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ORIGINAL ARTICLE

MR findings of endocardial fibroelastosis in children

Enno Stranzinger · Gregory J. Ensing · Ramiro J. Hernandez

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Abstract
Background Endocardial fibroelastosis (EFE) is characterized by a diffuse white fibrous tissue lining the endocardium. The diagnosis is difficult to establish because clinical symptoms and electrocardiographic findings are nonspecific. Surgical resection of EFE requires the establishment of the diagnosis and delineation of the extent of the fibrotic changes.
Objective To describe the use of MRI in the assessment of EFE in children.
Materials and methods Three children after surgery for aortic stenosis who were suspected of having EFE were evaluated by echocardiography and MRI. The MR evaluation consisted of black-blood, triple IR, bright-blood, perfusion and myocardial delayed-enhancement sequences. EFE was confirmed at surgery in all patients.
Results Echocardiograms demonstrated vigorous systolic function but substantial diastolic dysfunction of the left ventricle in all. Mild endocardial brightening of the anterior septum, anterior wall, or papillary muscles was present in two. No study was thought to be diagnostic of endocardial fibrosis. On MRI EFE manifested at the endocardial surface as a rim of hypointense signal in the perfusion sequences and as a rim of hyperintense signal in the myocardial delayed-enhancement sequences. The black-blood, triple IR, and bright-blood sequences were not diagnostic.
Conclusion The diagnosis of EFE is difficult to establish by echocardiography. MRI using perfusion and myocardial delayed enhancement can be useful in establishing the diagnosis.

Keywords Endocardial fibroelastosis · Cardiac MRI · Perfusion · Myocardial delayed enhancement · Children

E. Stranzinger (✉) · R. J. Hernandez
Section of Pediatric Radiology,
University of Michigan Health System,
1500 E. Medical Center Drive,
Ann Arbor, MI 48109-0252, USA
e-mail: enno@med.umich.edu

G. J. Ensing
Department of Pediatrics and Communicable Diseases,
Pediatric Echocardiography Laboratory,
University of Michigan Health System,
Ann Arbor, MI, USA

Introduction

Endocardial fibroelastosis (EFE) is characterized by a diffuse white fibrous tissue lining the endocardium. Mortality in EFE is very high, with surgical excision of the EFE being the only effective therapy. The diagnosis of EFE is sometimes difficult to establish because clinical symptoms and electrocardiographic findings are nonspecific. Echocardiography findings suggestive of EFE include increased endocardial echo-brightness and a globular shape of the left ventricle (LV) [1]. However, there is a weak association between the echocardiographic appearance of echo-brightness of the myocardium and histologic evidence of EFE at autopsy [2]. Recent reports have described the utility of myocardial delayed-enhancement MRI in establishing the diagnosis of EFE [3, 4]. We describe here our experience with MRI in establishing the presence of EFE in three children.

Materials and methods

Three children after balloon valvuloplasty for severe aortic stenosis and persistent pulmonary hypertension were suspected of having EFE. The ages of the children were 1 month, 11 months, and 2½ years. The 2½-year-old child had also undergone autograft replacement of the aortic valve (Ross procedure). All children had undergone cardiac catheterization and balloon angioplasty but had persistently elevated LV filling pressures and pulmonary hypertension. LV injection was performed only in one child. The appearance of the ventricular cavity was unremarkable. Multiple echocardiographic evaluations had been performed as part of their clinical care. The children were scanned under general anesthesia in a 1.5-T magnet (Philips, Best, the Netherlands) with a SENSE cardiac coil. All sequences were ECG-triggered and the children were imaged with a breath-hold technique under general anesthesia. Black-blood and triple IR sequences were obtained in the axial plane. A bright-blood sequence using a balanced fast field echo (bFFE) was used to obtain short-axis views of the heart. Perfusion and myocardial delayed-enhancement sequences were performed in the short-axis plane after the injection of gadopentetate dimeglumine 0.2 mmol/kg (Magnevist; Berlex Laboratories, Wayne, N. J.) through a peripheral vein. Perfusion imaging was performed using turbo field echo-planar imaging. The myocardial delayed-enhancement sequence consisted of an inversion recovery-prepared, electrocardiographically triggered, fast gradient-recalled echo pulse sequence (TR/TE 4.0/1.2 ms, inversion time 180–250 ms, acquisition matrix 224×200 mm, flip angle 15°, field of view 350×350 mm, matrix 256×256, slice thickness 5 mm). The optimal inversion time to determine the best myocardial signal was selected once, 10 min after contrast agent injection, using the Look-Locker technique. The myocardial delayed-enhancement sequence was obtained every 10 min for approximately 40 min. This study met the requirements of the Internal Review Board of the University of Michigan.

Results

Multiple two-dimensional, Doppler, and tissue Doppler echocardiograms were acquired in each child. Echocardiogram at the time of MRI demonstrated mean systolic

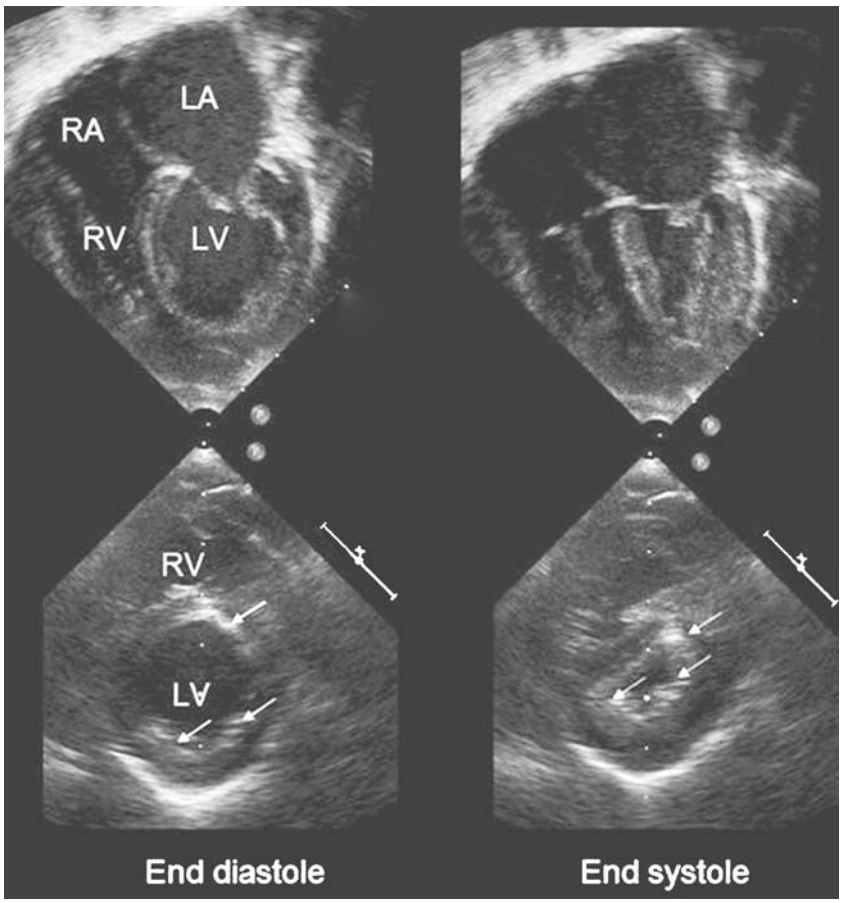

Fig. 1 Echocardiographic apical four-chamber (*top*) and parasternal short-axis (*bottom*) systolic and diastolic frames in the 1-month-old infant with the most prominent EFE changes. Note left atrial enlargement, hyperdynamic systolic function and echo-bright endocardial regions (*arrows*), especially in the anterior septum and adjacent to the LV papillary muscles

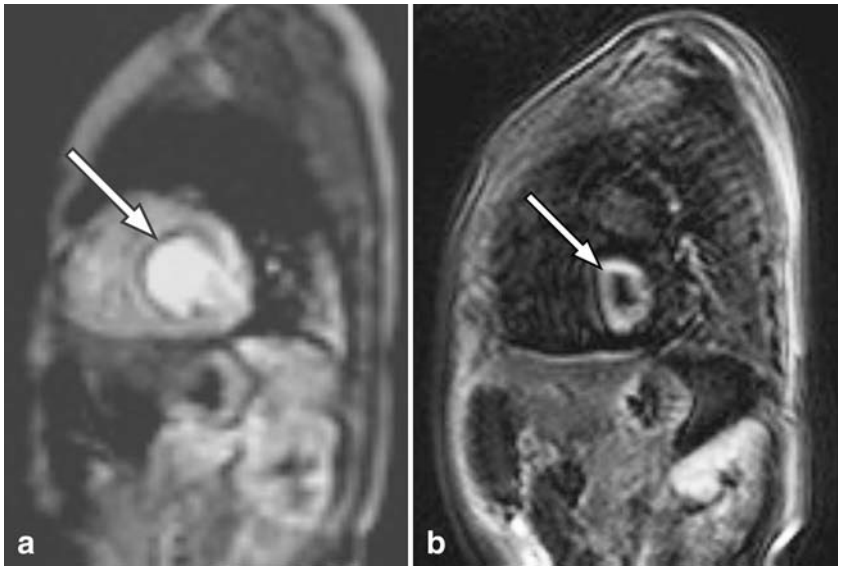

Fig. 2 A 15-month-old infant with aortic stenosis after balloon valvuloplasty as a neonate, who developed progressive pulmonary hypertension. **a** Hypointense inner layer (*arrow*) of the endocardium on early perfusion. **b** The 29-min viability sequence shows delayed enhancement of EFE (*arrow*)

gradients across the aortic valve of 12, 28, and 3 mm Hg, respectively. Aortic regurgitation was mild, mild, and trivial, respectively. Right ventricular pressure was estimated by tricuspid regurgitation velocity or septal position as greater than three-quarters systemic in one patient and systemic in two patients. All children had hyperdynamic LV systolic function with apical four-chamber Simpson's single-plane ejection fractions of 82%, 80%, and 65%, respectively (Fig. 1). All children had evidence of LV diastolic dysfunction with substantial left atrial enlargement, peak early/peak atrial velocity ratio greater than 2, and/or mitral spectral E to lateral annular tissue Doppler E velocity ratio greater than 10.

Assessment of the LV endocardium demonstrated very mild brightening of the anterior septum and papillary muscles in two patients while one had minimal brightening. While one of the three had images thought suggestive of LV endocardial fibrosis, none was thought diagnostic.

Fast spin-echo with and without fat suppression and steady-state free precession imaging did not distinguish between EFE and uninvolved myocardium. In the early perfusion sequences a hypointense inner layer of the endocardium was noted in all three children. Myocardial delayed enhancement demonstrated hyper-enhancement involving the endocardial surface of the LV. Best signal contrast between EFE and myocardium was again noted on the delayed viability sequences, 20–30 min after GD/DTPA administration. The delayed enhancement sequence requires imaging after a delay following the gadolinium injection. High signal from GD/DTPA administration within the ventricle is noted up to 15 min after contrast agent injection. Best discrimination between the enhancement of EFE was noted after 20–30 min, when the signal of the ventricle is noted to be hypointense again. The imaging at other times was highly suggestive for EFE. The optimal inversion time ranged between 180 and 250 ms.

All three children had surgical confirmation and removal of EFE (Figs. 2 and 3). One child had also pathologic confirmation of EFE.

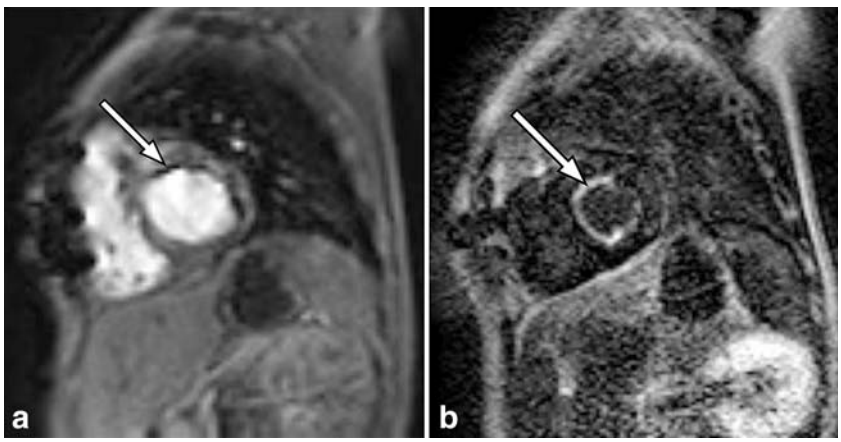

Fig. 3 A 2-year-old boy with history of critical aortic stenosis, after balloon valvuloplasty, who underwent a Ross procedure for significant aortic insufficiency. **a** Sagittal T1-weighted early perfusion sequence shows a hypointense inner layer (*arrow*) of the endocardium. **b** Sagittal delayed viability sequence shows a bright endocardial rim (*arrow*)

Discussion

We describe here our experience with MRI in establishing the diagnosis of EFE in three children. In the perfusion sequence EFE manifested as a hypointense layer at the endocardial surface, which became white in the myocardial delayed-enhancement MR sequence manifested as a hyperintense layer at the endocardial surface. The proposed mechanism for this enhancement is that the fibrous membrane, because of its avascular nature, will appear as low signal in the perfusion sequence. In the delayed enhancement sequence, similar to the findings seen after myocardial infarction, the membrane would exhibit increased signal.

Children with EFE present clinically with signs of congestive heart failure and elevated pulmonary pressure. The etiology of EFE is unknown [5–15]. Although EFE might be present without any structural anomaly of the heart, it is most commonly seen with obstructive lesions of the LV such as aortic stenosis. EFE is characterized by a diffuse white fibroelastic tissue that lines the endocardium and spares the myocardium [1]. The lining of the endocardium by the EFE leads to partial obliteration of the ventricle with decreased ventricular distensibility and impaired diastolic filling. Involvement of the chordae tendineae can lead to mitral or tricuspid regurgitation [16].

Children with EFE have a poor prognosis depending on the etiology of the underlying disease and the severity of the fibrotic process. The treatment of choice is endocardial resection of one or both ventricles [17]. Surgery is the only therapeutic option for those children with clinical manifestation of EFE (pulmonary hypertension). Reversal of diastolic dysfunction with an increase in cardiac output can be observed in most children, and is associated with an improvement in symptoms [16].

Echocardiography findings suggestive of EFE include increased endocardial echo-brightness and a globular shape of the LV [1]. However, previous reports have revealed a weak association between the echocardiographic appearance of echo-brightness of the myocardium and the histologic evidence of EFE at autopsy [2]. US findings consistent with EFE, the hyperreflective endocardium, might be influenced by instrument settings and the angle of incidence between the US beam and the endocardial surface. Surgical resection of EFE requires the establishment of the diagnosis and delineation of the extent of the fibrotic changes greater than can be provided by echocardiography.

Although the myocardial delayed-enhancement MR sequence was originally documented to be useful in visualizing fibrosis and myocardial infarctions [18, 19], a recent report described its utility in the diagnosis and delineation of EFE [4]. In this technique, the optimal inversion time is selected to null the signal of the myocardium. When the optimal inversion time is determined, imaging is obtained with the inversion recovery-prepared, electrocardiographically triggered fast gradient-recalled echo pulse sequence. Optimal signal contrast between EFE and myocardium occurred approximately 20–30 min after GD/DTPA administration. The membrane lining the myocardium exhibited hyperenhancement in comparison to the low signal of the myocardium.

Conclusion

EFE is often responsible for significant morbidity and mortality in children after technically successful intervention for LV outflow obstruction. Surgical excision, perhaps the therapy of choice, requires an accurate preoperative assessment. US has significant limitations. MRI using perfusion and myocardial delayed-enhancement sequences is a useful imaging technique for the diagnosis of EFE.

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