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Non-surgical and surgical interventions for airway obstruction in children with Robin Sequence

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ABSTRACT

There is widespread lack of consensus regarding treatment of airway obstruction in children with Robin Sequence. This study aimed to systematically summarize outcomes of non-surgical and surgical options to treat airway obstruction in children with Robin Sequence. The authors searched the Medline, EMBASE and CENTRAL databases. Studies primarily on mandibular distraction were excluded. Study quality was appraised with the Methodological Index for Non-Randomized Studies (MINORS) score. Forty-eight studies were included, of which 45 studies had a retrospective non-comparative set up, two studies had a prospective design and one study was a clinical trial. The mean MINORS score was 7.3 (range 3–10). The rates of successful relief of the airway obstruction (SRoAO) were: not available for orthodontic appliance (2 studies, n = 24), 67–100% for nasopharyngeal airway (6 studies, n = 126); 100 % for non-invasive respiratory support (2 studies, n = 12); 70–96% for tongue-lip adhesion (11 studies, n = 277); 50–84% for subperiosteal release of the floor of the mouth (2 studies, n = 47); 100% for mandibular traction (3 studies, n = 133); 100% for tracheostomy (1 study, n = 25). The complication rate ranged from zero to 55%. Although SRoAO rates seemed comparable, high-level evidence remains scarce. Future research should include description of the definition, treatment indication, and objective outcomes.

1. Introduction

Robin Sequence (RS) is a congenital facial condition occurring in 1 in 8,500 to 1 in 30,000 newborns (Bushe, 1983; Tolarova and Harris, 1995; Printzlau and Andersen, 2004). The French stomatologist Pierre Robin originally defined RS in 1923 as a triad of mandibular hypoplasia, glossoptosis and airway obstruction. Some clinicians also include cleft palate as part of the definition. However, there is no clear, unanimous definition of RS.

The main problems in RS include airway obstruction and feeding difficulties, both occurring with varying degree of severity. In this review we focus on airway obstruction. Airway obstruction may vary from virtually non-existing to apneas, increased activity of breathing muscles, failure to thrive, cyanosis and ultimately respiratory insufficiency. Patients with RS are frequently diagnosed with obstructive sleep apnea (OSA), which in turn is associated with considerable morbidity. The prevalence of OSA has been reported to be between 46 and 100 % in children with RS (Gilhooly et al., 1993; Wilson et al., 2000; Bravo et al., 2005; Anderson et al., 2011). The current gold standard to diagnose OSA is a nocturnal polysomnography (Section on Pediatric Pulmonology, 2002).

A number of treatment options are available to treat airway obstruction in RS, but there is currently no widely accepted guideline or treatment algorithm. Most clinicians agree that prone

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positioning is the treatment of choice for mild cases, but a large variety of treatment options exist for cases in which prone positioning fails. There is an obvious need for a more evidence-based approach to treatment of children with RS.

The aim of our study was to systematically summarize outcomes of non-surgical and surgical interventions for airway obstruction in children with RS based on effectiveness and safety. This review intends to inform clinicians about the current state of evidence in literature and to highlight research gaps, thereby functioning as a guide in the set-up of future clinical studies.

2. Material and methods

The Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) statement was adhered to as much as possible in the preparation of this review. No approval was necessary by an institutional review boards due to the nature of this study.

2.1. Search strategy

A detailed systematic review protocol was prepared by all authors. The review was conducted using detailed search and extraction methods for the MEDLINE, EMBASE and CENTRAL databases aimed at studies published after January 1st 2000. The reference list of included studies was checked for additional eligible studies.

2.2. Eligibility criteria

Studies were considered eligible for inclusion if: 1. Study participants had a diagnosis of RS; 2. Study participants were below the age 18 years old; 3. Studies had more than 5 participants; 4. Studies had a focus on non-surgical and/or surgical intervention(s) to manage the airway obstruction; 5. Studies contained original data on treatment outcomes; 6. The study was published in English.

The diagnosis of RS was author-defined to avoid excluding relevant studies. Given the ongoing debate on specific, more or less obligatory features of RS, all definitions were accepted. Children with a diagnosis of mandibular hypoplasia and airway obstruction were also considered to have RS. Children with both isolated and non-isolated forms of RS were included.

Since studies specifically on mandibular distraction in children with RS already have been extensively covered in reviews by Ow, Bookman and Paes, we decided to exclude articles solely on mandibular distraction (Ow and Cheung, 2008; Bookman et al., 2012; Paes et al., 2013).

Study quality was appraised with the Methodological Index for Non-Randomized Studies (Slim et al., 2003) and the Oxford Centre for Evidence-Based Medicine (CEBM) scale. MINORS consists of a 12-item checklist. The first eight items focus specifically on non-comparative studies. Each item is scored 0 (not reported), 1 (reported, but inadequate) or 2 (reported and/or adequate). The maximum score is 16 for non-comparative studies and 24 for comparative studies. Primary outcomes included successful relief of the airway obstruction without necessity for further treatment (SROAO), the obstructive apnea hypopnea index (oAHI) and mortality (not disease specific). Secondary outcomes included side effects, complications and improvement of oxygen saturation.

2.3. Selection of studies

Initially, all papers were independently examined on titles and abstracts by two authors (MvL and MvdS). Afterward, the full text manuscript was assessed for eligibility on basis of the defined criteria by the same authors. Any disagreements were resolved by discussion between the two review authors and if needed by involvement of another author of our review group.

2.4. Data extraction and quality appraisal

Data extraction of the manuscripts was performed independently by two authors (MvL and MvdS) using a customized data collection form.

3. Results

Forty-eight studies were included in the qualitative synthesis. All studies except Buchenau et al. were Oxford CEBM Level type IV. We did not find any studies that focused specifically on prone-positioning. The mean MINORS score was 7.3 (range 3–10). Reported outcome measures differed and included: clinical signs of airway obstruction, overnight polysomnography outcomes (oAHI, mixed-obstructive apnea index (mOAI), central apnea index (CAI), oxygen desaturation index (ODI), capillary blood pH, CO_2 pressure), weight velocity, body weight, oxygen saturation, growth, (in-patient) hospital stay, complication rate, need for additional surgery, need for tracheostomy, questionnaires on satisfaction, maxilla-mandibular discrepancy and death. Eleven studies mentioned the use of polysomnography in their clinic, but specific data were not always available (Fig. 1).

3.1. Orthodontic appliance (Table 1) (Two studies with 24 patients in total) (Buchenau et al., 2007; Bacher et al., 2011)

Two studies of the same group on the use of an orthodontic appliance were found. In a prospective observational study and a randomized clinical trial, the study group of the Tuebingen Hospital in Germany described the use of an intraoral orthodontic appliance with velar extension shifting the tongue anteriorly, thereby widening the hypopharyngeal space (Buchenau et al., 2007; Bacher et al., 2011). In the study of Buchenau et al. in 90% of the children in the pre-epiglottic plate group an improvement of mOAI was observed, compared to only 36% of infants in the control group who received a conventional appliance. In the study of Bacher et al. a significant decrease in mean mOAI was noted at the three-month follow-up.

Reported side-effects included soft tender spots. In both studies only children with an isolated RS participated.

3.2. Nasopharyngeal airway (Table 2) (Six studies with 126 patients in total)

Techniques differed in the six available studies, but in general a nasopharyngeal airway was created by modifying an endotracheal tube and position of the distal end of the tube on top of the larynx, bypassing the tongue base. The nasopharyngeal airway permits the child to breathe through the tube, and may break the seal between the posterior placed tongue base and the pharynx wall. The mean duration of the use of a nasopharyngeal airway ranged from 44 days to 8 months. The SROAO rates ranged from 67 to 100%. In the study of Wagener four complications were reported: three patients developed a chest infection and one patient developed right nostril stenosis (Wagener et al., 2003).

3.3. Non-invasive respiratory support (Table 3) (Two studies with 12 patients in total) (Leboulanger et al., 2010; Girbal et al., 2014)

Only two studies were found on non-invasive respiratory support (Leboulanger et al., 2010). Non-invasive respiratory support