Subglottic stenosis in infants and children

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ABSTRACT

Introduction: This study aimed to examine the epidemiology and outcome of subglottic stenosis in infants and children, and to evaluate the current techniques used in its diagnosis and management at the KK Women's and Children's Hospital, Singapore.

Methods: A retrospective review and long-term follow-up was conducted in all infants and children diagnosed with subglottic stenosis between January 1997 and December 2008.

Results: A total of 18 patients (nine male and nine female) with a median age of 7.5 months were identified. Two patients were diagnosed with definite congenital stenosis and 16 patients with acquired stenosis. The majority had Grade I stenosis (55.6 percent), followed by Grade II (27.8 percent) and Grade III (16.7 percent). None had Grade IV stenosis. 17 patients were intubated, and seven underwent tracheostomy. The most common surgical intervention performed was microlaryngoscopy and bronchoscopy with bougie dilation. The other surgical interventions included cricoid split, laryngotracheal reconstruction and cricotracheal resection. As of December 2008, the median duration of treatment was four years, with an overall recovery rate of 66.7 percent. The successful decannulation rate was 57.1 percent. Two mortalities were reported due to reasons unrelated to subglottic stenosis. Two patients were still undergoing treatment at the time of the study, and two were lost to follow-up.

Conclusion: Conservative management alone may be required in the majority of Grade I stenosis cases. We observed that the mean number of reconstructive procedures performed per patient increased with the increase in the severity of stenosis. Each laryngeal framework procedure has to be customised to suit the individual.

Keywords: laryngotracheal stenosis, larynx, paediatric, retrospective, subglottic stenosis

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INTRODUCTION

Subglottic stenosis (SGS) is a type of laryngeal stenosis that occurs below the glottis and above the first tracheal ring. Although rare, it is one of the most common causes of chronic upper airway obstruction in infants and children. Congenital SGS is the second most common cause of stridor in neonates, infants and children. Acquired SGS is the most common acquired anomaly of the larynx in the paediatric age group, and is the most common abnormality necessitating tracheotomy in children below one year of age.[1] The prognosis of acquired SGS in infants and children is significantly graver than that of the congenital type.

KK Women's and Children's Hospital (KKH) is a major tertiary referral centre for paediatric otolaryngological problems in the Asia Pacific region and the largest children's hospital in Singapore. This gives us the opportunity to examine the overall picture of paediatric subglottic stenosis. Our aims were to study the epidemiology and outcome of subglottic stenosis in infants and children at KKH and to evaluate our current techniques in its diagnosis and management. Since SGS can present as a life-threatening emergency and its management is challenging, such a study will help to manage future patients with this condition.

METHODS

All infants and children who were diagnosed with SGS between January 1997 and December 2008 were identified. All the cases were retrospectively reviewed and followed up, in particular, for parameters such as age, gender, race, the classification and grade of SGS, clinical presentation, history of intubation and tracheostomy, findings of airway endoscopy, procedures performed, complications and eventual outcome.

RESULTS

A total of 18 patients with SGS who required surgical intervention were identified during the study period. Two of the patients were referred from other hospitals. There were nine male and nine female patients. At the time of first presentation, the mean and median age of the patients was 24.9 months and 4.5 months, respectively (range one month to 13 years). The mean age at diagnosis of SGS was 29.4 months and the median age was 7.5 months (range one month to 13 years). The symptoms lasted an

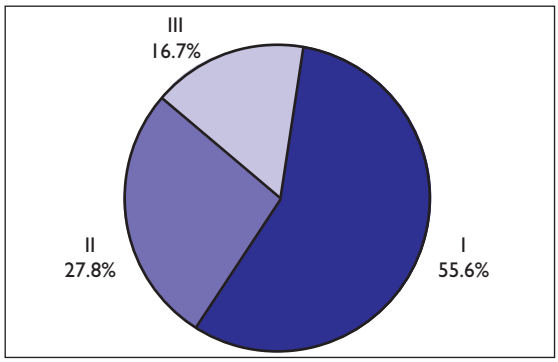

Fig. 1 Grades of subglottic stenosis.

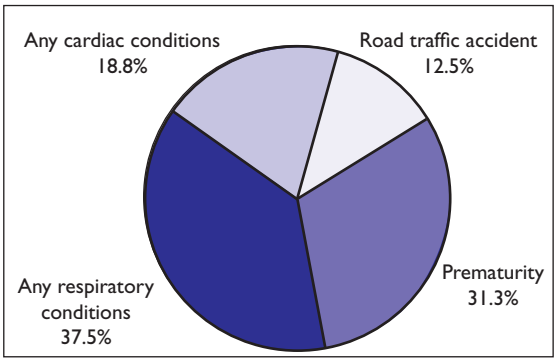

Fig. 2 Indications for ventilation via endotracheal tube for patients with acquired subglottic stenosis.

average of 4.53 months before diagnosis. There were ten Chinese, four Malay, two Indian and two Sri Lankan patients. Ten (55.6%) patients were born premature, with mean and median gestational ages of 27.3 weeks and 26 weeks, respectively (range 24–36 weeks).

Two patients developed symptoms prior to any previous intubation and were diagnosed as having a definite congenital stenosis. 16 patients were intubated prior to the presentation of symptoms and were classified under acquired SGS. The majority of patients, (10/18, 55.6%) had Grade I SGS, followed by five with Grade II (27.8%) and three with Grade III (16.7%) SGS. None of the patients had grade IV SGS (Fig. 1). Of the group of patients with acquired SGS, the majority (11/16, 68.8%) presented with symptoms of stridor. Other clinical manifestations included difficulty in breathing and severe episodes of apnoea. Both patients with congenital SGS also presented with stridor.

A total of 17 patients were intubated. The aetiology of acquired SGS in all 16 cases was related to intubation. Six (37.5%) patients required ventilatory support for respiratory conditions, including chronic lung disease, hyaline membrane disease, bronchopneumonia and apnoea. Five (31.3%) were ventilated for prematurity, three (18.8%) for cardiac conditions and two (12.5%) due to road traffic accidents (Fig. 2). The mean cumulative ventilatory time via endotracheal tube in all patients with acquired SGS was 3.14 (range 0.5–12) months. A total of seven (38.9%) patients required tracheostomy for relief of their airway obstruction, six of whom had acquired SGS and one had congenital SGS.

The diagnostic procedure of choice for all cases of SGS was microlaryngoscopy and bronchoscopy (MLB). The types of surgical interventions performed for all grades are listed in Table I. The most common surgical intervention performed was MLB with bougie dilation, with a total of 24 interventions performed on the 18 patients. Cricoid split was performed for two patients with Grade I stenosis and one patient with Grade II stenosis. Laryngotracheal reconstruction (LTR) was performed for one patient with Grade II stenosis and two patients with Grade III stenosis. One patient with Grade III stenosis underwent cricotracheal resection (CTR).

The mean and median duration of treatment was 4.63 and 4 years, respectively (range two days to 12 years), with an overall recovery rate of 66.7% (12 out of 18 patients). Two (11.1%) patients were still undergoing treatment when this study was being written and two (11.1%) were lost to follow-up. Two (11.1%) mortalities were reported due to reasons unrelated to SGS; one patient died of bronchopneumonia, while the other died of aspiration pneumonia. Four of the seven (57.1%) patients who underwent tracheostomy were successfully decannulated. Of the three patients who did not undergo decannulation, one would be due for decannulation in the near future, one was deemed unsafe for decannulation, and one died.

DISCUSSION

In the early 20th century, SGS was very rare. Figures from the literature showed a considerably varied prevalence of 1%–8%.[2-5] Since the 1960s, studies have reported an increase in the incidence of acquired SGS, with the age at presentation shifting toward young infants. This can be attributed to the advent of endotracheal intubation in neonatal medicine and the need for long-term ventilation in premature babies. Subsequent progress in ventilatory techniques and the widespread use of artificial surfactants have led to a decrease in the duration of intubation, and even its complete avoidance, thereby reducing the incidence of acquired stenosis. However, medical advances have had little impact on the incidence of congenital stenosis.[5-8]

In our study, a predominance (88.9%) of acquired stenosis was observed. There has been a general decrease in the number of cases of acquired SGS, from five in 1997 to a mean of 0.82 cases each year from January 1998 to

Table I. Total number of surgical interventions performed for each grade of stenosis in order of frequency.

Grade	No. of interventions	Type of intervention
I	5	MLB with bougie dilation
	4	CO_2 laser
	4	Tracheostomy
	2	Arytenoidectomy
	2	Cricoid split
II	12	MLB with bougie dilation
	6	CO_2 laser
	6	Mitomycin application
	5	Tracheostomy
	1	Diathermy resection of granuloma
	1	Cricoid split
	1	Laryngotracheal reconstruction with anterior graft
III	7	MLB with bougie dilation
	4	Tracheostomy
	3	Mitomycin application
	1	Cricotracheal resection
	1	Laryngotracheal reconstruction with anterior graft
	1	Laryngotracheal reconstruction with anterior and posterior grafts and stenting

MLB: microlaryngoscopy and bronchoscopy

December 2008, with an uncharacteristic spike in 2005. In contrast, no apparent trend has been observed in the number of cases of congenital SGS (Fig. 3). Acquired SGS is most commonly caused by direct trauma to the larynx during long-term intubation in the neonatal period, and is often associated with repeat and emergency situations.[8,9] This is reflected in our series by the aetiology of acquired stenosis in all 16 cases being related to intubation.

In all cases, the diameter of stenosis was estimated using endotracheal tubes during MLB, and the severity was objectively scaled using the Myer-Cotton Grading System, based on endotracheal tube sizing. The classification was Grade I for up to 50% lumen obstruction, Grade II from 51% to 70%, Grade III from 71% to 99% and Grade IV for complete obstruction.[10] Subsequent MLB enabled the clarification of previous findings and serial assessment after endoscopic procedures such as dilation by gentle bouginage, laser ablation and mitomycin application. Rational therapy is based on the accurate determination of the pathology and the precise location of the stenosis. An open reconstructive procedure was considered only in cases that showed no improvement with conservative endoscopic management.

The decision regarding the need for framework expansion was related to the grade of stenosis. Most patients with Grade I or Grade II stenosis were managed conservatively, with three out of ten (30%) and one out of five (20%), respectively requiring some form of reconstructive procedure. In Grade III stenosis, the proportion increased to three out of three (100%). The mean number of reconstructive procedures performed per patient increased with the increasing severity of stenosis (Fig. 4). Techniques for the treatment of SGS without tracheotomy include MLB with bougie dilation, CO_2 laser and anterior cricoid split. The more severe cases required LTR, CTR or long-term tracheostomy. The common complications of surgical intervention include failure of decannulation after LTR, granulation post grafting and dysphonia. CO_2 laser is mostly used for ablation of granulation tissue and laryngeal or subglottic webs.

Numerous studies have shown that mitomycin inhibits fibroblast and scar formation *in vitro* as well as *in vivo*.[11,12] In our study, mitomycin application was indicated in only two cases of Grade II and Grade III stenosis each. It was used in conjunction with bougie dilation, CO_2 laser and lysis of laryngeal web. There was significant reduction of granulation tissue or improvement in airway patency after mitomycin application in all nine instances.

In our study, cricoid split was indicated in two patients with Grade I stenosis. In one patient, there was a failed extubation attempt post operation with oxygen desaturation and CO_2 retention. However, after four days of administering dexamethasone, the patient was successfully extubated. The other patient had a failed decannulation attempt due to stridor, respiratory distress and possibly, bilateral vocal cord paralysis. A patient with Grade II stenosis also underwent a cricoid split after four failed attempts at extubation, even under dexamethasone cover. After three more failed extubation attempts, a decision was made to perform a tracheostomy in view of the glottic changes from intubation.

In another patient with Grade II stenosis, LTR with anterior costal cartilage graft was performed. Although there was empirical coverage with Augmentin post operation, the endotracheal tube showed a growth of *Pseudomonas aeruginosa*. Subsequent upper respiratory tract infection due to respiratory syncytial virus was observed. These two factors may explain the failure to decannulate in this patient. When this study was being written, the patient was on a tracheostomy speaking valve with no breathing difficulty, and was due for decannulation in the near future. A successful LTR with anterior costal cartilage graft was performed in a patient with Grade III stenosis. Post operation, the endotracheal tube showed growth of *Enterobacter aerogenes*. Airway patency improved from 50% pre-operatively to 80% after the procedure. However, the patient presented with stridor

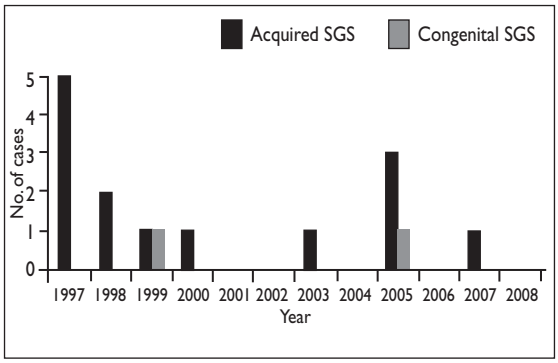

Fig. 3 Prevalence of congenital and acquired subglottic stenosis.

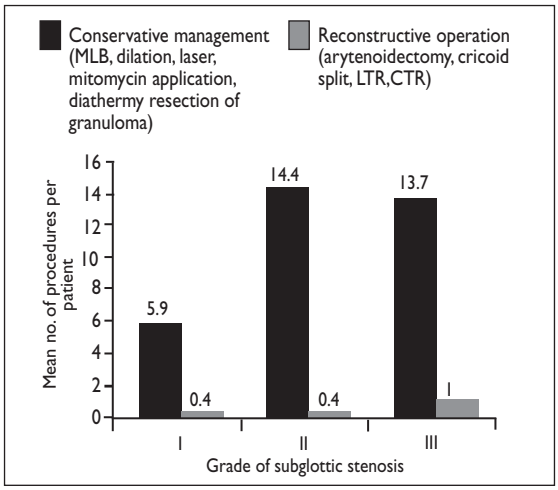

Fig. 4 Management of paediatric subglottic stenosis.

two months later due to a granuloma at the graft site, causing almost complete obstruction. Following excision of this healing granuloma, the patient made a complete recovery with 80%–90% airway patency and good voice production.

Untreated gastro-oesophageal reflux (GER) has been considered a risk factor for failure of LTR.[13-15] Walner et al recommended treating GER for one month before attempting reconstructive surgery.[14] A patient with Grade III stenosis had gross oedema of the supraglottis and glottis, with cobblestone appearance of the posterior pharynx after LTR with anterior and posterior grafts and stenting, indicating severe GER despite more than two months of medication. The oedema resolved completely only after a laparoscopic fundoplication, leaving 90% subglottic airway patency with epithelialisation of grafts. The patient eventually had a successful decannulation.

Before the 1990s, there was a general reluctance to use the technique of CTR in the paediatric population, stemming from concerns about the risk of growth abnormalities involving the laryngotracheal axis after this procedure, as well as due to the higher technical demand compared to standard LTR. However, there is emerging evidence showing the absence of growth abnormalities of the laryngotracheal axis post CTR. It is particularly suited in the management of severe Grade III and IV stenoses, as well as in salvaging laryngotracheoplasty failures.[6,7,16-19] Many authors have concurred that CTR is acceptable, and even preferred, for high-grade SGS, with the largest reported series of paediatric CTR[20] achieving long-term decannulation rates of more than 90%. In this study, only one CTR was indicated in a case of Grade III stenosis. Pre-operatively, normal vocal cord movements were observed in the patient. No significant complications were reported post operation, and after four MLBs and three serial bougie dilations, the patient was successfully decannulated. Interestingly, Grade I stenosis generally did not equate to a shorter duration of treatment than that required for the more severe stenoses. For Grade I stenosis, the mean and median duration of treatment was 5.97 years and five years, respectively (range six months to 12 years), for Grade II, it was 2.67 and 2.17 years, respectively (range two days to eight years), and for Grade III, the duration was 3.44 years and one year, respectively (range two months to nine years).

A retrospective medical record review has its limitations, but with the rarity and decreasing prevalence of SGS, it may be difficult to design an accurately powered prospective study. Nonetheless, our study does provide some insight into the current management techniques and outcome of SGS at KKH.

In conclusion, experience in airway endoscopy is crucial in the diagnosis and management of paediatric SGS. Conservative management alone may be sufficient in the majority of Grade I stenoses. More severe cases might warrant reconstructive surgery. LTR remains the intervention of choice, with CTR being a complement where indicated, such as in failure following multiple LTRs. However, we documented variable success for each type of reconstructive procedure in our study. The mean number of reconstructive procedures performed per patient increased with the increasing severity of the stenosis. With the decreasing incidence of acquired SGS due to improved medical care, major open surgical cases and experiences are now rarely encountered. Surgical intervention should not be rushed into until after all possibilities have been carefully considered. Each laryngeal framework procedure has to be customised to suit the individual, so that chances of resolution can be maximised without being detrimental to the patient.

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