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Brief Report

Preoperative diagnosis of co-existing divided left atrium and tetralogy of Fallot

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Abstract We present the eighth published case of divided left atrium co-existing with tetralogy of Fallot. This is the first report of preoperative echocardiographic diagnosis of this unusual combination of defects. Demonstration of a partition within the left atrium is imperative for successful repair of the combined lesions. We draw attention to the need for careful echocardiography in patients where an obstruction to pulmonary venous drainage would dramatically affect the outcome subsequent to surgical correction.

Keywords: Cor triatriatum; pre-surgical diagnosis; long-term follow-up

DIVISION OF THE LEFT ATRIUM, OR "cor triatriatum", is a rare cardiac malformation in which the left atrium is divided into two portions. In the most common variant, one part receives the blood from the pulmonary veins, while the other includes the atrial appendage and the vestibule of the mitral valve. Even less common is the association of divided left atrium with tetralogy of Fallot. To the best of our knowledge, only seven cases have previously been recorded, and all have been diagnosed intraoperatively or at autopsy.[1–7] This is the first report of a preoperative echocardiographic diagnosis of this unusual combination of congenital cardiac malformations in a patient who went on to successful surgical correction.

Case report

A 4-month-old white male infant was admitted to Children's Hospital of New York-Presbyterian on October 6, 1992 for repair of tetralogy of Fallot. The patient was born full term at a community hospital via repeat caesarean section after an uncomplicated pregnancy. The weight at birth was 8 pounds and 1 ounce, between the 50th and 75th centiles. The Apgar scores were 8 and 9. Physical examination initially was normal, but on the second day of life, a cardiac murmur was noted, albeit that the infant was acyanotic. The patient was discharged home on the fifth day of life without a specific cardiac diagnosis.

The infant was first seen at the Children's Hospital of New York-Presbyterian for cardiac evaluation at two months of age. Growth and development had been normal, and there had been neither tachypnea nor difficulties with feeding. The weight was now 7.12 kg, in the same centiles, heart rate was 126 beats/min, respirations 32 per minute, and blood pressures 105/59 as measured in the right arm, and 104/71 in the right leg. The patient appeared well developed, well nourished, alert and in no distress. Cyanosis was noted with crying. The cardiac examination revealed a prominent right ventricular impulse, with a normal first heart sound and a single second heart sound. A grade 3/6 harsh systolic ejection murmur was audible over the left sternal border. The liver was palpable 1 cm below the right costal margin. The lungs were clear. The limbs were warm and well perfused, with no oedema. The level of haemoglobin was 14 g/dl, with the haematocrit at 39 percent, white blood cell count 12,000, and platelets 446,000.

The chest x-ray demonstrated cardiomegaly with prominent pulmonary vasculature and a left-sided arch. There was normal abdominal arrangement. The electrocardiogram showed normal sinus rhythm, at a

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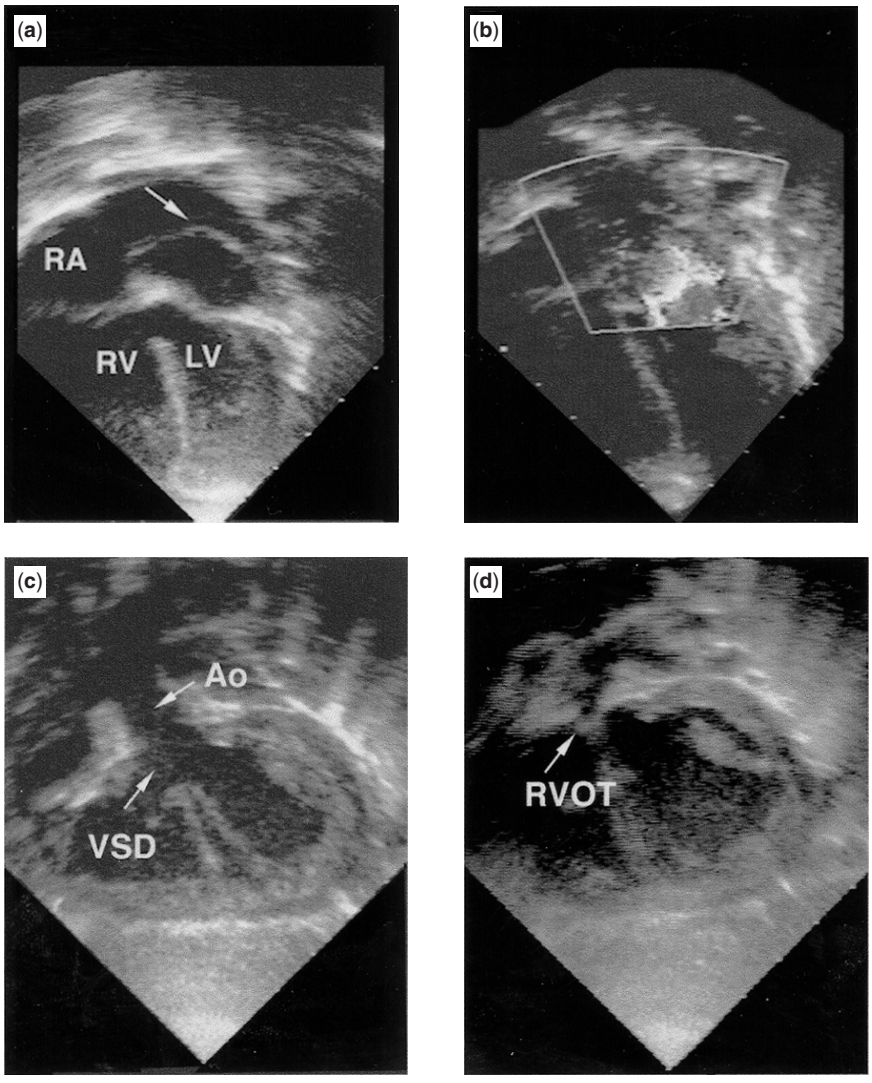

Figure 1.
*A preoperative echocardiogram demonstrates divided left atrium co-existing with tetralogy of Fallot. (**a**) Four-chamber view demonstrating the presence of a shelf in the left atrium (arrow). (**b**) Colour Doppler evidence of turbulence in the area of the left atrial shelf. (**c**) The ventricular septal defect (VSD) and overriding aorta (Ao) of tetralogy of Fallot. (**d**) The narrow right ventricular outflow tract (RVOT).*

rate of 154 beats/min, QRS axis of positive 105, and right ventricular hypertrophy. Echocardiography revealed usual atrial arrangement with right hand ventricular topology and normally related great arteries. A moderate sized subaortic ventricular septal defect (VSD) was identified, with colour Doppler evidence of left-to-right shunting. There was systolic doming of a thickened pulmonary valve. The diameter of the pulmonary valve was measured at 8 mm, while the left and right pulmonary arteries each measured 6 mm in diameter. A shelf was noted in the left atrium inferior to the orifice of the left atrial appendage, and the pulmonary veins were seen to be mildly dilated. The preoperative diagnosis, therefore, was tetralogy of Fallot with division of the left atrium (Fig. 1).

Surgical repair was carried out. A median sternotomy was performed and cardiopulmonary bypass was instituted. The aortic crossclamp time was seventy minutes. The entire operation to repair the tetralogy was performed through the tricuspid valve. The pulmonary outflow tract was enlarged, and the atrial septum opened in order to examine the left atrium. There was a thick shelf that separated the upper chamber, which received the pulmonary venous drainage, from the lower part of the left atrium. The shelf, which did not impinge on the normal mitral valve, was completely resected. Free flow from the pulmonary veins to the mitral orifice was thus established. The post-operative course was uneventful. The patient remained in the intensive care unit for three days, and was discharged on the tenth hospital day.

The child is currently 9 yr old, representing 8 and 3/4 yr follow-up relative to the surgical repair. He has done extremely well since surgery, with no symptoms of shortness of breath, fatigue, dyspnoea, cyanosis or exercise intolerance. He is a very active boy, with no limitations on his level of activity. Echocardiography now shows mild acceleration across the right ventricular outflow tract (RVOT), with a peak gradient

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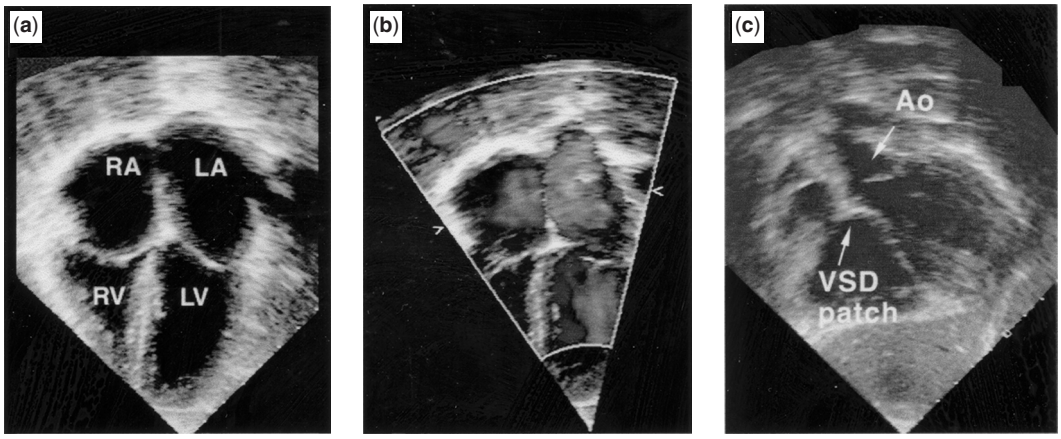

Figure 2.
*Post-operative echocardiogram. (**a**) Four-chamber view after removal of the left atrial shelf. (**b**) Colour Doppler evidence of laminar flow in the left atrium. (**c**) The angled patch closure of the VSD.*

of 4 mmHg and mild pulmonary insufficiency. The left atrium was unobstructed (Fig. 2). A progressive exercise test on the treadmill according to the standard Bruce protocol was normal for age, while a Holter monitor recording demonstrated sinus rhythm and occasional sinus pauses. There was no ectopy and no tachyarrhythmias.

Discussion

To the best of our knowledge, this is the first report of a pre-surgical diagnosis of tetralogy of Fallot associated with division of the left atrium (cor triatriatum). This rare combination of anomalies has previously been described in seven patients, with two being found at surgery and corrected,[2,4] and five diagnosed at autopsy.[1,3,5–7] In the past 16 years of echocardiographic diagnosis of divided left atrium, we could find no reports identifying a patient with concurrent tetralogy of Fallot by ultrasound examination.

The diagnosis of divided left atrium was often missed or delayed prior to the era of echocardiography. In the absence of murmur, the presence of pulmonary congestion on chest x-ray often led to suspicion of pulmonary parenchymal disease. Documentation of the divided atrial chamber was most often made at cardiac catheterisation on the basis of increased pulmonary capillary wedge pressure and increased pulmonary pressure with normal left ventricular end diastolic pressure. Demonstration of the divided left atrium by angiogram was difficult because of the overlapping of structures by contrast media. Only when the cineangiogram was recorded absolutely perpendicular to the intraatrial shelf would the diagnosis be made. Since the advent of cross-sectional echocardiography, however, the lesion can be readily diagnosed, especially in the presence of suggestive clinical findings. Obstruction to pulmonary venous drainage, still, may be difficult to diagnose in the presence of important associated defects. For example, the usual clinical signs of pulmonary venous obstruction and increased pulmonary venous pressure may not be apparent in the presence of tetralogy of Fallot. Pulmonary blood flow is decreased secondary to the pulmonary stenosis, and clinical and radiologic signs may be absent. When identified prior to surgery, successful repair of both defects can be carried out. Failure to diagnose the co-existence of the lesions prior to palliative or corrective surgery, in contrast, can result in death from pulmonary oedema, as reported in the cases identified at autopsy.[1,3,5–7] These patients died when surgical repair of tetralogy resulted in worsening of the previously unsuspected pulmonary venous obstruction associated with divided left atrium, since pulmonary blood flow was now unobstructed.

When pulmonary congestion is noted after creation of an arterial–pulmonary shunt in patients with the physiology of tetralogy of Fallot and undiagnosed division of the left atrium, pulmonary venous obstruction will often not be immediately suspected. Post-operative congestive heart failure will be attributed to an excessively large shunt, and surgical modification may be initially contemplated. It is important, therefore, also to consider the possibility of a left-sided obstructive lesion, especially if response to medical therapy is only marginal.[8] If the diagnosis of divided left atrium is not made preoperatively, further delay may result in a fatal outcome. Our case emphasizes the need for careful comprehensive echocardiography in patients with any congenital cardiac defect. Should an associated lesion be overlooked, the outcome may be profoundly affected.

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