Preoperative evaluation of the trachea in a child with pulmonary artery sling using 3-dimensional computed tomographic imaging and virtual bronchoscopy

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Abstract Pulmonary artery sling is frequently accompanied by tracheal anomalies other than local compression, including focal/segmental or extensive stenosis (ring-sling complex). Recent advances in computed tomography technology, such as multidetector computed tomography with 3-dimensional imaging (3-D) and virtual bronchoscopy (VB), offer important and accurate information of the entire tracheobronchial tree. We report on the value of 3-D imaging and VB in the preoperative assessment of a 4-year-old child with pulmonary artery sling and long segment tracheal stenosis owing to complete cartilaginous rings; we suggest that 3-D imaging and VB are important contributions in this assessment.

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The anomalous left pulmonary artery or pulmonary artery sling (PAS) is a rare congenital anomaly in which the left pulmonary artery arises from the right pulmonary artery and encircles the distal trachea, coursing between the trachea and esophagus to reach the hilum of the left lung. Pulmonary artery sling is often accompanied by tracheal stenosis that is either because of local compression by the arterial sling and malacia of the tracheal rings or because of short or long segment complete cartilaginous rings (up to 65% of cases, ring-sling complex) [1,2]; the presence of complete tracheal rings constitutes the major negative prognostic factor of this congenital malformation [2]. Wells et al have described 2 main types of PAS based on the level of the main carina: type I (vertebral bodies T4-T5) and type II (vertebral bodies T5-T6, associated with complete O-shaped cartilaginous rings, increased bifurcation angle, “bridging bronchus”) [3]. The preoperative evaluation of these patients includes [1-5] chest radiography, esophagography, echocardiography, fiberoptic bronchoscopy (FOB), computed tomography (CT) scan with use of contrast material, or magnetic resonance imaging (MRI); angiography clearly delineates the anomalous vessel but constitutes an invasive procedure and in most cases can be substituted by digital angiography and MRI. There are few publications on the application of recent advances in spiral CT technology, such as multidetector CT (MDCT), 3-dimensional (3-D) reconstruction, and virtual bronchoscopy (VB).
We report on the role of MDCT with 3-D reconstruction and VB in the preoperative evaluation of a child with PAS and tracheal stenosis.

1. Case report

A 4-year-old white girl presented with an episode of noisy breathing and respiratory distress. The plain anteroposterior chest x-ray raised the suspicion of distal tracheal stenosis and a wider-than-normal angle of the bifurcation of the mainstem bronchi. The patient was first evaluated at 5 months of age when she was admitted with tachypnea, chest retractions, inspiratory and expiratory monophonic wheezing, prolonged expiratory phase, “brassy” cough, and capillary hemoglobin oxygen saturation in room air of at least 95% associated with an upper respiratory tract infection. The diagnosis of primary or secondary tracheomalacia and/or bronchomalacia was considered at that time; but the parents declined further workup, and the patient was lost to follow-up until her second presentation at the age of 4 years. At least 5 similar episodes—two of which required admission—were diagnosed and managed as “viral croup.”

During her readmission, investigations included triplex echocardiography, cardiac catheterization, cardiac MRI, CT scan (including MDCT 3-D reconstruction and VB), and FOB. Cardiac MRI and catheterization demonstrated the anomalous left pulmonary artery arising from the right pulmonary artery—which evolved as a continuation of common pulmonary trunk—via a stenotic stem encircling the trachea posteriorly and enlarging into a normal-sized peripheral vessel. The trachea was evaluated under sedation by FOB and by MDCT with 3-D imaging and VB using a low-dose protocol (volume-rendering techniques via Sensation 16 scanner; Siemens Medical System, Erlangen, Germany).

The 3-D reconstruction demonstrated a long segment carrot-shaped tracheal stenosis that reached its smallest diameter at the main carina covering a length of 4.5 to 4.9 cm (Fig. 1); axial-source images helped quantitate the grade of tracheal stenosis. The shortest diameter of the C-shaped tracheal portion was 9.5 mm, and the cross-sectional area was 77.5 mm² (Fig. 1, line A); those of the O-shaped (stenotic) portion were 3.4 mm and 15.5 mm², respectively (Fig. 1, line B). The 2 mainstem bronchi formed a wider-than-normal angle (108°) and appeared larger than the distal tracheal segment (Fig. 1). Pulmonary lobulation and tracheal angulation were normal.

Fiberoptic bronchoscopy, under deep sedation, using a 3.6-mm fiberoptic bronchoscope (Olympus BF3C4, Tokyo, Japan) with video imaging revealed a long segment funnel-like tracheal stenosis that extended from approximately the entrance of the trachea into the thoracic cage, down to its bifurcation (Fig. 2A). Complete cartilaginous rings were visualized along the stenotic portion. The fiberoptic bronchoscope could not be passed through the stenotic segment; the orifices of the mainstem bronchi, partially visualized from a distance, appeared to be of normal size (Fig. 3A). The VB images confirmed the concentric tracheal stenosis (Fig. 2B), the O-shaped cartilaginous rings, and the normal lobar orifices (Fig. 3B, C). Based on the findings of preoperative evaluation, PAS repair and slide tracheoplasty were planned.

2. Discussion

Symptomatic patients with the ring-sling complex require surgical reimplantation of the anomalous vessel.
and restoration of the stenotic tracheal lumen during the same operative procedure. Surgery should be performed early after the diagnosis is made, especially in patients with stridor, apnea, or other symptoms of respiratory distress [9,10]. Many surgeons seek detailed preoperative knowledge of the anatomy of the tracheobronchial tree [1,2]. We report on the preoperative evaluation of a symptomatic 4-year-old girl with ring-sling complex ("stove-pipe trachea," "funnel-" or "carrot-like" stenosis) to undergo corrective surgery. We compare 3-D CT and VB with FOB findings of the trachea and mainstem bronchi.

Multidetector CT technology offers 3-D imaging of the trachea and its branches, and VB has made possible the accurate noninvasive acquisition of information by "navigating" through the reconstructed inner surface of the airways. It has been shown that the availability of axial, 3-D, and VB images raises the precision, accuracy, and sensitivity of radiological reports and provides detailed anatomical information that is more "user friendly" to the bronchoscopist and the surgeon [6]. In our patient, 3-D images that were generated using volume-rendering techniques helped measure the length of the stenosis and provided a detailed view of the trachea.

The present report is the first in the pediatric literature that shows VB images of the narrowed O-shaped trachea owing to ring-sling complex. Virtual bronchoscopy is a noninvasive method that can demonstrate the inner surface of the trachea, mainstem, and lobar bronchi. It is not time-consuming, and there is occasionally need for sedation in the pediatric population. Virtual bronchoscopy image manipulation is performed after the examination is concluded and offers flexibility in assessing the airways; in addition, it can provide the clinician with retrograde views of a stenotic airway without additional acquisitions. The disadvantages of the method, as compared with FOB, include

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**Fig. 2**  
A, Fiberoptic bronchoscopy image of the trachea. The tip of the bronchoscope is at the level of the normal-sized extrathoracic portion (C-shaped trachea). The posterior membranous wall is visualized (arrowhead). The narrowed O-shaped intrathoracic tracheal lumen is visualized distally (arrow). B, Virtual bronchoscopy image showing progressively worsening stenosis of the lumen.

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**Fig. 3**  
A, Fiberoptic bronchoscopy visualization of the carina and takeoff of the 2 mainstem bronchi. The orifices, seen from a distance, appear to be of normal size. B, Virtual bronchoscopy image obtained immediately above the carina at the level of maximal stenosis with partial visualization of the mainstem bronchial orifices. The virtual bronchoscope has bypassed the tracheal stenosis. C, Virtual bronchoscopy image of the right (R) and left (L) mainstem bronchial orifices.
**Table 1** Comprehensive review of publications of the use of VB in pediatric patients in the English literature

<table>
<thead>
<tr>
<th>Author (reference)</th>
<th>Year</th>
<th>No. of patients</th>
<th>Age (mean)</th>
<th>Clinical presentation</th>
<th>Imaging technique</th>
<th>Summary of results</th>
<th>Limitations and pitfalls</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Adaletli et al [11]</td>
<td>2006</td>
<td>37</td>
<td>4 mo-10 y (32 mo)</td>
<td>SFBA</td>
<td>Low-dose MDCT, VB</td>
<td>82% positive predictive value</td>
<td>Not all patients underwent CB. The surgeons who performed CB were aware of the findings of MDCT and VB.</td>
</tr>
<tr>
<td>2. Kocaoglu et al [12]</td>
<td>2006</td>
<td>21</td>
<td>8 mo-7 y (3.5 y)</td>
<td>SFBA</td>
<td>Thin-section axial MDCT, VB, MPR</td>
<td>88.9% sensitivity</td>
<td>VB did not provide additional information over MPR images.</td>
</tr>
<tr>
<td>3. Honnef et al [13]</td>
<td>2006</td>
<td>12</td>
<td>4 d-3 y (0.65 y)</td>
<td>Suspected tracheobronchial stenosis</td>
<td>16-slice MDCT, VB, VBG</td>
<td>11 of 12 stenoses were depicted by VB and VBG.</td>
<td></td>
</tr>
<tr>
<td>4. Briganti et al [14]</td>
<td>2005</td>
<td>25</td>
<td>Not reported</td>
<td>Tracheomalacia owing to esophageal atresia</td>
<td>CT, VB, VBG</td>
<td>VB confirmed CB findings</td>
<td></td>
</tr>
<tr>
<td>5. Kosucu et al [15]</td>
<td>2004</td>
<td>23</td>
<td>9 mo-13 y (3.3 y)</td>
<td>SFBA</td>
<td>Low-dose MDCT, VB</td>
<td>100% sensitivity</td>
<td>VB was unable to show the segmental and subsegmental tracheobronchial system.</td>
</tr>
<tr>
<td>6. Heyer et al [16]</td>
<td>2004</td>
<td>1</td>
<td>13 y</td>
<td>Tracheal bronchus</td>
<td>Low-dose MDCT, VB, CT, VB</td>
<td>VB confirmed CB findings</td>
<td></td>
</tr>
<tr>
<td>7. Haliloglu et al [17]</td>
<td>2003</td>
<td>23</td>
<td>8 mo-14 y (2.4 y)</td>
<td>SFBA</td>
<td>CT, VB</td>
<td>100% sensitivity</td>
<td>VB cannot detect endobronchial lesions smaller than 2-3 mm.</td>
</tr>
<tr>
<td>8. Sorantin et al [8]</td>
<td>2002</td>
<td>19</td>
<td>15 symptomatic: 5 wk-24.2 y (6.3 y)</td>
<td>Variety of reasons</td>
<td>VB compared with axial CT and MPR</td>
<td>Simultaneous display of axial CT, MPR, and VB:</td>
<td></td>
</tr>
<tr>
<td>9. Lam et al [7]</td>
<td>2000</td>
<td>11</td>
<td>1 d-5 y (2 mo)</td>
<td>Esophageal atresia and tracheal stenosis</td>
<td>3-D CT and VB</td>
<td>100% sensitivity</td>
<td>Small sample size</td>
</tr>
<tr>
<td>10. Burke et al [18]</td>
<td>2000</td>
<td>10</td>
<td>6 mo-17 y (6.22 y)</td>
<td>Upper airway obstruction</td>
<td>Helical CT, VB</td>
<td>VB evaluation was accurate in accessing degree and length of stenosis of fixed airway lesions.</td>
<td></td>
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<tr>
<td>11. Konen et al [19]</td>
<td>1998</td>
<td>8</td>
<td>6 mo-15 y (5.75 y)</td>
<td>Suspected compression or narrowing of the trachea and main bronchi</td>
<td>Helical CT, VB</td>
<td>VB imaging was feasible in all patients. Navigation with the virtual camera was accomplished to the level of at least the 3rd-generation bronchi.</td>
<td>Small sample size</td>
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</table>

MPR indicates multiplanar reconstructions; VBG, virtual bronchography; CB, conventional bronchoscopy; SFBA, suspected foreign body aspiration.

*a Limitations of VB applicable to all the studies are addressed in the discussion.*
its inability to provide texture and color details of the bronchial mucosa, overestimation of stenosis in the presence of thick mucus secretions in the lumen, as well as the radiation burden to the patient. Its inability to obtain biopsy specimens does not truly constitute a disadvantage in small children because biopsy is practically not feasible with the fiberoptic instrument either. The major advantage of FOB is that it provides the opportunity to assess dynamic changes of the airways during respiration (tracheomalacia or bronchomalacia) and to recognize external pulsation of an adjacent artery (vascular ring). A comprehensive review of the publications of the use of VB in the English pediatric literature is shown in Table 1. In the case of our patient, CT scanning was performed under a low-dose protocol; and a compromise was reached between image quality and scanning parameters, conforming to the As Low As Reasonably Achievable recommendation. Because of the patient’s age, sedation was required.

Three-dimensional imaging and VB enabled us to accurately assess the length and degree of the stenosis in planning the surgical repair of ring-sling complex. Unlike conventional bronchoscopy, VB “bypassed” the stenotic segment of the tracheal lumen; and normal-sized orifices of the main bronchi were clearly demonstrated. Therefore, we recommend 3-D imaging and VB as important tools in the preoperative planning of infants and young children with anomalous left pulmonary artery.

References