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Speech prognosis and need of pharyngeal flap for non syndromic vs syndromic Pierre Robin Sequence

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Abstract

Background: The aim of this retrospective study was to evaluate speech outcome and need of a pharyngeal flap in children born with nonsyndromic Pierre Robin Sequence (nsPRS) vs syndromic Pierre Robin Sequence (sPRS).

Methods: Pierre Robin Sequence was diagnosed when the triad microretrognathia, glossoptosis, and cleft palate were present. Children were classified at birth in 3 categories depending on respiratory and feeding problems. The Borel-Maisonny classification was used to score the velopharyngeal insufficiency.

Results: The study was based on 38 children followed from 1985 to 2006. For the 25 nsPRS, 9 (36%) pharyngeal flaps were performed with improvements of the phonatory score in the 3 categories. For the 13 sPRS, 3 (23%) pharyngeal flaps were performed with an improvement of the phonatory scores in the 3 children. There was no statistical difference between the nsPRS and sPRS groups ($P = .3$) even if we compared the children in the 3 categories ($P = .2$).

Conclusions: Children born with nsPRS did not have a better prognosis of speech outcome than children born with sPRS. Respiratory and feeding problems at birth did not seem to be correlated with speech outcome. This is important when informing parents on the prognosis of long-term therapy.

Pierre Robin Sequence (PRS) occurs once in every 8500 births. The term sequence is related to a cascade of events during embryological developmental starting with mandibular hypoplasia (micrognathia), which causes the abnormal posterior placement of the tongue (glossoptosis), which in turn prevents full closure of the palate (cleft palate [CP]). Palatal clefting can be U-shaped or V-shaped and can include the soft palate only or both the soft and hard palates [1]. Most teams consider that respiratory and feeding problems at birth

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are part of the sequence, but the severity of airway obstruction varies considerably in infants with PRS [2-4]. Severe cases of PRS show respiratory distress owing to glossoptosis resulting in carbon dioxide retention at birth and failure to thrive. After birth, rapid mandibular catch-up growth and improved coordination of the velopharyngeal muscles reduce airway obstruction, and the prognosis for these children is good. Caouette-Laberge et al [5] classified 3 categories of patients based on their respiratory and feeding problems at birth: adequate respiration in prone position and adequate bottle feeding (category I); adequate respiration in prone position but feeding difficulties requiring gavage (category II); respiratory distress requiring endotracheal intubation and feeding difficulties requiring gavage (category III).

The speech of children born with CP can be characterized primarily by abnormal nasal air emission. It is due primarily to velopharyngeal insufficiency (VPI), which diminishes oral volume and can cause articulation errors and misarticulations that decrease overall intelligibility. Evaluation of speech problems and especially VPI can be done subjectively by perceptual evaluation or measured by pressure-flow technology. The ideal method of evaluation must be reliable, reproducible, capable of grading the severity of VPI, practical, and noninvasive. No single technique provides reproducible results, and many teams evaluate the children using a combination of perceptual evaluation, nasometry, hearing records, and videonasopharyngeal endoscopy (VNPE) [6,7]. In French-speaking countries, the reference for the evaluation of VPI or nasal air emission is usually the Borel-Maisonny score (Table 1) [8].

It is generally believed that treatment of velopharyngeal dysfunction should be initiated as early as possible. Exercises to strengthen the velopharyngeal muscles can be performed with children as young as 12 months ("guidance"). If speech therapy is unsuccessful, velopharyngeal dysfunction can be treated prosthetically or surgically [9]. Surgical procedures include the enlargement of the posterior pharyngeal wall with various injectable materials, the lengthening of the palate by pushback palatoplasty and pharyngeal flap, sphincter pharyngoplasty or pharyngeal flap alone [10,11]. Pharyngeal flaps can be based superiorly, inferiorly, or laterally [12,13]. Indications for surgery must take into account the social impact of the child's speech and the age of the child because the recovery period is quite uncomfortable.

Table 1 Borel-Maisonny classification

Type 0	No phonation
Type 1	Excellent phonation, no nasal air emission
Type 1/2	Good phonation, intermittent nasal air emission, good intelligibility
Type 2	Phonation with continuous nasal emission
Type 2B	Phonation with continuous nasal emission but good intelligibility and no social discomfort
Type 2M	Phonation with continuous nasal emission, poor intelligibility
Type 2/3	Phonation with continuous nasal emission with compensatory articulation, poor intelligibility
Type 3	Continuous compensatory articulation, no intelligibility

The purpose of this study is to evaluate and compare speech outcome in children born with nonsyndromic PRS (nsPRS) vs syndromic PRS (sPRS), including those who required cranial-based pharyngeal flap.

1. Methods

We identified all children with a PRS born between 1985 and 1998 in our hospital. Pierre Robin Sequence was diagnosed when the triad microretrognathia, glossoptosis, and CP were present. We divided our patients in 2 groups: nonsyndromic and syndromic patients. The definition of syndromic patient was based on the presence of other anomalies. All children, whether syndromic or nonsyndromic, were classified at birth in 3 categories: category I, those with slight respiratory and/or feeding problems; category II, those with adequate respiration in prone position but feeding difficulties; category III, those with severe respiratory and feeding problems requiring pharyngeal tube. The classification of the children in 1 of the 3 categories was based on a combination of clinical observation, continuous need of oxygen, and monitoring.

All children were fitted with a removable palatal appliance within 1 week after birth. The palatal clefts involved the velum and the hard palate to varying degree. The first incision was done along the free margin of the cleft anterior to posterior to the uvula. The second lateral incision passed from the region of the pterygoid hamulus (which was fractured) transversely along the palatine shelf and parallel to the cleft anteriorly. The width of the 2 flaps of palatine mucosa and periostum was measured so as to lift the minimum necessary to rotate and close the cleft. The anterior tips of the flaps were left attached to improve vascularization of the flaps. A 2-layer suture was performed in all cases where the width of the cleft was not excessive and nasal mucosa was of good quality, sometimes using vomerian mucosa.

They were seen by the pediatric surgeon 6 months after surgical closure of the palate, and then once a year up to the age of 3.5 years. Hearing was checked once a year by the pediatric ENT specialist. When the child was 1 year old, parents were provided with strategies to encourage babbling and early verbal communication. Pediatricians were instructed in the special care needed to avoid chronic serous otitis media and risk of hearing loss in their cleft patients and were asked to perform routine otoscopy and tympanometry. At the age of 3.5 years, the child was evaluated by our multidisciplinary cleft team. The team was composed of a pediatric surgeon, 2 pediatric ENT specialists, a craniofacial