Cardiology in the Young

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Successful childbirth in a Fontan with protein losing enteropathy

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Brief Report

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Abstract

As survival rates and functional status of the adult single ventricle population have grown, some may become pregnant inadvertently or against our advice. The outcomes are often poor, being worse for the fetus/baby rather than the mother with a Fontan circuit. No reports of a successful delivery of a healthy baby to a Fontan mother with protein losing enteropathy were found in the literature. We present one such case.

As survival rates and functional status of the adult single ventricle population have grown, some may become pregnant inadvertently or against our advice. The outcomes are often poor, being worse for the fetus/baby rather than the mother with a Fontan circuit.

The fetal complications seen in mothers with Fontan circulation are the most alarming. The fetal demise rate in recently published literature is more than 50% and those that do survive having complications due to prematurity and intrauterine growth restriction.[1,2]

The maternal morbidity with pregnancy includes heart failure, arrhythmias, and post-partum haemorrhage.[2] A Fontan patient with protein losing enteropathy, with successful pregnancy, has not previously been reported.

Case report

We present a 21-year-old patient with Fontan circulation, recently diagnosed with protein losing enteropathy, who became pregnant and successfully delivered a healthy baby. The patient was born with unbalanced atrioventricular canal defect with hypoplastic left ventricle and hypoplastic aortic arch. She eventually underwent a fenestrated, lateral tunnel Fontan in which the fenestration spontaneously closed. She developed protein losing enteropathy 18 years after her Fontan, about 18 months prior to pregnancy. She had a history of atrial re-entry tachycardia with two prior cardioversions and sinus node dysfunction.

Prior to pregnancy she underwent cardiac catheterisation following her protein losing enteropathy diagnosis. Her Fontan pressures were favourable at 11 mmHg and trans-pulmonary gradient of 4 mmHg. Cardiac output was 3 L/min/m^2. Baseline saturation on room air was 90%. Her ventricular function on echocardiogram was normal. Due to her protein losing enteropathy and arrhythmias there was a discussion about proceeding to pacemaker placement. Her albumin prior to pregnancy was around 20 g/L. During pregnancy she was anti-coagulated with clopidogrel. This was chosen as it has not been associated with fetal or maternal complications. She had not had any thrombotic complications so low-molecular weight heparin was not started.

She presented at 8-weeks gestation. At 20-weeks gestation she was first hospitalised due to orthopnoea and oedema. She was discharged after administration of intravenous albumin, intravenous immunoglobulins, and diuretics. The fetus was monitored and showed re-assuring fluid levels and Dopplers. The fetal echocardiogram was normal.

At 24-weeks gestation she presented with poor home medication compliance and again had orthopnoea and oedema. She was admitted until delivery due to the high risk of fetal demise in a now viable fetus. Her labs were monitored closely and she was given albumin twice per week with a goal of keeping it above 2 g/dl and intravenous immunoglobin weekly. She was placed on oxygen for saturations in the high 80s with sleep. Arrhythmias during admission included sinus bradycardia, sinus pauses, and one episode of non-sustained ventricular tachycardia.

The fetus developed intrauterine growth restriction and was followed closely with fetal heart rate monitoring, umbilical artery Dopplers, and serial ultrasounds. The patient completed a course of betamethasone as well as Rhogam prior to delivery. The fetus developed abnormal Dopplers and delivery was planned by primary caesarean section. The patient developed pre-eclampsia without severe features prior to delivery. She underwent primary caesarean section at 32-weeks and 2-days gestation with epidural anaesthesia, which was converted to general anaesthesia for not achieving high enough level of neuroblockade.

Delivery was complicated with maternal hypotension and bradycardia which responded well initially to epinephrine infusion. This was likely contributory to the development of atrial

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re-entry tachycardia. During the post-partum time we were able to avoid positive pressure ventilation which we felt was best given her Fontan haemodynamics. She was discharged on post-operative day #7.

The baby's birth weight was 895 g and Apgars were 7 and 8 at 1 and 5 minutes. This was small for gestational age <10th percentile. She had a relatively uneventful neonatal course and was discharged at 2 months of age at a weight of 2010 g. She continues to do well with good weight gain and no structural heart disease.

The patient continued to have challenges with sinus node dysfunction and protein losing enteropathy post-discharge and underwent epicardial pacemaker placement 8 weeks after delivery. She has remained in sinus or atrial paced rhythm with an increased dose of Sotalol. She continues to be treated for protein losing enteropathy in Fontan with arrhythmias.

Discussion

There is a growing experience with Fontan patients and pregnancy. There is a high rate of fetal complications as well as an increase in maternal complications during pregnancy. To our knowledge, this did not include Fontan patients with protein losing enteropathy. This group is presumably at an even higher risk of complications. The largest risk is to the fetus with greater than 50% fetal demise. The fetus was small for gestational age, as is typical for Fontan pregnancies, and to date no additional adverse effects have been seen in the infant. One factor that may have been in the fetus' favour was that maternal oxygen saturations were 90% and we attempted to maintain this even with sleep with supplemental oxygen. Cyanosis is associated with poor fetal outcomes in other conditions. Certainly, it is too early to determine developmental outcome; however, fetal surveillance with electronic fetal monitoring and biophysical profile was always reassuring and post-natal growth has been excellent.

Maternal risks are less than that for the fetus. The patient developed more symptomatic oedema with the pregnancy; her own nutrition and non-fluid weight gain with the pregnancy were poor. Her arrhythmias though would be consistent with other reports of increased arrhythmias[2] with pregnancy and congenital heart disease. She appears to be back to her previous state of health, failing Fontan with symptomatic protein losing enteropathy, as to before pregnancy.

Conclusion

While we are not recommending pregnancy for Fontan patients with protein losing enteropathy we report a successful pregnancy in just such a case. Mom has returned to her previous state of health and the infant has not shown any other adverse effects other than being small for gestational age. A multi-disciplinary team working collaboratively and sharing their expertise is imperative to successful outcomes in pregnancies complicated by maternal congenital heart disease with multiple co-morbidities.

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Conflict of Interest. None.

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