Complete congenital foetal heart block: a case report

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Abstract:
Congenital heart block (CHB) is the most severe manifestation of neonatal lupus which can develop into a lethal atrioventricular (AV) block. Complete congenital foetal heart block related to maternal anti-Ro/SSA autoantibodies typically develops between 20 and 24 weeks of gestation. CHB with a structurally normal heart is frequently associated with maternal autoantibodies to Ro/SSA and La/SSB. We are presenting a case of foetal complete CHB with high maternal Ro/SSA and La/SSB titre with favourable outcome.

Key words: Congenital heart block, SLE, Ro/ SSA, La/ SSB, Dexamethasone.

Introduction
Isolated congenital heart block is defined as congenital heart block found in a structurally normal heart and when it is present with congenital heart disease known as complex congenital heart block. CHB with a structurally normal heart is frequently associated with maternal autoantibodies to Ro/SSA and La/SSB. Ro/SSA women have a 2% risk getting a child with CHB and the risk is about 20% among women who previously have had a child with CHB (Buyon et al., 2009). It is important to distinguish these two forms of CHB because they differ not only in their pathogenesis and in their rate of recurrence, but also in the prognoses of children affected. In fact, infants with CHB associated with severe structural heart disease have a poorer prognosis than infants with isolated CHB (Rosenthal, 2003) while the risk of recurrence is higher in mothers who have tested positive for anti-Ro/SSA antibodies. A mosaic of maternal, foetal, and possibly environmental factors might be involved in inducing CHB, but also the combination of such factors might be the way to induce the onset of CHB (Carolis et al., 2010).

Case report
A 23 years old G2P1L1 lady reported to our antenatal OPD at 24 weeks period of gestation (POG). In her last pregnancy, she underwent an emergency caesarean section at term for breech presentation and delivered a healthy male baby weighing 2.7 Kg. She also mentioned preeclampsia developed at 36 week period of gestation for which she was monitored by daily blood pressure and biochemical and haematological parameters.

On examination, she was averagely built with a BMI of 24.8 Kg/m² without any pedal oedema and normotensive with normal vital parameters. On abdominal examination, uterine height corresponds with POG and foetal heart sound was 66/min on Doppler (Fig. 1).

Her antenatal biochemical and haematological parameters were within normal limits. TSH was 3.08 and DCT was negative. Immunological tests revealed serum ANA moderately positive and SS-A (Ro) antibodies and SS-B (La) antibodies strongly positive. Ultrasound examination revealed a single live intra uterine foetus with normal liquor at 24w2d POG with FHR of 56-60/min. Foetal echocardiography showed complete heart block with structurally normal heart. Maternal echocardiography was within normal limits. She was started on dexamethasone (4mg/day), iron and calcium supplements. She was followed up by weekly ultrasound to follow the FHR pattern and FHR was stable at 52-56 beats/ min with no features of hydrops.
At 35w3d POG she underwent a caesarean section for preterm labour with post caesarean status and delivered a female baby weighing 2100 grams. The heart rate at birth was 60 beats/minute and APGAR score at 1 minute and 5 minutes was 7 and 9 respectively. Post delivery at NICU, the baby was stable at room air with heart rate of 60 beats/min. ECG showed a heart beat of 54/ min and echocardiography did not reveal any structural abnormality (Fig. 2). The baby was discharged after one week.

Fig 1. — Doppler showing foetal heart rate of 66/min

Fig 2. — Post delivery ECG showing heart rate of 54/ min
and carefully followed up on a weekly basis. Four months after delivery the baby did not require a pacemaker yet.

Discussion

CHB carries a significant mortality (20-30%, primarily foetal/ neonatal) and morbidity (67% require permanent pacing before adulthood) (Buyon et al., 1998). During pregnancy, the maternal autoantibodies cross the placenta and bind to cardiomyocytes, the atrioventricular (AV) conduction system is disrupted by inflammation with subsequent fibrosis and calcification leading to a complete AV block. A life-threatening cardiomyopathy (Nield et al., 2002) may be present in 10-15% cases. The most important risk factors for death in these patients are low birth weight, premature gestation, hydrops foetalis, endocardial fibroelastosis and diminished ventricular function. Patients who are diagnosed and treated in the neonatal period have a survival rate of 94% (Gupta et al., 2011).

Biomarkers such as prolongation of the foetal Doppler mechanical PR interval have not convincingly demonstrated utility in predicting advanced block (Friedman et al., 2008). Consistent with the fibrotic replacement of the atrioventricular node (AV) observed in autopsy studies from foetuses dying with CHB, reversal of a third degree block has never been achieved (Friedman et al., 2008). Current prophylactic and treatment strategies for CHB include maternal steroids, plasmapheresis, sympathomimetics, and in utero cardiac pacing (Buyon et al., 2009).

In most of the studies or case reports, the mother was treated with dexamethasone 4 mg daily after detection of foetal heart block and continued until the end of pregnancy. We also followed the same protocol with foetal echocardiography which showed a structurally normal heart. Although maternal tolerability of dexamethasone in our patient was excellent, dexamethasone may be associated with a structurally normal heart. Although maternal steroids, plasmapheresis, sympathomimetics, and in utero cardiac pacing (Buyon et al., 2009).

In conclusion, patients who are at high risk of developing CHB, frequent surveillance at 16-20 weeks of gestation is required because steroids may improve the outcome of the foetus in first and second degree heart block. The delivery should be planned in a tertiary care centre where pacemaker placement facility is available, when needed.

References

Friedman DM, Kim MY, Copel JA et al. Utility of Cardiac Monitoring in Fetuses at Risk for Congenital Heart Block.