International Journal of Pediatric Otorhinolaryngology 86 (2016) 98–103

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

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

Quality of life in children with Robin Sequence

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ARTICLE INFO

Article history:
Received 6 January 2016
Received in revised form 31 March 2016
Accepted 23 April 2016
Available online 27 April 2016

Keywords:
Robin Sequence
Infant
Child
Quality of life

ABSTRACT

Introduction: Parents may play an important role during the management of children with Robin Sequence (RS). However, so far only one study has been done on both parent-reported health-related quality of life (HRQoL) and obstructive sleep apnea (OSA) symptoms in children with RS.
Methods: Overall, 63 children with RS, aged 1 and 18, were included in this cross-sectional study. Fifty-three parents of children with RS with a median age of 8.9 [IQR 5.1–12.7] completed questionnaires on HRQoL (OSA-18) and symptoms of OSA (the Brouillette score) in their child with RS. Ten children between 12 and 18 years filled out the self-reported HRQoL questionnaire OSA-12.
Results: At cross-section, 10 children still had respiratory problems. Overall, parents of children with RS reported a lower HRQoL in their child compared with parents in the general population. Parents of children with RS who still had respiratory problems, i.e. OSA or airway support, reported significantly worse OSA-18 scores compared with parents of RS children without OSA. Children with RS themselves (n = 10) reported less physical suffering and less emotional distress on the OSA-12 compared with children in the norm population. Parental perceptions of OSA-related symptoms were accurate, except for the incidence of snoring. In assessing snoring, the multidimensional OSA-18 sleep domain was more informative.
Conclusions: Parents of children with RS reported a lower HRQoL in their child compared with parents in the general population. Parental perceptions of health and HRQoL in children with RS might have an additional value to recognize and treat respiratory problems.

1. Introduction

Robin Sequence (RS) is characterized by a sequence of clinical events with mandibular hypoplasia as the inciting anomaly. Due to the mandibular hypoplasia the tongue becomes posteriorly displaced (glossoptosis) and obstructs the airway. RS occurs in about 1 in 5600–30,000 newborns [1–5]. Patients with RS also present a cleft palate in 80–90% of the cases [4,6,7]. Children with RS are at high risk of developing obstructive sleep apnea (OSA) [8], with reported OSA prevalence rates between 46 and 100%, depending on the criteria used [8–12]. When RS is untreated, airway and feeding problems may be sustained, resulting in a range of morbidities, e.g. failure to thrive, cyanosis, cerebral hypoxia, and even death.

Health-related quality of life (HRQoL) has become an important measure throughout the diagnosis and management of children with obstructive sleep disordered breathing [13]. HRQoL is considered to be a multidimensional concept relying on the patient's subjective evaluation of physical, social, and emotional aspects of a patient's well-being [14] that are relevant to health and/or illness. HRQoL questionnaires can be categorized into generic versus disease-specific, focusing on disease symptoms per se versus on the subjective evaluation of these symptoms, and self-reported versus proxy-reported HRQoL, for example a parent that assesses the HRQoL in his/her child [15].

Pediatric OSA has been associated with an impaired HRQoL assessed with both generic and disease-specific questionnaires (Child Health Questionnaire and OSA-18) [16]. In a mixed group of 79 children with OSA, the OSA-18 score was found to be an important tool to identify the impact of OSA on children with OSA and their families [17]. However, in relation to PSG findings, the sensitivity and the negative predictive value (NPV) of OSA-18 for OSA were low. However, the positive predictive value (PPV) was high. Therefore, it was recommended that OSA-18 be used as a quality-of-life

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http://dx.doi.org/10.1016/j.ijporl.2016.04.030
0165-5876/

indicator and not as a reliable substitute for PSG [17,18]. In an RCT regarding early-adenotonsillectomy versus watchful-waiting, children with OSA significantly improved their PSG scores after early-adenotonsillectomy. In addition, parents also reported improvements in HRQoL (e.g. assessed with the OSA-18) in their child with OSA after early-adenotonsillectomy [16,19,20].

Little is known about the impact of RS on HRQoL in children. Therefore, the aim of this study was to perform a cross-sectional study in children who were born with RS in order to assess parent-reported HRQOL and parent-reported symptoms of OSA in their child with RS. Besides, these outcomes were related to the current respiratory status of the child.

2. Methods

All children, aged between 1 and 18 years, with RS who were treated at the Dutch Craniofacial Center, Erasmus MC – Sophia Children's Hospital, between 2012 and 2015 were eligible for this cross-sectional study. RS was defined as the presence of mandibular hypoplasia and airway obstruction [21,22].

2.1. Assessment procedure

The ethics committee review board of the Erasmus MC (MEC-2012-048) approved the research protocol. All eligible patients and/or parents were approached in a standardized way through a patient information letter. Written informed consent was obtained from all parents and patients above 12 years.

If possible, children underwent polysomnography (PSG) in the hospital or at home. Data regarding medical history were collected from the patients' medical record. For psychological assessment, parents were asked to fill out the Brouillette score, a parent-report of symptoms of OSA in their child, and the parent-reported Obstructive Sleep Apnea survey-18 (OSA-18), a questionnaire in which parents report on the impact of OSA in their child with RS. Children between 12 and 18 years old filled out the self-reported Obstructive Sleep Apnea survey-12 (OSA-12) regarding the impact of OSA on them.

2.2. Polysomnography

PSG was done either in an ambulant sleep study at home; level III, with data recorded by the Embletta Portable Diagnostic system, or in a clinical sleep study in the hospital; level I, i.e. attended PSG including medical and technical support. During these sleep studies a variety of cardiorespiratory variables were assessed, including nasal airflow, chest and abdominal wall motion, and arterial oxygen saturation. Data were analyzed using Somnologica for Embletta software 3.3 ENU (Medcare Flage, Reykjavik, Iceland) for ambulant studies, and Shell + BrainRT Software Suite Version 2.0 (O.S.G. Rumst, Belgium) for clinical studies.

For analysis, we aimed for a total sleep time (TST) of at least 360 minutes, free of artifact. The scoring for respiratory events and calculation of the obstructive apnea–hypopnea index (oAHI) were done as described by Spruijt et al [23]. OSA was defined as an oAHI ≥ 1 per hour [24].

2.3. The Brouillette score

Parents were asked to fill out the Brouillette score, a questionnaire to screen for the presence of OSA [25]. The score was calculated using the following formula: 1.42D + 1.41A + 0.72S – 3.83. D stands for difficulty in breathing, A for apnea, and S for snoring. For D and S, caregivers could choose between the options never = 0, sometimes = 1, often = 2, and always = 3. For A, caregivers could score 0 if no apneas occur or 1 if they do. A Brouillette score of >3.5 predicts the presence of OSA, a Brouillette score between –1 and 3.5 is suggestive of OSA, and a score <–1 predicts the absence of OSA. In analyses, parent-reported OSA was defined as a Brouillette score ≥ –1.

2.4. Parent-reported Obstructive Sleep Apnea-18 (OSA-18)

The Dutch version of the parent-reported disease-specific quality of life questionnaire OSA-18 was used to assess parental perceptions of the impact of OSA on the child with RS [26]. The OSA-18 is the most widely used HRQoL questionnaire in pediatric OSA [16]. The Dutch version has been validated in 459 parents of healthy children and in 119 parents of children with syndromic craniosynostosis [26]. The OSA-18 consists of 18 age-independent items grouped into five domains: sleep disturbance, physical suffering, emotional distress, daytime problems, and caregiver concerns. Parents were asked to report how often during the previous 4 weeks their child has had specific symptoms, using a response scale from 1 (never) to 7 (always). The total OSA-18 score ranges from 18 to 126, with a higher score indicating a worse outcome. Scores less than 60 suggest a small impact on health-related quality of life, scores between 60 and 80 suggest a moderate impact, and scores above 80 suggest a large impact. The parent-reported OSA-18 scores of the impact of OSA in children with RS were compared with those of 459 parents in the general population [26].

2.5. Child-reported Obstructive Sleep Apnea-12 (OSA-12)

Children aged 12–18 completed a comparable OSA questionnaire, the self-reported OSA-12, to assess the impact of OSA on them. Children were asked to report how often during the previous 4 weeks they have had specific symptoms, using a response scale from 1 (never) to 7 (always). This questionnaire consists of 12 questions, comparable to those of the OSA-18 with the following exceptions: 2 questions of the sleep disturbance domain and the total caregiver concerns domain were excluded. The total OSA-12 score ranges from 12 to 84. The OSA-12 has been validated in n = 162 children from the general population and in n = 29 children with craniosynostosis. OSA-12 scores of children with RS were compared with those of 162 children, aged 12–18 years, in the general population [26].

2.6. Visual analogue scale (VAS)

In addition, parents and children were asked to indicate the child's HRQoL on a visual analogue scale (VAS). The VAS consists of a Likert scale from 0 to 10, which was adjusted from the EQ-5D [27]. Scores ranged from 0, worst imaginable HRQoL, to 10, best imaginable HRQoL. A higher score indicated a better HRQoL.

2.7. Statistical analysis

To compare baseline characteristics between participants and non-participants, Pearson's Chi Square tests for dichotomous data and non-parametric Mann–Whitney U tests for continuous non-normally distributed data were used. In order to determine the median age at the time of cross-section in the group of non-participating patients we used the date halfway our study inclusion period as the date of cross-section.

To compare the mean OSA-18 results of the RS population with the mean in the general population, sample T-tests were used. Spearman correlation was calculated between OSA-18 and the visual analogue scale. Median OSA-18 scores were compared between three groups, divided for treatment history and the presence of OSA/currently receiving airway support, using Kruskal–Wallis H tests. A p-value of <0.05 was considered statistically significant.