A novel case of congenital fetal heart block in a mother with Addisons disease
Orefice R1 Paoletti D1 Robertson M 1,2
1Fetal Medicine Unit, Centenary Hospital for Women and Children, The Canberra Hospital, Canberra, Australia
2Australian National University Medical School, Canberra, Australia

A 33 year old G2 P1 was referred to our unit for management due to maternal Addisons disease. At 28 weeks during a planned fetal growth scan an unexpected persistent fetal sinus bradycardia was identified at rates of 45-70bpm. (Figure 1). The heart appeared structurally normal but enlarged (LV minor Z score 3.5 and RV minor Z score 2.6) with some trabeculation visible within the ventricular wall. (Figure 2).

A full autoimmunie antibody screen was performed and confirmed negative status for anti-Ro (SSA) and anti-La (SSB) antibodies. A full course of corticosteroids was given with a plan for a trial of salbutamol if the ventricular rate decreased further, and to deliver if there were signs of hydrops on ultrasound.

Fetal activity, growth and ventricular rate remained stable for the remainder of the pregnancy. Following a spontaneous pre labour rupture of membranes at 37 weeks and 4 days gestation an oxytocin induced labour resulted in a NVD of a 3304g LMI in good condition. The newborn was admitted to the Neonatal Intensive Care Unit for cardiac monitoring and was fitted for a pacemaker day three of life. He is currently thriving and meeting developmental milestones.

Addisons disease was once considered a contraindication to pregnancy 1. Due to treatment with corticosteroid supplementation, maternal mortality has been reduced from 45% in 1930 to 0.7% in 2000.2

Congenital fetal heart block is a rare condition affecting approximately 1 in 15 000 to 1 in 20 000 live births and is defined as a complete failure of normal conductions of atrial impulses to the ventricles.3 Approximately half of the cases diagnosed prenatally are due to structural abnormalities such as atrial septal defects and atrial isomerism 4

Congenital fetal heart block is strongly associated with the presence of maternal autoimmune antibodies which can cross the placenta, in particular, anti-Ro(SSA) and anti-La(SSB) antibodies, present in autoimmune conditions such as systemic lupus erythematosus, Sjogrens syndrome and rheumatoid arthritis 5. Ultrasound plays an important role in diagnosis and surveillance of complete heart block. Diagnosis is made by M mode (motion mode) or spectral Doppler techniques. M mode displays atrial and ventricular wall motion relative to time, thereby allowing determination of the relative timing of cardiac events.4

When the M mode line passes through the most contractile parts of the heart namely the free wall of the left ventricle and the right atrial appendage, the best trace will be achieved (Figure 3).

Treatment of the fetus with complete atrioventricular AV block is primarily expectant. In the absence of structural abnormalities, fetuses generally tolerate arrhythmias well when ventricular rates are greater than 60bpm 5. Attempts have been made to increase the fetal heart rate with administration of maternal beta-adrenergic agents and has been reserved for cases of FHR <55bpm or signs of decompensation, though this approach has not been evaluated by comparative studies.

To our knowledge, this is the first documented case of maternal Addisons disease with negative antibodies resulting in congenital fetal heart block. This case presented multiple diagnostic and management challenges and raised additional clinical questions. Congenital fetal heart block has never been reported in a case of Addisons disease, a known complex autoimmune disease, which raises the question whether there are other antibodies that may cause heart block. We hope that this case can help guide conservative management with close monitoring in cases of congenital heart block to avoid the short and long term complications of iatrogenic preterm birth.

References