IMAGES in PAEDIATRIC CARDIOLOGY

Management of congenital tracheal stenosis: a multidisciplinary approach

A Dodge-Khatami,* VT Tsang,** DJ Roebuck,*** and MJ Elliott**

*Senior Surgical Registrar, Cardiothoracic Unit, Great Ormond Street, London
**Consultant Paediatric Cardiothoracic Surgeon, Cardiothoracic Unit, Great Ormond Street, London
***Consultant Interventional Cardiologist, Cardiothoracic Unit, Great Ormond Street, London

Contact information: Ali Dodge-Khatami, Senior Surgical Registrar, Cardiothoracic Unit, Great Ormond Street Hospital NHS Trust, Great Ormond Street, London WC1N 3JH, UK ; Email: adodgekhatami@yahoo.com

Abstract

Congenital tracheal stenosis is a rare but underdiagnosed anomaly which can present as life-threatening respiratory insufficiency in neonates and infants. Initial control of the airway is mandatory. Surgical correction is the mainstay of therapy and is achieved with low mortality. The type and extent of repair depends largely on the length of stenosis. Cardiac anomalies are frequently associated and may be addressed at the time of tracheal surgery. Despite initial satisfactory results, post-operative morbidity due to persistent granulation tissue is substantial. It is through a multidisciplinary approach and close follow-up of the repaired airway that these demanding patients are best cared for. The long-term quality of life remains uncertain.
MeSH: Congenital tracheal stenosis, Multidisciplinary management, Heart defects, congenital, Prognosis

Introduction

Long-segment congenital tracheal stenosis associated with complete tracheal rings can cause life-threatening respiratory distress in neonates and infants. Surgery usually represents the ultimate therapy for this anomaly, particularly because of associated vascular and cardiac lesions. Initial emergent control of the airway is required with intubation and mechanical ventilation when possible, while planning a definitive surgical correction. If adequate oxygenation and ventilation cannot be achieved before operative repair, extracorporeal membrane oxygenation (ECMO) or intraluminal tracheal

stenting may be life-saving procedures as a bridge towards surgery. Post-operative therapy may involve prolonged ventilation and follow-up of the airway with bronchoscopy, during which granulation tissue removal and stenting may be performed as required. As these various treatment modalities imply, the care of the patient with congenital tracheal stenosis involves an initial emergent life-saving procedure, as well as a long-term strategy incorporating nursing, medical, surgical and interventional radiology teams. This case illustrates how these rare but demanding patients may be best cared for through combined multidisciplinary dedication, and the need for close post-operative follow-up.

Case

Baby CF was transferred emergently to our institution following elective caesarean delivery for failure to progress. At birth, he was in severe respiratory distress with no breath sounds entering either lung field and had decompensated respiratory acidosis. Endotracheal (ET) intubation was achieved with great difficulty with a size 3.0 mm tube before transfer from the outside hospital, although this was the expected size tube for the patient's weight. After failing conventional and then oscillatory mechanical ventilation, he was placed on veno-arterial ECMO at one day of life. Chest x-ray showed a tiny central airway shadow and bilateral whitening of the peripheral lung fields (Fig. 1).

Figure 1 Pre-repair chest x-ray after admission showing diffuse bilateral white-out of lung fields and veno-arterial ECMO cannulae in position

A bronchogram confirmed the suspicion of long-segment tracheal stenosis (Fig. 2). Echocardiography was performed and showed no intracardiac abnormality. After improvement of his peripheral perfusion, the lung fields gradually cleared following ECMO and continuous veno-venous hemofiltration

(CVVH). He achieved a satisfactory fluid balance and was deemed suitable for tracheal repair.

Figure 2 Pre-repair bronchogram demonstrating the string-like long-segment tracheal stenosis down to the carina. Note the relatively wider distal airway, highlighting the difficulty in diagnosing bronchomalcia
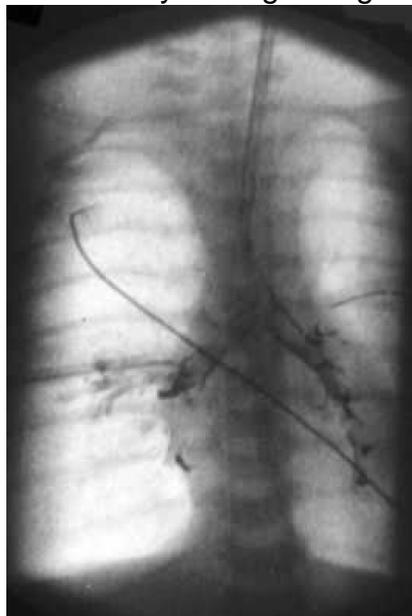
On day 6 of life and still on ECMO support, he underwent repair of his long-segment tracheal stenosis with an anterior autologous suspended pericardial patch, extending from cricoid to carina. The diagnosis of congenital complete tracheal rings was confirmed intraoperatively. A satisfactory repair was achieved, as assessed by intraoperative bronchoscopy, with a wide open patent patched trachea. Intraoperatively, a previously undiagnosed pulmonary artery (PA) sling was discovered, which was repaired by disconnecting the left PA and reanastomosing it to the main pulmonary artery anterior to the trachea. In the operative room, he was weaned from ECMO and decannulated. A period of 10 days to 2 weeks of endotracheal stenting with the ET tube is usual to allow for healing and consolidation of the pericardial patch to prevent collapse of the repair after extubation. In this case, the patient required a longer period of ventilation and relatively high mean airway pressures and PEEP to prevent atelectasis. Bronchoscopy and bronchography both diagnosed recurrent distal tracheal stenosis and unsuspected distal bronchomalacia, explaining the difficulty in weaning from the ventilator.

He underwent multiple bronchoscopies revealing persistent troublesome granulation tissue at the lower end of the pericardial patch repair. These granulations required repeat debridement and/or laser vaporization. After 7 weeks of continuous mechanical ventilation and repeat bronchoscopies, he underwent one further laser bronchoscopy of granulation tissue and balloon dilation of the distal trachea. His condition improved dramatically with marked reduction in ventilatory requirements (lowered mean airway pressures, FIO2 and rate), and it was decided to apply a more 'permanent' intraluminal dilating procedure. Thus, a balloon-expandable intraluminal metallic Palmaz

endovascular stent (8-12mm diameter, 30mm length) was inserted into the trachea under fluoroscopic guidance (Fig. 3). This procedure was uneventful. Following stenting, he was extubated to a nasal CPAP prong after 3 days. The CPAP was weaned over 2 weeks and he tolerated decreasing oxygen to room air.

Figure 3 Post-operative chest x-ray and intra-bronchial contrast material with initial tracheal stent in place. Note the pulmonary hyper inflation

Despite a satisfactory clinical condition, control bronchoscopy performed 3 weeks post stent placement revealed recurrence of granulation tissue, which was lasered. Two days later, he underwent balloon dilation of his stent (Figs. 4-6) and residual granulation tissue (balloon size 8). Finally, after two and a half months of hospitalization, he was breathing comfortably on room air, was afebrile, had a clear chest auscultation and x-ray image with well expanded lungs beyond a well-positioned Palmaz stent (Fig. 7).

Figure 4 Intraluminal ballooning of Palmaz stent to a wider diameter

Figure 5 Result of ballooned stent. Note the persistent malacia of the left mainstem bronchus visible on the bronchogram (as compared to Figure 2)
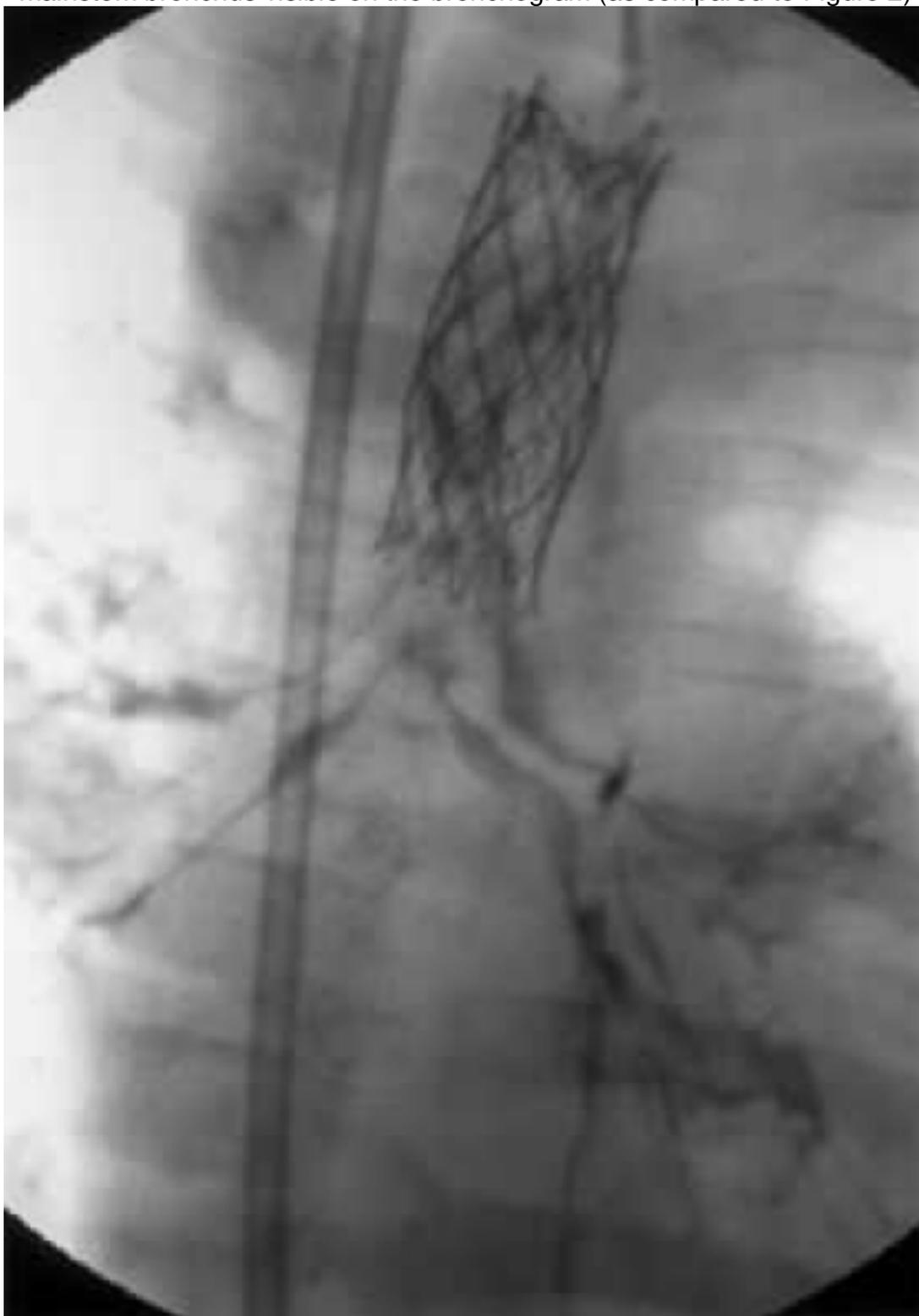
Figure 6 Final result showing a dilated stent and widely patent distal airway (as compared to Figure 5)

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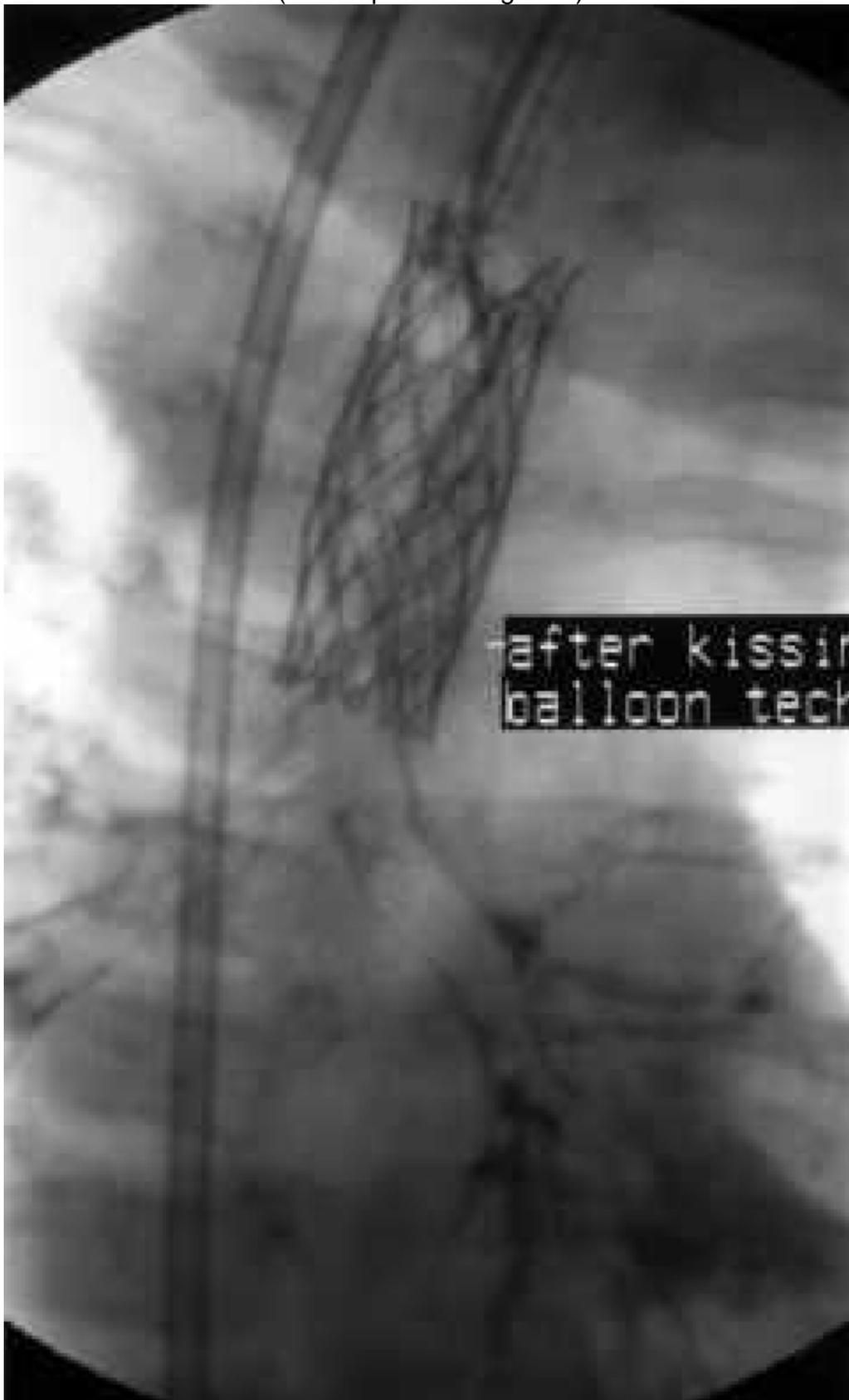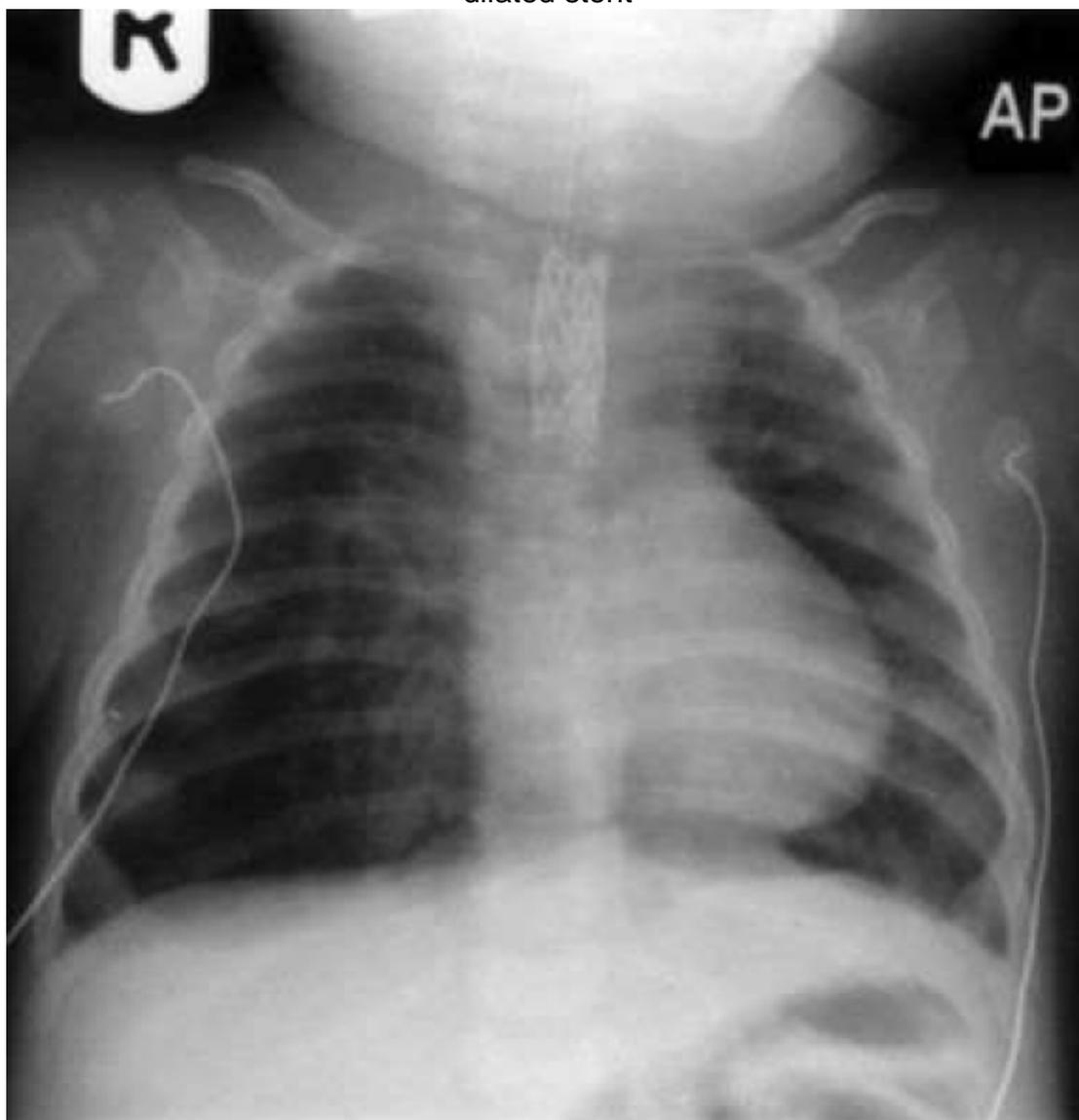
Figure 7 Chest x-ray prior to last discharge with clear distal lung fields and dilated stent

One week after discharge, he was readmitted to the hospital for cyanosis and respiratory collapse requiring emergent intubation. Bronchoscopy disclosed a large protruding polyp of granulation tissue at the carina, which was vaporized, and an otherwise patent stent. Subsequently, he was rapidly weaned and extubated with good recovery. Obvious close follow-up with bronchoscopy is planned since this last admission (Fig. 8).

Figure 8 Flexible bronchoscopy image of intra-luminal tracheal Palmaz stent in place
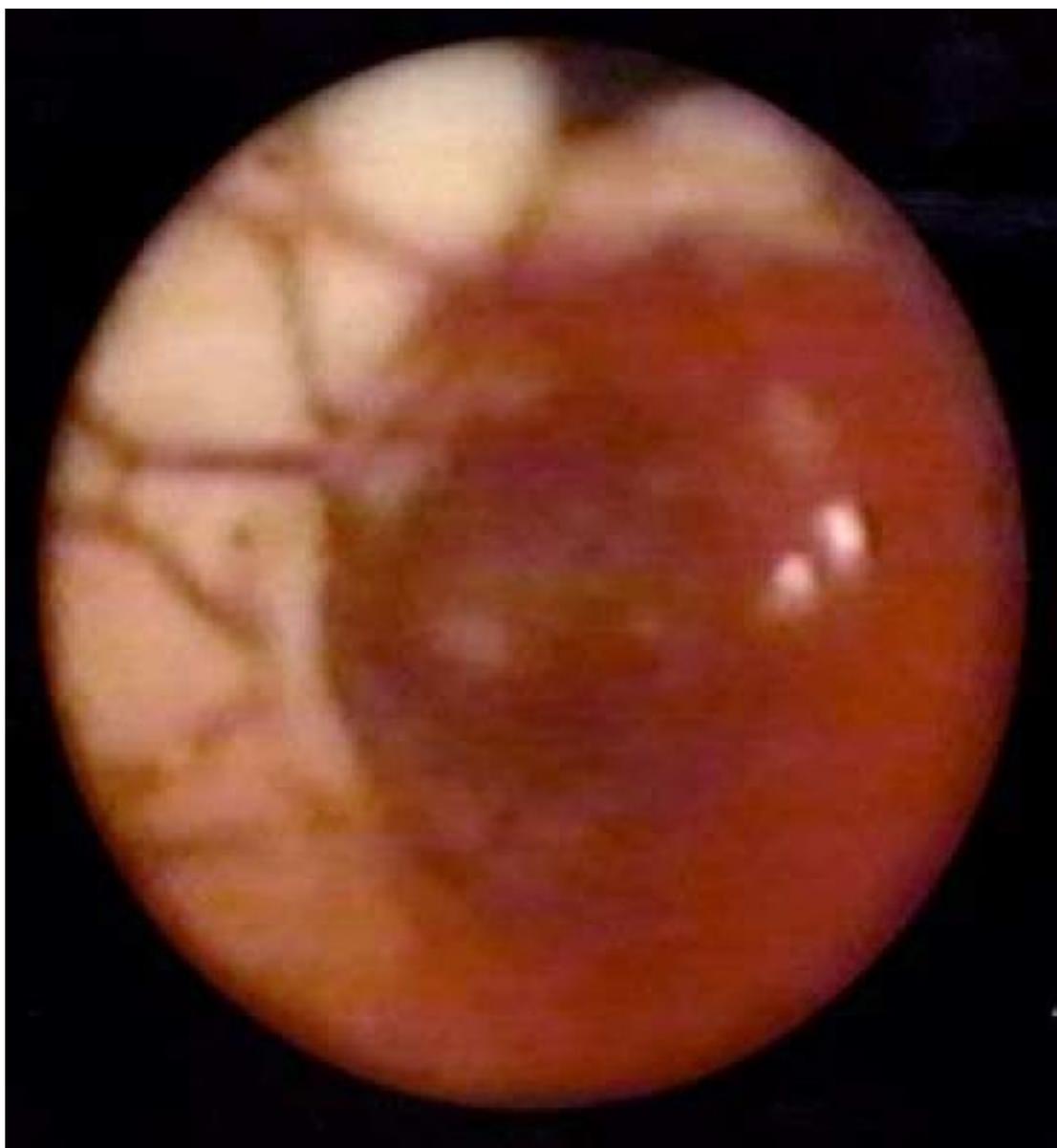
Discussion

Congenital tracheal stenosis associated with complete tracheal rings is a rare anomaly whose true incidence is unknown, as many infants expire before the diagnosis is made.[1] The absence of the membranous portion of the trachea may create local or generalized stenosis. Cantrell *et al* classified tracheal stenosis into three categories: segmental, funnel-like or generalized hypoplasia.[2] This anomaly is associated with a PA sling in 25-30% of cases[3] and 70% of patients with a PA sling have associated congenital tracheal stenosis. In our unit, 40% of patients with complete tracheal rings have had a PA sling. Associated cardiac anomalies are common, up to 20%, mostly represented by atrio-ventricular septal defects (AVSD) and tetralogy of Fallot.[3]

The clinical presentation may vary from stridulous cough and recurrent lower respiratory tract infections to acute and life-threatening respiratory insufficiency. Initial diagnostic workup includes a chest x-ray, and a barium swallow is performed to disclose an associated PA sling, seen as an anterior indentation on the oesophagus, if the patient is stable enough. A spiral CT scan of the chest will allow for reconstructive images of the trachea and assess the degree and length of tracheal stenosis, and will also document the PA sling, if present. Micro-laryngo-brochoscopy (MLB) can document the extent of tracheal disease. A bronchogram may reveal distal tracheo-bronchomalacia and is very useful when an MLB cannot reach the distal involvement of the anomaly. In order to reduce the occurrence of reactive airway disease, we use iotrolan (240 mg/ml) as a contrast agent. Finally, given the high prevalence of associated intracardiac anomalies, an echocardiogram should complete the preoperative assessment. Although our preoperative echocardiogram failed to pick up the PA sling, an echocardiogram is an accurate way of diagnosing a PA sling.

Non-operative management carries a dismal survival, with mortality ranging from 17-75%.[4] Nonetheless, temporary measures include ventilation with helium-oxygen, avoidance of a tracheostomy, and sometimes emergency endotracheal stenting or ECMO as a life-saving bridge to surgery.[4]

Classically, surgery represents the ultimate form of treatment for this anomaly, and techniques of repair have evolved over time. Ultimately, it is the length of the diseased trachea which dictates the type of reconstruction. Primary reconstruction with end-to-end anastomosis is possible with short segment stenosis,[5] but longer narrowing of the trachea involves various autologous or heterologous materials to bridge the defect. Bryant *et al*[6] introduced the pericardial patch tracheoplasty technique, modified by Idriss[7] using cardiopulmonary bypass (CPB) in 1984. Advantages of this graft material include the ready availability of pericardium, an airtight repair, the absence of rejection from an autologous tissue, a low metabolic rate requiring minimal blood supply and hence rapid healing and resistance to infection.[8] Systematic covering with the specialised ciliated respiratory epithelium occurs by migration of adjacent tracheal cells.[8]

Tsang *et al*[9] described the slide tracheoplasty technique in 1989, involving transection and spatulation of the proximal and distal ends through the stenosed area, with anastomosis of the two spliced extremities. Various other autologous graft materials have been used in conjunction with pericardium or alone, such as periosteum and costal cartilage.[10,11] More recently, tracheal reconstruction has been performed using cryopreserved scapular allografts,[12] cadaveric tracheal homografts[3] and cryopreserved tracheal allografts.[12,13] To enhance further the effect of cryopreservation by reducing allogenicity and hence rejection of these materials, some groups have attempted high dose irradiation[14] and photodynamic therapy[15] of these grafts.

The latest technique, described by Backer *et al*, involves the use of a free tracheal autograft, with or without pericardium.[16] The autograft is fashioned by tailoring the portion of stenosed trachea that would otherwise be discarded, and is used as an anterior patch. Theoretical advantages of this technique include the use of living autologous tissue already lined with respiratory epithelium, and autograft rigidity requiring less stenting by the ET tube, hence potentially reducing intubation times as compared to pericardium. The

potential for growth and technical facility are other attractive points of this operation.[16] Operative mortality of all types of tracheal reconstruction is low, varying between 0[9,10] - 17%,[3,17] and 5 year survival of up to 84% is reported.[17] The main concern with these patients is the significant morbidity, as demonstrated by their prolonged intubation times, intensive care stay, inhospital stay, and their need for very close follow-up requiring multiple bronchoscopies, stenting and ultimately reoperation. The common denominator of all postoperative morbidity is the formation of excessive granulation tissue at the repair site, i.e. the anastomotic line between the native trachea and graft materials. The protruding tissue causes various degrees of airway obstruction, failure to clear secretions, atelectasis and respiratory distress. In view of this frequent complication, or rather natural history of post-operative tracheal healing, the mainstay of follow-up is bronchoscopy. This may be either flexible for diagnostic and documentary purposes, or rigid for therapeutic granulation tissue debridement[16] and laser vaporization to free the airway.

All grafts may get infected, necrose or dehisce, with a resultant potential for air leak and mediastinal emphysema requiring reoperation. Indications for reoperation include requirement for frequent bronchoscopic dilations, reintubation for respiratory distress and failure to wean from mechanical ventilation.[18] The reconstructive technique in reoperation often involves repeat use of pericardium if any is still available, or autologous cartilage grafts.[18] The incidence of reoperation after pericardial patch repair is as high as 29%,[18] but is achieved with low mortality.

Intraluminal stents have gained recent popularity in the management of congenital tracheal stenosis, either preoperatively as a life-saving bridge to surgical reconstruction, or postoperatively in the event of a failed tracheoplasty procedure.[19] Various types of stents exist, including the Dumon silastic stent and the metallic Palmaz stent. Dumon stents are more frequently used in the setting of tracheal neoplasm or corrosive strictures, mold the preexisting tracheal form, are readily removable, but cannot be dilated. They have a continuous wall which does not allow cross-ventilation into branch bronchi. More commonly employed are the balloon-expandable metallic wire Palmaz stents. They are dilatable up to a certain degree and have a meshwork which allows ventilation across them into smaller branching bronchi. Disadvantages include incorporation into the tracheal wall after reepithelialization and extreme difficulty in removal.[19] Despite initial satisfactory enlargement of the airway, granulation tissue does develop and protrudes through the stent meshwork into the lumen. This may be managed by repeat ballooning which serves to compress the bulging granulations against the tracheal wall. Further dilatation of the stent is possible. Intraluminal stents represent a useful addition to our options, but their long-term role remains to be determined. Absorbable stents are currently under trial and may prove a valuable future asset.

Current research is being undertaken on improvements in tracheal healing, more specifically aimed at the reduction in granulation tissue formation, which universally complicates the repair anastomotic site. Vascular endothelial growth factor (VEGF) is an experimental protein which is implicated in wound healing,[20] is angiogenic,[20] increases vascular permeability, stimulates nitric oxide formation by the endothelium and hence vasodilation in the setting of

tissue hypoxia and ischemia.[20,21,22] In a rabbit model of tracheal reconstruction using tracheal autografts, pretreated with topical VEGF, autograft healing was shown to be enhanced.[23] Topical VEGF reduced granulation tissue formation, induced less inflammation of the free tracheal autograft, minimized anastomotic fibrosis and hence airway obstruction.[23] The future will also involve tissue engineering to grow trachea from the patients' own cell lines.

Conclusions

Congenital tracheal stenosis from complete tracheal rings presents a rare but very challenging group of patients. Treatment requires a multidisciplinary approach, with close coordination and collaboration amongst the various teams. Non-operative therapy carries a high mortality, but initial stabilization by agressive medical management, intraluminal stenting or ECMO may buy enough time to allow a full diagnostic assessment prior to surgery.

Surgical repair represents the mainstay of therapy and is achieved with low mortality, with initial good anatomical and functional results. However, morbidity is substantial and recurrent difficulties with the airway remain a constant threat. Besides the prolonged hospital stay, outpatient and home care place the families of these patients under considerable stress. Thus, the initial care 'contract' with the involved families should be explicit about the degree of uncertainty. The long-term quality of life of these patients is as yet unknown, and the degree and intensity of morbidity will be better indicators of outcome than simple mortality figures.

Close follow-up with bronchoscopy is mandatory and interventional procedures such as granulation tissue debridement, vaporization, balloon dilation or intraluminal stenting should be available and considered as required. "Virtual bronchoscopy" may also be obtained using three-dimensional reconstructive imaging of a spiral CT scan of the trachea.

Future advances may evolve around optimal tracheal healing and better control of inflammation, fibrosis and granulation tissue formation around the surgical repair site. VEGF and tissue engineering may constitute the major breakthroughs at a cellular level to improve the clinical results and postoperative care after surgical repair of congenital tracheal stenosis.

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