Staged pacemaker implantation in a preterm with hydrops fetalis due to complete heart block

Sir,

The outcome of the preterm fetus with hydrops fetalis due to congenital complete heart block (CHB) is frequently associated with significant morbidity and mortality. The management of CHB in premature, low-birth-weight infants is challenging. In addition to the technical difficulties in pacemaker implantation in these small babies, there are issues regarding long-term outcome, especially in those with hydrops fetalis. We report our experience of treating one such patient with temporary epicardial pacing followed by permanent epicardial pacing.

A 39-year-old lady, previously diagnosed to have systemic lupus erythematosus (SLE), presented to us with diamniotic dichorionic twins, for fetal echocardiography at 28 weeks of gestation. She had past history of recurrent spontaneous abortions and was detected to have high titers of anti-SSA/Ro antibodies. This pregnancy was the result of assisted reproductive techniques after 9 years of (nonconsanguineous) marriage. Fetal echocardiography demonstrated Twin A to have a heart rate of 30 beats/min, cardiomegaly with poor left ventricular (LV) function, bilateral pleural effusion, ascites, and skin edema suggestive of hydrops. Twin B had a heart rate of 52 beats/min and LV ejection fraction of 60%, with no other features of hydrops. Both twins had no structural cardiac anomaly. She had received dexamethasone for the fetal heart block, in addition to oral beta agonist therapy.

She was admitted for conservative care; however, she needed emergency cesarean section at 29 weeks of gestation in view of intrauterine fetal death of Twin A. Twin B was born with a weight of 1490 g and an APGAR score of 4 and 6 at 1 and 5 min, respectively. He was intubated in view of respiratory distress in the operating room and transferred to the neonatal intensive care unit. Physical examination revealed mild skin edema and pallor. There were no skin lesions suggestive of neonatal lupus erythematosus. His blood pressure was 70/43 mm Hg and heart rate was 50 beats/min. Chest X-ray showed mild cardiomegaly with clear lung fields and no pleural effusions [Figure 1].

A 12-lead electrocardiography confirmed complete atrioventricular (AV) block with ventricular rate of 52 beats/min (narrow complexes) [Figure 2]. Echocardiography demonstrated dilated left ventricle with mild LV systolic dysfunction (ejection fraction of 50%), and a small patent ductus arteriosus (PDA) with left to right shunt. There were no other associated heart defects. He received two doses of surfactant in prophylactic doses and was put on positive pressure ventilation. Intravenous immunoglobulin was given at 2 g/kg and dopamine infusion was started at 5 μg/kg/min. Despite adequate preterm care, his subsequent arterial blood gas showed rising lactates indicating poor peripheral perfusion. Tissue edema of hydrops is known to interfere with the threshold of permanent pacemaker;[1] hence, staged pacing was planned using temporary epicardial pacing.

On day 2 of life, two temporary pacing wires were placed on the epicardium over the right ventricle through a small midline incision over the xiphoid area. The external pacemaker was set at a rate of 120 beats/min with a threshold of 5 mA and an output of 5 mV. Features of hydrops fetalis started resolving gradually (body weight decreased to 1350 g, anasarca subsided, and serum lactates normalized). Follow-up echocardiography on day 8 of life demonstrated a closing PDA and normal LV systolic function. His pacing output slowly increased to 8 mV over the next few days. On day 10 of life, permanent pacemaker (St. Judes, VVIR) was implanted via a left 5th intercostal space anterior thoracotomy incision with a single epicardial lead placed on the LV apical wall and the rate was set at 120/min. Lead was tunneled subcostally and additional loop of the lead was taken to increase the longevity with somatic growth [Figure 3]. The external oblique muscle was incised and a pocket was made in between the layers of external and internal obliques, lateral to the rectus abdomini. There were no complications related to the procedure.

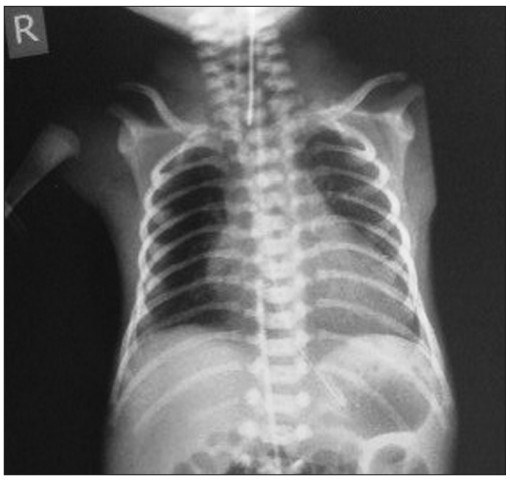

Figure 1: Chest X-ray showing mild cardiomegaly with clear lung fields and no pleural effusion

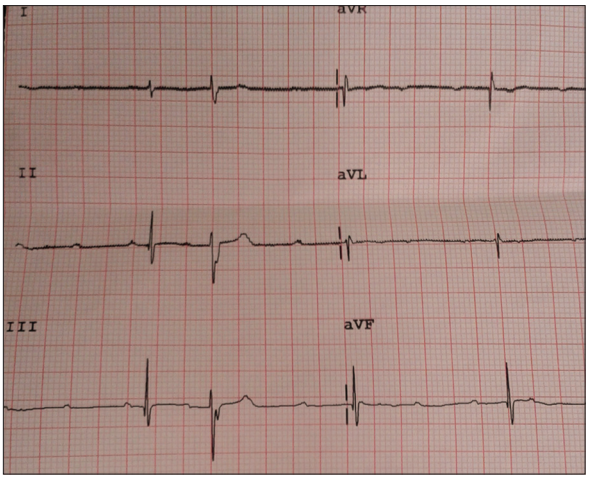

Figure 2: Limb lead ECG showing complete heart block with a heart rate of 50 beats/min

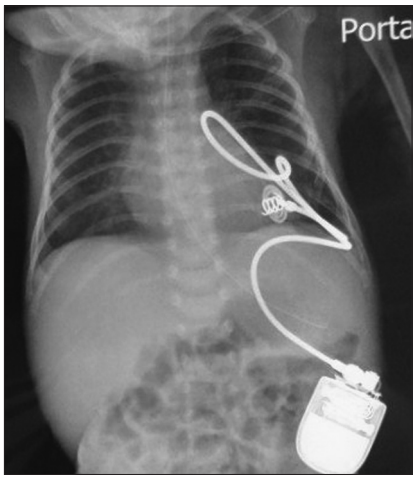

Figure 3: Chest radiograph showing position of the pacemaker lead and the generator in the abdomen

Baby remained hemodynamically stable thereafter and was extubated on day 13 of life. He received routine preterm care and was discharged home with a weight of 2 kg on day 45 of life.

Congenital CHB occurs in 1 in 14,000–20,000 live births. Transplacental passage of maternal anti-Ro and/or anti-La autoantibodies accounts for 90-95% of all cases.[2] The immune-mediated damage of the cardiac conduction system ultimately ends with its substitution with fibrotic tissue.[3] Prenatal diagnosis of CHB, when associated with hydrops fetalis and low ventricular heart rate (<55 beats/min), has very poor prognosis with fetal and neonatal mortality exceeding 80%.[4] Jaeggi *et al.*, studied 102 cases of congenital CHB and identified the risk factors associated with high mortality rates in this subgroup of neonates. Poor prognostic markers included early intrauterine diagnosis, hydrops fetalis, delivery <32 weeks, and postnatal ejection fraction of ≤40%.[5] In this report, a 29-week preterm with hydrops due to CHB was effectively managed with staged pacing. Deloof and colleagues[6] used a staged approach in two hydropic fetuses due to CHB, with good results. A similar approach was used by Khotiseth *et al.*,[1] in the management of a 33-week-old hydropic preterm. This approach helps in nullifying the interference to permanent pacemaker implantation produced by tissue edema in hydrops.

In summary, staged pacing was demonstrated as a novel approach in the management of congenital CHB. Short-term temporary pacing, in a neonate with hydrops, prior to permanent pacemaker implantation helps in improving the hemodynamics, as well as decreases the tissue edema, thus reducing the interference and complications of a permanent pacemaker implantation.

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REFERENCES

1. Khositseth A, Samankatiwat P, Withurawanit W, Khowsathit P. Pacing in preterm with hydrops fetalis due to congenital complete heart block. Asian Cardiovasc Thorac Ann 2006;14:428-31.
2. Silverman E, Mamula M, Hardin JA, Laxer R. Importance of immune response to the Ro/La particle in the devepment of congenital heart block and neonatal lupus erythematosus. J Rheumatol 1991;18:120-4.
3. Friedman DM, Rupel A, Buyon JP. Epidemiology, etiology, detection, and treatment of autoantibody-associated congenital heart block in neonatal lupus. Curr Rheumatol Rep 2007;9:101-8.
4. Groves AM, Allan LD, Rosenthal E. Outcome of isolated congenital complete heart block diagnosed in utero. Heart 1996;75:190-4.
5. Jaeggi ET, Hamilton RM, Silverman ED, Zamora SA, Hornberger LK. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. A single institution's experience of 30 years. J Am Coll Cardiol 2002;39:130-7.
6. Deloof E, Devlieger H, Van Hoestenberghe R, van den berghe K, Daenen W, Gewillig M. Management with a staged approach of the premature hydropic fetus due to complete congenital heart block. Eur J Pediatr 1997;156:521-3.

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