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ORIGINAL ARTICLE

WILEY Congenital Heart Disease

The utility of cardiac magnetic resonance imaging in post-Fontan surveillance

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Abstract

Objective: Gated cardiac MRI offers the most detailed and accurate noninvasive method of assessing cardiac anatomy, particularly in patients with complex congenital heart disease. The proposed benefits of using cMRI as a routine screening tool in the Fontan population include early recognition of asymptomatic, postoperative anatomic and physiologic changes. In 2011, we therefore instituted at our center a recommended practice of cMRI screening in patients with Fontan physiology at 3 and 8 years post-Fontan operation. The purpose of this study was to determine the impact of this standardized practice of cMRI screening on the clinical management of a Fontan population.

Design: We retrospectively reviewed charts from our institutional Fontan database to determine which patients were eligible for cMRI under the current guidelines and who underwent imaging from November 2002 to June 2015. We reviewed the frequency of cMRI and number of changes in management based on the results. Statistical significance was determined using a chi-square test.

Results: There were 141 cMRIs performed on 121 patients who met inclusion criteria. The odds of a change in management were significantly greater after clinically indicated cMRI compared to screening cMRI (OR = 3.79, 95% CI: 1.48-9.66, P = .004). There were near significant odds of change in management if the cMRI occurred <8 years after Fontan regardless of whether it was for screening or clinically indicated purposes (OR = 2.43, 95% CI: 0.97-6.08, P = .052). The most frequent change in management was referral for catheterization with pulmonary artery angioplasty.

Conclusions: There is an important role for cMRI in routine surveillance of post-Fontan patients. Screening cMRI performed less than 8 years after Fontan palliation offers increased utility compared to studies performed later. The optimal timing of such imaging after Fontan palliation remains unclear.

KEYWORDS

cardiac MRI, Fontan palliation, outcomes, surveillance

1 | INTRODUCTION

For more than 40 years the Fontan procedure has played an important role in surgical palliation for patients with a functional single ventricle. While perioperative mortality has dramatically decreased as the procedure has been modified, significant long-term morbidity remains, including heart failure, thrombi, conduit stenoses, and hepatic congestion.[1-6] For these reasons, some have advocated for regular cardiac MRI (cMRI) surveillance of this patient population.

Cardiac MRI has established itself as a reliable and noninvasive method for detecting and tracking the progression of morbidities commonly found both early and late after the Fontan procedure.[3,7] This imaging modality has also emerged as an alternative to echocardiography given the limitations of echocardiography to clearly delineate the complex anatomy in this patient population. Furthermore, cMRI has demonstrated the ability to identify potential disease among Fontan patients even in the asymptomatic stages such as "silent" pulmonary emboli.[8-11] For these reasons, cMRI has become a preferred method of surveillance for some clinicians seeking serial imaging of patients with Fontan physiology.[3]

While the benefits of cMRI in this population are clear, the frequency with which to conduct such testing remains unclear. Accordingly, the objective of this study was to determine the utility and optimal timing of cMRI at various points in time following Fontan palliation. To achieve this objective, we assessed the frequency of changes in management and their relationship to the timing of cMRI after a Fontan procedure.

TABLE 1 Fontan population characteristics

Demographics	n (%)
Race/ethnicity	
White non-Hispanic	76 (62.8%)
Black non-Hispanic	32 (26.4%)
Asian non-Hispanic	7 (5.8%)
Hispanic or Latino	2 (1.7%)
Other	4 (3.3%)
Sex	
Female	56 (46.3%)
Male	65 (53.7%)
Age at Fontan	
Median (25th-75th)	2.3 y (2.0 to 3.2 y)
Minimum-maximum	4.2 m to 11.2 y
Age at first cMRI	
Median (25th-75th)	10.3 y (6.6 to 13.4 y)
Minimum-maximum	2.4 to 21.2 y
Type of Fontan	
Intracardiac	69 (57.0%)
Extracardiac	50 (41.3%)
Y graft	1 (.8%)
Unspecified	1 (.8%)
Single ventricle classification	
Left predominant	59 (48.8%)
Right predominant	53 (43.8%)
Unspecified	9 (7.4%)

2 | METHODS

We performed a retrospective cohort study of patients who underwent a Fontan palliation procedure between 1994 and 2015. Patients were included in the study if they underwent cMRI any time after discharge following a Fontan procedure. Patients were excluded if there was no recorded reason for obtaining the cMRI. In 2011, our institution implemented a standardized protocol for care of children with Fontan palliation. This protocol included a screening cMRI at 3 years and 8 years post-Fontan palliation. The present study was approved by the Institutional Review Board at Children's Healthcare of Atlanta.

2.1 | Clinical variables

After identifying Fontan patients who had undergone cMRI, we determined if a change in management had been made following the imaging study. A clinician was considered to have made a "change in management" if the clinic note following cMRI acknowledged the recent imaging study and indicated that he or she was changing medical or interventional treatment strategy or ordering further diagnostic testing based on the results of the cMRI.

The reason for obtaining a cardiac MRI was noted as either for "screening purposes" in accordance with our institution's 2011 protocol, "screening" based on the clinicians' notes preceding the imaging order, or as "clinically indicated" based on the clinicians' notes preceding the imaging order. It is in this way that many cMRI studies performed before the implementation of the 2011 protocol are included in this study and classified as either screening or clinically indicated. Over the duration of the study period, all included cMRI studies were performed as standard of care clinical studies, though the specific pulse sequences and data obtained during the study period did evolve. All studies included steady state free precession cine imaging for anatomic assessment and ventricular functional analysis as well as 2D phase contrast imaging of the systemic arterial, branch pulmonary artery, bidirectional Glen, and Fontan baffle. In later study years, delayed enhancement imaging to assess for myocardial fibrosis, additional 2D phase contrast of the pulmonary veins to quantify collateral burden, and 4D phase contrast imaging was also performed in some, but not all, patients.

Some screening studies were performed outside the three- and eight-year intervals after Fontan surgery as recommended by our institution's 2011 protocol due to physician preference or that many patients were already several years post-Fontan when the protocol was implemented. Due to the variability in timing of cMRI after Fontan, we also divided the data into studies that occurred relatively early (<8 years) or late (≥8 years) after Fontan palliation.

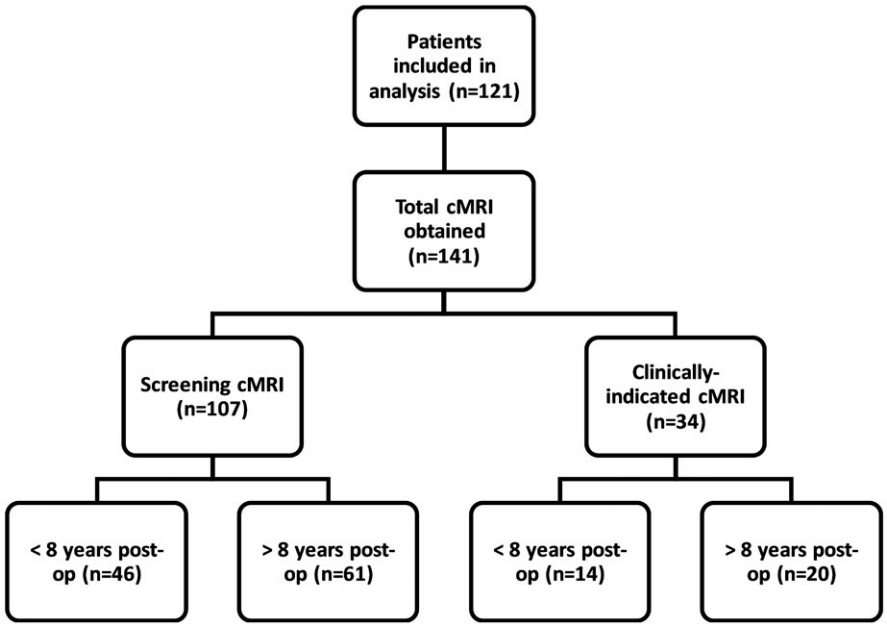

FIGURE 1 Numbers of cMRI obtained in relation to time since Fontan palliation

2.2 | Statistical analysis

Categorical variables are represented by frequencies and percentages. The number of management changes made after "screening" cardiac MRI's and "clinically indicated" cardiac MRI's were compared using chi-squared tests when appropriate.

The number of management changes in the "<8" and "≥8" screening MRI groups was compared using a chi-square test. There were often multiple cMRI's performed on a single patient. In order to analyze multiple "changes in management" from a single patient as separate events, each cMRI was considered as independent cMRIs for the analysis.

3 | RESULTS

3.1 | Population characteristics

Of 425 Fontan patients in our institutional database, 148 underwent cMRI and 121 met inclusion criteria. The majority of included patients were white, male, and had an intracardiac Fontan (Table 1). The median age at Fontan was 2.3 years with a median time of first cMRI at 10.3 years of age. The 121 patients included in our analysis underwent a total of 141 cMRI's, 107 of which were performed for screening purposes and 34 for clinical indications (Figure 1).

3.2 | Screening cMRI at <8 years and ≥8 years after Fontan

Of the 107 screening cMRI's performed after Fontan palliation, 12 (11%) resulted in a management change. Of the 46 cMRI's obtained <8 years after the surgery, 8 (17%) resulted in a change in management. The average interval between Fontan palliation and first screening MRI was 41 months with a median of 40 months. Of the 61 cMRI's obtained >8 years after a Fontan, 4 (7%) resulted in a change in management (Figure 2). There was a near -significant greater odds of a change in management following a screening MRI obtained <8 years after Fontan as compared to ≥8 years (OR = 3.00, 95% CI: 0.84-10.67, *P* = .079).

3.3 | Clinically indicated cMRI at <8 and>8 years after Fontan

Of the 34 clinically indicated cardiac MRI's performed after Fontan palliation, 11 (32%) resulted in a management change. Of the 14 cMRI's obtained <8 years after the surgery, 6 (43%) resulted in a change in management. The average interval between Fontan palliation and clinically indicated cMRI was 41 months with a median of 25 months. Of the 20 cMRI's obtained ≥8 years after a Fontan, 5 (20%) resulted in a change in management (Figure 2). There was no significant difference in the odds of management changes following clinically indicated cMRI obtained <8 years after Fontan as compared to ≥8 years (OR = 2.25, 95% CI: 0.52-9.73, *P* = .273).

3.4 | Management changes after screening cMRI compared to clinically indicated cMRI, at any time

Twelve of all 107 screening cardiac MRI's resulted in management changes (11%). Eleven of all 34 clinically indicated cardiac MRI's resulted in management changes (32%). There was a significantly greater odds of a change in management following clinically indicated cMRI after Fontan as compared to screening cMRI (OR = 3.79, 95% CI: 1.48-9.66, *P* = .004). These findings are summarized in Figure 3.

3.5 | Management changes regardless of purpose for imaging

A comparison was also made between groups with the only variable being time <8 years or ≥8 years after Fontan palliation. Fourteen (23%)

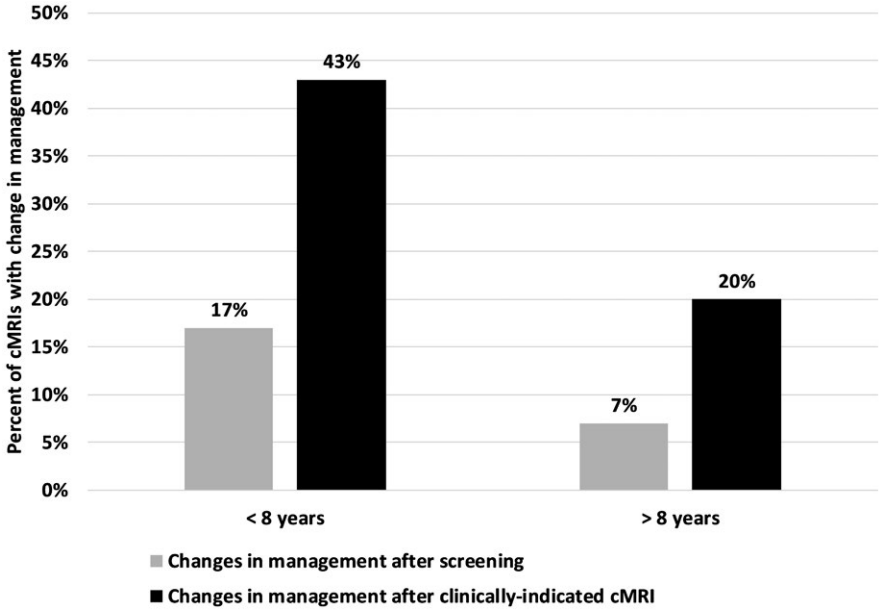

FIGURE 2 Percent of cMRI's with change in management, based on time since Fontan (P = .079)

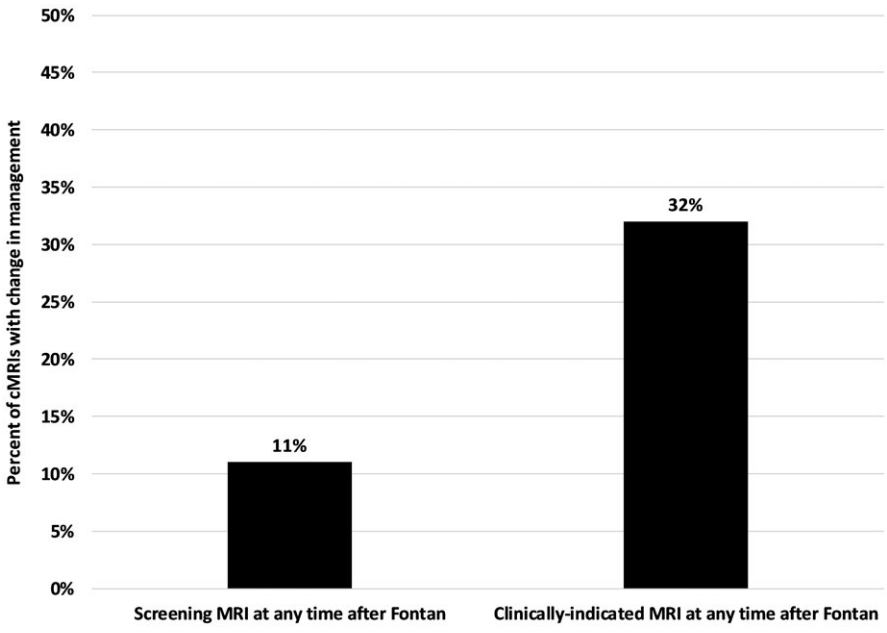

FIGURE 3 Percent of cMRI's leading to a change in management, based on indication (P = .004)

of 60 cardiac MRI's performed <8 years after Fontan resulted in a change in management. Nine (11%) of 81 cardiac MRI's performed ≥8 years after Fontan palliation resulted in a change in management. There was a near significant greater odds of a change in management following a cMRI obtained <8 years after Fontan as compared to ≥8 years (OR = 2.43, 95% CI: 0.97-6.08, P = .052). These findings are summarized in Figure 3.

3.6 | Types of management changes made after cMRI

The documented changes in management after cMRI are summarized in Table 2. These include changes to medications, cardiac catheterization, more imaging studies, pacemaker placement, and cardiac surgery. The most frequent single change in management across all groups was cardiac catheterization with angioplasty. While screening cMRI's did not appear to directly lead to any cardiac surgeries, this was a common outcome in clinically indicated cMRI ≥8 years after Fontan completion (Figure 4).

4 | DISCUSSION

In this large review of one center's experience with screening cMRI's for post-Fontan patients, we found that changes in management

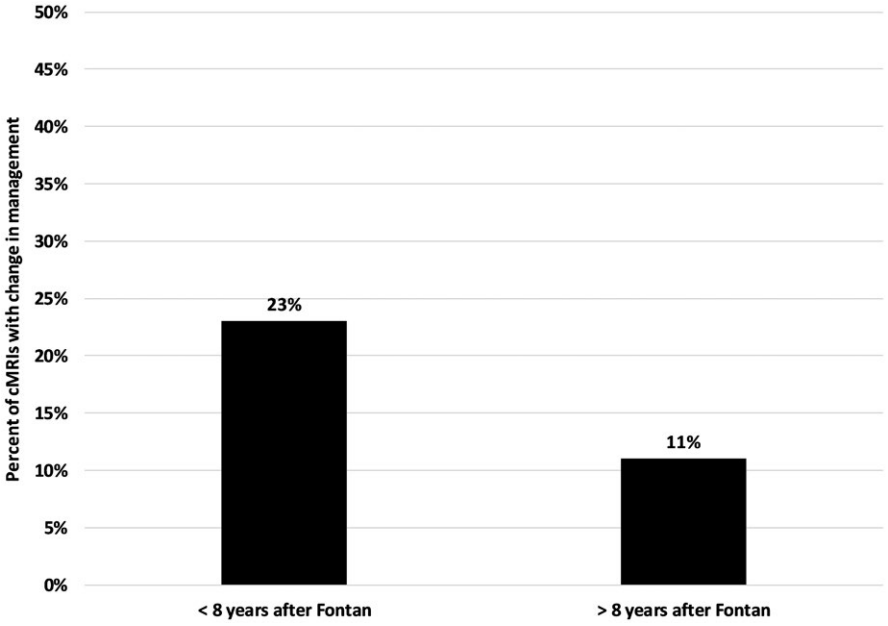

FIGURE 4 Percent of cMRI leading to a change in management, based on time since Fontan (P = .052)

TABLE 2 Summary of management changes after cMRI

Timing of management change	Management changes after clinically indicated cMRI	Management changes after screening cMRI
<8 years after Fontan	n = 6 (out of 14)	n = 8 (out of 46)
	• Hemodynamic catheterization (1) • Catheterization to close Fontan fenestration (1) • Catheterization for angioplasty (1) • Catheterization for dilation of aortic arch (1) • Unspecified catheterization (2)	• Catheterization for angioplasty (1) • Unspecified catheterization (3) • Increased frequency of cMRI (1) • Begin diuretic therapy (1) • Lung perfusion scan (2)
≥ 8 years after Fontan	n = 5 (out of 20)	n = 4 (out of 61)
	• Two-ventricle repair (2) • Fontan revision (1) • Pacemaker placement (1) • Fontan takedown (1)	• Catheterization for angioplasty (1) • Increased frequency of cMRI (2) • Hemodynamic catheterization (1)

occurred about three times as often after clinically indicated cMRI's but were not infrequent after screening cMRI. The reason for cMRI appears to influence the utility, as clinically indicated studies resulted in significantly more management changes than screening ones. Notably, the time since completion of a Fontan procedure is not a factor in this particular comparison.

The timing of cMRI appears to play some role in the utility of the study among our patient population. Imaging performed less than 8 years after Fontan palliation resulted in more management changes than imaging done more than 8 years after the surgery. This difference approached but did not reach statistical significance (P = .052). Notably, this comparison does not include the reason for obtaining a cMRI.

Among those cMRI's obtained for screening purposes, there appear to be more management changes when screening occurred less than 8 years after the procedure, with a P value approaching statistical significance (P = .079). One may compare this to the subset of cMRI's obtained only for clinically indicated purposes, in which there is no statistical difference (P = .273) in the number of management changes based on timing after Fontan completion. Given the potential benefit of identifying significant disease before it leads to symptomatic illness, the routine use of screening cardiac MRI should be considered, particularly in the first several years after the surgery. The exact timing and interval of such screening studies remains unclear.

As more patients with Fontan palliation reach their fourth decades of life it is increasingly important to rapidly diagnose or even preempt potential complications related to their palliated physiologic state in order to minimize their potential morbidities. The goal to screen at intervals after surgery is supported by prior studies showing

that risk of death in Fontan patients increases in both a constant and step-wise way after surgery.[1,5] For this reason, interval screening is the only way to identify new threats, such as emboli and stenosis.

We found that the most common change in management across all groups was referral for cardiac catheterization for pulmonary artery angioplasty after a finding of pulmonary artery stenosis. Prior studies have shown that leaks and stenoses are not uncommon at Fontan-related anastomotic sites[3] and that narrowing of the pulmonary arteries during palliation are correlated with worse outcomes.[12-15] Specifically, stenosis at these sites appears to be a key contributor to increased pulmonary artery pressures, a factor thought to be associated with development of plastic bronchitis among Fontan patients.[16] Tanase et al reported a case in which relief of pulmonary artery stenosis was strongly associated with relief of plastic bronchitis in Fontan patients.[17] Additionally, the distal branch pulmonary arteries are traditionally very difficult to fully visualize with echocardiography, particularly as patients age. It is possible that uncovering these stenoses before they became symptomatic may have prevented or delayed the onset of plastic bronchitis or other morbidities for these patients.

Our findings suggest that there is increased utility in screening cMRI performed less than 8 years after Fontan palliation when compared to screening exams done later. This is consistent with recent studies by Khairy et al and D'Udekem et al who examined causes of death among Fontan patients and suggested that the cumulative risk of death from thromboembolism increases during the first three years after Fontan palliation.[1,2] The authors did also note that this risk increases again 15 years after surgery but the data presented here on older Fontan patients is insufficient to either refute or support this hypothesis. Other authors[8] have reported that silent thromboemboli occur in about 17% of adult Fontan survivors. Given MRI's ability to detect emboli, future studies of our population may elucidate the benefits of screening MRI in the second decade after Fontan palliation.

Cardiac MRI appears to offer valuable information about anatomy and function in patients who are post-Fontan and frequently informs decisions about ongoing management in these complex patients. While cMRI is gaining in popularity as a method of disease surveillance, its role alongside other cardiac imaging modalities continues to evolve. The information obtained by cardiac MRI often complements that of both echocardiography and catheterization, signaling that it may one day replace catheterization as a surveillance tool when no acute intervention is needed.

5 | LIMITATIONS

Our study has several limitations. First, given the small numbers of overall events, we did not construct multivariable models controlling for various factors such as type of Fontan palliation. In our study, these were predominantly intracardiac and extracardiac baffles. If one type of Fontan were to have more complications, then more frequent or earlier imaging may be warranted for that subgroup. Second, while our protocol calls for cMRI's at three and eight years post-Fontan, the protocol was implemented in 2011, so we do not yet have any individuals who had screening cMRI's at each of those recommended intervals. In the future, we may be able to analyze the pros and cons of each specific timepoint. Third, 27 patients who had cMRI's were excluded, primarily because their cMRI happened in the immediate postoperative period. The reason for the early MRI could be either for a clinical indication or as part of a research protocol (eg, for many Y-graft recipients). It is unclear how this early imaging may impact the need for future imaging. Finally, unclear documentation led to some patients being excluded from the study and may have led to the inadequate capture of some management changes following cMRI.

6 | CONCLUSIONS

Cardiac MRI plays an important role in the management of the post-Fontan patient. For those with clinical indications, cMRI can clearly help identify important treatment strategies that may be needed. Our study shows that there is also an important role for cMRI in the routine surveillance of post-Fontan patients as well, even in the absence of symptoms. Future studies are needed to determine the optimal timing of such imaging in the long-term care of the post-Fontan patient.

CONFLICT OF INTEREST

There are no conflicts of interest to disclose.

AUTHOR CONTRIBUTIONS

Neil Zaki performed data collection and analysis, statistics, drafting article, and critical revision.

Michael Kelleman performed data analysis and statistics.

W. James Parks involved in the approval of the article, critical revision, and project concept.

Timothy Slesnick performed data interpretation, critical revision of the article.

Michael McConnell involved in the approval of the article, critical revision, and project concept.

Matthew Oster performed project design, data analysis, drafting article, critical revision, and approval of the article.

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