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Review Article

A general review of the otolaryngologic manifestations of Down Syndrome

Christopher J. Chin [a], Maria M. Khami [b], Murad Husein [a,*]

[a] Department of Otolaryngology – Head and Neck Surgery, Western University, London, Canada
[b] Schulich School of Medicine & Dentistry, Western University, London, Canada

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ABSTRACT

Objective: Down Syndrome (DS) is the most common chromosome abnormality in liveborn children. Otolaryngologists frequently encounter these patients in their practice; in one survey, 50% of DS patients had been seen by Otolaryngology at least once. As such, it is essential that the practicing Otolaryngologist is aware of the otologic, rhinologic, and laryngologic manifestations of this complex disease and comfortable in the management and treatment of these unique patients. Our goal was to provide this information in a concise and definitive document.

Methods: A comprehensive literature review using PubMed was completed. The terms "Otolaryngology", "Head and neck", "Ear, nose, throat", "Down Syndrome", and "Trisomy 21" were searched in various combinations. Applicable articles that discussed the Otolaryngologic manifestations of Down Syndrome were included.

Results: In total, fifty articles were included for review. The Down Syndrome child tends to have smaller external ear canals, have higher rates of chronic ear disease, and may present with conductive, sensorineural, or mixed hearing loss. As such, DS patients should receive behaviouralaudiological testing every 6 monthsand annually after the age of three in addition to closer follow-up by an Otolaryngologist if tympanic membranes cannot be visualized or if the external auditory canals are significantly stenosed. Management should involve close follow-up and a low threshold for PE tube placement to reduce the risks for speech and language delay. Chronic rhinitis in the Down Syndrome patient is common. Retrognathia, hypotonia, and macroglossia can all cause obstructive sleep apnea (OSA) in this population and therefore each DS patient should get an overnight polysomnograph. Subglottic stenosis, vocal cord paralysis and laryngomalacia are not infrequently seen in the Down Syndrome patient. To reduce acquired subglottic stenosis, endotracheal tubes that are at least two sizes smaller than what is appropriate for the patient's age should be used.

Conclusion: Down Syndrome is common and there are many Otolaryngologic manifestations. We recommend that this patient population visit an Otolaryngologist on a regular basis and that the practicing Otolaryngologist is comfortable with the management and treatment of the unique challenges faced with these children.

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* Corresponding author at: Department of Otolaryngology – Head and Neck Surgery, London Health Sciences Center, B3-453, 800 Commissioners Road E., London, ON N6A 5W9, Canada. Tel.: +1 519 685 8500xX58184.
E-mail address: Murad.Husein@lhsc.on.ca (M. Husein).

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1. Introduction

Down Syndrome (DS) is the most common chromosome abnormality in liveborn children with an incidence of 1 in 700 live births [1]. Down Syndrome was first described in 1866 by John L. Down, and is most often caused by a nondisjunction error that results in three copies of chromosome 21 ('Trisomy 21') [2,3]. Many of the anatomic changes in DS are in the head and neck region and cause sequelae amenable to treatment by an Otolaryngologogist. These anatomic abnormalities can prevent DS patients from reaching their full developmental capacities and can hinder their quality of life. In one survey, 50% of DS patients had been seen by an Otolaryngologist at least once [4]. There are also indications that perhaps some of the medical conditions in DS are being under-recognized and undertreated by physicians and parents; in a retrospective study by Roizen et al., over 40% of DS patients had some degree of sclerosis of the mastoid air cells, suggestive of an incompletely treated otitis media with effusion (OME) [5]. A prospective case control by Marcus et al. showed that 60% of children who had negative histories for OSA were found to have abnormal polysomnograms (PSG) [6]. As medical science advances and these patients continue to live longer [7], the Otolaryngologic care of the DS patient is no longer isolated to the Pediatric Otolaryngologist. Therefore, it is important for otolaryngologists to be comfortable in managing and treating these patients. The goal of our article is to provide a concise updated review of the common Otolaryngologic manifestations of this common entity.

2. Methods

A literature review of Pubmed (http://www.pubmed.org) was performed. The terms "Otolaryngology", "Head and neck", "Ear", "Nose", "Throat", "Down Syndrome", and "Trisomy 21" were searched in various combinations. Inclusion criteria included being published in the English literature and focusing on either the otologic, rhinologic, laryngologic, or perioperative aspects of Trisomy 21. Exclusion criteria included isolated case reports. The references of applicable articles were also analyzed to ensure that topical items that were missed in the literature search were included.

3. Results

In total, 50 articles met criteria for inclusion. Articles ranged from 1970 to 2014 with the majority being published after 2000. The most applicable findings are presented below, divided by anatomical subsite.

3.1. Otologic

The pinna in the DS child is, on average, 2 standard deviations smaller than the general population [8]. This has the effect of decreasing sound localization and sound concentration. Not only is the pinna small, but the external auditory canal is often stenosed (in up to 50% of children with DS) [9]. This can make examination of the ear difficult for the General Practitioner leading to missed effusions, and also predisposes the child to cerumen impaction, which can lead to a conductive hearing loss (CHL). As such, Shott [10] recommended that children with stenotic canals be assessed by an Otolaryngologist every three months until the age of 2–3 when their canals have grown to ensure effusions are not missed. In addition, regular cerumen debridement of ears in patients where cerumen impaction poses a problem is recommended to improve visualization of the tympanic membrane.

Down Syndrome children have an increased rate of chronic ear disease [9,10]; the causes are likely multi-factorial. The incidence of chronic ear disease in the normal pediatric population has not been well defined. Casselbrant et al. conducted monthly examinations using otoscopy and tympanometry in children between the ages of two and six and found that 53% of the children had at least one middle ear effusion in the first year; this had increased to 61% in the second year [11]. A recent study showed the prevalence of OME at age one is up to 93% while at age 5, the prevalence is up to 68% in the DS population [12]. In DS patients, up to 80% will have conductive hearing loss as a result of OME [13]. Park et al. showed a 37.9% rate of CHL due to serious otitis media in a study analyzing 323 DS patients [14]. A prospective observational study by Schwartz et al. concluded that the Eustachian tube has been found to be smaller on average in DS children [8]. As well, the generalized hypotonia in DS patients impacts the function of the tensor veli palatini muscle, which is felt to lead to poor Eustachian tube opening and subsequent poor aeration of the middle ear space [8,10]. This also predisposes to OME, which in turn can cause a significant CHL.

Sensorineural hearing loss (SNHL) can also be an issue with a child who has Down Syndrome. The incidence is not clearly defined; the incidence ranges from 4 to 55% in DS children [15–17]. The hearing loss is usually progressive in nature and in the high-frequency range. In Park et al.'s study, 26.2% of the 323 DS patients did not pass their newborn hearing screening and the average time to diagnose a SNHL was 485 $\pm$ 601 days [14]. Saliba et al. showed a SNHL rate of 41% in their Down Syndrome group [18]. Inner ear abnormalities that can be seen in DS include internal auditory canal hypoplasia, enlarged vestibular aqueducts, cochlear nerve hypoplasia among others [17]. When these abnormalities are present along with persistent OME, cerumen impaction, or a different cause of CHL, a mixed hearing loss will be present and can be seen on the audiogram. Early and accurate diagnosis, combined with aggressive medical and surgical interventions in the DS population soon after birth has led to decreased rates of hearing loss [10,17]. Shott et al. showed that only 2% of DS patients had persistent hearing loss after treatment of hearing loss secondary to chronic otitis media with pressure equalization (PE) tubes [10]. However, conflicting studies have also showed that DS patients did not experience the same benefit from PE tubes as a control group [19]. Similar to the general population, observation is the recommended first line treatment for the first 3 months after diagnosis. PE tubes are indicated in cases of severe hearing loss, if there was a risk of pathological changes of the eardrum, or if there is failure of the effusion to clear after 3 months [19]. Often, insertion of PE tubes is delayed until the ear canal is large enough to allow for insertion [19]. Paulson et al. recently showed that

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