

A Case Report on the Perinatal Management of a 30-week Preterm Baby with Congenital Complete Heart Block

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Abstract

Introduction: Congenital complete heart block is an uncommon condition in the newborn, but is known to occur with maternal systemic lupus erythematosus. **Clinical Picture:** This paper presents one such baby with complete heart block who was born premature (after a gestation of 30 weeks) and weighing 759 g. **Treatment:** Continuous isoprenaline infusion was initially used to support the baby while her other neonatal problems were treated. A Medtronic VVI pacemaker was subsequently inserted to maintain a heart rate that would be more physiologically acceptable for the patient. **Outcome:** This baby is currently thriving well, having been followed up for one year. **Conclusions:** The management issues, encompassing maternal and neonatal problems, and a review of current literature on this condition are discussed.

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Introduction

The association between congenital complete heart block (CHB) and maternal autoimmune disorders^{1,2} has been described for many years. Maternal systemic lupus erythematosus (SLE) is the commonest of these disorders, and a leading cause of heart block in newborns. Extensive clinical, immune and histopathologic research^{3,4} have been conducted for this interesting phenomenon. The pathogenesis is related to the passive transfer of auto-antibodies across the placenta, which presumably destroy the conduction system. The consequence of this is a wide spectrum of clinical presentation⁵ ranging from hydrops, stillbirths to newborns with congestive heart failure from inadequate systemic output.

The management of these pregnancies presents a challenge to the team and usually calls for close multi-disciplinary collaboration involving the obstetrician, rheumatologist, neonatologist and paediatric cardiologist. The heart block may present as early as the second trimester of fetal life. Together with this, and secondary features such as the accumulation of fluid in the third space, the baby may be disadvantaged even at birth. Complications

such as premature delivery and the attendant problems add to the complexity of the issues. The case described below features one such baby with CHB and the various problems associated with prematurity which compounded her management.

Case Report

This infant was born after a gestational period of 30 weeks to a mother with SLE as the fetus developed complete heart block and there was evidence of severe intrauterine growth retardation. The diagnosis of maternal SLE was made 7 years ago when she was 22 years of age. There was a previous bad obstetric history of a miscarriage and a stillbirth at 30 weeks. Close obstetric monitoring was therefore provided throughout this pregnancy and she was on low dose prednisolone for control of the SLE.

An early onset of intrauterine growth retardation in this baby was reported in the second trimester and congenital CHB was diagnosed at 28 weeks. At 29 weeks, oligo-hydramnios and growth retardation were present, and these became progressively severe requiring the baby to be delivered a week later. Two doses of intramuscular

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dexamethasone were given prior to delivery in an attempt to accelerate fetal lung maturation.

At birth, the baby weighed 759 g. The APGAR score was 2 and 3 at 1 minute and 5 minutes, respectively; and ventilatory support was commenced. CHB was confirmed on a postnatal ECG with a ventricular rate of approximately 42 beats per minute. The baby was not hydropic and there was no accumulation of excess third space fluid in the serous cavities. We started her on intravenous isoprenaline, initially at a dose of 0.2 mcg/kg/minute and this was gradually increased to 0.6 mcg/kg/minute. This brought the basal ventricular rate up to a maximum of 65 beats per minute.

The baby developed problems of prematurity with respiratory distress syndrome manifesting shortly after birth. Intratracheal surfactant was given with good results, and weaning of the ventilator settings could be achieved, with the baby taken off mechanical ventilation at day 13 of life.

Echocardiography performed at the time of birth showed normal cardiac connections. There was a patent foramen ovale and a large patent ductus arteriosus. The myocardium was hypercontractile with a fractional shortening of 45%. The ductus was successfully closed with a course of indomethacin which reduced the left to right shunting.

During the first month, there were complications of sepsis and necrotising enterocolitis. Which were treated aggressively with antibiotics. The baby was given intravenous nutrition and started on feeds from the third week. Finally, these problems resolved and full feeds were established by the sixth week.

The baby also had thrombocytopenia with a platelet count of 70,000/mL. This may be a haematologic manifestation⁶ of the neonatal lupus. Platelet transfusion was given intermittently to boost the count. There was also deranged liver function with raised transaminases and a

conjugated hyperbilirubinaemia.⁷ Several factors may also be responsible for this, apart from lupus involvement. These included cholestasis from prolonged parenteral nutrition, sepsis and enterocolitis. Fortunately, the problem resolved with time.

Meanwhile, the baby had been maintained on a continuous infusion of isoprenaline. The dose was brought down from 0.6 to 0.1 mcg/kg/minute to maintain a heart rate of approximately 60 beats per minute (Fig. 1a). When the baby became more stable on the sixth week, the infusion was discontinued as intravenous access became difficult, and the baby had remained relatively stable. Plans for an insertion of an epicardial pacemaker were arranged and her parents were consulted. She was observed for cardiac decompensation but showed no signs of heart failure, except for cardiomegaly that was present from birth.

A permanent epicardial pacemaker was surgically implanted at 2 months of life. Her weight had by then reached 2 kg. The surgery was carried out under general anaesthesia and was uncomplicated. A Medtronic VVI pacemaker was inserted (Fig. 2) and set at a rate of 90 beats per minute (Fig. 1b). The epicardial pacing threshold was 0.5 volts. The baby was discharged a week later. She remained well and was established on full oral feeding. Follow up revealed a healthy baby who is growing appropriately.



Fig. 1a. 12-lead ECG of baby with complete heart block on isoprenaline (ventricular rate of 60/minute).



Fig. 1b. 12-lead ECG of same baby after pacemaker implantation (rate set at 90/minute, note pacemaker spikes).

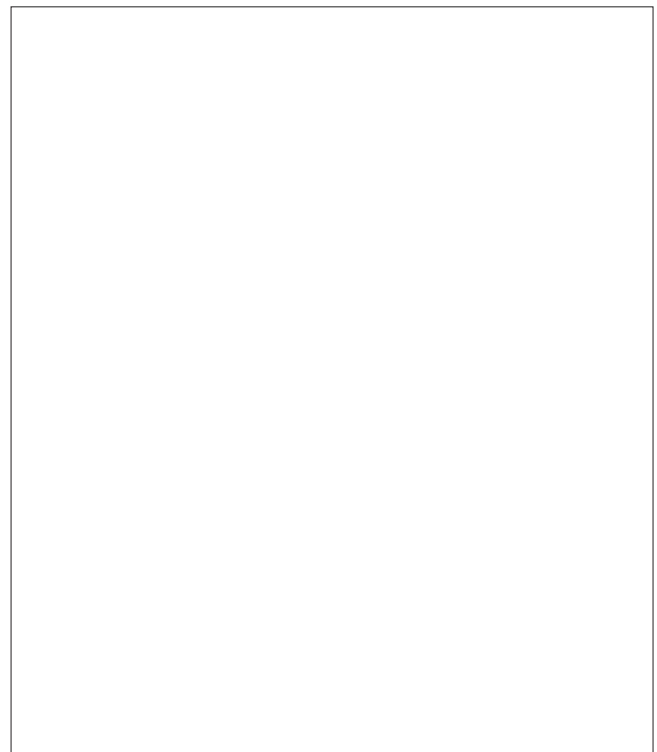


Fig. 2. X-ray demonstrating position of VVI pacemaker in-situ and position of the epicardial lead.

Discussion

Maternal SLE, with the production of autoantibodies, in particular anti-SSA/Ro and anti-SSB/La, can cause deleterious effects on the fetus, resulting in the neonatal lupus syndrome.⁸ It is believed that these antibodies cross the placenta and may affect the heart, skin, liver and blood. When antibodies attack the conducting system of the fetus, heart block results. This is an irreversible feature compared to the non-cardiac manifestations which are generally more transient and tend to resolve at 6 months of life. This also approximates the time when maternal autoantibodies disappear from the infant circulation.

Interestingly, for the same autoantibodies that may be present, there is a wide spectrum of clinical severity. Why certain systems are affected in some babies but not in others is not certain. It is also unknown why the antibodies, which have a predilection for the fetal conduction tissue, seem to spare the maternal heart.

CHB in infants of SLE mother is associated with a definite incidence of morbidity and mortality.⁹⁻¹¹ The risk of mortality in the hydropic baby with an extremely low heart rate is even higher. Permanent pacemakers are required in well over 50% of these patients in the newborn period in several series published.^{8,12} There is also a significant mortality rate.^{8,13} To improve the outcome of these babies, the approach to management should begin with pregnancy. Advances in imaging technology allow the obstetrician to detect fetal well-being and growth. On a general note, the growth of the fetus should be monitored closely. More specific to the cardiovascular system, a fetal echocardiogram¹⁴ is essential in diagnosing cardiac involvement. Heart rate and rhythm, size, structure, contractility and the presence of a pericardial effusion are important features. This should be performed at around 16 to 18 weeks of gestation. Serial evaluation in the follow-up is extremely valuable.

Several therapeutic regimes have been tested in pregnancy, including the use of steroids,¹⁵⁻¹⁷ sympathomimetics,¹⁸ immunoglobulins¹⁹ and plasmapheresis.^{10,20} Dexamethasone given to the mother orally becomes available to the fetus in an active form as it is not metabolised by the placenta. Another consideration is whether the CHB may be a result of a concurrent myocarditis.^{21,22} Suppression of the inflammatory reaction is a plausible explanation, and resolution of pleural, pericardial effusion and ascites have been reported following its use. However, it has not prevented or reversed CHB. The efficacy of dexamethasone, sympathomimetics and/or plasmapheresis, however, has not been clearly demonstrated to justify its routine use in all mothers with SLE in pregnancy. In the absence of controlled studies, these therapeutic regimes are probably still experimental and best reserved for the severely affected

and hydropic fetus.

In a recent report by Deloof and workers,²³ a staged approach to the premature, hydropic fetus has been proposed. In the two cases described, intravenous isoprenaline was used to increase the heart rate, followed by insertion of temporary epicardial leads. This was in the interim period while awaiting growth before placement of a permanent pacemaker. Fetal ventricular pacing²⁴ and staged pacing therapy²⁵ are further discussed as therapeutic strategies in other reports where hydrops was secondary to CHB.

In this case, management was made more difficult by her low birth weight, premature state and the various complications associated with prematurity. The attendant problems of prematurity had to be dealt with on top of a compromise from a slow ventricular rate. The low weight also made surgical epicardial implantation of pacemaker technically more demanding. The size of the pacemaker and the leads in proportion to the small size of the preterm baby would be significant. An important difference from the report by Deloof is that this baby was not hydropic and did not require temporary pacing in the first few weeks. This possibly improved her prognosis.

We opted to deal with the baby's complications of prematurity first. These included, among others, hyaline membrane disease and necrotising enterocolitis. The heart rate was boosted using a continuous intravenous isoprenaline infusion to a maximum of about 65 beats per minute. Closure of the patent ductus arteriosus was considered a priority as it was likely to aggravate the respiratory distress, may predispose to necrotising enterocolitis and was an added stress on the heart. Indomethacin was used, with surgical option kept in mind if necessary. Fortunately, the arterial duct closed with medical treatment, and the other problems of prematurity also resolved.

The use of a continuous isoprenaline infusion was a temporary measure to support the baby as she was too small and too ill in the initial weeks for a permanent pacemaker insertion. Temporary measures such as transcutaneous, transoesophageal pacing are also difficult and may pose significant morbidity. We therefore opted to adopt a more conservative treatment of boosting the heart rate with an isoprenaline infusion while carefully looking out for any cardiac decompensation from a low output, in which case temporary epicardial pacing as discussed above^{23,25} may be necessary. The practical problem of an intravenous access was a real one for those looking after these babies. As a result of this, we have had to stop the infusion.

Ambulatory holter monitoring done during this period demonstrated a basal heart rate of 40 to 45 beats per minute. This was deemed inadequate for a growing preterm baby

and the decision was made for an epicardial pacemaker to be implanted. This is one of our smallest babies to require implantation of an epicardial pacemaker.

It appears that prematurity and hydrops are 2 poor prognostic factors in these infants. Fortunately, although this baby was born premature, she was not hydropic at birth and this improved her chances. We managed to get away with temporary epicardial pacing. It should be emphasised however, that the strategy of management should be individualised according to the merits of circumstances as the outcome is variable.²⁶ Having said so, guidelines for management of such cases may be useful. Such a call for more collaborative and interdisciplinary work would therefore extend to all those looking after mothers and babies with this uncommon disorder.

Conclusion

Maternal SLE is a known cause of congenital CHB. This may be associated with morbidity and mortality. Predictive factors such as the presence of autoantibodies to the Ro and La ribonucleoproteins are important in detecting the fetus at risk. Advances in obstetric care and fetal monitoring have enabled physicians to detect problems early. The treatment of the mother, fetus and newborn with CHB continue to present challenges to the team involved in the care of both mother and child. While it would be useful to pool resources and collaborate to better optimise management strategies in this rare disorder, proper attention directed toward the medical needs of each individual baby with CHB cannot be overemphasised.

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