

TEACHING FILES (GRAND ROUNDS)

A 9 MONTHS OLD CHILD WITH ASYMPTOMATIC BRADYCARDIA

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Clinical Problem

A 9-month old boy, born of a non-consanguineous marriage was referred for bradycardia. The child was otherwise asymptomatic. On examination, his heart rate was 60 bpm, with no apex pulse deficit. Other systems were normal. Investigations showed complete heart block on electrocardiograph (ECG) and echocardiography was normal

How should this child be managed?

Complete heart block, also known as complete atrio-ventricular (AV) block, is a condition in which the electrical impulses generated by the sino-atrial node in the atria of the heart are not conducted to the ventricles. Due to this, the intrinsic electric rhythm of the ventricular muscle takes over the pacemaker role for the ventricles. As the intrinsic rhythm of the ventricles has a much lower frequency (about 40 beats/minute), compared to that of the atria (100 beats/minute)¹, this condition manifests clinically as bradycardia. The ECG shows a complete dissociation between the P-waves (representing the atrial rhythm) and the QRS complexes (representing the ventricular activity). This condition manifests in about 1 in every 20,000 live births.² Complete congenital AV block (CAVB) may be diagnosed during fetal life or after birth. It may be isolated or occur in association with congenital structural defects of the heart, such as atrio-ventricular discordance or AV canal defects. It may also be associated with heterotaxy.¹

A strong association has been recorded between the incidence of congenital heart block and the presence of maternal anti-Ro/SSA autoantibodies, which may or may not be associated with maternal autoimmune disease- usually systemic lupus erythematosus (SLE) or Sjogren's syndrome. About 1-2% of mothers with anti Ro/SSA antibodies give birth to children with CAVB, with a 12-20% recurrence rate in subsequent pregnancies.³ Other possible etiologies include myocarditis, and cardiomyopathy, including congenital conditions such as Hurler cardiomyopathy.¹ CAVB has a good prognosis. However, this depends on a

number of factors, such as intra-uterine detection at an early gestational age and the presence of associated structural defects. The estimated mortality of non-paced patients is 8%-16% in infants and 4%-8% in children and adults. Instances of patients surviving up to the fourth to six decades of life without pacemaker implantation have been recorded.⁴

A complete work-up of a child diagnosed with CAVB is required. Twenty-four hour (Holter or ambulatory) ECG monitoring is recommended to determine the nature of the block, i.e. if it is continuous or intermittent, as well as to check for any other electrocardiographical abnormalities.^{1,5} Chest radiography must also be performed to check for cardiomegaly and pleural effusions.¹ Exercise tolerance cannot be deduced from the resting heart rate, so exercise testing must be conducted, as ectopy with exercise is frequent in these patients.⁵ Routine electrolyte assays and complete blood counts must be performed in case of any metabolic derangements or blood cell deficiencies, especially thrombocytopenia. Assessments for evidence of organ damage must also be performed.¹ The mother should be evaluated for connective tissue disorders, presence anti-Ro antibodies, and other signs of autoimmunity if this has not been performed previously.^{1,5}

The approach to the therapy of CAVB can be classified into pharmacological and pacemaker therapy. Pharmacological therapy is more useful as a prophylactic measure as it does not reverse third degree heart block, but may be useful in preventing progression to complete heart block, if first- or second-degree heart block is detected in utero. These therapies include plasmapheresis, immunoglobulins, and corticosteroids.¹ Transplacental administration of dexamethasone is the current treatment modality for fetuses, however, recent studies show no significant reduction in the occurrence of hydrops fetalis.⁵

Most children with CAVB will inevitably require permanent pacemaker insertion at some point. This is indicated in all symptomatic patients with syncope, congestive heart failure (CHF), or chronotropic incompetence limiting physical activity.⁵ However, some patients may be asymptomatic or exhibit mild and general symptoms such as frequent nightmares, naps, tiredness or unsatisfactory growth. In such cases, pacemakers may be inserted if the cause for these symptoms can be strongly associated with bradycardia. Other parameters are based on ventricular dysfunction

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or dilatation, heart rate (<55 in neonates and <50 in children, adults and neonates), pauses during spontaneous rhythm (duration >3 seconds), QRS duration (>120 ms), complex ventricular ectopy, and prolonged QT interval. However, exact cut-off points within these parameters are difficult to establish, and often conflict.⁵

Complications such as CHF may follow even after the timely insertion of a pacemaker. Biopsies reveal interstitial fibrosis, myocyte degeneration or hypertrophy. Thus, close follow up of cardiac function, especially ventricular function, is necessary.⁶ The mean 5-year survival rate of patients with CAVB, post implantation of pacemaker is about 76-89%. Long-term complications include high-grade stenosis or occlusion of the great cardiac veins (25%) as well as infectious complications.⁵ Currently, alternative therapies are being explored in the form of robot-assisted pacemaker implantation, which reduces the risks of traditional implantation (thrombosis and infections), and is also minimally invasive, thus cosmetically sparing. As a number of complications of pacemakers are also thought to be caused by the presence of leads, leadless pacemakers are also being developed, in hopes of improving outcome.⁵

Compliance with ethical standards

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