Congenital Complete Heart Block

a report by
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White and Eustis were the first to document congenital complete heart block (CCHB) with electrocardiogram in 1921. Until the 1950s, CCHB without structural heart disease was considered rare and benign. CCHB is estimated to complicate one in 15,000–22,000 live births; however, the true incidence of CCHB is known as there is an association with structural congenital heart disease (CHD) with a resultant high foetal mortality. Maternal autoimmune disease is often associated with isolated CCHB (not associated with underlying structural congenital heart defect). Antibodies cross the placenta and are thought to cross-react with the developing conducting system, leading to injury.2 Pregnancies complicated by mothers positive for anti-Ro/La antibodies result in CCHB in 1–5% of live infants, with a familial recurrence rate estimated at 15–20%. Some studies have estimated that as many as 20–30% of these infants will suffer foetal or neonatal deaths, and an estimated 10% of children with antibody-positive CCHB will be born with hydrops or congestive heart failure (CHF) secondary to intra-uterine myocarditis and/or severe brachycardia.1,4 While CCHB in the absence of immunological exposure is recognized, patients with antibody-mediated CCHB have been found to require pacing earlier in life and follow a more malignant disease course than antibody-negative patients.5 These antibody-positive infants experienced a higher risk of developing dilated cardiomyopathy (DCM) with clinical CHF with signs and symptoms. CCHB also occurs in the setting of complex structural heart disease (most frequently heterotaxy syndromes and/or congenitally corrected transposition of the great arteries). The combination of heterotaxy syndrome and CCHB has an extremely high mortality rate, even with pacemaker and surgical treatments.

Combining several CCHB studies,6–9 risk factors for CHF, Stokes Adams attacks or sudden death include:

- low newborn ventricular rate (<55bpm);2
- low foetal atrial rate (<120bpm);6
- wide QRS complex on electrocardiogram (EKG);10
- corrected QT interval prolongation >460ms;10
- atrial enlargement on EKG or cardiomegaly by chest X-ray (risk factor for CHF);1,11
- ventricular ectopy in combination with a wide QRS complex or structural heart disease;12 and
- presence of complex structural heart disease.6–9

Many of these risk factors were defined in an era predating foetal echocardiography and current surgical and pacemaker therapies; however, they remain pertinent today for guiding patient care decisions. The indications for pacemaker implantation continue to be debated, with practice patterns varying from institution to institution. No large prospective trials have addressed the ideal timing or indications for device implantation and recommendations have been forged by consensus conferences. The most recent guidelines for pacemaker implantation published in 2002 by the American College of Cardiology (ACC)/American Heart Association (AHA)/North American Society of Pacing and Electrophysiology (NASPE)/Heart Rhythm Society (HRS) since 1991 and a member of the Pediatric Electrophysiology Society (PES). Prior to joining the faculty at Emory University, he was an Associate Professor and Director of the Pediatric Electrophysiology laboratory at the University of Michigan.

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Margaret Strieper is Director of Pacing and Electrophysiology at Children’s Healthcare of Atlanta Sibley Heart Center and an Assistant Professor of Pediatrics at Emory University School of Medicine. She is a Fellow of the American Academy of Pediatrics (AAP) and the American College of Cardiology (ACC), and a member of the American Heart Association (AHA), the North American Society of Pacing and Electrophysiology (NASP/Heart Rhythm Society (HRS) and the Pediatric Electrophysiology Society (PES). Her research interest is in the field of pediatric electrophysiology, in particular cardiac resynchronisation, ablation and syncope. She has published extensively and presented at multiple national meetings, including the AHA, the ACC, the HRS and the AAP, within the field of pediatric electrophysiology.

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2. Waltuck J, Buyon J, 11. Sholler GF, Walsh EP, 1. White P, Eustis R, changes following chronic pacing for CCHB and subsequently reported Moak et al.19 reported a group of 16 patients with CCHB who developed developments resulting in smaller-diameter pacing leads,14,15 transvenous function, i.e., any patient with congenital heart block”. Despite recent changes following chronic pacing for CCHB and subsequently reported transvenous Moak et al.19 reported a group of 16 patients with CCHB who developed function, i.e., any patient with congenital heart block”. Despite recent changes following chronic pacing for CCHB and subsequently reported transvenous Moak et al.19 reported a group of 16 patients with CCHB who developed function, i.e., any patient with congenital heart block”. Despite recent changes following chronic pacing for CCHB and subsequently reported transvenous function, i.e., any patient with congenital heart block”. Despite recent changes following chronic pacing for CCHB and subsequently reported transvenous function, i.e., any patient with congenital heart block”. Despite recent changes following chronic pacing for CCHB and subsequently reported transvenous function, i.e., any patient with congenital heart block”.

While dual-chamber pacing restores AV synchrony and affords physiological ventricular rates and variability, RV pacing desynchronises the ventricles. Therefore, the possibility of alternate site and or biventricular pacing in CCHB patients who will require a lifetime of pacing has recently been raised. For adults with CHF and ventricular dysynchrony, cardiac resynchronisation pacing has been shown to improve ventricular function, decrease symptoms and improve exercise performance. The authors’ group has promoted the term ‘prosynchronisation’24 (dual-chamber biventricular pacing) as a means of maintaining a more co-ordinated activation pattern in patients with CCHB and possibly preventing the deleterious effects induced by RV pacing. Acute animal studies,24,25 using a radiofrequency ablation-induced AV block model of CCHB, have demonstrated improved myocardial performance with biventricular compared with single-site ventricular pacing in mature and immature myocardium. Myocardial performance was assessed using pressure volume loops and tissue Doppler imaging techniques. These animal studies are consistent with a 2006 report demonstrating that biventricular resynchronisation pacing can benefit paediatric CCHB patients who develop ventricular dilatation and dysfunction, with clinical CHF signs and symptoms, following RV pacing.26

Moak et al.19 reported a group of 16 patients with CCHB who developed ventricular dysfunction (mean left ventricular shortening fraction 9±5%) following implantation of dual-chamber, single-site ventricular pacing systems. Karpawich20,21 reported histological degenerative myocardial changes following chronic pacing for CCHB and subsequently reported decreased systolic and diastolic ventricular performance in 24 patients who had undergone single-site ventricular pacing for a median of 10 years. Newer imaging techniques have been used to assess for mechanical dyssynchrony. Cummings et al.22 used tissue Doppler imaging to examine the time-course of ventricular dysfunction after the induction of RV pacing in 12 chronically paced patients (mean age 8.5 years; average length of pacing 4.2 years) with CCHB and structurally normal hearts. This study suggested that ventricular remodelling occurs early and may not initially manifest as decreased ejection fraction. This would support the notion that chronically RV-paced paediatric patients need close follow-up due to the possibility of developing a clinically significant cardiomyopathy.

Mechanical ventricular dyssynchrony is not synonymous with a wide QRS EKG complex. Non-invasive diagnostic studies in paediatric patients have shown that dyssynchrony may be present in patients with isolated DCM despite the presence of a narrow QRS complex on a surface EKG. Alderson and associates23 demonstrated mechanical dyssynchrony in 11 paediatric patients with DCM and a narrow QRS complex.

Other studies have suggested that pacing from alternate sites in the ventricular myocardium may be more haemodynamically beneficial than RV apical pacing. Pacing from the high RV septum has been shown to result in acute haemodynamics similar to those seen during sinus rhythm. Parahissian27 pacing, with the possibility of stimulating the His-Purkinje system high in the ventricle, should also result in maintenance of ventricular synchrony. While pacing lead placement has proved difficult to date, recent experience with this approach seems promising.25 Prinzen and colleagues24 have proposed left ventricular apical pacing as an optimal strategy. Data supporting the long-term benefits of resynchronisation pacing in paediatrics are limited. Strieper et al.20 reported a decrease in hospitalisations due to CHF exacerbation after an upgrade from a conventional pacemaker to a resynchronisation device. In this study, seven of nine patients were removed from transplant consideration due to improved symptoms and cardiac function after the pacing system upgrade. The decision to implant a prosynchronisation or resynchronisation pacing system, with additional leads, potential access issues and increased costs, must be carefully considered. Studies to define the role of resynchronisation pacing or alternate site pacing will be required before these options can be considered as a clinical standard of care for CCHB patients.
ESC Congress 2008

Abstract deadline
14 February 2008

30 August
3 September