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Turner Syndrome with Aberrant Right Subclavian Artery and Partial Anomalous Pulmonary Venous Return

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A 6-year old female with Turner syndrome presented with recent onset of non-specific gastrointestinal symptoms and chest pain. An echocardiogram from the referring hospital was reported to be normal including a trileaflet aortic valve. Subsequently, an esophagram (Figure 1) was performed to evaluate for gastroesophageal reflux, which suggested the presence of a vascular ring (Figure 1A). The lateral view demonstrated a smooth indentation to the posterior upper thoracic esophagus suggesting an aberrant right subclavian artery (Figure 1B). A contrast-enhanced, high-pitch, 128-slice, ECG-gated dual source CT (DSCT) scan, performed at 218 mAs and 80kV (estimated effective radiation dose 0.25 millisieverts (mSv), confirmed the presence of a left sided aortic arch with an aberrant right subclavian artery (Figure 2, Movie I). In addition, there was an incidental observation of partial anomalous pulmonary venous return with the right superior pulmonary vein draining directly into the superior vena cava (Figure 3). No atrial septal defects were noted. No other cardiovascular abnormalities, including bicuspid aortic valve or aortic coarctation were noted.

Individually, these are relatively common congenital cardiovascular anomalies associated with Turner syndrome but are rarely observed in combination. A review of literature has shown only 2 other cases of Turner syndrome associated with aberrant right subclavian artery and partial anomalous pulmonary venous return as diagnosed by magnetic resonance angiography.¹ This case illustrates the value of using high-pitch DSCT in pediatric patients, where diagnostic images can be acquired with minimal radiation exposure below 1 mSv.

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Figure 1. Vascular ring. Esophagram in the anteroposterior view (A) demonstrates an oblique indentation to the superior esophagus (arrow). (B) Lateral view shows this indentation to be along the posterior wall of the esophagus (arrow) suggesting the presence of a vascular ring, most likely an aberrant right subclavian artery.

Figure 2.

Anatomic assessment of the aberrant right subclavian artery. Coronal (A) and axial-oblique (B) maximum intensity projection images of a DSCT scan of the heart, following IV contrast at the level of the aortic arch demonstrating a left sided aortic arch with an aberrant origin of the right subclavian artery which arises from the distal arch (A, arrow) and crosses to the right side of the chest behind the trachea and esophagus (B, arrow). Posterior view of the volume rendered 3-D image, (C) again showing the right subclavian artery arising from the distal arch.

Figure 3.

Partial anomalous pulmonary venous return demonstrated by maximum intensity projection images obtained on axial (A) and coronal oblique (B) views of a contrast enhanced DSCT scan of the chest, to show the right superior pulmonary vein draining anomalously into the posterior aspect of the superior vena cava (arrow).