



Congenital subglottic stenosis: An occasional concern in severe croup

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Abstract

We reported a 54-day old infant suffering from congenital subglottic stenosis presenting as croup, subsequently requiring endotracheal intubation, tracheostomy and successfully managed by bronchoscopic balloon dilatation.

Keywords: Bronchoscopic balloon dilatation, Croup, Subglottic stenosis, Tracheostomy

Introduction

Croup is a common cause of stridor in the young child, especially between 6 and 36 months of life. It is estimated that by age 2 years, 5% of all children will have had croup. Population-based data indicate that 1-5% of children with croup are admitted to hospital. Most responded well with one or two doses of corticosteroid and could be discharged shortly. Need for intubation is infrequent, estimated to be less than 3% of those admitted, and mortality appears to be rare, with no more than 1 in 30,000 cases from mixed reports data.¹⁻⁴ Occasionally, there are alternative diagnoses that mimic croup or unmasked during an episode of croup.

Case report

A 54-day old boy was admitted from Accident & Emergency Department (A&E), Alice Ho Nethersole Hospital for stridor for 3-4 days. He was born full term by normal vaginal delivery, with birth weight 2.965 kg. Antenatal and birth history was uneventful with no neonatal resuscitation, intubation and mechanical ventilation needs. He had mild neonatal jaundice which responded to phototherapy and was subsequently followed up in Maternity & Child Health Clinic (MCHC) for infant routine care and immunisation. He was feeding and thriving well but noted to have mild noisy breathing since day 40 of life. There was no cyanosis or choking episode and he remained well until recent 3-4 days when he developed hoarse voice with increasing noisy

breathing, despite no fever or other coryzal symptoms. There was no sick contact in the family and travel history was negative. He was seen by private doctor, with symptomatic medications given for upper respiratory tract infection. However, his condition got worse, with decreased feeding and reduced urine output. He was taken to A&E and then admitted to our department for croup.

On admission, he appeared well nourished (body weight 4.605 kg), was afebrile and there was no skin rash or haemangioma. However, he was tachypnoeic (breath rate 52/min) and tachycardic (pulse rate 162/min), with mild oxygen desaturation (SpO₂ 90% in room air) which was improved by 1 L/min nasal cannula oxygen to SpO₂ 98%. Mild respiratory distress was noted on crying, with hoarse voice, inspiratory stridor and insucking chest. Cardiovascular, neurological and abdominal examinations were unremarkable.

He was kept nil by mouth, given intravenous fluid and one dose of 3 mg intramuscular dexamethasone. Chest X-ray showed a rotated film with non-specific bilateral lung haziness and intravenous amoxicillin-clavulanic acid was empirically started for possible pneumonia. Blood tests subsequently showed normal complete blood counts, glucose, electrolytes and C-reactive protein. Stridor and respiratory distress persisted and nebulised adrenaline (1:1000) 2.5 ml was added, which resulted in mild transient improvement. Flexible fiberoptic examination showed normal choanae and hypopharynx, mobile vocal cords but did not visualise well the subglottic area. Urgent computerised tomography study of neck and thorax revealed subglottic focal tracheal stenosis of 2 mm width (Figure 1) and bilateral lower lobe apical and postero-basal consolidations.

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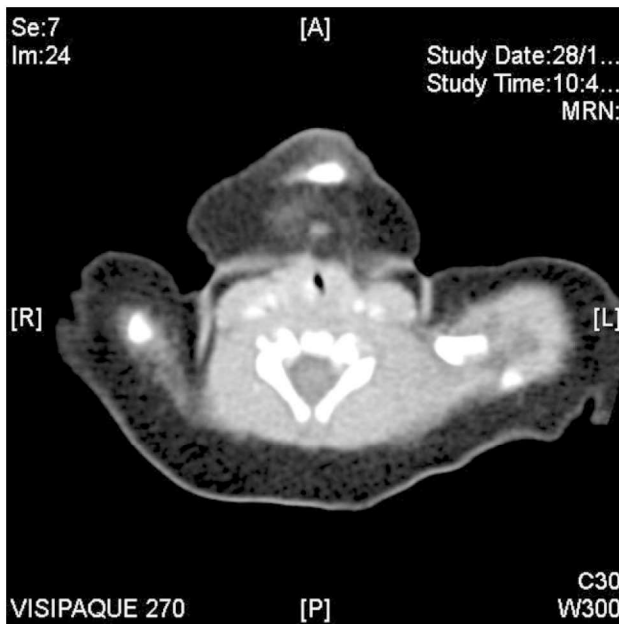


Figure 1. Computerised tomography study of neck.

He was transferred to a Paediatric Intensive Care Unit (PICU) for stabilisation. Emergency tracheostomy was performed during which the ENT surgeon revealed subglottic stenosis with transverse diameter smaller than 2.5 mm, which failed to pass the examining telescope further. There was a cricoid deformity with a small anterior notch. Subsequent elective laryngotracheobronchoscopic examination confirmed Cotton Myer's grade 3 subglottic stenosis with an elliptical slit-like opening passing only size 2.7 mm endoscope and the stenotic segment measured 11 mm. He was subsequently treated by balloon laryngoplasty with dilatation and adjuvant injections of steroid and mitomycin every 2 to 3 months. With 5 sessions of therapy, his airway size improved to grade 1, easily allowing passage of 3.5 mm bronchoscope (outer-diameter 5.7 mm), and was successfully decannulated at age 1 year after clearing up some granulation tissue by laser therapy. He remained well with no stridor at rest although occasional mild exacerbation was noted during respiratory infections. Feeding was normal and he was growing well and started kindergarten although assessed to have a slight 3-month speech delay.

Discussion

Our case illustrated that there are occasional patients we need to worry in children presenting with croup. In

those not responding to standard therapy¹⁻⁵ or appear extremely sick, clinicians should consider an alternative cause of airway obstruction (Tables 1 & 2).

Table 1. What / When to worry in children with croup

- Pre-existing upper airway narrowing e.g. subglottic stenosis
- History of endotracheal intubation, mechanical ventilation
- Previous admissions with severe croup
- Age below 6 months, especially younger than 3 months
- No improvement following corticosteroid treatment
- More than 2 doses of nebulised adrenaline required
- Require care exceeding level of comfort of local hospital

Table 2. Differential diagnosis of stridor

Common

- Croup

Less common

- Bacterial tracheitis
- Epiglottitis

Rare

- Upper airway abscess: peritonsillar, retropharyngeal
- Foreign body aspiration
- Allergic reaction, angioneurotic oedema
- Laryngeal diphtheria
- Congenital airway malformations
 - Laryngomalacia
 - Vocal cord paralysis
 - Subglottic stenosis
 - Laryngeal web, cleft, atresia
 - Tracheo / bronchomalacia
 - Subglottic haemangioma
- Tumour

Subglottic stenosis (SGS) is the third most common congenital laryngotracheal anomaly (after laryngomalacia and vocal cord paralysis) but the most common laryngotracheal anomaly requiring tracheostomy in infants.

For diagnosis, SGS is defined as a subglottic airway diameter of 4 mm or less in a full-term neonate or a diameter of less than 3.5 mm in a premature neonate.⁶ It can be acquired or congenital. Majority of cases of SGS are acquired and it is considered congenital only if there is no other apparent cause of stenosis e.g. intubation, trauma. If a congenitally misshapen larynx is intubated, inflammation and scarring may occur even though an age-appropriate size endotracheal tube is used. Therefore the true incidence of congenital SGS is difficult to determine since this cannot be clearly

distinguished congenital or acquired once a child has been intubated. Suspected incidence of congenital SGS is about 5% of all SGS cases.⁶⁻⁸ Histologically, SGS can result from cartilaginous defects of the cricoid or tracheal ring (usually in congenital SGS) (Figure 2), or soft tissue abnormalities (usually in acquired SGS) (Table 3).

Table 3. Histopathologic classification of subglottic stenosis

1. Cartilaginous stenosis (usually congenital)
 - a. Cricoid cartilage deformity
 - i. Normal shape, small size
 - ii. Abnormal shape
 - Elliptic
 - Cleft (partial, submucosal, incomplete)
 - Flattened
 - Other (including acquired lesions)
 - Fragmented, distorted cricoid
 - Cricoid ossification
 - Thickened cricoid
 - b. Trapped first tracheal ring
2. Soft tissue stenosis (usually acquired)
 - a. Submucosal mucous gland hyperplasia
 - b. Ductal cysts
 - c. Submucosal fibrosis (fibrous connective tissue)
 - d. Granulation tissue

Data from Holinger PH, Kutnick SL, Schild JA, Holinger LD. Subglottic stenosis in infants and children. *Ann Otol Rhinol Laryngol* 1976;85(5 Pt.1):591-9.

In addition to X-ray neck and chest often performed already in the initial acute situation, detailed diagnostic evaluation often requires computerised tomography (CT) scan or magnetic resonance imaging (MRI).⁹ Although MRI carries no radiation hazard, CT scan is often more readily available in the emergency setting. Modern CT skills enable fast scanning speed to acquire good quality images despite fast breath rates in the infant and virtual bronchoscopy could be performed with 3-dimensional reconstruction techniques to assess the length of the stenotic segment, facilitating further invasive investigations or operative procedures. Both, however, require sedation, which might be contraindicated in a very sick child with a compromised airway. When vascular compression is also suspected, barium oesophagram and echocardiogram may be required.¹⁰⁻¹¹

The gold standard of diagnosis is direct laryngoscopy and bronchoscopy under general anaesthesia. The outer diameter of the largest bronchoscope that can pass through the stenosis and the length as well as location of the stenotic segment should be noted. It also allows dynamic assessment of the cartilaginous support and presence of any tracheomalacia. The severity of obstruction is evaluated generally by the Cotton-Myer grading, although there is yet any ideal grading system and it does not prognosticate decannulation (Figure 3).

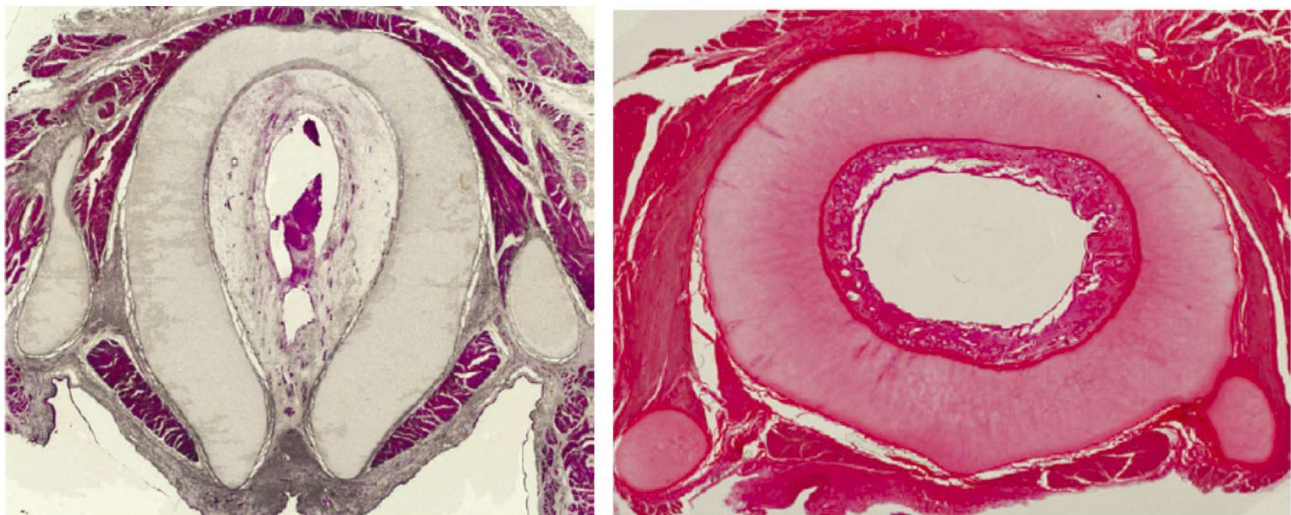


Figure 2. Misshapen cricoid cartilages: elliptical (left), flattened (right).



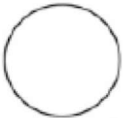





Classification	From	To
Grade I	 No Obstruction	 50% Obstruction
Grade II	 51% Obstruction	 70% Obstruction
Grade III	 71% Obstruction	 99% Obstruction
Grade IV	No Detectable Lumen	

Figure 3. The Cotton-Myer grading of subglottic stenosis.

Definitive management is airway surgery and classically external approaches have been the classical techniques of repair. With surgical and anaesthetic advances, endoscopic approaches have evolved (Table 4). The length of stenotic segment, any associated abnormalities and patient condition dictate the surgical treatment option which has to be individualised.¹²⁻¹⁸

Table 4. Surgical treatment of subglottic stenosis

- Endoscopic approaches
 - Dilatation with rigid dilator
 - Balloon laryngoplasty
 - Laser resection – KTP or CO₂ laser
 - Stent placement
- External approaches
 - Cricoid split
 - Cricotracheal resection
 - Laryngotracheal reconstruction (LTR)
 - Slide tracheoplasty

Given the rarity of congenital SGS, there is still limited literature available on use of balloon laryngoplasty as primary treatment for it.^{13,17-20} In a study of its use in infants with acquired Cotton grade 2 or 3 subglottic stenosis 70% success rate was reported.¹³ A long-term follow up study in adults reports it as a safe temporary measure providing immediate symptomatic relief for tracheal and bronchial stenosis but long-term definitive or additive treatment with laser or stent is required.²¹ A recent study in paediatric laryngotracheal stenosis¹⁸ concludes that dilatation with adjuvant topical therapy is successful as primary therapy in the majority of low-grade obstruction but it could not be determined whether balloon or rigid dilatation was superior to another.

Conclusion

An infant with congenital subglottic stenosis was presented, illustrating his acute presentation, clinical course and outcome. While the baby with mild stenosis and minimal symptoms may need only monitoring airway growth and expectant management, the severely sick infant requires prompt recognition of the problem, timely resuscitation and investigations. Availability of modern PICU and nursing support,²² integrated multi-disciplinary team-based care, together with surgical advances have greatly improved the survival and quality of life of children with congenital airway malformations.²³ Endoscopic approaches in appropriately selected cases is a promising surgical treatment modality which may obviate the need of open laryngeal surgery by 70-80%.²⁴

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