International Journal of Pediatric Otorhinolaryngology 90 (2016) 270–275

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: http://www.ijporlonline.com/

Severity of clinical manifestations and laryngeal exposure difficulty predicted by glossoptosis endoscopic grades in Robin sequence patients☆

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ARTICLE INFO

Article history:
Received 28 August 2016
Received in revised form
24 September 2016
Accepted 26 September 2016
Available online 28 September 2016

Keywords:
Endoscopy
Intubation
Laryngoscopy
Mandible
Pierre Robin syndrome

ABSTRACT

Objective: To evaluate the performance of two glossoptosis airway obstruction classifications in predicting symptom severity and laryngeal exposure difficulty in Robin Sequence (RS) patients.
Setting: Public tertiary hospital otolaryngology section (Hospital de Clínicas de Porto Alegre - HCPA).
Patients: All RS patients diagnosed at HCPA from October 2012 to February 2015 were enrolled, a total of 58 individuals. They were classified in isolated RS, RS-Plus and syndromic RS.
Intervention: Patients were submitted to sleep endoscopy and a score was attributed according to Yellon and de Sousa by a blinded researcher. Symptom severity evaluation was performed as defined by Cole classification.
Main outcome measure: Association between endoscopic findings and clinical symptoms severity and laryngeal exposure difficulty.
Results: Twenty four patients were identified as isolated RS (41.4%), 19 patients presented as RS-Plus (32.7%) and 15 patients had well defined diagnosed syndromes (25.9%). Concomitant airway anomalies were found in 18 patients (31%). Specifically 17.4% in isolated RS, 55.6% in RS- Plus and 28.6% in the syndromic group had such anomalies (P = 0,03). Probability of presenting severe clinical symptoms as graded by Cole was higher in grade 3 Yellon classification (68.4%, P = 0.012) and in moderate and severe de Sousa classification (61.5% and 62.5%, respectively, P = 0.015) than in milder grades of obstruction. This findings were considered significant even after controlling for patient age. Laryngeal exposure difficulty was correlated with de Sousa and Yellon (Rho = 0,41 and Rho = 0,43, respectively; P < 0,05).
Conclusion: Patients with higher degrees of obstruction in sleep endoscopy had a higher probability of presenting a more severe clinical manifestation and a more difficult laryngeal exposure. Since the number of patients included in this study was small for subgroup analyses, it is not clear if this association is restricted to a specific group of RS.

1. Introduction

The Robin Sequence (RS) is defined as the concomitant presentation of micrognathia, glossoptosis and respiratory distress with or without a cleft palate [1,2], while glossoptosis is defined by Pierre Robin as a backward and downward fall of the tongue base

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http://dx.doi.org/10.1016/j.ijporl.2016.09.036
0165-5876/

[2], leading to airway obstruction, which can be visualized through flexible fiberoptic laryngoscopy (FFL). Glossoptosis is a prevalent finding in micrognathic patients but it cannot be systematically presumed. Therefore, FFL has an important role on the identification of presence of glossoptosis in these patients. Moreover, FFL can aid in the evaluation of associated airway disturbances of interest. Often concomitant neurological diseases or even other miscellaneous disorders may cause hypotonia, leading to pharyngomalacia, laryngomalacia or tracheomalacia. Besides that, malformations like cysts, stenosis, vocal cord paralysis and many others can also coexist, aside from glossoptosis. Identification of parallel airway abnormalities is crucial to predict treatment outcomes [3]. In face of all these possible contributions, a diagnostic airway endoscopy is probably warranted in all RS patients. Another possible use of FFL is in outcome monitoring of osteogenesis advancement process [4].

Another valuable information FFL can provide is evaluation of the degree of airway obstruction. Notwithstanding, the method of respiratory obstruction evaluation is still subject to controversy. Different glossoptosis classification systems have already been reported. Sher [5,6] classified airway obstruction in awake RS patients, depending on the cause of the obstruction, attributing a score from 1 (true glossoptosis) to 4 (pharyngomalacia). More than a decade later, de Sousa et al. [7] classified glossoptosis in awake patients as mild, moderate and severe (Fig. 1). Afterwards, Yellon [8], performing FFL under light sedation, classified epiglottic and base of tongue prolapse from 0 to 3, ranging from normal to complete obstruction of the pharynx (Fig. 2). A uniform characteristic of these classifications is that they define progressive degrees of airway compromise, and as a consequence one could expect that clinical manifestations in different subsets of patients would also show a continuing severity spectrum. Although tempting, the demonstration of this correlation was not consistently verified in research until today. Indeed, this limitation eventually compromises the usefulness of those classifications in clinical practice.

Albeit use of these classifications can be sporadically seen in research, it is far from being consensual. There is certainly a complex background that could disentangle the various reasons why this is true. Obviously, scarce applicability in a clinical context would be one of the most relevant. Furthmore for this unique type of patients, a more practical use of this research information would be valuable.

Accordingly, the primary goal of this study was to test the performance of de Sousa and Yellon classifications [7,8] to predict severity of symptoms [9] and laryngeal exposure difficulty in a cohort of RS patients. Other characteristics such as concomitant airway lesions and associated malformations were also reported.

2. Material and methods

2.1. Site of study

This prospective study took place at Hospital de Clínicas de Porto Alegre (Rio Grande do Sul, Brazil), from October 2012 to February 2015, and was submitted to Institution's Research Ethics Committee approval prior to initiation. All newly diagnosed RS patients at our institution during study period were enrolled. Written informed consent was obtained from parents or legal guardians of all enrolled patients. No exclusion criteria were defined.

2.2. Diagnostic workup

Diagnosis of RS was established based on a multidisciplinary evaluation. Patients were required to meet all of the following criteria:

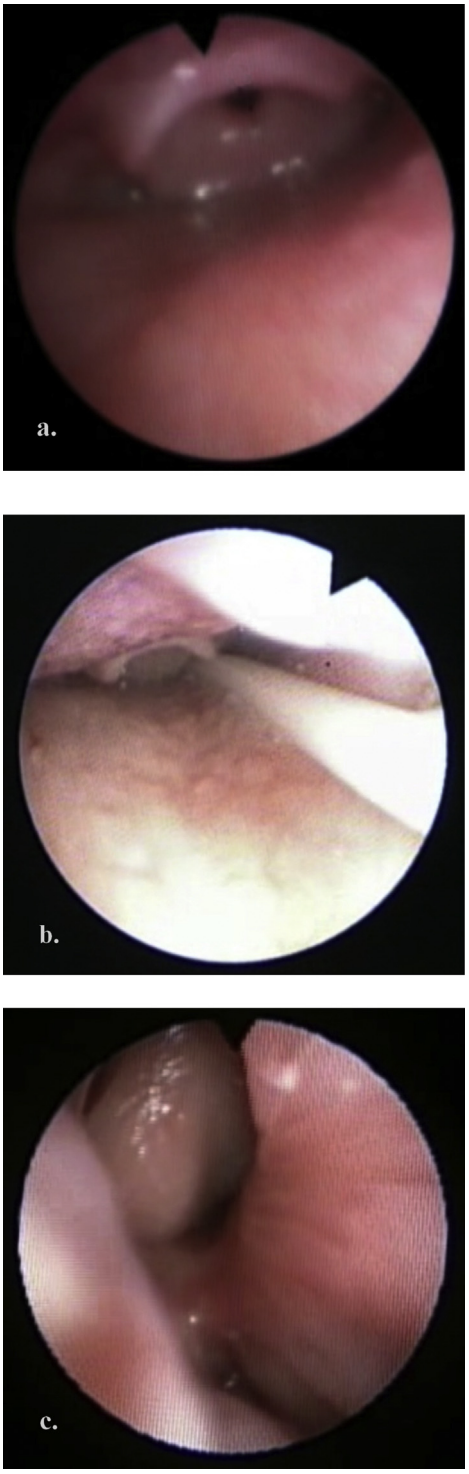

Fig. 1. Endoscopic evaluation of glossoptosis. De Sousa classification illustrated: **a**. Mild: most of the time the posterior region of the tongue does not touch the posterior wall of the pharynx; **b**. Moderate: the posterior region of the tongue touches the posterior wall of the pharynx but does not pressure it; **c**. Severe: the posterior region of the tongue pressures the posterior wall of the pharynx and sometimes the tongue remains in the nasal cavity.

a) Respiratory dysfunction: defined as any respiratory symptom of airway obstruction, whether while on activity engagement, specific positioning, feeding or at rest (reported by caretaker or observed by hospital staff) confirmed by polysomnography.