

## Images in Congenital Heart Disease

### Cantrell's syndrome

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**W**E PRESENT THE CASE OF A 3-MONTH-OLD dyspnoeic child who was referred with a large umbilical hernia. Clinical examination revealed indeed a large omphalocele (OP), with a pulsatile diverticulum (D) present at its superior border (Fig. 1). A systolic murmur, of grade 2 from 6, was also heard in the right parasternal area. The chest x-ray showed a moderate enlargement of the cardiac silhouette, which was in the middle of the chest, with signs of pulmonary hypertension (Fig. 2a). Echocardiography demonstrated the presence of a large unrestrictive ventricular septal defect, and a diverticulum (Div) extending from the apex of the left ventricle (LV) (Fig. 2b). The contraction of the diverticulum (Fig. 2c-d:diastole, d-systole) was synchronous with the rest of the ventricular muscle. Because of the impossibility to close the ventricular septal defect, and the size of the omphalocele, we did not undertake surgery. Five months later, he is still alive and doing well, albeit with his functional improvement almost certainly being secondary to increased pulmonary resistances. We did not observe any complications from the parietal defect.

Diverticulus differ from ventricular aneurysms by having a narrow communication with the ventricular cavity, the presence of all cardiac layers, and a synchronous contraction with the rest of the ventricle. The association of a congenital ventricular diverticulum, always apical, with a midline thoraco-abdominal defect was described by Cantrell and colleagues in 1958.<sup>1</sup> This uncommon syndrome usually involves projection of an apical portion of the

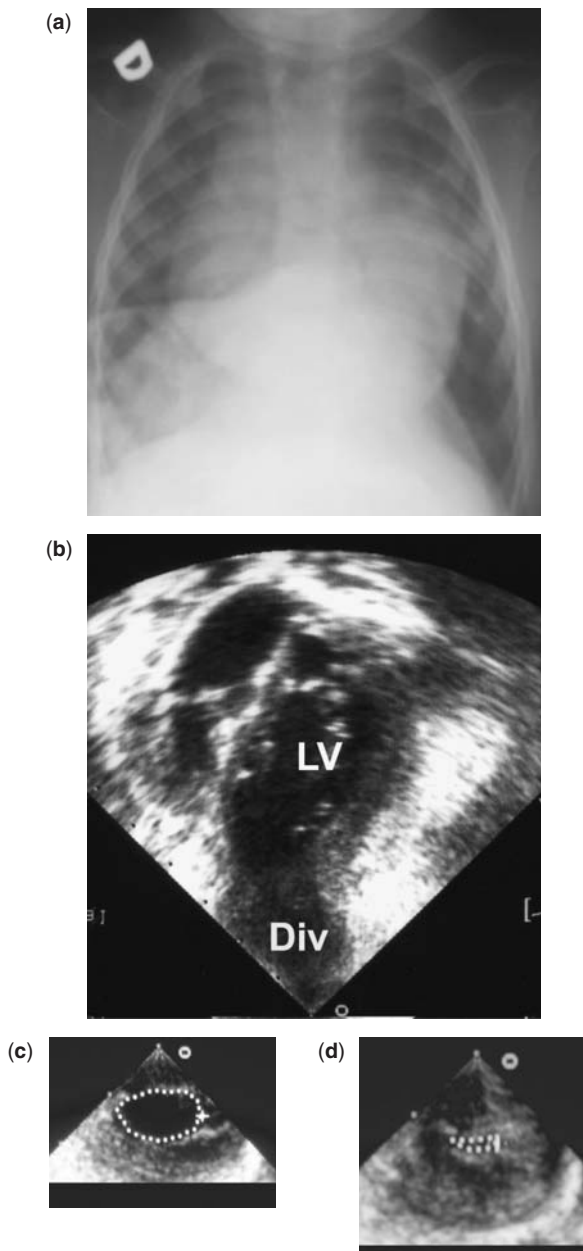


**Figure 1.** Typical clinical aspect of Cantrell's syndrome with a large omphalocele (OP) and a pulsatile diverticulum (D) originating of the apex of the heart.

ventricle through the weakened membranous part of the diaphragm, and an abnormal location of the heart. This syndrome is often associated with other cardiac malformations, the most common being a

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**Figure 2.**

*Chest x-ray showed a mesocardia and signs of pulmonary hypertension (a). Echocardiography demonstrated the presence of a large apical diverticulum of the left ventricle (LV) (b), which presented a synchronous contraction with the rest of the ventricular muscle: diastolic (c) and systolic (d) motion.*

ventricular septal defect. Our illustration showing the large ventricular diverticulum in the epigastric area as part of a giant omphalocele is exceptional.

### Reference

1. Vazquez-Jimenez JF, Muehler EG, Daebritz S et al. Cantrell's syndrome: a challenge to the surgeon. *Ann Thorac Surg* 1998; 65: 1178–1185.