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# Primary aerodigestive presentations of Pierre Robin sequence/complex and predictive factors of airway type and management☆

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ABSTRACT

*Objective:* To document the mode and age of primary aerodigestive presentation of Pierre Robin sequence/complex (PRS/C) children to the otolaryngologist and to explore predictive factors of upper airway type and management.
*Methods:* This is a retrospective cohort study conducted in a tertiary pediatric referral center. A prospective surgical database was searched for children who were diagnosed with PRS/C. Demographics, presenting complaint, secondary diagnoses, type of upper airway obstruction, secondary airway lesions, presence of cleft palate, and airway interventions were collected. Multiple linear regression analysis was performed to predict upper airway obstruction type and intervention.
*Results:* Seventy-seven potentially eligible patients were identified. Forty-six were included (20 females). Mean age at presentation was $20.4 \pm 36.9$ months (range 1–191.25 months). Twenty-three primarily presented with respiratory failure, 14 with sleep disordered breathing, and nine with swallowing dysfunction. Children with presentations other than respiratory failure were older ($p = 0.004$). Nineteen were syndromic. Overt cleft palate was more common in those presenting with respiratory failure ($p = 0.01$). The type of airway obstruction encountered and use of tracheostomy were positively predicted by the primary presenting feature of respiratory failure ($p < 0.05$) and male gender ($p < 0.05$).
*Conclusion:* A substantial number of PRS/C patients present later than the neonatal period with presentations other than respiratory failure. Both male gender and presentation with respiratory failure predicted a more severe airway obstruction type and the need for tracheostomy.


## 1. Introduction

Pierre Robin sequence or complex (PRS/C) describes a triad which includes retro or microagnathia, glossoptosis, and airway compromise [1]. Pierre Robin sequence (PRS) is thought to consist of multiple secondary anomalies derived from a single primary anomaly or mechanical factor, retrognathia [2]. The term Pierre Robin complex (PRC) describes the presence of this triad secondary to micrognathia as one of several congenital anomalies. Although not originally described by Robin as part of the sequence, cleft palate is often described as a defining feature [1,3–5]. Several authors have described variations in the definitions of PRS/C, all of which required mandibular deficiency but some of which did not include cleft palate or airway obstruction [6,7]. PRS/C is not only causally heterogeneous, but pathogenetically and phenotypically variable [7]. Children with PRS/C may have a variety of presentations to an otolaryngologist including hearing deficits [8], swallowing dysfunction (SD) [9,10], and sleep disordered breathing (SDB) or obstructive sleep apnea (OSA) [11–13]. The age at which these children present is variable.

The management of PRS has been described in numerous case series [1,4,5,14–18]. Despite agreement on the general principles

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of management, there is a great degree of variability in the specifics related to controlling the airway between centers. This ranges from positioning strategies to surgical interventions from tongue–lip adhesion, subperiosteal release, distraction osteogenesis, use of mandible traction wires, all the way to tracheostomy. These children also often encounter difficulty with swallowing and may require nasogastric tube feeds or gastrostomy tube placement.

While the literature contains a plethora of articles on airway management and swallowing issues, epidemiological information concerning the first presentation and its timing are scarce. Our aim was to report on the primary aerodigestive presentation and age at diagnosis of a consecutive series of patients diagnosed with PRS/C referred to a single tertiary care pediatric otolaryngology practice. We also sought to identify predictive factors for airway type and need for tracheostomy.

## 2. Materials and methods

A retrospective cohort study was conducted at a tertiary pediatric center (the Stollery Children's Hospital in Edmonton, Alberta, Canada) from the case-load of one pediatric otolaryngologist (HE). Approval for the study was obtained from the University of Alberta Research Ethics Board. Patients were identified from a prospectively kept surgical database (Microsoft Access 2002). This database contains demographic information, diagnoses, operative procedures performed, anesthetic mode, and complications of patients undergoing any procedure in the operating room (all surgical patients in the practice). The time period between 25th June 2002 and 27th May 2010 was searched. Both electronic and a manual searches were performed to ensure accurate identification and to avoid duplication.

All children (aged 17 or younger) who were diagnosed with PRC/S or a condition known to be associated with it [7] were *eligible*. As part of routine care, airway endoscopy is performed on these children in this pediatric otolaryngologist's practice. Only cases that underwent airway endoscopy, had documented micro/retrognathia, and airway compromise, with or without cleft palate [19], were *included*. The patients whose diagnoses were not confirmed by reports and airway endoscopy or had insufficient data for the main outcome measures were *excluded*.

Data was collected from three sources: the surgical database, hospital charts, and private medical records (including operative records and digital video recordings). The patients' age at diagnosis, gender, and mode of presentation at the time of first consultation (airway compromise, swallowing dysfunction (SD), or sleep disordered breathing (SDB)) were recorded. Other variables collected included whether the patient qualified for a complex or sequence diagnosis, the presence or absence of overt or submucous cleft palate, secondary diagnoses (otolaryngological, secondary, or syndromic), sleep study results, type of airway compromise, secondary airway lesions, and airway management (conservative, tongue–lip adhesion, or tracheostomy). The data collected was entered into individual forms and then transferred to relational database software spreadsheets.

The mode of presentation was defined as the primary presenting symptom that was of greatest concern to the parents, care givers, and/or their referring physician. SD was defined as choking, coughing, congestion, or change in respiratory status during feeds, with or without failure to thrive. SDB was defined as snoring, disrupted sleep related to breathing difficulty, and other nocturnal or diurnal symptoms. Respiratory failure (RF) included those patients failing to maintain respiration without intervention. Airway compromise was categorized as one of four types according to flexible endoscopy findings (awake or sedated and breathing spontaneously). The types were defined according to Sher et al. [11] as tongue collapse against the posterior pharyngeal wall (type 1), with or without trapping of the soft palate (type 2), and lateral (type 3) or circumferential (type 4) pharyngeal collapse. This grading assumes increasing severity from 1 to 4. Documentation of airway endoscopy findings was confirmed by reviewing video records for individual subjects.

Conservative management options included positioning, the use of nasopharyngeal tubes, alternate route of feeding, or no intervention. Those who failed these measures were offered tongue–lip adhesion. If the patient failed the latter options, had an obvious concomitant neurological or respiratory pathology that deemed respiratory failure a certainty, or had multi-level airway pathology, a tracheostomy was offered. In principle, the cornerstone of this practice is to avoid a tracheostomy if possible.

### 2.1. Outcome measures

#### 2.1.1. Primary

1. The distribution of patients by primary presentation and age.
2. The presence of overt and submucosal cleft palate, airway types, and distribution into complex or sequence, by primary presentation.
3. Comparison of proportions of early and late presenters among overt cleft and submucous cleft or intact palate children.

### 2.2. Secondary

Identify potential predictors of:

1. Severe airway compromise.
2. Requirement of tracheostomy.

### 2.3. Statistical analysis

Descriptive statistics were calculated. Comparison of proportions was performed using $\chi^2$ or Fisher's exact test. Student's *t*-test was used to compare means. Multiple linear regression analysis was performed. The independent variables used were type of airway compromise (1–4) and whether tracheostomy was required, respectively. The dependent variables included secondary airway lesions (absent or present), other associated diagnoses (absent or present), primary presentation (respiratory failure, swallowing dysfunction, or sleep disordered breathing (ordinal scale 1–3), age in months, and gender (male or female).

Statistical significance was defined as $p < 0.05$ in all cases. Data analysis was performed on SigmaStat and SigmaPlot version 3, Systat Software Inc, San Jose, CA, and http://www.quantitativeskills.com/sisa/.

## 3. Results

Over the 8-year period, 4576 cases were prospectively logged into the surgical database. Of these, a total of 77 consecutive patients were identified in whom the primary diagnosis was PRS/C or a syndrome known to be associated with PRS/C [7]. Of these, 46 patients met our inclusion criteria (Fig. 1). Thirty-one were excluded: one due to incomplete data, and 30 due to not meeting the inclusion criteria. A large number of patients did not meet the inclusion criteria as the potentially eligible group included patients with syndromes associated with PRS/C who did not subsequently receive a PRS/C diagnosis. In the 46 included patients, the mean age was 20.4 ± 39.6 months (range from 0.5 days to 15 years). Twenty-six (57%) were male and 20 were female, giving a male-to-female ratio of 1.3:1. Nineteen were identified as PRC, of which five had