Otolaryngologic Manifestations in Children with Down Syndrome

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Introduction

It is common for children with Down Syndrome (DS) to have ear, nose and throat problems. (Mitchell 2003, Vernail 2004, Shott 2006a). This includes a high incidence of chronic ear infections and associated conductive hearing loss, as well as sensorineural hearing loss. Obstructive sleep apnea and sleep disturbed breathing are also very common, occurring in as many as 60% of children as young as 3-4 years of age (Shott 2006b). This incidence increases to as hight as 90-100% as the children grow older (Marcus1991, Dyken 2003). The subglottic and tracheal airways of children with DS are smaller than in other children of the same age (Shott 2000b). This is of particular importance if surgery under general anesthesia with intubation is needed. Tracheomalacia, with partial collapse of the trachea with respirations and stridor is not uncommon. Because of delay in development of the immune system and the midfacial hypoplasia with smaller nasal passages and smaller paranasal sinuses, young children are more prone to upper respiratory tract infections and rhinorrhea (Gershwin 1977, Spina 1981, Strome 1981).

Chronic Otitis Media

Chronic ear infections are common in children with DS, occurring in up to 95% of children (Strome 1981, Shibahara and Sando, 1989; Yamaaguchi 1990; Mitchell 2003, Lee 2007). Etiologic factors include mid-face hypoplasia with a contracted nasopharynx, an abnormal shape in the Eustachian tube (Shibahara 1989), a higher incidence of upper respiratory infections due to immaturity and delay in the immune system (Gershwin 1977; Spina 1981), as well as poor function of the tensor veli palatine muscle of the soft palate, the muscle responsible for opening and closing the Eustachian tube (Strome1991).

We currently do not have data as to the length of time that this persistent Eustachian tube dysfunction lasts but it appears that the chronic ear infections continue much longer and to an older age in children with DS compared to typical children.

Studies have shown that aggressive medical and surgical management of the chronic otitis media results in better hearing levels and a lower occurrence of tympanic membrane perforations and cholesteatoma (Shott 2001, Shott 2003, Lee 2007).

Children with DS should have an initial audiologic evaluation in the first month of age following Universal Newborn Screening and AAP guidelines (Amer Acad Ped 2007). If a hearing loss is identified, referral to an Otolaryngologist is needed (See Audiology Section).

Up to 50% of newborns with DS will have stenosis of the external auditory canals (Strome 1992). Although the canals slowly grow during the first 3 years of life, until routine office examination of the tympanic membranes is possible, many children will need examination of the ears under a microscope with removal of cerumen in order to adequately rule out otitis media, both acute infections and middle ear effusions. It is suggested that for children with small ear canals, these examinations be done every three months to guarantee that the child does not go for an extended period with unrecognized middle ear fluid or infection. For most, this will require a referral to an Otolaryngologist.

For children with normal size ear canals that allow for examination of the tympanic membranes with an otoscope, ears should be monitored for both recurrent acute otitis media and chronic middle ear fluid. Changes in the tympanic membrane such as retraction and retraction pockets should be monitored with referral to an Otolaryngologist if retraction fails to improve.

Placement of pressure equalization tubes (PET's) should be considered when fluid has not resolved within three months and/or if the child has had three consecutive acute ear infections (AAFM, AAO,AAP guidelines 2004). It has been shown that children with DS with PET's in place have a 3.6 times higher chance of having normal hearing compared to audiograms done when PET's were not in place (Shott 2003). It has also been shown that there is a high incidence of under-diagnosis and under-treatment of ear infections in children with DS (Roizen 1994).

Due to **more prolonged Eustachian tube dysfunction**, multiple sets of PET's may be needed for the individual child. **Children should continue to have their ears monitored at least every six months** as long as chronic retraction of the ear drums, ear effusions, and/or recurrent infections persist. For children with minimal ear problems and normal hearing, any changes in previously normal hearing tests should be evaluated to rule out temporary hearing loss from either middle ear fluid or otitis infection. Referral to an Otolaryngologist may be needed for further evaluation if the cause in the hearing loss change cannot be determined or does not respond to medical management. A recent study by Lee et al. showed that will aggressive medical surveillance and pro-active surgical treatment of chronic ear disease, the need for more extensive otologic surgery can be minimized (Lee 2007). More frequent examinations and continued monitoring as the child grows will be needed for children with persistent chronic ear problems and hearing loss compared to children with normal hearing and minimal ear infections.

Audiologic Testing

Children with DS have a three times higher incidence of chronic ear disease and secondary hearing loss from chronic ear infections than other children with developmental delays (Brooks1972, Dahle 1986). Blaser et al. have shown through evaluations of CT scans and MRI a high incidence of inner ear abnomalities in DS (Blaser 2006). Multiple studies have shown a relationship between even mild hearing loss and educational, language, and emotional development (Holm 1969, Brooks 1972, Balkany 1979a). Even mild hearing loss can affect a child's articulation skills (Dobie 1979, Bess 1985). Initial studies

evaluating the incidence of hearing loss in children with DS reported rates as high as 78% (Balkany 1979a, Balkany 1979b). However, with more fastidious monitoring and more aggressive management, both medical and surgical, this incidence can be much lower (Shott 2001, 2003). Thus monitoring of hearing levels in addition to frequent ear examinations are imperative in order to maximize the child's otologic and audiologic status. Audiologic testing can be an important tool to identifying reversible hearing loss, especially in the very young children when ear canal stenosis has its greatest effect and physical examination is often difficult.

The current health care guidelines for children with DS, as prescribed in 2001 by the American Academy of Pediatrics Committee on Genetics recommend audiologic testing at birth and then every six months up to age three years, with annual testing after three years of age (American Academy of Pediatrics, 2001). Studies show that reliable, ear specific results by behavioral audiometry cannot usually be achieved in children with DS younger than 3½ years of age (Maurizi 1985). This was further confirmed by Shott, Heithaus et al. In their study, at 3 years of age, only 12% of the children with DS were able to do ear specific testing and at 4 years, only 41% were able to achieve ear specific results (Shott 2005). Testing protocols should therefore be determined by the child's developmental levels and not by chronological age. The goal is to have ear specific measurements of pure tones as well as speech awareness levels, and eventually speech reception levels and a determination of speech discrimination abilities. Therefore hearing evaluations should continue every six months until ear specific testing is achieved. If the hearing is normal, then annual follow-up hearing tests would be adequate. More frequent audiologic evaluations are necessary in the presence of a hearing loss.

Amplification with hearing aids should be considered even if there is only a mild hearing loss, especially in view of the data linking mild hearing loss with delays in educational, emotional, and language development (Holm 1969; Brooks 1972; Balkany et al., 1979a). This is of particular importance for children with DS where the expressive language skills are delayed compared to cognitive abilities. Statistically significant differences in I.Q. levels were demonstrated between children with mild hearing loss due to otitis media and matched controls. These studies, however, were all done on otherwise normal children with hearing loss. It follows as a reasonable assumption that the developmental problems associated with hearing loss may have a greater effect in children with the mental and physical handicaps associated with Down Syndrome (Balkany et al., 1979).

Due to the higher incidence of chronic Eustachian tube dysfunction and more prolonged problems with chronic otitis media in DS and the older age that children with DS "grow out" of their chronic ear problems, hearing tests may be needed more frequently if the child continues to experience chronic otitis media, including acute infections and chronic Eustachian tube dysfunction with chronic middle ear fluid and/or retraction of the tympanic membranes. Audiologic evaluations would become annual with the resolution of the middle ear pathology and once ear specific testing is achieved and normal or stable.

Obstructive Sleep Apnea Syndrome

Studies report a **50-100% incidence of obstructive sleep apnea syndrome in individuals with DS with almost 60% of children with DS having abnormal sleep studies by age 3.5-4 years** (Shott 2006b). Evidence shows that these numbers increase as children grow older. (Marcus 1991, Levanon 1999, Dyken 2003). Fitzgerald et al showed a 97% incidence of OSA in children with DS who snored, ages 0.2 to 19 years (4.9 years mean age) (Fitzgerald 2007). **Predisposing factors include midface hypoplasia, mandibular hypoplasia, a relative macroglossia, medially displaced tonsils, adenoids sitting in a contracted nasopharynx and thus causing more obstruction, as well as hypotonia of the upper airway with resultant collapse at multiple levels of the airway during sleep. Increased upper airway infections and nasal secretions, obesity and hypotonia further contribute to both oropharyngeal and hypopharyngeal collapse and obstruction with sleep. A high incidence of gastroespophageal reflux (GER) occurs in children with DS**. The GER can cause edema of the posterior pharyngeal area, decreasing the overall size of the airway and contribute to the sleep apnea.

Unfortunately, the ability of parents to predict sleep abnormalities in their children with DS has been shown to be poor (Shott 2006b, Marcus 1991, Ng 2006). Sleep disturbed breathing has been shown to affect cognitive abilities, behavior, growth rate and more serious consequences of pulmonary hypertension and cor pulmonale (Rowland 1981, Levine 1982, Southall 1987, Marcus 1991 Bonnet, 1989). Because of the high incidence of underlying congenital cardiac anomalies in individuals with DS, there is a higher risk of development of the more severe complications (Jacobs 1997).

A sleep study or polysomnogram continues to be the gold standard test from which to evaluate sleep disturbed breathing and sleep apnea.

Starting at birth, primary care physicians should actively inquire about restless sleep, snoring, respiratory pauses or apneas, and/or increased respiratory effort during sleep. Sleep positions should also be discussed such as sleeping sitting up, sleep with the neck hyperextend, or sleeping bent forward at the waist in a sitting position. These questions should continue through out the individual's life. If there is any question of airway disturbances during sleep, a referral to an Otolaryngologist should be done to determine if a sleep study and/or surgical intervention is needed. **Because of the higher rate of respiratory complications after removal of the tonsils and adenoids (T&A) in children with DS, overnight observation in the hospital after this surgery is recommended** (Bower 1995). Although T&A is the most common initial surgical intervention, studies have shown that **residual airway obstruction after this surgery is possible** and further interventions may be needed, both surgical and medical (Merrell 2006, Shott 2006b, Donaldson 1988, Jacobs 1996, Shott and Donnelly 2004). This has recently also been shown to be more common than previously believed in typical children with pre-operative sleep apnea. Mitchell in 2007 showed a **10-20% incidence of persistent sleep apnea in a group of 79 typical children after T&A**. Tauman et al., using a much more strict definition of

surgical cure showed **complete normalization of all components evaluated in a sleep study in only 25%** of their test population of 'typical' children (Tauman 2006). This compares to the 5% total success rate seen in the paper by Shott, Amin et al. where a similarly strict definition of "cure" was used in a group of children with DS (Shott 2006b). If 'cure' is more akin to the definitions used in the study by Mitchell, **50-70% of the children with DS in this study continued to have OSA after T&A.**

Surgical options include procedures addressing macroglossia and glossoptosis. Radiographic studies show that the **base of tongue as well as adenoid regrowth are some of the most common sites of residual obstruction in individuals with DS despite previous T&A** (Donnelly 2004). Medical options include weight loss, oxygen use during sleep, or continuous positive pressure application through CPAP or BiPap. Tracheostomy may also need to be considered in cases of severe sleep apnea with associated pulmonary hypertension, severe hypoxemia and/or cardiac complications.

Postoperative sleep studies or at a minimum, an overnight oximetry study, should be done following surgical intervention to ensure adequate treatment and rule out persistent sleep disturbed breathing as it has been shown that **at least 70% of children with DS will continue to have some degree of upper airway obstruction despite removal of the tonsils and adenoids** (Donaldson 1988, Jacobs 1996, Shott and Donnelly 2004, Merrell 2007).

Airway abnormalities – Laryngeal and Tracheal

In addition to the small oropharynx and nasopharynx, children with DS have a smaller subglottic and tracheal airways than typical children. An association between DS, stridor and/or subglottic stenosis is commonly discussed in the anesthesia literature (Sherry 1983). It has been shown that **children with DS**, when compared to other children, matched for age and weight, **require an endotracheal tube that is two sizes smaller when intubated appropriately, such that there is an air leak around the tube (Shott 2000b). The hypotonia seen in DS, also affects the larynx and the trachea, and laryngomalacia and tracheomalacia with associated stridor is not uncommon**. Post-operative airway complications are higher in children with DS than typical children, usually caused by airway issues. (Borland 2004, Mitchell 1995, Sherry 1983). **The combination of the smaller airway, a high incidence of gastroesophageal reflux and a high percentage of children with DS procedures done under general anesthesia places them at higher risk for development of subglottic stenosis** (Boseley 2001).

Although no ongoing monitoring is required, primary care physicians need to be aware of potential airway problems when their patient is scheduled for surgery, especially when intubation is expected to take place, especially if the child undergoes surgery in a center not accustomed to caring for children with DS. **Recurrent croup should be treated aggressively**, especially since it is most likely that the child is starting off with a subglottic airway smaller than usual. **Treatment of gastro-esophageal reflux (GER) should be considered, especially in cases of recurrent croup or if significant edema is seen in the**

posterior oropharynx. Mitchell found a high incidence of GER in his review of a cohort of children with **DS and GER has been linked as a risk factor for recurrent croup** (Mitchell 2003). There is also a higher incidence of GER in children who have laryngomalacia (Halstead 1999). Although initial reports suggested a lower success rate in surgical treating subglottic stenosis in children with DS, more recent reports have shown similar success rates to other children (Boseley 2001).

Because of the higher potential for post-operative airway problems after a general anesthesia, especially if intubation of the airway occurs, overnight hospitalization, even after a short procedure should be considered in children with DS (Moos 2005, Mitchell 2003) and is strongly advocated after adenotonsillectomy (Bower 1995).

Chronic rhinitis / sinusitis

The **chronic runny nose or chronic rhinitis** was in the past frequently referred to as something just "part of Down syndrome" and was noted to **improve as the child got older** (Strome 1981). However, the chronic rhinitis should not be accepted as an inevitable and untreatable condition and evaluation and management of any chronic rhinitis should be addressed aggressively. Radiographic studies have shown **abnormal development of the frontal, maxillary, and sphenoid sinuses, including hypoplasia or lack of pneumatization of the paranasal sinuses (Miller 1986). The delay in the development of the immune system also contributes to chronic problems with rhinitis in DS (Gershwin 1977, Spina 1981).The use of saline nasal drops, sprays and irrigations has long been used for treatment of chronic rhinitis** and recently further confirmed as an effective treatment in adults (Pyonnen 2007). For children with midface hypoplasia and thus smaller than usual nasal passages, this is an invaluable and effective treatment. Further evaluation and treatment of chronic sinusitis should follow similar pathways of treatment as would be used in typical children.

Education of parents should also include discussions on the **need to eliminate any cigarette exposure,** especially in view of the delay in the immune system development and higher incidence of upper respiratory infections in children with DS.

Other causes of **nasal obstruction such as adenoid hypertrophy or nasal turbinate enlargement should be considered**. Studies have shown that adenoid regrowth despite a history of previous adenoidectomy is not uncommon in DS (Donnelly 2004). Family history, in regards to environmental allergies should be discussed and allergy testing be considered. This should include an **assessment of the immune system, including IgG (both total IgG and subclass levels, IgA, IgM, IgE, titers to diphtheria, tetanus and *S. pneumoniae*).**

Medical management of rhinitis/sinusitis includes the use of appropriate **antibiotics, nasal steroid sprays, saline nose sprays, antihistamines and decongestants**. Similar to typical children, if infections continue despite medical management, referral to allergists and/or otolaryngologists may be needed to assess the need for further treatment including immunotherapy and surgical interventions.

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