Reoperation to correct unsuccessful vascular ring and vascular decompression surgery

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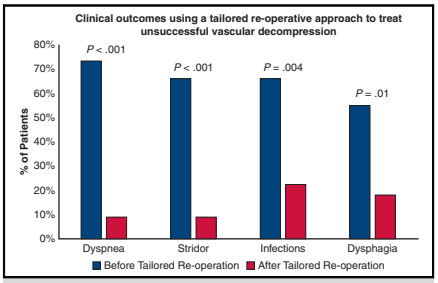
ABSTRACT

Objective: Although most children do well after operations to relieve vascular compression of the esophagus and airway, many will have persistent/recurrent symptoms. We review our surgical experience using a customized approach to correct various etiologies of failure after vascular ring/decompression surgery.

Methods: Our institutional database identified children who underwent reoperation for persistent/recurrent symptoms after vascular ring or aberrant arterial decompression surgery between January 2014 and December 2019. Charts were reviewed for operative approaches and clinical data. Findings were analyzed by Fisher exact test for comparison between groups.

Results: Twenty-seven children required reoperative surgery. Detailed preoperative workup identified 5 etiologies of failure for a customized approach. Residual scarring was corrected by lysis and rotational esophagoplasty (n = 23/27); fibrotic bands re-creating a ring were divided (n = 11); ongoing vascular compression was addressed by descending aortopexy (n = 19), aberrant subclavian division (n = 7), aortic uncrossing procedure (n = 4), and Kommerell resection (n = 8); anterior aortopexy (n = 6) and anterior tracheopexy (n = 9) corrected cartilage malformation; and tracheobronchomalacia was addressed with posterior airway pexy (n = 26). At available short-term follow-up (median 1 year), 21 of 22 patients (95%) had symptom improvement, and on bronchoscopy, the average number of airway sections with severe tracheobronchomalacia decreased from 2.8 ± 1.7 to 0.5 ± 0.9 ($P < .001$).

Conclusions: Persistent/recurrent symptoms after release of vascular compression are frequently caused by 5 different etiologies. A multidisciplinary strategy for workup and a customized operative approach can effectively treat these cases and may suggest opportunity at the index surgery to prevent reoperation and achieve optimal outcomes. (J Thorac Cardiovasc Surg 2022;164:199-207)



Clinical improvement in children after reoperation for unsuccessful vascular decompression.

CENTRAL MESSAGE

For children still symptomatic after vascular ring/decompression operations, a customized operative approach can be tailored to the patient based on preoperative workup for optimal treatment.

PERSPECTIVE

Although operations to relieve vascular compression of the airway/esophagus in children usually have good results, at least 10% will continue have obstructive symptoms frequently with unclear etiology, which poses a reoperative challenge. For these patients, we use a tailored approach to identify and correct the underlying etiology of failure for optimal outcomes.

See Commentaries on pages 208 and 209.

Vascular rings and related vascular compression syndromes are caused by abnormal embryonic development of the great vessels and their immediate branches, resulting in esophageal and airway compression.[1,2] Surgical correction, first performed by Gross[3] in 1945, involves releasing the compressed mediastinal structures usually via vascular or ligamentum division. Results from this approach have been acceptable, with 70% to 86% of children experiencing full resolution of symptoms.[4-10]

However, approximately 5% to 10% of patients need a repeat operation for persistent or recurrent symptoms.[9-11] From our experience, these treatment failures are caused by some combination of 5 different etiologies: scar tissue causing ongoing mediastinal constriction, contractile fibrotic bands re-creating a vascular ring, ongoing vascular compression, cartilaginous airway malformation, and tracheobronchomalacia (TBM). Management of underlying TBM in particular can be

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Institutional Review Board P00004344; approval August 11, 2020.

Read at the 101st Annual Meeting of The American Association for Thoracic Surgery: A Virtual Learning Experience, April 30-May 2, 2021.

Received for publication April 29, 2021; revisions received July 26, 2021; accepted for publication Aug 13, 2021; available ahead of print Nov 11, 2021.

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0022-5223/$36.00

https://doi.org/10.1016/j.jtcvs.2021.08.089

CONG

Abbreviations and Acronyms
CT = computed tomography
L1 = left mainstem bronchus
R1 = right mainstem bronchus
TBM = tracheobronchomalacia

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challenging and, when seen preoperatively with vascular compression, is associated with increased rates of treatment failure and reoperation.[7-10,12-15]

By recognizing what etiology of failure is driving symptoms, a customized and targeted treatment plan can be created for the patient. We present the reoperative outcomes using this approach in children with persistent or recurrent symptoms after the initial vascular release operation.

MATERIALS AND METHODS

We conducted a retrospective review of all children who underwent surgery for vascular ring or innominate artery/aberrant right subclavian compression syndromes at our institution between January 2014 and December 2019, identifying those who were undergoing reoperation (Institutional Review Board P00004344). These redo cases were all referred to and followed by our Esophageal and Airway Treatment Center, which is a multidisciplinary group composed of pediatric cardiovascular and general surgeons, pediatric otolaryngology, pediatric gastroenterology, and pediatric pulmonology teams.

Patient charts were reviewed for original vascular anatomy, index ring operation, and clinical data at the time of referral and the most recent available postoperative follow-up. Clinical data included any preoperative evaluation, patient demographics, and assessment of 4 primary symptoms: dyspnea on exertion, noisy breathing/barking cough, recurrent/persistent respiratory infections, and dysphagia. Heights and weights were converted to z-scores.[16,17] Need for positive pressure ventilatory support, supplemental oxygen, cyanotic episodes, or brief resolved unexplained events in infants were noted. Length of hospitalization and any perioperative complications were recorded.

Operative reports were reviewed for surgical details along with bronchoscopic description of the airway before repair, immediately after repair, and at most recent bronchoscopic follow-up. TBM was classified by dynamic airway collapse as mild (50%-65% collapse), moderate (66%-80%), or severe (>80%) in the upper trachea (T1), mid trachea (T2), lower trachea (T3), carina, right mainstem bronchus (R1), or left mainstem bronchus (L1).[18,19] Presence of thick secretions or cobblestoning, indicative of obstruction and inflammation, was also noted.[20]

Preoperative workup is a point of emphasis and guided by the multidisciplinary team: dynamic bronchoscopy is almost always performed, with the patient coughing to assess degree of TBM, airway malformation, and presence of external airway compression; computed tomography (CT) angiography helps assess ongoing vascular or scar tissue–related compression; and esophagram or upper endoscopy identifies presence of esophageal stricture/narrowing contributing to dysphagia.

Surgical approach varied depending on the patient's index operation and identified etiology(ies) of failure, as determined by preoperative workup. Incision was usually a redo left thoracotomy, but at times a sternotomy was required if any anterior airway or great vessel work was anticipated. Extensive mediastinal lysis of adhesions was performed on all patients, and in many the esophagus was dissected completely free with a rotational esophagoplasty: The esophagus is freed from any compressive scar tissue and rotated to the right of the spine into the pleural space, exposing the anterior spine for subsequent tracheopexy (see below). Contractile fibrotic bands causing a reformed vascular ring were divided, and the descending aorta was pexied posteriorly to prevent reformation of the band. Persistent vascular compression was treated via division of an aberrant subclavian artery (usually with reimplantation), resection of large Kommerell diverticulum, descending aortopexy for left mainstem relief,[21] or aortic uncrossing procedure for right-arch mediated compression.[22] Permanent cartilaginous airway deformation was corrected with direct anterior tracheopexy by passing pledgeted polypropylene suture through the cartilage ring and pexying to the sternum, using pericardium as a strut (by placing the strip of pericardium in between the trachea and spine, pexy conformation is smoothed and over-tension prevented) [14]; this was preceded by anterior aortopexy if there was ongoing vascular airway intrusion. For this anterior work, a sternotomy with thymectomy was usually performed for optimal exposure and, in severe cases, pexies were deferred and an external splint was instead used for airway stabilization.[23] Posterior membrane intrusion due to regional airway weakness (TBM) was addressed with posterior tracheobronchopexy, again using pledgeted sutures combined with a pericardial strut to secure the posterior membrane to the anterior spinal ligament.[14,15] Rotational esophagoplasty was almost always performed in these standard situations for appropriate exposure. Airway work was guided intraoperatively by flexible bronchoscopy for targeted therapy, with later patients in the series having negative pressure applied to the airways (up to −50 cm H20) near the end of the operation to test the effectiveness of the repair and evaluate for areas of unaddressed malacia (effectively simulating a cough during dynamic bronchoscopy under general anesthesia).[24]

Follow-up consisted both of clinical data at their most recent visit along with findings on dynamic bronchoscopy, generally performed as part of routine surveillance.

Statistical Analysis

To assess symptom and TBM resolution, Fisher exact test was used to compare preoperative and postoperative categorical data. Student *t* test was used to compare continuous data.

RESULTS

Of the 255 patients undergoing vascular decompression operations at our institution over the 6-year period, 27 had been referred for persistent or recurrent symptoms. These referrals were a combination of internal (n = 7), national (n = 18), and international (n = 2), giving an institutional reoperation rate of 3.0% (n = 7/235).

Median age at index operation was 4.5 months (2 days to 15 years), with 6 children undergoing multiple operations prior to referral. Eighteen were male. Eleven patients (41%) had significant comorbidities, most frequently syndromic/genetic (n = 9); another 5 patients had a prior esophageal atresia or tracheoesophageal fistula repair (2 with VACTERL association), and 1 patient had a connective tissue disorder (Ehlers-Danlos).

The index operation was usually division of a double aortic arch (n = 12) or ligamentum/persistent ductus

TABLE 1. Original anatomy, index operation, and reoperation

Anatomy	Index operation	No.	Reoperation	No.
Double aortic arch n = 12			**Rotational esophagoplasty**	**11**
	Arch division*	**12**	**Division fibrous band**	**5 (+1 division L subclavian†)**
	D/LA division*	**4**	**Aortic uncrossing**	**3**
	Anterior aortopexy*	**2**	**Anterior aortopexy**	**5**
	*		**Descending aortopexy**	**8**
	*		Anterior tracheopexy	5
			Posterior tracheopexy	11 (+1 tracheal splint)
			R1 Pexy	7
			L1 Pexy	7
	Reoperative approach:		*Sternotomy*	*7 (3 CPB)*
			Left thoracotomy	*3*
			Right thoracotomy	*2*
R arch with aberrant L subclavian n = 10			**Rotational esophagoplasty**	**8**
	D/LA division	**10**	**Division fibrous band**	**5**
	Division left subclavian	**3**	**Division left subclavian**	**6†**
			Anterior aortopexy	**1**
	Descending aortopexy	**2**	**Descending aortopexy**	**8**
			Anterior tracheopexy	3
	Posterior tracheopexy	1	Posterior tracheopexy	8 (+1 tracheal diverticulectomy)
			R1 Pexy	6
			L1 Pexy	4
	Reoperative approach		Sternotomy	3 (2 CPB)
			Left thoracotomy	7
R arch with circumflex aorta and mirror imaging n = 2			**Rotational esophagoplasty**	**2**
	D/LA division	**2**	**Division fibrous band**	**1**
			Aortic uncrossing	**1**
	‡		**Descending aortopexy**	**1**
			Anterior tracheopexy	1
			Posterior tracheopexy	2 (+1 tracheal resection and +1 diverticulectomy)
			R1 Pexy	1
			L1 Pexy	1
	Reoperative approach		*Sternotomy*	*1 (1 CPB)*
			Left thoracotomy	*1*
Innominate compression n = 2			**Rotational esophagoplasty**	**1**
	Anterior aortopexy	**2**	**Anterior aortopexy**	**0**
			Descending aortopexy	**1**
			Posterior tracheopexy	2 (+1 tracheal diverticulectomy)
			R1 Pexy	1
			L1 Pexy	1
	Reoperative approach		*Right thoracotomy*	*2*
Aberrant R subclavian n = 1			**Rotational esophagoplasty**	**1**
	Division R subclavian	**1**	**Descending aortopexy**	**1**
			Posterior tracheopexy	1
			L1 pexy	1
	Reoperative approach		*Left thoracotomy*	*1*
Diverticulum of Kommerell n = 13	**Kommerell resection**	**1**	**Kommerell Resection**	**8 (+1 pexy)**

Bold indicates procedures for external compression; italics indicates airway procedures. *D/LA*, Ductus/ligamentum arteriosum; *R1*, right mainstem; *L1*, left mainstem; *CPB*, cardiopulmonary bypass. *Five children with double aortic arch underwent subsequent operations before referral: ligamentum division with anterior aortopexy; aortic uncrossing procedure; descending aortopexy; resection of arch remnant/scar tissue and anterior aortopexy; and descending aortopexy with tracheo- and bronchopexies. †Five of 7 children who underwent division of an aberrant left subclavian had it reimplanted into the carotid. ‡One patient had a subsequent descending aortopexy.

(n = 15) with occasional resection of a Kommerell diverticulum (n = 2/13) and tracheopexy (n = 1). Table 1 shows the operative details by original anatomy.

On referral to our center, children were a median of 2 years from their index operation (range, 23 days to 17 years). Ten children had no symptom resolution after their original operation, and the other 17 children had a period of improvement, but then their symptoms recurred. The most frequent symptom was dyspnea (n = 20, 74%) (Table 2). Nine children required supplemental oxygen or positive pressure with 4 tracheostomies, and 9 children had experienced a recent brief resolved unexplained event or cyanotic episode.

All children underwent a dynamic bronchoscopy to evaluate for TBM and airway deformation before surgery. All children also had dynamic CT angiogram to identify evidence of ongoing vascular compression (seen in 15), and if dysphagia was present, an upper endoscopy or fluoroscopic esophagram was performed.

Twenty-six patients had moderate to severe TBM (the other patient had no airway symptoms and had re-presented with dysphagia alone) (Table 3). Of the 6 evaluated airway sections (T1-3, carina, L1, R1), there were an average of 2.8 (±1.7) sections with severe disease per patient. Twelve patients (44%) also had a laryngeal cleft.

Reoperation was performed at a median of 4.5 years old (2 months to 17 years), with an average weight and height (normalized to z-score) at that time of −0.2 ± 1.5 and −0.6 ± 1.6, respectively; 6 patients weighed less than 10 kg. Operations were designed to address each patient's individual etiology of failure, correcting causes of extrinsic constriction (scar tissue, fibrotic band, vascular compression) and intrinsic airway disease (airway deformation, TBM) (Table 1). Children were discharged a median of 7 days after surgery (3 days to 2 months), and 9 patients experienced complications: temporary laryngeal nerve dysfunction in 5 and chyle leak in 2, both of which resolved without intervention; postpericardiotomy syndrome in 1; and 1 patient had an acute subclavian vein thrombus requiring takeback and loosening of an anterior aortopexy. Four children required an additional operation for an anterior tracheopexy (n = 3), anterior aortopexy (n = 2), or R1 pexy (n = 2).

Clinical follow-up was available on 22 children; unfortunately, 1 child died after a hypoxic arrest when her tracheostomy tube dislodged 2 months after repeat operation for anterior tracheopexy, and 4 other patients, all national referrals, have not been seen in clinic. At a median of 1.1 years after surgery (3 months to 3 years), there was significant improvement in symptoms (Table 2 and Figure 1), most markedly dyspnea going from 74% to just 9%, although 7 of 22 patients were still having mild respiratory symptoms (4 with dyspnea/stridor, 3 with recurrent infections, and 2 with both). Dysphagia had significantly improved as well, although many (n = 15) required endoscopic balloon dilation for persistent mild to moderate stenosis. Of the 4 children with ongoing dysphagia, all required balloon dilations, 2 have required laryngeal cleft repairs, and 1 had a complex esophageal injury during atresia repair at an outside institution.

TABLE 2. Pooled symptoms pre- and post-redo operation

Symptoms	Pre-reoperative (n = 27)	Post-reoperative* (n = 22)	*P* value
Dyspnea	20 (74%)	**2 (9%)**	**<.001**
Noisy breathing/barking cough	18 (67%)	**2 (9%)**	**<.001**
Recurrent/persistent infections	18 (67%)	**5 (23%)**	**.004**
Dysphagia	15 (55%)	**4 (18%)**	**.01**
Height <5th percentile	8 (30%)	2 (9%)	.15
Weight <5th percentile	4 (15%)	1 (5%)	.36

Statistically significant differences in bold. *After final operation.

TABLE 3. Pre-reoperative versus post-reoperative airway malacia

Bronchoscopic findings	Pre-reoperative (n = 27)	Post-reoperative* (n = 20)	*P* value
T1 (any malacia)	2 (7%)	1 (5%)	1.0
Mild	1	1	1.0
Moderate	0	0	1.0
Severe	1	0	1.0
T2 (any malacia)	11 (41%)	**2 (10%)**	**.02**
Mild	0	1	.4
Moderate	1	1	1.0
Severe	10	**0**	**.003**
T3 (any malacia)	26 (96%)	**5 (25%)**	**<.001**
Mild	2	2	1.0
Moderate	4	1	.4
Severe	20	**2**	**<.001**
Carina (any malacia)	25 (93%)	**5 (25%)**	**<.001**
Mild	4	3	1.0
Moderate	2	1	1.0
Severe	19	**1**	**<.001**
L1 (any malacia)	19 (70%)	**6 (30%)**	**.009**
Mild	5	1	.2
Moderate	3	4	.4
Severe	11	**2**	**.02**
R1 (any malacia)	22 (81%)	**6 (30%)**	**<.001**
Mild	4	0	.1
Moderate	4	1	.4
Severe	14	5	.08
Secretions/cobblestoning	21 (78%)	**5 (25%)**	**<.001**

Statistically significant differences in bold. *T1-3*, Upper, middle, and lower trachea; *L1*, left mainstem; *R1*, right mainstem. *After final operation.

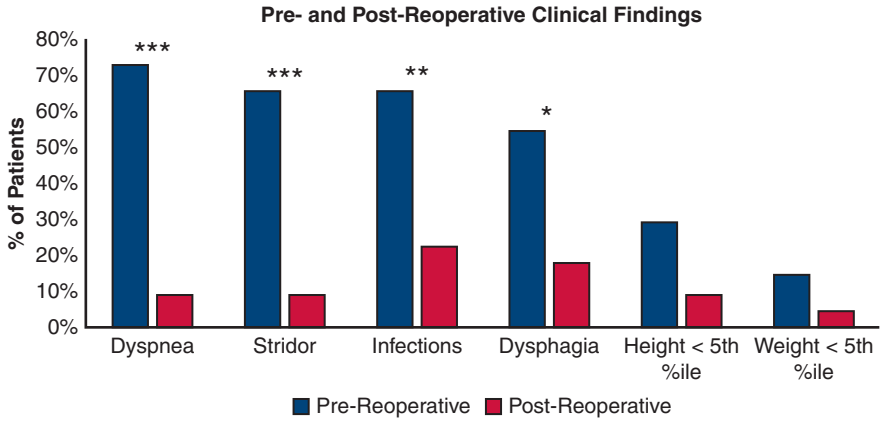

FIGURE 1. Clinical findings pre- and postsurgical reintervention. ***$P < .001$, **$P < .01$, *$P < .05$. *%ile*, Percentile.

Overall, 21 of 22 patients had subjective improvement in at least 1 symptom, the outlier being the aforementioned child with esophageal atresia who had iatrogenic tracheal and esophageal injuries (whose treatment is ongoing). No children were having cyanotic episodes or requiring home oxygen, and of the tracheostomies, 1 is followed at their home institution, whereas the other 2 have required laryngeal reconstruction and have been decannulated or are tolerating their capping trials.

Bronchoscopic findings improved in all children evaluated (n = 20, Table 3 and Figure 2). Although severe malacia persisted in 7 children, the overall disease burden significantly improved with an average of just 0.5 (±0.9) airway sections affected with severe disease per patient ($P < .001$ compared with preoperative). Severe R1 malacia proved most difficult to treat, with only 9 severe cases (64%) resolving in those evaluated with follow-up bronchoscopy. However, all children with unresolved right bronchomalacia had presented with recurrent respiratory infections (n = 4) or had a connective tissue disorder (n = 1).

For the 7 children with ongoing respiratory symptoms after reoperative surgery, all but 1 had a single persistent area of severe malacia: T3 in 1, L1 in 1, and R1 in 4 (the other, who had resolved stridor but ongoing respiratory infections, has a normal bronchoscopy). There was no relationship between persistent symptoms and diverticulum of Kommerell at birth, age at index operation, history of laryngeal cleft, or age at referral ($P = .5$ to 1.0). However, there was a significant relationship between severe preoperative bronchomalacia and ongoing symptoms at follow-up (7/14 patients with severe right or left bronchomalacia on presentation had persistent symptoms vs 0/8 without, $P = .02$); although there was a trend toward higher rates of ongoing symptoms with right arch anatomy, this did not reach significance (4/8 with right arch vs 3/14 with left arch, $P = .3$).

DISCUSSION

Unresolved or persistent symptoms after surgery for vascular ring or arterial compression pose a clinical dilemma that can be difficult to manage. Symptoms such as dyspnea, stridor, or dysphagia suggest residual obstruction, but the exact etiology is often difficult to parse by the lay clinician, and the child is left in a diagnostic quagmire. However, our results show that by advocating for specialized follow-up and using multidisciplinary workup with a combination of bronchoscopy, esophagoscopy, and CT angiography, the etiology of failure can be determined, allowing treatment to be tailored to the individual needs of the patient for optimal outcome. By identifying the contribution of both external compression (residual scar tissue limiting mediastinal growth, contractile band[s] reforming a vascular ring, residual arterial compression) and intrinsic airway disease (cartilage deformation and TBM), the operation can be planned to directly correct each of these underlying etiologies of failure as indicated.

Management of TBM in children is a debated topic. Although there is the opinion that malacia resolves and the airway will "firm up" as the child grows,[4,25,26] a large number of children have persistent respiratory symptoms and require surgical intervention. Children with vascular ring/arterial compression are particularly vulnerable to TBM-related complications due to the multifactorial nature of their airway weakness: Compression during organogenesis causes both intrinsic airway maldevelopment and structural deformation, which further contributes to a widened and flaccid posterior membrane.[27] In fact, 26 of 27 of our patients were found to have moderate to significant TBM as an etiology of failure requiring intervention.

Historically, the most common approach to treat both TBM and airway cartilage deformation in children after vascular ring division is an anterior aortopexy; however,

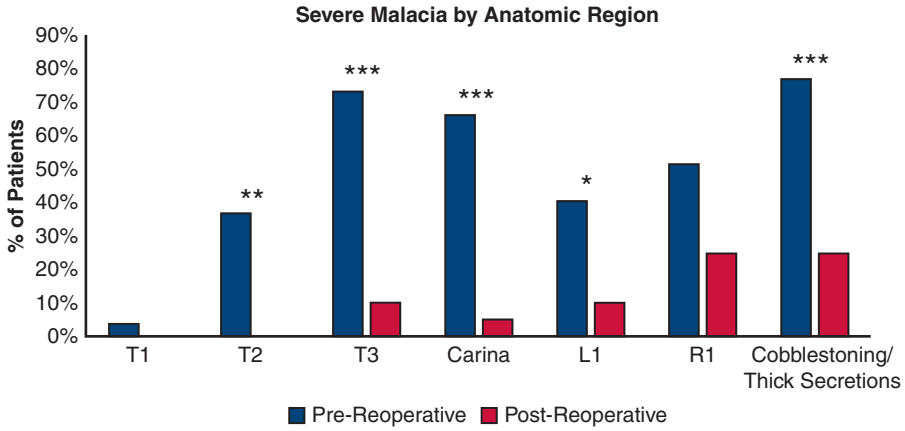

FIGURE 2. Bronchoscopic findings pre- and postsurgical reintervention for severe malacia (>80% collapse on dynamic bronchoscopy). ***$P < .001$, **$P < .01$, *$P < .05$. *T1-3*, Upper, middle, and lower trachea; *L1*, left mainstem; *R1*, right mainstem.

these results are often unsatisfying, and in smaller series with known preoperative TBM, symptoms persist in 32% to 50%.[8,13,28] These rates are higher compared with aortopexy for TBM associated with tracheoesophageal disease, where 0% to 15% have residual symptoms.[8,13] This is perhaps due to differences in the underlying disease etiology as discussed earlier, with tracheoesophageal disease TBM being primarily intrinsic, whereas vascular compression results in both intrinsic and structural airways defects making it less suitable to aortopexy.[14] Furthermore, children requiring a reoperation for TBM can often expect to have increasingly weakened airways from persistent infections and inflammation, which further complicates treatment.[25]

We have demonstrated that a tailored approach to address both external compression and intrinsic airway disease, guided by multidisciplinary workup, can effectively treat these complicated children who are still symptomatic after vascular decompression operation (Figures 1 and 2).

Several important surgical strategies were used. First, for TBM the floppy and intrusive posterior membrane was pexied to the spine's anterior longitudinal ligament often using pericardial struts (done in nearly all patients, n = 26/27). This prevents airway collapse from membrane intrusion during cough and Valsalva, which is not addressed by an anterior aortopexy.[14,15] Anterior aortopexy was still occasionally performed, but only in an effort to relieve external tracheobronchial compression. This anterior compression frequently caused deformation of the cartilaginous rings, which contributed to airway coaptation. Therefore, anterior tracheopexy or bronchopexy was performed to support the cartilage and encourage cartilage growth in a more open conformation.[18]

Residual external compression on the left mainstem/L1 was often caused by posterior impingement from the descending aorta; by pexying the descending aorta as posteriorly as possible, compression on L1 can be relieved.[21] Likewise, a circumflex right aortic arch can put pressure on the mid-to-distal trachea, which poses surgical challenges. By performing an uncrossing procedure or posterior descending aortopexy, the airway compression can be effectively treated.[22] Notably, 4 of 7 of the children with ongoing respiratory symptom had a right arch, and none underwent an uncrossing procedure (the others who underwent uncrossing were asymptomatic); this perhaps suggests a greater role for such procedures.

A significant risk of damage to the recurrent laryngeal nerve exists when operating the aortic arch, with paresis reported in approximately 22% of cases in adults.[29] Reoperation further increases this risk of injury due to scar formation and distortion of the nerve's expected path. Therefore, to avoid iatrogenic injury, intraoperative recurrent laryngeal nerve monitoring is now routinely used at our institution to identify and protect the nerve.[30] This is done by sensing vocalis activity, using electrodes placed onto the endotracheal tube (larger patients) or directly onto the vocalis muscle (any size patient); dissection through scar tissue is done only after carefully stimulating the area and monitoring for vocalis activation (or lack thereof), confirming the recurrent nerve is not present. With this method, the rate of laryngeal nerve dysfunction/vocal cord paresis in this series was 5 of 27 (19%) despite the hostile operative field, and injury only involved temporary stunning that resolved within weeks.

Finally, performing all maneuvers with bronchoscopic visualization is essential to target the appropriate region of the airway and avoid full-thickness intraluminal passage of the sutures. Through relief of any external compression and direct tracheobronchopexy, the airway can be made anatomically normal by the end of the case. Furthermore, a dynamic bronchoscopy can be recreated under general anesthesia by applying negative pressure to the airway,

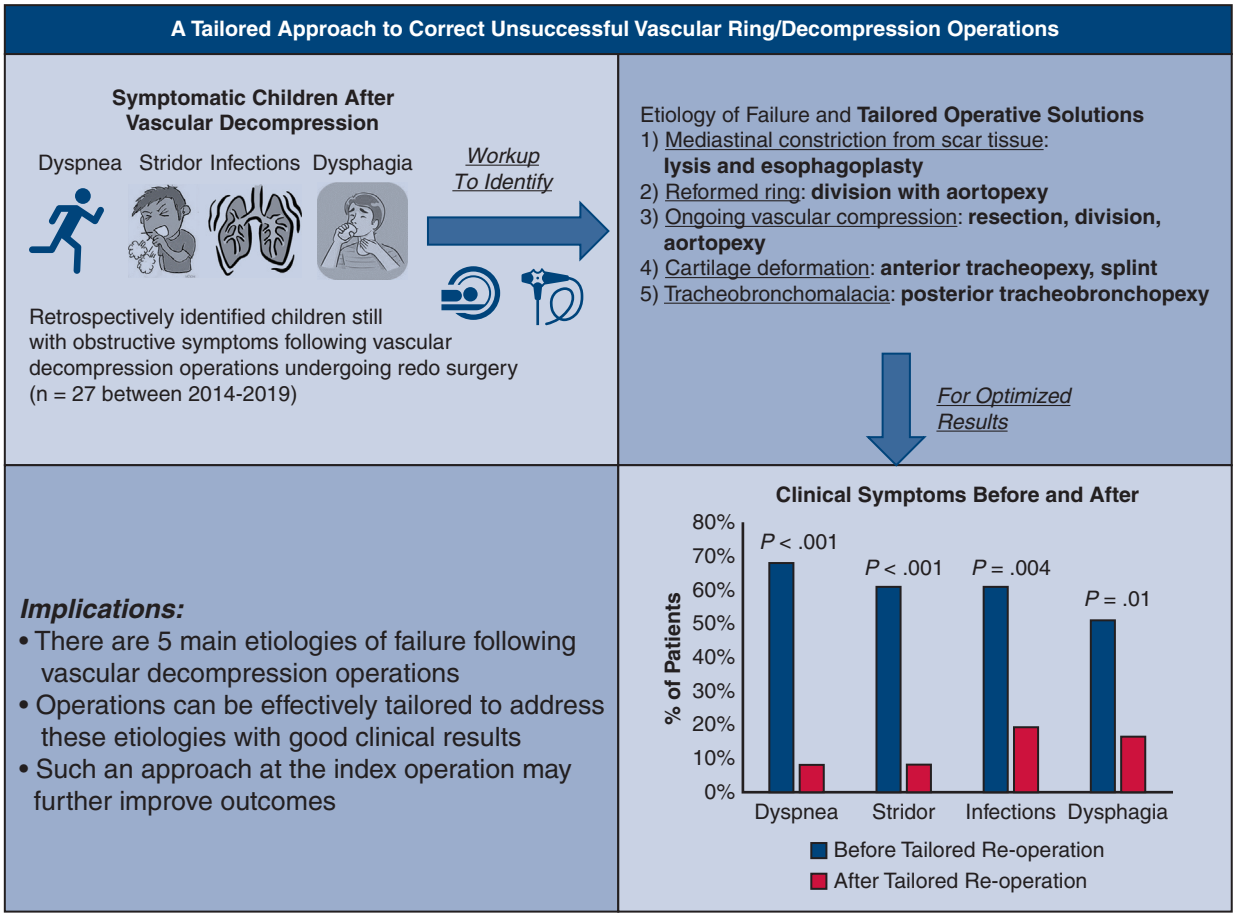

FIGURE 3. The methods (*top left*), the operative strategy based on etiology of failure (*upper right*), and the clinical results of this approach (*bottom right*). The future implications of this research are suggested (*bottom left*).

testing the durability of the repair and identifying any areas of residual disease.[24]

However, a few children had persistent symptoms or malacia, which seemed to be closely associated with bronchomalacia preoperatively, similar to what has been seen at other centers.[31] Most frequently, persistent disease involved the right mainstem/R1, and all likely had exacerbated airway weakness due to recurrent infections or connective tissue disorder. Alternative approaches, such as stents or external splints, may be useful for persistent bronchomalacia.[19,23]

Recurrent infections and dysphagia, although significantly improved from preoperatively, seemed to persist in approximately 20% of children. Esophageal strictures can form from prolonged compression, and motility can be affected as well,[32] whereas recurrent infections can impair pulmonary epithelial function.[33] Therefore, chronic primary disease of both the lung and esophagus may contribute to persistent symptoms, which highlights the need for ongoing multidisciplinary management and prompt intervention.

Of note, only 1 of 13 children with a diverticulum of Kommerell underwent resection at their initial operation. This has been found to be a source of ongoing compression[11] and likely continues to propagate TBM. We directly addressed the diverticulum in 9 of 12 patients, all with right arch and aberrant left subclavian; the 3 remaining were related to double aortic arch. It has been suggested that all patients with a large diverticulum of Kommerell associated with vascular rings should undergo resection and subclavian reimplantation at their index operation.[2,11] Likewise, there were 7 aberrant left subclavian arteries divided after being identified as an ongoing source of compression, and primary division of the subclavian has been advocated when approaching a right arch with aberrant left subclavian ring.[34]

Study Limitations

This study was not without its limitations. As a retrospective review at a single quaternary referral center, it is subject to the usual biases of patient selection and varying demographics. Given the challenging and unique patient population, there is also no good control cohort with which we can compare operative outcomes. The clinical follow-up was available in 81% (22/27) of our cohort, although loss

can be expected with such a large number of out-of-state referrals. The follow-up duration was also short at a median of only 1.1 years. The true test of the repair will be monitoring for recurrence as the children grow and become more active.

CONCLUSIONS

We have identified 5 etiologies for recurrent or persistent symptoms after vascular ring or arterial decompression operations and proposed solutions to successfully address these (Figure 3). Such approaches could be effective at the index operation in a subpopulation of these patients to prevent recurrence and achieve optimal airway outcomes.

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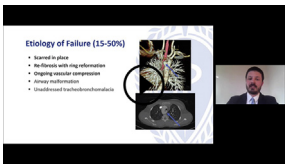

Conflict of Interest Statement

The authors reported no conflicts of interest.

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Key Words: reoperation, tracheobronchomalacia, vascular ring

Discussion

Presenter: Dr Daniel Francis Labuz

Dr Roosevelt Bryant III *(Cincinnati, Ohio)*. I am a congenital heart surgeon at Phoenix Children's Hospital. This is an important series because it continues to raise awareness regarding failed symptom resolution after vascular ring repair. As you know, Dr Backer and colleagues at Lurie Children's initially addressed this issue with their seminal report describing resection of the diverticulum of Kommerell in patients with persistent symptoms, particularly dysphagia, some 20 years ago.

We know from other single-center reports that 45% to 65% of patients with a single aortic arch or double aortic arch variant have symptom persistence after initial repair. The 10-year freedom from reoperation is only 86%. Where I believe your study is unique is its emphasis on the management of airway pathology as an adjunct to the re-repair of patients with persistent symptoms. This was done through a variety of airway pexy procedures, tracheal resection, and tracheal splints. You demonstrated a substantial reduction in symptoms after re-repair. However, 22 of 27 patients presenting with persistent or recurrent symptoms continued to have residual symptoms after re-repair; 23% had recurrent or persistent respiratory infections, and 18% had dysphagia.

My first question is: What should the expectations be after re-repair in terms of symptom resolution, and how do you counsel families in this regard?

Dr Daniel Francis Labuz *(Boston, Mass)*. Expectation should be set based on the patient's anatomy according to the preoperative workup. In our experience, in some patients who have significant and diffuse airway disease, sometimes it might take more than 1 operation to address everything. Sometimes, in terms of their anatomy, we have to do a staged approach starting with a posterior approach and then an anterior approach if the posterior approach is not enough. Also, with patients who are having early infections and issues with dysphagia, there's just intrinsic issues with the esophagus or even pulmonary epithelial dysfunction that can lead to ongoing infectious and dysphagia type symptoms. This is one of the reasons we really advocate for a multidisciplinary care center to fully take care of these patients and address all their unique needs.

Dr Bryant. Can you expand on what you think some of the additional reasons were for why 22 patients had residual symptoms and what might be done differently in the future to address that? You mentioned some patients needing anterior or posterior approaches and different operations. Was there anything else that you thought perhaps contributed to the persistent symptoms?

Dr Labuz. Just to clarify, 22 patients did not have persistent symptoms. There were 7 total with persistent symptoms of the 22 who we were following up on. So, it was only approximately 30% who were having persistent symptoms. And even out of those, like I mentioned in the presentation, 95% overall had some sort of symptom improvement.

In terms of the reason for the ongoing symptoms, you alluded to it. Sometimes the posterior pexy is not enough, and we have to proceed with the second stage and do an anterior pexy. Sometimes doing them at the same time might not be fully efficient. Sometimes we have to do a little bit better exposure for the left mainstem and use different approaches just based on patients' anatomy. The important point is customizing our approach based on the patient's anatomy.

Dr Bryant. Given our increasing understanding of the complexity of these patients, do you think we should be moving toward these multidisciplinary team models for addressing all patients with vascular ring anomalies? It seems particularly important, given the use of intraoperative bronchoscopy, to guide your interventions on the airway disease.

Dr Labuz. Yes. I would advocate for the multidisciplinary approach for focusing on preoperative workup including a workup that would identify all the factors associated or related to the clinical problems. This would potentially include gastroenterology, otolaryngology, pulmonary, general surgery, and a specialized bronchoscopist with a true understanding of malacia. Many of these patients with unidentified malacia or another unidentified airway or gastrointestinal deformity could benefit from just more complex workup and dedicated intraoperative evaluation to make sure you are appropriately addressing any residual disease that they may have.

Dr Bryant. One patient in your series had a tracheal splint. Can you expand a bit on the role of external tracheal stenting in your practice for patients who present initially for vascular ring repair?

Dr Labuz. Yes, in younger patients we are kind of going more toward a bioresorbable splint that we can mold intraoperatively and then it dissolves within about 1 year. We have a series that we're working on publishing, that hopefully is coming out soon. Other centers have also reported the use of customizable airway splints with positive results.

Dr Bryant. I really enjoyed your series. I think this will be an important contribution.