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Brief Report

Spontaneous improvement of severe right ventricular dysfunction in the setting of hypoplasia of the left heart

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Abstract Right ventricular dysfunction is known to occur after the first stage of the Norwood sequence for treatment of patients with hypoplasia of the left heart. In a subset of patients, such ventricular dysfunction occurs without a specific anatomical cause. We describe two such cases with severe right ventricular dysfunction. In both cases, magnetic resonance imaging was used accurately to measure ventricular function and assess the arterial trunks. In both cases, cardiac transplantation was considered, but right ventricular function improved without invasive management. Transient right ventricular dysfunction in these cases may be due to the reduced ability of the right ventricle to adapt to the systemic vasculature. The improvement in ventricular function in these two cases may be due to delayed adaptation.

Keywords: Hypoplastic left heart; magnetic resonance imaging; right ventricular function

The use of the Norwood operation for palliation of patients with hypoplasia of the left heart has significantly improved prognosis in this condition.[1] Improvements in surgical techniques, as well as improvements in pre and perioperative management, have led to a reduction in immediate postoperative mortality.[2,3] Nevertheless, complications do occur late in a proportion of patients, and are associated with significant morbidity and mortality. Right ventricular dysfunction is the common end-point of complications such as pre-coronary stenosis, neo-aortic obstruction, and tricuspid regurgitation.[4,5] In a subset of patients, no anatomical cause is found to explain the ventricular dysfunction,[4] and it has been speculated that this is secondary to the right ventricle being "unprepared" to supply the systemic vasculature.[4] Such cases may be refractory to therapeutic interventions, and cardiac transplantation may be the only therapeutic option. Cardiac transplantation, however, is not without risk in children.[6] It is important, therefore, to assess the progression or regression of ventricular dysfunction before deciding to proceed to cardiac transplantation. Cardiovascular magnetic resonance imaging has become a useful tool in the assessment of patients with congenital cardiac disease.[7] It is able to accurately assess ventricular function, valvar regurgitation, and vascular obstruction. It is, therefore, an excellent modality for assessing patients with hypoplasia of the left heart. We present two patients with hypoplastic left hearts who had severe right ventricular dysfunction, sufficient to warrant assessment for heart transplantation, in whom right ventricular function spontaneously improved.

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Case 1

Our first patient was diagnosed antenatally with hypoplastic left heart syndrome. She underwent the Norwood procedure on the first day of life, which was complicated by right atrial thrombosis and a pericardial effusion. She was re-admitted 6 weeks after the Norwood procedure with signs of right ventricular failure. Cardiac catheterisation revealed no coronary arterial abnormalities, no angiographic obstruction of the reconstructed aortic arch, nor any pressure gradients. Ventricular function improved

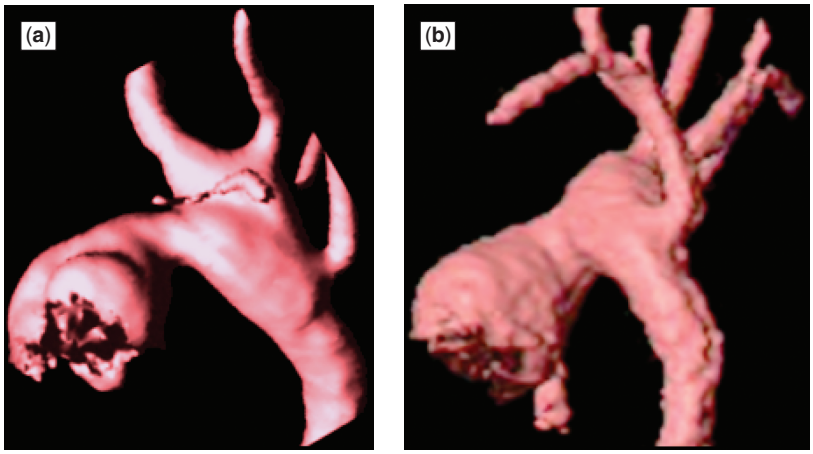

Figure 1.
*Gadolinium enhanced magnetic resonance angiogram of the aorta of our first patient showing proximal narrowing at the first scan, prior to balloon dilation of the aortic obstruction (**a**), and at the second scan when ventricular function had improved (**b**).*

slightly on medical management with captopril, frusemide and digoxin. She subsequently underwent a bi-directional Glenn procedure at the age of 2 months to reduce the volume circulating through the heart, with the hope of improving ventricular function. Post-operatively, right ventricular function was poor, and there was evidence of proximal obstruction of the transverse aortic arch on echocardiographic and magnetic resonance imaging (Fig. 1). Right ventricular ejection fraction, measured using short axis, multi-slice, multi-phase, steady state free precession imaging, was 28 percent. She therefore underwent balloon dilation of the neo-aortic arch. At catheterisation, a narrowing measuring 5 millimetres in diameter was visualised in the proximal transverse arch, and there was a systolic pressure gradient of 9 millimetres of mercury. Unfortunately, right ventricular function was still depressed after this procedure. Doppler echocardiography revealed a pressure drop of 21 millimetres of mercury across the narrowed area and the aortic arch. At this stage, we considered two therapeutic options, intervention on the slight narrowing of the aortic arch or cardiac transplantation. Further opinions were sought on the optimal management. It was felt that the degree of narrowing of the ascending aorta was not sufficient to account for the degree of right ventricular dysfunction, and that transplantation rather than interventional cardiac catheterisation would be more appropriate. The parents declined the option of transplantation because of the associated mortality, and the child continued on medical therapy. Over the following 9 months, there was a spontaneous improvement in right ventricular function as judged echocardiographically. Repeated magnetic resonance imaging demonstrated a right ventricular ejection fraction of 55 percent, with no tricuspid regurgitation (Fig. 2). There was still mild obstruction at the level of the proximal transverse arch, which now measured 12 millimetres in diameter, although there was some improvement in the appearance. The velocity across the narrowing, measured using phase contrast magnetic resonance, was 1.4 metres per second.

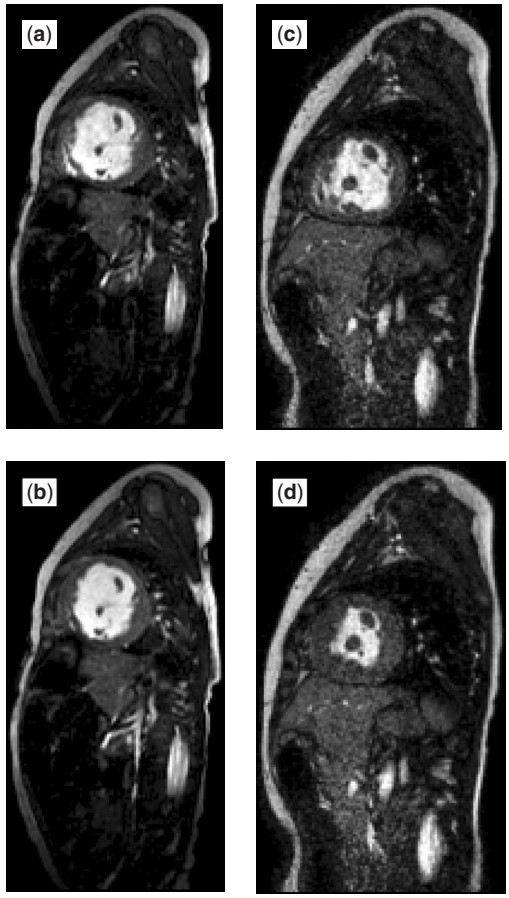

Figure 2.
*Steady state free precession end systolic and diastolic images profiled in short axis. End diastolic (**a**) and systolic (**b**) images at the first scan of our first patient. End diastolic (**c**) and systolic (**d**) images at the second scan.*

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The patient is currently well at the age of 2 years, with good right ventricular function, and is undergoing assessment for total cavo-pulmonary connection.

Case 2

Our second patient was also diagnosed antenatally with hypoplastic left heart syndrome. He underwent the Norwood procedure on the third day of life, which was complicated by a pericardial effusion. He was discharged 12 days post-operatively, at which time an echocardiogram showed good ventricular function and no neo-aortic obstruction. At the age of 3 months, he was re-admitted with physical and echocardiographic signs of right ventricular failure. At this time, he underwent cardiac magnetic resonance imaging to assess ventricular function, and to look for any evidence of neo-aortic obstruction. This revealed very poor right ventricular function, with the right ventricular ejection fraction measured at 25 percent, with no evidence of neo-aortic obstruction or significant tricuspid regurgitation. He was treated with captopril, frusemide, and digoxin. He was referred for cardiac transplantation, and placed on the waiting list. While on the active list, however, over the course of 5 months there was echocardiographic evidence of improving ventricular function. This improved to such an extent that he was deemed too well for cardiac transplantation, and was removed from the list. He went on to have the hemi-Fontan procedure at the age of 9 months, with no significant complications. Currently, he is well at the age of 16 months, with good right ventricular function on echocardiography.

Discussion

We report 2 patients with severe right ventricular dysfunction in whom ventricular function improved without invasive therapeutic interventions. This is an important finding, implying that in a sub-group of patients with hypoplasia of the left heart, cardiac dysfunction is a transient phenomenon, not requiring catheter or surgical intervention. In both cases, we considered cardiac transplantation. Cardiac transplantation in children is limited by difficulties acquiring suitable donor organs, significant post-operative morbidity and mortality, and the limited lifespan of the transplanted heart.[6,8] In our patients, with hindsight, it is evident that cardiac transplantation would not have been the best option at this age. In the assessment of such children, the possibility of spontaneous improvement of ventricular function should be considered and balanced against the risk of transplantation.

The aetiology of right ventricular dysfunction in these cases is unknown. It may be secondary to the reduced ability of the right ventricle to adapt to the systemic vasculature.[9] The improvement in ventricular function in our cases may be due to delayed adaptation.

Cardiac magnetic resonance has an important role in assessment of patients with hypoplasia of the left heart. It is able accurately to delineate the anatomy of the arterial trunks, to quantify tricuspid regurgitation, and to assess ventricular function. In both our patients, information from cardiac magnetic resonance imaging made an impact on decisions concerning management.

Ventricular dysfunction in patients with hypoplasia of the left heart, therefore, can be a transient phenomenon. Cardiac magnetic resonance imaging is an excellent modality with which to assess this problem.

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