International Journal of Pediatric Otorhinolaryngology 77 (2013) 499–503

Contents lists available at SciVerse ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

# Airway, feeding and growth in infants with Robin sequence and sleep apnoea

M. Daniel [a,b,*], S. Bailey [c], K. Walker [d,e], R. Hensley [c], C. Kol-Castro [c], N. Badawi [d], A. Cheng [a], K. Waters [c,e]

[a] Paediatric Otorhinolaryngology, Children's Hospital, Westmead, Sydney, Australia
[b] NIHR Nottingham Hearing Biomedical Research Unit, Nottingham, UK
[c] Sleep Medicine, Children's Hospital Westmead, Sydney, Australia
[d] Grace Centre for Newborn Care, Children's Hospital Westmead, Sydney, Australia
[e] University of Sydney, Sydney, Australia

ARTICLE INFO

*Article history:*
Received 29 August 2012
Received in revised form 13 December 2012
Accepted 15 December 2012
Available online 11 January 2013

*Keywords:*
Robin sequence
Airway
Sleep study
Continuous positive airway pressure
Child
Feeding
Obstructive sleep apnoea
Growth

ABSTRACT

*Objective:* Robin sequence (RS) is associated with airway abnormalities that result in functional problems of obstructive sleep apnoea (OSA), feeding difficulties, and consequent poor growth. We evaluated the relationships between OSA severity, airway and feeding interventions, and weight at 12 months in infants with RS and OSA.

*Methods:* Retrospective notes review of children with RS managed at our neonatal unit (1998–2010, inclusive).

*Results:* Of 39 infants studied, 10 (25.6%) had mild/moderate OSA, and 29 (74.4%) severe. Infants with severe OSA required more airway interventions in hospital (82.8 vs 30.0%, $p = 0.004$) and at discharge (72.4 vs 20.0%, $p = 0.007$) than those with mild/moderate OSA; 30.0% of infants with mild/moderate OSA required continuous positive airway pressure (CPAP) during admission and 20.0% on discharge, but amongst those with severe OSA 82.8% required airway interventions as an inpatient, 17.2% underwent mandibular distraction osteogenesis, and 55.2% required CPAP on discharge. Those with severe OSA were also more likely to require tube feeding on discharge (89.7 vs 50.0%, $p = 0.02$). Overall, children were on a lower weight centiles at discharge compared to birth (−10.2 centiles) and at 12 months of age compared to birth (−14.8 centiles), but this occurred irrespective of OSA severity or need for airway interventions or tube feeding.

*Conclusions:* Infants with RS commonly have OSA, feeding and airway difficulties. Weight at 12 months appeared not to be influenced by OSA severity, feeding or airway problems, suggesting that current intervention/management strategy results in the severely affected infants growing as well as those affected less severely.



## 1. Introduction

Robin Sequence (RS) is a triad of micrognathia, glossoptosis and respiratory distress [1]. Its clinical features stem from a mandible that is small, leading to posterior displacement of the tongue (glossoptosis), and thus pharyngeal obstruction with consequent airway and feeding difficulties. A cleft palate is often present and may contribute to feeding difficulties. Just under half of neonates have additional associated abnormalities [2]. The clinical features are variable, ranging from mildly affected neonates requiring no support, to severely affected ones that have airway obstruction and are unable to feed. Due to this clinical variability it has been difficult to study RS infants grouped together as a whole, with most reports focussing on the severely affected children requiring more medical input. What is clear, is that they are best managed in a multidisciplinary setting by an experienced team [3]. In addition to feeding and airway difficulties, neonates with RS also frequently suffer with OSA [4], with up to 85% affected in some studies.

Airway, sleep, feeding, and growth are intimately interlinked [1,4–6]. Airway obstruction may lead to OSA, poor feeding and therefore poor growth. We aimed to evaluate the severity of sleep apnoea, airway management, and feeding management in children with RS, and to determine if there was any relationship between these factors and growth at 12 months of age.

## 2. Methods

With institutional Research Ethics Committee approval, a retrospective review was undertaken at Children's Hospital Westmead, a large tertiary hospital that is part of the Sydney

* Corresponding author at: Paediatric Otorhinolaryngology, Children's Hospital Westmead, Hawkesbury Av, Sydney 2145, Australia. Tel.: +61 2 9845 3253; fax: +61 2 9845 2078.
*E-mail address:* matija.daniel@nottingham.ac.uk (M. Daniel).

0165-5876/$ – see front matter © 2012 Elsevier Ireland Ltd. All rights reserved.
http://dx.doi.org/10.1016/j.ijporl.2012.12.019

**Table 1**
Clinical characteristics and their relationship to OSA severity (mild/moderate OSA: AHI ≤ 10, severe > 10). The 1st admission refers to duration of in-patient stay following birth. Airway interventions include any treatments other than prone positioning. *P* values refer to differences between children with mild/moderate OSA and those with severe OSA.

| Features | OSA severity | | | |
|---|---|---|---|---|
| | All children | Mild/moderate | Severe OSA | *p*-Value |
| Number (%) of children | 39 (100) | 10 (25.6) | 29 (74.4) | |
| Number (%) female | 23 (59.0) | 7 (70.0) | 16 (55.2) | NS |
| Associated abnormality N (%) | 24 (61.5) | 7 (70.0) | 17 (58.6) | NS |
| Gestation weeks mean (range) | 38.9 (32–42) | 39.7 (38–41) | 38.6 (32–42) | NS |
| 1st admission mean days (range) | 41.8 (7–133) | 28.9 (16–69) | 46.3 (7–133) | NS |
| Airway intervention-inpatient N (%) | 27 (69.2) | 3 (30.0) | 24 (82.8) | 0.004 |
| Airway intervention-discharge N (%) | 23 (59.0) | 2 (20.0) | 21 (72.4) | 0.007 |
| CPAP on discharge | 18 (46.2) | 2 (20.0) | 16 (55.2) | NS |
| Mandibular distraction | 5 (12.8) | 0 | 5 (17.2) | NS |
| Tube feeding in hospital | 32 (82.1) | 6 (60.0) | 26 (89.7) | NS |
| Tube feeding on discharge | 31 (79.5) | 5 (50.0) | 26 (89.7) | 0.02 |
| Extra calories on discharge | 20 (51.3) | 5 (50.0) | 15 (51.7) | NS |

NS: not statistically significant; CPAP: continuous positive airways pressure; OSA: obstructive sleep apnoea.

Children's Hospital Network serving in excess of 5 million people. All infants who were admitted to the Neonatal Unit for RS management between 1st January 1998 and 31st December 2010 were included. Data on weight at birth, on discharge, and at 12 months of age (chronologic) were collected, as was information on associated features, and management of airway and feeding in hospital and on discharge.

Sleep apnoea was assessed using polysomnography (PSG) with transcutaneous $O_2/CO_2$ monitoring for a period of at least 4 h of sleep time. Studies completed prior to January 2008 were recorded using Compumedics Profusion PSG (Version 1.01, Compumedics Limited, Abbotsford, VIC, Australia) with subsequent studies recorded on the Sandman Digital 32+ Amplifier Sandman Elite Sleep Diagnostic Software (version 9.0, Nellcor Puritan Bennett (Melville) Ltd., Kanata, ON, Canada). Sleep staging for infants <6 months of age was completed using the criteria outline by Anders, Emde & Parmalee [7], with American Academy of Sleep Medicine criteria applied for those ≥6 months. Respiratory events were considered significant if they lasted ≥2 respiratory cycles and were terminated by an arousal and/or desaturation of ≥3%. Briefly, obstructive apnoeas were defined as the cessation/decrease in airflow to ≤20% baseline amplitude, and hypopnoeas were defined as a decrease in airflow to 20–50% of baseline amplitude. Arousals were defined as changes in ≥2 independent channels, with at least a 10 s period of sleep prior to the change and a disturbance lasting for >1 s. In addition to the apnoea hypopnea index (AHI), defined as the average number of respiratory events (central and obstructive apnoeas, plus hypopnoeas) per hour of sleep time, information was also collected on the obstructive AHI (OAHI, average number of obstructive or mixed respiratory events per hour), OAHI during rapid eye movement (REM) or active sleep, OAHI during slow wave (SW) or quiet sleep, mean duration of apnoeas and hypopnoeas, and $O_2$ and $CO_2$ levels.

AHI was used to categorise severity of sleep apnoea [8], with an AHI of 1–5 considered mild, 6–10 moderate, and >10 indicating severe sleep apnoea. There were two exceptions to this rule. The first was a child with an AHI of 12.6, but an obstructive AHI of only 3.3 and mild symptoms, who was classified as moderate OSA. The second child was clinically severely affected requiring mandibular distraction osteogenesis, with OAHI during REM of 36.2. However, the overall AHI was only 3.3, but this required significant active observation and intervention with prone positioning. Because of clinically severe symptoms and high OAHI he was classed as having severe OSA. His AHI following mandibular distraction osteogenesis was 10.9.

Statistical analysis was performed using SPSS 19, analysing categorical data with Chi squared test and numerical data with Student's t test.

## 3. Results

A total of 39 infants, aged at least 12 months at time of review, were included in the study. Of these, 17 (43.6%) had an associated cleft palate. The timing of their sleep studies ranged between 5 days of age and 141 days (only 3 studies were performed after 3 months of age, and those were all >10 years ago). All had OSA. The 5 infants with mild OSA were grouped together with those 5 with moderate OSA for analysis purposes. Table 1 shows various clinical characteristics and their relationship to sleep apnoea severity. Table 2 details PSG results.

Of 39 infants, 24 (61.5%) had other abnormalities, including 7 with Stickler syndrome, 4 with chromosomal abnormalities, 7 with dysmorphic or syndromic features, a further 4 with cardiac abnormalities, 1 with strabismus and 1 with mild global developmental delay.

There were no statistically significant differences in the proportion of infants with severe OSA amongst those with isolated RS (80.0% having severe OSA) or those with additional abnormalities (70.8%). There were also no statistically significant differences with respect to proportion requiring airway interventions other than prone positioning in hospital (53.3 vs 79.2%, respectively) or on discharge (46.7 vs 66.7%), or the proportion requiring tube feeding in hospital (73.3 vs 87.5%) or on discharge (73.3 vs 83.3%)

Airway interventions other than prone positioning were required in 27 (69.2%) infants in hospital and in 23 (59.0%) on discharge. CPAP was the most frequently used modality, alone or in combination with other treatment, in 25 children during admission, and in 18 infants on discharge. Five children (all with severe OSA) underwent mandibular distraction osteogenesis. A nasopharyngeal tube was used in only one child that also had CPAP and mandibular distraction osteogenesis.

Overall, children were on a lower weight centile at discharge than at birth (−10.2 centiles, standard deviation 23.2, $p = 0.01$), at 12 months of age compared with birth (−14.8 centiles, standard deviation 34.4, $p = 0.01$), and at 12 months of age compared with discharge (−4.6 centiles, standard deviation 34.7, not statistically significant). To examine effect of clinical characteristics on weight, we determined the changes in weight centile between birth and discharge, birth and 12 months, and discharge and 12 months. The influence of various clinical features on weight centiles change is