Alagille syndrome in pregnancy complicated by intrahepatic cholestasis and chronic hypertension with superimposed preeclampsia

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Background

Alagille syndrome is a rare multisystem autosomal dominant disorder characterised by dysmorphic features, cardiac defects, and hepatic, renal, skeletal and ophthalmic abnormalities. The few published cases in pregnancy have demonstrated a strong association with fetal growth restriction, pre-eclampsia and preterm delivery.

Aim

- To highlight the challenges associated with managing a pregnancy in Alagille syndrome

Case & Results

We present a 30-year-old primipgravid with known Alagille syndrome and multisystem involvement including spinal canal stenosis, pulmonary artery stenosis with preserved right ventricular function, chronic kidney disease (stage 2) and cholestasis with intractable pruritis. Pre-pregnancy biochemistry included bile acid 9.2 µmol/L, ALP 408 U/L, GGT 691 U/L, ALT 106 U/L, AST 77 U/L, creatinine 80 µmol/L and urine PCR 13.2 mg/mmol. Her pregnancy was conceived by IVF with pre-implantation genetic diagnosis.

Antenatally, she experienced worsening pruritus refractory to all medical management. This coincided with peak bile acids of 34.1 µmol/L at 31+1 weeks. In addition to this, she was diagnosed with chronic hypertension in the first trimester and commenced on low-dose aspirin and labetalol. She developed superimposed pre-eclampsia at 28+5 with increasing anti-hypertensive requirements and creatinine rise to 102 µmol/L.

At 31+5, serial ultrasounds noted slowing growth velocity of the abdominal circumference (from 74% to 25%), intermittently raised UAPI with cerebroplacental ratio on the 5th centile and increased uteroplacental resistance. Due to this, she underwent elective c-section with steroid cover at 32+1. A healthy live male infant was born with birthweight 1,849 grams and APGAR 9 at 1- & 5-minutes.

Discussion

- Management of Alagille syndrome in pregnancy requires multidisciplinary input from gastroenterology, nephrology and cardiology
- Obstetric complications such as intrahepatic cholestasis of pregnancy and pre-eclampsia can be challenging to diagnose due to pre-existing biochemical derangements
- Comprehensive pre-pregnancy counselling and vigilant antenatal surveillance is vital to the optimisation of obstetric outcomes