Introduction

Subglottic stenosis may be congenital (primary) or acquired (secondary). Congenital stenosis may be caused by either a small cricoid cartilage or a thick submucosa due to trauma associated with other laryngeal anomalies. Acquired subglottic stenosis (ASGS) is caused by soft-tissue stenosis due to trauma following prolonged endotracheal intubation. Congenital subglottic stenosis (CSGS) is a well-known cause of stridor in infancy. It usually occurs sporadically and familial occurrence is rare.

Materials and methods

Main body text fit closely to the printing area. Three children who presented with subglottic stenosis born to consanguineous parents were identified. For the literature research, reports published from 1950 to June 2011 were retrieved. We also screened bibliographies of the collected articles to identify pertinent reports. They consisted mainly of single case reports and case series. A summary of evidence from the literature is discussed.

Results

Three Asian children were born with congenital subglottic stenosis for closely related consanguineous parents. Their presentation, family tree, genetic tests, associated features and treatment are discussed.

Case reports

Case 1
A boy (FH, 3 years) was born at 40 weeks preterm by normal birth and his birth weight was 3.2 kg. At 2 months he presented with noisy breathing while feeding and was admitted at 3 months of age due to failure to thrive. At the age of 3 months he was diagnosed with grade II subglottic stenosis. He had noisy breathing while feeding and was admitted at 3 months of age due to failure to thrive. The family history included failure to thrive and rickets. He did not have any dysmorphic features and developmentally was normal. He had normal dietary assessment with appropriate intake but with poor weight gain. He continued to be small for age and failure to thrive did not improve. At the age of 35 months he was diagnosed with complete growth hormone deficiency and been prescribed growth hormone injections. He was managed conservatively until four years of age when he was managed with repeated balloon dilation. He developed harsh inspiratory noise and mild recession without tracheal tug. Of relevance his mother has a history of renal impairment and hypertension due to dysplastic kidneys. He also suffered with poor weight gain. He was of proportionate small stature and his growth was along the 0.4th centile. He was diagnosed with grade III subglottic stenosis. He was managed with repeat balloon dilation. He had laryngotracheal reconstruction with anterior and posterior rib graft at 33 months. He is currently symptom free without treatment.

Case 2
A boy (FH, 3 years) was born at 38 weeks preterm by normal birth and his birth weight was 2.58 kg. Antenatal and postnatal history was unremarkable and no history of previous intubation or special care unit admission. He had emergency tracheostomy for failed intubation during congenital inguinal hernia repair and at the age of 5 months subsequently he had bilateral inguinal hernia repair and religious circumcision. He suffered with renal impairment and hypertension due to dysplastic kidneys. He also suffered with poor weight gain. He was of proportionate small stature and his growth was along the 0.4th centile. He was diagnosed with grade III subglottic stenosis. He had managed with repeated balloon dilation. He had laryngotracheal reconstruction with anterior and posterior rib graft at 33 months. He is currently symptom free without tracheostomy.

Case 3
A boy (ZH, 3 years) was born at 38 weeks preterm by normal birth and his birth weight was 2.3 kg. He had tracheostomy for poor fetal growth and maternal diabetes. From birth he had noisy breathing while feeding and was admitted at 3 months of age due to failure to thrive. He had laryngotracheal reconstruction with anterior and posterior grafted cartilage. He was symptom free until the age of 2 when the anesthetist noticed subglottic stenosis during intubation for religious circumcision. The procedure was abandoned and postoperatively he developed harsh inspiratory stridor and was referred to an otolaryngologist when a diagnosis of grade II subglottic stenosis was made. Intermittently he was having harsh inspiratory noise and mild recession without tracheal tug. Of relevance his medical history included failure to thrive and rickets. He did not have any dysmorphic features and developmentally was normal. He had normal dietary assessment with inappropriate intake but with poor weight gain. He continued to be small for age and failure to thrive did not improve. At the age of 35 months he was diagnosed with complete growth hormone deficiency and been prescribed growth hormone injections. He was managed conservatively until four years of age when he developed noisy breathing during upper respiratory infections. He had adenotonsillectomy at age of 5 years for sleep disordered breathing. At the age of 6 years, he was breathless on exertion and still had failure to thrive. Single stage anterior graft laryngotraheal reconstruction was carried out at the age of 7 years and 4 months. Postoperatively he developed granulation tissue around the graft. This was managed conservatively. The shortness of breath on exertion abated following surgery and he was discharged one year after surgery.

Conclusions

Congenital familial subglottic stenosis in unrelated parents has been reported previously but not in consanguineous parents. The genetic work up of these families is still incomplete although the family history suggests a genetic aetiology. Further genetic research is needed to assess the mode of possible inheritance in these cases.

Acknowledgement

Mr Bull – Involved in the management of case 1

References