Introduction

Congenital isolated CAVB (1/14,000 - 20,000 live births) is present in systemic lupus erythematosus (SLE) or Sjögren syndrome. CAVB is rare, i.e. 1-2% in anti-Ro antibodies positive women; in presence of positivity for anti-SLE antibodies, echocardiograms should be performed every 2 weeks from the 16th week of gestation, to detect early foetal abnormalities (premature atrial contractions), that might precede complete atrioventricular block, and to start therapy with betamethasone (2), even if controversial (10).

CABV is often associated with asymptomatic maternal SLE, and require implantation of a pacemaker immediately after birth. SLE antibodies provoke damage to the A-V conduction tissue in the foetus, causing total heart block. When signs of cardiac heart failure (CHF) appear in the foetus, a decision–making approach to deliver the neonate in advance, and to implant a pacemaker, is started.

Since late 60’s, the field of pacing has experienced a significant improvement (3,4). Despite this, pacemaker implantation in small infants remains a challenge. The main problems in implantation of generators is firstly the generator size (and the wires length) compared to the body surface area of the neonate, and, secondly, the energy consumption of the generator; thankfully, dependable advances in lead technology, programmability and generator miniaturization have been achieved during the last years (Fig. 1).

We present a case of a premature newborn, with CAVB diagnosed at 29 weeks of gestation, in which implantation of a pacemaker at 1295 grams body weight has been without unexpected events.

**SUMMARY:** Pacemaker implantation in a premature low weight newborn with critical congenital atrioventricular block.

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We describe a case of a preterm newborn with congenital complete atrioventricular block (CAVB), delivered for distress by cesarean section, and undergone, at 1295 grams of weight, to permanent pacemaker implantation. The pacemaker has been inserted between the upper left rectus abdominal muscle and the posterior rectus sheath and connected by two epicardial leads on the left ventricle wall. The procedure has been successful.

**KEY WORDS:** Congenital block - Premature - Pacemaker.

**RIASSUNTO:** Impianto di pacemaker definitivo in un neonato prematuro di basso peso corporeo con blocco atrioventricolare completo congenito critico.

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Ripportiamo il caso di un neonato pretermine con blocco atrioventricolare totale nato prematuramente da parto cesareo per severo distress clinico e sottoposto a impianto di pacemaker permanentemente ad 1,295 kg di peso corporeo. Il generatore è stato inserito tra il muscolo retto addominale sinistro e la fascia del retto posteriore e poi connesso tramite due elettrodi epicardici sulla parete del ventricolo sinistro. La procedura è stata condotta con successo e senza eventi collaterali.
Case report

A premature newborn from mother with asymptomatic SLE was referred to our institution after diagnosis of foetal bradycardia at 29 weeks. As foetal distress syndrome developed at the 31st week, the baby was delivered by cesarean section with a heart rate of 50 bpm and A-V block, and supported with mandatory ventilation for 15 hours.

The echocardiography showed a decreased left ventricular function with mild dilatation. Dobutamin and furosemide administration was unsuccessful to improve cardiac function. The anti-SLE antibodies ratio mother/neonate was 1/1.

At the age of 7 days and 1295 grams body weight, we decided to implant a pacing system (1). A bipolar catheter with two steroid eluting Medtronic® Capsure leads “epi 1033 and 4968” (Medtronic Inc. Minneapolis, USA) was fixed with prolene, through a left 5th anterior thoracothomy (Fig. 2), on the antero-lateral wall of the left ventricle, following longitudinal pericardiotomy; the Y wire of 35 cm was placed into the left pleural cavity, and wires placed through the subcutaneous fascia toward the abdominal pocket, where the pulse generator St. Jude Medical® Microny SR II has been placed, between the left rectus abdominal muscle and the posterior sheath. The subxyphoid approach has not been considered because too narrow to settle the generator and the wires, and because the high risk to open the peritoneum (which could cause ileus, mediastinal or abdominal infections or hernia development) (3). So, in our personal opinion, the best approach seemed to be the combined abdominal plus thoracothomy, with tunnelization of wires between the two sites, avoiding in a very small patient, to collect in the same site lead wires and generator, which produces high risk for leads rupture and decubitus.

At 5 months follow up, the baby is in very good conditions, with a 5500 grams weight and no signs of infections, or dislocation of the device. The growth appears regular.

Discussion

Benefits derived from permanent pacing in congenital atrioventricular block are known, and placement techniques established (3-5, 6, 7); however, problems and disadvantages in low weight prematures remain. The procedure poses challenges in finding a suitable pocket for the generator, due to a mismatch between the weight of the patient and the generator size. Furthermore, the length of wires presents beyond measure. Various techniques could be considered safe. We have reported the case of a low weight premature newborn with heart failure and bradycardia since the 29th week of gestation in which we have observed the heart parameters first; then, SLE serology confirmed the etiology of CAVB, so we have decided for a pacing system with a bipolar leads catheter and a selfprogrammable autocaptured pulse generator (8-9).

We have approached to newborn through the 5th left intercoste space to insert epicardial electrodes, placing the generator between the left rectus abdominal muscle and the posterior sheath. The subxyphoid approach has not been considered because too narrow to settle the generator and the wires, and because the high risk to open the peritoneum (which could cause ileus, mediastinal or abdominal infections or hernia development) (3). So, in our personal opinion, the best approach seemed to be the combined abdominal plus thoracothomy, with tunnelization of wires between the two sites, avoiding in a very small patient, to collect in the same site lead wires and generator, which produces high risk for leads rupture and decubitus.

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Conclusion

In conclusion, our management of this patient has been focused on selecting one of the smallest pulse generator (12.8 grams x 5.9 cm x 6 mm thin). A double approach, abdominal plus thoracothomy, is safe and reproducible, and decreases risk of secondary infection of incisions, makes the risk lower of leads fractures and wires kinking, and is a procedure in compliance with the body surface area of the baby (1-7, 8, 9).

In addition, it’s easier to find areas of lower threshold on the anterolateral wall of left ventricle rather than on the diaphragmatic wall of the right ventricle. The Microny® St. Jude Medical SR II setter offers a total programmable feature and stable sensing. The procedure was safe and without collateral events. X-rays have showed a good placement of the pacemaker and leads (Fig. 3).

References