Congenital Complete Heart Block A Case Report

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Abstract:
Congenital complete heart block or third-degree congenital atrioventricular block (CAVB) is seen in a fetus or in a neonate younger than 28 days. CAVB can occur in a structurally normal heart (isolated CAVB) or with congenital heart disease (complex CAVB with congenital heart defects). Congenital complete heart block occurs in approximately 1 per 15,000-20,000 live births. Structural congenital heart block is rare, but with a higher proportion of fetal loss. Patients with isolated CAVB typically have a better overall outcome than do infants with complex congenital heart disease isolated CAVB. The prognosis in complete (complex CAVB with congenital heart defects) CAVB is relatively good but may be influenced by the patient's age at presentation. Patients presenting as fetuses or at birth have significantly higher morbidity and mortality rates than do patients presenting later in childhood. Hereewith we are presenting a rare case of congenital complete heart block reported in a tertiary care hospital in Chennai.

Case report:
A 10 days old full term newborn female child weighing 3 kg was referred as having low heart rate and admitted with respiratory distress. The child was delivered by emergency caesarean section and cried immediately after birth. During the antenatal ultrasonography, fetal bradycardia was made out. The mother can...
which mother was investigated. She was positive for anti SSA(Ro) and anti SS-B(La) antibodies but was asymptomatic. There was no history of consanguinity, maternal fever, rashes, joint pain or drug intake. The patient was first in order of birth and there was no history of abortion or still birth. On examination, the child was pink and normothermic. There was marked respiratory distress. Respiratory rate was 68/min with intercostal and substernal retractions. Heart rate was 42 per minute.

The child was warm and all peripheral pulses were palpable. Cardiovascular system examination revealed the apex beat on left side in the fourth intercostal space. A pansystolic murmur of grade III intensity was audible all over precordium. Liver was 2 cm below left costal margin and the liverspan was 6.5 cm. Bilateral crepitations were heard in the chest. No other obvious congenital anomaly was found. The oxygen saturation was 94%.

Chest X-ray showed levocardia with cardiomegaly. On electrocardiogram, atrial rate was 150/min, and ventricular rate was 40/min with complete A-V dissociation. 2D-Echo revealed dilated cardiomyopathy, gross LA/LV dilatation, mild MR, pulmonary hypertension, physiological and global hypokinesia. The baby was negative for anti SS-A and anti SS-B antibodies. The child was diagnosed as a case of complete congenital heart block without congenital heart defect (isolated CAVB). The baby was managed with warmer care, oxygen inhalation, iv fluids and Inj. Isoprenaline(0.05 micrograms/kg/min) as stop gap procedure. Baby underwent pacemaker implantation. The baby is now on followup without any specific new complaints.

**Discussion:**

The incidence of congenital complete heart block has been estimated to be about 1 out of 15,000 to 22,000 live births (4,5). There is a tendency for female preponderance in CAVB (6). Approximately 25% to 33% of all congenital complete heart blocks are associated with congenital heart disease like ventricular septal defect, endocardial cushion defect, patent ductus arteriosus, mitral incompetence, persistent foramen ovale, transposition of great arteries and Ebstein's anomaly. The most common among these associated lesions is L-transposition of the great arteries. Our case did not have any heart defects. The association of collagen vascular disease in mothers of infants with congenital complete heart block is significant (7). About 60% of mothers who deliver children with CAVB have anti SSA and anti SSB antibodies. Mothers with SLE who have had one child with congenital heart block are at risk of having subsequent offspring with heart block (6). Maternal Lupus may not become manifest for years even after the birth of an infant with CAVB. In our case also the mother was asymptomatic. Maternal lupus is known to influence fetal and neonatal outcomes and is associated with increased incidence of obstetric complications such as stillbirth, abortion, prematurity, intrauterine growth restriction (IUGR), and neonatal complications such as neonatal lupus erythematosus syndrome, which is characterized by transient lupus dermatitis, hepatic and haematological abnormalities like hemolytic anemia, leucopenia, thrombocytopenia and or isolated CAVB, CCF, endocardial fibroelastosis (8). Skin rash, hepatitis and thrombocytopenia generally resolve without sequel. By contrast, the heart block is permanent and
requires a pacemaker in about 66% of cases (8). The presence of anti SS-A or SS-B antibodies in mothers and neonates with complete heart block has been clearly documented (9). These antibodies are IgG against SSA and SSB ribonucleoproteins, can cross the placenta and appear in the fetus at around the 16th week of gestation and affect the conduction system, cause sclerosis of the AV node. All fetuses are not affected. Other factors that could be responsible for the development of congenital complete heart block are HLA type, timing of antibody transfer, and in utero environment. The presence of certain HLA types (HLA-DR3, B8, DRW52, and DQW2) in the mother and fetus increase the possibility of complete heart block. The clinical features of the neonate depend on the effect of heart rate on cardiac output. At birth, the infant may present with congestive heart failure, anasarca, hepatomegaly and metabolic acidosis requiring emergency pacing. Isolated CAVB is only rarely accompanied by CCF (6) which was observed in our case. It is becoming increasingly recognised that dilated cardiomyopathy (DCM) either fetal or postnatal is a rare but serious outcome of autoimmune congenital complete heart block. Our case had dilated cardiomyopathy. The cause of the cardiomyopathy is unclear but may be related to an autoimmune myocarditis occurring in utero rather than primarily caused by bradycardia. The SSA/Ro and SSB/La antibodies have been suggested to be a major determinant (10,11) in the patients with CAVB with DCM. Evidence for a potential immunopathologic role of the SSA/Ro and SSB/La antibodies in CAVB is described in several studies (12,13). Immunofluorescent studies have shown IgG and IgM deposition throughout the myocardium on postmortem examination (14,15). However, affected newborns often appear asymptomatic and may have accelerated ventricular rates approaching those of healthy newborns. An associated finding in isolated CAVB may be the presence of discoid skin lesions. Our case did not have any skin lesions. Children with structural heart defects may present with cyanosis, failure to thrive or recurrent pneumonias or may be completely asymptomatic in childhood (such as children with L-transposition of the great arteries and intact ventricular septum). Prolonged QTc is not a constant finding in congenital block but is common with associated malformations and symptomatic patients. In asymptomatic patients, prolonged QTc may herald the onset of symptoms (16). There is an approximately 95% twenty year survival of patients without anatomic heart defect (17). After birth, electrocardiography (ECG) is recommended to assess for CAVB and to assess the QT interval that can be prolonged. Neonatal assessment should include a measurement of anti-Ro and anti-La antibody levels. Echocardiography should be performed initially and in periodic follow-up care in affected fetuses, infants and children to assess ventricular function and size and to rule out congenital or acquired cardiac malformations or valve dysfunction (18). In utero management of fetuses with transuterine fetal cardiac pacing have been tried without success (19). Administration of steroids, immunoglobulins and plasmapheresis in the mother are useful in first and second degree heartblocks. The use of intravenous dopamine and isoproterenol in babies has shown some success. Pacemaker therapy is needed in symptomatic babies. Temporary pacing can be done by transcatheter, transesophageal and transvenous routes. However permanent pacemaker is eventually needed for most infants. It is of two types namely

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epicardial and endocardial pacing. The indications for pacing are HR < 50 per minute, atrial rate > 140 per minute, atrioventricular block pauses lasting > 3 seconds, features of CCF, low cardiac output, ventricular dysfunction, post surgical CAVB.

References:


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