Vascular Compression of the Airway

Establishing a Functional Diagnostic Algorithm

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Importance: Pediatric imaging carries the risk of radiation exposure. Children frequently undergo computed tomography with angiography (CTA) for findings on bronchoscopy with limited knowledge regarding the necessity of such imaging.

Objective: To report our experience with all pediatric patients at our institution over an 8-year period with airway symptoms warranting bronchoscopy followed by CTA for potential vascular anomaly. Goals were to report the percentage of positive findings seen on CTA leading to surgery; discuss relative radiation exposure risk and sedation risk for additional radiologic studies; and propose a functional diagnostic algorithm.

Design, Setting, and Participants: Retrospective chart review of 42 children aged 2 months to 11 years with tracheomalacia who underwent CTA between 2004 and 2012 in our tertiary aerodigestive center.

Interventions: Bronchoscopy and CTA.

Main Outcomes and Measures: Presence of vascular anomaly and need for thoracic surgery.

Results: Of these 42 children, 21 (50%) had a vascular anomaly identified on CTA. Of these 21, 17 (81%) had innominate artery compression; 1 (5%) had double aortic arch; 1 (5%) had right aortic arch; 3 (14%) had bronchial compression by pulmonary artery; and 1 (5%) had dextrocardia with duplicated vena cava. Six (29%) of these 21 had clinical symptoms and CTA findings requiring thoracic surgery. The most common symptoms in children requiring thoracic surgery were cough, cyanosis, and stridor.

Conclusions and Relevance: Deciding when to obtain imaging for bronchoscopic findings suggestive of vascular compression remains challenging. A diagnostic algorithm is proposed as a means to provide the best clinical care while weighing risks of additional radiation exposure vs sedation and exposure to general anesthesia.

Eggy in the pediatric population. Even if MRI is performed instead of CTA to work up patients with suspected vascular anomalies, patients often need a second general anesthetic, which is not without risks.

The mere presence of a vascular anomaly does not always necessitate thoracic surgery. An anomalous innominate artery may cause few if any symptoms, or it may produce severe tracheal compression with associated symptoms.4 Myer et al5 and others have reviewed absolute and relative criteria for surgery to correct innominate artery compression.6 Complete vascular rings, such as the double aortic arch, and pulmonary artery slings usually require surgical correction.

In light of the varied findings on bronchoscopy and the increased risks associated with CTA and additional general anesthetics, the purpose of this study was to report our experience with all pediatric patients at our institution over a period of 8 years who had tracheomalacia.
The Massachusetts Eye and Ear Infirmary institutional review board approved this study. Included patients were children 18 years or younger who underwent bronchoscopy first for airway symptoms then had a CTA for findings during the bronchoscopy. Exclusion criteria were age older than 18 years, extraluminal or intraluminal mass, retroesophageal subclavian artery, or presence of an additional airway lesion.

Between 2004 and 2012, 48 children had a CTA for suspected vascular anomaly on operative bronchoscopy at our institution. One patient was excluded because of supraglottic stenosis, 1 for subglottic stenosis, 1 for a subglottic hemangioma, 1 for a large subglottic cyst, and 1 for an intraluminal lesion of a bronchus. One patient with a right aberrant subclavian artery was excluded because of the inherent lack of tracheal or bronchial compression associated with this lesion. The resulting total number of patients included in this study was 42.

Symptoms that led to bronchoscopy included severe stridor and only mild laryngomalacia on flexible fiberoptic laryngoscopy, chronic cough unresponsive to empirical reflux and asthma treatment, cyanosis, recurrent bronchopneumonia, and failure to thrive due to aerodigestive symptoms. A CTA was ordered for patients with clinically significant expiratory stridor (where significant was defined as "enough to warrant bronchoscopic evaluation") with a short-segment tracheomalacic segment more than 50% obstructing with expiration or if there was any degree of short-segment combined anterior and posterior compression (suggestive of complete vascular ring).

Due to the intimate relationship of the thoracic vasculature with the trachea and bronchi, any variation in this...
et al12 showed that 96% of children with vascular anomaly; and 1 (17%) had a right aortic arch. Erwin et al27 found that 96% of children with vascular rings and found the most common presenting symptom in all the children with a vascular anomaly and in those who required thoracic surgery was cyanosis and stridor. Tracheal compression also impairs mucociliary clearance, which may lead to recurrent bronchopneumonia and chronic cough.4 The tracheal compression may be severe enough to completely collapse the airway and cause cyanosis or death spells.8,10 In general, complete vascular rings and pulmonary artery slings present earlier in life because they are tighter and produce more compression.10,11 Roesler et al12 showed that 96% of children with vascular anomalies present by age 6 months, but 25% of children had at least a 6-month delay from symptom onset to surgical intervention. In our population, the most common presenting symptom in all the children with a vascular anomaly may produce compression of the airway. Stridor, particularly during expiration, occurs due to higher intrathoracic pressure than atmospheric pressure, which causes narrowing of the airway.9 Any narrowing, whether from intrinsic tracheal or bronchial weakness or from external compression, may cause stridor. Tracheal compression also impairs mucociliary clearance, which may lead to recurrent bronchopneumonia and chronic cough.4 The tracheal compression may be severe enough to completely collapse the airway and cause cyanosis or death spells.8,10 Recent reports of the dangers of radiation from imaging in the pediatric population have forced health care providers to reevaluate the use of CT. Boone et al,15 on behalf of the American Association of Physicists in Medicine (AAPM), determined that CT scans in the pediatric population may result in up to 2.79 times the radiation dose of that in adults, depending on the type of CT scanner used and the size of the child.15 Frush et al2 and Brody et al16 have demonstrated the need for health care providers to follow the ALARA principle as a dose-reduction strategy in the pediatric population. Many centers use MRI to evaluate for potential vascular anomalies,14,17 which represents an attractive alternative to CTA, especially if it can be performed under the same general anesthetic, because it involves no radiation. However, at our center, children are unable to go directly from their bronchoscopy to get an MRI under the same general anesthetic, but they may be transported directly for a CTA. This means scheduling a separate sedation and/or general anesthetic for a longer radiologic examination, often in children with a tenuous airway. Additional general anesthetics in these children are not without risk. Girshin et al18 found that the mortality rate for general anesthesia performed on children in the MRI suite was 1 in 3900, which is approximately twice the rate for general anesthesia performed in the operating room. Aside from the obvious risks of airway obstruction, studies have shown that exposure to anesthetics in young children may lead to behavioral disturbances and learning disabilities.19,20

### Table 3. Characteristics of Children Who Required Thoracic Surgery

<table>
<thead>
<tr>
<th>Patient No./ Age, mo</th>
<th>Syndrome</th>
<th>Stridor</th>
<th>Cyanosis</th>
<th>Failure to Thrive</th>
<th>Recurrent Bronchopneumonia</th>
<th>Cough</th>
<th>Operative Bronchoscopy</th>
<th>CTA</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/2</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>80% Short-segment anterior mid-tracheal compression</td>
<td>43% Innominate artery compression</td>
</tr>
<tr>
<td>2/M/4</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>70% Short-segment right anterior and posterior distal tracheal compression</td>
<td>26% Innominate artery compression, right lateral trachea completely collapsed</td>
</tr>
<tr>
<td>3/F/4</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>75% Short-segment mid-tracheal compression</td>
<td>57% Innominate artery compression</td>
</tr>
<tr>
<td>4/M/5</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>100% Short-segment mid-tracheal compression</td>
<td>48% Innominate artery compression</td>
</tr>
<tr>
<td>5/M/14</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>80% Short-segment distal left tracheal compression, left bronchial narrowing</td>
<td>Right aortic arch with mirror image branching</td>
</tr>
<tr>
<td>6/F/15</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>90% Short-segment distal anterior and posterior tracheal compression</td>
<td>Double aortic arch</td>
</tr>
</tbody>
</table>

Abbreviation: CTA, computed tomography with angiography.

The clinical presentation of a child with a vascular anomaly causing extrinsic tracheomalacia may be quite
would need thoracic surgery should a vascular anomaly be found. All complete vascular rings and pulmonary artery slings undergo thoracic surgery. For innominate artery compression and other partial vascular rings, many children may be observed carefully owing to the self-limiting nature of these anomalies. However, some patients will require thoracic surgery to correct the vascular anomaly. Mustard et al,6 in 1969, described their indications for thoracic surgery as reflex apnea and recurrent bronchopulmonary infections. In 1971, MacDonald et al22 divided surgical criteria into absolute and relative criteria while adding failure of medical management, greater than 50% tracheal compression, and associated airway or lung abnormalities to the existing criteria. Myer et al1 reviewed absolute and relative criteria for thoracic surgery in 1989. Absolute criteria included reflex apnea, failure of medical management of severe respiratory distress after 48 hours, and prolonged intubation. Relative criteria were recurrent tracheobronchitis or bronchopneumonia; exercise intolerance; significant dysphagia or failure to thrive; and coexistence of subglottic stenosis, asthma, cystic fibrosis, or prior tracheoesophageal fistula repair.

We developed a diagnostic algorithm to provide the best clinical care while weighing the risks of additional radiation vs additional sedation and exposure to general anesthesia (Figure 4). When a child presents with symptoms concerning for airway obstruction severe enough to warrant further workup, first a thorough history is taken and physical examination performed followed by flexible laryngoscopy to assess for laryngomalacia, vocal fold mobility, and other lesions. If the child demonstrates severe stridor and only mild laryngomalacia, recurrent bronchopneumonia despite adequate treatment, failure to thrive due to aerodigestive symptoms, or if endoscopy is needed to evaluate recalcitrant reflux and/or reflux, then operative bronchoscopy is performed.

If the tracheal compression or collapse is short-segment type involving 33% or less of the tracheal length (as defined by the senior author C.J.H.), this is suggestive of extrinsic compression. It would seem that external compression by the caliber of a large blood vessel would result in a short length (≤33%) of trachea being compressed and that intrinsic tracheomalacia would involve more of the trachea (>33%). In addition, if the compression involves the right anterior and posterior tracheal walls or mainstem bronchus (fishmouth deformity), this may be from a double aortic arch, other complete vascular ring, or even a pulmonary artery sling and should be imaged. Symptoms are less important in these patients because a complete vascular ring will eventually require surgery. In the remainder of the short-segment tracheal compression group, if greater than 50% anterior or posterior tracheal compression is found, the child should undergo a CTA or MRI. We believe that if the collapse is greater than 50%, these patients are more likely to have severe symptoms (possibly requiring surgery).

If the tracheal compression or collapse is less than 50%, the patient’s symptoms become more important in the workup. In patients with less than 50% collapse, we place more emphasis on symptoms to ensure we do not miss a vascular ring or sling in these patients. If the primary symptoms are chronic cough, cyanosis, and stridor as seen similar to that of a child with intrinsic tracheomalacia. These children will usually undergo operative bronchoscopy at some point owing to the severity of their symptoms, and this is the clinical decision point where the surgeon must determine whether to further image the child. One must remember that the decision to image for a potential vascular anomaly must be based on both the clinical symptoms and the findings on bronchoscopy. For example, 1 of the children in our study was found to have a right aortic arch on CTA, which required thoracic surgery, but the patient had not developed stridor and cyanosis by presentation. In addition, children may be quite symptomatic and have severe tracheal compression on bronchoscopy, but the CTA may underestimate the severity of innominate artery compression. Moës et al21 stressed that the decision for surgery should be based on the severity of symptoms and not just the degree of airway compression.21 Some CTAs must be performed while the patient is intubated or by using a laryngeal mask airway with positive-pressure ventilation, which likely stents the trachea open. Also, the CTA must catch the images in the expiratory phase to fully evaluate the amount of extrinsic compression. Although effort is made to image the trachea during expiration, a second acquisition of images to ensure the patient was indeed in the expiration phase is unlikely due to radiation risks.

Another important concern when evaluating children for potential vascular anomalies is knowing who would need thoracic surgery should a vascular anomaly

Figure 4. Diagnostic algorithm for a child with possible vascular anomaly. Short-segment compression is defined as 33% or less of the tracheal length; long-segment compression, more than 33% of the tracheal length. CTA indicates computed tomography angiography of the neck and chest; DLB, direct laryngoscopy with bronchoscopy; GERD, gastroesophageal reflux disorder; and MRI, magnetic resonance imaging.
in our patients with vascular anomalies that required thoracic surgery, these patients should undergo a CTA or MRI. If these symptoms are not present, one should treat other causes first, before considering additional imaging. Likewise, if the tracheal compression is long-segment type, involving greater than 33% of the tracheal length (as defined by the senior author C.J.H.), these patients should also be treated for other causes before further imaging. If patients are still quite symptomatic despite appropriate treatment, one should consider a CTA or MRI because vascular anomalies may compress a longer length of the trachea than is actually in contact with the vessel.15 If patients continue to improve after treating other causes, they should not be imaged.

Our study has a few limitations. It is a retrospective review, so bias was difficult to completely exclude. Although 42 children who underwent CTAs for suspected vascular anomalies were included in this study, only 6 required thoracic surgery. Moreover, we were only able to evaluate children who were determined to need bronchoscopy and a CTA, introducing selection bias of the senior author (C.J.H.), who made the determination. Due to the radiation and anesthesia risks in children, it is unfeasible to image every child who has findings suggestive of a vascular anomaly on bronchoscopy. Likewise, more self-limiting partial vascular rings, such as an aberrant innominate artery, may be found, which may not necessitate thoracic surgery.

In conclusion, children with vascular anomalies causing extrinsic compression of the airway often present similarly to those with intrinsic tracheomalacia. In light of recent well-documented concerns regarding increased cancer risk associated with CT and the risks of additional general anesthetics, one must determine the appropriate patient for additional imaging. Clinical symptoms and findings on bronchoscopy may predict who is likely to have a vascular anomaly or who may need thoracic surgery to correct it. We developed a diagnostic algorithm in an attempt to provide the best clinical care while weighing the risks of additional radiation vs additional sedation and exposure to general anesthesia.

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Author Contributions: Dr Hartnick had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Study concept and design: Rogers, Cunnane, and Hartnick. Acquisition of data: Rogers, Cunnane, and Hartnick. Analysis and interpretation of data: Rogers, Cunnane, and Hartnick. Drafting of the manuscript: Rogers. Critical revision of the manuscript for important intellectual content: Rogers, Cunnane, and Hartnick. Administrative, technical, and material support: Rogers, Cunnane, and Hartnick. Study supervision: Hartnick.

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REFERENCES


