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Double outlet right ventricle with infective endocarditis

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ABSTRACT Double outlet right ventricle (DORV) is an uncommon congenital heart disease with a poor prognosis. We report a rare case of a girl with untreated DORV who survived until adolescence, but then developed infective endocarditis with florid complications and succumbed to it. Although infective endocarditis is seldom encountered in children in developed countries, a high index of suspicion is required for those with congenital heart disease. The roles of cross-sectional imaging are demonstrated, including the usefulness of magnetic resonance imaging not only in evaluating congenital heart disease, but also in detecting incidental lesions in the extracardiac structures.

Keywords: cerebral embolism, double outlet right ventricle, infective endocarditis, pulmonary embolism
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INTRODUCTION

Double outlet right ventricle (DORV) is an uncommon congenital heart disease with a poor prognosis. We report a rare case of a girl with untreated DORV who survived until adolescence, but later developed infective endocarditis with florid complications.

CASE REPORT

A teenage girl from a neighbouring country was diagnosed in childhood with DORV and patent ductus arteriosus (PDA). She had not previously undergone corrective or palliative surgery. She was initially asymptomatic but started to have decreased effort tolerance in the past year. Seven months prior to admission, she had intermittent fever following a dental procedure. The fever abated transiently with oral antibiotics, only to recur every time the medication was stopped. This continued for several months until she deteriorated acutely and was admitted to a local hospital. She presented with breathlessness, headache and drowsiness. Following admission, she had generalised tonic-clonic seizures. She was intubated, ventilated and subsequently transferred to our institution.

On examination, the patient was cyanosed and clubbed. There was a parasternal heave as well as a displaced apex beat. The second heart sound was single and there was a grade 4 systolic murmur over the upper left sternal edge. There was also an early diastolic murmur over the upper right sternal edge associated with a bounding pulse and prominent carotid pulsation. The patient had hepatosplenomegaly. There were no other peripheral stigmata of infective endocarditis. Her right pupil was dilated and non-reactive to light, while her left pupil was reactive. She had increased tone in her lower limbs, and had right lower limb hyperreflexia with an upgoing plantar response. No facial dysmorphism was noted. Her total white cell count was 22.1 × 10^9/L (93.3% neutrophils), C-reactive protein (CRP) level was 119 mg/L and erythrocyte sedimentation rate (ESR) was 27 mm/hr. Pan-sensitive *Streptococcus* and coagulase-negative *Staphylococcus* were grown from the blood cultures.

Two-dimensional echocardiography showed DORV with sub-aortic ventricular septal defect (VSD) and pulmonary stenosis, as well as a PDA with left-to-right flow. Large vegetations, noted on the aortic and tricuspid valves (measuring 10 mm × 13 mm and 5 mm × 6 mm, respectively), were associated with severe aortic regurgitation and moderate tricuspid regurgitation (Figs. 1 & 2). This confirmed the diagnosis of infective endocarditis (IE). Cardiac magnetic resonance (MR) imaging was performed to fully delineate the underlying cardiac anatomy, as there was limited acoustic window for echocardiography. The above findings were confirmed, and the large vegetation on the aortic valve was clearly seen on MR imaging (Fig. 3). Incidentally, a filling defect in the mid to distal left pulmonary, in keeping with pulmonary embolus, was noted. There were also multiple peripheral opacities in both lungs, indicating septic emboli, some of which showed cavitation (Fig. 4).

Computed tomography of the patient's brain revealed multiple hypodense lesions in the brain in keeping with infarcts (Fig. 5), and further evaluation with MR angiography demonstrated the presence of emboli in the mid to distal basilar artery (Fig. 6). Cytogenetic and fluorescence *in situ* hybridisation analyses were done, which showed normal female karyotype with no deletion of the *TUPLE1* gene at 22q11.2, hence excluding DiGeorge syndrome and major chromosomal abnormalities.

Despite maximal medical support in the intensive care unit, including mechanical ventilation, inotropic support and appropriate antibiotic therapy, the patient succumbed to the disease before surgical intervention was possible.

DISCUSSION

DORV is a heterogeneous group of conotruncal abnormalities in which both the great vessels arise entirely or predominantly from the right ventricle.[1] The location of the VSD is the basis for

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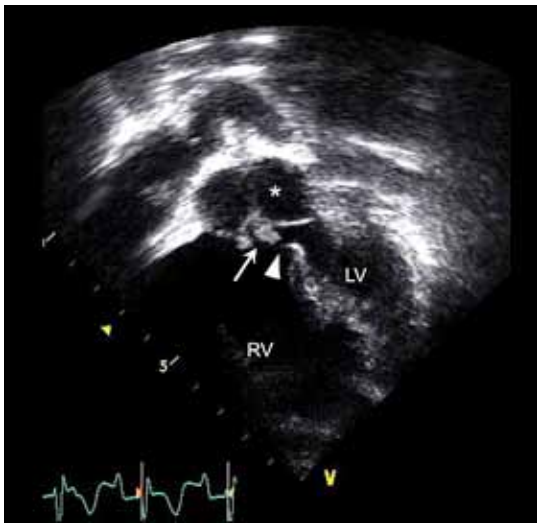

Fig. 1 Echocardiography image of a 5-chamber view of the heart shows a large ventricular septal defect (arrowhead) with an overriding aorta (*). A vegetation is seen on the aortic valve leaflet (arrow). LV: left ventricle RV: right ventricle

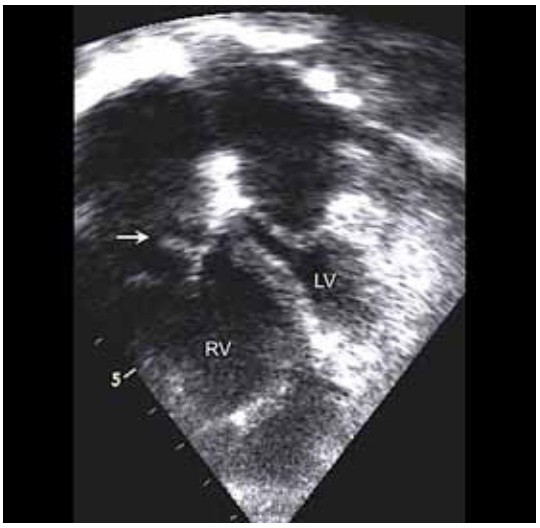

Fig. 2 Echocardiography image of a 4-chamber view of the heart shows a vegetation on the tricuspid valve leaflet (arrow). LV: left ventricle RV: right ventricle

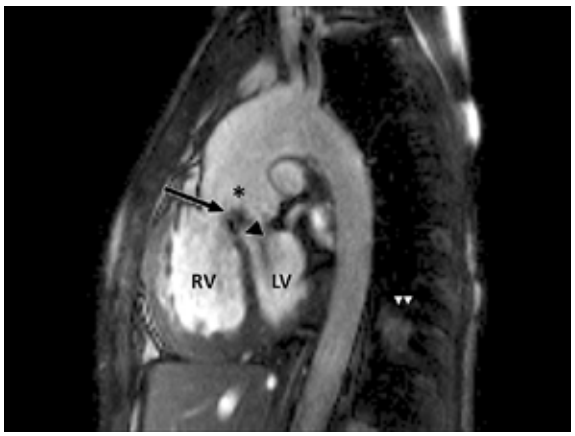

Fig. 3 Cine steady state free precession (SSFP) image of the left ventricular outflow tract cross-cut view shows a small vegetation at the aortic leaflets (thin arrow), overriding the aorta (*) and an outlet ventricular septal defect (VSD) (black arrowhead). There is also a lesion in the lower lobe of the left lung due to pulmonary infarct from septic embolus (white arrowheads). LV: left ventricle RV: right ventricle

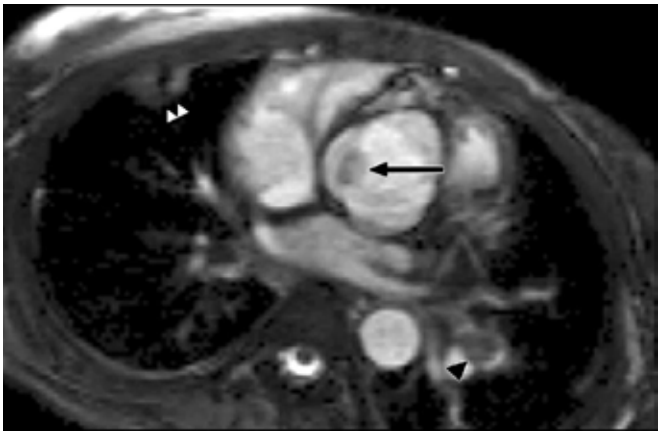

Fig. 4 Cine steady state free precession (SSFP) image of the enface view of the aortic valve shows a small vegetation at the non-coronary leaflet (thin arrow), a filling defect in the left lower lobar pulmonary artery (black arrowhead) and a pulmonary infarct at the periphery of the middle lobe with cavitation (white arrowheads).

the classification of DORV into various subtypes, the commonest being that with a sub-aortic VSD and pulmonary stenosis. DORV is rare and its overall prognosis is poor.[2-5] It is unusual to encounter patients with untreated DORV who survive to adolescence and beyond, such as in this case.

IE is an uncommon infection in children and occurs mainly in those with congenital heart disease (CHD). Advances in medical science and health care have led to a decrease in the incidence of IE and a shift in its demographics. In developed countries, there is a substantial decrease in the incidence of rheumatic heart disease as a predisposing cause of IE. At the same time, there is improved survival of children with CHD, thereby leading to an increase in the number of children who are at risk for IE.[6] The risk for IE is decreased in children with CHD after successful surgical repair, such as Tetralogy of Fallot repair, VSD closure and PDA ligation. This risk, however, remains if there are residual valvular lesions, conduits, aortopulmonary shunts and congenital collaterals.[7]

The emphasis of the new guidelines from the American Heart Association (AHA) has shifted toward maintenance of oral hygiene, as it is believed that the majority of IE cases are caused by bacteraemia from daily activities. AHA only recommends antibiotic prophylaxis for a select group of heart diseases (including cyanotic CHD) at high risk of developing infective endocarditis due to the higher risk of adverse outcomes in this group.[8,9] This patient's severe manifestation of IE underscores the importance of antibiotic prophylaxis in this group of patients. Oral hygiene must also be emphasised to high-risk patients, as this would reduce the likelihood of occurrence of IE.

The early recognition of IE is also important following a procedure with a high risk of bacteraemia. This may occur even if antibiotic prophylaxis is given. Recurrent fever without any obvious source following such a procedure should raise suspicion of IE. Investigations such as complete blood count, ESR, CRP and a series of blood cultures should help clinch the

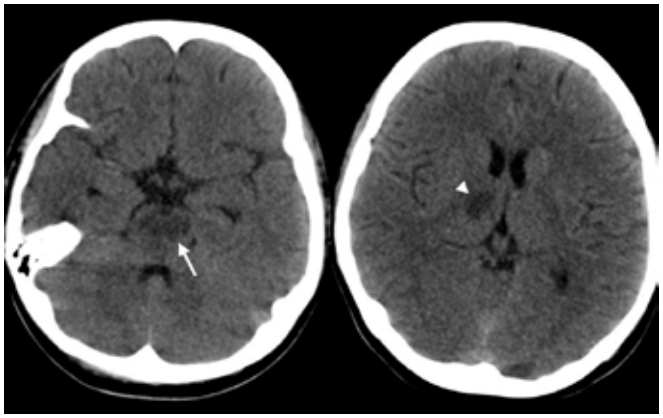

Fig. 5 Axial CT sections of the brain show hypodense lesions representing subacute infarcts in the pons (arrow) as well as the right thalamus and posterior limb of the right internal capsule (arrowhead).

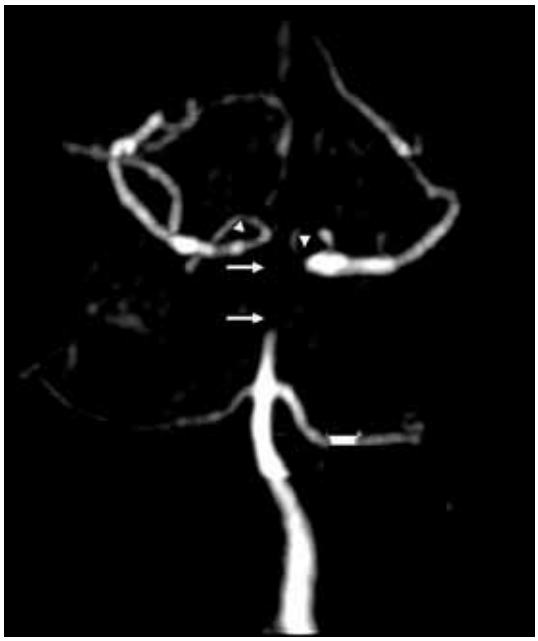

Fig. 6 Time-of-flight MR angiography of the vertebrobasilar system shows absent signal from the mid to distal basilar artery (arrows) in keeping with septic embolus. The posterior cerebral arteries (arrowheads) are supplied by their respective posterior communicating arteries. Incidentally, the right vertebral artery is hypoplastic.

diagnosis. Vegetations are usually easily demonstrated on transthoracic echocardiography in children. Transoesophageal echocardiography may be recommended for older children and adolescents if the transthoracic acoustic window is poor. Our patient's symptoms went unrecognised for several months until she finally presented with a cerebrovascular event.

The main cause of morbidity and mortality in IE is congestive heart failure due to valvular destruction, embolic events and sepsis. Destruction of the left-sided native valves with consequent severe regurgitation is an independent predictor of mortality, but early surgery may improve outcome.[10] Systemic embolic events occur in 30%–40% of left-sided IE, with cerebral embolism as one of the major causes of morbidity among all systemic embolic events as well as a predictor of in-hospital mortality.[11] Tricuspid valve endocarditis is associated with a high incidence of pulmonary embolism (75%–100%). However, most of the cases can be treated medically.[12]

At the outset, this patient had a high risk of IE with predictors for poor outcome. Diagnosis and definitive treatment were delayed for several months, allowing florid complications, including left-sided valvular destruction with regurgitation, cerebral embolism, congestive cardiac failure and pulmonary emboli to develop, all of which portend a poor prognosis.

In conclusion, this is a rare case of a patient with untreated DORV, who survived till adolescence but developed IE with florid complications following a dental procedure. The many radiological features of the complications of IE are well demonstrated in this case. Patients with cyanotic CHD have a high risk of IE, and the need for high clinical suspicion by the treating physician and patient education cannot be emphasised enough.

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