Cardiol Young 2002; 12: 284–285

ISSN 1047-9511

Images in Congenital Heart Disease

Contiguous but non-fused ventricles in conjoined thoracopagus twins

Colin J. McMahon, G. Wesley Vick, Michael R. Nihill

Lillie Frank Abercrombie Division of Pediatric Cardiology, Texas Children's Hospital and Baylor College of Medicine, Houston, TX, USA

Female thoracopagus twins were delivered at 37 weeks' gestation by caesarean section with a combined weight of 4000 g. They were electively intubated and ventilated at birth, and subsequently referred to our institution for cardiac assessment at three days of age. Cardiac catheterisation was performed on the fifth day of life to delineate the degree of cardiac fusion between the twins, in addition to the cardiac anomalies of each respective twin. The first twin, labelled "twin A", had a right-sided heart, with diverticulums from both left and right ventricles, but otherwise normal intracardiac anatomy. The second twin, labelled "twin B", had coarctation (coarct) of the aorta and a small perimembranous ventricular septal defect, with ventricles positioned superiorly and inferiorly (Fig. 1). The twins shared the pericardial sack, with muscular fusion of the hearts at ventricular level. There was also extensive hepatic fusion. Twin B underwent angioplasty of the coarctation, a 5 mm–2 cm balloon delivered via the umbilicus being

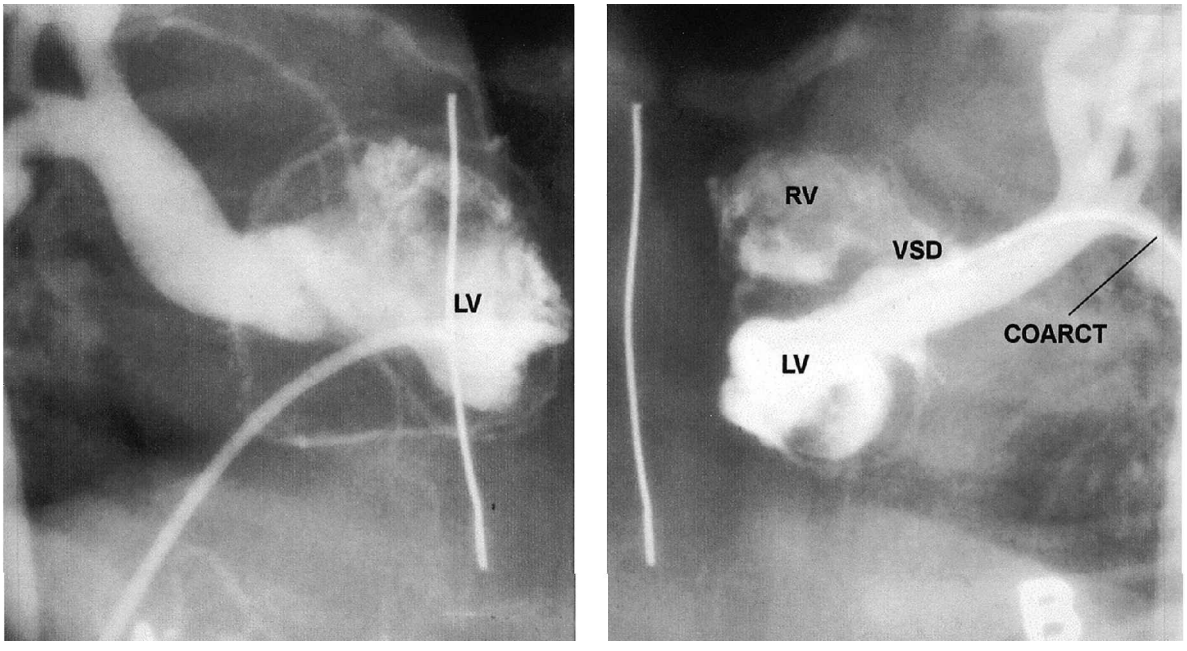

Figure 1.

Correspondence to: Colin J. McMahon MRCP (UK), Division of Pediatric Cardiology, Texas Children's Hospital, 6621 Fannin, MC 2-2281, Houston, TX 77030, USA. Tel: (832) 824 5659; Fax: (832) 825 5630; E-mail: cmcmahon@bcm.tmc.edu

Accepted for publication 23 August 2001

https://doi.org/10.1017/S1047951102000604 Published online by Cambridge University Press

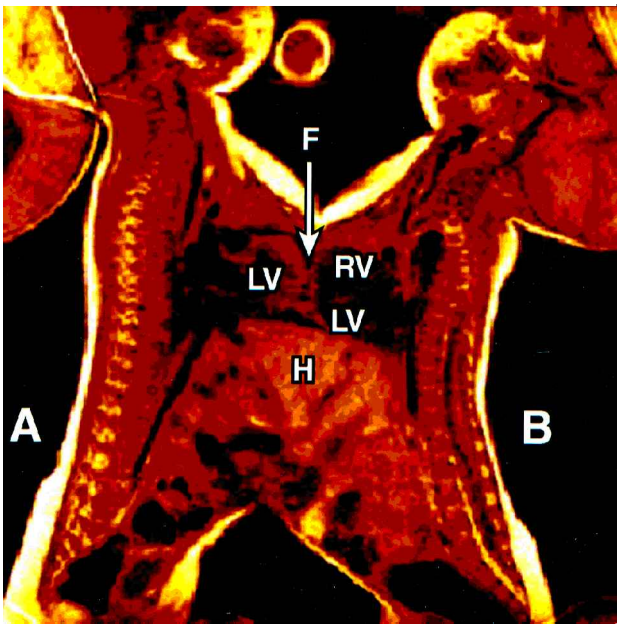

Figure 2.

inflated three times at the same catheterisation. This reduced the gradient from 20 to 7 mmHg. Repeated dilation at the age of 7 months, again with a 5 mm–2 cm balloon, produced a further reduction in gradient from 29 to 8 mmHg. Preoperative assessment confirmed fusion of the hearts at ventricular level, both on echocardiography and angiography. Magnetic resonance imaging of the thorax was performed (Fig. 2), which demonstrated two well-developed ventricles in each twin, again confirming the muscular fusion at ventricular level (F) together with extensive hepatic fusion (H) (Fig. 2).

The family agreed to proceed with surgical separation at 17 months of age. At surgical separation, the two hearts were found to be joined by a fibrous band of 1 cm diameter connecting the atrioventricular grooves. This was easily divided, allowing separation of both hearts without performing a ventriculotomy. Although there was no obvious plane of dissection between the ventricles preoperatively, the ventricles were found to be contiguous but not fused. The twins had a prolonged stay in intensive care, with twin A intubated for three weeks, and requiring tracheostomy for a further eleven months for chronic lung disease. The ventricular septal defect in twin B was restrictive, prompting treatment with digoxin and frusemide. These twins were alive and well on their last visit to the clinic 5 years following surgery. The case highlights the potential difficulties in determining fully the degree of cardiac fusion in the setting of thoracopagus twins, and distinguishing between external muscular fusion and contiguous but individual ventricles.

https://doi.org/10.1017/S1047951102000604 Published online by Cambridge University Press