Transposition with intact septum diagnosed at nine months: arterial switch?
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Transposition with intact septum diagnosed at nine months: arterial switch?

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Abstract
A 9-month-old infant presented with transposition of the great arteries and intact ventricular septum. His left ventricle was thin and deconditioned. He underwent an arterial switch operation and was electively supported with a left ventricular assist device for 8 days. He subsequently made a full recovery. Elective use of left ventricular support allows an extension of the age limit for the arterial switch operation.

Keywords
heart-assist devices, transposition of great vessels, vascular surgical procedures, ventricular function, left

Introduction
Transposition of the great arteries is best treated with an arterial switch operation within the first 3 weeks of life, before the left ventricle becomes deconditioned. A case of late presentation of transposition of the great arteries with a deconditioned left ventricle was treated with an arterial switch operation and elective support with a left ventricular assist device.

Case report
A 9-month-old infant from Iraq was admitted to the Royal Children’s Hospital in Melbourne, Australia, with transposition of the great arteries. He was clinically well and weighed 9 kg, the interventricular septum was intact, and good mixing of the blood could be demonstrated at the atrial level. The left ventricle was deconditioned: it had a thin wall and there was no gradient in the outflow tract. Cardiac magnetic resonance imaging (Figure 1) showed his left ventricular mass index to be $52 \text{ g} \cdot \text{m}^{-2}$. His parents rejected the offer to perform a Senning procedure, because of their uncertainties about providing adequate care in the forthcoming decades if the child deteriorated. A decision was taken to proceed with an arterial switch procedure. Before initiation of cardiopulmonary bypass, the measured mean pulmonary artery pressure was $8 \text{ mm Hg}$. At the end of the procedure, the patient was electively supported by a left ventricular assist device consisting of a Rotaflow centrifugal pump (Jostra Medizintechnik AG, Hirrlingen, Germany) placed between the left atrium and the aorta. He was weaned from the device after 8 days, and his chest was closed the following day. There were some episodes of pulmonary edema and he remained on mechanical ventilation for 25 days postoperatively. He was discharged from hospital 45 days after the operation. Magnetic resonance imaging (Figure 1) showed that his left ventricular mass index had increased to $61 \text{ g} \cdot \text{m}^{-2}$ at 35 days after surgery. At his 3-month follow-up visit, he had recovered normal ventricular function on echocardiography, with a shortening fraction of 33%.

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Discussion

Success with an arterial switch after a few months of age has been occasionally reported. In a series reported from Great Ormond Street Hospital, a 6-month-old infant underwent a successful switch operation, but his preoperative left ventricular pressures were found to be elevated at 80% of that of the right ventricle.1 

After 3 months of age, the Senning procedure is a viable alternative, but the progressive attrition observed in these patients after the second decade deters most families and practitioners from this option.2 The most crucial decision that the surgeon needs to take before performing a late switch operation in a child with a deconditioned left ventricle is whether to train the left ventricle with banding prior to the switch operation. While this strategy has been successful, the results of banding are often unpredictable; the patient may be unstable postoperatively, and may sometimes require the addition of a systemic-pulmonary shunt. Moreover, it has been demonstrated that these trained left ventricles may develop late diastolic dysfunction.3

A third option is to perform an arterial switch operation and then support the left ventricle postoperatively with a centrifugal pump.4,5 The cut-off age for this strategy was thought to be low. We have hereby demonstrated that, with elective left ventricular support, the age limit for an arterial switch can be extended up to 9 months. The life expectancy of this child is now back to normal. We believe that this approach may offer new prospects to children with a late diagnosis of transposition of the great arteries.

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Conflicts of interest statement

None declared.

References


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