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The prevalence of airway problems in children with Down's syndrome

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ABSTRACT

Aim: Airway disorders are common in children with Down's syndrome. We report the findings on airway endoscopy in a birth cohort of children from a well-defined geographical area, in order to estimate true population prevalence of airway problems in children with Down's syndrome.
Method: Retrospective case note review over a 20-year period between 1993 and 2013 for all children in Greater Glasgow born with Down's syndrome, identified through the hearing surveillance programme. All children undergoing airway endoscopy under general anaesthesia for investigation of potential airway symptoms (stridor, hoarseness, recurrent croup and difficulties with intubation/extubation) were studied in detail to identify the number with laryngeal, tracheal or bronchial pathology.
Results: All 239 children (F:M = 1.15:1) were reviewed. Of these, 39 (16.3%) underwent microlaryngoscopy-bronchoscopy under general anaesthesia for airway symptoms. The main presentations were stridor (9), extubation problems (12) and exacerbations of recurrent croup (7). Thirty-three were found to have at least one airway diagnosis (13.8%) including trachaeobronchomalacia (17), laryngeal cleft (2), laryngomalacia (2), tracheal compression (2), vocal cord paralysis (1), acquired tracheal stenosis (2) and symptomatic subglottic stenosis (14).
Conclusion: Laryngo-tracheo-bronchial pathology is much more common in children with Down's syndrome than in the general population, particularly subglottic stenosis and tracheal problems.

1. Introduction

Down's syndrome (Trisomy 21) is the most common genetic disorder in humans, with an incidence rate of 1/670–770 births [1,2]. These children are predisposed to a number of systemic clinical problems, including cardiac and gastrointestinal abnormalities. Problems affecting the ear, nose and throat are very common [3] and their early identification and management can have significant benefits for health and well-being.

It is known that children with Down's syndrome have a higher incidence of otolaryngological problems than the general paediatric population, particularly hearing impairment. 50% of children with Down's syndrome having been seen by an otolaryngologist at least once [4]. Airway obstruction at the pharyngeal level is common: by age 5, 79% have either current symptomatic upper airway obstruction, or a history of adenotonsillectomy [5] and the prevalence of obstructive sleep apnoea on polysomnogram is 57% [6]. The aetiology is multifactorial, including a narrowed airway [7] but may include jaw abnormalities, macroglossia, adenotonsillar hypertrophy, soft palate hyperplasia and hypotonia [2,8,9].

Airway disorders at the laryngeal and tracheal levels are also common, particularly tracheobronchomalacia [See Fig. 1], subglottic stenosis (congenital and acquired) and tracheal stenosis [10,16]. Every child with Down's syndrome has a degree of subglottic stenosis as a fundamental feature of the syndrome and most of these are asymptomatic. As the trachea is smaller in children with Down's syndrome, paediatric anaesthetists tend to routinely use smaller endotracheal tubes in children with Down's syndrome than they would for other children of the same age, especially in children with cardiac problems. Failure to do so can lead to a high incidence of post-extubation stridor [11–13] and may lead to the development of a more severe, symptomatic airway stenosis. The incidence of symptomatic subglottic stenosis is 6% in children with Down's syndrome compared to only 0.63% in the general paediatric population [14,15]. Children with Down's syndrome make up 4% of all paediatric subglottic stenosis cases

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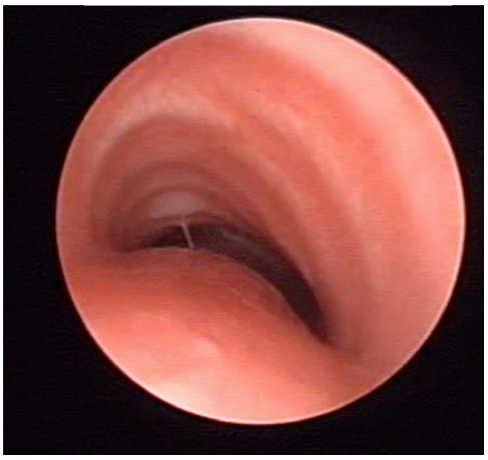

Fig. 1. Appearance of the lower trachea at microlaryngoscopy bronchoscopy under general anaesthesia, showing severe tracheobronchomalacia in a child with Down's syndrome.

[16], and 24% of these stenoses are congenital with no history of intubation [17].

Laryngomalacia is often described as being common in children with Down's syndrome and has been described as the second commonest reason that they require otolaryngological servies [18]. Laryngomalacia in these children has been attributed to the hypotonia and gastro-oesophageal reflux disease [19].

Most studies on airway problems in children with Down's syndrome are cross-sectional studies of children, referred into a hospital department, which makes it impossible to make any realistic comment on population prevalence. For this you need to know the size of the population of children with Down's syndrome that the referrals have come from, and also that you have captured all the referrals. In Glasgow, we have a community-based, hearing surveillance clinic for children with Down's syndrome. It is the only service where such children are seen and referred after identification by geneticists, neonatologists, paediatricians (in hospital and community), audiologists and colleagues in otolaryngology. As such, we are confident that our identification of children with Down's syndrome is complete. We have a stable, well-defined population from a specific geographical area with very little migration in or out. In addition, our Scottish National Paediatric Airway Service sees all the children with major airway disorders in Scotland, and it is the only otolaryngology service that sees children in the Glasgow area.

Thus, we have a well-defined, complete birth cohort of all children who have been diagnosed with Down's syndrome in the Greater Glasgow area. We can be confident that we have captured all the children with airway problems, and hence we are able to calculate the true population prevalence of airway disorders in children with Down's syndrome, which is novel.

2. Method

Our study population comprised all children diagnosed with Down's syndrome born over a 20-year period between July 1993 and July 2013 in Greater Glasgow, as identified from the audiology records of our Down's syndrome hearing surveillance clinic. This was then cross-referenced against the Scottish National Airway Service database and personal operative logs of the airway surgeons to identify any children undergoing airway endoscopy under general anaesthesia. A retrospective case note review was undertaken to identify personal and clinical data.

Children seen in the outpatient clinic and managed conservatively are not been identified in this study. We chose to study only those who undergoing airway endoscopy under anaesthetic as this means, we are only studying those with definite symptoms deemed severe enough to require formal investigation. Our prevalence figures will therefore be conservative, as some children with very mild symptoms may have managed without recourse to general anaesthesia. As this is a national hearing surveillance clinic, we are certain that we have covered 100% of the local Down's syndrome population. According to the National Records of Scotland for 2014 [20], the total number of children aged 0–15 years in Greater Glasgow (Glasgow City plus East Renfrewshire and East and West Dumbartonshire) is 149,009—with Down's syndrome making up a proportion of 1.6 children per 1000 in Glasgow.

Retrospective case review studies such as, this are not deemed to require the full Research Ethics Committee approval process in our institution, but the study is registered with the hospital's clinical governance committee.

3. Results

239 children with Down's syndrome were identified over the 20-year period, comprising 127 girls and 112 boys (F:M = 1.15:1). Of these, 39 (16.3%) underwent microlaryngoscopy–bronchoscopy under general anaesthesia for airway symptoms. The presenting symptoms were stridor (9, 23%), recurrent episodes of croup (7, 17.9%), chronic aspiration (1, 0.03%), and failed or difficult extubation after surgery (12, 30.7%).

Thirty-three children were found to have at least one airway pathology (13.8%). The airway diagnoses identified on microlaryngoscopy-bronchoscopy are shown in Table 1; of course, the numbers in the table add up to more than 33 because some children had more than one airway pathology. Some co-existing conditions included one child with complex cardiac problems, with severe subglottic stenosis [See Fig. 2], tracheobronchomalacia and laryngomalacia, requiring tracheostomy. Subglottic stenosis and tracheobronchomalacia were found together in 5 children (15.1%).

Not all of the pathology seen was benign and a significant proportion required intervention. One of the laryngeal clefts required endoscopic repair, and both the children with laryngomalacia required supraglottoplasty. The child with acquired midtracheal stenosis from intubation injury [See Fig. 3] has managed successfully with endoscopic balloon dilatation. Of the 17 children

Table 1
Airway diagnoses after microlaryngoscopy–bronchoscopy in the cohort of 239 children with Down's syndrome.

Diagnosis	Number	% of those with pathology ($n = 39$)	% of cohort ($n = 239$)
Subglottic stenosis	14	42.4	5.9
Congenital (no intubation)	3	9	1.3
Acquired (history of intubation)	11	33.3	4.6
Laryngeal cleft	2	6	0.8
Grade 1	1	3	0.4
Grade 2	1	3	0.4
Laryngomalacia	2	6	0.8
Vocal cord paralysis	1	3	0.4
Tracheal compression	2	6	0.8
Aberrant subclavian artery			
Acquired tracheal stenosis	2	6	0.8
Tracheobronchomalacia	17	51.5	7.1
Trachea only	7	21	2.9
Bronchi only	3	9	1.3
Trachea and bronchi	7	21	2.9