Sinus node function in children with congenital complete atrioventricular block

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Aims

Children with congenital complete atrioventricular block (CCAVB) often need pacemaker therapy. In these children, it may be preferable to use single-lead VDD pacing, but for VDD pacing a normal sinus node function is required. Our aim was to study sinus node function in children with CCAVB.

Methods and results

We longitudinally evaluated sinus rate in 36 children with CCAVB and normal anatomy of the heart. The rate of sinus rhythm on a 12-lead ECG, in Holter recordings, and exercise tests were evaluated at regular intervals. Age at the first visit of the children was 2.5 ± 3.3 years (mean ± SD). Follow-up was 10.6 ± 7.3 years. The rate of sinus rhythm on a 12-lead ECG was at every age within the normal values for age (e.g. 0 – 1 year: 153 ± 24 bpm, and 17 – 18 years: 76 ± 4 bpm). Lowest and highest sinus rates in the Holter recordings were normal. During exercise, mean sinus rate in the total group of children increased from 92 ± 8 at rest to 171 ± 9 bpm at maximal exercise.

Conclusion

We conclude that sinus node function is normal in children with CCAVB. Because of the normal increase in sinus rate during exercise, a single-lead VDD pacemaker can be safely implanted in these children.

KEYWORDS

Arrhythmias; Complete heart block; Congenital; Pacemakers; Paediatrics; Sinus node

Introduction

Children with a congenital complete atrioventricular block (CCAVB) will need pacemaker implantation early in life. Some need a pacemaker already in infancy, but the majority in adolescence or early in adulthood. Especially, since Michaelsson et al.1 published the 30-year follow-up of patients with CCAVB with a high incidence of Adams–Stokes attacks, mortality and mitral insufficiency, early prophylactic pacemaker implantation in adolescence is recommended. This means many decades of pacing in these young patient group. Therefore, the (paediatric) cardiologist should consider implantation techniques that save the subclavian veins, as much as possible.

One of the risks of endocardial pacing is the development of thrombosis and obstruction of the subclavian veins. The incidence of abnormal venography, mentioned in the literature, varies from 23 to 64%. Total occlusion was found in 6–25%.2–7 Risk factors, from the literature, for venous occlusion are contradictory and unclear, although some authors found a relation between multiple leads and obstruction and between the cross-sectional area of leads indexed to body surface and obstruction.6,7

To save the subclavian veins, we are in favour of starting with epicardial pacing leads and when endocardial pacing is preferred or needed a single lead should be considered. That means a VVIR or VDD pacing mode. VDD pacing is preferable because of the AV synchrony and the normal heart rate response to physiological stress. Single-lead VDD pacing has been proved to be safe and reliable in adults as well as in children.8,9

A prerequisite for VDD pacing is a normal sinus node function. However, there is only sporadic information about sinus node function in patients with CCAVB. Menon et al.10 found a diminished rate response of the sinus node during exercise in 3 out of 28 patients with CCAVB. Pathologists showed small abnormalities of the sinus node in the hearts of some patients with CCAVB because of anti-Ro antibodies.11,12

The aim of our study was to answer the question if children with CCAVB have a normal sinus node function. Therefore, we longitudinally evaluated all children referred to our hospital with CCAVB and a normal heart since 1976 by yearly ECG, 24-h Holter recording and (if old enough) exercise test.

Methods

Patients

Thirty-six children with CCAVB and normal anatomy of the heart were admitted to our hospital since 1976. All parents provided informed consent for clinical evaluation. Complete AV block was defined as complete dissociation of the P wave and the QRS

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complex with a higher atrial than ventricular rate. Mean age of the 36 children (25 females and 11 males) at the time of diagnosis was $2.5 \pm 3.3$ years (mean $\pm$ SD) with a range from in utero at a gestational age of 20 weeks to 14.2 years. RoSSA antibodies were present in 9 of the 17 children tested for this. Seventeen children were treated with a pacemaker.

Figure 1 | Data (mean $\pm$ SD) from a 12-lead ECG at rest in children with CCAVB. (A) $P$ rate. The lines represent the 5th, 50th, and the 95th percentile curves of normal values for age.$^{13}$ (B) $QRS$ rate. (C) $P/QRS$ ratio.
Monitoring parameters

We longitudinally evaluated the children during follow-up by history, in particular of complaints like syncope or exercise intolerance, physical examination, ECG, 24-h Holter recording, and an exercise study. Only values obtained before pacemaker therapy were included.

ECG

In a 12-lead ECG (rest; supine position), P rate (=sinus rhythm) and QRS rate (=AV nodal escape rhythm) were evaluated and compared to normal values.\textsuperscript{13} P/QRS ratios were calculated by dividing P rate by QRS rate.

Holter recording

In the Holter, lowest, mean, and highest sinus rates and lowest, mean, and highest QRS rates were evaluated yearly and compared to normal values.\textsuperscript{14} Also P/QRS ratios were calculated.

Exercise test

After the age of 7 years, the lowest and highest sinus and QRS rates during an exercise test (bicycle) were measured two-yearly. Also P/QRS ratios were calculated.

Results

Patients

Age at the first visit of the children was 2.5 ± 3.3 years (mean ± SD). Follow-up was 10.6 ± 7.3 years.

12-lead ECG

The P rate on a 12-lead ECG at rest was at every age within the normal values for age according to Davignon (Figure 1A; e.g. 0–1 year: 153 ± 24 bpm, and 17–18 years: 76 ± 4 bpm). From the age of 4 years, the sinus rate is between the 5th and 50th percentile of normal. The QRS rate is

![Figure 2](https://academic.oup.com/europace/article-abstract/9/9/844/517139)

Figure 2 Data (mean ± SD) from Holter recordings in children with CCAVB. (A) Highest (solid squares) and lowest (solid circles) P rate. The lines represent normal values for age.\textsuperscript{14} (B) Highest and lowest QRS rate.
shown in Figure 1B. P/QRS ratio was greater than 2.0 at age <3 years; thereafter less than 2.0 (Figure 1C).

Holter
Lowest, mean, and highest P rates in a Holter recording were normal (Figure 2A). Lowest and highest QRS rates are shown in Figure 2B. P/QRS ratios of lowest and highest heart rates did not alter with ageing during childhood.

Exercise study
During exercise, mean rate in the total group of children increased from 92±8 at rest to 171±9 bpm at maximal exercise. Mean QRS rate increased from 51±4 at rest to 90±8 bpm at maximal exercise. P/QRS ratios are similar as found on the Holter recording. The maximal reached exercise capacity was 85.8±8.9% of the normal value for age and length.15

Discussion
Sinus node function
We found a, totally, normal sinus node function on rest ECG and 24-h Holter monitoring in this longitudinal study of 36 children with CCAVB with a follow-up of 10.6±7.3 years. The maximal heart rate during exercise was only slightly below normal, but only 85.8±8.9% of the normal value for age and length of the maximal exercise capacity was reached. We assume that the maximal exercise capacity is lower in CCAVB because of a limitation in increase in AV nodal escape rate. If we take into account the lower exercise capacity, the maximal sinus rate at 100% exercise capacity will be as high as 199 bpm, which is normal.14

Escape rhythm
This study gave us the opportunity to study the ‘natural history’ of the AV nodal escape rhythm. This gives information to determine if the escape rhythm of future patients is within normal limits at rest, during daily activities, or at maximal exercise. The decline in QRS rate over the ages follows the pattern of normal sinus rhythm. The P/QRS ratio is mostly ≤2 except for the first 3 years.

Pacemaker therapy
Life-long pacing obliges the (paediatric) cardiologist to choose the pacing methods very carefully to prevent major problems and risks for the future. Comparison of single-lead (VVI, VDD) pacing with dual chamber pacing showed a significantly higher operation time and complication rate in DDD pacing. There were no differences between VVI and VDD pacing.9 Furthermore, some authors found a relation between the vein obstruction and the number of pacing leads or the cross-sectional area of leads indexed to body surface.6,7 Single-lead VDD pacing is preferable to VVI(R) pacing because of the restoration of AV synchrony and the physiologic rate response. Complete dislodgement of the free floating atrial dipole in VDD pacing is rare in adults and the risk is lower than of atrial lead malfunction in DDD pacemakers.9 In children, a small amount of extra lead is left in the atrium to allow for somatic growth. This could increase the risk of atrial malsensing later, but in the majority AV synchrony remains intact. If there will be loss of atrial sensing in the long-term, than the VVIR mode of pacing can be programmed and the system can, if needed, always be updated to a DDD system by placement of an extra atrial lead. An other argument to limit the number of pacing leads is the fact that, although the experience in intravascular lead extraction has increased considerably over the years, it remains associated with the risk of major complications.16

Conclusion
We conclude that sinus node function is normal in children with CCAVB. Therefore, a single-lead VDD pacemaker can safely be implanted in these children and young adults. This is preferred over dual chamber pacing, in view of the long period of pacing ahead.

Conflict of interest: none declared

References