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Brief Report

Pregnancy in atriopulmonary connection and total cavopulmonary connection – a comparison of two cases

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Since Fontan and Baudet[1] described the radical palliation for patients with tricuspid atresia, there have been many modifications to the principle and extension to other conditions where biventricular repair is not possible. There are reports of successful pregnancy in women having had Fontan-type surgery but the different techniques used are not clearly separated.[2] This article reports two pregnancies with a Fontan-type circulation from our centre, one in a patient with total cavopulmonary connection and the other with atriopulmonary connection, comparing and contrasting the complications.

Case reports

Patient 1

S.S., an unmarried 27-year-old woman, presented to Grownup Congenital Heart clinic in Malta when she was 14 weeks pregnant. She was born with double outlet right ventricle, uncommitted ventricular septal defect, and subvalvular pulmonary stenosis and had a total cavopulmonary connection with intra-atrial tunnel at Great Ormond Street Hospital, London, at the age of 8 years. She led an asymptomatic normal life thereafter, taking aspirin 75 milligrams daily.

An echocardiogram early in pregnancy showed normal left ventricular systolic function, no atrioventricular valve regurgitation, and unobstructed flow through the intra-atrial conduit and caval veins. Echocardiograms throughout pregnancy showed no change in the left ventricular dimensions or function and no inferior caval vein dilatation. Apart from bleeding at 6 weeks, her pregnancy was uneventful and she denied symptoms at monthly reviews. Aspirin was continued up to the delivery date. Her oxygen saturations were 95% on room air. Foetal growth on serial ultrasound scans was normal.

Following joint consultation, the obstetrician decided on elective Caesarean section to allow for optimal control, although normal vaginal delivery was considered possible. This was performed under epidural anaesthesia with intravenous unfractionated heparin cover at 36 weeks of pregnancy. A healthy girl weighing 2.4 kilograms was delivered with an Apgar score of 9 at 1 minute. The patient was discharged on subcutaneous enoxaparin for 1 month postpartum and then returned to aspirin. Postpartum echocardiograms showed normal ventricular function and no clot formation. To date, she remains well and symptom free.

Patient 2

S.P., with tricuspid atresia and normally related great arteries, had an atriopulmonary connection in Sydney, Australia, at the age of 8 years. She was on aspirin 75 miligrams daily since 11 years of age. She had a normal life with good exercise tolerance and no arrhythmias until 19 years of age when she developed atrial flutter. Transoesophageal echocardiogram showed two right atrial thrombi, and thus she was changed over to warfarin, keeping an

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Table 1. Summary of the characteristics of the patients before, during, and after pregnancy.

	Patient 1	Patient 2
Baseline characteristics (before pregnancy)		
Congenital defect	Double outlet right ventricle; ventricular septal defect; pulmonary stenosis	Tricuspid atresia
Surgical palliation	Total cavopulmonary connection (aged 8 years)	Atriopulmonary connection (aged 8 years)
Functional status	New York Heart Association class I	New York Heart Association class I
Saturations on room air	95%	91%
Arrhythmias	–	Atrial flutter
Ventricular function	Normal	Normal
Anti-thrombotic treatment	Aspirin	Warfarin
Pregnancy and delivery		
Age (years)	27	28
Drugs	Aspirin, unfractionated heparin peripartum	Sotalol, furosemide, enoxaparin
Cardiac complications	–	Left ventricular dilatation + increasing mitral regurgitation, arrhythmias, general weakness
Mode of delivery	Planned Caesarean section under epidural anaesthesia	Planned Caesarean section under epidural anaesthesia
Birth weight (g)	2400	2100
Postpartum		
Functional status	New York Heart Association class I	New York Heart Association class II
Arrhythmias	–	Persistent atrial flutter needing catheter ablation

international normalised ratio of 2 to 3. Later she underwent radiofrequency isthmal ablation but the flutter recurred 4 years later. She was started on oral flecainide, which helped maintain sinus rhythm, but she eventually stopped it as she remained in sinus rhythm without attacks of palpitations.

She sought preconceptual advice from the Grownup Congenital Heart disease clinic at the age of 28 years. Echocardiogram at the time showed normal left ventricular dimensions and function with trivial mitral regurgitation. Warfarin was changed to aspirin and she became pregnant. After an episode of self-terminating atrial flutter at 8 weeks of gestation, she was changed over to subcutaneous enoxoparin. She complained of further short-lived episodes of palpitations associated with facial flushing often precipitated by retching, and thus sotalol was started at a dose of 40 milligrams twice daily and the attacks stopped. An increase in the dose was attempted later in pregnancy but was not tolerated because of light-headedness and postural blood pressure drop. Despite this, she kept feeling "low" and generally unwell.

Monthly transthoracic echocardiograms showed progressive left ventricular dilatation and worsening mitral regurgitation. At 30 weeks of gestation, low-dose furosemide was added because of rising venous pressure and breathlessness. Her oxygen saturations on room air were in the low 90s throughout pregnancy. Liver enzymes and electrolytes remained normal. Serial foetal ultrasounds showed satisfactory growth.

The obstetrician opted for Caesarean section under epidural anaesthesia. This was performed at 37 weeks of gestation and a healthy boy weighing 2.1 kilograms was delivered with an Apgar score of 9 at 1 minute. She was discharged on enoxaparin and was changed back to warfarin after 1 month. Postpartum echocardiograms showed slow improvement in the left ventricular dimensions and degree of mitral regurgitation over a period of 12 months. Frequent short-lived palpitations continued and were partially controlled by sotalol. At 18 months after delivery, with normal left ventricular function when in sinus rhythm, she was referred for catheter ablation of her atrial arrhythmias.

The patient characteristics before and during pregnancy are summarised in Table 1.

Discussion

Pregnancy, with the increased blood volume and tendency for thrombosis, is a risk for patients with Fontan-type physiology. This can result in deterioration in ventricular function and increasing atrioventricular valve regurgitation. The inevitable elevation in right atrial pressure predisposes to tachyarrhythmias.[3–5] Reports show added risks to the foetus with an increased rate of spontaneous abortions and reduced intrauterine growth.[5]

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Owing to the tendency for sluggish blood flow in the Fontan circuit and the prothrombotic state of pregnancy, thromboembolic risk is considered high, although it could be lower in total cavopulmonary connection. Some authors suggest anticoagulation in most cases.[6,7] In retrospect, patient 2 was at higher thromboembolic risk in view of her atriopulmonary Fontan, past history of atrial flutter, and documented right atrial thrombi and would have benefited from being started on low-molecular-weight heparin as soon as she became pregnant.

Total cavopulmonary connection does not involve the right atrium, and therefore this is not subject to pressure or volume increases. In fact, long-term, total cavopulmonary connection has been shown to be associated with less atrial tachyarrhythmias.[8,9] Thus, one might anticipate that patients with total cavopulmonary connection would tolerate pregnancy better, which in our report appears to be the case. There are other factors that could contribute to a less favourable outcome. Patient 2 had a past history of atrial flutter before pregnancy and the arrhythmia recurred once the haemodynamic load started to increase at 8 weeks of gestation.

Although ventricular functioning at the beginning of both pregnancies was normal, the ventricle dilated in case 2 and perhaps there was also a fall in output, particularly when atrial flutter recurred. Pre-pregnancy exercise data could have given us insight into her cardiac reserve.

One cannot conclude that total cavopulmonary connection is a better operation for the woman and the pregnancy on the basis of single cases, but this comparison in women of similar age suggests that it may be. Arrhythmias already occurring before pregnancy are an adverse factor for deterioration and are likely to recur during pregnancy.

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