# Airway Management in Patients with Robin Sequence

# 11

Kevin D. Han, Mitchel Seruya, Diego A. Preciado, and Albert K. Oh

## Abbreviations

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| CNMC | Children's National Medical Center |
| CPT | Current Procedure Terminology |
| ICD – 9 | International Classifications of Diseases Version 9 |
| LOS | Length of hospital stay |
| NPA | Nasopharyngeal airway |
| RS | Robin sequence |
| SPSS | Statistical Program for Social Sciences |
| TLA | Tongue-lip adhesion |

K.D. Han, M.D. • M. Seruya, M.D.
Department of Plastic Surgery, 1PHC,
Georgetown University Hospital,
3800 Reservoir Rd, NW, Washington, DC 20007, USA
e-mail: kdh8@gunet.georgetown.edu; mseruya@cnmc.org

D.A. Preciado, M.D., Ph.D.
Department of Otolaryngology, Pediatrics,
and Integrative Systems Biology,
The George Washington University School of Medicine,
Washington, DC, USA

Division of Pediatric Otolaryngology, Department of Otolaryngology, Children's National Medical Center,
111 Michigan Ave NW, Washington, DC 20010, USA
e-mail: dpreciad@cnmc.org

A.K. Oh, M.D. (✉)
Department of Surgery and Pediatrics,
The George Washington University School of Medicine,
Washington, DC, USA

Department of Plastic and Reconstructive Surgery,
Children's National Medical Center,
111 Michigan Ave NW 4th floor West Wing,
Washington, DC 20010, USA
e-mail: aoh@cnmc.org

## 11.1 Introduction

Robin sequence (RS), the clinical triad of micrognathia (small jaw), glossoptosis (downwardly displaced tongue), and upper airway obstruction, affects approximately 1 in 8,500 births (Bush and Williams 1983; Sadewitz 1992). Cleft palate is also noted in up to 90.4 % of patients (Caouette-Laberge et al. 1994). Infants may present with a wide phenotypic variability, ranging from infrequent episodes of airway obstruction and/or feeding difficulty to severe crises of asphyxia and failure to thrive (Marques et al. 2001). The latter group of patients is at particular risk for hypoxic brain damage, impaired mental development, pulmonary hypertension, aspiration pneumonia, and failure to thrive (Tomaski et al. 1995; Hoffman et al. 1965; Kapp-Simon and Krueckeberg 2000). Despite advances in critical care medicine, mortality is not inconsequential and ranges from 0 to 13.6 % (Sadewitz 1992; Marques et al. 2001; Caouette-Laberge et al. 1994; Cruz et al. 1999; Dykes et al. 1985). Such high morbidity and mortality have been attributed to late diagnosis, delayed airway protection, and multisystem disorder. Furthermore, care of infants with RS can require prolonged hospitalization, with averages ranging from 10 to 60 days and translate into increased costs (Bull et al. 1990; Cruz et al. 1999; Matsas et al. 2004; Wagener et al. 2003).

Several modalities have been proposed to address airway obstruction in infants with RS. Options range from conservative management, namely, prone positioning, nasopharyngeal airway (NPA) placement, and orthopedic devices to

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operative interventions, including subperiosteal release of floor of mouth, tongue-lip adhesion (TLA), tracheostomy, and mandibular distraction osteogenesis (lengthening of the mandible).

The decision on how to manage these infants is often based on the experiences of the provider and the practices at a particular center. Therefore, it is not surprising to see the management of such infants continues to be controversial. Prior studies have attempted to establish evidence-based parameters to help clinicians to devise management plans for these infants. Typically, the management of infants with RS focuses on avoiding tracheostomy either conservatively or surgically, providing adequate respiration and nutrition and preventing long-term sequelae and death. However, by bypassing the obstruction, tracheostomy remains the most definitive treatment for infants with severe respiratory obstruction that is not compatible with life.

Although it is known that tracheostomy in these infants is a long-term commitment with a reported average age of 3.1 years at decannulation (Tomaski et al. 1995; Moyson 1961; Sadewitz 1992), studies on the chronology of decannulation in patients with RS are lacking overall. It is still unclear whether natural "mandibular growth" and time allow for decannulation in patients without further intervention can occur. As part of an overall institutional effort to establish evidence-based guidelines for surgical intervention in patients with RS, we present here a cohort of patients with severe upper airway obstruction treated with tracheostomy to determine length of time for decannulation without further surgical intervention (i.e., "natural" decannulation) and to investigate potential factors associated with successful "natural" decannulation.

## 11.2 Methods

The records were reviewed for patients who underwent primary airway management at Children's National Medical Center (CNMC) from 1994 to 2010. All study subjects were identified within the accounting departments of *Otolaryngology* and *Plastic and Reconstructive Surgery* using International Classifications of Diseases Version 9 (ICD-9 524.00, 524.06, and 524.10) related to diagnosis of anomaly of jaw size and Current Procedure Terminology (CPT 41510, 20690/20692, and 31600/31603/31605) which include tongue-lip adhesion (TLA), mandibular distraction, and tracheostomy.

Inclusion criteria were patients with documented micrognathia, glossoptosis, and respiratory obstruction who have received a tracheostomy at Children's National Medical Center. The following children were excluded: (a) respiratory difficulties other than upper airway obstruction, (b) patients who were treated at CNMC but with missing or incomplete medical records, and (c) patients of CNMC but who have received definitive airway management at another hospital.

Within the subset of the patients who have received tracheostomy, their demographics, nutritional and respiratory status, laboratory values, and polysomnographic (sleep) studies were reviewed. Perioperative and postoperative complications include tracheitis (inflammation of the trachea), pneumonia, breakdown, stoma infection, hematoma, reoperation, and finally long-term outcomes such as developmental delay, organ systems dysfunction (neurogenic, gastrointestinal, and/or cardiopulmonary), and death were recorded. Finally, the timing of events was investigated. Length of hospital stay (LOS) subdivided into pre-tracheostomy, postoperative hospital stay, and total length of hospital stay was noted. The duration of tracheostomy to "natural" decannulation was also recorded.

Data were analyzed with Statistical Program for Social Sciences (SPSS) version 16.0 software (Chicago, IL) and Microsoft Excel version 2008 software (Redmond, WA). Univariate analysis included chi-squared and Fisher's exact tests for contingency data. Kaplan–Meier curves with log-rank (Mantel–Cox) test were used to estimate the percent of patients on tracheostomy as a function of a time.

## 11.3 Results

Of 61 infants with RS, 25 infants received a tracheostomy. The other 36 infants were managed with lateral/prone positioning, noninvasive

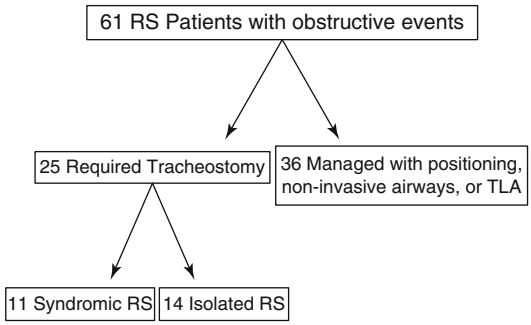


**Fig. 11.1** Of 61 patients with obstructive events, 25 required tracheostomy, of which 11 were syndromic (Stickler syndrome (a group of genetic disorders affecting connective tissue), Treacher Collins (congenital craniofacial syndrome), and others) and other 14 patients had isolated Robin sequence

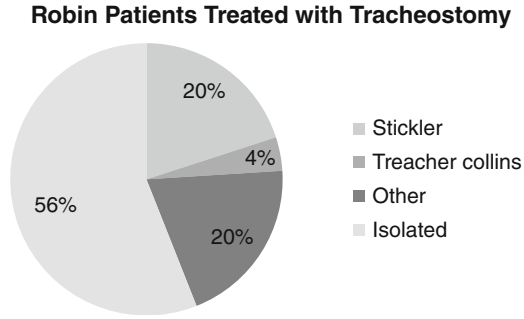


**Fig. 11.2** Of 25 patients treated with tracheostomy, 44 % (11 cases) were syndromic, of which 45 % (5 cases) was either Stickler or other syndromes and 10 % (1 case) was Treacher Collins and the other 56 % (14 cases) had isolated RS

oxygen supplement, or TLA in Fig. 11.1. Among the cohort requiring tracheostomy, 14 patients (56 %) were isolated and 11 patients (44 %) were syndromic as shown in Fig. 11.2. Overall, the median time to decannulation was 97 months in Fig. 11.3. The few patients with syndromic RS who were successfully decannulated required a median time >73 months as compared to patients with isolated RS who had a median time to decannulation of 19 months in Fig. 11.4. In total, 13 out of 25 infants (52 %) were successfully decannulated without further surgical intervention; only two patients were syndromic and the remaining 11 patients were isolated.

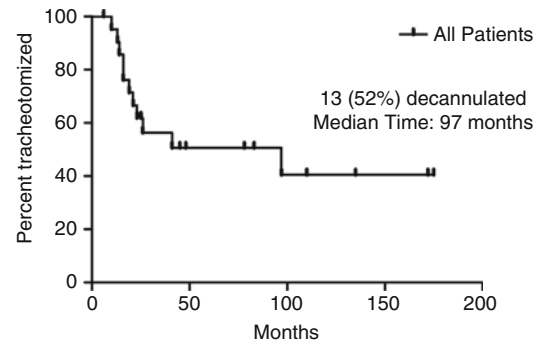


**Fig. 11.3** Of 25 infants treated with tracheostomy, 13 infants (52 %) were eventually decannulated with median time to decannulation of 97 months

At mean follow-up of 4 years, the rate of tracheostomy-specific complications (e.g., cannula obstruction or accidental decannulation, delays in speech and language development, tracheomalacia (flaccidity of the tracheal support cartilage)/tracheitis, pneumonia) was 52 % and tracheostomy-specific mortality was 8 %. Patients with syndromic RS stayed in the hospital significantly longer than patients with isolated RS (50 versus 28 days, respectively). There was one death in each group; however, patients with syndromic RS had significantly more events of end-organ dysfunction (neurogenic, gastrointestinal, and/or cardiopulmonary) per patient than patients with isolated RS (2.08 versus 0.69, $p=0.005$).

## 11.4 Discussion

Based upon the literature, approximately 2/3 of patients with RS can be successfully managed with conservative therapies (Kochel et al. 2011; Gozu et al. 2010; Evans et al. 2006; Horikiri et al. 2010). Prone positioning is often sufficient in less severely affected patients but may result in prolonged hospitalization (Kochel et al. 2011; Sher 1992; Bhat et al. 2006). NPA can be effective in during the early clinical course to avoid emergent tracheostomy, but difficultly maintaining proper position for an extended period of time has limited its use (Chang et al. 2000; Sher 1992; Kochel et al. 2011; Masters et al. 1999; de Buys Roessingh et al. 2007). Lastly, orthopedic devices have been

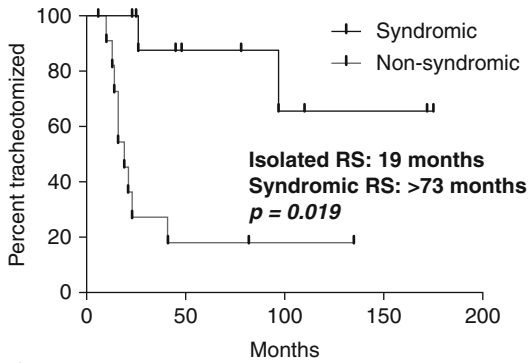


**Fig. 11.4** Patients with syndromic RS had a median time to decannulation of >73 months versus patients with isolated RS had a median time to decannulation of 19 months, $p = 0.019$

tried with success in a few small studies but these devices are often expensive (Hotz and Gnoinski 1982; Buchenau et al. 2007; Kochel et al. 2011). More recently, Kochel et al. described the usage of newer orthopedic devices as another noninvasive way to treat upper airway obstruction. Three major types (plate with a posterior wire spur, plate with a posterior acrylic extension, and plate with a pharyngeal tube) of orthopedic devices were used in seven patients based on their mechanisms of the obstruction (Sher's classifications; see Sect. 11.4). All of the seven patients had normal oxygen saturation upon discharge and at the end of study period, five out seven patients tolerated the removal of the orthopedic devices (Kochel et al. 2011).

Surgical modalities such as glossopexy or TLA, mandibular distraction, mandibular extension, and subperiosteal release of floor of mouth muscle have been studied extensively for their effectiveness in avoiding or delaying tracheostomy, improving polysomnographic results, facilitating feeding, and bypassing or correcting the anomalous anatomy. TLA, subperiosteal release of floor of mouth muscle, and mandibular distraction have been described most frequently in the recent literature. TLA temporarily bypasses the obstruction by creating a stable airway with reported success rates between 70 and 100 % (Denny et al. 2004; Bijnen et al. 2009; Kirschner et al. 2003). Dehiscence (breaking down of the incision) still ranges between 0 and 57 % with a mean of 30 % (Sher 1992; Marques et al. 2001; Bookman et al. 2011). Based on a few studies, the need for a secondary invasive airway procedure such as tracheostomy after subperiosteal release ranges between 10 and 100 % (Delorme et al. 1989; Caouette-Laberge et al. 1996; Breugem et al. 2008; Siddique et al. 2000). Mandibular distraction has been shown to improve polysomnographic outcome, to avoid or delay tracheostomy, and to expedite decannulation of a tracheostomy (Scott et al. 2011; McCarthy et al. 1992; Denny et al. 2001; Lin et al. 2006; Schaefer et al. 2004; Schaefer and Gosain 2003). Nevertheless, complications such as infection, device failure, and nonunion occur from 2.5 to 52 % (Caouette-Laberge et al. 1994; McCarthy et al. 2002; Shetye et al. 2009). One large retrospective review of 141 infants with RS who underwent mandibular distraction reported a 52 % overall complication rate with a 5 % major complication rate – a complication that necessitated a secondary invasive therapy (Shetye et al. 2009).

Indications for surgical airway management are also highly debated. While many clinicians rely on a "gestalt" impression of airway obstruction, some have proposed data-driven clinical parameters for airway management. Caouette-Laberge et al. and Cole and colleagues proposed grading systems based on the settings of respiratory obstruction (Caouette-Laberge et al. 1994; Cole et al. 2008). Caouette-Laberge grouped 125 infants with RS into three categories: (1) adequate respiration in prone position and regular bottle-feeding, (2) adequate respiration in prone position and difficulty with feeding, and (3) endotracheal intubation (Caouette-Laberge et al. 1994). Parsons and Smith provided rule of thumb criteria for TLA in infants with RS; those who had progressive weight/strength gain over a 7-day period did not require TLA, while those infants who needed more than 3 days of endotracheal airway support should receive TLA (Parsons and Smith 1982). Freed et al. reported on the use of bedside monitoring and polysomnography to objectively guide airway management for infants with RS (Freed et al. 1988). Criteria for TLA included (1) an average transcutaneous $O_2$ level below 60 mmHg or transcutaneous $CO_2$

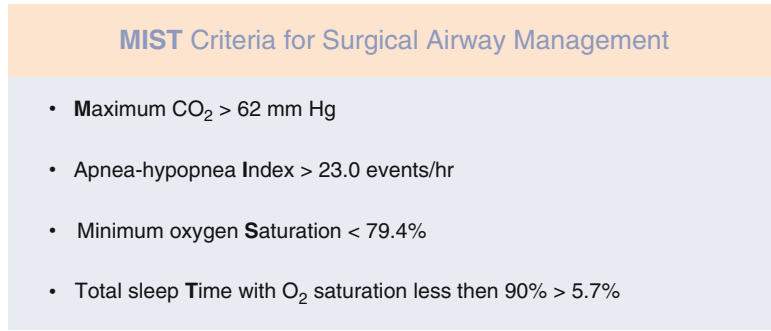


**Fig. 11.5** *MIST* criteria for surgical airway management of patients with Robin sequence (*RS*)

level over 50 mmHg for a minimum of 8 h, (2) obstructive episodes on sleep study, and (3) oxygen saturation below 80 %.

More recently, Rogers et al. described the GILLS scoring system where one point was assigned to each of the five variables: *g*astroesophageal reflux, *i*ntubation preoperatively, *l*ate operation, *l*ow birth weight, and *s*yndromic diagnosis. TLA was 100 % successful in infants with a GILLS score of 2 or less but failed in 43 % of infants with a score of 3 or more (Rogers et al. 2011). Sher detailed the use of flexible fiber optic endoscopy to guide the form of surgical treatment in infants with RS failing conservative measures. Based upon nasopharyngoscopy and identification of the site of obstruction, infant airways were classified into four groups. Type I represented airway obstruction solely due to glossoptosis, while types II–IV had additional components of obstruction. Regardless of airway classification, all patients were initially treated with NPA placement for up to 8 weeks. If conservative therapy failed, type I patients received TLA and types II–IV underwent tracheostomy (Sher 1992). Schaefer et al. described an algorithm for approaching respiratory and nutritional dysfunction in infants with RS. Management decisions were based upon the ability to maintain progress on the growth curve, continuous pulse oximetry and bedside polysomnography, and the site of airway obstruction (Schaefer et al. 2004). Finally, data from our own institution (Fig. 11.5) from a retrospective review of airway management in patients with RS from 1994 to 2010 found that four clinical factors (*MIST* criteria) were most associated with surgical airway management: *m*aximum $CO_2 > 62$ mmHg, apnea–hypopnea *i*ndex (index of sleep apnea severity) > 23.0 events/h, minimum $O_2$ *s*aturation < 79.4 %, and greater than 5.7 % total sleep *t*ime with $O_2$ saturation less than 90 %. Each of these parameters identified operative intervention with 75–85 % accuracy (Seruya et al. 2011).

Despite the various procedures described for surgical airway management, tracheostomy remains the most definitive treatment for infants with severe respiratory obstruction that is not compatible with life. In many cases, tracheostomy is believed to be a temporary measure until "natural" mandibular growth permits decannulation. The topic of mandibular growth in patients with RS has been heavily debated yet remains poorly defined. Some authors have documented diminished mandibular size and proportions compared age-adjusted norms while others have cited evidence to the contrary (Shen et al. 2010; Hermann et al. 2003; Daskalogiannakis et al. 2001; Figueroa et al. 1991). Rogers et al. found that mandibular length was shorter in all patients with RS irrespective of the type of airway management, and the differences in both mandibular length and sagittal position varied significantly among all the syndromic subtypes (Rogers et al. 2009). Maalouf et al. found that 60 % of patients who received bilateral mandibular distraction maintained proportionate facial symmetry at the median follow-up time of 57 months; mandibular size, however, was not evaluated (Maalouf and Lehman 2011). Finally, Pruzansky and Richmond demonstrated the opportunity for "catch-up" growth of the mandible without the need for invasive procedures aside

from a temporary tracheostomy (Pruzansky and Richmond 2005). Overall, these studies highlight the phenotypic heterogeneity of patients with RS, which may stem from the dissimilar mandibular growth kinetics of isolated versus syndromic patients.

In our retrospective study of 61 infants with RS, 25 patients required tracheostomy (14 cases of isolated RS and 11 cases syndromic RS). This is one of the largest published cohorts of RS children with tracheostomy that have been followed to analyze rates of eventual decannulation due to mandibular growth without other ancillary surgical interventions. The median time to decannulation in our cohort was longer (97 months) than what other smaller series have reported (Tomaski et al. 1995; Demke et al. 2008). Patients with syndromic RS largely contributed to this extended time course, as most of them could not be decannulated by the completion of the study. Outcomes following tracheostomy were significantly poorer in patients with syndromic RS as compared to those with isolated RS: patients with syndromic RS had longer hospital stay and more long-term complications as compared to patients with isolated RS.

## Summary and Conclusion

Airway management in patients with RS and severe airway obstruction remains controversial. The benefits of various surgical airway procedures, as well as indications for intervention, are unclear and based upon imperfect data. Our experience with these complex patients has documented some factors associated with the need for surgical intervention, but these criteria await validation in a randomized prospective trial. We have also found that although tracheostomy was intended to be a temporary airway for these patients, the time to natural decannulation was longer than expected. This may be partially explained by the mandible's inability to "catch up" in growth, especially in syndromic patients. We are planning further study into these issues by means of a prospective trial that will incorporate serial lateral cephalograms to document mandibular growth as well as offer mandibular distraction as a surgical modality to patients with severe airway obstruction. Based on our experience, we believe that all potential treatment options should be exhausted before offering tracheostomy to syndromic patients with Robin sequence.

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**Conflict of Interest**
None

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