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CARDIOVASCULAR FLASHLIGHT

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Heart with a trunk: form fruste of Cantrell's Syndrome

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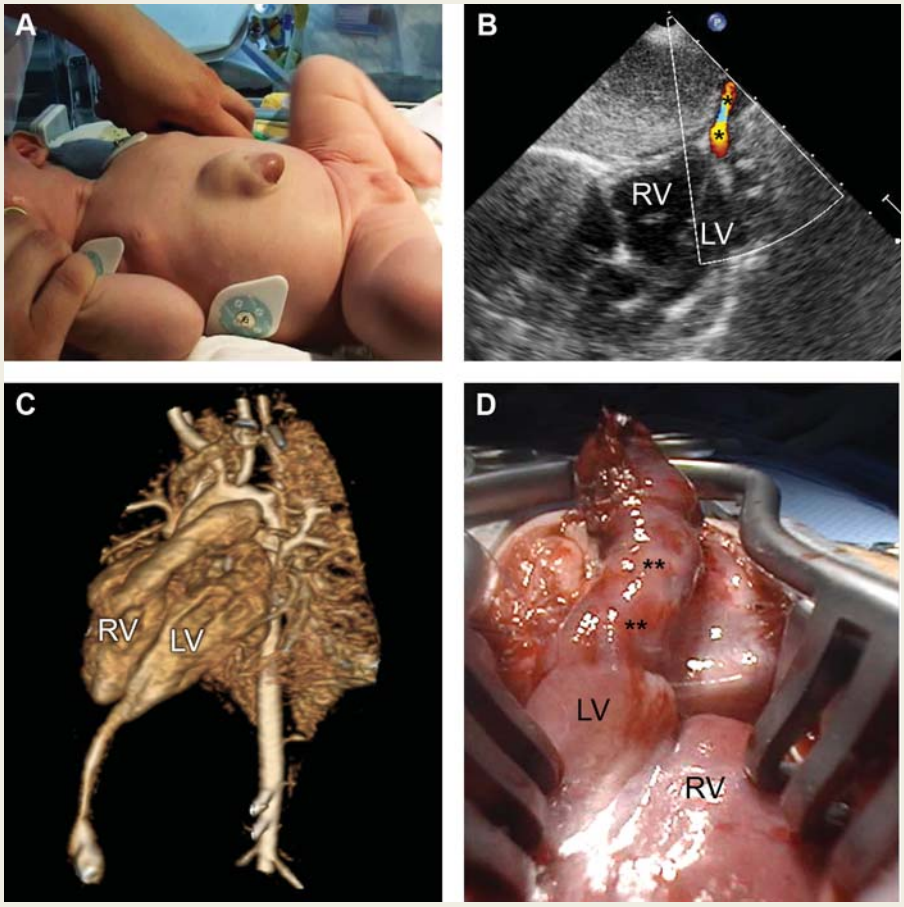

A newborn presented with a prominent pulsatile supraumbilical tumour which was initially diagnosed as umbilical hernia by the caring paediatrician. On referral, clinical examination showed a subcutaneous pulse-synchronous pulsatile mass with diastasis of m. rectus abdominis (*Panel A*). Echocardiography demonstrated situs solitus with laevocardia and a secundum atrial septal defect. The apex of the left ventricle (LV) continued into a funnel-like diverticulum running caudally and ending supraumbilically in a pulsatile bulbous vesicle (*Panel B*). These findings were confirmed by cardiac magnetic resonance (CMR), where the anatomical course of the LV diverticulum was clearly delineated (*Panel C*), and a defect in the anterior midline abdominal wall, just covered by the skin, visualized. Echocardiography and CMR demonstrated that the diverticulum was entirely covered by myocardium, and therefore pulsatile. Surgical resection of the LV diverticulum (*Panel D*) was performed without the use of cardiopulmonary bypass through a partial inferior sternotomy, at the age of 3 weeks. Overlapping reconstruction of the abdominal wall without the use of any prosthetic material was performed. Postoperative course was uneventful. The patient was discharged in good clinical condition and with a normal cardiac function.

Cantrell's pentalogy is a midline defect, including midline, suprapumbilical abdominal wall defects, a defect of the lower sternum, a deficiency of the anterior diaphragm, a defect of the diaphragmatic pericardium, and congenital heart disease. Clinical presentation can range from mild forms (fruste) similar to the case presented, to severe defects with omphalocoele, ectopia cordis, and more severe congenital heart defects, such as tetralogy of Fallot.

Panel A: Clinical picture showing a diastasis of the m. rectus abdominis and a subcutaneous supraumbilical mass.

Panel B: Echocardiographic subcostal view showing the apex of the left ventricle continuing in a funnel-like diverticulum (**). Colour Doppler demonstrates blood flowing into the diverticulum.

Panel C: 3DThree-dimensional reconstruction of cardiac magnetic resonanceCMR angiography delineates the entire course of the LVleft ventricular diverticulum.

Panel D: Intra-operative view of the LVleft ventricular diverticulum before surgical resection.