*Cardiology in the Young* (2011), **21**, 235–237
doi:10.1017/S1047951110001770


*Brief Report*

# Pentalogy of Cantrell with double-outlet right ventricle: a case of surgical correction

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**Abstract** Pentalogy of Cantrell is characterised by a combination of severe defects in the middle of the chest including the sternum, diaphragm, heart, and abdominal wall. Mortality rate after cardiac surgery is usually high. We report a successful total correction of the cardiac defects in a case of Pentalogy of Cantrell with a double-outlet right ventricle prior to abdominal wall defect repair.

Keywords: Congenital cardiac defect; cardiac surgery; omphalocele

Received: 9 June 2010; Accepted: 31 October 2010; First published online: 21 December 2010

Pentalogy of Cantrell was first reported in 1958.[1] This rare disorder typically shows two or three of the birth defects like: a defect in the middle part of the abdomen above the belly button – omphalocele; a deficiency in the front part of the diaphragm; a defect in the lower part of the sternum – sternal cleft; a defect in the pericardium; and an intracardiac defect – often a ventricular septal defect. There are limited numbers of reports describing open cardiac surgery for this complicated disorder. Mortality rate after cardiac surgery is usually high in this disease.[2–8]

We describe a successful total correction of the cardiac defects in a case of Pentalogy of Cantrell with a double-outlet right ventricle associated with pulmonary stenosis.

## Case reports

A 37-week gestation female infant presented with a ventral wall hernia with abdominal contents and apex of the heart visualised beneath the skin covering. Ventilatory support was needed for 5 days. Echocardiography revealed a double-outlet right ventricle with d-malposed great arteries, large inlet ventricular septal defect, atrial septal defect, pulmonary stenosis, ventricular diverticulum, and dextrocardia. The ventricular diverticulum was positioned anteriorly and extended caudally through a diaphragmatic hernia (Fig 1). The ventricular diverticulum consisted of a thick cardiac muscular wall having sufficient contraction. Oxygen saturation was greater than 90% on room air when she was 2 months old; however, it gradually dropped to 80% in 6 months. Pre-operative cardiac catheterisation also revealed stenosis of the left pulmonary artery and a small persistent left superior caval vein accompanied by a normal-sized bridging vein. There was no other evidence of atrial isomerism. Complete repair was performed when she was 7 months old (5.4 kilograms in weight). There was no xyphoid process, and the sternum was short and thin. The diverticulum portion of the ventricle extended through a diaphragmatic hernia into the abdomen (omphalocele) and was severely adherent to the surrounding tissue. The pericardium had a partial defect and was adherent to the sternum. There was anti-clockwise rotation of the heart. Total cardiopulmonary bypass was established between the ascending aorta and both caval veins. Myocardial protection was achieved using moderate systemic hypothermia, topical cooling with iced saline, and antegrade cold blood cardioplegia, repeated every 20 minutes. A right

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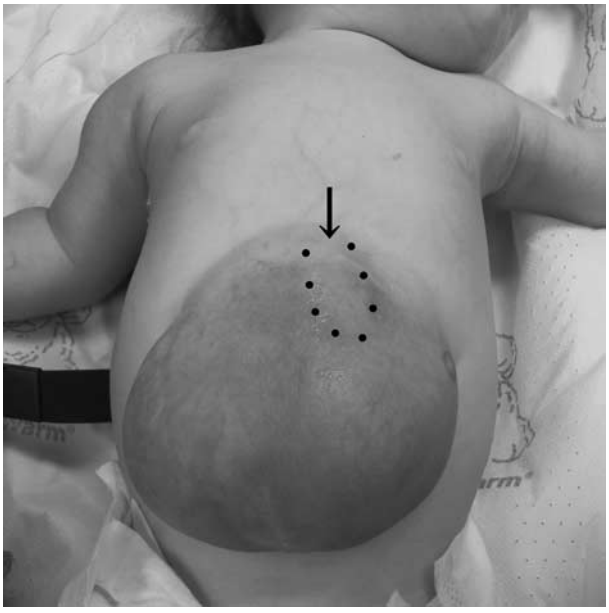

**Figure 1.**
*Large omphalocele. Ventricular diverticulum through a diaphragmatic hernia is shown by an arrow.*

infundibulotomy was made and a large inlet ventricular septal defect visualised. Intracardiac baffle repair was performed using a polytetrafluoroethylene patch. The left pulmonary artery was enlarged using a glutaraldehyde-treated autologous pericardium. The right ventricular size and the tricuspid valve were relatively small. A pulmonary homograft was implanted between the right ventricle and the pulmonary artery to provide normal haemodynamics to the right heart. The atrial septal defect was closed via a right atriotomy. A small left superior caval vein was ligated. The ventricular diverticulum and omphalocele were left unrepaired at this time and cardiac repair was covered with a polytetrafluoroethylene membrane. Post-operative course was uneventful. She was weaned from the ventilator 3 days after the surgery and discharged from the hospital 13 days post-operatively. Surgery for the omphalocele with/without resection of the ventricular diverticulum is being planned when the patient reaches two years of age.

## Discussion

The estimated incidence of the Pentalogy of Cantrell is 1 in 65,000 live births, according to the literature.[2] Ventricular septal defect is the most common cardiac defect (72%), followed by atrial septal defect (34.6%), left ventricular diverticulum (32.3%), and pulmonary stenosis or atresia (31.5%).[2] Takaya et al[5] reported a palliative surgery with a Blalock–Taussig shunt for a 5-month-old patient with a double-outlet right ventricle. To our knowledge, our case is the only surviving case after complete repair surgery for a double-outlet right ventricle associated with pulmonary stenosis.

Pentalogy of Cantrell represents high mortality and morbidity. In a literature review of 153 patients in 1998, Vazquez-Jimenez et al[2] reported that only 57 patients (37.3%) survived, 79 patients (51.6%) died after operation, and the outcome was not reported for 17 patients (11.1%). In a literature review between 1987 and 2007, Van Hoorn et al[4] also reported that 37 of 58 patients (63.8%), including patients in whom pregnancy was terminated, died within days after birth. O'Gorman et al[8] reported seven cases of Pentalogy of Cantrell who underwent cardiac surgery in 2009. They reported that post-operative care was complicated by the prolonged need for ventilatory support – average of 170.4 days, ranged 4–485 days – and multiple post-operative complications. Despite all patients surviving cardiac surgery, care was withdrawn from three patients due to failure to wean from the ventilator. Of the four patients who survived, two required ventilation by tracheostomy. In our experience, complete repair of the cardiac defects present in Pentalogy of Cantrell can be safely performed at an early age and prior to abdominal wall defect repair, which is often recommended at more than 2 years of age. Early extubation may be beneficial in the post-operative period and is associated with better outcomes. Strategies for surgical correction of the intracardiac defects as well as omphalocele might be important for this complicated disease.

Patients with Pentalogy of Cantrell often face complications with ventricular diverticulum (20–35%).[2,4] Resection of the ventricular diverticulum was recommended because of the risk of spontaneous or traumatic rupture and sudden death by tachyarrhythmia (6–20%);[2] however, our patient had a thick diverticulum wall and normal contraction. Korver et al[6] reported a case where the left anterior descending coronary artery extended towards the diverticulum. They preserved the proximal portion of the diverticulum at the time of cardiac surgery and concluded that complete resection of the diverticulum was not mandatory as there was normal contractility at the remnants of the diverticulum. Careful follow-up of the remaining ventricular diverticulum using an echocardiogram might be needed to determine further treatment.

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https://doi.org/10.1017/S1047951110001770 Published online by Cambridge University Press

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