A full-term female newborn with prenatal diagnosis of ventricular asymmetry was referred to our department for postnatal echocardiographic evaluation. Two-dimensional echocardiography revealed a coarctation of the aorta (Fig. 1A), a bicuspid aortic valve (Fig. 1B) and a double-orifice mitral valve (DOMV) (Fig. 2A, Video 1). Three-dimensional echocardiography showed two distinct orifices of unequal size divided by a complete fibrous bridge (Fig. 2B). The smaller orifice was situated laterally (Fig. 2A and B). There was no mitral stenosis or insufficiency. The mitral annulus was of normal size and two papillary muscles were clearly distinguished. Surgical repair of the mitral valve was not performed. The coarctation of the aorta was treated surgically at day 7 with the conventional technique of resection and end-to-end anastomosis (Crafoord’s operation). The postoperative course was uneventful.

DOMV is a rare congenital anomaly, usually found in association with another congenital cardiac anomaly, such as atrioventricular septal defect (52%) or obstructive left-sided lesions (41%), but may also be isolated. A complete or partial fibrous bridge, defining the ‘bridge type’, separates the mitral orifice into two orifices, which are usually of unequal size; the medial orifice is usually larger. The ‘hole type’ is characterized by a secondary orifice in the lateral commissure of the mitral valve. Mitral stenosis and/or regurgitation occur in 60% of cases. The clinical presentation closely depends on the associated lesions. Mitral valve reconstruction is required only when mitral stenosis or insufficiency is significant. In such cases, a detailed anatomical description is necessary to guide the surgeon.
Figure 1. Two-dimensional echocardiographic views showing: (A) the coarctation of the aorta; (B) the bicuspid aortic valve in diastole. Ao: aorta; LA: left atrium; RA: right atrium; RV: right ventricle.

Figure 2. Double-orifice mitral valve seen: (A) on a two-dimensional parasternal short-axis view; (B) on a three-dimensional echocardiographic view. The mitral valve is completely divided into two orifices by an accessory bridge of fibrous tissue. The lateral orifice is smaller than the medial orifice. LO: lateral orifice; MO: medial orifice.

and can be assessed by two- and three-dimensional echocardiography.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.acvd.2010.09.007.