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# Parental perceptions and morbidity: Tracheostomy and Pierre Robin sequence

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Pierre Robin tracheostomy and quality of life

**Summary**

*Objective:* Evaluate parental perceptions associated with tracheostomy morbidity and quality of life in the management of Pierre Robin Sequence (PRS).
*Study Design:* Retrospective review/survey.
*Methods:* 42 Pierre Robin patients were identified, records were reviewed and airway assessments evaluated relative to airway compromise. Twenty patients had undergone tracheostomy. Perceptions of quality of life/morbidity related to tracheostomy were assessed using parental surveys.
*Results:* 31/41 (76%) patients participated in the survey. 15/31 (48%) of survey participants required tracheostomy and were decannulated after a mean of 28 months. Of the patients who had undergone tracheostomy, 10/15 (67%) had isolated Pierre Robin (iPRS) and the remaining 5/15 (33%) had syndromes associated with Pierre-Robin (sPRS). 9/10 (90%) iPRS and 4/5 (80%) sPRS families' expectations were met regarding expected duration of tracheostomy although 3/5 (60%) sPRS, and 8/10 (80%) iPRS described the overall experience as difficult. Of the 2/15 patient's families who were dissatisfied 1 patient had iPRS and the other sPRS. 9/15 (60%) required multiple ($\geq$3) hospitalizations. 3/13 (23%) reported airway problems after decannulation and 2/15 (13%) remained tracheostomy dependent at the time of survey. Prolonged tracheostomy duration represented a significant parental concern.
*Conclusions:* A subset of patients required extended duration of tracheostomy; some continued to have airway problems after decannulation and/or distraction. Although some patients benefit from early mandibular distraction other Pierre Robin patients have multi-level obstruction requiring additional therapies and often tracheostomy. Parental concerns and perceptions relative to tracheostomy have not been

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adequately studied for Pierre Robin airway obstruction. Of those responding to this survey, the majority of parents' expectations were met regarding tracheostomy. Of those whose expectations were not met, it seems that better pre-intervention counseling regarding length of tracheostomy tube dependence, as well as a discussion about potential complications and hospitalizations frequently associated with prolonged tracheostomy, may lead to improved parental expectations.


## 1. Introduction

Pierre Robin sequence (PRS), initially described by Pierre Robin in 1923 and more fully depicted in 1926, is estimated to occur in 1 in 2000 to 1 in 50,000 live births [1]. Characteristic features include mandibular deficiency (micrognathia, or retrognathia), glossoptosis, or retropulsion of the tongue, which together often lead to airway obstruction. It is thought that these events lead to the classic U-shaped cleft palate as a superiorly pushed tongue inhibits the normal fusion of descending palatal shelves. Cleft palate was added to the constellation of findings in 1934, but is absent in many descriptions of PRS. Untreated, airway and feeding difficulties in the neonatal and postnatal period may result in failure to thrive, cyanosis, cerebral hypoxia, $CO_2$ retention, cor-pulmonale, pulmonary hypertension, heart failure, pulmonary morbidity and death.

The inciting event in the Pierre Robin sequence, mandibular hypoplasia, occurs between week 7 and 11 of gestation, leading to glossoptosis and subsequently to persistent clefting of the palatal shelves. PRS may be found as an isolated anomaly, or as part of a syndrome or complex including Stickler syndrome, velocardiofacial, cerebro—oculo—facial, and trisomy 18 [2]. Although the precise etiology and subsequent variable pathophysiology of PRS are not fully understood, the inciting events may be different in isolated (iPRS) versus syndromic Pierre Robin sequence (sPRS). The incidence of sPRS is significant and is reported in the literature to range from 30% to 80% [2,3]. Upper airway obstruction is generally more severe in sPRS, and patients frequently have concomitant systemic disabilities and multi-level airway obstruction. The prognosis for "catch up" growth, in which significant native mandibular growth takes place, ranges from the first 6 months to 2 years but may take 5—6 years to fully take place in PRS patients [4,5]. This is less likely in children with sPRS who will be more likely to require surgical management, often tracheostomy, to achieve a stable airway [5]. Of note, the long-term results of mandibular distraction in such syndromic patients may prove inadequate because of underlying mandibular growth abnormalities that persist beyond distraction [6].

Timing of airway compromise may be unpredictable, with presentation at any time during the first month of life; sudden infant death has been reported from day of life 13 to 95 [7]. Although respiratory distress is most frequently secondary to tongue base obstruction, other factors may also be involved, including neurologic issues affecting muscular coordination and tone.

A tenuous airway may be optimized with measures such as prone or lateral positioning, nasopharyngeal airway stents, positive pressure mask ventilation, temporary intubation, and glossopexy. These temporizing measures seek to stabilize the airway, with the anticipation that ultimately native-mandibular growth will improve the dimensions of the upper airway. However, children with severe PRS will often fail conservative management and have been traditionally treated with tracheostomy, with an expected duration of about 2—3 years. Increasingly, mandibular distraction has been gaining favor as an early treatment that helps avoid tracheostomy and its attendant morbidity and complications. We sought to evaluate the impact and morbidity of tracheostomy on patients and their families, and to discern characteristics that could predict a longer course of cannulation to assist in family counseling.

## 2. Methods

Institutional Review Board approval was obtained through the University of North Carolina, Chapel Hill Office of Human Research Ethics. PRS patients actively followed by the UNC-Craniofacial Center were identified and their medical records reviewed from July 2005 to June 2006 ($n = 41$). The families were retrospectively questioned using a survey regarding airway interventions, difficulties in daily care, expectations with regards to tracheostomy and decannulation, and overall impact on family dynamics. Patient charts were also retrospectively reviewed, and data regarding complications, hospitalizations, and bronchoscopic findings were collected. Pharyngoscopy with laryngoscopy and bronchoscopies were performed every 6 months on average, and findings were reviewed to identify the factors that correlate with a need for prolonged