Case Report 1

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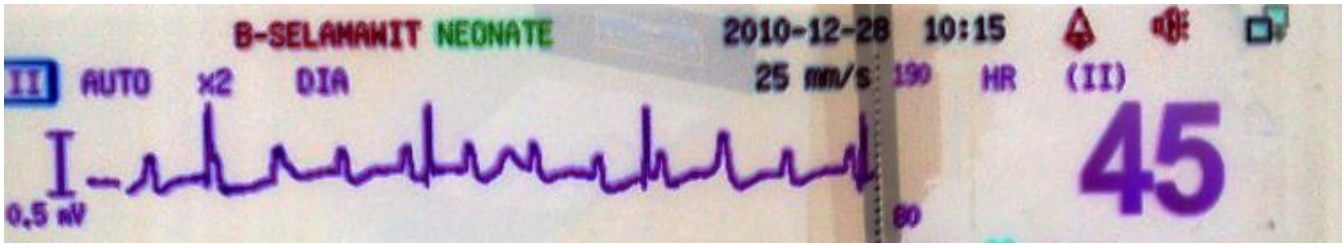
This case report is about a female neonate who was admitted to the NICU of Girum General Hospital at the age of 3hr on Dec, 28, 2010 after she was delivered at another private hospital.

The baby was born to 25 years old para I lady at gestational age of 34^{+5} week by date. Delivery was by emergency C/S for severe maternal preeclampsia. It was a twin pregnancy and this baby is twin B with birth weight of 1680gm and apgar score of 6 and 7 at 1st and 5th minute respectively. Twin A is also an alive male baby with birth weight of 2.5kg. The pregnancy was uneventful till two weeks back before the delivery when the mother was diagnosed to have hypertension. Since then the mother was taking methyl dopa for the hypertension.

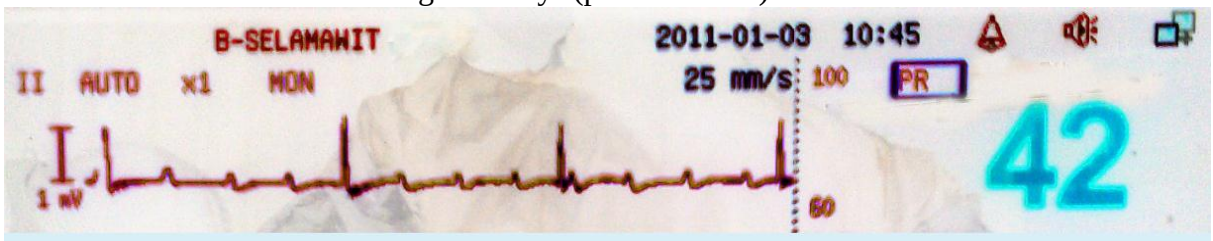
This baby was referred to our NICU for respiratory distress and admitted to our NICU at the age of 3hours on 28/12/2010/. At admission she had heart rate of 37beat per min and respiratory rate of 68 breath per min with mild intercostals retraction. There was no organomegaly and edema. She was alert and had appropriate neonatal reflexes.

After detecting the brady cardia, ECG was taken and revealed third degree AV block. Neonatal serum electrolytes, Echocardiography, CBC, and maternal and neonatal ANA and rheumatoid factors were determined and found to be normal. From this Isolated congenital heart block was considered. After communicating with pediatric cardiologist, atropine, drenaline and dexamethasone were given with the intention to raise the heart rate; but the heart rate persisted with in the range of38 to 45 beats per minute despite the effort. The baby stayed for a week in the NICU and discharged after writing a case report for possible pace maker implantation abroad and arranging follow up at the pediatric cardiologist. The purpose of the case report is to bring the case in to medical professionals attention and so that better ideas will be forwarded on how to manage and handle such cases in our set up. Besides this, isolated congenital heart block is a rare condition and this will give us the chance to review literatures on the issue.

- ECG Taken at admission to the NICU i.e. on Dec 28/2010 (picture below)



ECG Taken on 3/1/2011 at the age of 6days (picture below)



Literature Review

The incidence rate of congenital heart block is about 1 in 11000 to 1 in 20000 per live births (1). Autopsy studies of fetal hearts with congenital heart block and born to mothers having auto antibodies indicated that there is an exaggerated apoptosis most pronounced in regions containing conductive tissue and this is probably thought to be caused by IgG binding and also ingestion by macrophage initiated by the maternal antibodies. Persistence of this pathologic change after birth may be is the cause of heart block seen in infants postpartum (2). In a mother with autoimmune disease, the maternal immune system forms antibodies which includes anti-sjogren's syndrome A(SSA/Ro) anti-sjogren's syndrome B(SSB/La), ribonuclear protein (RNP) and DNA anti bodies. These auto anti-bodies can induce inflammatory damage to tissues which can lead to clinical autoimmune diseases. Auto antibodies can cross the placenta and target fetal or neonatal antigens leading to neonatal lupus syndrome (3).

Isolated Congenital heart block occurs commonly in babies born from mothers with rheumatic disease like SLE and sjogrens syndrome even though some of the mothers can be initially asymptomatic and later develops the symptoms (4). Follow up of babies born to mothers with connective tissue disease and positive autoantibodies showed that they are at risk of developing neonatal lupus syndrome in addition to cardiac rhythm alteration (5). Rashes, cytopineas and hepatobiliary disease are the most common clinical manifestations as compared to heart block and cardiomyopaties which are known to pose significant morbidity and mortality (3). In a pregnant lady with SLE during pregnancy, the maternal serum levels of anti-Ro/SS-A and anti-Ro-B auto antibodies do not predict exactly the occurrence of congenital heart block in the fetus **(6).**

Congenital heart block is known to occur due to many reasons. Cardiac malformation, presence of maternal antinuclear antibodies and reasons other than these two have been mentioned as possible causes (7)**.** Review of the Finish hospital registry revealed that most babies with congenital heart block (90%) are born to mothers with antibodies to SSA or SSB (8). With proper follow up during the pregnancy, the median time for the detection of bradyarhythmia due to the congenital heart block is about 23 weeks of gestational age **(9).** Presence of hydrops and cardiac malformation found to predict poor out come. Sympatomimetic treatment for the cases with cardiac malformation and hydrops didn't show any benefit (8).

Screening for auto immune disease has to be considered for those mothers who have clinical clues of auto immune disease or if there is strong family history of auto immune disease. Pregnancy related – hormones can also trigger auto immune disease. Evaluation for maternal auto antibodies also has to be done when there is any clinical evidence suggestive of neonatal lupus syndrome. These screening tests include ANA, SSA and SSB. Similar tests has to be done for the infant if neonatal lupus is suspected beside other tests like complete blood count, liver function test, ECG and Echocardiography (3). In fetuses at risk to develop heart block, fetal kinetocardiogram can be used for accurate measurement of AV conduction time and detection of first degree AV block and helps to decide early management (10).

Recent review of papers indicates that the efficacy of steroid treatment is inconclusive in treating congenital heart block in the fetus (1). But previous reports had shown that dexamethasone was successfully used in treating fetal myocardial dysfunction and

dysrhythmia in a mother with sjogrens syndrome (11). Similar finding was also reported about the beneficial effect of corticosteroids in improving the fetal cardiac hemodynamics and conduction system in the presence of maternal auto antibodies **(12).** On the other hand, fluorinated steroids were shown to be useful for fetuses with incomplete heart block and hydropic changes as compared to those who have received no intervention (10, 13). The risk of developing antibody mediated congenital heart block in the new born was reduced by administration of steroids starting early in pregnancy (14).

Analysis of the Research Registry for the Neonatal lupus of New York University School of Medicine found that the cumulative probability of three year survival to be 79% after following a cohort of 113 infants who have congenital heart block born to mothers whose sera contain anti-SSA/Ro or anti-SSB/La antibodies, or both. Of those who are live born, 67% of them required pace makers (9). Isolated congenital heart block due to maternal rheumatic disease can lead to death in the early neonatal period of which up to one third of them found to die during this period and those who survive, most of them require pace maker (4). Myocardial dysfunction due to cardiomyopathy is one cause of death due to congestive heart failure. This can occur even with early implantation of cardiac pace maker (15). Factors like presence of maternal auto antibodies, increased heart size at initial evaluation and failure to improve after pace maker implantation are found to be predicting factors for the development of cardiomyopathy **(16).** Progression of incomplete heart block to higher degrees of heart block years after the neonatal period is also possible. Other ECG abnormalities in the neonate include transient sinus bradycardia, QT interval prolongations, and wolff-parkinson-white syndrome (3). Babies with auto anti body mediated second degree and above heart block are reported to have retarded growth with no catch up growth during the infancy period **(17).** Review of a 30 years experience of a single institution has shown that patients with isolated congenital atrioventricular block have different mortality rates depending on the time of diagnosis of the block. Patients with the heart block having diagnosed during fetal time have higher mortality rate in the first two decades of life than diagnosis during neonatal and child hood time. Similar difference in the need of pace maker implantation among the groups was also observed in the same study (18). Congenital heart block associated with structural heart defect as well as presence of hydrops, and lower atrial and ventricular rates in isolated congenital heart block were associated with higher fetal or neonatal deaths (19, 20, 21).

The congenital heart block has a risk of recurrence in the subsequent pregnancy with two to three folds higher in a mother who had an affected baby as compared to those who never had an affected child **(9).** But in another different study the risk of recurrence in such condition is found to be low (22).

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