European Journal of Cardio-Thoracic Surgery Advance Access published October 15, 2016

European Journal of Cardio-Thoracic Surgery 0 (2016) 1–7
doi:10.1093/ejcts/ezw299

ORIGINAL ARTICLE

Cite this article as: Serio P, Nenna R, Fainardi V, Grisotto L, Biggeri A, Leone R *et al.* Residual tracheobronchial malacia after surgery for vascular compression in children: treatment with stenting†. Eur J Cardiothorac Surg 2016; doi:10.1093/ejcts/ezw299.

Residual tracheobronchial malacia after surgery for vascular compression in children: treatment with stenting†

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Received 10 May 2016; received in revised form 25 July 2016; accepted 2 August 2016

Abstract

OBJECTIVES: Surgery for vascular anomalies can occasionally fail to relieve symptoms, especially when severe tracheobronchial malacia persists. We studied outcomes in children who underwent airway stenting for severe post-surgical airway malacia and tested known clinical and surgical prognostic factors.

METHODS: Among 257 children evaluated for tracheobronchial vascular compression, we reviewed the clinical charts for the 59 patients (23%) who underwent surgery. After surgery, children in whom severe malacia and respiratory symptoms persisted underwent airway stenting.

RESULTS: Among the 59 patients (boys: 58.1%, median age: 6.4 months, age range: 0.1–182.8 months), 79.7% had major comorbidities, 39% additional upper airway anomalies and 15.2% abnormal bronchial anatomy. Diagnostic imaging identified seven vascular anomalies: innominate artery compression 27.1%, left bronchial compression from the descending aorta and left pulmonary artery 20.3%, lesions associated with right aortic arch 22.0%, double aortic arch 13.6%, bovine arch type 10.2%, aberrant right subclavian artery 5.1% and pulmonary artery sling 1.7%. For severe residual malacia and severe persistent respiratory symptoms, 20 patients (33.9%) had silicone or metallic stents inserted. The most frequent indication for stenting was double aortic arch ($P = 0.02$ by chi-square test). A clinical prognostic factor for permanent stenting was left bronchial compression from the descending aorta and left pulmonary artery [odds ratio (OR): 14.667, 95% confidence interval (CI): 2.881–74.659], particularly if associated with congenital heart disease (OR: 30.00, 95% CI: 4.349–206.927). All silicone stents but one were successfully removed; metallic uncovered stents were patent and completely re-epithelialized.

CONCLUSIONS: When surgery leaves severe airway obstruction and respiratory symptoms unchanged, children with tracheobronchial malacia can safely undergo airway stenting.

Keywords: Vascular anomalies • Aortic arch anomalies • Stent • Airway obstruction • Children

INTRODUCTION

Vascular airway compression needing surgical correction is an especially challenging problem in infants and young children. Because the thoracic vasculature lies close to and supplies the trachea and bronchi, eventual anatomical variations especially in children can compress the airway, cause malacia and reduce the airway lumen. Although about 3% of the childhood population has vascular anomalies, these lesions rarely cause symptomatic compression, requiring a surgical approach. Despite successful surgical airway decompression residual tracheobronchial malacia, whose causes remain unclear[1, 2], can severely complicate the child's postoperative course by delaying recovery [3], hindering extubation [4], increasing the need for asthma medications [1] and sometimes requiring repeated hospitalization for severe persistent respiratory symptoms [5].

†Presented at the 24th International Congress of the European Respiratory Society, Munich, Germany, 6-10 September 2014.

Whether and if so how to treat children with residual tracheobronchial malacia and severe respiratory symptoms remains controversial. The currently preferred options include prolonged intubation or tracheostomy [6]. Surgical options include various procedures [7], such as slide tracheoplasty [8, 9] whenever the child's clinical conditions allow. More recently proposed therapeutic techniques are external tracheobronchial suspension within a ring-reinforced polytetrafluoroethylene prosthesis [10], anterior tracheal suspension [11] and airway stenting [12]. Although tracheobronchial stents are an attractive conservative option for restoring lumen patency in adults their use in children remains debatable given that the child's constantly growing body makes it difficult to predict the device's outcome in the airway [13]. Most paediatric series therefore consider stenting procedures as the last option for critically ill children [14, 15]. Having reliable evidence-based information from a centre specialized in interventional airway endoscopy would possibly help thoracic surgeons, paediatric surgeons, airway endoscopists and others who are less experienced in managing these rare post-surgical airway diseases in children.

This retrospective single-centre study conducted in our large-volume tertiary referral centre was designed to find out whether airway stenting could effectively and safely manage post-surgical persistent severe malacia manifesting with severe respiratory symptoms, in a series of children with tracheobronchial vascular compression. To do so we assessed the indications for stenting and complications during a long-term endoscopic follow-up (up to 8 years). As our secondary objective, we used chi-square test to evaluate whether five known baseline factors (type and severity of vascular anomalies, presence and type of comorbidities, age, sex and surgical variables) have prognostic value for severe persisting malacia requiring stenting.

MATERIALS AND METHODS

From a consecutive series of 257 young children with tracheobronchial vascular compression seen from 2006 to 2014 at the Respiratory Endoscopy Unit, A. Meyer Children's Hospital (Florence, Italy), we reviewed the clinical records for the 59 (23%) who underwent surgical correction. The diagnosis of vascular compression was clinically suspected and confirmed by flexible (Karl Storz®, Germany; Ø 2.5/3.7/5.2 mm) or rigid (Karl Storz®, Germany; Ø 3.0/3.5/4/4.5/5/5.5 mm) bronchoscopy and by chest computed tomographic angiography (CTA). The diagnostic work-up to rule out coexisting cardiac anomalies included echocardiography. We defined severe respiratory symptoms as dependence on ventilation, increased work of breathing or recurrent infections that required frequent prolonged hospitalizations. Surgical planning envisaged a customized approach (with or without cardiopulmonary bypass) for each patient according to the specific type of vascular anomalies (Supplementary Table S1.). After surgery, before and immediately after chest closure, the tracheal or bronchial lumen was visualized with a rigid bronchoscope.

According to the residual tracheal or bronchial obstruction and whether symptoms persisted after surgical correction, to assess the indications for stenting, patients were divided into three groups. Children with mildly reduced (<50%) or moderately reduced lumen (50–70%) and symptom improvement underwent clinical and endoscopic follow-up. Children with moderately reduced lumen (50–70%) and severe persistent respiratory symptoms but no vascular collision and in whom CTA confirmed sufficient space between the airway and the previously dissociated vessel, underwent airway stenting. Symptomatic patients with permanent vascular collision were scheduled for further surgical correction. Children who had severely reduced lumen (>70%) had an airway stent placed soon after surgery.

Stent placement

Silicone (Poliflex, Rusch® and Dumon, Novatech®) or covered Nitinol stents (Silmet®) were preferred for children with tracheomalacia and stainless steel metallic stents (Jostent, Abbott®) for those with bronchomalacia. Stents were placed in the operating theatre using a rigid bronchoscope with patients under general anaesthesia (sevoflurane inhalation ± intravenous propofol) and radiological guidance, as previously described [16].

Children who had silicone and covered Nitinol stents—inserted as non-permanent devices for subsequent removal—underwent bronchoscopic follow-up every 2, 4 or 6 months depending on whether granulation tissue tended to develop. To minimize the risk of granulation, all patients received inhaled corticosteroids. If granulation tissue formed causing airway obstruction, it was removed by laser. During follow-up, if rigid bronchoscopy showed that the stent was smaller in diameter than the trachea, the stent was removed and airway patency re-evaluated. If malacia remained unchanged a larger-diameter stent was inserted to allow for airway growth. If malacia improved the endoscopist attempted to remove the stent and the patient continued clinical follow-up.

Children who had stainless steel metallic stents—inserted and intended to remain as permanent devices—underwent bronchoscopic follow-up every 4–6 months up to 1 year and then at prefixed times to calibrate stent diameter finally according to airway growth.

Technical success was defined as effective stent insertion in the appropriate site in a single bronchoscopic session. Clinical success was considered as clinical improvement when the child was weaned off mechanical ventilation or respiratory symptoms dramatically improved or respiratory infections diminished after the procedure.

Statistical analysis

To seek possible prognostic factors related to persisting malacia requiring permanent stenting we used Pearson's chi-square test with odds ratios (OR) and 95% confidence intervals (CI) to test known baseline clinical and surgical variables including the type of vascular anomalies and presence and type of comorbidities [1]. Continuous variables are reported as means ± standard deviation (SD) or median and range. *P* values equal to or more than 0.05 were considered to indicate statistical significance, all tests were two-tailed. Data were analysed using the SPSS system (version 22.0; SPSS Inc., Chicago, Illinois, USA).

The study was approved by the research and institutional review board at Meyer Children Hospital, Florence, Italy (146/2014).

RESULTS

The hospital records for the 257 children showed that 59 consecutive children (boys: 58.1%, median age: 6.4 months, range: 0.1–182.8 months) underwent surgery to remove airway compression. In these 59 children, diagnostic imaging including

bronchoscopy distinguished seven vascular anomalies: innominate artery compression in 16 patients (27.1%), left bronchial compression from descending aorta and left pulmonary artery in 12 (20.3%), lesions associated with right aortic arch in 13 (22.0%), double aortic arch in 8 (13.6%), bovine arch type in 6 (10.2%), aberrant right sub-clavian artery in 3 (5.1%) and pulmonary artery sling in 1 (1.7%). Of the 59 children, 47 (79.7%) had major comorbidities (24 had oesophageal atresia with tracheoesophageal fistula; 18 congenital heart disease; 2 congenital lobar emphysema; 1 tracheal hypoplasia; 1 visceral angiomatosis and 1 Rubinstein Taybi syndrome). In addition, 23 (39%) patients had airway anomalies: 8 vocal cord anomalies, 7 sub-glottic stenosis, 5 choanal atresia, 1 laryngotracheal cleft, 1 global laryngomalacia, 1 laryngeal web and 9 (15.2%) bronchial anomalies. The reports for 52 (88.1%) patients mentioned previous surgical treatments for various reasons: 20 congenital heart disease (Supplementary Table S2), 23 oesophageal atresia or tracheoesophageal fistula, surgical corrections, 2 choanal dilation, 2 vocal cordotomy, 6 laryngotracheoplasty, 2 supraglottoplasty, 3 laryngotracheal cleft repair, 1 lobectomy and 9 Nissen fundoplication.

Surgery in 32 of the 59 children (54.2%) achieved mild lumen reduction with clinical improvement, in 12 (20.4%) residual moderate lumen reduction (5 asymptomatic and 7 symptomatic) and in 15 (25.4%) persistent severe lumen reduction (Supplementary Table S3). The 37 children in whom respiratory symptoms and luminal loss improved after surgery (32 with mild lumen reduction + 5 with moderate lumen reduction without symptoms) underwent a clinical follow-up.

Of the seven children with persistent moderate lumen reduction and severe respiratory symptoms four underwent aortopexy. Among them, despite undergoing the second operation, one of the patients required a metallic stent inserted into the left bronchus and one an hourglass-shaped silicon stent placed for 2 months in the trachea (Tables 1 and 2).

Of the remaining 18 children who underwent airway stenting, in 3 children stenting was indicated for persistent moderate lumen reductions and severe respiratory symptoms and in 15 for persistent severe lumen reduction.

In all 20 children who required stenting we obtained technical and clinical success immediately after tracheobronchial stenting. After stenting, all the six children were successfully weaned off mechanical ventilation and are in spontaneous breathing. The three patients who underwent tracheotomy were extubated.

Overall, 2 covered Nitinol and 26 silicone stents were placed in the trachea in 12 children and 10 ballon expandable metallic stents (Jostent, Abbott®) in the left mainstem bronchus in 8 patients. Of the 26 silicone stents used, 12 were cylinder-shaped (*n*: 5 Poliflex, and n: 7 Dumon®), 10 hourglass-shaped (Dumon®) and 4 Y-shaped (Dumon®).

In one patient with pulmonary sling and tracheal stenosis (Table 1, Patient 12), as the preferred surgical approach we corrected the sling and repositioned it close to the main pulmonary artery and simultaneously undertook a tracheoplasty. Tracheoplasty involved reconstructing a pericardial patch with cartilage strips. In our patient, the pericardium used for enlargement collapsed during extubation and we had to insert a tracheal stent.

The 11 patients whose silicone stents were removed after a median 4.5 months (0.5–24 months) completed follow-up after a median 2.9 years (range 0.5–7.7 years), except for two children who were lost to follow-up after 0.1 and 4.4 years. Patients with a stent still in place (1 silicon and 8 metallic stents) were followed for a median 5.3 years (range 3.2–8.1 years).

Among minor silicone stent-related complications, granulations (removed by laser) were found in 8/21 Dumon (38.1%) and 3/5 (60%) Poliflex stents. The only major silicone stent-related complication was a tracheal dislocation in one child. When the device was removed 6 months later, chest CTA and bronchoscopy showed new previously non-existing pulsatile compression by the pulmonary artery and descending aorta (Tables 1 and 2, Patient 4). The child underwent a second posterior aortopexy and a Y-stent (Dumon®) was inserted to treat the residual severe tracheal and left bronchial malacia.

Balloon expandable metallic stents required a mean (SD) 4.5 (3.7) calibrations or dilatations, are still in place in all eight children at a mean (SD) 4.9 (2.3) year-follow-up and are completely re-epithelialized. Ovalizations requiring dilations were found in 6/10 (60%) children with Jomed stents. One child with left bronchial compression from the descending aorta and the left pulmonary artery underwent a posterior aortopexy 26 months later to correct continuous stent ovalization related to the persistent compression by the descending aorta. Granulations (removed by laser) were found in the two children with Nitinol stents. None of the children who had metallic stenting experienced bronchial stenosis.

Data for the endoscopic follow-up showed that the 19 children who had an airway stent placed after surgery had corrected vascular compression are all alive and their respiratory symptoms have improved. Only one patient died of complications related to her visceral angiomatosis but stent placement reduced her respiratory symptoms (Tables 1 and 2).

When we analysed outcomes according to the type of vascular compression, 75% of children with double aortic arch, 50% with left bronchial compression from the descending aorta and the left pulmonary artery, 30.8% with right aortic arch anomalies, 16.7% with bovine arch type, 12.5% with innominate artery compression and the child with pulmonary artery sling required silicon or metallic stents ($P = 0.02$ by chi-square test). In 50% of children with left bronchial compression from the descending aorta and left pulmonary artery, 15.4% with right aortic arch anomalies and 6.3% with innominate artery compression, a stent is still in place ($P = 0.017$). Data for comorbidities showed that more children with than without coexisting congenital cardiac disease still have a stent in place (35.3% vs 7.1%; $P = 0.013$ by chi-square test).

In answer to our secondary objective, among vascular anomalies, chi-square test identified as a prognostic factor for persistent severe malacia requiring permanent stenting the presence of left bronchial compression from the descending aorta and left pulmonary artery (OR: 14.667, 95% CI: 2.881–74.659), particularly when combined with coexisting congenital heart disease (OR: 30.00, 95% CI: 4.349–206.927). No relationships were found between the other clinical and surgical variables tested including age, other major comorbidities and previous surgical treatment and airway stenting.

DISCUSSION

Our new findings in this retrospective study from a single referral centre specialized in interventional airway endoscopy suggest that airway stenting can successfully manage post-surgical persistent severe malacia in children with tracheobronchial vascular compression and severe respiratory symptoms. Our endoscopists do about 1200 airway procedures per year (about 200 new

Table 1: Demographic characteristics, coexisting diseases and type of vascular compression in children who required stent placement in trachea or in left mainstem bronchus

Pt	Age (y)	Sex	Coexisting diseases	Vascular compression
1[a]	0.2	Boy	–	Double aortic arch
2[a]	0.5	Boy	Congenital heart disease	Innominate artery compression
3[a]	0.1	Girl	–	Double aortic arch
4[a]	6.0	Girl	Tracheoesophageal fistula	Compression from descending aorta and left pulmonary artery
5[a]	1.6	Girl	–	Right aortic arch + ALSA
6[a]	0.4	Boy	Tracheoesophageal fistula	Bovine arch type
7[a]	0.3	Boy	–	Double aortic arch
8[a]	0.6	Boy	–	Double aortic arch
9[a]	0.1	Girl	Congenital heart disease	Double aortic arch
10[a]	2.4	Boy	Tracheoesophageal fistula	Innominate artery compression
11[a]	0.1	Boy	–	Double aortic arch
12[a]	1.4	Boy	Tracheal hypoplasia	Pulmonary sling
13[b]	0.9	Girl	Tracheoesophageal fistula + laryngotracheal cleft	Innominate artery compression
14[b]	0.5	Girl	Congenital heart disease	Compression from descending aorta and left pulmonary artery
15[b]	0.7	Boy	Laryngotracheal cleft	Right aortic arch + ALSA
16[b]	0.3	Girl	Congenital heart disease	Compression from descending aorta and left pulmonary artery
17[b]	0.1	Boy	Congenital heart disease	Compression from descending aorta and left pulmonary artery
18[b]	0.6	Boy	Congenital heart disease	Compression from descending aorta and left pulmonary artery
19[b]	0.4	Girl	Congenital heart disease	Right aortic arch + ALSA
20[b]	7.0	Girl	Congenital heart disease	Compression from descending aorta and left pulmonary artery

ALSA: anomalous left subclavian artery.
[a]Stent placement in trachea.
[b]Stent placement in left mainstem bronchus.

patients) often for children with severe airway malformations and comorbidities. Our study therefore provides previously unavailable evidence-based information from a large case series on these rare vascular anomalies. Although the considerable differences in age and co-morbidities make our findings hard to interpret they closely reflect this heterogeneous childhood population. Among the 59 children with vascular anomalies who underwent surgery to treat severe respiratory symptoms, one-third required stent placement for residual severe tracheobronchial malacia and unchanged severe respiratory symptoms. Few children who underwent vascular surgery (15.2%) in this series still have a stent in place. After a long follow-up (up to 8 years), endoscopy documented a dislocated or ovalized stent, each complication in one child. The vascular lesion associated with the highest risk of leaving a stent in place was left bronchial compression from the descending aorta and left pulmonary artery, particularly if combined with congenital heart disease.

No information yet explains why post-surgical persistent malacia arises. In our series, none of the reported potential causes of residual tracheobronchial malacia such as severity and duration of extrinsic compression and age at the time of surgical correction [1], were significantly associated with malacia. In 37% of the children who had their vascular anomalies surgically corrected in our series, the diagnostic work-up described severe malacia and severe persistent respiratory symptoms. Long-term follow-up studies after surgery for vascular compression report different degrees of airway obstruction, in variable rates ranging from 9 to 14% of children [17, 18]. In their retrospective study, Humphrey *et al.*, [1] reported chronic postoperative symptoms in 80% of patients who had the vascular compression before 6 months of age. Tracheobronchial malacia might persist when an insult damages the developing cartilage at a critical moment thus precluding future recovery [2].

When we assessed how we managed residual malacia and severe respiratory symptoms in the 59 children who underwent surgery for vascular collision, we found that one-third had a permanent or non-permanent airway stent placed. Growing evidence suggests airway stenting as a conservative treatment in children [19] to allow earlier extubation and relieve compressive symptoms [4, 20]. Nicolai [14] indicated this treatment only in extremely severe cases (long-lasting failure to wean from ventilation due to airway obstruction or 'dying spell' on ventilation due to dynamic over inflation). Our experience in treating these children suggests these major questions to ask before undertaking stenting: Does the patient have residual obstruction that surgery left unchanged? Does residual obstruction affect respiratory status or prolong the need for ventilation? and Which kind of stent should be used?

When we assessed the indications for stenting, a finding that seemingly justifies tracheobronchial stenting is that surgery for vascular airway compression left the severity of the preoperative endoscopic findings unchanged (>70% lumen reduction). In our series, patients with critical residual malacia underwent stenting soon after surgery. Although this timely choice might seem unnecessarily invasive, Vinograd *et al.*, [21] have already tried a prompt approach treating patients under the same anaesthesia with a prosthetic airway splinting procedure when aortic suspension failed to prevent tracheal collapse. Similarly, others in the

Table 2: Indications for stenting, stent characteristics and follow-up for children who required stent placement

Pt	Indication for stenting	Stent type	Stent removal (months)	Outcome	Follow-up (years)
1[a]	>70% lumen reduction	D 7 × 5×7	2	Resolution	Completed (0.5)
2[a]	Ventilator-dependent	D 7 × 20 D 7 × 5×7	1	Weaned Decannulated	Completed (6.1)
3[a]	Ventilator-dependent	D 7 × 30 D 10 × 30	1.2	Weaned	Completed (7.7)
4[a]	Persistent symptoms	D 12 × 10×12 N 12 × 30 Posterior aortopexy D 12-8-8 D 13-9-9	Patent	Resolution	(4.5)
5[a]	>70% lumen reduction	D 9 × 30 D 11 × 30 D 12 × 10×12	12	Resolution	Completed (1.2)
6[a]	>70% lumen reduction	P 8 × 20 P 8 × 30	4.5	Resolution	Completed (2.1)
7[a]	>70% lumen reduction	P 8 × 20 P 10 × 30 P 12 × 20	4.5	Resolution	Completed (4.5)
8[a]	>70% lumen reduction	D 8 × 6×8	4	Resolution	Lost (4.4)
9[a]	>70% lumen reduction	D 7 × 20 D 7 × 5×7	0.5	Weaned	Lost (0.1)
10[a]	>70% lumen reduction	D 11 × 9×11 D 12 × 10×12	1	Resolution	Completed (1.2)
11[a]	>70% lumen reduction	D 6 × 20 D 8 × 6×8 Anterior aortopexy	5	Weaned	Completed (2.9)
12[a]	>70% lumen reduction	D 5 × 7×5 N 8 × 20 D 7-5-5 D 8-5-5	24	Resolution	Completed (4.5)
13[b]	Persistent symptoms	Posterior aortopexy J 17-6/12	Patent	Decannulated	(4.6)
14[b]	>70% lumen reduction	J 12-6/12	Patent	Resolution	(5.5)
15[b]	>70% lumen reduction	J 12-6/12	Patent	Decannulate Weaned	(8.1)
16[b]	>70% lumen reduction	J 17-6/12	Patent	Resolution	(6.5)
17[b]	>70% lumen reduction	J 17-6/12	Patent	Weaned	(5.0)
18[b]	>70% lumen reduction	J 12-6/12 J 12-6/12	Patent	Resolution	(6.2)
19[b]	>70% lumen reduction	J 12-6/12	Patent	Death	(0.4)
20[b]	>70% lumen reduction	J 17-6/12 J 12-6/12 Posterior aortopexy	Patent	Resolution	(3.2)

Outcome resolution = clinical improvement: outcome improved when the child was weaned off mechanical ventilation or respiratory symptoms dramatically improved or respiratory infections decreased after the procedure.
D**:** Dumon stent; N: Nitinol stent; P: Palmaz; J: Jomed stent.
[a]Stent placement in trachea.
[b]Stent placement in left mainstem bronchus.

same operative setting treated severe residual tracheobronchial malacia within a ring-reinforced polytetrafluoroethylene prosthesis and anterior tracheal suspension [10, 11]. We used tracheoplasty to treat severe malacia after surgery for pulmonary sling in one child, as already described [22]. Even though stenting can be a hazardous undertaking if done by inexperienced staff, a centre with wide expertise in using these devices can undertake the procedure easily and safely [16].

The indication for stenting in patients with moderate residual airway malacia and persistent respiratory symptoms are less clear. In our patients, when surgery failed to resolve the initial symptoms, stenting dramatically and rapidly improved their respiratory status and restored spontaneous breathing. Others have already successfully used tracheal stents in patients with persistent and severe tracheomalacia to allow earlier extubation after surgery and relieve compressive symptoms [15, 23].

An equally important choice in planning stenting is the type of stent to use. Metallic balloon expandable stents are a major cause for concern given that once inserted they must be considered as a permanent device [12, 14]. Despite concern, they have great advantages. Apart from ensuring accurate placement and precise luminal diameter, they leave mucous clearance unchanged and rarely migrate [12, 14, 16, 19]. Several reports describe severe complications such as excessive granulation tissue formation, arteriobronchial fistulas, airway perforation or bronchial stenosis [13, 15, 21]. In our series, with a long-term follow-up, all the metallic uncovered stents are still in place, completely re-epithelialized and have led to few minor complications and persistent ovalization in

only one child. We attribute these promising results to the fact that rather than considering stenting as an alternative to aortopexy, we reserve stent placement to patients without CTA-documented residual vascular compression, as already described [24]. Equally important, we closely follow these children with scheduled endoscopic evaluations, that considered the child's growth.

For use in children with tracheomalacia to facilitate our patients' spontaneous improvement, we preferred silicon or Nitinol-covered stents both easily removable devices. All the children were eventually successfully weaned off mechanical ventilation, are in spontaneous breathing and had their tracheal stents successfully removed after a median 4.5 months. Hence silicon stents and Nitinol-covered stents function as non-invasive, non-permanent devices that can temporarily overcome the child's symptoms, are relatively easy to place and remove and are well tolerated. A disadvantage is that they can become obstructed by mucus and frequently migrate [18] as happened in one child in our series. Two studies conducted in recent years described the use of biodegradable stents also in children with residual tracheobronchial obstruction after surgery to remove vascular compression and showed promising results [25].

When we analysed the hospital reports to understand the specific type of vascular compression that required stenting, we found that the most frequent indication was double aortic arch. In children with a double aortic arch, dividing the anterior arch sometimes fails to relieve the obstruction [26] and leaves the underlying tracheomalacia untreated. As many as 50% of these children appear to have residual symptoms secondary to malacia after surgical treatment [27]. In two children in our series residual symptoms secondary to post-surgical malacia required an aortopexy, as others have underlined [26, 27]. In six children in our series who had a double aortic arch, tracheal stents rapidly resolved the respiratory symptoms and the improvement persisted.

In our study, extending current reports, the prognostic factor most closely related to persistent severe malacia requiring permanent stenting was left bronchial compression from the descending aorta and left pulmonary artery, particularly if combined with congenital heart disease. The high percentage of residual left bronchial malacia after surgery in this type of vascular anomaly accords with evidence that aortopexy can leave bronchial malacia unchanged [28]. When the surgical option fails in these children some recommend stenting [23, 29]. Managing residual bronchial obstruction in these severely ill patients, particularly ones with coexisting congenital heart disease can be difficult insofar as children with impaired cardiovascular physiology poorly tolerate hypoventilated or atelectatic lung segments [30].

This study has some limitations. Most patients described in this series were in severe clinical conditions and had severe comorbidities and airway anomalies. Both factors could have acted as confounding bias for the initial and post-surgical symptoms. Another limitation is that we evaluated symptomatic improvement after stenting on clinical criteria alone without lung function tests. Nor did we enrol a control group without airway stenting to compare the clinical outcome in patients with residual malacia or persistent respiratory symptoms or both, because for ethical reasons we were unable to abstain from treating children with severe respiratory symptoms and severe malacia.

CONCLUSION

When surgical correction leaves airway obstruction causing chronic severe respiratory symptoms definitively unresolved, airway stenting done in specialized centres provides a safe alternative option a longer follow-up would be necessary to assess the effects of these devices in the growing child. Future research might seek further information on risk factors for respiratory symptoms in children who have undergone surgery to correct vascular anomalies.

SUPPLEMENTARYMATERIAL

Supplementary material is available at *EJCTS* online.

ACKNOWLEDGEMENT

The authors thank Alice M Crossman for help with English style.

Conflict of interest: None declared.

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