International Journal of Pediatric Otorhinolaryngology 115 (2018) 77–81

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

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

Association between symptomatic submucous cleft palate and otologic disease: A retrospective review

Adam Kwinter[a], Ann Dworschak-Stokan[b], Josee Paradis[a], Murad Husein[a,*]

[a] Department of Otolaryngology- Head and Neck Surgery, Children's Hospital, 800 Commissioners Rd E, London, ON, N6A 5A5, Canada
[b] Thames Valley Children's Center, 779 Base Line Rd E, London, ON, N6C 5Y6, Canada

ARTICLE INFO

Keywords:
Submucous cleft palate
Otologic disease
Otitis media with effusion
Velopharyngeal insufficiency
Velopharyngeal dysfunction

ABSTRACT

Introduction: Submucous cleft palate (SMCP) is the most common congenital malformation of the posterior palate and characterized in part by palatal muscle diastasis. Patients with SMCP are considered symptomatic when suffering from velopharyngeal insufficiency which leads to hypernasality of speech and excess nasal air emission. SMCP results from defects in the muscles thought to be responsible for regulating the patency of the eustachian tube which can lead to a number of middle ear pathologies and ultimately hearing loss. Hearing loss, especially at a young age, can make speech acquisition a challenge. The purpose of this study was to review patients with symptomatic SMCP (SSMCP) who have presented to our center and report on the association between SSMCP and otologic disease.

Methods: This study was a retrospective review of all patients presenting to our center between 2004 and 2016. Patients with SSMCP were identified through a patient database maintained on site. Inclusion criteria were diagnosis of SMCP and documentation of the presence or absence of otologic disease. Once identified patient records were reviewed and various demographic and patient factors were recorded. Patients were then grouped into cohorts of those with and without otologic disease and those requiring tympanostomy tubes and not. T-tests were then used to compare various factors between cohorts.

Results: A total of 73 patients were identified and 58 met inclusion criteria (mean age 5.8 years, male:female 36:22). 27.5% of patients were diagnosed with a genetic condition. 69% of patients had otologic disease and 47% required at least one set of bilateral tympanostomy myringotomy tubes. The cohort with ear disease tended to have worse scores on ACPA testing though this was not statistically significant. There were no significant demographic differences between the cohorts.

Conclusions: Patients with symptomatic SMCP suffer from a higher incidence of otologic disease and bilateral myringotomy tube placement than the general population. This is an important potential comorbidity in SSMCP patients as it can act as a further barrier to speech acquisition. Further research is needed to determine if otologic disease can be used for prognostication or treatment purposes.

1. Introduction

Submucous cleft palate (SMCP) is the most common congenital malformation of the posterior palate [1]. It is characterized by the classic triad of a bony notch in the posterior edge of the hard palate, bifid uvula, and palatal muscle diastasis [2]. Clinically palatal muscle diastasis is seen as a central divot on nasopharyngoscopy or zona pelucida on intra-oral exam. This condition is unique among malformations of the palate in that a layer of mucous membrane overlies the submucous cleft palate. As such, diagnosis is often delayed until speech development. While most individuals with SMCP are asymptomatic, when symptomatic SMCP can cause inability of the velopharyngeal sphincter to perform effectively (termed velopharyngeal insufficiency or VPI) leading to hyper nasality of speech and excess nasal air emission [1]. Patients are deemed to be symptomatic when they have any degree of VPI.

Due to the anatomical abnormalities present in SMCP it is reasonable that a large number of SMCP patients would suffer from concomitant eustachian tube dysfunction (ETD). This is secondary to the dysfunction of the tensor veli palatini muscle which is thought to be involved in regulating the patency of the pharyngotympanic tube (ET) [3]. Decreased patency of the ET leads to a failure of the middle ear to ventilate and can cause build up of fluid (effusion), negative pressure and tympanic membrane contractions termed otitis media with effusion

* Corresponding author. Children's Hospital, LHSC Victoria Hospital, 800 Commissioners Rd E, London, ON, N6A 5A5, Canada.
E-mail address: Murad.husein@lhsc.on.ca (M. Husein).

https://doi.org/10.1016/j.ijporl.2018.09.022
Received 28 April 2018; Received in revised form 19 September 2018; Accepted 21 September 2018
Available online 22 September 2018
0165-5876/

(OME) [4]. Additionally, ETD is also associated with ossicular fixation, tmypanosclerosis and higher levels of hearing loss [5]. While recurrent middle ear disease can be highly uncomfortable for the patient it can more importantly lead to reversible or irreversible hearing loss if not appropriately managed [4]. This is especially significant in the formative years when children are acquiring speech. Even more significantly, given the anatomical challenges that children with SSMCP must overcome in order to become proficient with language, an additional insult in the form of hearing disability can be catastrophic [6,7]. As such, ear pathologies affecting hearing are an important consideration when treating patients with symptomatic submucous cleft palate (SSMCP).

While the incidence and to some extent natural history of ear pathology is well documented among the overt cleft population, there is little research in this area among the SMCP population. Ha in 2013 reported on a population of 92 patients with SMCP and found that 56% of patients had recurrent otitis media, 49% had recurrent OME and 67% of patients required at least one set of bilateral myringotomy tubes (BMTs) [8]. Harris 2013 looked at the incidence of cholesteatoma among all cleft patients and found that 1% of 194 SMCP patients did have cholesteatoma [9]. Zheng 2009 found that 12/40 SMCP patient ears had hearing loss on pure tone average (PTA) [10]. Reyes 1999 found in a study of 66 patients that 44% of SMCP patients had middle ear disease and 35% had conductive hearing loss (CHL) [11].

The purpose of this study was to review patients with SSMCP who have presented to our centre and report on the association between SSMCP and otologic disease.

2. Methods

A retrospective consecutive cohort was evaluated. The population was all patients presenting with SMCP to the Otolaryngology - Head and Neck Surgery service at Children's Hospital London Health Sciences Center between 2004 and 2016. Patients were identified through a database of patients with SMCP maintained on site. Inclusion criteria were a diagnosis of SMCP and documentation of the presence or absence of otologic disease.

Charts were reviewed to obtain data for age of presentation, presenting complaint, medical and genetic comorbidities, pre and post-operative American Cleft Palate Association (ACPA) perceptual assessment score, pre and post-operative nasometry scores, pre and post-operative audiometric data, surgical history and history of OME or other otologic disease.

The patients were subsequently grouped into cohorts of no otologic disease (nOD) and otologic disease (OD). The OD cohort was further divided into patients requiring bilateral myringotomy tympanostomy tube placement (BMT) and those who did not require BMT placement (nBMT). Paired t-tests were used to compare various values between the groups.

The study was approved by the Western University Research Ethics Board (HSREB 108883).

3. Results

Seventy-three patients were identified as having presented to our center with a SSMCP during the study period and 58 met inclusion criteria (see Table 1). Average age at presentation was 5.8 years and ranged from birth-15 years of age. Thirty-six patients were male and 22 were female. Sixteen suffered from some form of genetic condition which is listed in Table 2. As we are a referral center all patients were symptomatic from their SMCP having some degree of VPI, with presenting complaints of either speech dysfunction (usually excess nasal air emission), nasal regurgitation or both (see Table 3).

There were 40 patients in the OD cohort (summarized in Fig. 1) as follows: Otitis media with effusion (OME) 30, tympanic membrane (TM) perforation 1, ETD without OME 1, myringostapediopexy 1, OME with congenital sensorineural hearing loss (SNHL) 3, recurrent acute otitis media without effusion 4. Of these 40 patients, 31 received at least one set of BMTs at an average age of 2.9 years, 1 patient underwent myringotomy without tube placement and 8 needed no surgical management of their otologic disease. Of the patients with SNHL there was no underlying cause identified. No patients had vestibular pathology.

Table 1
Demographic data of patients presenting to with SSMCP.

	n	Gender	Average Age (years)	Patients with a Genetic Syndrome	Pre-Op PTA (dB)
All	58	M:36 F:22	5.8	16 (27.5%)	15.8
nOD	18 (31%)	M: 12 F: 6	6.8	7 (38%)	6.6*
OD	40 (69%) BMT: 31 (47%) nBMT: 9 (15%)	M: 24 F: 16	5.3	9 (22.5%)	18.3*

nOD – no otologic disease, OD – otologic disease. BMT – bilateral myringotomy tubes, nBMT – no bilateral myringotomy tubes. *p-value < .05 on two-tailed *t*-test.

Table 2
Summary of genetic conditions among SSMCP patients.

	nOD	OD	Total
22q11.2 Deletion	**4** (10%)	**3** (16.7%)	**7** (12%)
Treacher Collins Syndrome	**1** (2.5%)	**1** (5.5%)	**2** (3.4%)
Klinefelter Syndrome	**0**	**2** (11.1%)	**2** (3.4%)
Kabuki Syndrome	**0**	**1** (5.5%)	**1** (2.5%)
Mobius Syndrome	**0**	**1** (5.5%)	**1** (2.5%)
Unspecified skeletal dysplasia	**1** (2.5%)	**0**	**1** (2.5%)
Stickler Syndrome	**1** (2.5%)	**0**	**1** (2.5%)
Facioscapulphumerol muscular dysplasia	**0**	**1** (5.5%)	**1** (2.5%)

With respect to genetic syndromes 49% (n = 7) of the patients in the nOD group had a syndrome while only 22% (n = 9) of the patients in OD cohort had a syndrome.

Not all patients underwent audiometric testing. Six patients from the nOD group underwent audiometric testing and had a PTA of 6.6 dB. Fifteen of the patients in the OD cohort underwent audiometric testing pre-BMT and submucous cleft correction and had a PTA of 18.3 dB. Of the OD patients with pre-op testing 9, had post-op audiometric testing with a PTA of 12.5 dB. Four patients from the OD cohort without pre-operative testing had post-operative testing for a total of 13 patients in the OD cohort having post-operative audiometric testing with a PTA of 11.2 dB.

With respect to management, all patients received speech language pathology assessments and treatment. Forty-six patients required surgical management with 42 undergoing a furlow palatoplasty, 1 underwent a primary pharyngeal flap (p-flap) procedure, 10 did not require surgical correction, 2 were awaiting surgery, and 1 patient transferred their care to another center and was subsequently lost to follow-up before undergoing surgery. Nine patients required a secondary procedure in the form of a p-flap procedure (see Table 4). Of the patients who did not require surgical correction 4 were in the nOD group.

Of the patients with OD at least 83% had full resolution of otologic symptoms (see Table 5). Of the patients who did not have resolution of their symptoms, 1 patient suffered from recurrent cholesteatoma, 1 had chronic tube otorrhea and 1 suffered from a myringostapediopexy. Two patients were lost to follow-up.

Amongst the OD cohort there was a higher rate of BMT placement in those who underwent a palatoplasty than those who did not have