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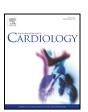
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Letter to the Editor

Giant left atrial appendage mimicking a mediastinal mass in a new diagnosis of atrial septal defect and pulmonic stenosis

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To the Editor:

We present the case of a 44 year old female, with a history of Down syndrome and a cardiac murmur diagnosed in childhood, who presented to our institution after she was hospitalized at an outside facility for new onset of heart failure. Her physical exam was significant for a Grade III/VI midsystolic murmur heard best at the left upper sterna border which accentuated with inspiration, a loud, fixed P2 component of her S2 heart sound. No significant right ventricular heave was felt upon examination. Mild digital clubbing without cyanosis was also seen. A 12-lead electrocardiogram was significant for right axis deviation and met criteria for right ventricular hypertrophy. A transthoracic echocardiogram revealed right sided enlargement with significant right to left interatrial shunting during agitated saline contrast injection, consistent with a possible atrial septal defect (ASD). Pulmonic valve stenosis was

also seen. A chest radiograph showed an enlarged cardiac silhouette and a soft tissue bulge along the left heart border concerning for a mediastinal mass. Dual source 128-slice cardiac multidetector computed tomography (MDCT) revealed a large, L-shaped left atrial appendage (LAA) extending over the left lateral aspect with no thrombus which correlated with the suspected mediastinal mass (Fig. 1). No coronary artery disease or anomalous anatomy was seen. A large secundum ASD was seen that was confirmed on subsequent transesophageal echocardiography (TEE); on TEE, the extent of the LAA could not be well seen (Fig. 2, Movie 1). The patient later underwent surgical closure of her ASD and LAA excision without complications.

Congenital cardiac defects are known to affect up to 50% of Down syndrome patients, warranting routine cardiac screening [1]. Cardiac computed tomography has been increasingly used in assessing LAA anatomy and thrombus with excellent diagnostic accuracy [2] and preplanning for percutaneous LAA closure [3]. In this situation, MDCT provided more accurate anatomic definition compared to TEE of the patient's LAA, excluded malignancy, and provided invaluable preoperative visual information for surgical repair.

Supplementary data to this article can be found online at http://dx.doi.org/10.1016/j.ijcard.2014.04.122.

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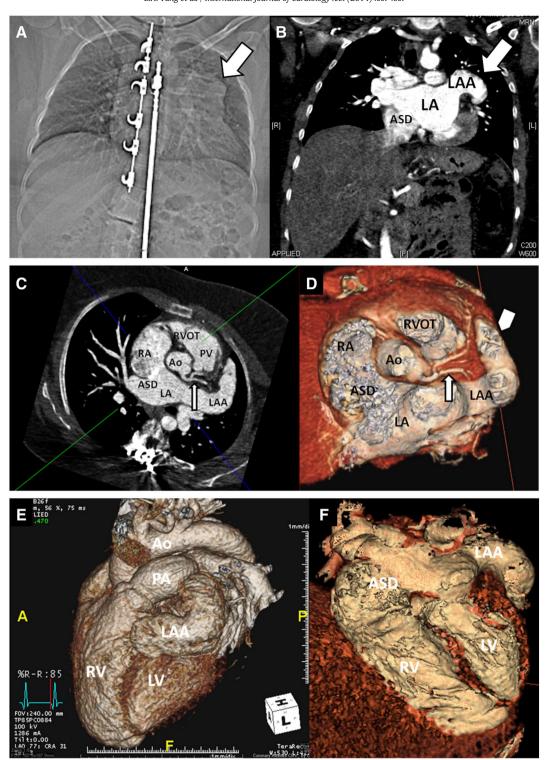


Fig. 1. Panel A: Cardiac computed tomography scout film and corresponding coronal view (panel B) showing the contour of the left atrial appendage (arrow) being responsible for the mediastinal abnormality seen on panel A (arrow). Part of the secundum atrial septal defect can be seen in panels C–D. Panel C: Dual source 128 slice multidetector cardiac computed tomography (MDCT), axial view. All relevant cardiac findings are seen in this view, with an L-shaped large left atrial appendage (LAA) measuring 95 mm in length. The superior aspect of the secundum atrial septal defect (ASD) is seen as well as diastolic doming of the pulmonic valve (PV). The left main coronary artery and proximal left anterior descending and circumflex artery are also visualized (arrow). Panel D: 3-D volume rendering reconstruction, short axis view of the aortic valve. Cross section of the LAA shows prominent pectinate muscles in this view (arrowhead). Panel E: Dual source 128 slice MDCT with 3 dimensional volume rendering, left anterior oblique cranial view of the large left atrial appendage (LAA) is seen. Panel F: Cross section of the heart showing the large secundum atrial septal defect and LAA. LA = left atrium, LAA = left atrial appendage, RA = right atrium, RV = right ventricle, RVOT = right ventricular outflow tract, PV = pulmonic valve, LV = left ventricle, PA = pulmonary artery, Ao = Aorta, ASD = atrial septal defect.

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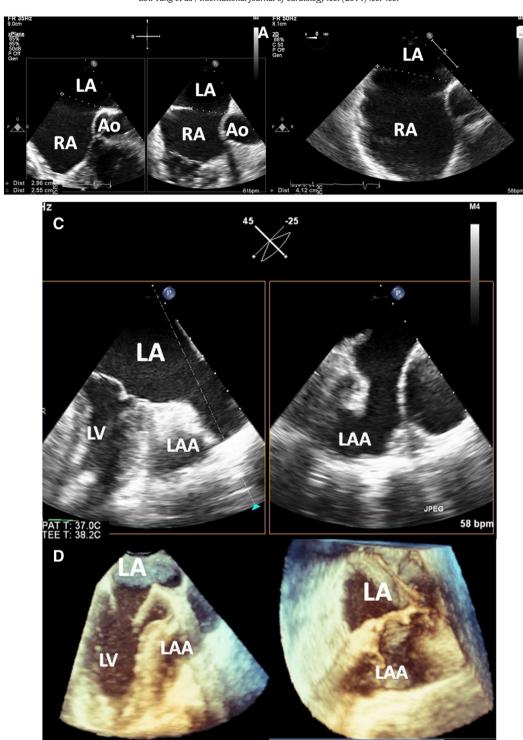


Fig. 2. Panels A–B: Transesophageal echocardiography (TEE) visualizing the secundum atrial septal defect (dotted line). A large secundum atrial septum defect is seen, measuring 41 mm × 29 mm × 33 mm with no retroaortic rim, warranting surgical repair. Panel C: 2-D TEE with biplane views visualizing the large left atrial appendage. LA = left atrium, LAA = left atrial appendage, LV = left ventricle. Panel D: Two views of 3 dimensional TEE visualizing the left atrial appendage showing the extension over the lateral portion of the left ventricle (left image), and visualizing the left atrial appendage from a caudal view of the left atrium (right image), showing the wide ostium of the left atrial appendage and its extent over the lateral portion of the left ventricle. RA = right atrium, LA = left atrial appendage, LV = left ventricle, Ao = aortic root.