Journal of Cranio-Maxillo-Facial Surgery 45 (2017) 210–215

Contents lists available at ScienceDirect

Journal of Cranio-Maxillo-Facial Surgery

journal homepage: www.jcmfs.com

The role of flexible fiberoptic laryngoscopy in Robin Sequence: A systematic review

Denise Manica [a, b, *], Cláudia Schweiger [a, b], Leo Sekine [c], Simone Chaves Fagondes [d], Gabriel Kuhl [a, e], Marcus Vinicius Martins Collares [e, f], Paulo José Cauduro Marostica [e, g]

[a] Otolaryngology and Head and Neck Surgery Department, Hospital de Clínicas de Porto Alegre, Brazil
[b] Programa de Pós Graduação em Saúde da Criança e do Adolescente, Universidade Federal do Rio Grande do Sul, Brazil
[c] Programa de Pós Graduação em Epidemiologia, Universidade Federal do Rio Grande do Sul, Brazil
[d] Pneumology Department, Hospital de Clínicas de Porto Alegre, Brazil
[e] Professor of Medicine, Universidade Federal do Rio Grande do Sul, Brazil
[f] Plastic Surgery Department, Hospital de Clínicas de Porto Alegre, Brazil
[g] Pediatric Pneumology Department, Hospital de Clínicas de Porto Alegre, Brazil

ARTICLE INFO

Article history:
Paper received 22 April 2016
Accepted 10 November 2016
Available online 22 November 2016

Keywords:
Endoscopy
Intubation
Laryngoscopy
Mandible
Pierre Robin syndrome

ABSTRACT

Objective: Systematically search literature for flexible fiberoptic laryngoscopy (FFL) use in Robin Sequence (RS) patients, in diverse clinical scenarios.
Data sources: Pubmed, LILACS and SCIELO.
Review methods: Systematic review using a sensitive search strategy focused on RS patients and FFL.
Results: There were 48 full text articles included in this systematic review. No summary meta-analytic measurement could be calculated due to heterogeneity of interventions and outcomes. FFL approaches were grouped in five topics, as follows: *Endoscopic classification*: no evidence on superiority of awake over light sedation and correlation of grading scales with symptom severity. *Airway abnormalities:* high incidence of concomitant lesions besides glossoptosis. *Swallowing evaluation:* no validation against fluoroscopy (gold standard) yet. *Intubation aid for mechanical ventilation:* ultra-thin bronchoscopes improve success rates of intubation. *Treatment outcome monitoring:* no consensus on ideal parameters to be checked.
Conclusion: Some applications have their roles already well established in the management of RS patients, like the evaluation of glossoptosis and associated lesions and as an intubation assistance tool, while others need to be the subject of further research, like the exact method of evaluation, its association with clinical manifestations, its role in swallowing investigation and as a postoperative success predictor.

1. Introduction

Robin Sequence (RS) is a craniofacial abnormality involving mandibular hypoplasia and glossoptosis (with or without cleft palate), leading to life-threatening obstructive apnea and feeding difficulties during the neonatal period or even later in life. Respiratory disorders, generally associated with posterior displacement of the tongue and airway obstruction, require careful management and, in severe cases, may require extended treatment in neonatal intensive care units and surgical interventions to relieve airway obstruction. Those feeding and respiratory difficulties frequently continue well into childhood, affecting not only growth and development, but also impacting on long-term intellectual achievements. The diagnosis of RS depends on clinical features that are often easily recognizable, although eventual clinical manifestations can be very heterogeneous. Because symptom severity is variable, treatment standardization is truly a challenge.

There have been reviews on RS (Cielo et al., 2016; Cote et al., 2015; Kochel et al., 2011; Marques et al., 2005; Ow and Cheung, 2008; Rothchild et al., 2008; Schweiger et al., 2016; Tahiri et al., 2014), but none of them focused on endoscopic evaluation of

* Corresponding author. Rua Ramiro Barcelos, CEP 90035-903, 2350, Porto Alegre, RS, Brazil. Fax: +55 51 3359 8314.
E-mail address: denisemanica@gmail.com (D. Manica).

http://dx.doi.org/10.1016/j.jcms.2016.11.008
1010-5182/

these patients. The importance of discussing this matter is emphasized on a 2015 European survey (van Lieshout et al., 2015) that revealed that, over 101 European clinics researched, only 63% used FFL and 16% used rigid laryngoscopy as a diagnostic tool in RS. In the present article, we highlight the role of flexible fiberoptic laryngoscopy (FFL) in RS in different management approaches described in the literature, concerning evaluation of glossoptosis itself and its classification, evaluation of airway abnormalities, swallowing evaluation, intubation aid and treatment outcome monitoring.

2. Material and methods

We have conceived a search strategy using the PICO (Population, Intervention, Control and Outcome) framework on different reference databases, using terms referring to RS, and FFL application. Databases included Pubmed (Medline), LILACS and SCIELO. Also, retrieved articles' references lists, textbooks, abstracts from congresses and meetings were searched, and medical experts on the subject were consulted for ongoing relevant research. Search strategies for each database are available in Table 1.

Search results were peer reviewed by title and abstract by authors M.D. and S.C. separately. References considered to be included in full text appreciation were compared and agreed upon among reviewers. Where no agreement could be reached a third author (S.L.) took the final decision on inclusion. Included references were retrieved as full text articles to be further analyzed. After full text consideration, articles to be finally included on systematic review were again agreed upon by S.C. and M.D., with the aid of a third reviewer where necessary.

Included articles were then separated by FFL application and their contents were summarized altogether. As different methods, purposes and outcomes for the use of laryngoscopy were the aim of the various studies included it was not possible to compile data in a conventional meta-analysis. Therefore, a narrative discussion on separated topics was chosen as the format of this systematic review.

3. Results

There were 48 full text articles included in this systematic review. The search process summary flowchart, as recommended by PRISMA (Moher et al., 2009), is shown in Fig. 1.

Five main areas of application of FFL in RS patients were identified. Articles were grouped according to these criteria. They are discussed in separate sections ahead.

3.1. Classification of endoscopic findings

Glossoptosis is defined by the backward and downward fall of the base of the tongue, and this diagnosis is usually made during FFL with the patient breathing spontaneously either awake or lightly sedated. This is a subjective criterion, and no objective way to measure it is yet reported. The classification of this defining aspect of RS is fundamental since it allows for an evaluation of surgical results among different patients and different surgeons. Besides that, a thorough classification would permit the discernment of clinical abnormalities, polysomnographic findings and swallowing characteristics. Classifications systems found while searching the literature are described in detail in Table 2. Studies are scrutinized ahead.

Sher et al. (Sher, 1992; Sher et al., 1986) described four mechanisms of obstruction in 33 patients with craniofacial anomalies, including RS, from birth to 24 years of age, examined while awake. Sher emphasized that other mechanisms of upper airway obstruction were involved in RS, not only glossoptosis. The mechanism of airway obstruction could be useful to select the modality of therapy. This classification of airway findings (not glossoptosis) was used by other authors (Argamaso, 1992; Kochel et al., 2011; Marques et al., 2001; Shprintzen and Singer, 1992) and was considered crucial in the treatment choice.

de Sousa et al. (2003) evaluated 56 children with RS, also examined while awake, without any type of sedation. They described a poor correlation between the severity of glossoptosis and the severity of clinical manifestations.

Schaefer et al. (Schaefer and Gosain, 2003; Schaefer et al., 2004) performed nasoendoscopy and bronchoscopy in RS patients under sedation in patients with desaturation during sleeping, feeding or wakefulness. They stratified these patients into three subdivisions, without a specific classification for glossoptosis.

Sorin et al. (2004) evaluated 20 patients with RS with anesthesia by spontaneous breathing. The average compiled preoperative airway obstruction scores in decannulated patients were compared to those who remained tracheostomy dependent and were found not significantly different. They concluded that preoperative airway

Table 1
Search strategy (accessed on 04/17/16).

Database	Search strategy	References retrieved
Pubmed	((“Myopathy, congenital nonprogressive with Moebius and Robin sequences” [Supplementary Concept] OR “Pierre Robin Sequence with Facial and Digital Anomalies” [Supplementary Concept] OR “Robin Sequence with Distinctive Facial Appearance and Brachydactyly” [Supplementary Concept] OR “Corpus Callosum, Agenesis of, with Facial Anomalies and Robin Sequence” [Supplementary Concept] OR “Thrombocytopenia Robin sequence” [Supplementary Concept] OR “Ventricular extrasystoles perodactyly Robin sequence” [Supplementary Concept] OR “Radial defect Robin sequence” [Supplementary Concept] OR “Pierre Robin sequence with pectus excavatum and rib and scapular anomalies” [Supplementary Concept] OR “Robin sequence and oligodactyly” [Supplementary Concept] OR “Richieri Costa Pereira syndrome” [Supplementary Concept] OR “TARP syndrome” [Supplementary Concept] OR “Pierre Robin Syndrome” [Mesh] OR “Robin Sequence” OR “Pierre Robin Sequence” OR “Robin Syndrome”) OR (“Micrognathism” [Mesh] OR “Clavicular Hypoplasia, Zygomatic Arch Hypoplasia, and Micrognathia” [Supplementary Concept] OR “Limb Deficiencies, Distal, with Micrognathia” [Supplementary Concept] OR “Trigonobrachycephaly, Bulbous Bifid Nose, Micrognathia, and Abnormalities of the Hands and Feet” [Supplementary Concept] OR “Corpus Callosum, Agenesis of, with Mental Retardation, Ocular Coloboma, and Micrognathia” [Supplementary Concept] OR “Ectrodactyly of Lower Limbs, Congenital Heart Defect, and Micrognathia” [Supplementary Concept] OR “Genito palato cardiac syndrome” [Supplementary Concept] OR “Micrognathia”) OR (“Glossoptosis” [Mesh] OR “Cri-du-Chat Syndrome” [Mesh] OR “Glossoptosis”) OR (“Mandibular Distraction Osteogenesis” OR “Mandibular Distraction”)) AND (“Endoscopes” [Mesh] OR “flexible fiber optic nasopharyngoscopy” OR “flexible fiberoptic nasopharyngolaryngoscopy” OR “microlaryngoscopy” OR “bronchoscopy” OR “nasopharyngoscopy” OR “direct laryngoscopy” OR “airway endoscopy” OR “diagnostic laryngoscopy” OR “flexible fiber optic endoscopy” OR “flexible fiberoptic endoscopy” OR “endoscopic” OR “endoscopy”)	118
LILACS	Pierre Robin AND endoscopy	2
SCIELO	Pierre Robin	21