarsus adductus, and a right preauricular skin tag. In addition, the child was small for gestational age. Oral feedings were attempted but not tolerated, and the child developed moderate respiratory distress when placed in the supine position. A diagnosis of Pierre Robin sequence was made, and the child was transferred to our hospital for evaluation by regional Cleft Palate Team.

Admission examination was consistent with the Robin sequence, and the infant was placed on a transcutaneous monitor for determination of T\textsubscript{O\textsubscript{2}} and T\textsubscript{CO\textsubscript{2}} trending. A 12-hour trending revealed an average T\textsubscript{O\textsubscript{2}} of 54 mm Hg and T\textsubscript{CO\textsubscript{2}} of 57 mm Hg. The next day a modified
polysomnographic (sleep) study was performed that showed numerous obstructive episodes and \( O_2 \) saturations as low as 60 percent. The child was then intubated with great difficulty and taken to the operating room 36 hours after admission for a tongue-lip adhesion. The surgery was well tolerated, and the infant was extubated 72 hours postoperatively. Oral feedings, which were begun after extubation, were easily handled. Postoperative histograms revealed \( O_2 \) in the 70s and \( CO_2 \) in the 30s. The repeat sleep study was free of obstructive episodes and the oxygen saturations were now in the 80 to 96 percent range. Ten days following surgery (12 days after admission) the child was discharged home on bottle feedings.

**Case 2: Baby KK**

This patient, a white female, was the 3,880g product of a full term pregnancy. The infant was delivered by cesarean section to a 21-year-old \( G_1 P_0 \) female. The initial examination in the nursery revealed that the child had a small chin and a cleft palate. The child was transferred to our institution for evaluation by the regional Cleft Palate Team.

Admission examination was consistent with the Pierre Robin sequence. The patient was placed on a transcutaneous monitor, and an 8-hour histogram revealed \( O_2 \) levels in the 70s and \( CO_2 \) levels in the 30s. A modified sleep study showed no obstructive episodes or oxygen desaturation in the lateral, prone, or supine positions. As a result, it was felt that surgical intervention was not indicated. Bottle feedings were started and were initially supplemented with nasogastric feedings. By the fourth day after the institution of feedings, the infant was taking 4 oz over a 30-minute period. She was discharged from the hospital 8 days after admission.

**DISCUSSION**

Upper airway obstruction presenting only during sleep has been recognized in adults since the 1960s. This obstructive apnea, which appears to be entirely functional and specifically induced by sleep, has been referred to as a "sleep apnea syndrome." In fact, most apnea occurring in adults is obstructive in nature. Term infants, unlike adults, rarely have obstructive apnea in a "normal" state. Instead, their apnea is usually centrally mediated. When obstructive apnea does occur in infants, especially in the presence of known upper airway anomalies (e.g., glossoptosis or micrognathia) a potentially dangerous situation exists and must be treated. The marked decrease in oxygen saturation that occurred in association with obstructive episodes is an indication of their pathologic nature. Although no standard exists for what is a normal \( O_2 \) saturation in a newborn, we decided to define any level
The reliability of using skin surface oxygen and carbon dioxide tensions in the evaluation of obstructive apnea has been demonstrated (Rowe et al., 1980). In each instance we found, as did the earlier studies, that those infants with $T_O_2$ below 60 or $T_CO_2$ above 50 on the histograms had obstructive episodes and had $O_2$ saturations below 80 percent on the sleep studies. Table 1 shows $T_O_2$ and $T_CO_2$ results pre- and postsurgery in addition to the pre- and postsurgical $O_2$ saturations. Those infants with abnormal $T_O_2$ levels and $T_CO_2$ levels uniformly had oxygen saturations below 80 percent during obstructive events on sleep studies.

Most of the literature currently available describes only subjective criteria as to when surgical intervention is indicated in the treatment of the Robin sequence. Such criteria as the presence of respiratory distress, color changes, or progressive gain in weight and strength are used (Parsons and Smith, 1982). To date, no clear objective criteria have been found. We have found that by using the aforementioned objective criteria: (1) average $T_O_2$ below 60 or $T_CO_2$ above 50 over a minimum of 8 hours, (2) obstructive episodes on sleep study, and (3) oxygen saturations below 80 percent, a determination can be made as to when to proceed with surgery (Rowe et al., 1980).

On the basis of these criteria, four of the six neonates evaluated received a tongue-lip adhesion to stabilize the airway (see Table 1). The infants were taken to the operating room where endotracheal intubation was achieved using a Jackson laryngoscope according to technique described by Handler and Kean (1983). With the airway stabilized and anesthesia induced, a tongue-lip adhesion was fashioned as described by Parsons and Smith (1980). The patients remained intubated for 72 hours postoperatively. The traction suture and submental button were removed on the seventh to tenth postoperative day.

Seventy-two hours postoperatively the infants were extubated (once airway edema had decreased) and oral feedings were started shortly thereafter. The length of hospitalization was determined by the infant’s ability to tolerate the oral feedings and ranged from 11 days to 54 days (average 23 days, although 4 of 6 infants were discharged in 18 days or less).

The same protocol was employed to evaluate the airway before scheduling the child’s palate repair at about 1 year of age. Following palatoplasty (approximately 6 weeks after surgery) the children were reevaluated without evidence of obstruction. Similar findings were noted after the tongue-lip adhesion was released, thus confirming our confidence in this technique to evaluate and predict the safe timing of surgical intervention.

In summary, we have found the technique of polysomnography to be of great help in the management of neonates born with the Pierre Robin sequence. This technique has given us a reliable method for determining the need for initial surgical intervention and for helping us plan subsequent procedures.

REFERENCES


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