

# Central Airway Obstruction: Diagnostic Challenges and Management Strategies in Subglottic Stenosis and Poly Chondrites with Case Studies

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## ABSTRACT

**Tumours or strictures compressing the trachea /bronchial tree cause central airway obstruction (CAO). Subglottic stenosis are a cause of respiratory symptoms and distress. Idiopathic sub glottic stenosis (ISGS) is a rare disease occurring mainly in women and has a history of recurrences. Endoscopic laser microsurgery and balloon dilatation are used in the management of strictures. Recurring strictures may require tracheoplasty. Poly chondrites is a rare disorder causing poly arthritis and inflammation of cartilaginous tissue. Inflammatory changes in the lung cause tracheobronchomalacia (TBM) and lung collapse. Relapsing polychondritis (RP) effecting the airway is life threatening as there is dynamic airway closure during expiration. The medical therapy in RP includes steroids, disease modifying agents (methotrexate) and biologics. Surgical interventions include tracheostomies and tracheobronchial stents for severe forms of the illness.**

**Keywords:** Tracheal stenosis, endoscopic laser microsurgery, balloon dilatation, relapsing polychondritis, biologics, stents.

## INTRODUCTION

Central airway obstruction (CAO) is caused by strictures and compression from benign and non-benign tumours. The majority of strictures are due to post intubation trauma or post tracheostomy stenosis. Thyroid enlargement or malignancy related causes include locally advanced lung cancers, secondary metastasis from a breast and thyroid cancer (1). These cause tracheomalacia and airway obstruction. The symptoms of CAO are shortness of breath, hoarseness, cough and wheezing. They mimic those of asthma and chronic obstructive pulmonary disease (COPD) resulting in delayed diagnosis. RP (relapsing polychondritis) is a rare form of an immune mediated illness causing polyarthritis and inflammation of cartilaginous tissue (2). RP affecting the airways is debilitating and life threatening. This paper discusses two case studies of CAO, the diagnostic challenges and management strategies as they relate to sub glottic stenosis and relapsing polychondritis (RP).

## CASE STUDIES

### Case 1

A female age 65 presented with respiratory difficulties. She was dyspnoeic with poor effort tolerance, stridor and a choking feeling. She had a history of gastrointestinal reflux disease (GERD) and was on anti-reflux medication. Cardio- pulmonary examination was non-

contributory. She had no history of any previous anaesthesia or long-term intubation. Treatment was symptomatic and included bronchodilators and steroids with little improvement.

Otolaryngologist opinion was sought and a flexible bronchoscopy under topical anaesthesia revealed a slight narrowing below the chords (subglottically).

The patient required laryngoscopy and bronchoscopy under general anaesthesia.

This procedure requires unimpeded surgical access to the airway and a shared airway with the anaesthesiologist. TIVA (total intravenous anaesthesia) and (HPSV) high pressure source jet ventilation was used to maintain oxygenation. Standard monitoring was done and included a cerebral function monitor (Connex) to titrate the depth of anaesthesia and analgesia.

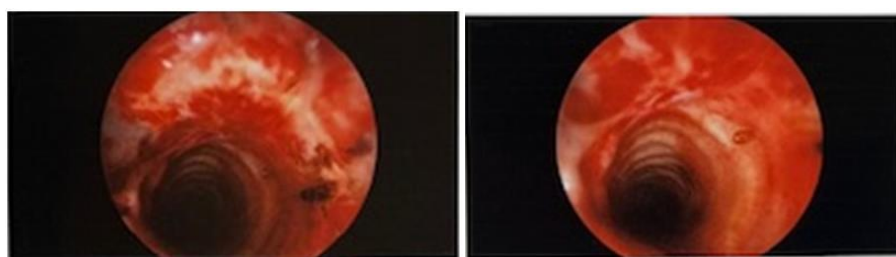
Laryngoscopy revealed significant narrowing subglottically. Endoscopic laser microsurgery and balloon dilatation of the stricture was successfully done; the procedure being uneventful and uncomplicated.

Post operative management in the ward included adrenaline nebulisation and a short course of steroids. Improvements in patient symptoms were observed in the ensuing period. Her breathing became easier with no choking feeling, her effort tolerance improved as did her peak flow (PF) measurements. The patient was discharged and asked to return for regular following up or should she suffer breathing difficulties. Reviewed several months later, a repeat procedure was required at seven months. Recurrences and restenosis are not uncommon and regular following up required should patients become symptomatic.

A diagnosis of idiopathic tracheal stenosis was made having excluded other likely causes such as collagen vascular disease, relapsing polychondritis and tracheomalacia (5) Gelbard et al.



**Pre-dilatation**



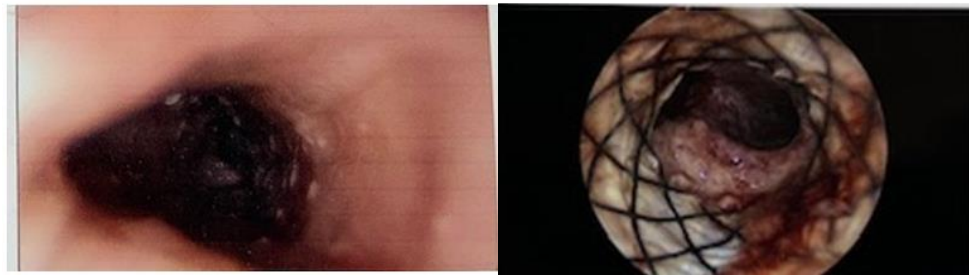
**Post-dilatation**

## Case 2

A 39-year-old female with a history of polychondritis was referred from (Zambia) a neighbouring sub-Saharan country. She presented with a tracheostomy in situ following failed attempts at extubation. Diagnosed at a young age (20 years) with Polychondritis when she presented with auricular and nasal inflammation. Treatments over the years included anti-inflammatory (NSAIDS), prednisolone, intermittent methotrexate and dapsone. Her past illness had resulted in a flattened nasal bridge deformity. Owing to respiratory difficulties that were life threatening a tracheostomy was deemed necessary. On examination cardio/ pulmonary function testing was non-contributory and could not account for her failed extubation. These included CXR, blood gasses, and echocardiogram.

Blood indices showed a mildly raised CRP of 11 mg/L (normal < 5) and PCT 0.11 ng/ml (normal < .05). The immunology screen was negative. The otolaryngologist recommended an examination under anaesthesia (EUA) of the airway.

Laryngoscopy and bronchoscopy were in keeping with tracheomalacia. There was dynamic airway collapse during expiration. The area below the tracheal stoma but proximal to the carina were more compromised and flaccid, with both main bronchi spared. In view of her past history, a diagnosis of relapsing polychondritis (RP) with tracheomalacia was confirmed.



**TM and airway collapse during expiration Tracheal stent in situ.**

To prevent a collapse of the airway lumen a tracheal stent was inserted endoscopically. Stents improve the dyspnoea and lung function, but have inherent risks (1)(3). They can migrate, cause infection and mucus plug formation. They also cause a proliferation of granulation tissue and airway obstruction. Medical therapy was optimised to include (NSAIDS) for pain, corticosteroids and methotrexate to control the inflammatory process, as well as its steroid sparing effects.

The patient had a fenestrated tracheostomy tube inserted enabling her to speak and communicate. Importantly she was advised on proper stomal hygiene; the use of mucolytics and expectorants to clear secretions and preventing stent blockage. The patients' respiratory symptoms improved. She was comfortable at rest, mobile and no longer in respiratory distress. Her need for the tracheostomy will be reviewed on her following visits with the aim of weaning and extubation.

## **DISCUSSION**

Tracheomalacia (TM) is a common tracheal defect seen in infants and toddlers and is due to immature cartilage formation around the trachea. Common presenting symptoms are cough, stridor and respiratory distress (4). Treatment includes respiratory support, instrumental

intervention or a tracheostomy. The vast majority of patients with tracheostomies are successfully weaned and grow into adulthood with little or no respiratory symptoms. A laryngeal stenosis (LS) in children is also a congenital anomaly (4).

A subglottic stenosis (SGS) can be (a) acquired from Iatrogenic injury, long term endotracheal intubation (70%), caustic injury, infection or tumour. (b) autoimmune conditions associated with collagen vascular disease (15 %) Wegeners granulomatosis. (c) idiopathic sub glottic stenosis (iSGS) with no known cause (15 %). This condition commonly occurs in women (98%) who are adults above 40 years and white. It is a recurring fibro inflammatory disease requiring repeat procedures (5).

Idiopathic (iSGS) is a rare condition of unknown aetiology. Gastrointestinal reflux disease (GERD) may be a risk factor. Recommendations include gastroscopy or pharyngeal PH testing to exclude reflux disease (6). To reduce the likelihood of recurrence aggressive medical management should include inhaled corticosteroids and anti-reflux measures (7).

Serial Intralesional Steroid Injection (SILSI) with triamcinolone acetate helps reduce airway restenosis (8). They dampen inflammation and inhibits scar formation. At initial balloon dilatation, it is injected into the sub glottis with repeat injections required as per protocol (8). The availability of a trachealator a novel a non-occlusive balloon dilator is advantageous over intermittent dilatation done over short apnoeic periods. This treatment strategy allows continuous ventilation and dilatation lasting up to 10 minutes, the disadvantage however is the risk of epithelial damage to the airways (9).

Patients requiring frequent procedures (12-15) yearly can benefit from a single stage laryngotracheal resection and reconstruction procedure that may be a preferred option (10).

Relapsing Polychondritis (RP) is a rare multi organ disease characterised by inflammation in the cartilaginous tissue of the body. Auricular and nasal chondrites and/or polyarthritis are the commonest clinical presentation. The painful inflammatory condition may subside spontaneously but cause deformities in affected structures. RP is immune mediated and associated with other autoimmune diseases. Rheumatoid arthritis (RA) occurs in 30% of patients (2)

Airway involvement in RP is incapacitating and life threatening. Cardiovascular complications are the second most frequent cause of mortality. Arthropathy frequently occurs and mild ocular disease is present in half of all RP patients (2).

RP is diagnosed clinically and diagnostic criteria developed share common features. Additional modification has been described. Mc Adam et al. (1976); Damiani and Levine (1979); Michelle et al. (1986) (2)(11).

### **Airway Involvement in RP**

Mild and non-severe forms of the disease will require control of pain and inflammation. Treatment will include NSAID, Dapsone and Colchicine (2)(12).

RP can cause airway inflammation and severe tracheobronchomalacia (TBM) where dynamic airway collapse occurs during expiration (12). Common symptoms of RP are wheezing, stridor, shortness of breath, chronic cough and recurrent respiratory infections (4). Sub glottic and trachea- broncho stenosis are also known complications (12). Therapy includes systemic steroids, disease modifying agents (Methotrexate) and the early use of biologics (infiximab). Surgical interventional procedures include tracheostomy for sub glottic stenosis and the insertion of tracheobronchial stents (12).

RP effecting the airway can be confirmed during chest CT or bronchoscopy where collapse of the airway occurs due to weakness of the cartilage. This is due to increased intra -thoracic pressure occurring during expiration. Raised inflammatory markers include ESR, CRP and Procalcitonin but no diagnostic blood markers are known (12).

Therapeutically invasive (IPPV) and non-invasive (NIPPV) positive pressure ventilation is required during acute episodes of RP. Airway stenting prevents airway collapse should other treatment modalities fail. The early use of biologics in RP with infiximab and tocilizumab prevents severe airway deterioration and reduce mortality (12). Stenting with self-expanding metal or silicone stents improve lung functions and quality of life but are not without complications.

## CONCLUSION

Central airway occlusion from tracheal stenosis is common following post intubation and post tracheostomy events. Idiopathic tracheal stenosis occur in adults, is rare and presents with respiratory symptoms that mimic asthma and COPD. EAU and bronchoscopy are helpful diagnostic aids and treatment requires endoscopic laser microsurgery and balloon dilatation.

RP is a rare autoimmune condition affecting cartilaginous structures in the airway that can be life threatening. It causes TBM and laryngeal stenosis hugely impacting on health-related quality of life (HRQoL).

There are presently no established treatment guidelines. Recommendations are early use of biologics, NSAID, steroids and methotrexate. Ventilation as needed and tracheostomy may be required. Stenting is a last resort for unstable respiratory conditions (12)(2).

Patients with tracheal stents requiring surgery need careful airway evaluation. Second generation supraglottic devices are recommended and should intubation be required fibre optic visualisation will ensure correct placement.

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