Editorial

Congenital complete atrioventricular block. A pace in time saves lives (?)

The study by Balmer et al[1] in the present issue of Europace lends support to our clinical notion that early intervention is beneficial even to the young individual. The children in this study that received a pacemaker had signs and/or symptoms such as bradycardia, exercise intolerance, syncope or heart failure that were ameliorated by pacemaker implantation. Three patients received a pacemaker within the first month of life. The common experience that the prognosis is worse when congenital complete atrioventricular block (CCAVB) is associated with cardiac malformation was not confirmed in this study. Interestingly there was no difference in heart rate between symptomatic and asymptomatic children before pacemaker implantation. The clinical consequence is that the resting heart rate does not help in determining the right time for intervention. Their main conclusion is that pacemaker therapy reduces morbidity and mortality in children with CCAVB when compared with natural history data. This information is valuable but there is still room for argument about when to intervene.

In their study all symptomatic children received a pacemaker whereas asymptomatic ones did not. But is there an asymptomatic child with CCAVB? Is the breastfed baby with CCAVB symptomatic if feeding time is somewhat prolonged? Is the 7-year-old with CCAVB symptomatic if he does not want to play soccer? Is the 13-year-old computer-enthusiast with CCAVB symptomatic? My point is that our tools to detect symptoms at present are too blunt.

We really need to know when to treat the (?) asymptomatic child and what pacemaker mode to choose. There is evidence that when the patient has passed the mid-teens, pacemaker implantation reduces mortality and morbidity[2]. Although symptoms and signs such as premature ventricular complexes, wide QRS escape rhythm, fatiguability and effort dyspnoea are poor indicators of future severe symptoms[2], they are used as an indication for pacemaker implantation! Furthermore, signs such as prolongation of QTc and progress of mitral regurgitation are prognostically bad signs — most likely late manifestations of a slowly beating heart — and may only indicate that we are intervening on a downhill course! The finding that there was no difference in heart rate between symptomatic and asymptomatic children[1] is intriguing. What does this reflect, differences in the extent of myocardial damage by foetal exposure to maternal antibodies, differences in myocardial excitation pattern or autonomic influence?

Like many institutions, without scientific evidence, we implant epicardial electrodes and subclavicular pacemakers within the first month in all infants with CCAVB, symptomatic or not on the hypothesis that AV sequential pacing is advantageous and that epicardial leads carry a lower risk of complications in early life.

There are additional problems to resolve — the time of diagnosis and the issue of defining congenital heart block. It has been proposed that we should distinguish between congenital blocks, i.e. those that are diagnosed in utero or within the first month of life, and those detected later in life, because the aetiology of complete heart block in the two categories may very well be different[3].

The aetiology of CCAVB relates in most cases to maternal rheumatic disease[4] with the presence of autoantibodies that can traverse the placenta and accumulate in the foetal heart predominantly in the AV node. The foetus usually shows complete block in gestational weeks 20–24. Successful steroid treatment of pregnant women has shown that it may be possible to prevent progression to complete block[5]. Refined methods using foetal doppler indicate that it is possible even to diagnose first-degree block that may help us to find the right moment to intervene and prevent the development of CCAVB[6].

The rapid development of pacemaker leads and technology has given us the tools for early pacemaker-implantation but there is little consensus.

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how lead(s) should be applied — endocardial vs epicardial — and where the pacemaker should be implanted — abdominal or sub clavicular. Our hope is that these patients will have a life expectancy corresponding to that of the normal population with as few leads and pacemaker implantations as possible and that treatment should be initiated well ahead of any symptoms, whatever they may be!

Friedman wrote an editorial [7] — ‘Pace me now or pace me later’ — on this issue in 1995 and the questions he posed remain essentially unanswered. The study by Balmer et al. [1] does give, however, some support for intervention at an earlier age than in the study by Michaelsson et al. [2].

When facing the ECG of a child with dissociated atrial and ventricular rhythms it is relieving to know that we have the technological means to intervene and more or less establish ‘normal function’ of the heart. Pacemaker technology is progressing rapidly providing us with new pacemaker leads and pacemaker units while clinical research rarely has the time necessary to evaluate the quality of each technological improvement. This problem becomes evident for those relatively rare disorders such as CCAVB that occur early in life and require life-long treatment.

The basic question still remains unanswered — should all patients with CCAVB, irrespective of age, have a pacemaker implanted? Maybe it is time for a multicentre study to address these issues.

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References