Original Article

Evaluation of the Extracardiac Conduit Modification of the Fontan Operation for Thrombus Formation using Magnetic Resonance Imaging

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Background: **The prevalence of thrombosis after the Fontan procedure depends upon the surgical technique used and the method of detection employed. Current investigations for thrombosis lack sensitivity and specificity or, in the paediatric population, require a general anaesthetic. We undertook a study to examine the feasibility of using magnetic resonance imaging (MRI) to detect thrombosis within the conduit, cardiac chambers and pulmonary arteries after the extracardiac conduit modification of the Fontan procedure.**

Methods: **Of the 50 children who had undergone this procedure at our institution between 1997 and 2002, 26 were eligible for, and 13 underwent, MRI study. The mean age was 10.2 years (range 8.2–16.8 years, median 9.5 years) and the average time from operation was 63 months (range 29–79 months, median 68 months). The mean age at Fontan operation was 4.9 years (range 2.1–10.5 years). Ten were on low dose aspirin, two were on warfarin and one was not anti-coagulated. In all cases, satisfactory imaging of the venous pathways and pulmonary arteries was obtained and there were no thrombi detected.**

Conclusions: **We conclude that MRI is a potentially useful tool for the detection of thrombus in patients who have undergone the Fontan operation.**

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Keywords. Fontan; Thrombosis; Magnetic resonance imaging

Introduction

Since the original Fontan operation was performed in 1971, several surgical modifications have been devised to improve haemodynamic efficiency and reduce the risk of complications.[1–6] The extracardiac conduit (ECC) is the latest modification, where the inferior vena cava is connected to the pulmonary arteries with a polytetrafluoroethylene conduit.

The presence of thrombus and embolic events after the Fontan operation has been reported with a variable prevalence depending upon the surgical technique, the population of patients studied and the modality of diagnosis used.[7–13] Transthoracic echocardiography for the detection of thrombus in this setting lacks sensitivity but is non-invasive. Transoesophageal echocardiography is more sensitive but in the paediatric population requires a general anaesthetic and for this reason it is not suitable for the routine surveillance of thrombosis.[14,15] The uncertainty regarding the prevalence of thrombosis after the Fontan operation has made the systematic evaluation of the role and efficacy of anti-coagulants difficult.

We describe a cross-sectional study to evaluate the potential of MRI to obtain images of sufficient quality to detect mural thrombi within the venous pathways, cardiac chambers and pulmonary arteries of paediatric patients who have undergone the ECC modification of the Fontan.

Materials and Methods

Eligible patients who had undergone an ECC Fontan were identified from the cardiology database at The Children's Hospital at Westmead, Sydney. All patients had a staged surgical repair, having previously undergone a bi-directional Glenn shunt. Children who were at least eight

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Table 1. *Details of Enrolled Patients*

Sex	Age (years)	Age at Fontan (years)	Months since Fontan	Diagnosis	Operations Prior to the Fontan
M	9.5	3.2	76	DILV, d-TGA	PAB, BDG, DKS, AS
F	8.5	4.7	46	MA, DORV, PA	BTS, BDG, AS
F	10.6	6.4	51	TA, PA	BTS, BDG
F	8.3	2.1	75	PA, IVS, HRV, ASD	BTS, BDG
F	10.6	5.9	56	DILV, d-TGA	PAB, BDG
M	8.3	5.9	29	l-TGA, VSD, DORV, HRV, PA, ASD	BTS, BDG
M	16.8	10.5	76	AVSD, HLV, PA, bilateral SVC	BTS, bilateral BDG
F	9.4	3.7	68	TA, VSD, PS	BTS, BDG
F	10.9	5.7	63	DILV, HRV, d-TGA, VSD, COA	PAB, CR, BDG, DKS, AS
F	11.3	5.2	73	DILV, d-TGA, VSD, HRV, COA	PAB, CR, AS, BDG, DKS
M	9	4.3	56	DILV, HRV, PA, VSD, ASD	BTS, BDG
M	8.7	3.0	68	TA, d-TGA, VSD, HRV	PAB, BDG, DKS
F	10.2	3.6	79	dextrocardia, HRV, CC-TGA, VSD	BTS, BDG

M: male; F: female; DILV: double inlet left ventricle; TGA: transposition of the great arteries; MA: mitral atresia; DORV: double outlet right ventricle; TA: tricuspid atresia; PA: pulmonary atresia; IVS: intact ventricular septum; HRV: hypoplastic right ventricle; ASD: atrial septal defect; VSD: ventricular septal defect; AVSD: atrio-ventricular septal defect; HLV: hypoplastic left ventricle; SVC: superior vena cava; PS: pulmonary stenosis; COA: coarctation of the aorta; cc: congenitally corrected; PAB: pulmonary artery band; BDG: bi-directional Glenn shunt; AS: atrial septectomy; BTS: modified Blalock Taussig shunt; DKS: Damus Kaye Stanzel procedure; CR: coarctation repair.

years old were selected to avoid the need for a general anaesthetic during the MRI scan. The study protocol was approved by the institutional ethics committee. Informed parental consent and patient assent was obtained.

The medical records were reviewed for the anatomical diagnosis, previous surgery, surgical technique and the type of anti-coagulation used.

MRI Protocol

MRI was performed on a Phillips© Intera 1.5 Tesla magnet machine. A Phillips© synergy phased array body coil was used. All imaging was done with quiet respiration. Cardiac triggering was used for all black blood T1 weighted sequences.

A series of T1 weighted (spin echo–echo planer imaging, echo time = 25 ms, repetition time = R–R interval) black blood images were acquired through the thoracic cavity. The cardiac chambers, ECC, pulmonary trunk, superior vena cava (SVC) and both pulmonary arteries were identified. A series of white blood images (single shot, balanced fast field echo) were acquired in the transverse and oblique coronal planes to demonstrate the ventricles, and each pulmonary artery in relation to the attached SVC and ECC. Flow dynamics were surmised by the flow artefacts on the coronal and sagittal white blood images. Only two children consented to the insertion of an intravenous line and the injection of contrast to perform CINE images. Thrombus was defined as any filling defect or wall irregularity within the conduit, pulmonary arteries or cardiac chambers and appearing bright on T1 sequences. The in-plane resolution was 0.6 mm × 0.8 mm. The MRI images were independently reviewed by one paediatric cardiologist (GFS) and one paediatric radiologist (EO).

Results

The details of the enrolled patients are outlined in Table 1.

Fifty children had undergone the ECC modification of the Fontan operation at our institution between 1997 and 2002. Twenty-six of these were eligible and thirteen agreed to participate. The mean age at the time of the MRI was 10.2 years (range 8.3–16.8 years, median 9.5 years). Eight were female and five were male. The mean time since the Fontan operation was 63 months (range 29–79 months, median 68 months). Ten were on low dose aspirin (3–5 mg/(kg day)), two were on warfarin and one patient was not compliant with anti-coagulation. Three patients had fenestrations performed at the time of initial Fontan surgery.

None of the patients had documented arrhythmia.

The mean saturation was 94% (range 85–98%).

The black blood images obtained showed good anatomic detail of the venous pathways and pulmonary arteries (Figs. 1 and 2). There was no evidence of conduit obstruction. No thrombi were detected within the conduit, pulmonary arteries or within the cardiac chambers.

Figure 1. *Coronal black blood image through the extracardiac conduit, superior vena cava and left pulmonary artery.*

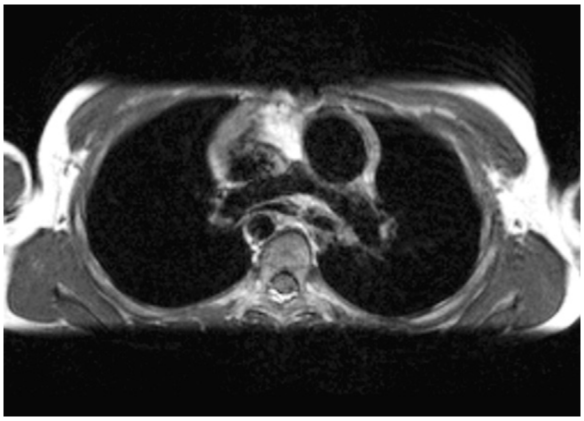

Figure 2. *Transverse black blood image through the pulmonary arteries.*

Figure 3. *Coronal white blood image through the extracardiac conduit, superior vena cava and left pulmonary artery.*

On the white blood sequences, the flow within the conduit was laminar and in 11 patients there was evidence of preferential streaming of blood from the conduit into the left pulmonary artery and from the SVC to the right pulmonary artery (Figs. 3 and 4). In the other two patients, who were the earliest studied, the images acquired did not allow assessment of flow patterns.

Discussion

There are a number of potential causes of thrombosis after the Fontan operation including slow or turbulent blood flow within the venous pathways, the use of thrombogenic foreign material in the circulation and a prothrombotic coagulation profile.[16–19]

The potential adverse effects of thrombosis after the Fontan operation are systemic embolisation and stroke,

Figure 4. *Transverse white blood image through the pulmonary arteries.*

major pulmonary embolus and recurrent pulmonary micro-embolisation possibly causing an elevation of pulmonary pressures and the subsequent failure of the Fontan circulation.

Studies on the prevalence of thrombosis after the Fontan operation and on the benefits of prophylactic anti-coagulation are hampered by the lack of a sensitive, non-invasive tool for the detection of thrombosis in this setting. Transthoracic echocardiography lacks sensitivity and whilst transoesophageal echocardiography has better sensitivity, it does not give good visualisation of all the venous pathways and pulmonary arteries and, in the paediatric population, requires a general anaesthetic.[14,15]

There has been one case report of the detection of a large intra-cardiac thrombus after the Fontan procedure by MRI.[20] However, there has been no systematic study of the use of MRI to detect thrombus in the venous pathways, cardiac chambers and pulmonary arteries.

Numerous in vitro studies and computational models of the fluid dynamics within the various modifications of the Fontan circulation have been reported.[3,21–23] Be'eri et al. describe an *in vivo* study, using phase velocity MRI (PV-MRI), of the flow dynamics after the atrio-pulmonary and total cavopulmonary connection types of Fontan operation.[24] PV-MRI allows the generation of flow vector maps and it also provides a quantitative analysis of flow dynamics within the Fontan pathways. It shows promise as a research tool to refine the haemodynamic efficiency of Fontan pathways and its use after the ECC modification should be analysed.

Fogel et al. have analysed the pulmonary blood flow dynamics after the lateral tunnel Fontan completion using CINE MRI.[25] Their technique of applying a presaturation pulse on the blood entering the image plane in order to produce a signal void, demonstrated preferential flow from the inferior vena cava to the left pulmonary artery and from the superior vena cava to the right pulmonary artery. In our study, the use of the white blood technique has confirmed this preferential streaming in the ECC Fontan and that the blood flow within the ECC is laminar. The white blood technique shows promise as a non-invasive clinical tool to examine the flow within the Fontan pathways.

The ability of MRI to detect thrombus was not compared with transoesophageal echocardiography in our study. Ethical considerations precluded such a comparison in otherwise asymptomatic children. The small patient numbers and the analysis at only one time point meant that the prevalence of thrombosis was not able to be determined. In addition the presence of extracardiac sources of thrombosis was not evaluated in this study. The presence of cardiac thrombi below the limit of resolution of MRI cannot be excluded. However, these limitations did not affect the aims of establishing a readily usable MRI protocol and evaluating the potential of MRI to detect cardiac thrombi of a size likely to be clinically important.

MRI provides good quality, high resolution imaging of the venous pathways, cardiac chambers and pulmonary arteries after the ECC modification of the Fontan operation.

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