



International Journal of Cardiovascular Diseases & Diagnosis

Research Article

MR-Conditional Cardiovascular Implantable Electronic Devices in Patients With Congenital Heart Disease: A Real Gain? -

Grace HT Kwok^{1*}, Bram Ruijsink², Srinivas Narayan¹, Kuberan
Pushparajah¹, Henry Chubb¹, Yaso Emmanuel¹, Harith Alam¹, Eric
Rosenthal¹ and Alessandra Frigiola^{1,2}

¹Guy's and St. Thomas' NHS Foundation Trust, Westminster Bridge Road, Lambeth, London, SE1 7EH

²King's College London, Guy's Campus, Great Maze Road, London, SE1 1UL

***Address for Correspondences:** Grace HT Kwok, Guy's and St. Thomas' NHS Foundation Trust,
Westminster Bridge Road, Lambeth, London, SE1 7EH, E-mail: Grace.kwok@doctors.org.uk

Submitted: 26 February 2021; **Approved:** 08 March 2021; **Published:** 11 March 2021

Cite this article: Kwok GHT, Ruijsink B, Narayan S, Pushparajah K, Chubb H, Emmanuel Y, Alam H,
Rosenthal E, Frigiola A. MR-Conditional Cardiovascular Implantable Electronic Devices in Patients
With Congenital Heart Disease: A Real Gain? Int J Cardiovasc Dis Diagn. 2021 Mar 11;6(1): 017-024.

Copyright: © 2021 Kwok GHT, et al. This is an open access article distributed under the Creative
Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any
medium, provided the original work is properly cited.

ABSTRACT

Background: MR-conditional Cardiovascular Implantable Electronic Devices (CIED) enables patients with CIED to have MRI scans. While the safety profile of MR-conditional CIEDs is encouraging, artefacts from CIED are unavoidable. This study aims to provide a qualitative evaluation on the extent of CIED artefacts for patients with Congenital Heart Disease (CHD) undergoing Cardiac MRI (CMR).

Methods: This is a retrospective study of all patients with CHD and MR-conditional CIEDs who underwent CMR between 10/10/2011 and 04/09/2018 at our level one surgical centre for CHD. Patients were included in the study if they have a structural CHD, had a CIED inserted and had a CMR scan with the CIED in situ. Images were acquired on a 1.5T MRI Scanner. Low Specific Absorption Rate (SAR) mode was activated on the MRI scanner to keep SAR levels below 2 Watts/kg. Retrospective gated steady-state free precession cine MRI of the heart were acquired in vertical long-axis, 4-chamber view and the short-axis view covering the entirety of both ventricles (9 to 12 slices). The extent of artefacts on diagnosis were graded from 1 to 4 in cardiac anatomy, flows, volumes and overall diagnostic value by two cardiologists.

Results: 17 patients received a CIED at a median age of 18 years (range 3-56 years) and underwent a CMR scan within the studied timeframe. 59% of the scans were of acceptable to good diagnostic quality (grades 1/2, n = 10). Assessment of cardiac volumes was most affected (53% of scans graded 1/2), followed by cardiac anatomy (59% grades 1/2). Flow analysis was most robust (81% grades 1/2). Contralateral PM sites appeared to be associated with overall better quality.

Conclusion: Over half of the patients had an acceptable quality scan. Anatomy and volumes were significantly affected by CIED artefacts, which may have implications for accurate assessment.

Keywords: Congenital heart disease; Cardiovascular magnetic resonance; Cardiovascular implantable electronic devices; Fontan; Artefacts

ABBREVIATIONS

AOO: Asynchronous Atrial Pacing Mode; B-SSFP: Balanced Steady-State Free Precession; C/Contra: CIED Implanted on the Contralateral Side of the Heart; Cctga: Congenitally Corrected Transposition of the Great Arteries (Cctga); CHD: Congenital Heart Disease; CIED: MR-Conditional Cardiovascular Implantable Electronic Devices; CMR: Cardiovascular Magnetic Resonance Imaging; DILV: Double Inlet Left Ventricle; EF: Ejection Fraction; FFE: Fast Field Echo Sequence; HLHS: Hypoplastic Left Heart Syndrome; I/Ipsi: CIED Implanted on the Ipsilateral Side of the Heart; ICD: Implantable Cardioverter Defibrillator; IVC: Inferior Vena Cava; LPA: Left Pulmonary Artery; MRI: Magnetic Resonance Imaging; PA: Pulmonary Artery; PM: Pacemaker (Used Interchangeably With CIED in this Paper); RPA: Right Pulmonary Artery; RVOT: Right Ventricular Outflow Tract; SAR: Low Specific Absorption Rate; SV: Stroke Volume; SVC: Superior Vena Cava; TE: Time to Echo (Time Between Delivery Of The Radiofrequency Pulse To Receipt Of The Echo Signal); TGA: Transposition of the Great Arteries; ToF: Tetralogy Of Fallot; TR: Receptive Time (Time Between Successive Pulse Sequences Applied to The Same Slice); VA: Ventriculo-Arterial; VSD: Ventricular Septic Defect

INTRODUCTION

Cardiovascular Magnetic Resonance (CMR) has significantly improved the ability to quantitatively assess cardiac volumes, function, blood flow and interrogate whole heart and great vessel anatomy, without the limitation of body habitus and acoustic windows or exposing the patients to high doses of radiation or invasive procedures [1]. These advantages have significantly promoted the use of CMR for assessment of patients with Congenital Heart Diseases (CHD), who are generally left with residual haemodynamic lesions following initial repair and often requiring re-interventions during their life time. The ability to accurately and reproducibly assess volumes, function and anatomy are crucial for optimal timing of re-intervention and to guide on the technique of choice (surgical or percutaneous).

In addition to structural abnormalities, CHD patients have a high

burden of arrhythmias. Some of these arrhythmias may be intrinsic to the structural condition itself, for example, complete heart block in Congenitally Corrected Transposition of the Great Arteries (ccTGA) and supraventricular tachycardia in Ebstein's anomaly due to the presence of accessory pathways. Both post-operative complete heart block and atrial arrhythmias from surgical scarring lead to the implantation of pacemakers or implantable cardioverter defibrillators (Cardiac Implanted Electronic Devices (CIED)) in a relatively large group of patients [2].

Previous studies have suggested that over three-quarters of patients with a CIED will have a clinical indication for MRI over the lifetime of their device [3]. Having a CIED was once seen as an absolute contraindication for magnetic resonance imaging (MRI). However, advances in medical technology have enabled patients with implantable devices to undergo MRI scans [4,5]. These devices, named as MR-conditional CIEDs, have been promoted as safe alternatives for those who would otherwise have been deterred from MRI scans [6,7]. Albeit being safe, little has been discussed regarding the clinical utility of CMR in those with MR conditional devices. In patients with CHD, whom require regular scans for clinical follow up and monitoring purposes, the implications of artefacts arising from CIEDs can be significant and significantly hinder the clinical usefulness of CMR for assessment of the heart.

This study aims to provide a qualitative evaluation of the extent of CIED artefacts on CMR in CHD patients and to assess the clinical usefulness of the acquired information. Furthermore, we hypothesised that a CIED contralateral or away from the heart would yield fewer artefacts, therefore making the MRI assessment more accurate.

MATERIALS AND METHODS

All subjects (and/or a parent or guardian) gave informed consent for the anonymised use of their data. The local research ethics committee of the hospital trust has approved this study.

All CMR scans were performed at a major tertiary hospital in London, United Kingdom. Our study period was from 10/10/2011 to 04/09/2018. Patients were included in the study if they had been

clinically diagnosed with a structural CHD, had a MR-conditional CIED inserted and had a CMR scan with the CIED in situ during the study period. All ages, sex, indications for CMR and models of MRI-conditional CIEDs were included. Patients were excluded if they had not been formally diagnosed with a CHD or had MRI scans for non-cardiac reasons.

Cardiac MR imaging protocol

Prior to imaging, CIED were set to AOO mode (atrial pacing, no sensing) by the pacemaker technician. This mode is regarded as the 'MRI safe mode'. The CIED-technician was present throughout the study. All images were acquired on 1.5T MRI Scanners (PHILIPS, THE NETHERLANDS AND SIEMENS, GERMANY). Low specific absorption rate (SAR) mode was activated on the MRI scanner to keep SAR levels below 2 Watts/kg.

Cine imaging

Retrospective gated steady-state free precession cine MRIs of the heart were acquired in the vertical long-axis, 4-chamber view and the short-axis view covering the entirety of both ventricles (9 to 12 slices). Image parameters were TR 3.3 ms; TE 1.67 ms; flip angle 60°; slice thickness 10 mm; matrix 196 x 149; field of view typically, 300 x 360 mm; and temporal resolution 30-50 phases acquired during a single breath-hold per slice. Typical SAR was <50%. Assessment of Left Ventricular (LV) and RV volumes was performed by manual segmentation of short-axis cine images at end diastole and end systole (PHILIPS VIEWFORUM). End diastolic and end systolic volumes were calculated by use of Simpson's rule for each ventricle, and from these volumes, Stroke Volume (SV) and Ejection Fraction (EF) were calculated.

If the metal artefacts were significantly affecting the image quality and thus preventing accurate assessment, T1 FFE gradient echo sequences were performed. Typical parameters were TR 5.1 ms, TE 3.0 ms; flip angle 15°; slice thickness 8 mm; matrix 156 to 288; field of view typically, 300 to 350 mm, with a typical SAR < 15%.

3D imaging

3D volume imaging was performed using a b-SSFP sequence. Typical imaging parameters for 3D imaging were: TE 2.4 ms; TR 4.8 ms; voxelsize 1.6 mm isotropic; matrix 180 x 208 ; field of view typically 288 x 332mm.

Black blood imaging was performed to assess cardiac anatomy, especially for the right ventricular outflow tract and branch pulmonary arteries when stents were present or if no sufficient quality was achieved by the above mentioned sequences. Sequence parameters were: TE 30 ms; TR 1200 ms; flip angle 90°; slice thickness 7-8 mm; matrix 224 x 102; field of view typically, 163 x 358 mm, with a typically SAR < 10%.

Flow imaging

PA and aortic flow data were acquired by use of a flow-sensitive gradient-echo sequence. Parameters were TR 4.4 ms; TE, 2.7 ms; flip angle, 15°; slice thickness, 10 mm; and matrix, 136 x 130 during free breathing, typical SAR was < 5%. Image planes were located at the midpoint of the main Pulmonary Artery (PA), and just above the sinus level of the ascending aorta; similarly through plane flow data was acquired in the branch pulmonary arteries, Inferior Vena Cava (IVC), Superior Vena Cava (SVC), and in the abdominal aorta using two perpendicular planes in order to establish the correct

position. Through-plane flow data (30 phases per cardiac cycle) was acquired by use of retrospective cardiac gating. Arterial blood flow was calculated from phase contrast images by use of a semiautomatic vessel edge-detection algorithm (PHILIPS VIEWFORUM) with operator correction.

All volume and flow measurements were indexed for body surface area and expressed in mL/beat/m². After MRI scanning, the CIED was set back to the original mode and a CIED check was subsequently performed.

Artefacts assessment

The extent of artefacts and its implications on diagnostic abilities were assessed in four domains – cardiac anatomy, flows, volumes and overall diagnostic impression. 'Cardiac anatomy' refers to the visualisation of various cardiac structures; 'flows' refers to the feasibility of calculating the blood flow in the aorta, main PA and branch pulmonary arteries, SVC, IVC, Left and Right Pulmonary Arteries (LPA and RPA); 'volumes' refers to the volumetric assessment of the right and left ventricles in biventricular patients and the dominant ventricle in single ventricular patients; 'overall diagnostic impression' refers to the subjective impression of the overall adequacy of the CMR, and to what extent the CMR answered the clinical indication of the study. The quality of the CMR has been graded from 1 to 4 in each of the four domains by two cardiologists. The decision was mutually agreed in all of our patients.

Grade 1 – Minimal artefacts, good diagnostic quality;

Grade 2 – Moderate artefacts, acceptable diagnostic quality;

Grade 3 – Significant artefacts, limited diagnostic value;

Grade 4 – Severe artefacts, unable to interpret data.

DATA ANALYSIS

CMR scans with a score of 1 or 2 were regarded as diagnostically useful, while CMR scans with a score of 3 and 4 were regarded as scans with significantly impaired diagnostic use. Due to the small number of patients, in a number of cases only a single patient in a category, we refrained from statistical analysis and only presented descriptive data.

RESULTS

Patient characteristics

A total of 17 patients were included in the study (12 males, 5 females). The patients had a CIED inserted at a median age of 18 years (range 3-56 years) and had a CMR performed at a median age of 20 years (range 5-57 years). The main CHD diagnoses included Tetralogy of Fallot (ToF), ccTGA, ventricular septal defect (VSD), dextrocardia, hypoplastic left heart syndrome (HLHS), double inlet left ventricle (DILV) and various valvular defects. Indications for a CIED included sinus bradycardia, tachy-brady syndrome, atrial flutter, prolonged QT interval and complete heart block (see table 1). The main indication for CMR was for assessment of cardiac anatomy and function.

Pacemaker characteristics

7 of the inserted pacing devices were of the single chamber type, whilst 10 were dual chamber devices (including 1 Implantable Cardioverter Defibrillator (ICD)). 11 pacemakers were inserted in the left subpectoral region, 2 in the right subpectoral region and 1 in the abdominal region. All pacemakers except the one in the abdomen

were endocardial pacemakers. Considering the fact that 2 of the CHD patients had dextrocardia and 1 had mesocardia, a total of 6 pacemakers were inserted on the contralateral side or away from the dominant ventricle. Pacing device models included MEDTRONIC ADVISA DR MRI A3DR01, MEDTRONIC ENRHYTHM MRI SURESCAN EMDR01 and MEDTRONIC EVERA S DR SURESCAN DDMC3D4 (see table 1).

Prior to CMR, all of the CIEDs were configured in the MR safety mode. No device failure, defuncting of the devices or arrhythmic changes were noted after the scan. Additionally, none of the patients experienced any complications or severe discomfort at the device site after the scan. In Fontan patients (except the patient with an epicardial CIED), a dummy second lead was used to allow MRI compatibility despite pacing in single chamber mode. The dummy lead was coiled behind the generator for these patients.

Impact of artefacts

On overall diagnostic quality: CIED artefacts were visible in all scans, however, this did not significantly interfere with the assessment of cardiac structures or major vessels in 59% of the scans (grades 1, n=0; grade 2, n = 10). Outcomes of the quality assessment of all images are reported on table 2 figures 1, 2, 3 demonstrate the typical impact of CIED artefacts on cardiac imaging.

On cardiac anatomy, cardiac volumes and blood flow assessment: With regard to the assessment of cardiac anatomy, 59% of the scans were ranked as diagnostically useful (grade 1, n = 0; grade 2, n = 10); 41% of the scans were severely impacted by artefact rendering them unuseable for diagnostic purpose (grade 3, n = 5; grade 4, n = 2). The main reason for poor grading based upon the differences in magnetic susceptibilities of the implanted metal device

Table 1: Clinical characteristics of study participants.

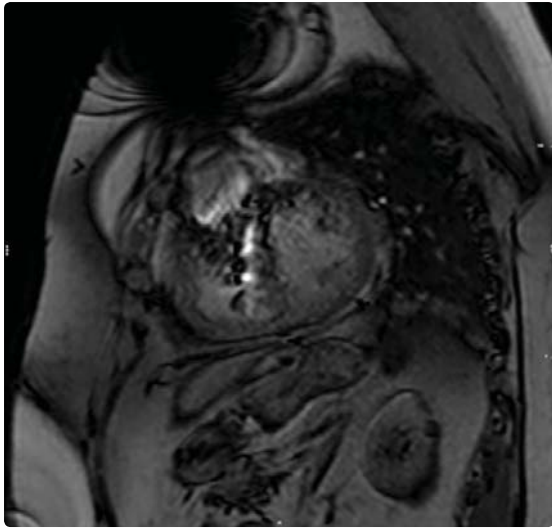
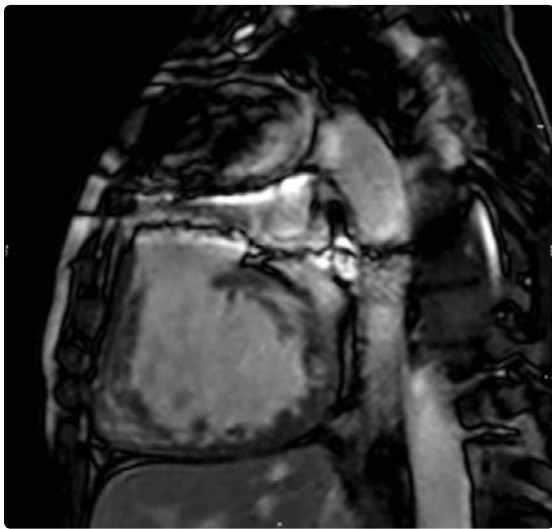
	Principle diagnoses	Main surgical procedure	PM Model	Indications for PM	Site of PM	Ipsi (I)/ Contra (C)	Type of PM	Indications for CMR	Overall grade
1	TOF	TOF Repair	Advisa	Sick sinus syndrome	Left subpectoral	I	Dual	Dyspnoea, tachycardia	2
2	ccTGA, dextrocardia, multi-level pulmonary stenosis	Fontan	Advisa	Sinus bradycardia	Left subpectoral	C	Single	Assessment of Fontan circulation patency, ventricular function	2
3	TGA	Senning	EnRhythm	Tachy-brady syndrome, atrial flutter, AF	Left subpectoral	I	Dual	Assessment of anatomy, function and atrial scar	2
4	TGA, VSD, dextrocardia	Fontan	Advisa	Unknown	Left subpectoral	C	Single	Assessment of Fontan circulation patency, ventricular function	2
5	Tricuspid atresia, VSD	Fontan	Advisa	Sinus bradycardia	Left subpectoral	I	Single	Worsening exercise intolerance	3
6	TOF	TOF Repair	Advisa	Tachy-brady syndrome	Left prepectoral	I	Dual	Recent atrial arrhythmias	2
7	ccTGA	Fontan	Advisa	Prolonged QT interval with ventricular arrhythmias; Sinus bradycardia on betablockers	Left subpectoral	I	Single	Anatomy and function assessment	4
8	Bicuspid aortic valve	ROSS	Advisa	Complete heart block	Left subcutaneous	I	Dual	Anatomy and function assessment	3
9	Hypoplastic heart, left atrial isomerism	Kawashima procedure	Advisa	Atrial flutter	Left subpectoral	I	Single	Anatomy and function assessment	2
10	Interrupted aortic arch type B	Aortic arch repair	Advisa	Post-operative heart block	Left submuscular	I	Dual	Assessment of aortic root dilatation and aortic arch	4
11	Hypoplastic left heart	Fontan	SureScan Not the true model name	Tachy-brady syndrome	Right subpectoral	C	Single	Worsening exercise intolerance	2
12	Congenital aortic stenosis	Ross operation	Advisa	Ventricular tachycardia	Right subpectoral	C	Dual, ICD	Anatomy and function assessment	3
13	ccTGA	ccTGA repair	Advisa	Complete heart block	Left subpectoral	I	Dual	Assessment of ventricular function and quantification of AVV regurgitation fraction	4
14	Double inlet left ventricle with VA discordance	Fontan	Advisa	Complete heart block	Sub rectus (Epicardial)	C	Dual	Anatomy and function assessment	2
15	Bicuspid aortic valve	Aortic valve replacement	Advisa D MRI A3DR01	Complete heart block	Left subpectoral	I	Dual	Anatomy and function assessment	2
16	ccTGA	VSD Repair	Advisa D MRI A3DR01	Complete heart block	Left subpectoral, mesocardiac	C	Dual	Non-specific fatigue and chest pain	2
17	Ebstein's anomaly	Epstein repair	Pacesetter St Jude Accent MRI PM2224	Atrial arrhythmia after radiofrequency ablation	Left subpectoral	I	Single	Dizziness and palpitations	3

Abbreviations: TOF: Tetralogy Of Fallot; PM; Pacemaker; Ipsi/I: Pacemaker on Ipsilateral Side of the Heart; Contra/C: Pacemaker on Contralateral Side of the Heart; CMR: Cardiac Magnetic Resonance Imaging; Cctga: Congenitally Corrected Transposition of Great Arteries; TGA: Transposition of Great Arteries; VSD: Ventricular Septal Defect; HLHS: Hypoplastic Left Heart Syndrome; DILV: Double Inlet Left Ventricle; VA: Ventrículo-Arterial

Table 2: Image quality scores and impact on diagnostic usefulness of CMR scans in patients with CIED.

Diagnostically useful?	Grade	Anatomy N = 17		Volumes N = 17		Flows N = 16		Overall N = 17	
		n	%	n	%	n	%	n	%
YES	1	1	41%	5	53%	4	81%	0	59%
	2	6		4		9		10	
NO	3	5	59%	5	47%	2	19%	5	41%
	4	5		3		1		2	

Impact on assessment of cardiac anatomy and quantification of cardiac volumes and blood flow separately and overall score for image quality of CMR scans. Expressed are n (%). See text for image score definition.

**Figure 1:** T1 imaging, short axis stack with significant pacemaker artifact over right ventricle.**Figure 2:** Right Ventricular Outflow Tract (RVOT) b-SSFP cine in tetralogy of Fallot patient. Note significant artefact of the PM impeding assessment of the RVOT dimensions.

and human tissue. Due to the ferromagnetic properties of the CIED and corresponding leads, local magnetic field inhomogeneities are created during CMR, causing severe distortion to the images.

Assessment of cardiac volumes was significantly impaired in 47% of cases (grade 3, n = 5; grade 4, n = 3), due to significant artefacts in

2D cine images, while in 53% of the cases cardiac volumes could be assessed reliably (grade 1, n = 5; grade 2, n = 4).

2D phase contrast flow imaging was the more robust among the four domains. Flow analysis could be performed with good or acceptable accuracy in 81% of the scans (grade 1, n = 4; grade 2, n = 9), whereas the remaining scans (19%) were of no clinical utility (grade 3, n = 2 or 4, n = 1) and did not allow accurate flow quantification. In one patient, phase contrast flow imaging was not performed as it was not indicated.

Site of pacemaker/ICD: 65% of patients had their CIED implanted on the ipsilateral side of the heart (n=11). 6 patients had the device implanted on the contralateral side or away from their heart: 2 patients had the device implanted in the right subpectoral site, 2 patients were dextrocardiac, 1 patient was mesocardiac and all three had the device implanted on the left subpectoral site. 1 patient had an epicardial pacemaker implanted in the abdomen. Image quality scores in patients stratified by location of the pacemaker is reported on table 3.

Out of the patients with an ipsilateral CIED, overall scan quality was diagnostically useful in 45% of cases, while for the contralateral group 83% of cases were useful. For cardiac anatomy, 27% of CMR scans in the ipsilateral CIED patients had sufficient diagnostic image quality (grades 1 and 2, n = 3 out of 11 ipsilateral CIED patients), compared to 67% in the contralateral group (grades 1 and 2, n = 4 out of 6 contralateral CIED patients). For cardiac volumes, quality of the CMR scans was sufficient for diagnostic purposes in 36% of patients with ipsilateral CIED (grades 1 and 2, n = 4), while 83% of cases with

**Figure 3:** Impact of artefact on black-blood imaging. Note that only limited artefact is seen.

Table 3: Impact of implantation side of CIED on image quality of CMR. Expressed are n (%). See text for image score definition.

Diagnostically useful?	Grade	Anatomy		Volumes		Flows		Overall									
		Ipsi (N = 11)	Contra (N = 6)	Ipsi (N = 11)	Contra (N = 6)	Ipsi (N = 10)	Contra (N = 6)	Ipsi (N = 11)	Contra (N = 6)								
YES	1	0	27%	1	67%	1	36%	4	83%	2	70%	2	100%	0	45%	0	83%
	2	3		3		1		5		4		5					
NO	3	4	73%	1	33%	4	64%	1	17%	2	30%	0	0%	4	55%	1	17%
	4	4		1		0		1		0		2		0			

Abbreviations: Ipsi: CIED implanted on the ipsilateral side of the heart ; Contra: CIED implanted on the contralateral side of the heart

a contralateral CIED had diagnostic useful CMR scans (grades 1 and 2, n = 5). Finally, assessment of blood flow was possible in 70% of ipsilateral CIED patients (grades 1 and 2, n = 7, out of 10), compared to 100% in the contralateral CIED group (n = 6). See figure 4 for an example of the decreased impact of artefact on cardiac imaging in a patient with a contralateral implanted CIED.

Fontan patients: 7 of our patients have had a Fontan procedure. In all patients, the grade assessors (cardiologists) were able to comment on the patency of the Fontan pathway. With regard to flow, 86% (n = 6) scans were graded 1 or 2 (grade 1 n = 1, grade 2 n = 5). While the flow in the aorta could be easily assessed, flows in the SVC, LPA and RPA were the most affected by susceptibility artefacts due to the pacing electrode. It was therefore not possible to comment of the presence of shunt through the Fontan fenestration and on collateral flow. However, it is worth noting that in one patient who had an epicardial pacemaker implanted in the abdomen, all flows could be seen reliably (all flows grade 1). For anatomy, however, only 57% (n = 4) of the scans were graded 1 or 2 due to PM artefacts (grade 1 n = 1, grade 2 n = 3). In terms of the overall assessment, 71% were graded 1 or 2 (grade 1 n = 0, grade 2, n = 5).

DISCUSSION

Need for lifelong monitoring

Recent medical advances in management of CHD have resulted in a larger proportion of CHD patients that are now able to live to adulthood. Nonetheless, patients with CHD often require multiple

interventions during their lifetime. Depending on the underlying anatomy and the extent of surgical repair, many of these patients develop long term complications years or decades after their initial procedures. The complexities of CHD and their surgical outcomes, coupled with the changing physiology from child to an adult, demand regular cardiac monitoring, including comprehensive imaging. Cardiac MRI is the current gold standard for quantification of cardiac function and great vessels flows and has taken a prominent place in monitoring patients with CHD, largely replacing diagnostic cardiac catheterisation.

The development of MR-conditional CIEDs has improved the safety profile of performing MR imaging in such patients [1]. Specific safety protocols, such as avoiding an MRI scan if the leads were implanted less than 6 weeks before, low SAR imaging and adaption of the pacing protocol to VOO/DOO in pacing dependent patients allow CMR imaging of patients with pacemakers[3].

However, the extent of imaging required in CHD, with multiple planes of imaging for functional, anatomical as well as flow assessment, raises the question as to whether CMR is suitable in CHD patients with MRI-conditional CIEDs for regular follow up.

In our study, all of our scans were affected by CIED-related artefacts. 57% of the scans obtained were of grade 1-2 diagnostic quality. As 43% of the scans were susceptible to a degree of diagnostic uncertainty (grades 3-4), this may hold clinical significance regarding future management plans.

Patients with single ventricle circulation

Although the main questions of interest were for assessment of anatomy and function, subgroups of various CHDs require different imaging strategies. Special considerations are required for patients with single ventricle physiology who were previously palliated with a Fontan circulation, a common pathway for various complex CHDs which would otherwise not be compatible with life. This subgroup, which is also commonly affected by arrhythmias, was the largest in our population.

Despite improved surgical techniques and short term outcomes, Fontan patients are subjected to long term cardiac, pulmonary and hepatic complications. Common cardiac complications include pulmonary arteries stenosis, ventricular failure, atrioventricular valve regurgitation, right atrium or lateral tunnel dilatation, pulmonary venous hypertension, and formation of blood clots and emboli [8]. These complications may be asymptomatic, or present as worsening exercise tolerance, dyspnoea and arrhythmias, warranting regular assessments of cardiac anatomy and function.

For a scan to be of good diagnostic value in these patients, the following parameters should be accurately assessed: (i) ventricular

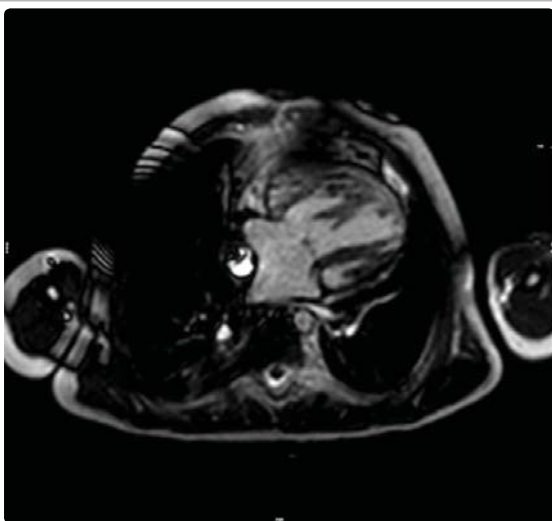


Figure 4: Four-Chamber view with contralateral pacemaker. Note that the artefact is away from the heart.

volumes and systolic function, (ii) patency of Fontan circuit inclusive of pulmonary arteries (iii) flow in the aorta, SVC, IVC, branch pulmonary arteries and in the pulmonary veins in order to calculate the shunt through the fenestration, amount of pulmonary-collateral flow, degree of aortic valve regurgitation and aortic valve function (iv) aortic arch anatomy in patients with hypoplastic left heart and previous arch reconstruction (v) exclude presence of cardiac clots [8,9].

It is worth noting that for all of our Fontan patients, a dummy lead has been used to allow MRI compatibility. To our knowledge, this technique of imaging Fontan patients has not been reported or extensively studied. Assessing and comparing the extent of artefacts between our 'true' single-chamber and 'dummy' single-chamber system would be useful for academic and clinical purposes. However, as only 1 of our patients had a 'true' single-chamber system, this would not be a representative comparison between the groups. Generally, our results have shown that while the flow in the aorta could be assessed in all of our Fontan patients, the SVC, LPA and RPA flows were severely affected by lead related susceptibility artefacts. This could be due to a combination of image distortion and slow blood flow. This poses key difficulties in assessing the status of the Fontan circuit. The artefacts from the CIED also affected the assessment of cardiac anatomy and volumes. In light of these, the usefulness of performing CMR in Fontan patients with a CIED is debatable. Therefore, such patients may have to undergo cardiac catheterisation for a more comprehensive assessment of the Fontan pathway patency, assessment of collateral flow and the quantification of shunt through fenestration.

Minimising the impact of artefacts

Site of CIED: Studies investigating patients with acquired heart diseases have reported good image quality when the pacemaker was located outside the view of the study [3]. Further to this, a previous study by Khan et al. suggested that right-sided implantation could minimise the degree of artefacts on cardiac imaging [10,11]. Abiding by the principle, it would appear to be sensible to implant the device on the left side of a patient with dextrocardia. In our study, 5 patients (36%) had a pacemaker on the contralateral side. The size of both groups is small and therefore statistical analysis would not be informative. Nonetheless, it is worth noting that 4 out of 5 patients with a pacemaker on the contralateral side had scans graded 1 or 2 (80%). On the other hand, only 4 out of 9 patients from the ipsilateral group had scans graded 1 or 2 (44%). Larger, multicentre studies would be able to provide a better assessment of the usefulness of contralateral implantation of devices on CMR quality in CHD.

As previously noted, CMR assessment of CHD often involves more comprehensive assessment of the circulation, including flow and anatomical imaging of the large vessels, when compared to acquired heart diseases. As artefacts do not only arise from the generator itself, but also from the sternal wires in patients who had previous surgical procedures, the observed benefit of contralateral placement of the CIED observed in acquired heart diseases cannot directly be translated to the CHD population. However, our results strongly suggest that contralateral placement of the CIED generator could be beneficial in patients with CHD.

Interestingly, the pacemaker implanted in the abdomen in one of the patients in this study resulted in the highest image score of all the Fontan patients. The lack of artefacts in PA and SVC flows

allowed for quantification of flow in the full Fontan circulation, enabling calculation of collateral blood flow. Although implantation in the abdominal wall may affect imaging of the abdomen, abdominal implantation may pose an advantage for those who may have to undergo ongoing extensive monitoring for cardiac complications, including developing of collateral arteries in Fontan patients. This epicardial device used endocardial leads sutured onto the epicardium as standard epicardial leads were not MRI-conditional. The off-label system in this patient was specifically chosen to allow MRI scanning and has proved useful. Longevity of the endocardial leads in this system is uncertain and therefore further experience is required before this can be widely recommended.

Pulse sequence: In the majority of our patients we used gradient echo imaging to obtain the 2D and 3D anatomical information. In very few patients we added non-balanced T1 FFE gradient echo imaging, however no significant improvement was seen in image quality. 3D volumetric imaging using balanced SSFP imaging is routinely used for anatomical assessment in congenital CMR. However, as its susceptibility to metal artefacts significantly disturbed the assessment in our patients with CIED, we added black blood spin echo imaging, which was less susceptible for artefacts. While maintaining a low SAR, this imaging allowed better assessment of vascular anatomy where needed.

Strengths and limitations: The purpose of our study was to evaluate the clinical utility of CMR scans in those with CIED in CHD, a group of patients requiring lifelong imaging and follow up appointments. Our study took place at a tertiary hospital with clinicians specialised in CHD. With a small group of targeted patients, we maximised our study period to 6 years and 6 months in order to include as many patients with structural CHD as possible. Nevertheless, our small cohort size of 17 patients in a single-centre study precluded statistical analysis, which limited the generalizability of these findings to other cohorts of CHD patients. Our study, however, would be invaluable as a descriptive study for the artefacts observed in those with CIED undergoing CMR scans.

CONCLUSIONS

In our study, CMR was safely performed in CHD patients with MR-conditional CIEDs regardless of site of implantation or Fontan circulation. However, artefacts significantly impact diagnostic quality of CMR scans in a large proportion of patients.

The utility of CMR scans for patients with a Fontan circulation is unclear based on this study, where physio-pathological interpretation of the scans was found to be difficult. Alternative CIED implantation sites should be considered for better image quality. Nonetheless, larger studies are required for further suggestions regarding its clinical usefulness.

REFERENCES

1. Ferreira AM, Costa F, Tralhão A, Marques H, Cardim N, Adragão P. MRI-conditional pacemakers: current perspectives. *Med Devices (Auckl)*. 2014 May 7;7:115-124. doi: 10.2147/MDER.S44063. PMID: 24851058; PMCID: PMC4019608.
2. Walsh EP, Cecyogeshksharma969@gmail.com, Chin F. Arrhythmias in adult patients with congenital heart disease. *Circulation*. 2007 Jan 30;115(4):534-545. doi: 10.1161/CIRCULATIONAHA.105.592410. PMID: 17261672.
3. Nazarian S, Beinart R, Halperin HR. Magnetic resonance imaging



- and implantable devices. *Circ Arrhythm Electrophysiol.* 2013 Apr;6(2):419-428. doi: 10.1161/CIRCEP.113.000116. PMID: 23592868.
4. Roguin A, Schwitter J, Vahlhaus C, Lombardi M, Brugada J, Vardas P, Auricchio A, Priori S, Sommer T. Magnetic resonance imaging in individuals with cardiovascular implantable electronic devices. *Europace.* 2008 Mar;10(3):336-346. doi: 10.1093/europace/eun021. PMID: 18308754.
 5. Yerra L, Reddy PC. Effects of electromagnetic interference on implanted cardiac devices and their management. *Cardiol Rev.* 2007 Nov-Dec;15(6):304-309. doi: 10.1097/CRD.0b013e31813e0ba9. PMID: 18090066.
 6. Harden SP. MRI conditional pacemakers: the start of a new era. *Br J Radiol.* 2011 Sep;84(1005):773-774. doi: 10.1259/bjr/86609066. PMID: 21849362; PMCID: PMC3473794.
 7. Nazarian S, Roguin A, Zviman MM, Lardo AC, Dickfeld TL, Calkins H, Weiss RG, Berger RD, Bluemke DA, Halperin HR. Clinical utility and safety of a protocol for noncardiac and cardiac magnetic resonance imaging of patients with permanent pacemakers and implantable-cardioverter defibrillators at 1.5 tesla. *Circulation.* 2006 Sep 19;114(12):1277-1284. doi: 10.1161/CIRCULATIONAHA.105.607655. Epub 2006 Sep 11. PMID: 16966586; PMCID: PMC3410556.
 8. Fredenburg TB, Johnson TR, Cohen MD. The fontan procedure: anatomy, complications, and manifestations of failure. *Radiographics.* 2011 Mar-Apr;31(2):453-463. doi: 10.1148/rg.312105027. PMID: 21415190.
 9. Geva T. Repaired tetralogy of Fallot: the roles of cardiovascular magnetic resonance in evaluating pathophysiology and for pulmonary valve replacement decision support. *J Cardiovasc Magn Reson.* 2011 Jan 20;13(1):9. doi: 10.1186/1532-429X-13-9. PMID: 21251297; PMCID: PMC3036629.
 10. Khan JN, Singh A, Pakkal MV, McCann GP. MRI-safe pacemakers and reduction of cardiac MRI artefacts with right-sided implantation. *Eur Heart J Cardiovasc Imaging.* 2013 Aug;14(8):830. doi: 10.1093/ehjci/jet022. Epub 2013 Feb 17. PMID: 23419285.
 11. Sasaki T, Hansford R, Zviman MM, Kolandaivelu A, Bluemke DA, Berger RD, Calkins H, Halperin HR, Nazarian S. Quantitative assessment of artifacts on cardiac magnetic resonance imaging of patients with pacemakers and implantable cardioverter-defibrillators. *Circ Cardiovasc Imaging.* 2011 Nov;4(6):662-670. doi: 10.1161/CIRCIMAGING.111.965764. Epub 2011 Sep 23. PMID: 21946701; PMCID: PMC3218212.