Fontan Circulation in Pregnancy: Principles of Management

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Introduction

- Fontan circulation results in systemic venous blood travelling to the lungs without passing through a ventricle
- As per the modified WHO pregnancy risk classification, pregnancy in a woman with a Fontan circulation is class III-IV (significant risk of maternal morbidity or mortality)
- There is no pre-pulmonary pump (ventricle) in a Fontan circulation → reliance on passive filling → difficulty coping with normal physiological changes in pregnancy → may lead to arrhythmias, cyanosis, heart failure and impaired exercise capacity
- High risk pregnancy complications in woman with a Fontan circulation: high risk of miscarriage, antepartum and postpartum haemorrhage, premature birth, small for gestational age and neonatal death

Case Presentation

A 30-year-old nulliparous woman, who resided in a rural area, had a background of tricuspid atresia and had a lateral tunnel Fontan procedure in early childhood. She was on long-term anticoagulation with warfarin prior to pregnancy. She had a previous suspected embolic event during attempted transition to rivaroxaban. She was commenced on prophylactic enoxaparin at the beginning of her pregnancy and then was changed to therapeutic enoxaparin at 10 weeks gestation. Her antenatal care was transferred to a tertiary centre at 15 weeks gestation.

Throughout her pregnancy, she had regular antenatal care, CARPREG II risk scoring, NT-proBNP levels, transthoracic echocardiograms and tertiary fetal growth ultrasounds.

Transthoracic echocardiography at 15, 21 and 28 weeks gestation had stable findings with normal phasic flow within the Fontan with a small fenestration, preserved systolic function and unobstructed left atrioventricular valve with trivial regurgitation and no outflow tract obstruction. Her NT-proBNP and oxygen saturations on room air remained stable throughout her pregnancy. However, her exercise tolerance decreased throughout pregnancy with development of orthopnoea. From 26 weeks gestation, she used a wheelchair due to breathlessness and fatigue secondary to decreased exercise tolerance.

Central placenta praevia was noted on the tertiary fetal morphology ultrasound, which was otherwise unremarkable. Fetal echocardiogram at 24 weeks gestation was normal. She attended four tertiary growth ultrasounds at 24, 28, 30 and 32 weeks gestation and there was appropriate interval growth, with normal amniotic fluid index and umbilical artery Dopplers throughout. Central major placenta praevia was demonstrated in all ultrasound scans.

Interdisciplinary team meetings were held with the maternal fetal medicine, anaesthetics, and cardiology teams in attendance and elective caesarean section was planned for 37 weeks gestation in the context of major placenta praevia. Both elective and emergency plans were formulated.

Delivery Management Plan	Rationale
Transition from therapeutic to prophylactic anticoagulation prior to delivery	Reduce intraoperative bleeding risk
Four units of packed red blood cells crossmatched at all times	Antenatal and peripartum bleeding risk
Continuous oxygen saturation monitoring (maintain above 90% with supplemental oxygen as required), telemetry, arterial line placement	Risk of decompensation during delivery
Low dose combined spinal epidural anaesthesia	Reduce risk of decreased preload
Strict fluid balance monitoring with fluid restriction	Risk of heart failure
Calf compressors	Venous thromboembolism prophylaxis
Use of double particle filters and careful priming of all intravenous lines	Reduce risk of air embolism
Three doses of intravenous ampicillin (first dose at time of delivery)	Endocarditis prophylaxis
Not for undiluted intravenous oxytocin	Prevent haemodynamic responses including hypotension, tachycardia and electrocardiography changes
Liaison with cardiac anaesthetist if general anaesthesia required to determine ventilation strategy	Venous return required to generate pulmonary blood flow in Fontan circulation
Close observation in high-dependency unit or Intensive Care Unit and arterial line to remain in situ for 24 hours postpartum	Reduce risk of heart failure after autotransfusion phenomenon, with uterine blood entering systemic circulation
Maintain haemoglobin level above 80 g/L	Reliance on increased haemoglobin levels for oxygen carrying capacity and peripheral saturation

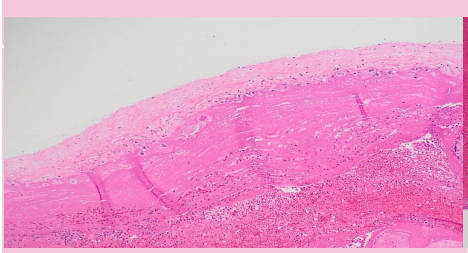
Delivery and Postpartum Course

The woman presented to Birth Unit with unprovoked painless antepartum haemorrhage at 31 weeks gestation and was admitted to the hospital. Decision was made to change from therapeutic enoxaparin to prophylactic unfractionated heparin due to its reversibility. However, she continued to have recurrent antepartum haemorrhage and, in view of persisting recurrent antepartum haemorrhage, decision was made for antenatal corticosteroids and then delivery. She gave birth to a 1670 g baby via caesarean section at 32 weeks gestation. Intraoperative estimated blood loss was 800 mL. Postoperatively, she was recommenced on therapeutic enoxaparin. She was transitioned to warfarin with bridging enoxaparin after a week. There was no cardiac decompensation during this period.

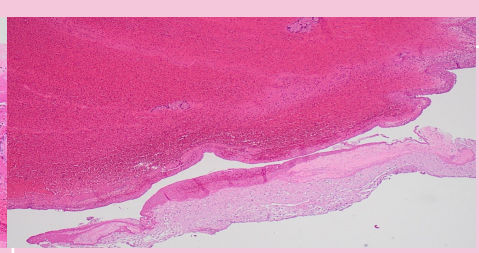
The baby was admitted to the Neonatal Intensive Care Unit for 14 days prior to being transferred to the regional hospital which was local to the mother's residence; the baby was on non-invasive ventilation for respiratory distress syndrome for four days and received caffeine for apnoea of prematurity.

Placental Histopathology

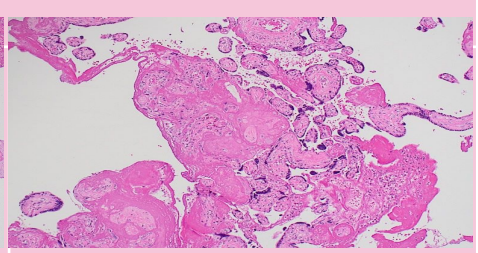
- Increased subchorionic, basal plate and perivillous fibrin deposition consistent with decreased flow and stasis in maternal circulation
- Haematoma formation consistent with recurrent antepartum haemorrhage
- Compensatory features of placental insufficiency/hypoxia with villous accelerated maturation (not shown)



Subchorionic fibrin



Basal fibrin with haemorrhage



Perivillous fibrin

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