# VACTERL association with double-chambered left ventricle: A rare occurrence

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## ABSTRACT

**VACTERL association is a non-random association of birth defects of unknown etiology derived from structures of embryonic mesoderm. The common cardiac defects seen with VACTERL association are ventricular septal defects, atrial septal defects, and tetralogy of Fallot. We present a 2-year-old child with VACTERL association in whom we detected double-chambered left ventricle on transthoracic echocardiography.**

**Keywords:** Double-chambered left ventricle, VACTERL association, birth defects

## INTRODUCTION

VACTERL association is defined by the presence of at least three of the following congenital anomalies: Vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities.[1] The incidence is 1 in 10,000 to 1 in 40,000 live-born infants.[1] In 40-80% of patients with VACTERL association, cardiac malformations are reported.[1] The common cardiac defects seen with VACTERL association are ventricular septal defects, atrial septal defects, hypoplastic left heart syndrome, patent ductus arteriosus, transposition of the great arteries, truncus arteriosus, and tetralogy of Fallot.[2,3] We present a 2-year-old child with VACTERL association in whom we detected double-chambered left ventricle on transthoracic echocardiography, which has not been reported previously in this syndrome.

## CASE REPORT

A 2-year-old male asymptomatic child came for echocardiogram referred from an orthopedic center. His past history was significant for various limb and urachal anomalies of VACTERL association including bilateral radial aplasia, club hand, absent thumbs, sacral agenesis, and urachal sinus. At 9 months of age, he was diagnosed to have apical muscular ventricular septal defects and secundum atrial septal defect by echocardiography done elsewhere. On examination, he had normal development and normal intelligence for his age. There were dysmorphic facial features with bilateral radial club hand, absent thumbs, and short upper and lower limbs [Figure 1a]. Chest examination was normal with cardiac auscultation revealing regular heart sounds with no murmur. His ECG and Chest X-ray were unremarkable. Sacral X-ray showed partial sacral agenesis [Figure 1b].

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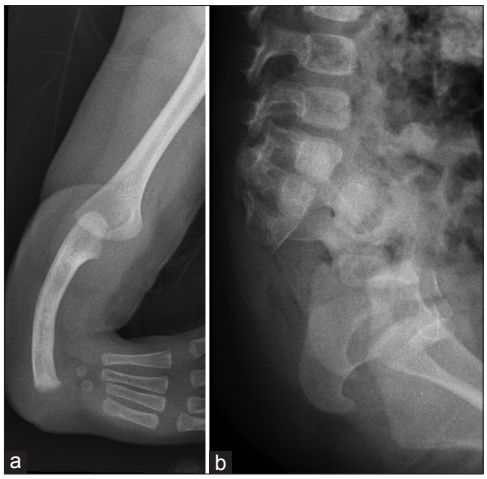


**Figure 1: X-ray of right upper limb demonstrating radial aplasia, ulnar bowing, absent thumb, and four metacarpals (a) and partial agenesis of sacrum (b) in a patient with VACTERL association and double-chambered left ventricle**

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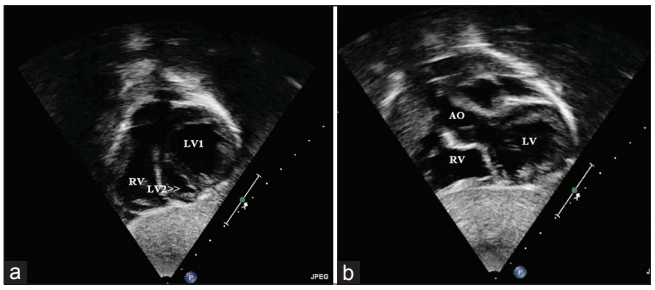


**Figure 2: Transthoracic echocardiography in short-axis inverted view showing a transverse muscle band traversing the left ventricle dividing the left ventricle into two halves producing a typical "figure-of-eight" appearance indicating a double-chambered left ventricle in a patient with VACTERL association (a), (b) Demonstrates the left ventricle in systole leading to aorta confirming double-chambered left ventricle. RV = right ventricle; LV = left ventricle chamber; LV1 = left ventricle chamber 1; LV2 = left ventricle chamber 2; AO = aorta**

Transthoracic echocardiography demonstrated situs solitus with normal atrioventricular and ventriculoarterial concordance. There were no ventricular septal defects. The left ventricle (LV) was abnormally configured with a muscle band extending from the interventricular septum toward the apex dividing the LV into two separate contracting LV chambers. A typical "figure-of-eight" appearance was seen in the parasternal short-axis view in diastole indicating double-chambered left ventricle (DCLV) [Figure 2a arrowheads, Video 1]. During systole, the LV was seen leading to aorta [Figure 2b]. There was laminar flow inside the LV with no intraventricular gradient. Rest of the cardiac examination was normal with good biventricular function.

## DISCUSSION

In this patient with VACTERL association, we detected DCLV, which is an uncommon association. DCLV was first conclusively described by Kay *et al.*, in 1983.[4] DCLV represents division of the LV into two chambers by abnormal muscular tissue similar to double-chambered right ventricle. However, the chambers of a DCLV are in parallel with no significant pressure gradient, as both chambers contract synchronously.[5] The pathogenesis of DCLV is less understood and it is thought to be congenital and non-progressive.[5] It was initially thought to be a cardiomyopathy,[4] but other authors describe it as a congenital anomaly due to abnormal septum or a muscle bundle across the LV between septum and left ventricular wall.[5-8] Congenital LV out-pouching like LV aneurysms or diverticula need to be differentiated from DCLV, and if in doubt, computed tomography scan, magnetic resonance imaging, or invasive angiography will aid in the diagnosis.[5-7] However, in this patient, there was clear separation of two chambers within the LV cavity which contract during systole. DCLV, by itself, may not require any treatment. If DCLV causes obstruction or tachyarrhythmias, then the muscle bundles need to be surgically excised.[5,6,8] B-blocker use or surgical resection has been reported to reduce the gradient if present.[8,9] In conclusion, this case report documents that patients with VACTERL association may rarely have a DCLV as the cardiac anomaly.

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