

## Brief Report

# Twisted atrioventricular valves in double inlet left ventricle

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**Abstract** Twisted atrioventricular connections usually occur in hearts with biventricular atrioventricular connections. Here, we describe a case of twisted atrioventricular valves associated with double inlet left ventricle and discordant ventriculo-arterial connections. Color Doppler echocardiography, and cine magnetic resonance imaging, clearly demonstrated that the right atrioventricular valve was located anterior and superior to the left atrioventricular valve, and that the axes of the two atrioventricular valves crossed each other within the dominant left ventricle.

Keywords: Criss-cross heart; functionally univentricular heart; magnetic resonance imaging

THE HEART WITH TWISTED ATRIOVENTRICULAR connections represents a rotational abnormality in which the systemic and pulmonary venous streams cross at the level of the atrioventricular valves.<sup>1,2</sup> With a greater degree of twisting, the two blood streams from the atriums to the ventricles appear to “criss-cross”. Such twisted arrangements are seen mostly in hearts with biventricular atrioventricular connections. Recently, Kim et al.<sup>3</sup> reported four cases of twisted atrioventricular valves in the setting of double inlet right ventricle. In contrast, to the best of our knowledge, there is only one postmortem description of twisted valves with double inlet left ventricle.<sup>2</sup> Here, we report a patient with double inlet left ventricle in whom the twisted atrioventricular valves were revealed by color Doppler echocardiography and cine magnetic resonance imaging.

### Case report

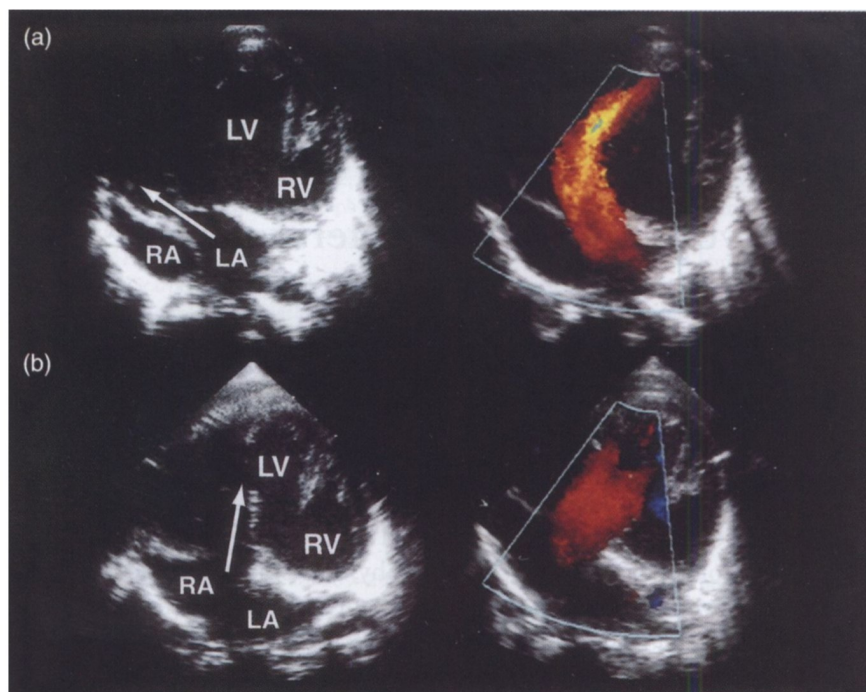
A 2-day-old female baby, weighting 2910 g, was admitted to our hospital because of cyanosis and a heart murmur. The oxygen saturation, as determined

by pulse oximeter, was 77%. On auscultation, a grade III/VI systolic murmur was heard along the left sternal border. The chest radiograph showed a left-sided heart, with a cardiothoracic ratio of 53% and slightly decreased pulmonary vascularity. Electrocardiographically, all precordial leads showed an RS pattern, and the QRS axis was 120°.

Cross-sectional echocardiography demonstrated double inlet left ventricle and discordant ventriculo-arterial connections, with usual arrangement of the atriums and thoraco-abdominal organs. The rudimentary right ventricle was seen to the left of the dominant left ventricle. Both atrioventricular valves opened into the right-sided morphologically left ventricle. In the short axis view of the ventricles, the orifices of the atrioventricular valves were antero-posterior to each other, rather than side-by-side. The pulmonary trunk arose from the right-sided left ventricle, and the pulmonary valve was stenotic. The anteriorly and leftward located aorta arose from the rudimentary right ventricle. The twisted atrioventricular valves were clearly demonstrated in the four-chamber view (Fig. 1), with the left atrioventricular valve located inferior to the right atrioventricular valve. The axis of the right atrioventricular valve was directed toward the apex of the heart, while that exiting from the left atrium extended more rightward. It was impossible to display the flow of blood from the two atriums in a single plane of the four-chamber

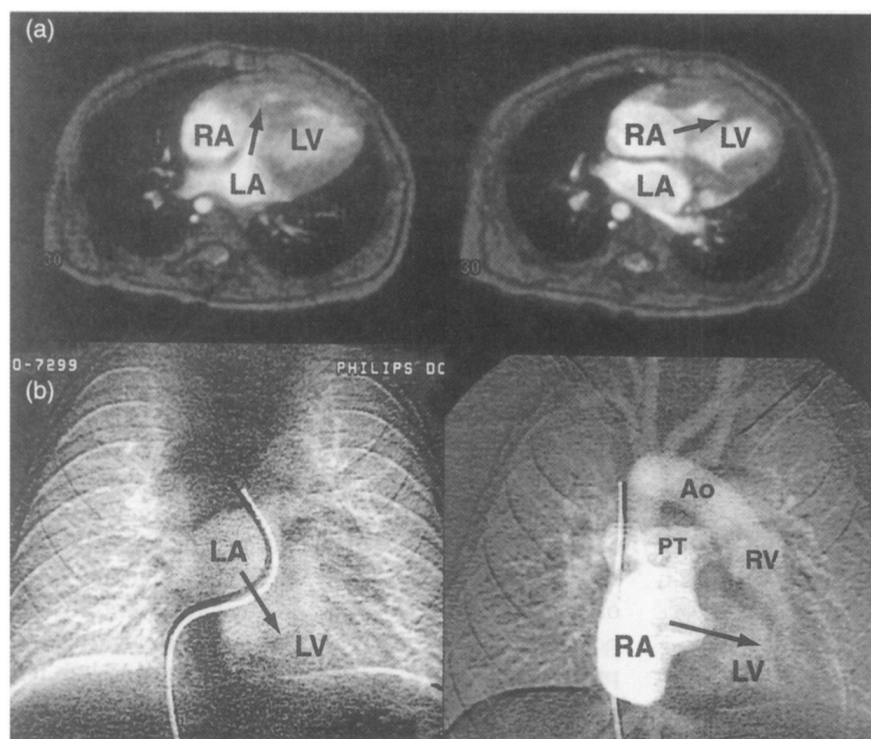
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**Figure 1.**

These transthoracic cross-sectional echocardiograms are taken in the four-chamber view. Panel (a) shows the left atrioventricular valve to be located inferior to the right atrioventricular valve. The axis of opening of this valve points rightward. In contrast, panel (b) shows the axis of the right atrioventricular valve to be directed towards the apex. The two atrioventricular valves have twisted relationships. The streaming of blood is confirmed by color Doppler echocardiography, as shown in the right panels. RA: right atrium; RV: right ventricle; LA: left atrium; LV: left ventricle.



**Figure 2.**

Transverse cine magnetic resonance imaging, as seen in panel (a), shows twisted atrioventricular valves (arrows). The left atrioventricular valve is inferior to the right atrioventricular valve. The atrial septum shows a curved configuration with its convexity towards the left atrium. The left panel (b) shows a levophase pulmonary arteriogram. The right panel (b) shows right atrioventricular arteriography. The leftward located aorta arose from the rudimentary right ventricle. The axes of opening of the two atrioventricular valves are twisted (arrows). Ao: aorta; PT: pulmonary trunk.

view. The atrial septum was abnormal, showing a curved configuration with the convexity toward the left atrium.

These findings were confirmed by biplane transesophageal echocardiography. In the longitudinal plane, color Doppler echocardiography clearly demonstrated flow of blood across the right atrioventricular

valve. Flow of blood from the left atrium, however, could not be visualized because the left atrioventricular valve opened almost perpendicularly to this plane.

Magnetic resonance imaging was performed with a 1.5-T unit when the baby was 5 months old. After anatomic cuts with electrocardiographically gated

spin-echo images, resonance images were obtained using the cine phase contrast method. The echo time was 8.2 ms, with a repetition time of 33 ms, a field of view of 240 mm, matrix size of  $256 \times 192$ , signal averages of 2, and slice thickness of 4 mm. In the transverse images, the left atrioventricular valve was inferior to the right atrioventricular valve, and their axes of opening were twisted (Fig. 2).

Cardiac catheterization and angiography conducted at 6 months of age confirmed the diagnosis of double inlet left ventricle, discordant ventriculoarterial connections, and pulmonary stenosis. The pulmonary arteries were well developed, and the pulmonary arterial pressure was 30/22 mmHg, with a mean of 24 mmHg. The oval foramen was closed. The directions of the contrast medium from the two atriums to the dominant ventricle were not parallel, but crossed each other (Fig. 2). At the age of 7 months, a hemi-Fontan operation was performed. The patient is doing well 5 months later, except for mild cyanosis with a pulse oximeter reading of 82%. She is waiting for completion of the Fontan circulation.

## Discussion

Usually, twisting of the atrioventricular connections occurs in a direction that places the morphologically right ventricle superior and anterior to the morphologically left ventricle. Looking at the heart from its apex, hearts with ventricular topology of right-hand pattern, or "d loop", usually are twisted in a clockwise direction, while hearts with left-hand ventricular topology, or "l loop", rotate in a counterclockwise direction.<sup>1,2</sup>

Kim et al.<sup>3</sup> reported 4 cases of double inlet right ventricle and twisted atrioventricular valves, all of which showed clockwise rotation. They predicted that this rule would also apply to double inlet left ventricle. So far, only one case of double inlet left ventricle and twisted atrioventricular valves has been described.<sup>2</sup> Our case is very similar to that one, in that the atrioventricular valves are in an anterior-posterior relationship rather than existing side-by-side, and that the rudimentary right ventricle is located just to the left of the left ventricle, not anteriorly. Because the right atrioventricular valve was anterior to the left atrioventricular valve, we believe twisting had occurred in a clockwise direction. This finding is in

contrast to cases with congenitally corrected transposition, in which twisting usually is counterclockwise. In our case, the atrioventricular connection remains double inlet despite the twisting. In other words, it is the atrioventricular valves which are twisted within the dominant left ventricle.

Although pulmonary arteriograms in the levophase are often necessary during angiocardiology to clarify the course of the stream of the blood from the left atrium to the ventricle, this sometimes remains obscure. On the other hand, both color Doppler echocardiography and magnetic resonance imaging can clearly demonstrate the cardiac segments, and determine the axes of opening of the atrioventricular valves.<sup>4-6</sup> Because magnetic resonance imaging provides a stack of truly parallel images, an abnormal spatial relationship of the atrioventricular valve is more accurately defined than is possible using cross-sectional echocardiography.<sup>6</sup> Cine magnetic resonance imaging demonstrates especially clearly the direction of the streams of blood through the atrioventricular valves.

Hearts with overriding or straddling atrioventricular valves, which are intermediate in the spectrum from biventricular to double inlet atrioventricular connections, are sometimes associated with twisted atrioventricular connections.<sup>2</sup> Our case confirms that twisted atrioventricular valves can also occur in the setting of double inlet left ventricle.

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