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Predictors of speech outcomes in children with Pierre Robin sequence

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ABSTRACT

Backgound: Pierre Robin sequence (PRS) has worse speech outcomes than isolated cleft palate. We aimed to search for possible associations of phonological outcomes with PRS status (isolated vs syndromic), clinical severity, soft palate muscles deficiency, or surgical procedure.
Methods: We designed a retrospective study of 130 children (male/female ratio: 0.4) with isolated (96) or syndromic (34) PRS with cleft palate. Grading systems were used to classify retrognathia, glossoptosis, and respiratory and feeding disorders. Electromyography was used to investigate levator veli palatini muscles. Hard cleft palate was measured using maxillary casts. Intravelar veloplasty was performed using the Sommerlad's technique. Phonological outcomes were assessed using the Borel-Maisonny classification.
Results: Cleft palate was repaired in one stage (65.5%) or hard palate closure was postponed (34.5%). Velopharyngeal insufficiency was more frequent in syndromic PRS (53%) vs. isolated PRS (30.5%) ($p = 0.01$), but was not statistically associated with clinical grade, hard cleft palate width, soft palate electromyography, and surgical procedure.
Conclusions: In children with PRS, anatomic variables, initial clinical severity, and soft palate muscle deficiency are not predictors of speech prognosis.

1. Introduction

Pierre Robin originally reported retrognathia and glossoptosis in infants with airway obstruction, and later mentioned cleft palate as an associated symptom (Robin, 1923, 1934). The term 'sequence' was introduced to reflect a presumed sequential developmental pathogenesis – the long-lasting upright position of the tongue leading to retrognathia and a characteristic U-shaped cleft palate (Hanson and Smith, 1975). Pierre Robin sequence (PRS) can occur as isolated or syndromic conditions, showing various degrees of the clinical triad, as well as respiratory, feeding, and speech disorders (Evans et al., 2006; Patel et al., 2012; Witt et al., 1997).

Patients with PRS and cleft palate have been shown to have worse phonological outcomes than patients with isolated cleft palate, although no strong prognostic factors have been identified (Hardwicke et al., 2016; Stransky et al., 2013). Comparisons between syndromic PRS and isolated PRS have not shown clearly distinct outcomes (de Buys Roessingh et al., 2008; Patel et al., 2012; Witt et al., 1997). We reviewed a large series of children with PRS and cleft palate, with the aim to compare their speech outcomes according to PRS status (isolated vs syndromic), and to analyze statistical associations of phonological evaluation with: (1) the morphological criteria of cleft palate, retrognathia, and glossoptosis; (2) the severity of initial respiratory and feeding disorders; (3) clinical evaluation and electromyography (EMG) of soft palate muscles; and (4) primary cleft palate repair procedures. Additionally, we aimed to evaluate phonological outcomes after secondary velopharyngeal surgical procedures.

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2. Material and methods

2.1. Patients

We reviewed the database of our maxillofacial surgery unit over a 13-year period (2000–2012) to identify patients who presented with PRS and cleft palate, with or without airway obstruction. We excluded five patients who had initially been managed in other hospitals, four patients who were lost to follow-up, and 19 patients with no available phonological evaluation, including five with tracheostomy tube. We finally included 130 patients, who were followed by our team over more than 3 years. Based on the results of multidisciplinary clinical assessment, complementary examinations, and genetic studies, we classified PRS as isolated or syndromic. The data were de-identified prior to analysis. Approval by an institutional ethics committee was waived by our Institutional Review Board.

2.2. Clinical assessment

Shortly after birth, retrognathia and glossoptosis were classified according to specific clinical examinations (Table 1). Clinical condition was graded as follows: grade I, when respiration was normal and oral feeding was safe at birth or within the first 2 weeks; grade II, when tube feeding was required for more than 15 days, with no respiratory care except prone position; and grade III, when respiratory support was needed, including endotracheal or nasopharyngeal tube, non-invasive mechanical ventilation, or tracheostomy. Cleft palate was qualified as 'complete' when it reached the anterior palatine foramen, and as 'partial' otherwise. The width of the soft palate cleft was visually evaluated as narrow, medium, or wide; the width of the hard palate cleft was measured using maxillary casts made just before cleft palate repair (Table 1; Fig. 1).

2.3. Soft palate muscle deficiency

Electromyography (EMG) of the soft palate was performed by one of us (FR) and was part of each patient's assessment. Conventional needle EMG was used to study the levator veli palatini muscle bilaterally, after local contact anaesthesia (Renault et al., 2011). Recordings were analyzed manually and classified as normal, neurogenic (i.e. single or reduced interference pattern), or low amplitude (reflecting muscle hypoplasia) (Renault and Quijano-Roy, 2006). In addition, we noted the presence or absence of muscle atrophy reported on the per-operative clinical evaluation chart.

2.4. Cleft palate repair

Three experienced surgeons on our team applied a unique investigating and decision-making protocol, and used the same surgical techniques. Primary cleft palate repair consisted of intravelar veloplasty using the Sommerlad's technique (Sommerlad, 2003). Partial clefts and narrow complete clefts were repaired in a single stage. For wide complete clefts, intravelar veloplasty was performed as a first step, followed by postponed hard palate closure. Lateral incisions of the palatal fibromucosa in addition to a vomerian flap were performed in case of extremely wide hard palate clefts. Secondary surgical treatments included oronasal fistula closure, secondary intravelar veloplasty, velopharyngeal autologous fat injection, and velopharyngoplasty using a superior pharyngeal flap.

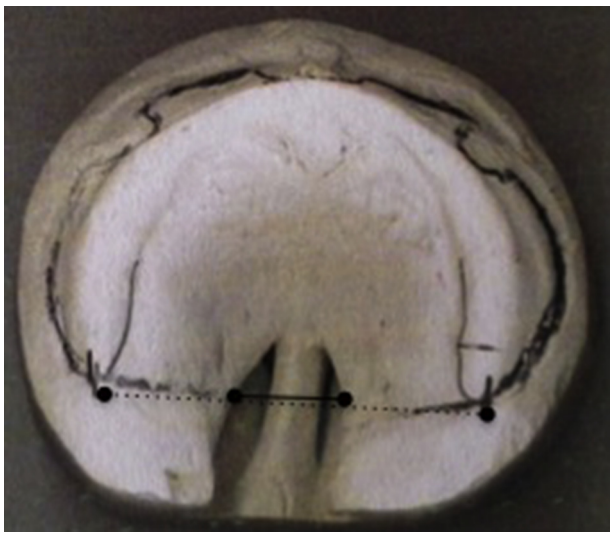

Fig. 1. Measurement of hard palate cleft width using maxillary casts. Measurements of width of the cleft (= A = solid line) and intermaxillary tuberosity distance (= B = dotted line) for calculating A/B ratio.

2.5. Outcome measures

Phonological function was assessed by a speech therapist using the Borel-Maisonny classification, which is the standard test in French-speaking countries for evaluating velopharyngeal function during spontaneous speech and in repetition (Launay and Borel-Maisonny, 1976).

For analyzing phonological outcomes, we took into account the results of three evaluations. The first one was performed after primary cleft palate repair before the onset of speech therapy. The two others were performed before and after secondary velopharyngeal surgery.

2.6. Statistical analysis

Statistics were performed with GraphPad Prism 5 (GraphPad Software, La Jolla, CA).

Data are given as numerical variables, percentages, mean ± SD, or median ± IQR or [25th-75th percentile]. We assessed contingency tables using the Chi-square test, where applicable, or the Fisher exact test. The t-test and Mann–Whitney U-tests were used to compare quantitative data for two groups of patients. p-values <0.05 were considered as significant.

Table 1
Grading system for retrognathia, glossoptosis, and cleft palate.

Morphological criteria	Grades		
Retrognathia	Mild: Red lower lip completely apparent	Moderate: Red lower lip partially apparent	Severe: Red lower lip covered by upper lip
Glossoptosis	Mild: Sublingual crest rise	Moderate: Posterior ptosis, but no vertical position of the tongue	Severe: Vertical and posterior position of the tongue
Hard palate cleft width[a]	Narrow: < 20%	Medium: 20–40%	Wide: > 40%

[a] Width of the cleft/intermaxillary tuberosity distance ratio (Fig. 1).

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