Parental perceptions and morbidity: Tracheostomy and Pierre Robin sequence

Joshua Demke a, *, Marc Bassim a, Mihir R. Patel a, Shay Dean b, Reza Rahbar b, J.A. van Aalst b, Amelia Drake a

a Department of Otolaryngology, Head and Neck Surgery, University of North Carolina School of Medicine, Chapel Hill, NC 27599-7070, USA
b Division of Plastic Surgery, Department of General Surgery, University of North Carolina, Chapel Hill, NC 27599, USA

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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 (>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
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, CO2 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
Pierre Robin Study: All patients | Date of Survey  
---|---
1. Patient’s name:  
2. Did your child have airway problems at birth? Y/N  
3. Did airway problems develop after birth? Y/N Age started (in months)  
4. What options were required to treat the airway problems? Circle all that apply  
   a. Positioning: Y/N  
   b. CPAP: Y/N  
   c. BIPAP: Y/N  
   d. Lip-tongue adhesion? Y/N  
   e. Intubation: Y/N  
   f. Tracheostomy: Y/N  
   g. Distraction: Y/N  
   h. Other:  
5. What options for treatment were discussed with you? Circle all that apply  
   a. Positioning: Y/N  
   b. CPAP: Y/N  
   c. BIPAP: Y/N  
   d. Lip-tongue adhesion? Y/N  
   e. Intubation: Y/N  
   f. Tracheostomy: Y/N  
   g. Distraction: Y/N  
   h. Other:  
6. Does your child have a cleft palate? Y/N  
7. Is the cleft palate repaired? Y/N  
8. Time of repair? (Age in months):  
9. Did airway problems worsen after cleft palate repair? Y/N  
11. Does your child still require an apnea monitor? Y/N  
12. Has your child required a sleep study? Y/N How many? (Exact number)  
13. First sleep study? (age in months)  
   Most recent? (age in months)  
14. If multiple sleep studies done, is the most recent sleep study  

Pierre Robin Survey | Tracheostomy Patients | Date of Survey  
---|---|---
1. Child’s Name:  
2. At what age was your child’s tracheostomy placed? (in months)  
3. Were other options discussed with you? Y/N  
   a. Position changes? Y/N  
   b. Lip-tongue adhesion? Y/N  
   c. Intubation? Y/N  
   d. Distraction of the mandible? Y/N  
4. How long was the tracheostomy in place? (in months)  
5. At what age was the tracheostomy removed? (in months)  
6. How long did you believe the tracheostomy would be in place? (in months)  
7. Were your expectations met with regard to length of tracheostomy? Y/N  
8. Were your expectations met with regard to difficulty with tracheostomy? Y/N  
9. What was the most difficult aspect of having the tracheostomy in place? (1–10, 1 is easy and 10 is difficult)  
   a. Dealings with equipment company?  
   b. Respiratory care?  
   c. Repeat hospitalizations?  
   d. Mechanical failures?  
   e. Family lifestyle?  
   f. Public outings?  
   g. Nursing care?  
10. If there were repeat hospitalizations? Y/N How many?  
   a. Cause and duration for each?  
   b. Cause and duration for each?  
   c. Cause and duration for each?  
11. On a scale of 1 to 5 how difficult was the tracheostomy to manage? 1 2 3 4 5 (1 being the least difficult, 5 being the most difficult)  

Fig. 1 Survey on the left sent to all patients (n = 41) and survey on the right sent to tracheostomy patients (n = 20).
3. Results

All patients surveyed had cleft palate, glossoptosis and micro/retrognathia. 31 of the 41 patients identified participated in the survey. 25 of those responding to the survey (81%) had airway problems, and of these 22/25 (88%) required airway interventions. The failure of positioning maneuvers and a nasopharyngeal airway prompted early discussions with family members regarding options for surgical intervention. Initial alternatives offered included tracheostomy 12/31 (39%), lip–tongue adhesion 8/31 (16%) and mandibular distraction 1/31 (3%). 15/31 (48%) patients required tracheostomy; 8 were male, the other 7 female. 10/15 (67%) had iPRS and the other 5/15 (33%) sPRS. The syndromic manifestations of the sPRS patients included Stickler syndrome, fragile X syndrome, femoral facial syndrome, trisomy 11/22 with Dandy Walker malformation and seizure disorder, and Hajdu Cheney syndrome. Among the group who underwent tracheostomy 7/15 (47%) of the patients required tracheostomy longer than initially anticipated by the family; however despite these differences in actual tracheostomy duration versus family’s expectations, only two families stated that their expectations were not met with regards to tracheostomy duration. Of the 7/15 whose did not accurately anticipate actual tracheostomy tube dependence the majority did not require prolonged tracheostomy tube dependence. Only 2/7 required tracheostomy ≥36 months, one of whom remained tracheostomy dependent at the time of this survey and had been for 72 months prior, whereas the remaining 6/7 had been decannulated at a mean of 25 months. 3/7 who expected a shorter course of tracheostomy tube dependence had sPRS and the other 4/7 iPRS; however, 5/7 stated that their expectations were met despite longer periods of tracheostomy dependence relative to expected duration of tracheostomy (see Table 1).

The mean time of actual tracheostomy cannulation for these 7 patients was 32 months versus the mean parental expectation of 17 months for that group giving credence to the fact that these patients actual cannulation times were not excessively long, rather patient’s families grossly underestimated actual length of tracheostomy dependence. 9/15 (60%) had multiple hospitalizations with a mean of 8 hospitalizations per patient.

Roughly two-third of families 11/15 (73%) stated that the difficulties with tracheostomy met their expectations based on the preoperative counseling by the involved teams. 3/15 (20%), all with iPRS, underwent mandibular distraction for successful decannulation. Of the distraction patients, one had pharyngomalacia and required CO2 laser excision of suprastomal granulation tissue, another had autism and laryngomalacia, and the third had significant subglottic stenosis ultimately undergoing laryngotracheal reconstruction with rib graft.
<table>
<thead>
<tr>
<th>Patient</th>
<th>Pierre Robin sequence</th>
<th>Syndrome/comorbidities</th>
<th>Age at tracheostomy (month)</th>
<th>Age of tracheostomy removal (month)</th>
<th>Tracheostomy duration (month)</th>
<th>Expected tracheostomy duration (month)</th>
<th>Suprastomal collapse/granulation tissue</th>
<th>Laryngomalacia/tracheomalacia/pharyngomalacia</th>
<th>Repeat hospitalization</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Isolated</td>
<td></td>
<td>Day of life 6</td>
<td>Present</td>
<td>15</td>
<td>18</td>
<td>Suprastomal collapse/granulation tissue</td>
<td></td>
<td>2</td>
</tr>
<tr>
<td>2</td>
<td>Isolated</td>
<td></td>
<td>Day of life 4</td>
<td>15</td>
<td>15</td>
<td>48</td>
<td>Laryngomalacia</td>
<td></td>
<td>10</td>
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<td>3</td>
<td>Isolated</td>
<td></td>
<td>3</td>
<td>21</td>
<td>18</td>
<td>12</td>
<td>Granulation tissue</td>
<td></td>
<td>5</td>
</tr>
<tr>
<td>4</td>
<td>Isolated</td>
<td></td>
<td>21</td>
<td>40</td>
<td>19</td>
<td>12</td>
<td>Laryngomalacia/tracheomalacia/pharyngomalacia</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>5</td>
<td>Isolated</td>
<td></td>
<td>2</td>
<td>24</td>
<td>22</td>
<td>36</td>
<td>Granulation tissue</td>
<td></td>
<td>5</td>
</tr>
<tr>
<td>6</td>
<td>Isolated</td>
<td>MR; anoxia; seizure d/o</td>
<td>Day of life 14</td>
<td>24</td>
<td>24</td>
<td>24</td>
<td></td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>Isolated</td>
<td></td>
<td>2</td>
<td>30</td>
<td>28</td>
<td>36</td>
<td>Granulation tissue</td>
<td>Pharyngomalacia</td>
<td>1</td>
</tr>
<tr>
<td>8&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Isolated</td>
<td>Autism</td>
<td>Day of life 24</td>
<td>34</td>
<td>30</td>
<td>12</td>
<td>Suprastomal collapse/granulation tissue</td>
<td></td>
<td>2</td>
</tr>
<tr>
<td>9&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Isolated</td>
<td>Autism</td>
<td>Day of life 25</td>
<td>36</td>
<td>36</td>
<td>40</td>
<td>Laryngomalacia</td>
<td></td>
<td>9</td>
</tr>
<tr>
<td>10&lt;sup&gt;b, **&lt;/sup&gt;</td>
<td>Isolated</td>
<td>Trisomy 11/22; bilateral TVC paralysis</td>
<td>1</td>
<td>Present</td>
<td>72</td>
<td>24</td>
<td></td>
<td></td>
<td>4</td>
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<tr>
<td>11</td>
<td>Syndromic</td>
<td>Trisomy 11/22; bilateral TVC paralysis</td>
<td>3</td>
<td>21</td>
<td>18</td>
<td>12</td>
<td></td>
<td></td>
<td>1</td>
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<tr>
<td>12</td>
<td>Syndromic</td>
<td>Femoral facial syndrome; seizure d/o</td>
<td>3</td>
<td>24</td>
<td>24</td>
<td>36</td>
<td>Suprastomal collapse</td>
<td></td>
<td>5</td>
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<tr>
<td>13</td>
<td>Syndromic</td>
<td>Hajdu Cheney syndrome</td>
<td>1</td>
<td>30</td>
<td>30</td>
<td>24</td>
<td>Bronchomalacia</td>
<td></td>
<td>9</td>
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<tr>
<td>14</td>
<td>Syndromic</td>
<td>Fragile X</td>
<td>Day of life 59</td>
<td>36</td>
<td>35</td>
<td>36</td>
<td></td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>15&lt;sup&gt;b&lt;/sup&gt;</td>
<td>Syndromic</td>
<td>Stickler syndrome</td>
<td>Day of life 17</td>
<td>36</td>
<td>36</td>
<td>24</td>
<td>Granulation tissue</td>
<td>Laryngomalacia</td>
<td>5</td>
</tr>
</tbody>
</table>

**Abbreviations:** MR, mental retardation; seizure d/o, seizure disorder; TVC, true vocal chord.

<sup>a</sup> Underwent mandibular distraction once
<sup>b</sup> Expectations not met.
<sup>**</sup> Underwent mandibular distraction twice.
All children who required tracheostomy had significant upper airway obstruction with persistent glossoptosis lasting over 6 months following tracheostomy. Eventual decannulation occurred when this finding improved. Among the 4 patients who required prolonged tracheostomy for ≥35 months, bronchoscopy continued to show severe airway obstruction, however all but one of these patients had been decannulated at the time of survey completion. Mean duration of tracheostomy was 28 months for the entire group of 15; the early decannulation group (n = 11) had a mean time until decannulation of 22 months; the late group (n = 4) had a mean decannulation time of 45 months; the distraction group (n = 3) had a mean tracheostomy time of 31 months (see Table 2). Mean time to decannulation in iPRS (10/15) was 28 and 29 months for the sPRS (5/15) group (p = 0.465).

Of these syndromic patients only 1/5 was in the late (≥35 month) decannulation group. Complication rate after tracheostomy was 9/15 (60%), mild suprastomal collapse was noted in 3/15 (20%) and CO₂ laser excision for tracheostomy-related granulation tissue was required in 5/15 (33%). Laryngotraheal stenosis requiring laryngeal reconstruction with rib grafts was required in 2/15 (13%), both iPRS patients; repeat tracheostomy in 1/15 (7%) in an iPRS patient with autism; and tracheocutaneous fistula requiring surgical repair in 5/16 (31%), 3/5 (60%) sPRS patients and 2/10 (20%) iPRS patients. The mandibular distraction group, all with iPRS, 3/15 (20%) had a similarly high-complication rate 2/3 (67%), including tooth loss (n = 1), repeat tracheostomy (n = 1) and need for repeat mandibular surgery (n = 1). Of the three mandibular distraction patients, two had laryngeal stenosis, and one required laryngeal reconstruction. Other sites of airway obstruction were identified aside from those associated with PRS. These included: laryngomalacia in 3/15, 2/10 (20%) with iPRS and 1/5 (20%) with sPRS; pharyngomalacia in 2/10 (20%), both with iPRS; tracheomalacia in 1/10 (10%), an iPRS patient; bilateral true vocal cord paralysis in 1/10 (10%) an iPRS patient. All in all, 12/15 (80%) of the patients presented with secondary airway abnormalities categorized as suprastomal collapse, bilateral true vocal chord paralysis, granulation tissue, laryngomalacia, tracheomalacia, pharyngomalacia, and bronchomalacia: 8/10 were iPRS patients, 4/5 were sPRS patients.

4. Discussion

The management of upper airway obstruction in PRS patients varies both with the severity of respiratory distress and institutional biases for certain interventions. The long-term airway management of infants with PRS is controversial and many centers have proposed algorithms which suggest an evaluation of mandibular hypoplasia, airway distress, anatomic obstruction, and feeding through a variety of measures followed by conservative nonsurgical intervention while progressively more invasive procedures with tracheostomy being reserved for persistent, severe, or multi-leveled obstruction or syndromic patients. However, no algorithm accounts for parental expectations or satisfaction. Cruz et al. described secondary respiratory abnormalities such as central apnea, laryngomalacia, tracheomalacia, bronchomalacia, and bronchial stenosis in 11/47 (23%) of PRS patients [8]. Sher et al. commented on four different patterns of airway obstruction in PRS including tongue base obstruction, elongated velum, and pharyngeal collapse [9]. Both these studies highlight the need to endoscopically localize potential anatomic and physiologic levels of collapse/obstruction in order to best direct treatment.

In the last 10 years many centers have also reported on their efforts to avoid tracheostomy. Tracheostomy is recognized as a long-term commitment with an average age of 3.1 years at decannulation as described by Tomaski et al. [1]. Different authors have described the need for tracheostomy or endotracheal intubation in 12, 23 and 42% of PRS patients [10]. Although surveyed almost 10 years ago, 91% of pediatric otolaryngology fellowship programs felt that tracheostomy provided the safest

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Actual duration of tracheostomy in Pierre Robin sequence patients compared to parent expectations of time required to maintain tracheostomy</th>
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<tbody>
<tr>
<td>Mean duration of tracheostomy (month)</td>
<td>Mean family expectation for duration of tracheostomy (month)</td>
</tr>
<tr>
<td>Total (n = 15)</td>
<td>28</td>
</tr>
<tr>
<td>Early decannulation (n = 11)</td>
<td>22</td>
</tr>
<tr>
<td>Late decannulation (n = 4)</td>
<td>45</td>
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<tr>
<td>Distraction (n = 3)</td>
<td>31</td>
</tr>
<tr>
<td>Isolated PRS (n = 10)</td>
<td>28</td>
</tr>
<tr>
<td>Syndromic PRS (n = 5)</td>
<td>29</td>
</tr>
</tbody>
</table>

However, tracheostomy is not without complications, including those encountered early such as bleeding, pneumothorax, or pneumomediastinum; as well as late complications such as laryngeal/tracheal stenosis, pneumonia, speech delay, recurrent airway infections, stomal granulation, tracheocutaneous and tracheo-innominate fistula, interference with speech and language development, as well as feeding and swallowing difficulties. The complication incidence is as high as 29% in some reports [12], although mortality in several large series of long-term pediatric tracheostomy patients has been reported at 0.5–4% and usually is the result of accidental decannulation or tracheostomy tube plugging [5,13]. Long-term speech, behavior and developmental problems have been associated with tracheostomy as well as moderate to severe intellectual and physical impairments [14]. Adult studies have demonstrated reduced body image perception and reduced life satisfaction after long-term tracheostomy [15].

We sought to evaluate parental perceptions via a retrospective parental survey regarding the risks and benefits of tracheostomy as well as other surgical procedures such as mandibular distraction and whether their perceptions matched their expectations regarding length of treatment and the attendant problems associated with such interventions. Interestingly, almost 50% of those who responded to the survey had children who underwent tracheostomy. Two-third of the patients from those with tracheostomy surveyed had iPRS whereas the remaining third had sPRS. Anecdotally, the incidence of multi-level obstruction was high in our iPRS group (as noted above) and several iPRS patients had multiple comorbidities including autism in a distraction patient, who required repeat tracheostomy and repeat mandibular distraction; as well as profound mental retardation and seizure disorder in another iPRS patient after an anoxic brain injury. The high percentage of tracheostomy patients in the survey may also be partly explained by inherent bias in a retrospective survey in which those most affected are more likely to respond.

When considering parental dissatisfaction, the cost of multiple hospitalizations as well as anesthesia and repeated bronchoscopies are not inconsequential factors. It is also clear from reviewing our data that parental expectations largely collectively approximated the actual duration of tracheostomy dependence and yet varied widely individually. These parental estimates ranged with 5/14 (36%) families estimating ≥36 months up to 48 months and at the other end of the spectrum 5/14 estimating <20 months, and 4/5 actually retrospectively felt that their child would only require 12 months of tracheostomy dependence. It seems clear in reviewing the literature that algorithms to manage airway obstruction in PRS often overlook parental concerns and perceptions regarding the morbidity and quality of life associated with treatment. In our series 9/15 (60%) tracheostomy patients had complications that included severe laryngotracheal stenosis 2/15 (13%) and suprastomal collapse or granulation tissue in 6/15 (40%).

Physicians that manage PRS patients need to make a more concerted effort to communicate the morbidities and the potential length of tracheostomy cannulation in order to achieve reasonable expectations for the parents. Given the unmet expectation of length of tracheostomy in our series of PRS parents, a patient handout that depicts the average timeline for tracheostomy cannulation and the typical timeframe in which complications typically arise may better educate PRS parents as well as assuage any discontent regarding tracheostomy management. Tracheostomy is an effective, time-proven method for airway management of PRS patients; and not withstanding these complications, the majority of those surveyed felt that their expectations were met in terms of timing of tracheostomy dependence and ultimate ability to achieve decannulation. Further research into prognostic factors relating to duration of tracheostomy tube dependence would be helpful to enable families to more accurately anticipate length of tracheostomy dependence.

5. Conclusions

Despite the largely satisfied number of PRS patient families who responded to our survey and their ability to collectively anticipate length of tracheostomy tube dependence and the associated difficulties with tracheostomy, it seems likely that improving informed consent and preoperative tracheostomy-related education will benefit families with PRS children. This may facilitate families to better understand and more accurately anticipate the duration/difficulty of their child’s tracheostomy. Tracheostomy was associated with significant morbidity in our patient population; late mandibular distraction also had a high morbidity, in part related to prolonged tracheostomy dependence. Parents should be involved in making informed decisions along the way, understanding the inherent risks and benefits of the differing nonsurgical and surgical treatment options as well as the timing of such interventions. Providing parents with information such as the likely duration of tracheostomy
dependence is crucial in establishing realistic expectations. It was also evident that parents’ expectations were not always realistic, and we attribute this, at least in part, to inadequate counseling on the part of the involved medical teams. Interestingly, of those who expected earlier decannulation than was actually achieved, the majority of these parents did not express dissatisfaction. Tracheostomy is life-saving and frequently necessary with syndromic PRS and patients with multi-level obstruction. This data is useful in adding to the body of literature since it highlights the importance that physicians who care for infants with PRS serve as the instruments to providing parents with an accurate perception to the challenges of PRS management.

References