A 33-year-old woman presented to our department with complaints of bluish discoloration of the body, thickening of the nail beds, and dyspnea on exertion for many years. She had history of repeated respiratory infections during infancy and early childhood. Her physical examination revealed cyanosis and clubbing with a pulse rate of 90 bpm, blood pressure of 128/80 mmHg, and left parasternal heave. Auscultation demonstrated a loud pulmonary component of the second heart sound and a short ejection systolic murmur over the second left intercostal space. Her arterial oxygen saturation was 82% at room air in all 4 limbs. Her chest radiograph showed a cardiothoracic ratio of 55% with a prominent main pulmonary artery segment (Figure 1A). Two-dimensional transthoracic echocardiography (TTE) revealed a large defect between the ascending aorta and the main pulmonary trunk, suggesting an aortopulmonary window (APW) (Figure 1B and 1C and Movies I and II in the online-only Data Supplement). Color Doppler showed bidirectional flow across this APW (Figure 1D and Movie III in the online-only Data Supplement). The entire aortic arch could not be visualized in the suprasternal view. Three-dimensional (3D) TTE further delineated the spatial geometry of the APW and its proximity to the 2 semilunar valves (Figure 2A–2D and Movie IV in the online-only Data Supplement). However, the issue of the arch was still not resolved. Computed tomography imaging was planned in view of the limitations of echocardiography. Computed tomography showed that this large APW created a common chamber comprising the ascending aorta and main pulmonary artery, and this chamber then gave rise to a right-sided common trunk that ultimately divided into the right brachiocephalic, left common carotid, and left subclavian arteries (Figure 3A–3D and Movie V in the online-only Data Supplement). There was an interruption of the aortic arch following that, and the descending aorta was filling through the patent ductus arteriosus originating from the pulmonary side of the common chamber. Both the right and left pulmonary arteries also originated from the pulmonary side of this common chamber. The differential computed tomography attenuation score helped to identify that the descending aorta was filling through the pulmonary artery and the common carotid trunk was filling through the ascending aorta. Thus, a diagnosis of APW with interrupted aortic arch with Eisenmenger syndrome was made. A conservative approach was planned for the patient.

APW is a rare form of congenital heart disease, accounting for 0.2% of all cardiac defects. The natural history of APW is similar to that of a large left-to-right shunt and congestive heart failure in early infancy. Survival of the infant can lead to severe pulmonary hypertension and Eisenmenger syndrome in the first or second decade. Associated cardiovascular anomalies are seen in more than half of all cases and include septal defects, interrupted aortic arch, tetralogy of Fallot, and aortic origin of the right pulmonary artery. Mori et al classified APW as proximal or type A, distal or type B, and total or type C defects. Our case represents type C or complete defect.

Interrupted aortic arch is defined as the complete separation of ascending and descending aorta. According to the classification of Celoria and Patton, interruption of arch can be type A if the interruption is distal to the left subclavian artery, type B if it is between the left subclavian and left common carotid arteries, or type C if it is between both carotid arteries. In our case, there was interrupted aortic arch type A. Diagnosis of APW can be made by 2-dimensional TTE, but it is delineated better by 3D TTE. In addition, 3D TTE is helpful in assessing the distance from the defect to the semilunar valves and the aortic origin of pulmonary artery and thus can help in the noninvasive assessment of APW and its classification. However, arch abnormalities cannot be accurately assessed with 3D TTE. Arch abnormalities can be identified with computed tomography and its 3D volume-rendered images. Cardiac catheterization is done to assess pulmonary artery pressure.

We have described a patient with a complete (type C) APW with interrupted aortic arch type A with Eisenmenger syndrome who presented in her fourth decade and was studied with multimodality imaging. It is rare for such a patient to present so late with a large APW.
Disclosures

None.

References


Figure 1. A, Chest x-ray, posteroanterior view, showing a prominent main pulmonary artery segment and an absence of cardiomegaly. B, Two-dimensional transthoracic echocardiography (2D TTE) in the modified suprasternal view showing a large aortopulmonary window (APW). C, 2D TTE in the modified parasternal short-axis view demonstrating an APW. D, Color Doppler in 2D TTE in the modified suprasternal view showing bidirectional flow across the APW. White arrows point to the APW. Ao indicates aorta; and PA, pulmonary artery.

Figure 2. Three-dimensional transthoracic echocardiography (3D TTE). A, Modified suprasternal view showing the aortopulmonary window (APW) and its spatial geometry. B, Tilted view of A showing both the aortic valve and the pulmonary valve and their relation to the APW. C, Modified parasternal short-axis view showing the APW. D, Color Doppler in the same view as A showing flow across the APW. White arrows point to the APW. Ao indicates aorta; AoV, aortic valve; PA, pulmonary artery; and PV, pulmonary valve.
Figure 3. A 64-slice computed tomographic angiogram.  
A, Three-dimensional volume-rendered image showing interrupted aortic arch (white arrow) with a common chamber, formed as a result of an aortopulmonary window (APW) and through which the common arterial trunk, the descending aorta (Dao), and both pulmonary arteries (PAs) originate. B, Cropped image in A showing that the descending aorta is filling through the patent ductus arteriosus (PDA; black arrow). White arrow points to the interrupted aortic arch. C, Transverse view showing the APW (black arrow). D, Transverse view at a higher location showing the common chamber and descending aorta filling through the PDA (black arrow). Ao indicates aorta; LPA, left pulmonary artery; and RPA, right pulmonary artery.
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**Movie Legend**

**Movie 1.** Two dimensional trans-thoracic echocardiography in modified suprasternal view showing the two great arteries and the aorto-pulmonary window. Best viewed with Windows Media Player.

**Movie 2.** Two dimensional trans-thoracic echocardiography in parasternal short axis view showing the aorto-pulmonary window between the proximal part of both great arteries. Best viewed with Windows Media Player.

**Movie 3.** Colour Doppler image in modified suprasternal view showing the bidirectional flow across the aorto-pulmonary window. Best viewed with Windows Media Player.

**Movie 4.** Three dimensional trans-thoracic echocardiography showing the three dimensional orientation of the aorto-pulmonary window and its proximity with the semilunar valves. Best viewed with Windows Media Player.

**Movie 5.** 64 slice computed tomography in transverse view showing both ventricles, both great arteries and the aorto-pulmonary window with the common chamber and the branches originating from it. Best viewed with Windows Media Player.