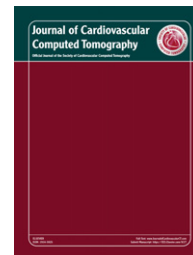




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Images in Cardiovascular CT

Multimodality imaging of an adult with Shone complex

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ABSTRACT

Shone complex is a rare combination of valvular or supra-avalvular aortic stenosis, supra-avalvular mitral membrane, parachute mitral valve, and coarctation of the aorta. This article presents an unusual case of a 40-year-old woman who presented with a history of progressive dyspnea. We describe the main imaging features and protocol used for visualizing this syndrome using various imaging modalities.

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A 40-year-old woman with Shone complex, a 4-part constellation of valvular or supra-avalvular aortic stenosis, supra-avalvular mitral membrane, parachute mitral valve, and coarctation of the aorta, presented with worsening dyspnea. The echocardiogram showed a 133-mm Hg peak gradient across the aortic valve (AV) (Fig. 1A). Planimetry, however, suggested moderate AV stenosis (Fig. 1B). The parachute mitral valve (MV) had mild stenosis. Aortic coarctation was present with a 40-mm Hg gradient (Fig. 2C and D).

To exclude any subvalvular or supra-avalvular membrane and to evaluate the aorta, the patient underwent an electrocardiogram gated, axial, 320-detector row cardiac CT. The MV showed parachute morphology (Fig. 3A and B). The basal

septum was thickened (16 mm), but no subvalvular obstruction was seen (Figure 3C; Supplemental Movie). No supra-avalvular membrane was detected. The bicuspid AV had an area of 1.2 cm² (Fig. 3D; Supplemental Movie). The narrowest diameter of the descending aorta was 16 × 12 mm (Fig. 2A). No thoracic arterial collateralization was seen.

The left heart catheterization showed an AV area of 0.96 cm² with peak gradient of 58 mm Hg. The coarctation had a gradient of 20 mm Hg (Fig. 2B). Collectively, the combination of the AV stenosis and the coarctation were felt to be the cause of the dyspnea. She underwent a percutaneous stent placement in the aortic coarctation (Fig. 4) and was discharged for routine follow-up.

Conflict of interest: The authors report no conflicts of interest.

Supplementary material for this article may be found at <http://www.CardiacCTjournal.com>.

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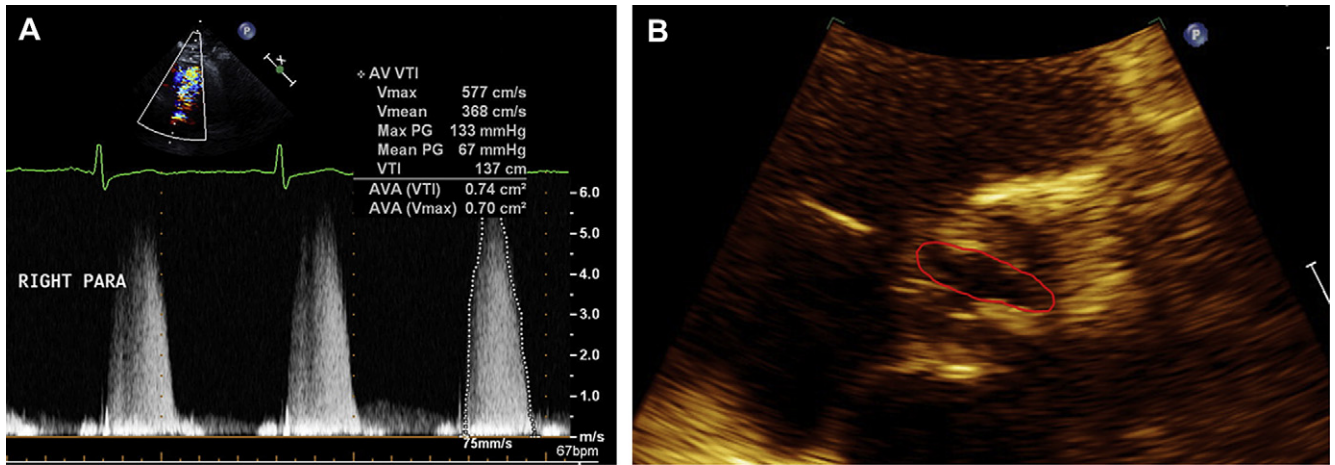


Figure 1 – Transthoracic echocardiogram. (A) Continuous wave Doppler from the left ventricular outflow tract shows a maximal velocity of 5.7 m/s, equivalent to a peak gradient of 133 mm Hg and a mean gradient of 67 mm Hg. (B) Aortic valve planimetry with an area of 1.2 cm².

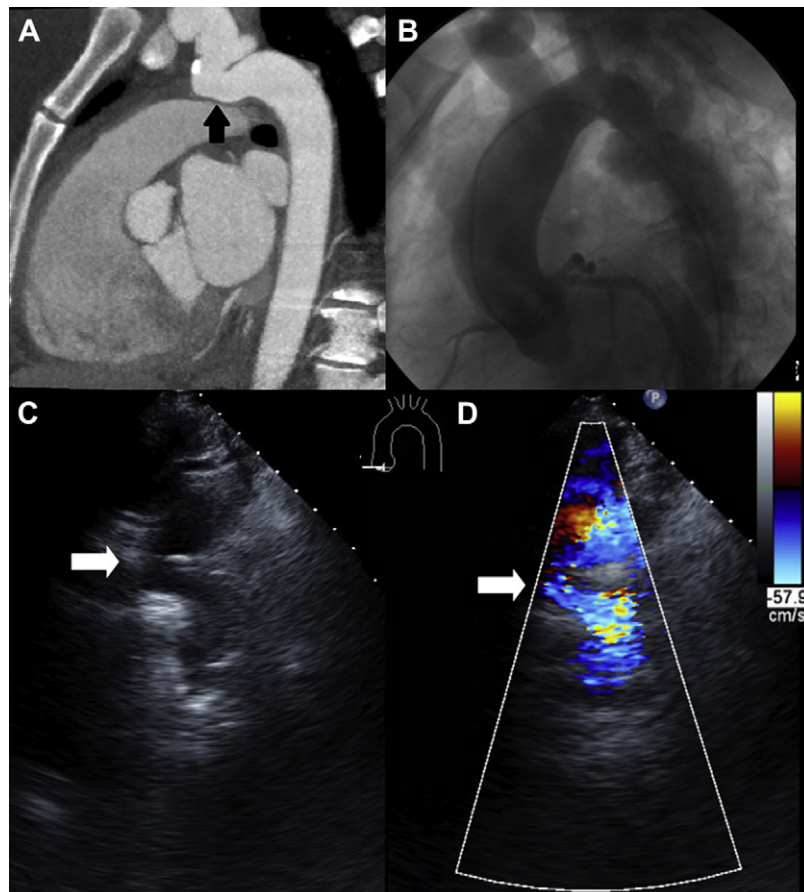


Figure 2 – (A) Shown is 5-mm maximum intensity projections (MIP) multiplanar reformat of the aortic arch. The cervical aortic arch extended superior to the manubrium, and a luminal reduction is associated with a tortuous segment and calcification of the distal aortic arch (arrow). (B) Invasive aortogram shows the same image as panel A. (C) Transthoracic image of the coarctation (white arrow). (D) Transthoracic image of the coarctation with color Doppler (white arrow).

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