Review of pediatric airway malacia and its management, with emphasis on stenting

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Summary

Malacia of the pediatric airway presents itself in a variety of clinical circumstances. Pediatric airway stenting is a more recent treatment modality. Complications may necessitate stent removal. This is usually performed bronchoscopically. We were forced to surgically remove a complicated airway stent. The Palmaz stent had been inserted for bronchomalacia presenting after interrupted aortic arch surgery in a 4-month old child with DiGeorge syndrome. This prompted us to review pediatric airway malacia, its management options and long-term outcomes, in an attempt to crystallise the current status of this relatively uncommon and difficult issue. The role of stents is analysed.

Keywords: Tracheobronchomalacia; Airway stenting; Aortopexy

1. Introduction

Very occasionally, difficult airway issues frustrate the results of otherwise successful and difficult cardiac surgery. Our experience is described below, which stimulated this review.

A full term male neonate was diagnosed with Type B interrupted aortic arch and a subaortic ventricular septal defect with a small ductus feeding the descending aorta. The child had chromosomal 22q11 deletion with a diagnosis of DiGeorge syndrome. We performed a staged correction as he was poorly nourished and sick on presentation. The aortic arch was reconstructed in the first week of life and a pulmonary artery band was placed. He was extubated on day 3. He presented with respiratory distress due to left bronchomalacia at 2 months. Following intubation, he remained ventilator dependant. At 4 months, a left bronchial Palmaz stent was inserted. The VSD was corrected when he was 23 months old and he has remained well. Symptomatic at three and a half years, bronchography revealed a near-total occlusion of the left bronchus proximally. The stent had migrated further into the left main bronchus with tissue obstruction proximally. Two wire points at the proximal stent orifice had also bent into the lumen. Stent dilatation was not feasible as the wire points would burst the expanding balloon and might snag the balloon afterwards, precluding a safe procedure. An earlier bronchoscopic attempt to remove the stent failed as it was firmly adherant to the bronchus distally and bled on manipulation.

We therefore decided to remove the stent surgically with cardiopulmonary bypass standby. Via a median sternotomy with endotracheal intubation, the carina and left main bronchi were exposed. The sites for aortic and atrial cannulation, if required were identified. The recurrent laryngeal nerve was avoided. The left bronchial anterior wall was incised transversely to expose the stent inside. Using an endarterectomy dissector, a plane was carefully developed circumferentially between the stent and the bronchial wall till it completely dislodged. Care was taken at the posterior pars membranacea. Bleeding was not excessive. The bronchotomy was closed with interrupted 5/0 prolene suture. There was no air leak on testing under water. Oxygenation was acceptable throughout and there was no need for cardiopulmonary bypass. Fibreoptic bronchoscopy confirmed a patent bronchus. Except for an early chest infection, he was uneventfully discharged.

Though successful, this potentially life threatening sequelae, raises the issue of long term results of initial viable treatment options. The aim of this review is to gather the worldwide-published experience in the English language on pediatric airway management. The emphasis is on malacia. Congenital stenosis and secondary laryngotracheal stenosis is another issue and is not dealt with here.

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Conservative, surgical and the evolving stent experience is collated and streamlined. We hope to provide a useful information source for this unusual issue, for which personal experience is often limited and the literature dispersed.

2. Material and methods

Published world-wide literature in the English language in the last 40 years was searched using PubMed and Medline Databases. Keywords used include airway malacia, tracheobronchomalacia, tracheobronchial obstruction, bronchomalacia, aortopexy, and airway stenting. Case series and case reports were used. Not many reviews existed on the topic (Table 1). The vast majority of airway stent experiences are reported in the adult. We included useful learning points from the adult to serve as a mould for the decision making process in the pediatric population.

2.1. Incidence

A few papers help provide an incidence perspective on the problem. The issue has been varying reported by Respiratory and Critical care physicians, Head and neck surgeons, and Cardiothoracic and Vascular surgeons. Masters [1] had 299 bronchoscopically confirmed pediatric cases of tracheobronchomalacia over a 10 year period. There was a 2:1 male to female ratio. 13.7% had congenital cardiac and cardiovascular anomalies, 9.7% was associated with tracheoesophageal fistula and tracheomalacia, while 8% had syndromic pathology. Jacobs [2] found a 15% incidence of primary tracheobronchomalacia among all bronchoscopies performed in infants for respiratory distress. The Great Ormond Street Hospital experience [3] provides an incidence seen in a specialised pediatric intensive care unit. Forty-eight patients (median age of 4 months) over a 5-year period were reported in 2001. Vascular anomalies comprised 18% while cardiac defects contributed to 35% of the heterogeneous case mix. Dedicated personal experiences vary widely. Backer, in a comprehensive review detailed 204 children (mean age 13 months) from 1947 to 1987 with vascular anomalies causing airway compression [4]. About 10% of vascular anomalies within the mediastinum led to residual malacia after primary surgical treatment. Series on airway issues secondary to specific congenital cardiac anomalies have fewer numbers. McEhinney [5] reviewed the pathophysiology with right-sided cardiac obstructive lesions with a case series of 5 infants. Khatami reported his 7-year experience on 11 patients with Tetralogy of Fallot and absent pulmonary valve [6].

2.1.1. Etiology

Hollinger first described Tracheobronchomalacia (TBM) [7]. Primary TBM is an inherent weakness in the structural integrity of the cartilaginous ring and arch [8] and may be associated with a widened posterior membranous wall. This may occur in isolation, with prematurity and bronchodysplasia, or secondarily, in association with various congenital abnormalities or genetic syndromes with craniofacial abnormalities [2,9]. Huang reviewed the spectrum of structural airway anomalies in the DiGeorge syndrome [10].

Tracheoesophageal fistula is a well-established cause of secondary malacia. It mainly affects the trachea [11,12]. Vascular mediastinal anomalies form another significant group [4,13,14]. In the seminal paper by Gross, aberrant great vessels in the superior mediastinum forming rings and slings were described, producing both tracheal and bronchial compression. These include double aortic arch, right aortic arch with a left ligamentum arteriosum (circumflex aorta), anomalous innominate artery, anomalous left common carotid artery and aberrant right subclavian artery. PA slings were described later.

Bronchomalacia is more commonly associated with congenital heart disease. Berlinger linked the anatomical relationships, predisposing the bronchus to compression from the adjacent dilated or hypertensive pulmonary artery and left atrium. This complicates acyanotic heart disease with large left to right shunts as in VSD and PDA [15].

Aneurysmal PA dilatation producing severe airway compression is typified in the Fallot with absent pulmonary valve [16]. The left lateral lower trachea is in intimate contact with the ascending aorta. Dilated ascending aorta producing tracheal and carinal compression is associated with pulmonary atresia type congenital heart disease. Right-sided cardiac obstructive lesions leads to shunting and flow related accelerated intrauterine growth and malposition of the aortic root [5].

Airway compromise has been reported after correction of congenital heart disease. The Lecompte manoeuvre of posterior transfer of a large ascending aorta in the arterial switch operation [17], the interrupted aortic arch repair, conduit reconstruction of pulmonary outflow tract and conduit shunt creation have been substrates [14].

Miscellaneous conditions such as mediastinal cysts, teratomas, neoplasms, thyrom hyperplasia, pectus excavatum [18] can congest the small infant mediastinum and compromise the airway. Pediatric lung transplantation has

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presented with the difficult airway issue [19]. Malacia should also be considered in the differential diagnosis of persistent wheezing in the young.

2.1.2. Pathophysiology

Infants are clinically more susceptible to airway compromise than older children and adults. As airway resistance correlates inversely with the fourth power of its radius, a slight calibre reduction of the small infant airway has a relatively large impact on airway resistance. The glottis in infancy has an antero-posterior diameter of 7 mm and a posterior transverse dimension of 4 mm. Mucosal edema of 1 mm will reduce the cross-sectional area by 35% [7]. The Venturi effect also diminishes the pressure within the malacic segment. They, therefore, easily become symptomatic. A small calibre bronchus is more easily compromised than the larger sized trachea. If interventions are to be successful, these luminal dimensions must be preserved post-therapy. Young airways remain prone to malacia even after repair and removal of any extrinsic compression. With the continued growth and development of the airway, the condition especially in the trachea is often self-limiting, usually by the age of 3 years. However, bronchomalacia may not resolve as completely with growth [20].

The left mainstem bronchus appears to be the more common site of isolated BM, possibly because it is longer without branch points than the right mainstem bronchus.

A variety of symptoms may be seen.

TBM is characterised by expiratory airway collapse, exacerbated by increased respiratory effort such as crying and coughing [4,13]. This can be so severe as to manifest as ‘dying spells’ in trachoesophageal fistula [21]. Sometimes it may even require deep sedation or paralysis and manual thoracic compression to overcome acute obstructive events [22].

Airway collapse leads to an inability to clear secretions, atelectasis, recurrent infections, hypercapnea, hypoxemia and respiratory failure. There may be a hesitation to swallowing. With tracheal involvement, the infants characteristically tend to lie with their heads hyperextended. This relieves the obstruction by stretching and splinting the trachea open [13].

With bronchial involvement, air trapping may result in lobar emphysema [23]. It commonly affects the upper lobes even when the main bronchi is involved in the pathology [24]. The expiratory forces acting on the lower lobes are much greater than those acting on the upper lobes. The upward rising diaphragm and more forceful movements of the lower ribs would more likely drive air out of the lower lobes past a bronchial obstruction. The upper lobe, acted upon more passively, is therefore more prone to obstructive emphysema. Rarely, unilobar distention may assume immense size leading to Nelson’s ‘tension emphysema’ [25].

Failure to wean from ventilation after successful correction of congenital heart disease or other neonatal surgical problems is another common presentation. The amount of airway pressure required for optimal ventilation is an indicator of severity.

2.2. Accurate physiologic and anatomic diagnosis

An accurate diagnosis is vital to plan the most effective treatment.

Masters provide a detailed description of the endoscopic diagnostic features in airway malacia at three levels—larynx, trachea and bronchi. Appearances such as ‘Eifel Tower’, ‘Inverted Teardrop’ and ‘Lunar crescent’ are illustrated [1].

Cheung et al. recently reported their experience in combining cine tracheobronchography and angiocardiology in the problematic airway, in comparison to flexible bronchoscopy or MRI [26]. The former provided detailed information on the spatial relationship between vascular and airway structures and allowed a dynamic assessment of airway malacia. This showed that all patients with an abnormal anatomy did not necessarily have actual vascular compression and instead had primary airway malacia. This facilitated the appropriate treatment of the airway instead rather than vascular correction. Tracheobronchography also allows precise measurement of the airway, facilitating precise stent calibration and avoiding the risks of increased compressive force on the mucosa from size mismatch.

With the advent of ultrafast computed tomography, dynamic 3D airway imaging is available to diagnose malacia within a few minutes. The state of the art imaging technology was recently described by Boiselle [27]. In patients being considered for stent placement, computed tomography allows (1) accurate representation of airway anatomy in three dimensions (2) measurement of airway diameter, (3) evaluation of airway anatomy distal to a narrowed segment that is invisible to bronchoscopy, (4) demonstration of dynamic changes in airway morphologic features during forced exhalation in patients with airway malacia, and (5) demonstration of focal or diffuse air trapping in lung peripheral to the abnormal airway. In patients who have had stent placement, computed tomography is valuable in assessing airway morphologic features and dynamics distal to the stent, and can be valuable in assessing stent dysfunction. However, Burke found that such ‘virtual endoscopy’ was not as sensitive as actual endoscopy in detecting dynamic airway obstruction [28].

Hertzog, in a single case report in a patient with airway malacia describe the use of CT technology in the ICU setting to determine the ventilatory settings for optimum lung volumes, with minimal disruption of patient care [29].

Though CT is non-invasive, bronchoscopy remains invaluable to monitor and assure the intraoperative results of pexy procedures.

3. Treatment options

3.1. Conservative therapy

The initial consideration in a primary pathology should be positive pressure ventilation and if prolonged, conversion to a tracheostomy. There are a few excellent reviews on tracheostomies in the first year of life [30,31]. Reassuringly the morbidity associated with the home care period was very low. The length of the tracheostomy period was greater than 12 months in 50%.
3.2. Surgery

3.2.1. Relief of external compression

The initial strategies focussed on tracheomalacia. Direct surgical correction of vascular anomalies [13] include division of the ring/ductus, aortic uncrossing [5], etc.

3.2.2. Pexy operations

The concept of arterio/aortopexy was initially described by Gross and Neuhauser for vascular tracheal compression [13]. The ascending aorta was lifted and adventitia sutured to the undersurface of the sternum. Gross’s concept was adapted later to tracheoesophageal fistula [12]. Though very effective in tracheomalacia, Filler reported an 80% failure rate when aortopexy was applied to tracheobronchial or bronchial involvement [11]. However, by relieving the associated element of tracheal collapse with a subsequent decrease in total airway resistance, aortopexy reduce the overall expiratory effort. This indirectly leads to some success in relieving bronchial collapse. Variations of the original technique such as pericardial flap aortoplasty minimise trauma to the aorta [32]. Thoracoscopic approach has also been described.

Bertlinger described pulmonary arteriopexy to relieve left bronchial obstruction in interrupted aortic arch and VSD [15]. Kamata et al. distinguished the roles of aortopexy versus PA pexy in a study of 14 children [33]. Their observations suggest that aortopexy is helpful to eliminate collapse of the distal trachea, the right main bronchus and the proximal half of the left main bronchus. On the other hand PA pexy was shown to eliminate collapse of the distal half of the left main bronchus. More aggressive management with aortic arch extension with conduit for bronchial compression after Interrupted aortic arch repair has been described [34].

In isolated bronchomalacia, bronchopexy is performed. Here, the left main bronchus is suspended to the ligamentum arteriosum [20,35]. This is certainly preferable to former methods such as a pneumonectomy.

In the absent pulmonary valve, extensive aneurysmorhaphy/plication rather than PA suspension lifted the PA bifurcation away from the carina to relieve the obstruction [36]. Litwin described transposition of the PA anterior to the aorta.

Aortopexy generally has excellent results for tracheomalacia. Filler [11] reported his 7-year experience with 21 patients while Vinograd [37] reported the 12-year results on 28 patients. In the latter, there was 1 post-operative death, 25 free of disease, and 3 on tracheostomy.

Austin reviewed the anaesthetic experience for these situations [9].

3.3. Stent

3.3.1. Hagl’s external stent and pexy hybrid

For a short segment malacia, resection and end-to-end anastomosis can be performed. However, the persistence of dysfunctional pars membranacea, the tendency for recurrent stenosis at the anastomosis due to tension and the reduced growth potential make this less attractive.

Hagl described the concept of external stabilisation using polytetrafluoroethylene PTFE [38]. In this, the stent is placed outside the trachea and the sutures are used to approximate the tracheal adventitia to the stent, thus holding open the lumen of the malacic segment. The reactive mucosa remains untouched and granulation tissue formation is less of an issue. Six tracheal and 2 left main bronchus suspensions were described. Seventy-five percent required extracorporeal circulation. The ages at bronchial stabilisation were 20 months and 4 years and with a reported follow-up of 25 and 54 months.

This technique anticipates the issue of growth by using oversized grafts; 20 mm diameter for the trachea and 14 mm for the bronchus. The authors emphasise that in the trachea, an anteroposterior diameter of 11 mm and a transverse diameter of 13 mm, seem sufficient; with symptomatic stridor expected at less than 6 mm even in the adult. The oversizing resulted in an immediate and significant reduction of airway resistance facilitating prompt extubation between 7 and 10 days.

3.3.2. Internal airway stenting

Against this background of surgical options, airway stenting has carved out a niche. Stenting aims to support the airway till it regains its structural integrity with growth. Stents have the apparent appeal of transbronchial insertion and avoiding another operation in the already ill patient.

Airway stenting in adults was first described in the trachea by Montgomery using silicone T-tubes in 1965. The larger size of silicone stents is not as adaptable in a smaller pediatric airway. The unfavourable wall to lumen ratio causes significant reduction in the cross-sectional diameter. The large continuous surface area of the stent does not epithelialise and interferes with the respiratory mucociliary function. They may repeatedly become obstructed from inspissated secretions and granulation tissue predisposing to secondary infections and bleeding. Stent migration [39] is another problem.

Since 1969, metallic stents have been pursued to supplement balloon angioplasty in the treatment of arterial occlusive disease. Wallace et al. in 1986 first adapted an expandable wire intravascular stent for use within the tracheobronchial tree [40]. Progressing from earlier experiments with coiled steel springs [41], Farga and Filler in 1997 provided an evidence-based understanding of the effects and feasibility of a metal stent in the airway [42]. In their study on the trachea of cats, granulation tissue was the commonest complication. The points of maximum movement at the upper and lower ends of the stent as well as stent overdistension with increased tension on the wall were the most important factors for excessive granulation tissue formation. Overdistention risks perforation and led to more fibrosis. The study did not address the issue of stent removal.

3.4. Infection

Granulation tissue associated with stents, is colonised by numerous bacteria, including Streptococcus viridans, Pseudomonas aeruginosa, non-hemolytic streptococci and Staphylococcus aureus [43]. This could lead to generalised sepsis in an already ill child. Santoro felt that the infective problems in their series were not related to stent implantation. More aggressive management of
tracheomalacia by earlier stent implantation instead, might have avoided prolonged assisted ventilation and lessened the likelihood of secondary infections [44]. This concept could lower the eventual morbidity and mortality. The permanence of a metallic foreign body may be undesirable in lung transplant recipients, who are receiving immunosuppressive medication.

Duda analysed the physical properties of 10 brands of metallic stents [45]. Respiratory stenting currently probably represent about 6% of the worldwide non-vascular stent practice [46].

3.5. Types of metal stents

The stents used in infant airways include the balloon expanded Palmaz, the self-expanding Wallstent, the Nitinol/ Ultraflex and occasionally the Gianturco stent.

3.5.1. Palmaz stent (Johnson and Johnson Interventional Systems, USA)

Robert M. Filler et al. at Toronto has the single largest reported experience to date with infant airway stenting. The balloon expanded Palmaz stent appears to be the most widely used and emerged in the late 1990s as a possible short-term infant airway solution.

The pioneering experimental work of Julio Palmaz on balloon expandable intravascular stent dates back to 1985 [47,48]. The Palmaz stent is constructed as a stainless steel tube with rectangular slots. The stent is balloon expanded to fit the desired cross-section and should exert no potentially tissue damaging outward pressure. The stent can be inflated to a diameter of 8-12 mm and is available in lengths of 10-40 mm. Conveninetly it is small enough to pass through a 3 or 3.5 mm endotracheal tube or bronchoscope.

3.5.2. Wallstent (Boston Scientific Corporation, USA)

Goldstraw was the first to use the Wallstent in the airway in the adult in 1988 [49]. Other reports followed [50,51]. The Wallstent is a braided tubular mesh made of cobalt-based alloy. It consists of 16-20 wires woven in a crisscross fashion, forming a cylindrical mesh. It is available in diameters from 8 to 24 mm and in lengths from 20 to 60 mm.

3.5.3. Nitinol/ Ultraflex stent (Boston Scientific Corporation, USA)

A novel technological advance was reported by Vinograd in a rabbit model in 1994 [52]. Nitinol, a nickel-titanium alloy exhibits a shape memory phenomenon known as the Marmen effect. This is a thermodependant metal-crystalloid structure change. At low temperatures or martensitic state, the alloy can be moulded to a straight compact form for deployment. When heated to a higher temperature or austenitic state, the stent can be made to memorise a helical shape. It regains its original shape when re-cooled. The austenitic characteristic is very important, because after expansion, the nitinol stent does not increase its pressure on the airway wall. This contrasts with the expandable metallic stent in which there is constant elastic pressure on the airway. Over-expansion is unlikely as body temperature does not normally rise beyond 41 °C. The stent can be tailored to various sizes during manufacturing to limit its expansion in the austenitic state. The stent can be inserted, fixed and removed easily in its martensitic state, even in a very small airway.

The elasticity of this alloy of 11%, biodynamically closely resembles that of the tracheobronchial tree. This is much greater than stainless steel at 0.5% [53]. Currently, only nitinol shows a hysteresis curve in stress-strain tests that approximates an elastic tissue such as cartilage. The return to the starting point after a change in size is delayed. The force exerted onto the airway by the stent therefore does not suddenly increase even when coughing increases its compression. This property prevents damage to the mucosa. It also has sufficient resistance to external pressure to avoid stent deformation. It is kink and fatigue resistant. Nitinol has high tissue biocompatibility [54]. Stainless steel in comparison, may be highly corrosive in the presence of chloride ions in the body fluids and can incite chronic inflammation, fibrosis, necrosis, etc. [55].

The Ultraflex is a self-expanding device made of nitinol [56]. It has a cylindrical open knitted loop design made from a single strand of nitinol wire and available in 8-20 mm diameter. It does not fully expand to its austenitic state immediately after release, allowing time to readjust the position. The flexibility also allows it to fit to complex stenotic shapes. It scarcely migrates and it can even be safely implanted in the subglottic region. It can be easily removed before complete epithelisation. This is unlike other stents, which are difficult to remove even if not covered by mucosa. Though not clearly visualised on CXR, it advantageously neither shows artefacts on CT nor problems with MRI [57]. Miyazawa published the first multicentre study in adults [58].

3.5.4. Gianturco stent (William Cook, Denmark)

The Gianturco stent has undergone several modifications [40]. The present Gianturco-Z stents for the airway possess diameters of 6-35 mm and lengths of 2.5-5 cm [59].

4. Comparative clinical analysis of airway stents

Various reports have highlighted the possible adverse events with stents, as documented mainly in the adult. These provide an insight into its physical limitations and perhaps represent a cautionary note for the enthusiasts faced with infants with dissimilar airway characteristics.

4.1. Deployment and repositioning

Palmaz stents can be precisely deployed, with minimal shortening on dilatation. The Gianturco stent does not shorten on deployment but tends to unpredictably spring open making it slightly more difficult to position precisely. Repositioning of Gianturco stents is limited by the fact that they have metallic hooks attaching it to the wall of the airway. Though Palmaz stent has no hooks, once it is against the wall of the airway, it is extremely difficult to move. Precise placement is not easily achieved with the Wallstent because of its significant shortening 20-40% [59] during deployment. A Wallstent cannot be advanced further.
into the airway but can be pulled back after being partially deployed. In the only available comparison between the Wallstent and Ultraflex done in an adult population, the Ultraflex stent was more flexible and easier to reposition or remove after deployment [60].

4.2. Stent interstices

Palmaz stents have a mesh size between the finer mesh of the Wallstents and the large interstices of Gianturco stents. The fine metallic mesh could become encrusted with secretions, causing airway obstruction. There have been two reports of the use of a bronchoscopic laser to remove the filaments of a Wallstent causing lobar obstruction in three patients [61,62]. Filler cautions against the use of laser, that essentially contributed to death in an infant [63]. An alternative to laser, is to pass a wire through the interstices of a Wallstent and dilate a hole in the side of the stent with use of a balloon catheter [64]. Being a multifilament in construction, fragmentation of the Wallstent wire filaments has been reported.

4.3. Stent rigidity, flexibility and complications with the Palmaz stent

The rigidity of Palmaz stents suggests the potential to provide a significant radial force following balloon expansion. Unfortunately, the longitudinal stiffness and lack of radial compliance permits deformation and collapse. When compressed or bent along their longitudinal axis, Palmaz stents do not recoil to their original expanded configuration. Perini reported the clinical course of 24 Palmaz stents inserted in 8 adults for bronchial anastomotic strictures and malacia after lung transplantation [65]. Deformation and crumpling of the bronchial stents were seen in 2 patients after 2.5 and 7 months. A deformed stent migrated from the left mainstem bronchus to the right lower lobe bronchus. None had extrinsic compression or had tracheobronchial suction. Stent failures presumably resulted from coughing, which generates high intrathoracic and transmural compressive airway pressures. The bronchial position is perhaps more vulnerable to stent migration due to its greater anatomical mobility and proximity to pulsatile great vessels. Stent collapse and migration could result in death secondary to airway obstruction. Perini has since stopped using Palmaz stents in the tracheobronchial tree. The airway pressure in infants probably does not reach these deforming levels, and no such complication has yet been reported.

Stent flexibility was addressed by Hautmann [66]. The lack of longitudinal flexibility is undesirable if the portion of the airway to be treated is non-linear [64]. The Gianturco stent has some degree of flexibility at the junction points of the individual stent bodies. The Wallstent is more flexible, easily able to accommodate acute angulation, and also shown to have a greater dynamic expansive force than the Ultraflex stent [60].

4.4. Hazard of periodic balloon expansion

Expandable stents require periodic inspection to exclude size mismatch from the continuing airway growth. Periodic balloon expansion lessens the potential to dislodge. This has its attendant procedural risks.

Miyamoto reported tracheopulmonary artery fistula in a 7-month baby following dilatation of a distal tracheal Palmaz stent, inserted following slide tracheoplasty for pulmonary artery sling. The baby was rescued with emergency cardiopulmonary bypass and repair. Stepwise stent expansion and migration brought the lower edge close to the inferior wall of the carina to eventually perforate it [67]. Slonin reported a bronchial mucosal laceration with dilatation in 1 patient [64]. Repeated bronchoscopy carries a risk of stent damage [63].

4.5. Intrinsic and extrinsic pressure erosion

Erosion and migration would be expected to be more common from the mainstem bronchus than the trachea because the airway wall is thinner and has less cartilage support. As the Palmaz stent is balloon expanded, it can be custom-sized to fit the airway without the risk of intrinsic pressure erosion. This complication has been reported with Gianturco stents [68,69]. Slonin reported a severe Palmaz bronchial stent deformation and extrinsic erosion from a thoracic aortic aneurysm compression [64]. This serves to reemphasise the importance of dealing with the primary vascular pathology in the first instance, except perhaps in unusual circumstances. A child with Ehlers Danlos syndrome and an aortic arch aneurysm was initially stented with a Wallstent at 2 months. Aneurysm surgery was deferred to a more mature age of 54 months [70]. With its greater flexibility and less radial pressure, the Wallstent is more suited in situations with extrinsic compression.

The potential for stent erosion is of particular concern in the cyanotic patient who has increased dilated bronchial collateral vessels in close proximity to the stent site. Wells reported major bronchial hemorrhage in such situations with a Palmaz stent and even a Wallstent. Nitinol stents are perhaps better suited here [71].

5. Major case series and results with pediatric airway stents (Table 2)

5.1. Palmaz stent

Filler and Forte et al. in 1998 reported the largest clinical experience with Palmaz stents to date with a 5-year follow-up of 30 stent insertions in 16 infants with a median age of 9 months [63]. The majority of the stents were tracheal. 40% of the stents were bronchial of which 80% (10 stents) were in the left bronchus. The longest follow-up in the series was 56 months for a tracheal stent and 25 months in the left bronchus. There was no stent related mortality. Safe removal of stents, almost entirely from the trachea was reported, at an average age of 14 months. These were notably only described in cases of tracheoesophageal fistulas. One tracheal stent was removed as late as after 4 years. The authors also describe a death after attempted bronchoscopic removal of a tracheal stent, which had laser treatment earlier for granulations. The stent was firmly incorporated into the tracheal wall and manipulation...
resulted in total airway occlusion. All stent removals were performed bronchoscopically. Stent migration notably complicated left bronchial stents in congenital heart disease. A 1-year-old left bronchial stent was found to cause a full thickness bronchial erosion, but with no evidence of perforation. The data therefore leaves the long-term safety of stents in the bronchial position uncertain.

Furman and Backer et al. in 1999 reported 12 Palmaz stent insertions in 6 infants [72]. There was equal number of tracheal and bronchial insertions. Mucosalisation was observed to occur between 20 and 90 days. Furman suggested that mucosalisation prevents stent removal and that such stents should be regarded as permanent. This is in contrast to Filler's series in which stents were removed successfully at a much later time.

Santoro reported the use of the Palmaz stent in 3 neonates with vascular anomalies [44]. Though all were promptly extubated, two died from sepsis eventually. Khatami et al. described the use of 4 Palmaz stents in 2 patients in a series of 11 infants undergoing complete repair of Tetralogy of Fallot with Absent Pulmonary valve. One child died of sepsis and the other is doing well at home at 2 years [33].

Geller reported the use of Palmaz stents in 9 children with tracheomalacia. There were 3 deaths from significant tracheal hemorrhage [73]. They cautioned stenting as a final resort.

5.2. Wallstent

Goldstraw reported the use of the Wallstent in infants. 10 Wallstents were used in 5 infants with congenital heart disease who had been ventilator-dependant [70]. These included 2 infants who had been ventilator-dependent and 1 infant who required reintubation for subglottic stenosis in 5 infants [22]. There were 2 tracheal, and 11 bronchial insertions in 1 infant, with 10 months follow-up. Granulation of the stent could be observed without replacement with forceps or rotation technique. All were removed and were found to be patent after 4 months. The longest follow-up was 54 months for a left bronchial stent. No stents were removed.

5.3. Nitinol/Ultraflex stent

Nicolai reported the first experience with 13 Ultraflex stents in 7 infants with structural airway pathology [22]. There were 6 tracheal, 1 left bronchial, and 6 bronchus intermedius stents. Granulation of the stent was observed without replacement with forceps or rotation technique. All were removed and were found to be patent after 4 months. The longest follow-up was 50 months for bronchial stent. No stents were removed.

5.4. Gianturco stent

Numerous reports in adult tracheobronchomalacia have shown a prohibitive risk of wire fracture and unraveling. Numerous reports in adult tracheobronchomalacia have shown a prohibitive risk of wire fracture and unraveling.

Table 2

<table>
<thead>
<tr>
<th>Ref</th>
<th>Author</th>
<th>Country</th>
<th>Year reported/</th>
<th>Number of patients</th>
<th>Median age/</th>
<th>Etiology</th>
<th>Stent type</th>
<th>Number of metal stents</th>
<th>Location of stent</th>
<th>Major complications</th>
<th>Longest follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>[63]</td>
<td>Filler</td>
<td>Toronto, Canada</td>
<td>1998/1992-1997</td>
<td>16</td>
<td>9 months</td>
<td>TBM/TEF, TS, Cong cardiac</td>
<td>Palmaz</td>
<td>30</td>
<td>Trachea</td>
<td>1 death on stent removal, 1 full thickness bronchial erosion</td>
<td>56 months with tracheal stent, 25 months with bronchial stent</td>
</tr>
<tr>
<td>[72]</td>
<td>Furman</td>
<td>Chicago, USA</td>
<td>1999/1994-1997</td>
<td>6</td>
<td>10 months</td>
<td>TBM, Post-tracheoplasty Cong cardiac</td>
<td>Palmaz</td>
<td>12</td>
<td>Bronchus</td>
<td>1 stent related death</td>
<td>86 months with tracheal, 30 months with bronchial in cyanotic CHD</td>
</tr>
<tr>
<td>[73]</td>
<td>Geller</td>
<td>California, USA</td>
<td>2004</td>
<td>9</td>
<td>3 months-8 months</td>
<td>Cong heart Cong lung TEF/LTEF meningomyelocele</td>
<td>Palmaz</td>
<td>13</td>
<td>Bronchus</td>
<td>3 deaths from Tr hemorrhage, 1 stent related death</td>
<td>86 months with tracheal, 30 months with bronchial in cyanotic CHD</td>
</tr>
<tr>
<td>[44]</td>
<td>Santoro</td>
<td>Italy</td>
<td>1993</td>
<td>3</td>
<td>Vascular anomalies</td>
<td>Tracheoesophageal fistula with absent pul valv</td>
<td>Palmaz</td>
<td>3</td>
<td>Bronchus</td>
<td>2 airway related deaths</td>
<td>2 years</td>
</tr>
<tr>
<td>[22]</td>
<td>Nicolai</td>
<td>Germany</td>
<td>2001</td>
<td>7</td>
<td>4 months to 9 years</td>
<td>Structural airway pathology</td>
<td>13 Nitinol/6 others</td>
<td>19</td>
<td>Bronchus</td>
<td>3 deaths from generalised airway disease</td>
<td>50 months bronchial stent, 54 months</td>
</tr>
<tr>
<td>[70]</td>
<td>Kumar</td>
<td>London, UK</td>
<td>2002</td>
<td>5</td>
<td>Infants</td>
<td>Cong cardiac</td>
<td>Wallstent</td>
<td>10</td>
<td>Bronchus</td>
<td>1 cardiac death with patent airway</td>
<td>50 months bronchial stent, 54 months</td>
</tr>
</tbody>
</table>

TBM, tracheobronchomalacia; TEF, tracheoesophageal fistula; TS, tracheal stenosis following surgery; LTEF, laryngotraceo-esophageal fistula; TOF, Tetralogy of Fallot; Cong, congenital; CHD, congenital heart disease; Pul, pulmonary; Tr, tracheal.
obstruction, perforation, and suction catheter entrapment [76] leading to death. Grasping and repositioning manoeuvres carry risks. Rousseau reported a complication rate as high as 31%. The occasional case of aortobronchial fistula [69] highlights the potential hazard. It has rarely been used in the child. An anecdotal case of the use of a Gianturco stent in a 6-month-old child for tracheobronchomalacia, 3 months following the Lecompte maneuver was reported. The child was well at 32 months follow-up [61]. Hirameic addressed the effects of disease etiology on stent behaviour [62]. The malacic trachea has an increased luminal diameter and wider excursions, with complete expiratory collapse during the respiratory cycle and coughing, in comparison to a narrowed stenotic, fixed trachea or even the malacic bronchus. These can lead to metal fatigue from repetitive stress-strain movements, as shown by Freitag [77]. Due to the reported prohibitive risk, Hirameic has recommended against the use of Gianturco wire stent in malacic tracheal pathology. It appeared safer in the bronchus.

5.5. Silicone stent

There have been no significant reports of traditional silicone stents in the native airway of children. Jacob’s report included the use of silicone stents in 7 children less than 1.75 years following reconstructive surgery tracheal stenosis. There was a high mortality of 40% [78].

6. Evolving technology and future prospects

Bioreosorable films can serve as anatomic support [79]. Novel expandable support devices (stents), developed from these films are being studied. Poly-L-lactic acid and poly-glycolic acid (PLPG) resorbable stents may offer a potential solution to the problem of tracheomalacia. Advantages of this material include its strength, its versatile shaping characteristics, and its resorbability, which would preclude surgical removal and allow for infant airway growth. Nalwa in 2001 reported an encouraging pilot study showing the usefulness of PLPG as ‘Hagl-like’ stents for temporary external airway stenting of tracheomalacia in a porcine model [80]. Sewall achieved superior efficacy with the PLPG stent against the Palmaz stent, in a piglet model [81].

Another development in self-expandable airway stents is the PolyFlex (Willy Rüsch AG; Kernen, Germany), which consists of a polyester wire mesh embedded in a thin silicone covering. With spikes on its outer surface and not at the ends, this new technology hopes to address the issue of granulation tissue, maintain a favourable wall:lumen ratio as well as combat stent migration [82].

7. Conclusion

Infant airway is a complex issue with uncertain long-term implications. Personal experience is often limited. An accurate anatomic and functional diagnosis is essential. The best possible long-term outcome prediction must guide the choice. One should resist the temptation of short-sighted solutions. An initial conservative ventilatory airway support after correction of precipitating anomalies should always be attempted. Every surgical option must be explored to the fullest. Bronchoscopy must always guide the results.

Stenting certainly offers the least immediate procedural risk, especially in an already sick child. However, the lessons learnt in the evolution of clinical experience have made the characteristics expected of a stent very demanding. Rafanan and Metha from the Cleveland Clinic have outlined the ideal stent exhaustively [59]. The nitinol stent possesses more physiology-friendly properties.

Most authors consider metallic stents permanent once inserted. With the smaller pediatric stents, airway growth would eventually lead to size mismatch in adulthood. In cadaveric studies, the tracheal external transverse diameter is about 2 cm in adult males, and 1.5 cm in adult females. The lumen in live adults is, however, about 12 mm in transverse diameter; the discrepancy being accounted for by the relaxation of the smooth muscle at its posterior aspect, after death [83]. Smaller stents inserted in the child therefore must require periodic maintenance, change or even removal if they are to survive into adulthood. These have their attendant risks.

Though stents improve the short-term results, what will happen as the child reaches adulthood is speculative at present. More long-term clinical follow-up and quality of life data needs to be available to shed more light on the issue. The longest reported follow-up currently is about 7 years. Newer developments will hopefully bring safer stent alternatives. Till then, airway stents in children, should be used only as a last resort.

Tracheomalacia generally has a better outcome than bronchomalacia. Primary airway malacia also has a better long-term result than secondary airway involvement. The care of the pediatric airway is a challenging arena.

References


