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Prognosis of airway obstruction and feeding difficulty in the Robin sequence☆

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Summary

Objective: To evaluate the course and prognosis of airway obstruction and feeding difficulty in the Pierre Robin sequence (PRS).
Methods: Retrospective review of 60 patients with PRS between 1993 and 2002 at the University of California, Davis Medical Center. Patients were placed into diagnostic subgroups: (1) Isolated PRS; (2) Syndromic PRS (known syndrome with PRS); (3) Unique PRS (unique anomalies with PRS). Data regarding severity, duration, and management of airway obstruction and feeding difficulty were collected.
Results: Airway obstruction requiring intervention beyond positional therapy was seen in 28% isolated, 42% syndromic, and 58% unique PRS. One-third of patients who failed positional therapy were temporarily stabilized with a nasopharyngeal airway or endotracheal intubation. The remaining two-thirds of patients, who failed positional therapy required a surgical airway procedure. Four patients underwent mandibular distraction osteogenesis, resulting in successful decannulation or avoidance of tracheostomy. Thirteen patients underwent tracheostomy; mean duration of tracheostomy-dependence was 17.0 months in Isolated PRS and 31.7 months in Unique PRS ($p < 0.01$). Successful decannulation by age of 3 years was confirmed in 85% of patients who underwent tracheostomy.

Tube feeding was required in 53% Isolated, 67% Syndromic, and 83% Unique PRS. Forty-two percent of PRS patients with a successful positional airway still demonstrated feeding difficulty. Short-term (0—3 months) and intermediate (4—18 months) tube feeding was more commonly required in Isolated and Syndromic PRS, while long-term (beyond 18 months) gastrostomy tube feeding was more commonly required in

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Unique PRS ($p < 0.01$). By 3 years of age, a successful oral diet was seen in 91% Isolated, 92% Syndromic, and 78% Unique PRS.
Conclusions: Diagnostic subgroups based on the presence of additional anomalies help families and physicians in understanding the severity and duration of feeding and airway difficulty in PRS. Two-thirds of PRS patients who fail positional therapy may ultimately require a surgical airway procedure. Feeding difficulty can be present in the absence of clinically significant airway obstruction. Families and physicians should be encouraged that by 3 years of age, most patients were successfully taking an oral diet without airway obstruction.

1. Introduction

Newborn infants with Pierre Robin sequence (PRS) demonstrate varying degrees of respiratory distress and feeding difficulty secondary to retrognathia and posterior displacement of the tongue base [1]. The severity of both airway obstruction and feeding difficulty in PRS is also related to the presence of additional congenital anomalies. Placing PRS patients into diagnostic subgroups based on the presence of additional anomalies helps the clinician predict the severity and duration of airway and feeding difficulty [2,3]. This study was undertaken in an effort to determine the prognosis of airway obstruction and feeding difficulty in patients with PRS at the University of California, Davis Medical Center.

2. Methods

A retrospective review was performed of patients diagnosed with PRS at the University of California, Davis Medical Center, between 1993 and 2002. Under the discharge diagnosis, ''Anomalies of the Skull and Facial Bones'', a total of 60 PRS patients were identified with medical records adequate for review. Data regarding the severity, timing, duration, and management of both airway and feeding difficulty were collected. The presence of additional congenital anomalies or syndromes was recorded. Patients with retrognathia or micrognathia and glossoptosis with cleft palate and/or airway obstruction were labeled 'Isolated PRS'. Patients with a known syndrome associated with PRS were labeled 'Syndromic PRS'. Patients who demonstrated a collection of otherwise unique congenital anomalies were labeled 'Unique PRS'. Data were analyzed in consultation with a biostatistician using Fisher's exact test (modified for 3×2 tables) and unpaired two-tailed Student's *t*-test. Approval of the study protocol was obtained from the Human Subjects Review Committee of the Institutional Review Board at the University of California, Davis.

3. Results

Sixty patients with PRS and adequate medical records were identified, with 60% Isolated PRS ($n = 36$; 19 females), 20% Syndromic PRS ($n = 12$; six females), and 20% Unique PRS ($n = 12$; six females) (Fig. 1). Named syndromes associated with PRS included Stickler syndrome ($n = 5$), fetal alcohol syndrome ($n = 2$), velocardiofacial syndrome, Treacher Collins syndrome, Nager acrofacial dysostosis, terminal chromosome 4q deletion, and methotrexate-associated dysmorphia. Unique PRS patients demonstrated a variety of abnormalities that did not meet criteria for named syndromes.

Sixty-three percent (38/60) of PRS patients demonstrated a successful prone positional airway. Airway obstruction requiring intervention beyond prone positioning was seen in 28% (10/36) Isolated, 42% (5/12) Syndromic, and 58% (7/12) Unique PRS (Fisher's exact test, $p = 0.15$; see Fig. 2). Twelve percent of PRS patients (two Isolated, three Syndromic, and two Unique PRS) failed positional therapy and were temporarily stabilized with endotracheal intubation or nasopharyngeal airway placement, thus avoiding a surgical airway procedure. A surgical airway procedure consisting of tracheostomy and/or mandibular distraction osteogenesis

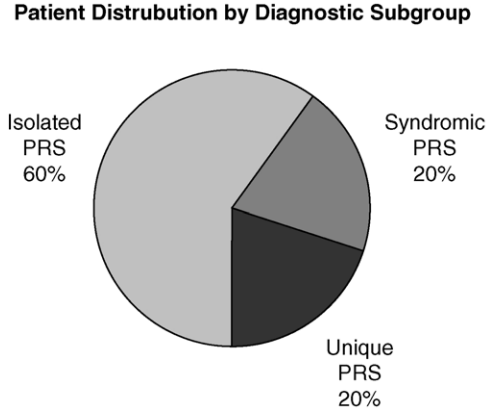

Fig. 1 Distribution of Pierre Robin sequence (PRS) patients by diagnostic subgroup.