Pregnancy in a Patient with Fontan Physiology: A Case Report

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Background

Since the introduction of the Fontan repair in 1971 and high survival rates of 60-85% at 20 years post-operative, it is likely that Obstetricians will encounter post-Fontan patients presenting for care during their pregnancy. It is important to be aware of the specific concerns of a pregnancy in a patient with Fontan physiology.

Case Presentation

A 19-year-old primigravida was transferred to a tertiary hospital with pre-term pre-labour rupture of membranes (PPROM) at 24+5 weeks gestation. She had a complex past medical history significant for congenital heart disease. She was born with an unbalanced atrioventricular (AV) septal defect with pulmonary atresia and underwent a neontatal systemic pulmonary artery shunt in early childhood followed by a bi-directional Glenn anastomosis and atrial septectomy at 2-years-old. She then had an 18mm extracardiac Fontan completion at 9-years-old.

Prior to pregnancy, the patient was on Lisinopril and Warfarin; she had no clinical signs of heart failure and led an active lifestyle. Her transthoracic echocardiogram showed moderate to severe AV valve regurgitation and severe left ventricular impairment but preserved contraction of the right ventricle with overall ejection fraction 40-45%. Her regular medications changed during pregnancy to aspirin 150mg daily and enoxaparin 40mg twice daily.

The patient was managed as per routine PPROM protocol with intravenous benzylpenicillin and oral erythromycin and received steroid loading. She had an antepartum haemorrhage at 28+5 weeks gestation with an estimated blood loss of 130ml with associated intermittent incoordinate contractions. At 29+0 weeks the patient went into pre-term labour. She had a slowly titrated epidural block with intravenous (IV) hydration and proceeded to have a normal vaginal delivery of a liveborn male infant. The duration of active second stage of labour was twenty minutes and there was active management of the third stage with the placenta delivered with controlled cord traction and misoprostol 800 micrograms per rectum and oxytocin 10 units given over one hour. A heparin infusion was commenced at 6 hours post partum and the patient was transferred to the Coronary Care Unit for monitoring. The postnatal period was uncomplicated.

Discussion

The Fontan repair is reserved for congenital heart defects characterised by one functional ventricle, including tricuspid atresia and hypoplastic left heart syndrome. It is characterised by creating a single ventricle serving systemic circulation whereby the right and left circulations are separated. During the procedure the right atrium is typically connected directly to the pulmonary arteries while the AV valve leading to the hypoplastic right ventricle is closed. In the absence of a functional right ventricle the right-sided circulation, responsible for perfusing the lungs, relies on a pressure gradient from the right to left atrium. This passive circulation is dependent on increased systemic venous pressures. Factors that can jeopardise this pressure system include increased pulmonary vascular resistance or poor vena caval return to due venous dilatation or hypovolaemia.

There are a myriad complications resulting from Fontan circulation, including peripheral oedema, hepatomegaly, ascites and pleural effusions from increased systemic venous pressure, atrial thrombus from non-pulsatile right-sided circulation, and arrhythmias from increased right atrial enlargement.

During pregnancy significant cardiovascular adaptation is required with cardiac output increasing thirty to fifty percent with associated vasodilatory effects of progesterone and prostaglandins resulting in decreased systemic vascular resistance. Due to the abnormal physiology created by the operation, these post-Fontan patients may be unable to meet the cardiovascular demands of pregnancy due to their inability to augment cardiac output resulting in heart failure. They may also develop arrhythmias in response to cardiac stretch, atrial thromboses due to the prothrombotic changes in pregnancy, or antepartum or postpartum haemorrhage in the setting of anticoagulation. The fetus of a mother with Fontan physiology is at risk of spontaneous miscarriage, intrauterine growth restriction, prematurity and a 5% risk of congenital heart disease.

During labour it is important to have peripheral IV access with line air filters and adequate hydration to avoid hypovolaemia. Vaginal delivery if preferred unless the woman has refractory symptomatic heart failure or arrhythmias, with a shortened second stage of labour and carefully titrated neuraxial anaesthesia. Management of the third stage should be active with preparation for post partum haemorrhage; it is recommended to avoid rapid boluses of oxytocin due to the risk of hypotension and arrhythmias, and ergometrine is contraindicated as it causes increased pulmonary resistance and systemic venous return. Specific anaesthetic concerns in this patient group include the avoidance of increased pulmonary vascular resistance, such as during general anaesthesia or nitrous-oxide induced atelectasis and hypercarbia, or compromising vena caval return due to supine positioning, venous dilatation due to epidural anaesthesia, or hypovolaemia secondary to dehydration.

With an estimated 70,000 post-Fontan patients worldwide, half of whom are women, it is expected that Obstetricians will be increasingly tasked with managing these patients through pregnancy.

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