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Review article

Cardiovascular, obstetric and neonatal outcomes in women with previous fontan repair

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ABSTRACT

Objectives: To determine cardiovascular, obstetric and neonatal outcomes of pregnancies in women who have a Fontan circulation.
Methods: A retrospective case note review of all women with a Fontan circulation who attended the joint obstetric cardiac antenatal clinic at St Mary's Hospital, Manchester (UK) between 2004 and 2016 was performed.
Results: In total, there were 19 pregnancies in 9 women with a history of Fontan repair. 23 women with univentricular physiology attended in this time period. 10 pregnancies (53%) resulted in live births; 1 in a stillbirth at 31 weeks gestation and 8 in miscarriage. Cardiovascular complications occurred in 2 pregnancies (11%). There were no thrombotic events, arrhythmias, myocardial infarction, or endocarditis in the antenatal or postnatal period. Obstetric complications included miscarriage (26% first trimester, 16% second trimester), along with premature delivery (24–36^{+6}) (80%) and fetal growth restriction (70%). The majority of women were delivered by caesarean section (60%).
Conclusions: Women who become pregnant following a Fontan repair carry an increased risk of cardiovascular complications. Fetal and neonatal complication rates are high and emphasize the importance of thorough, multidisciplinary, pre-conceptual assessment and counseling to allow patients to make informed decisions regarding future pregnancy.

Contents

Introduction

The Fontan surgical procedure has significantly improved prognosis and survival for patients with complex congenital heart

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disease characterised by univentricular physiology. Consequently, an increasing number of post Fontan repair patients reach childbearing age and contemplate pregnancy [1]. Pregnancy in post Fontan repair patients is associated with an increased risk of cardiovascular, obstetric and neonatal complications [1–7]. This increased risk may be attributable to cardiovascular changes associated with pregnancy; primarily volume loading and a hypercoagulable state. In order to enable post Fontan repair women to make informed decisions regarding family planning and pregnancy, it is vital that clinicians provide them with reliable and contemporary information [3].

Data from previous reports in this population of women is not recent and is derived from small case series [1–3], hence it is difficult to provide an accurate and validated risk assessment of pregnancy in such individuals, making pre-conceptual counseling challenging. To enable provision of appropriate information during pre-pregnancy discussions we reviewed the pregnancy outcomes of women who have undergone Fontan procedure and were managed in our tertiary centre.

Methods

Patient selection

All women with a history of congenital heart disease and univentricular physiology who attended the joint cardiac antenatal clinic at St Mary's Hospital, Manchester (UK) between 2004 and 2016 were identified from a local database. All women with an intrauterine pregnancy were included. Women attending only for pre-conception counseling and/or patients who had not undergone Fontan repair were excluded. Using our institutional medical records, we performed a retrospective case note review and data analysis to assess cardiovascular, obstetric and fetal outcomes relating to these women.

Data collection

Cardiac outcomes included arrhythmia, heart failure, deterioration in systemic ventricular function (defined as an echocardiographic reduction in systemic ventricular ejection fraction), myocardial infarction, endocarditis or thromboembolism.

Obstetric outcomes included miscarriage, fetal cardiac anomaly, pregnancy induced hypertension (PIH) (new onset hypertension: >140 mmHg systolic or >90 mmHg diastolic without significant proteinuria >20 weeks gestation), pre-eclampsia (PIH with clinically significant proteinuria: >30 PCR mg/mmol), postpartum haemorrhage (Vaginal delivery >500 mls, Caesarean section >1000 mls) and mode of delivery. Neonatal outcomes included prematurity (<37 completed weeks gestation), small for gestational age (birth weight <10th centile of that predicted for gestation, gender and ethnicity), stillbirth (fetal death in utero >24 weeks gestation) and neonatal death (within 7 days of birth).

Data analysis

All data are expressed as mean ± SD or median values. Incidence and 95% confidence intervals are included where appropriate.

Results

Between 2004 and 2016, 23 women with univentricular physiology attended the clinic. We excluded 12 women who attended for a preconception consultation but did not subsequently become pregnant. A further 2 women were excluded as they did not have a history of Fontan procedure. In total, 9 women with a total of 19 pregnancies were included in the final analyses.

Baseline obstetric data

Maternal age ranged from 19 to 34 years (median 23 years). Three women had a single pregnancy and the remaining 6 women had more than one pregnancy included in our cohort. There were 7 Caucasian and 2 Asian women. The range of body mass index was 22–31 (median 27). One patient was an active smoker and had 2 pregnancies. Two thirds of our women (67%) attended for pre conceptual counseling. Each pregnancy that reached the appropriate gestation had an anomaly scan at 20 weeks gestation (+/− 7 days) and a fetal echocardiogram in the antenatal period. All patients were followed up at regular intervals in the joint cardiac antenatal clinic for the duration of the pregnancy and were reviewed by an obstetric anesthetist with experience in the management of women with complex cardiac disease.

Baseline cardiac data (Table 1)

Of the 9 patients, 7 had a classical Fontan procedure with the remaining 2 having had a total cavo-pulmonary connection (TCPC). Mean age at Fontan surgery was 5.5 ± 3.4 years. In all patients, the morphological left ventricle was the systemic ventricle. Ventricular systolic function was preserved (LVEF >50%) in 7 patients, mildly impaired (LVEF 45–49%) in 1 patient (C) and moderately

Table 1
Baseline Cardiac Data.

Patient	Anatomy	Fontan Type	Systemic Ventricle	Ventricular Systolic Function	AV Valve Regurgitation	MVO2 (ml/kg/min)	NYHA Class	Previous VTE	Previous Arrhythmia	PPM	BB	ACE-I	VKA
A	TA, VSD	C-L	Left	Preserved	N	NA	1	N	N	N	N	Y	Y
B	TA	T	Left	Preserved	N	NA	1	Y	N	Y	N	Y	Y
C	TGA, HRV, VSD, PS	C-L	Left	Mildly impaired	N	18.9	1	N	N	N	N	Y	Y
D	DILV	C-R	Left	Preserved	Mild	14	1	N	Y	N	Y	N	Y
E	DOLV, CCTGA, PS	M?	Left	Preserved	N	NA	2	N	N	Y	N	N	N
F	DORV, VSD, PS	T	Left	Preserved	Mild	20.5	1	Y	N	N	N	N	N
G	PA, DEX, LAI	?	Left	Moderately Impaired	Moderate	9	1	Y	N	N	N	N	Y
H	TA	?	Left	Preserved	Mild	NA	1	N	N	N	Y	Y*	Y
I	PA, AVSD, TGA, APD, RAI	C-L	Left	Mildly Impaired	Mild	NA	1	N	Y	Y	N	N	Y

Anatomy: APD – Anomalous Pulmonary Venous Drainage, AVSD – Atrioventricular Septal Defect, CCTGA – Congenitally Corrected TGA, DEX – Dextrocardia, DILV – Double Inlet Left Ventricle, DOLV – Double Outlet Left Ventricle, DORV – Double Outlet Right Ventricle, HRV – Hypoplastic Right Ventricle, LAI – Left Atrial Isomerism, PS – Pulmonary Stenosis, RAI – Right Atrial Isomerism, TA – Tricuspid Atresia, TGA – Transposition of the Great Arteries, VSD – Ventricular Septal Defect.