Aortic uncrossing and tracheobronchopexy corrects tracheal compression and tracheobronchomalacia associated with circumflex aortic arch

Ali Kamran, MD, a Kevin G. Friedman, MD, b Russell W. Jennings, MD, a and Christopher W. Baird, MD c

ABSTRACT

Objective: Aortic uncrossing is an effective procedure for relieving the external airway compression from a circumflex aortic arch by transferring the aortic arch to the same side as the descending aorta. However, patients frequently have residual tracheobronchomalacia (TBM), which may result in persistent postoperative symptoms. We review a series of patients who underwent an aortic uncrossing and concomitant tracheobronchopexy to correct the airway compression and residual TBM.

Methods: Retrospective review of all patients who underwent aortic uncrossing and concomitant tracheobronchopexy at a single institution between September 2016 and March 2019. Preoperative evaluation included computed tomography angiography and rigid 3-phase dynamic bronchoscopy.

Results: Eight patients who ranged in age from 4 months to 15 years with significant respiratory symptoms underwent an aortic uncrossing procedure with concomitant tracheobronchial procedures. Mild hypothermic cardiopulmonary bypass (mean time, 105.6 ± 39.4 min) and regional perfusion (mean time, 44 ± 10 min) were used without circulatory arrest. Intraoperative bronchoscopy demonstrated no patients had residual TBM. There were no postoperative mortalities, neurologic complications, chylothoraces, coarctations, or obstructed aortic arches. Two patients required tracheostomy and gastrostomy for bilateral recurrent laryngeal nerve paresis (patients 2 and 3). One patient with bronchial stenosis after concomitant slide bronchoplasty required stenting. At a median follow-up of 22 months (range, 5-34 months), all patients were alive without evidence of significant respiratory symptoms.

Conclusions: The aortic uncrossing procedure can be performed safely in pediatric patients of all ages without circulatory arrest. Concomitant procedures addressing associated TBM can significantly improve respiratory symptoms. (J Thorac Cardiovasc Surg 2020;160:796-804)

CENTRAL MESSAGE

Aortic uncrossing without circulatory arrest and concomitant tracheobronchopexy is effective in treating symptomatic airway compression and tracheobronchomalacia associated with circumflex aortic arch.

PERSPECTIVE

Aortic uncrossing and concomitant tracheobronchopexy for treating severe airway compression and tracheobronchomalacia associated with circumflex aortic arch can improve clinical symptoms and degree of airway collapse on bronchoscopy. An individualized and flexible surgical approach guided by the precise vascular anomaly and intraoperative bronchoscopy is required to optimize the outcomes.

See Commentaries on pages 805, 806, and 807.
and aortic uncrossing where the retro aorta that crosses the trachea and esophagus is transected, mobilized, and anastomosed to the ascending aorta on the same side of the airway as the descending aorta. The aortic uncrossing procedure is focused on relieving the external airway compression but does not address the cartilage malformation and tracheal stenosis nor the tracheobronchomalacia (TBM), which frequently remains at the site of compression and/or the distal airways. Patients with residual TBM may continue to have postoperative respiratory symptoms and may require tracheostomy and/or reoperative surgery. The purpose of this study was to review our experience in a series of patients with a circumflex aortic arch causing severe airway compression and TBM who underwent an aortic uncrossing procedure and concomitant tracheobronchial procedures to address the external airway compression and residual TBM.

**METHODS**

**Patients**

All patients with a circumflex aortic arch causing severe airway compression undergoing an aortic uncrossing procedure at Boston Children’s Hospital between September 2016 and March 2019 were retrospectively reviewed under an approved institutional review board protocol (IRB-P00004344). Data, including demographic characteristics, vascular anomalies, comorbidities, prior operations, preoperative respiratory symptoms, surgical procedures performed, and postoperative outcomes were collected.

**Preoperative Evaluations**

Patients underwent a computed tomography (CT) angiogram with 3-dimensional reconstruction of the airway and the blood vessels to definitively delineate the vascular anomaly and the anatomic relationships of the trachea and main bronchi to the surrounding vasculature. Patients also underwent a rigid, dynamic 3-phase bronchoscopy to precisely assess the extent and severity of airway compression and associated TBM.

**Surgical Technique**

Following median sternotomy and subtotal thymectomy, the pericardium was opened. The extensive dissection and mobilization of the ascending aorta, transverse aortic arch, and branch pulmonary arteries were performed, taking care to identify and preserve the vagus and recurrent laryngeal nerves. The ductal ligament and smallest arch in patients with a double aortic arch were ligated and divided if present. The patient was heparinized, the brachiocephalic artery and right atrium were cannulated for mild hypothermic (25°C-30°C) regional cardiopulmonary bypass. The aortic cannula was placed at the base of the innominate or right carotid artery (Figure 2, A). While cooling, further mobilization of the aortic arch and its main branches was performed. The distal transverse aortic arch just beyond the subclavian artery was crossclamped and second crossclamp was applied 1 to 2 cm more distal, and the aorta was divided between the 2 clamps and the proximal end was oversewn. An ascending aortic crossclamp was then applied followed by the cardioplegic diastolic arrest, and regional perfusion was initiated via the innominate artery. The descending aorta was dissected from its posterior attachments, pulled from behind the trachea, brought lateral to the trachea, and anastomosed to the ascending aorta in an end-to-side fashion on the contralateral side (Figure 2, A). Frequently, an autologous pericardial patch was used to augment the aortic arch reconstruction. The aorta was de-aired, aortic crossclamp removed, and once the patient was re-warmed, cardiopulmonary bypass was weaned.

Rotational esophagoplasty was routinely performed to move the esophagus to the contralateral side of the newly positioned aortic arch to prevent...
recurrent posterior tracheal compression and relieve esophageal entrapment, when present (Figure 2). Based on the type, location, and severity of the residual TBM, pexy procedures, including aortopexy, tracheopexy (anterior and/or posterior), and/or bronchopexy (anterior and/or posterior), were performed under bronchoscopic guidance (Figure 2, B). Tracheobronchial resection with slide tracheobronchoplasty was also performed in patients with marked cartilaginous alteration.

RESULTS

Our single-center experience between September 2016 and March 2019 included 8 patients (aged between 4 months and 15 years) with a circumflex aortic arch with significant respiratory symptoms who underwent an aortic uncrossing procedure with concomitant tracheobronchial procedures for severe airway compression and TBM. All patients underwent preoperative CT angiogram and a rigid dynamic 3-phase bronchoscopy (Figure 3). Seven patients had a right circumflex aortic arch, and 1 patient had a left circumflex aortic arch. Of the 7 patients with a right circumflex aortic arch, 5 had a right-dominant double aortic arch. Three patients had prior operations, including the division of ductal ligament and/or double aortic arch division, without improvement in their respiratory symptoms. Patients’ demographic characteristics, vascular anomaly, comorbidities, prior operations, and respiratory symptoms are listed in Table 1.

The aortic uncrossing procedure was performed with a median sternotomy using mildly hypothermic cardiopulmonary bypass (mean time, 105.6 ± 39.4 minutes) and regional perfusion (mean time, 44 ± 10 minutes) without

FIGURE 2. The circumflex aorta (Ao) was transected and anastomosed to the ascending Ao on the same side of the airway as the descending Ao (aortic uncrossing). The esophagus was rotated to the right side (rotational esophagoplasty). The posterior tracheal membrane with severe dynamic intrusion was fixed to the anterior longitudinal ligament of the spine (posterior tracheopexy). A, Anterior view. B, Lateral view. PA, Pulmonary artery.

circulatory arrest. Only 1 patient required intermittent circulatory arrest for 3 minutes. Concomitant procedures were also performed in all patients to optimize each individual airway (Table 2). The intraoperative bronchoscopic evaluation following repair showed marked improvement in airway patency in all patients. Details of patient-specific vascular and tracheobronchial procedures are listed in Table 2. All patients underwent postoperative echocardiogram demonstrating an unobstructed arch.

There were no postoperative mortalities, esophageal dysfunction, neurologic complications, chylothoraces, or coarctations. Two patients with bilateral recurrent laryngeal nerve paresis required tracheostomy and feeding-tube placement (patients 2 and 3). One patient with bronchial stenosis after slide bronchoplasty required stenting (patient 1). At a median follow-up of 22 months (range, 5-34 months), all patients were alive without evidence of significant respiratory symptoms (Table 3).

**DISCUSSION**

In normal circumstances, the descending aorta persists on the same side as the aortic arch. However, a circumflex aortic arch is composed of an aortic arch with a retrotracheo-esophageal descending aorta crossing the midline to the contralateral side. A circumflex aortic arch can cause posterior compression by the descending aorta passing behind the trachea and esophagus as well as lateral compression where the aortic arch is tethered to the left pulmonary artery by the ductal ligament. Division of the ductal ligament releases the vascular ring but does not effectively relieve the tracheal stenosis due to cartilage malformation from the compressive aorta, nor the posterior airway

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**TABLE 1. Patient demographic characteristics and preoperative data**

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Sex</th>
<th>Age</th>
<th>Vascular anomaly</th>
<th>Comorbidities</th>
<th>Prior operations</th>
<th>Respiratory symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Male</td>
<td>4 mo</td>
<td>Left circumflex aortic arch: (left aortic arch with aberrant right subclavian artery, right descending aorta)</td>
<td>Right lung agenesis, tracheobronchial compression</td>
<td>None</td>
<td>Respiratory distress, noisy breathing</td>
</tr>
<tr>
<td>2</td>
<td>Male</td>
<td>7 mo</td>
<td>Right circumflex aortic arch: (right-dominant double aortic arch, left descending aorta, left ductal ligament)</td>
<td>EA/TEF, tracheal compression, TBM</td>
<td>EA/TEF repair (DOL 1), Division of left arch and ductal ligament (DOL 7)</td>
<td>Respiratory insufficiency requiring ventilatory support</td>
</tr>
<tr>
<td>3</td>
<td>Male</td>
<td>9 mo</td>
<td>Right circumflex aortic arch: (right-dominant double aortic arch, left descending aorta, left ductal ligament)</td>
<td>Tracheal compression, TBM</td>
<td>None</td>
<td>Noisy breathing, recurrent respiratory infection</td>
</tr>
<tr>
<td>4</td>
<td>Male</td>
<td>1 y</td>
<td>Right circumflex aortic arch: (right aortic arch, left descending aorta, left ductal ligament)</td>
<td>Tracheal compression, TBM</td>
<td>None</td>
<td>Noisy breathing, recurrent respiratory infection</td>
</tr>
<tr>
<td>5</td>
<td>Male</td>
<td>2 y</td>
<td>Right circumflex aortic arch: (right aortic arch, left descending aorta, left ductal ligament)</td>
<td>Tracheal compression, TBM</td>
<td>None</td>
<td>Prolonged and recurrent respiratory infection, exercise intolerance</td>
</tr>
<tr>
<td>6</td>
<td>Female</td>
<td>5 y</td>
<td>Right circumflex aortic arch: (right-dominant double aortic arch, left descending aorta, left ductal ligament)</td>
<td>Tracheal compression, TBM</td>
<td>None</td>
<td>Prolonged and recurrent respiratory infection, exercise intolerance, barking cough</td>
</tr>
<tr>
<td>7</td>
<td>Male</td>
<td>12 y</td>
<td>Right circumflex aortic arch: (right-dominant double aortic arch, left descending aorta, left ductal ligament)</td>
<td>Tracheal compression, TBM</td>
<td>Division of left arch (age 4 mo)</td>
<td>Exercise intolerance, recurrent respiratory infection</td>
</tr>
<tr>
<td>8</td>
<td>Female</td>
<td>15 y</td>
<td>Right circumflex aortic arch: (right-dominant double aortic arch, left descending aorta, left ductal ligament)</td>
<td>Tracheal compression, TBM</td>
<td>Division of left arch and ductal ligament (age 8 mo)</td>
<td>Noisy breathing, exercise intolerance</td>
</tr>
</tbody>
</table>

EA, Esophageal atresia; TEF, tracheoesophageal fistula; TBM, tracheobronchomalacia; DOL, day of life.
compression caused by the aorta crossing behind the lower trachea and/or main bronchi. Furthermore, a fibrous band may be formed between the divided ends of the ductus and/or divided aortic arch in double aortic arch patients that may contract, effectively reforming a vascular-fibrous ring and recapitulating the original problem.

In symptomatic patients, to completely address the external compression caused by a circumflex aortic arch, it is necessary to move the posterior aorta away from the tracheobronchial system and/or esophagus. This can be accomplished by an aortic uncrossing procedure in which the aortic arch is transferred to the same side as the descending aorta so that it runs parallel and lateral to the trachea and esophagus rather than directly crossing behind them. Planche and LaCoeur-Gayet\(^1\) first described the aortic uncrossing procedure in 3 patients, all of whom had previously

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Vascular procedures</th>
<th>Concomitant procedures</th>
<th>Circulatory arrest time (min)</th>
<th>Cardiopulmonary bypass time (min)</th>
<th>Crossclamp time (min)</th>
<th>Regional perfusion time (min)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Aortic uncrossing (left to right), ductal ligament division, left subclavian artery division</td>
<td>Tracheobronchial resection with slide tracheobronchoplasty, descending aortoplasty (posterior)</td>
<td>0</td>
<td>66</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Aortic uncrossing (right to left)</td>
<td>Tracheal resection with slide tracheoplasty, tracheoplasty (anterior, posterior)</td>
<td>3</td>
<td>110</td>
<td>66</td>
<td>41</td>
</tr>
<tr>
<td>3</td>
<td>Aortic uncrossing (right to left), ductal ligament division, left arch division</td>
<td>Tracheoplasty (anterior, posterior), left main bronchoplasty (posterior)</td>
<td>0</td>
<td>74</td>
<td>39</td>
<td>35</td>
</tr>
<tr>
<td>4</td>
<td>Aortic uncrossing (right to left), ductal ligament division</td>
<td>Tracheoplasty (anterior, posterior), left main bronchoplasty (posterior)</td>
<td>0</td>
<td>79</td>
<td>49</td>
<td>31</td>
</tr>
<tr>
<td>5</td>
<td>Aortic uncrossing (right to left), ductal ligament division</td>
<td>Tracheoplasty (anterior, posterior), ascending aortoplasty (anterior)</td>
<td>0</td>
<td>77</td>
<td>30</td>
<td>42</td>
</tr>
<tr>
<td>6</td>
<td>Aortic uncrossing (right to left), ductal ligament division, left arch division</td>
<td>Tracheoplasty (posterior), bilateral bronchoplasty (posterior), descending aortoplasty (posterior)</td>
<td>0</td>
<td>177</td>
<td>31</td>
<td>56</td>
</tr>
<tr>
<td>7</td>
<td>Aortic uncrossing (right to left), ductal ligament division</td>
<td>Tracheoplasty (anterior, posterior), left main bronchoplasty (anterior), right main bronchoplasty (anterior)</td>
<td>0</td>
<td>94</td>
<td>41</td>
<td>61</td>
</tr>
<tr>
<td>8</td>
<td>Aortic uncrossing (right to left)</td>
<td>Tracheoplasty (posterior)</td>
<td>0</td>
<td>118</td>
<td>63</td>
<td>43</td>
</tr>
</tbody>
</table>

**TABLE 3. Postoperative data and follow-up**

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Echocardiography</th>
<th>Hospital stay (d)</th>
<th>Complications during hospital stay</th>
<th>Further procedure</th>
<th>Respiratory symptoms</th>
<th>Follow-up (mo)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Unobstructed arch</td>
<td>131</td>
<td>Bronchial stenosis after slide bronchoplasty</td>
<td>Bronchial stenting and multiple dilations</td>
<td>Episodes of respiratory distress (improved)</td>
<td>31</td>
</tr>
<tr>
<td>2</td>
<td>Unobstructed arch</td>
<td>43</td>
<td>Bilateral vocal cord dysfunction</td>
<td>Tracheostomy Feeding-tube placement</td>
<td>None</td>
<td>31</td>
</tr>
<tr>
<td>3</td>
<td>Unobstructed arch</td>
<td>61</td>
<td>Bilateral vocal cord dysfunction</td>
<td>Tracheostomy Feeding-tube placement</td>
<td>None</td>
<td>14</td>
</tr>
<tr>
<td>4</td>
<td>Unobstructed arch</td>
<td>8</td>
<td>No complication</td>
<td>None</td>
<td>None</td>
<td>34</td>
</tr>
<tr>
<td>5</td>
<td>Unobstructed arch</td>
<td>6</td>
<td>No complication</td>
<td>None</td>
<td>None</td>
<td>29</td>
</tr>
<tr>
<td>6</td>
<td>Unobstructed arch</td>
<td>7</td>
<td>No complication</td>
<td>None</td>
<td>None</td>
<td>7</td>
</tr>
<tr>
<td>7</td>
<td>Unobstructed arch</td>
<td>4</td>
<td>No complication</td>
<td>None</td>
<td>None</td>
<td>5</td>
</tr>
<tr>
<td>8</td>
<td>Unobstructed arch</td>
<td>5</td>
<td>No complication</td>
<td>None</td>
<td>None</td>
<td>14</td>
</tr>
</tbody>
</table>
undergone ligamentum division and had persistent postoperative symptoms. In 2013, Russell and colleagues reported their experience of performing this procedure in 4 patients using cardiopulmonary bypass, deep hypothermia, and circulatory arrest.

Although the aortic uncrossing procedure can effectively relieve the external airway compression, it does not address the residual TBM at the site of airway compression and/or the branch bronchi. Additionally, the deformed tracheal rings may remain in the compressed shape after removing the external compression. Thus, in addition to relieving the external airway compression, an optimal outcome requires the residual TBM and/or tracheal cartilage deformation to be directly addressed; otherwise, it can be the reason for persistent and long-standing respiratory symptoms.3,4

Our multidisciplinary team routinely uses a standardized scoring system based on bronchoscopic evaluation of the airway anatomic regions.5-7 The greater the severity of airway collapse combined with the presence of clinical symptoms may indicate the need for surgical intervention. The bronchoscopic evaluation is performed in 3 phases to completely assess abnormalities in the airways. The first phase occurs while the patient is shallow breathing to reveal not only the airway compression but also the secretion accumulation and vocal cord dysmotility if present. The second phase is to induce coughing and Valsalva maneuvers while observing the entire airway, which demonstrates the maximum dynamic airway collapse and also helps accumulated secretions to move into the larger airways. Finally, the third phase is to distend the airways to 30 to 60 cm of water, which will reveal structures and lesions that may not normally be seen, including tracheoesophageal fistulas, tracheal diverticulum, and aberrant bronchi.

Surgical treatment options for the treatment of TBM include anterior and/or posterior tracheobronchoplasty, tracheobronchial resection with slide tracheobronchoplasty, and placement of internal stents and external splints. These often combined with anterior ascending aortopexy and/or posterior descending aortopexy. Anterior ascending aortopexy indirectly supports the anterior wall of the trachea through the attachment of the aorta to the back of the sternum but does not directly influence the posterior membranous intrusion, which is the most common cause of pediatric TBM. Also, the tracheal attachments to the aorta will grow and relax over time, which can limit the effectiveness of aortopexy.6,11 Anterior and/or posterior tracheobronchoplasty improve airway patency by directly addressing anteriorly compressed tracheal cartilages and posterior tracheobronchial membranous intrusion.7,12,13 Furthermore, for those with posterior compression of the left mainstem bronchus from the descending aorta, a posterior descending aortopexy is performed.

This article attempts to address the issue of balancing the risks of aortic uncrossing in symptomatic patients who have airway compression. We reported our experience concerning surgical correction of 8 patients with symptomatic airway compression from the circumflex aortic arch. Five of the 8 patients in this series had a double aortic arch that was right-dominant arch crossing to a left-sided descending aorta, 3 of whom had prior left aortic arch division and/or ductal ligamentum division but had persistent symptoms after vascular ring division due to continued airway compression from the right circumflex aorta and persistent untreated TBM. When the circumflex aorta crosses behind the lower trachea and/or main bronchi, it can act as a mass that posteriorly compresses the airway and covers the spine in such a way as to prevent posterior airway support (Videos 1 and 2). Viewing a CT angiogram often reveals the necessary maneuvers required to achieve an open airway; often the descending aorta needs to be moved, which necessitates an aortic uncrossing. We consider moving the descending aortic crossing point below the airways, as well as shifting the descending aorta to 1 side and supporting the airway next to the aorta—the problem with both is that the aorta tends to move back centrally and cause later airway posterior airway compression.

In this series, 2 patients had bilateral vocal cord paresis early in the experience that required tracheostomy and associated aspiration that required gastrostomy feeds, and 1 patient with unilateral lung agenesis required bronchial stent placement and dilation because of bronchial stenosis after slide bronchoplasty, which significantly improved over a period of several months. The remaining 5 patients had no complications and were rapidly discharged home without respiratory symptoms and no swallowing dysfunction. Our protocol is esophagram and vocal cord evaluation before discharge and close follow-up of all patients. If asymptomatic, they undergo an evaluation that includes bronchoscopy at 1 year. The patients addressed in this series did not require airway reintervention.

Identification of the vagus and recurrent laryngeal nerves should be performed during the dissection of the mediastinum to minimize the risk of their injury. The abnormal anatomic variations and somewhat unpredictable courses of the vagus and recurrent laryngeal nerves put them at risk, and experience is required to prevent nerve injuries. The 2 children with bilateral vocal cord injury were the first 2 patients in this series we operated on for severe airway compression with TBM and circumflex descending aorta. We have since improved the dissection technique to protect the vagus and recurrent laryngeal nerves. Also, we have started performing vocal cord assessments preoperatively and postoperatively to further identify the incidence of vocal cord injury during the procedure, and more recently, we have been utilizing recurrent laryngeal nerve monitoring during the dissection around the aorta and ductal ligament to reduce nerve injury.

We have a unique referral center for complex airway problems, and we receive a highly selected group of very
symptomatic patients. The majority of complex patients referred to us with previously divided vascular rings remain symptomatic from the residual TBM and may require further reoperation(s). We have developed an effective tool (dynamic 3-phase bronchoscopy) for detecting and classifying the regions and severity of airway compression and TBM, which are 2 different problems. Airway compression and/or cartilage malformation is a static narrowing of the airway and typically caused by great artery malposition and/or abnormalities; however, TBM is a dynamic narrowing of the airway due to excessive intrusion of the posterior membrane that is greatest during forced exhalation and periods of increased intrathoracic pressure. In a recent study at our center, Svetanoff and colleagues reported that, in more than 100 patients with vascular compression abnormalities and symptomatic airway narrowing, the majority of complex patients have not only airway compression and/or cartilage deformation from the great artery but also TBM in the airway distal to the compressed region.

Children with vascular rings but no symptoms usually do not need surgical treatment. However, those who have symptoms from vascular rings typically require surgery to relieve pressure on their airway and esophagus. In those cases, the vascular ring division relieves the external compression on the airway but does not address the deformed cartilage and/or residual TBM, which can be the reason for persistent clinically apparent airway collapse. Those who remain symptomatic typically still have a more than 50% airway narrowing after vascular ring division. Therefore, we do bronchoscopy before, during, and after the vascular procedure to evaluate the airway for residual TBM and/or cartilage malformation. Our preference is to correct all the airway lesions at the same operation as the vascular compression to prevent multiple reoperations with their attendant increased risks. The surgical plan should be based on the type and severity of airway narrowing, taking into account all the associated conditions, including vascular anomalies and cartilage deformation. It is important to customize the plan of care for each patient after complete assessment, including CT angiogram and dynamic 3-phase bronchoscopy, to correct all the problems in a single operation.

Aortic uncrossing is not necessarily needed in all patients with a circumflex aortic arch and should be reserved for patients with symptoms from the great vessel that is greatest during forced exhalation and periods of increased intrathoracic pressure. In a recent study at our center, Svetanoff and colleagues reported that, in more than 100 patients with vascular compression abnormalities and symptomatic airway narrowing, the majority of complex patients have not only airway compression and/or cartilage deformation from the great artery but also TBM in the airway distal to the compressed region. Children with vascular rings but no symptoms usually do not need surgical treatment. However, those who have symptoms from vascular rings typically require surgery to relieve pressure on their airway and esophagus. In those cases, the vascular ring division relieves the external compression on the airway but does not address the deformed cartilage and/or residual TBM, which can be the reason for persistent clinically apparent airway collapse. Those who remain symptomatic typically still have a more than 50% airway narrowing after vascular ring division. Therefore, we do bronchoscopy before, during, and after the vascular procedure to evaluate the airway for residual TBM and/or cartilage malformation. Our preference is to correct all the airway lesions at the same operation as the vascular compression to prevent multiple reoperations with their attendant increased risks. The surgical plan should be based on the type and severity of airway narrowing, taking into account all the associated conditions, including vascular anomalies and cartilage deformation. It is important to customize the plan of care for each patient after complete assessment, including CT angiogram and dynamic 3-phase bronchoscopy, to correct all the problems in a single operation.

Aortic uncrossing can be effective at relieving airway compression when combined with tracheobronchopexy and airway reconstruction procedures as needed to address residual TBM and/or deformation of tracheal cartilage. We have experience in the treatment of many forms of airway compression, including those caused by vascular rings, as well as TBM. We perform posterior tracheobronchopexy in symptomatic children with vascular rings who also have TBM distal to the vascular ring. The posterior tracheobronchopexy has been the preferred method at our institution to support the posterior membrane, allowing for multiple areas of airway collapse to be directly addressed during the same operation as well as a good approach to correct many vascular compression anomalies. A minority of these patients may also need anterior airway support (anterior tracheobronchopexy). Our goal is to achieve at least a 50% open airway. Resection of the trachea and/or main bronchus may be indicated in some types of short-segment tracheobronchial collapse, such as in cases with congenital absence of cartilage or severe cartilage deformation.

**VIDEO 1.** Preoperative computed tomography scan demonstrating right aortic arch and left descending aorta passing behind the trachea and esophagus, causing posterior compression. Video available at: [https://www.jtcvs.org/article/S0022-5223(20)31000-0/fulltext](https://www.jtcvs.org/article/S0022-5223(20)31000-0/fulltext).

**VIDEO 2.** Postoperative computed tomography scan following aortic uncrossing and posterior tracheopexy to correct airway compression and tracheomalacia associated with circumflex aortic arch. Video available at: [https://www.jtcvs.org/article/S0022-5223(20)31000-0/fulltext](https://www.jtcvs.org/article/S0022-5223(20)31000-0/fulltext).
This study is the largest review of aortic uncrossing in the literature and highlights the use of regional perfusion to avoid circulatory arrest and includes tracheobronchopexy and airway reconstruction procedures as needed to optimize airway outcomes. Given the complexity of this patient population, treatment and long-term follow-up is best done in multidisciplinary specialized centers for individualized patient care. This study is limited by the single-center, retrospective nature of the work and the relatively short-term and variable follow-up intervals. Further studies to follow long-term outcomes are certainly warranted and are ongoing.

CONCLUSIONS

Aortic uncrossing is indicated for patients with a circumflex aortic arch who require the aorta repositioning to relieve symptomatic airway compression. The aortic uncrossing procedure can be performed safely in patients of all ages without circulatory arrest. Concomitant procedures addressing the residual TBM and/or tracheal cartilage deformation may also be required to optimally open the airways.

Webcast

You can watch a Webcast of this AATS meeting presentation by going to: https://aats.blob.core.windows.net/media/19%-20AM/Saturday_May4/202BD/202BD/S35%-20-%20Airway%20Surgery%20in%20Children/S35_6_webcast_02445225.mp4.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The Journal policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References


Key Words: circumflex aorta, aortic uncrossing procedure, airway compression, tracheobronchomalacia (TBM), tracheobronchopexy

Discussion

Presenter: Dr Ali Kamran

Carl L. Backer (Chicago, Ill). Very nice presentation; thank you. Among the take-away messages here is that preoperative imaging is so very important. A circumflex aorta is a much more complicated vascular ring than a so-called simple right aortic arch. Dividing the ligamentum in a patient with a circumflex aorta will not necessarily resolve the patient’s symptoms. When the aorta crosses posteriorly above the carina, you need to consider the aortic uncrossing operation.

Our results are similar to yours, but our operative strategy was different. All of our aortic uncrossings have been done under deep hypothermic circulatory arrest, which I think
facilitates the operation. How do you do regional cerebral perfusion when you have 4 separate arch vessels; in other words, you don’t have an innominate artery. Do you just cannulate a carotid artery?

**Dr Ali Kamran (Boston, Mass).** The cannulation was on the ascending aorta at the take-off of the innominate or the right subclavian artery.

**Dr Backer.** So you cannulate both the right subclavian artery and the right carotid artery separately?

**Dr Kamran.** No.

**Dr Christopher W. Baird (Boston, Mass).** So for all of these patients, we cannulated the aorta at the base or the take-off of the innominate artery.

**Dr Backer.** Some of these patients don’t have an innominate artery because they’ve got 4 separate vessels coming out of the arch.

**Dr Baird.** Yes, but usually, at least in our experience, if you cannulate the distal ascending aorta, we can divide just below the take-off of the right subclavian. So in our experience, it really hasn’t mattered; you just cannulate as distal in the ascending aorta as you can and you’re going to perfuse all of the head vessels. The caveat to that is that if you have an aberrant right subclavian artery.

**Dr Backer.** Correct, most of my patients have had an aberrant.

**Dr Baird.** Then your right subclavian is going to come off more distal. And then in that case, we had I think 1 in this series, we divided the subclavian artery and reimplanted it in the innominate artery. So for the most part, we didn’t have a problem with perfusion. Another option is to just divide the aorta a little farther distal beyond the subclavian.

**Dr Backer.** The other issue is whether you really need to do the tracheopexy. In our 8 patients, there was only 1 patient who may have benefited from this. He came to me with a tracheostomy after 2 previous operations and stayed in the hospital for 30 days before we finally sent him home. All of our other patients have been extubated and gone home within about 6 to 8 days, and we haven’t had a problem with tracheomalacia. We also do a preoperative bronchoscopy on all our patients. With many of them, we’ve done postoperative imaging and postoperative bronchoscopies and the area of compression definitely improves. They get discharged without symptoms. So I don’t know if we have 2 different populations, or how to explain that.

**Dr Baird.** Yes. I suspect that is it. We don’t advocate doing the uncrossing operation if there are no tracheal symptoms preoperatively—so I think that’s first and foremost. We do intraoperative bronchoscopy. We also do a study in which you put negative pressure on the airway to see how much collapse there is. But I think the most important thing is, we try to evaluate the symptoms preoperatively. If the airway is not involved, it may not require tracheal intervention. So we’re not advocating for a tracheopexy in every patient who gets an uncrossing operation. However, if the trachea is involved, at least in our experience, it has been.

**Dr Backer.** I think in our series, there was probably just 1 patient who would have benefited from that. Final question: You have 2 patients with bilateral recurrent laryngeal nerve injuries. We had 1 in our series who had temporary bilateral recurrent laryngeal nerve palsy. Have you done anything differently since these 2 patients?

**Dr Baird.** Yes. That was probably related to being too aggressive with electrocautery. The left recurrent nerve is quite easy and obvious to see—whereas the right is sometimes a little bit higher and more difficult to see. There has been some suggestion about using a different type of cautery such as bipolar; however, I’m not sure if that’s necessary. But clearly, we were very aggressive on the mobilization before. Now we have toned down the aggressiveness with electrocautery. We have also recently employed the use of a nerve stimulator/monitor.

**Dr Backer.** I think this is something you really have to pay attention to, and even though we probably both pay attention to it, you have to be really careful. Thanks for a nice presentation.

**Dr Petros V. Anagnostopoulos (Madison, Wis).** Chris, do you guys do the tracheopexy through the sternotomy and the esophageal? Dissection through the sternotomy?

**Dr Baird.** Yes, actually most of that part is pretty straightforward through the sternotomy. Once you divide the aorta and pull it under the trachea—and again, I can’t emphasize enough the benefit of Dr Jennings’ experience with the esophagus; I don’t have to worry about how aggressive I can be; he can handle the esophagus, for lack of a better way to put it, and it does okay. So once we get the esophagus out of the way, it gives us complete access to the spine and the posterior trachea. It is a little tricky, because you’re trying to look down and around, under and up on the airway. That’s where the intraoperative bronchoscopy helps. So actually, you’re putting the stitches in, but looking at the bronchoscopy images on the screen to determine exactly where they go.

**Dr Anagnostopoulos.** I would imagine the double row of stitches, the left side of sutures, are difficult to see, right?

**Dr Baird.** Yes, it’s often very hard to gauge how far left or right you are.