Congenital absence of the left atrial appendage

A 3-month-old male child was referred to our tertiary-care center for respiratory distress. He was in very poor general condition and expired within 2 hours of admission. Before this referral, the child had been hospitalized twice in the recent past for similar complaint and had been administered antibiotics and inotropes. There were 3 two-dimensional echocardiographic reports which had suggested ostium secundum atrial septal defect (ASD) and large muscular ventricular septal defect in a setting of transposition of great arteries. There was an additional finding of left atrio-ventricular valvular atresia with a discrepancy in the type of ventricular looping (d-loop versus l-loop). A complete autopsy was requested. All organs were in *situs solitus* with normal bronchial morphology (eparterial right bronchus and hyparterial left bronchus). The cause of death was related to adenoviral interstitial pneumonitis with superadded confluent bronchopneumonia and diffuse alveolar damage. Detailed cardiac examination showed a single ventricle of right ventricular morphology with a double outlet [Figures 1 and 2], along with a large ostium secundum ASD and mitral valvular atresia. The aorta and pulmonary trunk were situated side-by-side with a right and left positions, respectively. The pulmonary trunk was larger than the ascending aorta. A hypoplastic left ventricle was not demonstrated. Interestingly, the left atrial appendage (LAA) was absent [Figure 1].

The appendage of the left atrium is the only portion that develops from the left wall of the primitive atrium as muscular finger-like projection.[1] Though it functions to some extent as a decompression chamber and elaborates the atrial natriuretic peptide, it has attained importance in adults in situations of non-valvular and valvular atrial fibrillation (AF).[1] In such patients, since the LAA serves as a trigger for AF and the site for thrombus, it is a seat for percutaneous or surgical techniques for LAA exclusion.[2] However, in children, its characteristic morphology plays a crucial role in the sequential segmental analysis used in the assessment of congenital heart disease (CHD), particularly when one encounters complex anomalies,[3] as seen in our case of single ventricle; however, the LAA was absent.

Absence of the LAA at its normal anatomical location (to the left of the root of the pulmonary trunk) has 3 distinct possibilities. The first is the congenital anomaly of right juxtaposition of the atrial appendages, where the LAA is on the right side, flanked by the right atrial appendage and ascending aorta. This is associated with complex cardiac malformations but is less common than left juxtaposition.[4] Another reason is spontaneous inversion of the LAA,[5] which also occurs with complex CHD. The final possibility is a true congenital absence (seen in the present case), which very surprisingly is seen as isolated finding in adults with fewer than 20 reports in literature.[6] To the best of our knowledge, this is the first true congenital absence of LAA being reported in a child with complex CHD.

**Declaration of patient consent**
The authors certify that appropriate patient consent was obtained.

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**Conflicts of interest**
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**Figure 1**: Heart and lung block in the fresh state, shows moderate cardiomegaly with side-by-side relationship of the great arteries. Note presence of single ventricular chamber (absence of clear-cut delineation of the interventricular septum by left anterior descending artery) and absence of left atrial appendage (LAA)*. [AA, ascending aorta; DTA, descending thoracic aorta; LCCA, left common carotid artery; LSA, left subclavian artery; PT, pulmonary trunk; RAA, right atrial appendage; RBCA, right brachiocephalic artery]

**Figure 2**: Single ventricle of right ventricular (RV) morphology with double-outlet (a) The right-sided outflow leading to the ascending aorta (AA); and (b) The left-sided outflow leading to the pulmonary trunk (PT) [AV, aortic valve; PV, pulmonary valve; TV tricuspid valve]
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