Journal of Cranio-Maxillo-Facial Surgery 43 (2015) 1626–1631

Contents lists available at ScienceDirect

Journal of Cranio-Maxillo-Facial Surgery

journal homepage: www.jcmfs.com

Robin sequence: A European survey on current practice patterns

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ARTICLE INFO

Article history:
Paper received 18 November 2014
Accepted 17 July 2015
Available online 31 July 2015

Keywords:
Survey
Robin sequence
Diagnosis
Treatment
Europe

ABSTRACT

To provide an overview of current practice patterns with regard to Robin sequence (RS) patients in Europe, a survey was conducted among European clinicians. This online survey consisted of different sections assessing characteristics of the respondent and clinic, definition, diagnosis, treatment, and follow-up. In total, surveys from 101 different European clinics were included in the analysis, and 56 different RS definitions were returned. The majority (72%) of the respondents used a sleep study system to determine the severity of the airway obstruction. A total of 63% used flexible endoscopy and 16% used rigid endoscopy in the diagnostic process. Treatment of the airway obstruction differed considerably between the different countries. Prone positioning for mild airway obstruction was the treatment modality used most often (63%). When prone positioning was not successful, a nasopharyngeal airway was used (62%). Surgical therapies varied considerably among countries. For severe obstruction, mandibular distraction was performed most frequently. Three-quarters of the respondents noted the presence of catch-up growth in their patient population. This first European survey study on definition and management of RS shows that there are considerable differences within Europe. Therefore, we would encourage the establishment of national (and international) guidelines to optimize RS patient care.

1. Introduction

Robin sequence (RS) is a condition classically characterized by micrognathia, glossoptosis, and airway obstruction (Robin 1923). Since this description in the original monograph of Pierre Robin, several authors have used modified definitions (Breugem and Mink van der Molen 2009; Breugem and Courtemanche 2010). RS has an estimated incidence from about 1 in 8000 to 1 in 20,000 newborns, depending on the criteria used to define RS (Bush and Williams 1983; Tolarova and Cervenka 1998; Printzlau and Andersen 2004; Vatlach et al. 2014). It can be divided into those with an isolated and those with a non-isolated condition, the latter being present in about 40% of the cases. Several problems, such as airway obstruction and feeding difficulties, may occur in children with RS. In severe RS cases, this may require long-lasting admission to the pediatric intensive care unit.

Frequently, authors have noted that care of children with RS should be multidisciplinary, with non-surgical and surgical disciplines involved, but there seems to be no consensus on the diagnostic work-up and treatment (Whitaker et al. 2003; Mackay 2011). Recent European literature describes a wide range of diagnostic and treatment modalities. We conducted a survey to provide an overview of current practice patterns of RS within Europe and to provide a springboard for future discussion. To our knowledge, this is the first European survey to focus on RS.

2. Material and methods

For this cross-sectional study, an online survey was constructed by the author panel, which was followed by pilot testing among eight local clinicians who were acquainted with RS. After further

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http://dx.doi.org/10.1016/j.jcms.2015.07.008
1010-5182/

refinement, the final survey consisted of 23–44 questions, depending on the answers given (Supplementary Material). Both multiple choice questions and open questions were included. Most questions were formulated in such a way that the respondent was asked to give a response for his or her clinic, assuming that definition and management were uniform within the clinic. The survey could be accessed in a secure survey environment (Lime Survey Version 1.91 + Erasmus Medical Center).

In total, 655 persons in the European network existing through contacts of the Cleft Center Rotterdam and the Dutch Craniofacial Center were invited. This network includes pediatricians, otolaryngologists, plastic surgeons, oral and maxillofacial surgeons, and nurse practitioners. All received an e-mail message with an online link to the survey. If the e-mailed person was not involved in care of children with RS, we asked them to send the e-mail address of the person who was involved in their clinic. This person was then invited. After the initial survey distribution we send out a maximum of two reminders to increase response rate. The survey period was from June 2013 until November 2013. The survey was available only in English.

Both complete and incomplete surveys were included in the analysis with the exception of surveys that were considered unreliable by all authors for various reasons (e.g., only completion of the respondent characteristics). If more than one survey per clinic was filled out, we included only the first submitted survey in the main analysis. Statistical analysis was performed using IBM SPSS version 20 (SPSS Inc., Chicago, IL, USA). We performed only descriptive statistical calculations. For this survey study, we obtained approval of the Medical Ethics Commission (MEC-2014-242) of the Erasmus Medical Center.

3. Results

3.1. Characteristics of the respondents

A total of 138 responses were collected (a response rate of 21%). Twelve surveys were excluded from the main analysis because they originated at clinics that had already returned a survey, and 25 surveys were excluded because only the respondent's characteristics were filled out. In total, 101 surveys from 24 European countries were included in the main analysis.

The response between countries was variable. There were five countries with more than five respondents: the United Kingdom (n = 26), Germany (n = 12), the Netherlands (n = 11), France (n = 8), and Sweden (n = 6). The other countries had fewer than 5 respondents: Ireland, Italy, Portugal and Spain had four respondents; Belgium, Romania, and Switzerland had three; Hungary had two; and Austria, Croatia, Denmark, Estonia, Finland, Greece, Latvia, Lithuania, Norway, Poland, and Turkey had one respondent.

The large majority of the respondents (72%) were specialists in the field of oral and maxillofacial surgery, plastic surgery, or otorhinolaryngology. Other fields included pediatrics (15%) and cleft nurse, orthodontist, or pulmonologist (10%). Three respondents did not answer on this item. Respondents worked mainly in university hospitals (70%) but also in general hospitals (21%), private practices (4%), or another type of clinic (6%). In most of the respondents' clinics, a specialized multidisciplinary cleft and/or craniofacial team was present: 35% had a cleft team, 6% had a craniofacial team and 45% had both a cleft team and a craniofacial team. Twelve percent did not have a cleft team or craniofacial team, and 3% did not answer on this question. The number of new RS children who were seen annually in the respondents clinic were more than five children (33%), five to 10 children (42%), 10 to 15 children (11%), and more than 15 children (11%).

3.2. Definition

In total, 56 different combinations of features necessary for a diagnosis of RS were returned (Table 1). About one in three respondents distinguished micrognathia, retrognathia, and mandibular hypoplasia. In the comments, some respondents noted that micrognathia is a small mandible (size), that retrognathia is a normal-sized, backwards-placed mandible (position), and that mandibular hypoplasia is the same as micrognathia. Others regarded the different terms as a grade of severity, with micrognathia being the most severe form. However, several respondents noted that the use of a certain term does not influence their management.

A cleft palate was considered an obligatory feature for a diagnosis of RS by 96% in the United Kingdom, 88% in France, 67% in Sweden, 58% in Germany, and 55% in the Netherlands. A clear distinction between isolated and non-isolated RS was made by roughly half of the respondents. Twenty-one of these respondents noted in the comments that this distinction influenced their management approach.

3.3. Diagnosis

Mandibular size was mainly assessed by clinical sight (Table 2). About one in three of the respondents used other diagnostic modalities to assess the mandible and airway such as X-ray, three-dimensional computed tomography, magnetic resonance imaging, esophageal pressure recording, sleep endoscopy, and blood-gas analysis. For additional screening, genetic analysis, hearing tests, gastroscopy, echocardiography and electrocardiography were used.

About three-quarters of respondents used a sleep study system. Flexible endoscopies were performed far more frequently than rigid endoscopies (63% vs 16%). Respondents considered presurgical assessment, severe airway obstruction, or suspicion of other airway anomalies to be the main indications for a rigid endoscopy.

Table 1
Definition of Robin sequence (RS).

Features obligatory for a diagnosis of RS	
Mandibular hypoplasia	39 (39%)
Retrognathia	52 (52%)
Micrognathia	65 (65%)
Cleft palate	75 (75%)
Glossoptosis	65 (65%)
Macroglossia	7 (7%)
Clinical airway obstruction	47 (47%)
Airway obstruction proven by a diagnostic modality	11 (11%)
Feeding difficulties	19 (19%)
Other	2 (2%)
Distinction retrognathia, micrognathia and mandibular hypoplasia	
Yes	35 (35%)
No	65 (66%)
Distinction between isolated and non-isolated RS	
Yes	54 (54%)
No	47 (47%)
Most often-mentioned combinations of features obligatory for a diagnosis of RS[a]	n = 101
Mandibular hypoplasia and/or retrognathia and/or micrognathia AND glossoptosis AND cleft palate	25 (25%)
Mandibular hypoplasia and/or retrognathia and/or micrognathia AND cleft palate	12 (12%)
Mandibular hypoplasia and/or retrognathia and/or micrognathia AND clinical airway obstruction AND glossoptosis AND cleft palate	10 (10%)

[a] Mandibular hypoplasia, retrognathia and micrognathia have been pooled together in this table.