Delayed-enhancement cardiovascular magnetic resonance identifies fibrous tissue in children after surgery for congenital heart disease

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Supplemental material is available online.

Objectives: To determine whether delayed-enhancement magnetic resonance imaging can identify fibrous tissue associated with patch reconstructions in postoperative patients with congenital heart disease. Additionally, to determine whether fibrous structures not directly related to the surgical reconstruction exhibited delayed enhancement.

Methods: Seventy-three patients underwent magnetic resonance imaging. Studies were retrospectively reviewed for the presence of delayed enhancement along the ventricular outflow tracts, cardiac valves, and where available, the ascending aorta. Three groups were identified. Group A patients ($n = 34$) underwent right ventricular outflow tract reconstruction and ventricular septal defect patch closure. Group B patients ($n = 33$) had never undergone cardiac surgery. Group C patients ($n = 6$) had functional single ventricle and underwent Norwood reconstruction.

Results: In group A, 31 of 34 patients had delayed enhancement of the right ventricular outflow tract, and 14 of 34 had delayed enhancement of the ventricular septal defect patch ($P < .001$). In group B ($n = 33$), 1 patient with arrhythmogenic right ventricular dysplasia had delayed enhancement limited to the right ventricular outflow tract. The remainder had no delayed enhancement of either outflow tract. Delayed enhancement of the aortic valve and ascending aorta was observed in 13 of 34 ($P = .002$) and 10 of 26 ($P = .05$) group A patients, respectively, compared with 2 of 33 and 3 of 24 group B patients. In group C, delayed enhancement of the Norwood reconstruction was observed in 5 of 6 patients ($P = .002$).

Conclusions: Delayed-enhancement imaging detects fibrous tissue along regions of reconstruction in patients who have had surgery for congenital heart disease. Furthermore, delayed-enhancement imaging detects fibrous tissue in regions not directly related to the reconstructive surgery, including cardiac valves and the wall of the ascending aorta.

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Patients with congenital heart defects such as tetralogy of Fallot (TOF) may require surgical intervention involving reconstruction of the right ventricular outflow tract (RVOT) and patch closure of a ventricular septal defect (VSD). Reconstruction of the RVOT is typically performed with cryopreserved homograft tissue as a transannular patch or a valved conduit.^{1,2} VSD patches are most commonly composed of artificial materials such as polytetrafluoroethylene or Dacron.^{2,3} Histologic evidence of fibrous tissue formation has been observed on the surface of homograft and artificial patch material in animal models^{4,5} and humans.^{6,7}

Delayed-enhancement (DE) magnetic resonance (MR) imaging can identify myocardial fibrosis.⁸⁻¹¹ DE occurs after intravenous administration of a gadolinium chelate, which primarily remains in the interstitial space. The proposed mechanisms involve delayed contrast washout and an increased volume of distribution present in fibrous tissue.¹²⁻¹⁴

Abbreviations and Acronyms

DE	= delayed enhancement
FLASH	= fast low-angle shot
MR	= magnetic resonance
RVOT	= right ventricular outflow tract
SSFP	= steady-state free precession
TOF	= tetralogy of Fallot
VSD	= ventricular septal defect

We hypothesized that DE should occur in regions directly associated with the surgical reconstruction where fibrous tissue formation occurs. Recently, DE of the RVOT and in the region of VSD repair was reported in adults with TOF.^{15,16} Our study differs in that it specifically investigated the pediatric population, in whom fibrous tissue may not have had the time to form. In addition, fibrous tissue formation in children may affect the surgical reconstruction differently than in adults, because the patients and their hearts continue to grow. Moreover, recent studies have shown that fibrous tissue is found within the aortic sinuses and the wall of the ascending aorta in patients with TOF, providing a possible mechanism for TOF aortopathy.¹⁷ Therefore, we expanded our DE MR study to determine whether fibrous tissue could be identified in regions not directly involved in the reconstructive surgical repair, such as in heart valves and within the wall of the ascending aorta. Finally, in this study, we confirmed our findings using a control population of patients who had never undergone cardiovascular surgery.

Materials and Methods**Patient Population**

We conducted a retrospective review of all pediatric DE MR studies performed at The Children's Hospital of Philadelphia between October 2003 and August 2005. Inclusion criteria required patients to be under age 21 years and to have a DE MR study covering the ventricular outflow tracts. All included studies were satisfactory for interpretation without significant artifact. Two studies were excluded owing to poor image quality. Three groups were identified. Group A patients ($n = 34$) had conotruncal anomalies and underwent both reconstruction of the RVOT and patch closure of a VSD. The mean age at the time of the patients' most recent surgical reconstruction was 1.6 ± 2.7 years. The time between surgery and the MR study was 8.9 ± 5.5 years (Table E1). Group B patients ($n = 33$) had never undergone cardiovascular surgery. Fifteen of these patients had congenital heart disease, 7 had acquired heart disease, and 11 had no evidence of cardiovascular disease confirmed by cardiac MR (Table E2). Group C patients ($n = 6$) had functional single ventricle and underwent Norwood aortic reconstruction with homograft patch (Table E3). All remaining MR studies that did not fall into one of these three categories were excluded. Group A ages ranged from 10 months to 20 years (mean 10.6 ± 5.2 years), group B ages ranged from 10

days to 20 years (mean 11.5 ± 6.7 years), and group C patient ages ranged from 2 to 18 years (mean 7.9 ± 7.4 years). The local institutional review board approved this retrospective study.

Cardiovascular MR

Studies were performed on 1.5-T Siemens Sonata or Avanto systems (Siemens Medical Solutions, Malvern, Pa). Group A patients' assessments routinely included evaluation of the RVOT and pulmonary artery architecture, as well as quantification of right ventricular volume, function, and pulmonary regurgitation. Group B patients presented most commonly for assessment of ventricular function. Studies were retrospectively reviewed for the presence of delayed gadolinium enhancement in the region of the ventricular outflow tracts, valve leaflets, and the ascending aorta.

Patients under 7 years of age were typically sedated for the MR study with pentobarbital, and midazolam, except for infants under age 1, who received chloral hydrate. No complications were observed.

Approximately 10 minutes after the administration of gadopentetate dimeglumine, (0.4 mmol/kg), and with the appropriate inversion time to achieve myocardial nulling, DE imaging was executed in the same long- and short-axis planes that were used for cine imaging. Comparison of the cine and DE images distinguished DE from other potential causes of bright signal, such as pericardial fat or fluid. DE studies consisted of steady-state free precession (SSFP) or fast low-angle shot (FLASH) inversion recovery sequence with and without phase-sensitive inversion recovery. Typical SSFP parameters were repetition time 10% of R-R interval, echo time 1.3 ms, inversion time 300 ms, flip angle 45° , segments 65, and voxel size $1.8 \times 1.3 \times 8.0$ mm. Typical FLASH parameters were repetition time 10% of R-R interval, echo time 4.4 ms, inversion time 260 ms, flip angle 30° , segments 25, and voxel size $1.7 \times 1.3 \times 8.0$ mm. Infant studies included single-shot acquisition, two averages, and thinner slices (5.0–6.0 mm). Group A studies were reviewed for the presence or absence of DE of the RVOT patch/conduit, VSD patch, valve leaflets, and where available, the ascending aorta. Group B studies served as controls. Group C studies were reviewed for DE of the aortic reconstruction.

Statistical Analysis

Data are expressed as mean and SD range as appropriate. The Fisher exact test was performed as appropriate.

Results**RVOT and VSD Patch**

In group A ($n = 34$), DE of the RVOT reconstruction occurred in 31 (91%) patients compared with 1 (3.0%) of 33 patients without surgery (group B) ($P < .001$) (Figure 1; Table 4). One patient, with the diagnosis of double-chambered right ventricle, underwent subpulmonary resection and VSD closure without homograft patching and still had DE in the region of the RVOT. The only group B patient to exhibit DE of the RVOT had arrhythmogenic right ventricular dysplasia.

In group A ($n = 34$), DE of the VSD patch occurred in 14 (41%) of 34 patients, compared with 0 of 33 group B

Figure 1. A 3-year-old girl after repair of truncus arteriosus at 1 week of age. **A**, Right ventricular outflow tract view, steady-state free precession (SSFP) cine image, and the corresponding SSFP phase sensitive inversion recovery image (**B**). Note the prominent delayed enhancement along the anterior wall of the right ventricular-pulmonary artery conduit (*open arrows*) and the ventricular septal defect patch (*long solid arrow*). Also, note delayed enhancement of the truncal (*dashed arrow*) and mitral valves (*arrowheads*).

Figure 2. A 16-year-old boy after tetralogy of Fallot repair in infancy. Delayed enhancement of the aortic valve (*dashed arrow*) and ventricular septal defect patch (*solid arrow*) are observed.

patients ($P < .001$) (Figures 1 and 2; Table 4). Fifteen (79%) of the 19 patients who underwent right ventricle-pulmonary artery conduit placement had at least qualitatively moderate obstruction in the region of the DE. Of the 3 group A patients who did not have DE of the RVOT, only 1 had a conduit, and the conduit was unobstructed. In group A, the mean right ventricular ejection fraction was $61\% \pm 9\%$. The mean pulmonary regurgitant fraction was $30\% \pm 16\%$.

Cardiac Valves

DE of the aortic valve occurred in 13 of 34 group A patients versus 2 of 31 group B patients (38% vs 7%; $P = .002$) (Figures 2 and 3; Table 4). DE of the mitral valve occurred in 22 (65.0%) of 34 group A patients and 14 (42%) of 33 group B patients ($P = .088$) (Figures 1 and 3; Table 4). DE of the tricuspid valve occurred in 25 (74%) of 34 group A patients versus 17 (52%) of 33 group B patients ($P = .080$) (Figure 4B; Table 4).

In group A, DE of the mitral (22/34, 65.0%; $P = .004$) and tricuspid valves (25/34, 74%; $P = .03$) occurred more frequently than DE of the aortic valve (13/34, 38%). In group B, DE of the mitral (14/33, 42%; $P = .001$) and tricuspid valves (17/33, 51%; $P < .001$) occurred more frequently than DE of the aortic valve (2/33, 7%). The pulmonary and conduit valves were rarely observed and therefore no comparison was possible.

DE of the Ascending Aorta

DE of the ascending aorta was observed in 10 (38%) of 26 group A patients and 3 (13%) of 24 group B patients ($P = .03$) (Figure 3; Table 4). The ascending aorta was not observed in 8 group A patients and 9 group B patients.

Figure 3. An 8-year-old girl after tetralogy of Fallot repair. **A**, Cine image of the ascending aorta (solid arrows), and the aortic (dashed arrows) and tricuspid (arrowheads) valves. **B**, Corresponding delayed enhancement image. Note the delayed enhancement of the ascending aorta, as well as the aortic and tricuspid valves. These structures exhibit delayed enhancement, even though they were not directly involved in the surgical reconstruction.

DE of Homograft Reconstruction of the Aorta

Five (83%) of 6 Norwood reconstructions compared with 3 (13%) of 24 group B patients showed DE ($P = .002$).

Structurally Abnormal Valves

In group A, 2 of the 5 patients with truncus arteriosus had DE of the truncal valve (Figure 1). In group B, there were 7 patients with bicuspid aortic valve. Of these, only 1 (14%) exhibited DE of the aortic valve and the ascending aorta. The remaining 6 had no DE of either the aortic valve or the ascending aorta. In group B, there was 1 patient with Ebstein anomaly who showed DE of the septal leaflet of the tricuspid valve (Figure E1, A, and E1, B).

Discussion

This cardiac MR study demonstrates that after intravenous administration of gadopentetate dimeglumine, DE is readily

observed on homograft patches/conduits and VSD patches. This study extends the recent findings in adult TOF patients^{15,16} to the pediatric population. In addition, our findings of DE MR are validated through comparing them with an age-matched control population (group B, $n = 33$) that never underwent cardiovascular surgery.

We studied younger patients to stress that fibrous tissue deposition is significant enough to image even among infants, children, and young adults. During these early post-operative years, patients generally do not exhibit right ventricular dysfunction from chronic valvular or conduit incompetence and the resultant volume overload. Our findings of frequent DE along RVOT reconstructions (31/34, 91%) occur in the context of a preserved mean right ventricular ejection fraction ($61\% \pm 9\%$). This is in contrast to the adult TOF patient studies,^{15,16} which reported signifi-

Figure 4. A 2-year-old patient after Norwood aortic reconstruction and Fontan operation for hypoplastic left heart syndrome. **A**, Note the delayed enhancement along the anterior (white arrows) and posterior walls (black arrows) of the Norwood reconstruction. **B**, Delayed enhancement confirmed in the coronal view. Interestingly, the tricuspid valve (black arrows) and the lateral wall of the lateral-tunnel Fontan (open arrows) also exhibit delayed enhancement. **LV**, Left ventricle; **RV**, right ventricle; **TV**, tricuspid valve.

cantly lower right ventricular ejection fractions in the populations that had DE. The differences in right ventricular function between the two studies suggest that the impact of DE differs between the pediatric and adult populations.

Identifying fibrous material in the pediatric context may be important, because the fibrosis may contribute to RVOT obstruction in the form of a fibrous peel within a conduit. Nearly half of our surgically treated patients had at least qualitatively moderate-range conduit stenosis. Conduit obstruction is not an uncommon finding in the pediatric population because patients outgrow conduits placed in infancy.

It is unclear why DE of the RVOT homograft reconstruction was more common than DE of the artificial VSD patch (91% vs 41%). It is possible that the original homograft collagenous skeleton provides the framework for fibrous tissue deposition through immune-mediated responses of the host to the donor tissue endothelial surface proteins, as has been postulated for cryopreserved homograft valve implant failure.⁷ Similar to RVOT homograft patch reconstructions, it is understandable that DE of Norwood homograft reconstructions is common when compared with normal aortas.

Interestingly, we observed that even structures not directly involved in the surgical repair may exhibit DE. For instance, patients with repaired conotruncal anomalies without homograft reconstruction of the ascending aorta exhibited DE of the ascending aorta more frequently than the control population (41% vs 13%; $P = .05$). DE of the aortic wall unrelated to surgical reconstruction has been reported in Takayasu arteritis,¹⁸ which is thought to result from aortic wall inflammation. Furthermore, DE of the aortic valve occurred more commonly in the group with repaired conotruncal anomalies than in the control group (37% vs 6%; $P < .01$). Dilation of the aortic root and ascending aorta frequently occurs in patients with TOF. Recently, the histologic basis of TOF aortopathy in infants and adults was described by Tan and associates.¹⁷ The findings included fibrosis, cystic medial necrosis, elastic fragmentation, and elastic lamellae disruption involving the aorta and the aortic sinuses. We speculate that these abnormal histologic findings could result in impaired gadolinium washout kinetics and provide a possible explanation for our observation of DE of aortas and aortic valve cusps in patients with conotruncal anomalies. Tan and colleagues¹⁷ also found earlier onset of elastin fragmentation and elastin lamellae disruption of the aortic root and ascending aorta in patients with TOF in contrast to those with a bicuspid aortic valve. Although not statistically significant, we found a trend among our pediatric study population showing that patients with repaired conotruncal anomalies exhibited DE of the aortic valve (14/34) and ascending aorta (10/26) more often than do patients with a bicuspid aortic valve (1/7). Nevertheless, it is possible that the fibrous tissue identified in the

ascending aorta could be related to a previous cannulation site or aortic crossclamping. However, the degree of DE observed was diffuse and not discrete and, therefore, would less likely be thought to result from aortic cannulation or crossclamping. Finally, though it had not previously been reported, we are reassured that our findings of DE of valve tissue are valid, inasmuch as it is present on DE images performed at other institutions.¹⁹

Though metabolically active, cardiac valves have been thought to be relatively avascular structures, possessing few capillaries proximally, receiving their blood supply primarily via diffusion.²⁰ Weind, Ellis, and Boughner²¹ demonstrated the existence of a more extensive vasculature than previously thought in the aortic valve of swine and suggested that cryopreserved homograft implants fail owing to the absence of this functioning vasculature. It is interesting that in our study atrioventricular valves exhibit DE more frequently than aortic valves, in both the repaired conotruncal and control groups. Though the reason for this finding is unclear, it suggests that different contrast washout kinetics operate between atrioventricular and semilunar valves. It is also interesting that the tricuspid valve was frequently enhanced in both the surgical and control groups (74% vs 52%), without statistical difference. Inasmuch as the DE was not confined to the septal leaflet of the tricuspid valve, one can reasonably consider that tricuspid valve DE is a property of its contrast washout kinetics and not the result of VSD closure.

Study Limitations

Preoperative cardiac MR studies were not available in any of our patients with conotruncal anomalies (group A). These data would be useful in further confirming the presence and extent of DE associated with surgical reconstructions. In addition, preoperative MR studies might clarify whether the increased frequency of DE of the aortic valve and ascending aorta in patients with repaired conotruncal anomalies was a result of the native disease or, in part, related to the postoperative state.

Conclusions

In conclusion, DE MR imaging identifies fibrous tissue associated with surgical patch reconstructions in pediatric patients undergoing surgery for congenital heart disease. In addition, DE imaging also identifies fibrous structures not directly involved in the surgical repair, such as the ascending aortas of patients with conotruncal anomalies and in cardiac valves. We believe that this imaging technique will prove useful in the assessment of fibrous tissue formation associated with congenital heart disease and its evolving treatments.

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TABLE E1. Group A patient characteristics and clinical data—after conotruncal surgery

Group A characteristics		
n	34	
Male	22	66%
Age at MRI (y)	10.6 ($\pm$ 5.2)	
Age at operation (y)	1.6 ($\pm$ 2.7)	
Time between operation and MRI (y)	8.9 ($\pm$ 5.5)	
Conduit	19	56%
Transannular patch	15	44%
Ventricular septal defect patch	34	100%
Conduit stenosis	15	44% (of total) 79% (of conduits)
Type of congenital heart disease		
Tetralogy of Fallot	27	79%
Truncus arteriosus	5	15%
Transposition of the great vessels	1	3%
Double-outlet right ventricle	1	3%
Physiology		
Right ventricular ejection fraction (%)	61 ($\pm$ 9)	
Pulmonary regurgitant fraction (%)	30 ($\pm$ 16)	

MRI, Magnetic resonance imaging.

TABLE E3. Group C patient characteristics and clinical data—after Norwood reconstruction

Group C characteristics		
<i>n</i>	6	
Male	3	50%
Age at MRI (y)	7.9 ($\pm$ 7.4)	
Age at Norwood	2-40 days	
HLHS	5	83%
DILV	1	17%

DILV, Double-inlet left ventricle; *HLHS*, hypoplastic left heart syndrome; *MRI*, magnetic resonance imaging.

TABLE E2. Group B patient characteristics and clinical data—patients not treated surgically

Group B characteristics		
n	33	
Male	19	58%
Age at MRI (y)	11.5 (±6.7)	
Congenital heart disease	15	46%
Acquired heart disease	7	21%
No heart disease	11	33%

MRI, Magnetic resonance imaging.

TABLE E4. Delayed enhancement differences between groups

	Group A (n = 34): Conotruncal anomaly			Group B (n = 33): No surgery		P value	Group C (n = 6): Norwood		
RVOT	31	91%		0		<.01	n/a		
VSD patch/LVOT	14	41%		0		<.01	n/a		
Aortic valve	13	38%		2	6%	<.01	n/a		
Tricuspid	25	74%		17	52%	.08	n/a		
Mitral	22	65%		14	42%	.09	n/a		
AAO/Norwood	10	38%	(n = 26)*	3	13%	.05	5	83%	P < .01

AAO, Ascending aorta; LVOT, left ventricular outflow tract; n/a, not available; RVOT, right ventricular outflow tract; VSD, ventricular septal defect.
*Retrospective delayed enhancement assessment was not possible in 8 group A and 9 group B patients because the ascending aorta was not imaged.

Figure E1. A 19-year-old patient born with Ebstein anomaly of the tricuspid valve. A, Cine image demonstrates the dilated atrialized portion of the right ventricle. Arrows point to the region where the rudimentary septal leaflet resides. B, The corresponding delayed enhancement image highlights the rudimentary septal leaflet.