Pacing Therapy in Infants and Children with Congenital and Acquired Complete Atrioventricular Block: Optimal Pacing Strategies, Management, and Follow-up.

Floris E.A. Udink ten Cate and Narayanswami Sreeram
Department of Paediatric Cardiology, Heart Center Cologne,
University Hospital of Cologne, Cologne,
Germany

1. Introduction

Bradycardia due to high-degree atrioventricular (AV) block remains the main indication for permanent cardiac pacing in childhood. Although there are major differences in the etiology of AV block in children and adults, the same pacing systems and leads are used in both age groups. The first pacemaker was implanted in a child in the late 1960s (Martin et al., 1966). Nowadays, implantation of a permanent pacing system is a straightforward and safe procedure with excellent pacemaker performance during follow-up, even in infants and small children (Welisch et al., 2010).

Applying permanent pacing therapy in the young can be challenging. Many paediatric issues, such as patient size, body growth, coexistence of congenital heart disease, presence of residual intracardiac shunts and life style, have to be considered. Selecting the best pacing system for a child requires a firm understanding of modern pacing design, pacing indications, advantages and drawbacks of epicardial and endocardial lead pacing, and possible complications. The aim of this chapter is to give a dedicated overview of the practical implications of pacing in children with complete AV block.

2. Etiology of atrioventricular block in childhood

Before we describe the techniques and management of pacing in children, an overview of the most important causes of complete AV block in children is given. Having a complete understanding of the etiology and prognosis of complete AV block in childhood should improve best pacing practice in the paediatric age group. A summary of the causes of complete AV block in childhood is presented in Table 1.

2.1 Postoperative complete atrioventricular block

Complete AV block is an important complication of surgical correction of congenital heart disease (Gross et al., 2006). Due to advances in surgical techniques and improved anatomic knowledge of the course of the conduction system in various congenital heart diseases, the

incidence of postoperative complete AV block has decreased significantly during the last decades (Gross et al., 2006). Although complete AV block was found in more than 10% of children in the 1960s, a study from the early 1970s reported that this incidence had fallen to around 2% (Fryda et al., 1970). In the current era, 1 – 3% of all children undergoing surgery for congenital heart disease develop complete AV block. Importantly, it has been shown that surgical complete AV block is associated with significant morbidity and decreased survival. (Gross et al., 2006; Driscoll et al., 1979). Therefore, all children should receive a pacemaker device when complete AV block does not resolve in the early postoperative course (Gross et al, 2006).

Surgical acquired AV block	VSD, AVSD, TOF LVOT obstruction, ccTGA discordant AV connections
Isolated congenital complete AV block	maternal anti-SSA/Ro – SSB/La antibodies, NLE abnormal development of AV conduction
AV block associated with structural cardiac abnormalities	ccTGA heterotaxy, left atrial isomerism single ventricle physiology, Fontan palliation D-TGA after Mustard or Senning operation
Infectious disease	bacterial viral EBV, varicella, Coxsackie B Rheumatic fever Chagas' disease Lyme disease HIV
Neuromuscular disease	Myotonic dystrophy Emery-Dreifuss muscular dystrophy Duchenne muscular dystrophy
Metabolic disease	Carnitine deficieny Kearns-Sayre syndrome
Syndromes	Holt-Oram syndrome 18p-syndrome
Others	long-QT syndrome post-Ablation hypertrophic cardiomyopathy percutaneous VSD closure myocarditis connective tissue disease sarcoidosis amyloidosis cardiac tumors

AVSD = atrioventricular septal defect, AV = atrioventricular, ccTGA = congenitally corrected transposition of the great arteries, D-TGA = dextro-transposition of the great arteries, EBV = Ebstein-Barr virus, HIV = human immunodeficiency virus, LVOT = left ventricular outflow tract, NLE = neonatal lupus erythematosus, TOF = tetralogy of Fallot, VSD = ventricular septal defect.

Table 1. Causes of complete atrioventricular block in neonates, infants and children.

The greatest risk for complete AV block is associated with corrective surgical procedures for ventricular septal defects (VSD), usually as part of more complex congenital heart disease, atrioventricular septal defects (AVSD), left ventricular outflow tract obstruction, L transposition of the great arteries, tetralogy of Fallot (TOF), and discordant atrioventricular connections (Batra et al., 2003; Gross et al., 2006). Most children who require pacing for surgically induced AV block are less than 1 year old.

Of importance, several studies report spontaneous resolution of complete AV block in the early postoperative period, most often occurring between 7 and 14 days. Complete AV block may resolve spontaneously in 43 – 92% of children (Gross et al, 2006). Factors associated with a spontaneous recovery of AV nodal function are currently not known. Hence, late recovery of complete AV block has also been reported in approximately 10% of cases (Batra et al., 2003). Recovery of AV conduction was identified within the first 30-day postoperative interval in most children. Recurrence of high-degree AV block or complete AV block is not observed in these children, although different studies have shown inconsistent findings (Batra et al., 2003; Gross et al., 2006). On the contrary, late onset complete AV block (> 30 days postoperatively) has been described after cardiac surgery for congenital heart disease (Goldman et al., 1985.; Liberman et al., 2008). Complete AV block was identified at a mean of 4.7 years after surgery in one study (Goldman et al., 1985), occurring as late as 16 years after cardiac surgery (Liberman et al., 2008). Close monitoring of AV conduction seems mandatory in all patients after surgery for congenital heart disease.

2.2 Congenital complete AV block

Congenital complete AV block (CCAVB) is a rare cardiac conduction disorder with an estimated incidence of 1 in 11.000 to 22.000 live births (Michaelson & Engle, 1972). In 25% of cases, coexisting congenital heart disease can be identified. Congenital heart diseases in which AV conduction is particularly at risk include heterotaxy syndrome with left atrial isomerism, atrioventricular septal defect with common atrioventricular junction, and congenitally corrected transposition of the great arteries (ccTGA) (Anderson et al., 1974; Stephenson & Kaltman, 2006). Complete AV block in these patients may be present at birth, or may develop later in life. When complete AV block is not present at birth, close monitoring of the cardiac conduction system is mandatory in these patients (Stephenson & Kaltman, 2006 & Graham et al, 2000). Notwithstanding, most patients with CCAVB have structurally normal hearts, and CCAVB in these cases is therefore referred to as 'isolated' CCAVB. Although abnormal embryological development of the cardiac conducting system have been identified as a cause of isolated CCAVB (Anderson et al., 1977; Lev et al., 1971), most patients have autoimmune, anti-SSA/Ro – SSB/La-antibody-induced CCAVB (Buyon & Winchester, 1990; McCue et al., 1977).

Since an association between maternal connective tissue disease and anti-SSA/Ro – SSB/La antibodies was suggested in the late 1970s (McCue et al., 1977), efforts in clinical and experimental studies have resulted in a broad understanding of the pathogenesis and clinical outcome of autoimmune-associated CCAVB (Boutjdir et al., 1998; Buyon & Clancy, 2005; Jaeggi et al., 2010). Maternal anti-SSA/Ro – SSB/La autoantibodies enter the fetal circulation between 16 and 24 weeks gestation (Buyon et al., 1995), and may induce injury to the developing cardiac conducting system and myocardial tissue in a subset of fetuses. Autoimmune CCAVB develops in 1 – 2% of these antibody-positive pregnancies. Of importance, the estimated recurrence risk of women who had a previous child with CCAVB is 16 – 25% (Buyon et al., 2009). Although 30 – 50% of these women have clinically manifest

connective tissue diseases, such as systemic lupus erythematosus or Sjögren's syndrome, the majority of antibody-positive women who gave birth to an infant with CCAVB, are asymptomatic (Julkunen & Eronen, 2001).

Anti-SSA/Ro – SSB/La antibody-induced CCAVB is associated with substantial morbidity and mortality during gestation and infancy, with more than 60% of infants requiring permanent pacemaker therapy in their first year of life (Buyon et al., 1998; Breur et al., 2002). The most common indication for pacemaker placement in the neonatal period is congestive heart failure (Buyon et al., 1998). The natural history of isolated CCAVB was addressed in a prospective multicenter study, involving 102 children and adults (Michaelsson et al., 1995; Michaelsson et al., 1997). All children were asymptomatic during their 15 years of life. Of concern, 10 patients without a pacemaker died, of which 6 CCAVB related deaths occurred without preceding symptoms. The mortality rate was significantly lower in paced compared to non-paced patients. Therefore, the authors concluded that the prognosis of isolated CCAVB may be improved when patients are paced earlier in life (Michaelsson et al., 1997). A recent study supported the concept of earlier 'prophylactic' pacing in children with isolated CCAVB (Balmer et al., 2002). At present, more than 94% of children with CCAVB are paced before they reach the age of 15 years (Villain et al., 2006).

Another important prognostic issue in children with autoimmune-associated CCAVB is the development of dilated cardiomyopathy (Moak et al., 2001; Udink ten Cate et al., 2001). We and others have demonstrated that as many as 6 – 11 % of paced children with autoimmune-associated CCAVB develop dilated cardiomyopathy during a follow-up period of 10 ± 7 years (Moak et al., 2001; Udink ten Cate et al., 2001; Kim et al., 2007). Risk factors may include presence of anti-SSA/Ro – SSB/La antibodies, increased heart size at initial evaluation and the absence of pacemaker-associated normalization of left ventricular size during follow-up (Udink ten Cate et al., 2001). Although pacemaker-induced ventricular dysfunction has been offered as an etiologic factor in the pathogenesis of dilated cardiomyopathy in these patients (Janousek et al., 2004), some children with anti-SSA/Ro – SSB/La antibody-induced CCAVB develop dilated cardiomyopathy before being paced (Nield et al., 2002; Villain et al., 2006). These data suggest that children with anti-SSA/Ro – SSB/La antibody-induced CCAVB may have preexisting myocardial damage, which increases their risk of pacemakerinduced dilated cardiomyopathy. Close monitoring of their ventricular function during follow-up is mandatory.

The prognosis of non-autoimmune CCAVB seems to be different compared to autoimmune-associated CCAVB (Cruz et al., 2004; Villain et al., 2006). Patients are commonly diagnosed later in life, the AV block is often progressive, and these children have a good prognosis after pacemaker implantation (Villain et al, 2006).

Moreover, connective tissue disease is not found in these mothers, and the recurrence risk in future pregnancies is low. Pacemaker-induced heart failure does not seem to occur during medium follow-up (Cruz et al., 2004). For these reasons, a new definition of congenital complete AV block was recently proposed: 'an AV block is defined as congenital if it is diagnosed in utero, at birth or within the neonatal period' (Brucato et al., 2003).

3. Pacing indications and patient selection

Specific recommendations for pacemaker implantation have been published in several guidelines by the American College of Cardiology, the American Heart Association, and the Heart Rhythm Society, with an update in 2008 (Epstein et al., 2008). The European Society of

Cardiology, and the European Heart Rhythm Association have also recently published a guideline for permanent pacing and cardiac resynchronization therapy (Vardas et al., 2007). Pacing recommendations for children are also a part of these published guidelines, and have been very useful in daily practice. It should be remembered however, that the level of evidence for these recommendations is low. Most recommendations for children requiring permanent pacing are not based on prospective clinical studies, but rather rely on expert opinion.

The main indications for pacing in children are (1) symptomatic sinus node dysfunction, (2) bradycardia-tachycardia syndrome, (3) congenital, surgical or acquired complete AV block, and (4) advanced second-degree AV block. Because this chapter is dedicated to AV block in children, pacing recommendations for this specific conduction disorder are summarized in Table 2.
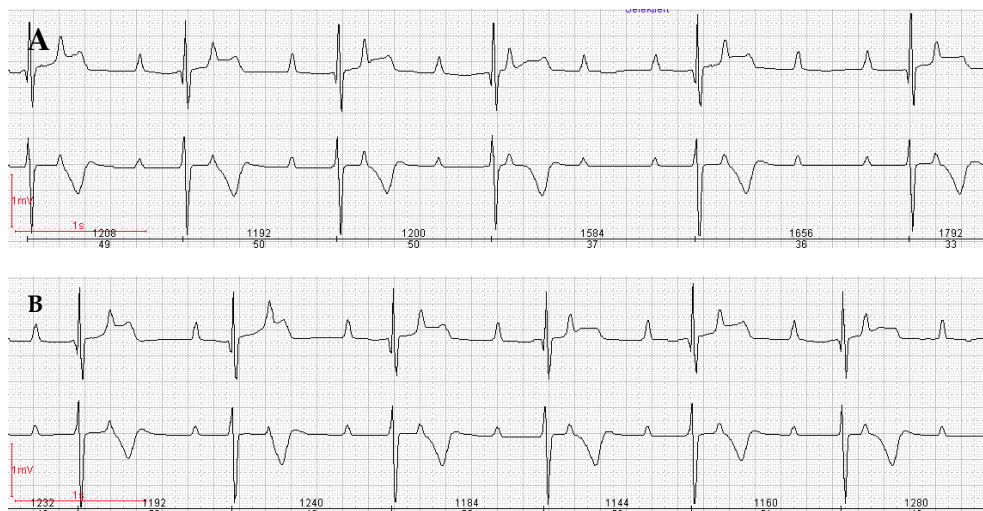

Fig. 1. Rhythm strip showing complete atrioventricular block (**A**) and second-degree atrioventricular block in a 2-year-old boy with bradycardia-related symptoms (**B**). The child underwent permanent pacemaker implantation (single chamber) using the transvenous approach. A lumenless 4.1-F endocardial lead inserted.

Surgical second-degree or complete AV block persisting beyond 7 days postoperatively, not expecting to resolve spontaneously, is considered to be a Class I indication for permanent pacemaker implantation (Vardas et al., 2007; Epstein et al., 2008). There are two important reasons for not delaying pacemaker implantation in these patients, although there is reasonable evidence that AV conduction may recover in the first 30 postoperative days (Batra et al., 2003). The prognosis of surgically induced complete AV block is poor (Gross et al, 2006, Simon et al., 1982). The second consideration is that there have been enormous improvements in pacing systems and battery longevity. A pacemaker can now be safely implanted in most small infants.

Advanced second-degree or complete CAVB with symptomatic bradycardia (Figure 1), ventricular dysfunction, or low cardiac output, is also considered to be a Class I indication for pacemaker insertion. One has to keep in mind that symptoms of bradycardia can be subtle in children, (Balmer & Bauersfeld, 2003). Therefore, many investigators have tried to

predict the need for future pacemaker therapy in isolated CCAVB, using diagnostic tests, such as serial ECG recordings and ambulatory Holter monitoring (Esscher & Michaellson, 1983; Dewey et al., 1987; Breur et al, 2006). Interpretation of these parameters remains difficult.

Class I, permanent pacemaker implantation is indicated for:

1. Advanced 2nd or 3rd AV block with symptomatic bradycardia, ventricular dysfunction, or low cardiac out put.

2. Postoperative advanced 2nd or 3rd degree AV block that is not expected to resolve or persists > 7 days after cardiac surgery.

3. Congenital complete AV block with a wide QRS escape rhythm, complex ventricular ectopy, or ventricular dysfunction.

4. Congenital complete AV block in the infant with a ventricular rate less than 55 betas per minute (bps) or with congenital heart disease and a ventricular rate les s than 70 bps.

Class IIa, permanent pacemaker implantation is reasonable for:

1. Congenital complete AV block beyond the first year of life with an average heart rate less than 50 bp m, abrupt pauses in ventricular arte that are 2 or 3 times the basic cycle length, or associated with sym ptoms due to chronotropic incompetence.

2. Congenital heart disease and impaired hemodynamics due to sinus bradycardia or loss of AV synchrony.

3. Unexplained syncope in the patient with prior congenital heart surgery complicated by transient complete heart block with residual fascicular block after careful eveluation to exclude other causes of syncope.

4. Long QT syndrome with 2:1 or complete AV block.

Class IIb, permanent pacemaker implantation may be considered for:

1. Transient postoperative third-degree AV block that reverts to sinus rhythm with residual bifascicular block.

2. Congenital complete AV block in asymptomatic children or adolescents with an acceptable rate, narrow QRS complex, and normal ventricular function.

3. Neuromuscular diseases with any degree of AV block (including first-degree AV block) due to risk of unpredictable progression of AV conduction disease.

Class III, permanent pacemaker implantation is not indicated for:

1. Transient postoperative third-degree AV block with return of normal AV conduction within 7 days.

2. Asymptomatic postoperative bifascicular block with and without first-degree AV block.

3. Asymptomatic type I (Wenckebach) second-degree AV block

* Adapted from Epstein AE, DiMarco JP, Ellenbogen KA, et al. ACC/AHA/HRS 2008 guidelines for device-based therapy of cardic rhythm abnormalities: executive summary. Heart Rhythm 2008;5:e1-62; and Vardas PE, Auricchio A, Blanc JJ, et al. Guidelines for cardiac pacing and cardiac resynchronization therapy: the task force for cardiac pacing and cardiac resynchronization therapy of the European Society of Cardiology. Developed in collaboration with the European Heart Rhythm Association. Eur Heart J 2007;28:2256-2295.

Table 2. Indications for permanent pacemaker implantation in children and adolescents with high-degree or complete AV block with or without congenital heart disease*.

Several studies have shown that monitoring of heart size and cardiomegaly in children with isolated CCAVB is of use in daily practice in deciding if a child requires a pacemaker (Beaufort-Krol et al., 2007; Breur et al., 2002; Sholler & Walsh, 1989). We favour implantation of a pacemaker in a child with isolated CCAVB in whom the heart size progresses during follow-up, even when the heart rate criteria for pacemaker implantation are not met.

4. Selecting a pacing system: single or dual chamber device?

The aim of permanent pacing therapy in patients with complete AV block is to restore heart rhythm and rate, relieve the patient from bradycardia-related symptoms, provide haemodynamic stability, and thereby improve patient well-being and clinical outcome. These objectives can only be met when the best of 'two pacing worlds' are combined: a single or dual chamber device with an epicardial or transvenous pacing lead. Selecting a pulse generator and pacing lead for a child who needs life-long pacing for complete AV block depends on several important factors. It should be remembered that the risks of pacemaker placement are related to the size of the patient, making patient size an important consideration in selecting an appropriate pacing system. Issues regarding the selection of a single or dual chamber are addressed in this part of the chapter. Considerations related to epicardial or endocardial lead system selection are subsequently discussed.

4.1 Modern pulse generators: size and location of pacemaker pocket
Many of the shortcomings of pacing in children have been overcome in the last two decades. Pulse generators have become smaller and battery longevity has increased. Also the programmability of many pulse generators has seen major advances.

Due to these advances, pacemaker therapy can be applied safely to small infants and even preterm infants weighing less than 2000 g (Ohmi et al., 1992; Kammeraad et al 2004,; Inoue et al., 2005). Although most of these infants will receive a single chamber device (VVI mode) with an epicardial pacing lead because of their small size, single lead endocardial pacing systems also have been successfully implanted with an abdominally located pacemaker pocket (Hoorntje et al., 2000).

The abdominal pocket is the preferred pulse generator location in neonates, infants and small children receiving an epicardial pacing system. Subpectoral and prepectoral subcutaneous pockets are used in children and adolescents for placement of their transvenous pacing system. In general, the subpectoral pocket is preferred in young active children in whom a pulse generator implanted in a prepectoral subcutaneous pocket is more prone to trauma. Moreover, the risk of deep pocket infections seem to be less when the pulse generator in inserted under the pectoralis major muscle (Gillette et al., 1991; Cohen et al., 2002).

4.2 Programming a pacing device: heart rate limits and rate-responsive pacing
Most modern pulse generators have a large flexibility in programming. Several pacemaker options are available which may help achieving better pacemaker performance, longevity, and optimize patient needs in daily life.

Upper and lower heart rate settings are important in children. Physiologically, the average heart rates in children differ from the adult heart rate requirements. An upper heart rate limit of 180 beats per minute is required in most small children. The selected pulse generator should be capable of providing high upper rate limits. Another important topic is the

programmed minimal heart rate. Particularly in neonates and small infants, programming the heart rates in the normal physiological range may induce ventricular dysfunction (Chen et al., 2008). After programming a pacemaker in a specific heart rate range, these settings should be re-evaluated at every pacemaker follow-up visit. Pacemaker programming must be tailored to the individual paediatric patient.

Another important function is the rate-responsive pacing mode. The advantage of rate-responsive pacing lies in the presumed augmentation of cardiac output with exercise. Ideally, rate-responsive or rate-adaptive sensors in pacemakers sense changes in activity level and adapt heart rate to the new level of activity. An adequate chronotropic response to exercise has been demonstrated in adult patients with a VVIR pacing mode (Oldroyd et al., 1991). Another study in adults showed that DDDR pacing resulted in a slight improvement in cardiopulmonary performance compared to VVIR pacing, probably due to a better atrial contribution to cardiac output on exercise in DDDR paced patients (Vogt et al, 1988). Although, a normal increase in heart rate during exercise is more important than AV synchrony, the addition of AV synchrony does provide an additional benefit, supporting the use of DDDR pacing mode in adults (Benditt et al., 1987).

As with many topics in paediatric cardiology, results from large adult studies are often extrapolated to paediatric patients. However, limited data regarding rate-responsive pacing exist in children. One study showed that minute ventilation sensors, used for sensing the chronotropic response, closely matched intrinsic sinus node function during exercise in healthy children, supporting the appropriateness of rate-responsive pacing in children requiring permanent pacing therapy (Cabrera et al., 2002). Ventricular rate-responsive pacing is feasible, effective and safe in children with complete atrioventricular block (Ragonese, et al., 1994). Therefore, most pacemakers in children with complete AV block are programmed to a rate-responsive mode.

4.3 Battery longevity and autocapture-controlled pacing

In general, battery longevity is good with modern pacemakers. A pacemaker battery may last for 5 – 10 years in most paediatric patients (Udink ten Cate et al., 2002; Welisch et al., 2010). Battery survival is influenced by many factors, of which pacing mode, programmed heart rate, pacing lead performance, and percentage of cumulative stimulation are the most important (Maginot et al., 2000.; Batra & Balaji, 2006). The higher heart rate requirements of infants and children compared with adults result in less battery durability. Moreover, epicardial pacing systems, which are frequently implanted in infants and small children, have higher acute and chronic pacing thresholds, further limiting battery longevity (Khairy et al., 2006).

An important tool in reducing energy drain and extending battery life in infants and children is the use of automatic algorithms for pacing threshold measurement, such as AutoCapture and Ventricular Capture Management (Bauersfeld et al, 1999; Cohen et al., 2004; Silvetti et al, 2007a). These algorithms measure atrial and/or ventricular pacing thresholds throughout the day. The system responds to increases and decreases of pacing thresholds and subsequently regulates battery output. These pacing algorithms can be used in both epicardial and transvenous lead systems. A recent study demonstrated that autocapture pacing in children with epicardial lead systems extended the calculated battery life up to 15% (Tomaske et al, 2007). Automatic pacing threshold determination might be an important technological tool to prolong battery longevity and increase pacemaker safety. Patients with high or fluctuating pacing thresholds benefit the most. However, not all pacemakers are equipped with this function.

4.4. Single and dual chamber pacemakers

The positive haemodynamic effects of AV synchronous pacing have been well described in adults. A properly timed atrial contraction, resulting in AV synchrony, augments ventricular filling and cardiac output through the Frank-Starling relationship, improves venous return, and assists AV closure (Buckingham et al., 1992). The value of AV synchrony was already recognized in the early 1970s in patients with AV block following cardiac surgery and acute myocardial infarcts (Chamberlain et al., 1970; Hartzler et al., 1977).

While evidence exists that DDD pacing might be superior to VVI pacing in adults, no similar data are available in children, in whom a single chamber device offers the advantage of lower cost, ease of implantation and extended battery longevity. With AV asynchronous pacing, the atria may contract against closed AV valves, causing atrial distension with subsequent increases in pulmonary wedge pressure and jugular venous pressure (Horenstein et al., 2004). This might provoke signs and symptoms of fullness, dyspnea, headache, fatigue, syncope and exercise intolerance, the so-called pacemaker syndrome (Furman, 1994).

Pacemaker syndrome however, is exceedingly uncommon in young children. The development of symptoms attributable to the pacemaker syndrome in children with single chamber devices is a time related event (Horenstein et al., 2003; Horenstein et al., 2004). Although more than 50% of children with single chamber devices may develop pacemaker syndrome, those children who became symptomatic had been paced for a median of 11 years. These data were confirmed in another study (Horenstein et al., 2003). Based on these findings, single chamber pacing device is preferably implanted as the initial pacing mode in young children, particularly in those with a structurally normal heart, isolated complete AV block and normal ventricular function. Early establishment of AV synchrony in this patient population offers no added benefit.

Some data exists on VDD pacing in children, a pacing mode which combines the advantages of AV synchrony and the smaller size of single chamber pacing systems (Rosenthal et al., 1997a; Seiden et al., 1997). In this pacing mode, a single implanted transvenous lead is used for AV synchronous pacing, with the extra proximally located electrode being used for atrial sensing. The acute results of this pacing mode are similar to others used in the conventionally used pacing modes (Rosenthal et al., 1997a; Seiden et al., 1997). There are two major practical difficulties in applying VDD pacing mode in children: (1) with growth, atrial sensing is lost in many patients due to loss of contact between the atrial sensing electrode and the myocardium, and (2) only large diameter adult leads are available. Therefore, most centers have abandoned this pacing technique in children.

4.5 Specific considerations in selecting single or dual chamber device
4.5.1 Ventricular dysfunction

An important consideration in selecting a pacing system is the presence of ventricular dysfunction. The ventricular dysfunction may worsen when a child receives a single chamber device. Therefore, a dual chamber system should be selected in patients with ventricular dysfunction with or without congenital heart disease.

4.5.2 Single ventricle morphology and Fontan palliation

Specific considerations are required for permanent pacing systems in patients after the Fontan procedure. Currently, the Fontan operation is the surgical approach of choice for

children with single ventricle physiology (Barber et al., 2005). Sinus node dysfunction and complete AV block are common indications for permanent pacing in patients after Fontan palliation. Studies have estimated that approximately 10% of these patients need pacemaker insertion during follow-up (Barber et al., 2005; Cohen et al., 2001a). Due to the limitations in venous access to the heart after the Fontan procedure, most patients receive an epicardial pacing system (Cohen et al., 2001a). Although various pacing modes have been described in these patients (Warfield et al., 1999), a recent haemodynamic study performed in the early postoperative period showed clearly that AV synchrony is vital in patients with Fontan physiology (Barber et al., 2005). Whether novel pacing techniques, such as multisite pacing to improve ventricular synchrony, have advantages over dual chamber pacing in these patients warrants further study.

5. Selecting a pacing system: epicardial or endocardial pacing leads?

Both transvenous and epicardial pacemaker leads are being used in paediatric patients with good early and long-term results. Because published guidelines on pacing indications in children and adults do not address selection of the type of pacing lead, choosing a transvenous or epicardial pacing lead depends on several considerations. An overview is presented on lead selection, the relative advantages of transvenous and epicardial pacing systems, complications and lead longevity.

5.1 Anatomic considerations

A unique aspect of pacing practice in children is that many patients have associated congenital heart disease. Coexisting anatomic vascular abnormalities in both surgically corrected and uncorrected congenital heart disease can influence the choice of the pacing lead system. Venous abnormalities associated with limited or difficult access to the heart include persistent left superior vena cava, various systemic venous abnormalities, single ventricle physiology after a modified Fontan procedure and presence of superior vena cava vein stenosis or obstruction in patients after Mustard or Senning operations for transposition of the great arteries. An epicardial lead system may be preferable in some of these patients.

However, several options for alternative vascular access have been proposed in various types of corrected congenital heart disease (Gillette et al., 1986; Rosenthal et al., 1995; Adwani et al., 1997; Emmel et al., 2007). Pacing following the Fontan procedure for single ventricular physiology is usually accomplished using an epicardial lead system. Some authors have implanted endocardial pacing leads in the coronary sinus or using the transhepatic pathway in patients with a Fontan circulation (Rosenthal et al., 1995; Adwani et al., 1997). However, these techniques can be used only in a limited subset of patients with single ventricular physiology.

Another example of transvenous lead implantation in a difficult venous anatomic setting is in patients with D-transposition of the great arteries palliated with a Mustard or Senning operation. Although progressive loss of sinus rhythm and sinus node dysfunction has been well documented in a significant proportion of patients following the Mustard or Senning operation, AV conduction abnormalities, higher-degree AV block and surgical complete AV block have also been noted at follow-up (Hayes & Gersony, 1986; Vetter et al., 1987). Superior vena cava obstruction appears postoperatively in 5 – 22% of patients with a Mustard or Senning type repair (Turley et al., 1988; Khairy et al., 2004). Because an obstruction in the venous pathway used for transvenous lead implantation can be present without signs and

symptoms, detailed venography, computerized tomographic scans or magnetic resonance imaging investigations are warranted in patients selected for an transvenous pacing system. When a venous stenosis or obstruction is encountered, implantation of a vascular stent may relief the stenosis, and allows implantation of a transvenous pacing system (Emmel et al., 2007). The intermediate term results of the 'stent-and-pace' procedure are good.

5.2 Intracardiac right-to-left shunting: a contraindication for transvenous pacing?

Thrombus formation and paradoxical embolization may adversely affect patients with transvenous pacing systems. Although risk factors and data on the prevalence of thrombus formation on endocardial leads in children are lacking, several studies support the concept of paradoxical embolization (Silka & Rice, 1991; Johnson & Galindez, 1998; Barakat et al., 2000). A recent study identified the risk of systemic thromboemboli associated with transvenous pacing systems in adult patients with intracardiac shunts (Khairy et al., 2006). This study showed that patients with transvenous pacing systems and intracardiac shunts have a greater than two-fold increased risk of systemic thromboemboli. Of concern was that there was no apparent protective value of chronic aspirin or warfarin therapy in these patients. Independent risk factors for paradoxical systemic embolic events were older age, atrial fibrillation or flutter, and ongoing phlebotomy. A limitation of the study was that standardised methods for assessing right-to-left shunting were not uniformly adopted in all participating centers.

The authors of this study suggested that efforts to eliminate right-to-left shunting should be pursued before transvenous pacing is applied, and if this may not be feasible, the epicardial pacing approach should be considered (Khairy et al., 2006).

5.3 Endocardial or epicardial pacing system?

The introduction of modern steroid-eluting bipolar leads has resulted in improved lead performance and survival of both epicardial and endocardial leads (Goldman-Cutler et al., 1997; Johns et al, 1992; Udink ten Cate et al, 2002). The addition of steroid elution limits the inflammatory response at the electrode-tissue interface, resulting in better acute and chronic stimulation thresholds and improved battery longevity (Mond & Stokes, 1992). Both pacing lead systems offer effective pacing options in infants and children. Moreover, implantation of a transvenous or epicardial pacing system are safe and feasible procedures in most children. Although, the epicardial approach remains essential in children with congenital heart disease in whom venous access is not available, a transvenous lead system is most commonly advocated in children of adequate size (Fortescue et al., 2004; Kammeraad et al., 2004).

An endocardial pacing system is preferably implanted in children weighing more than 15 kg (Fortescue et al., 2004; Alexander 2004), although implantation at a smaller patient size has been reported (Kammeraad et al., 2004; Sachweh et al., 2000). Several patient and lead related features should be weighed before an optimal decision regarding an epicardial or endocardial lead system can be made.

Advantages of the epicardial approach include the avoidance of vascular injury and risk of venous occlusion, absence of lead problems associated with somatic growth, and no risk of endocarditis (Cohen et al., 2001b; Fortescue et al., 2004). Moreover, a dual chamber device can be implanted more easily in small children with or without congenital heart disease using the epicardial approach. Transvenous pacing systems offer the advantages of avoidance of a surgical procedure (thoracotomy or sternotomy), excellent lead survival, and lower acute and chronic pacing thresholds, and thereby increased battery longevity (Kammeraad et al., 2004; Udink ten Cate et al., 2002).

Major concerns regarding epicardial and endocardial lead systems include lead survival in epicardial leads, and risk of venous thrombosis and long-term vascular integrity after endocardial lead placement (Cohen et al., 2001; Kammeraad et al., 2004). Although modern steroid-eluting epicardial leads offer acceptable acute and chronic performance, they are prone to exit-block or non-capture due to high thresholds at the lead tip (Alexander, 2004). Several possible reasons for epicardial lead failure have been suggested, such as recurrent minor traumas imposed on the leads due to the active lifestyle of children (Figure 2), and presence of scarred epicardium after surgery for congenital heart disease (Silvetti et al., 2007b). Current lead survival is 90% at 2 years after implantation, and 74% at 5 years for epicardial leads. A recent study showed a 5-year lead survival of 90% for endocardial leads (Fortescue et al., 2004). However, when a reanalysis was made of all leads placed in children who were less than 1 year of age at implant, the estimated 2-year and 5-year endocardial lead survivals were 90% and 78%, respectively.

Endocardial lead systems carry a significant risk of venous thrombosis in infants and small children. The concern of venous patency in children who need life-long pacing has been an argument for a more conservative approach of selecting a lead system, preferring the epicardial approach (Bracke et al, 2003; Bar-Cohen et al., 2006).

Complete venous obstruction has been documented in 11 – 21% of children at medium-term follow-up (Bar-Cohen et al., 2006; Figa et al. 1997, Kammeraad et al., 2004). Partial venous obstruction may be seen in another 12% of children receiving an endocardial pacing system
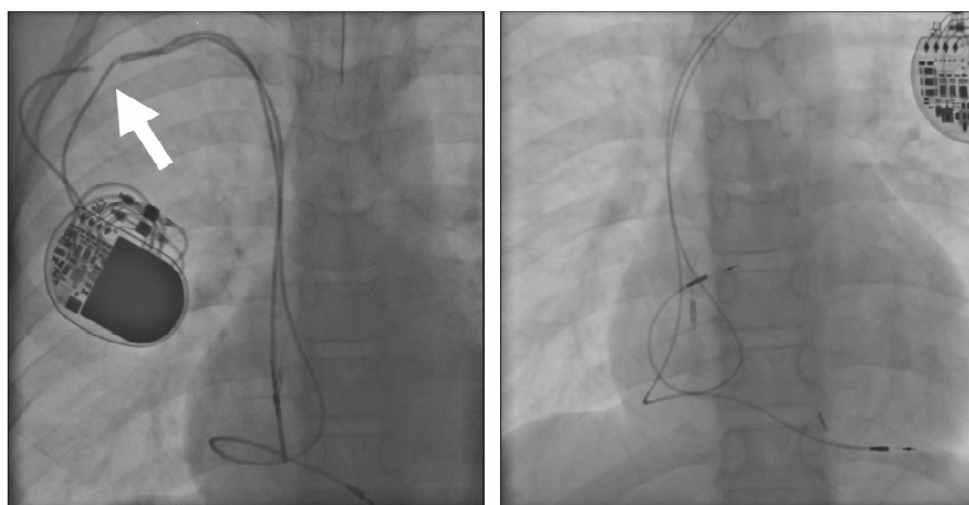

Fig. 2. In the example above, a 12 year old active boy presented with acute failure of the atrial lead 5 years following upgrade from single chamber to dual chamber pacing. The original ventricular lead had been partially abandoned in the heart. The atrial lead clearly shows a fracture, probably related to lead compression stress between the clavicle and the first rib. Four weeks later, the ventricular electrode had also failed, and he presented with a junctional escape rhythm at a rate of 35 to 40 beats per minute. The leads were entirely removed using a combined subclavian and femoral venous approach, and a new DDD system was implanted via the left subclavian vein, using 2 thin lumenless leads.

at young age. The location of thrombosis varied throughout the venous anatomy, with most venous obstruction occurring in the left innominate vein (Bar-Cohen et al., 2006). However, not all studies found venous obstruction in children after endocardial pacing (Gillette et al., 1988).

Venous thrombosis is most often asymptomatic (Bar-Cohen et al., 2006; Kammeraad et al., 2004). Nevertheless, it may cause superior vena cava syndrome in some patients. An important corollary, particularly in young patients requiring future lifelong pacing, is that every effort must be made to remove and replace nonfunctional leads in the vascular system. This often means the use of a variety of extraction systems, and occasionally transcatheter recanalization of stenosed or occluded vessels.

Risk factors for venous thrombosis are not yet known. A recent study showed that patient age, body size, and lead characteristics at implant did not clearly predict venous occlusion (Bar-Cohen et al., 2006). However, an earlier study concluded that if the lead size indexed to body surface area was more than 6.6 mm2/m2, venous thrombosis could be predicted with a sensitivity of 90% and specificity of 84% (Figa et al, 1997). The recent development of a lumenless 4.1 F endocardial lead is a promising new lead design, which may decrease the risk for venous occlusion in children (Chakrabarti et al., 2009).

In summary, many arguments exist for and against the use of epicardial or endocardial pacing systems in different subgroups of paediatric patients. In general, most neonates and small infants weighing < 10 kg receive initially an epicardial pacing system. At subsequent generator replacement, and depending on the child's size, an endocardial pacing system may be implanted.

5.4 Patient size and somatic growth issues in children

Transvenous pacing implantation in children requires special attention to their amazing somatic growth potential. It has been estimated that approximately 190 mm of additional right ventricular pacing lead in infants and 100 mm in 10-year-old children is needed to enable growth to adulthood (Gheissari, et al., 1991). Moreover, an 80-mm right atrial lead loop will allow 6 to 12 years of growth in infants and older children (Sanjeev & Karpawich, 2006).

To enable safe future growth in children with reliable transvenous single or dual chamber pacing, an additional amount of lead can be advanced into the heart during pacemaker implantation. Several options for creating such a redundant lead loop have been described (Rosenthal et al., 1997b; Gasparini, et al., 2000). A redundant lead loop may be placed in the right atrium or in the inferior vena cava. An example of a redundant lead loop in the inferior vena cava is shown in Figure 3.

If necessary, additional lead can be advanced during an elective pulse generator replacement. When transvenous pacing is applied in neonates or small infants, the length of the redundant lead loop is insufficient for most of these patients to grow into adulthood. Elective lead advancement is often needed. Lead advancement during the first 2 years after pacemaker implantation was necessary in 4/36 (11%) infants weighing < 10 kg who received transvenous leads (Kammeraad et al., 2004). In one of these, lead advancement failed and the lead was replaced.

5.5 Active or passive lead fixation

A stable position of the lead tip is pivotal to provide excellent acute and chronic thresholds and lead performance. The development of passive and active fixation mechanisms has
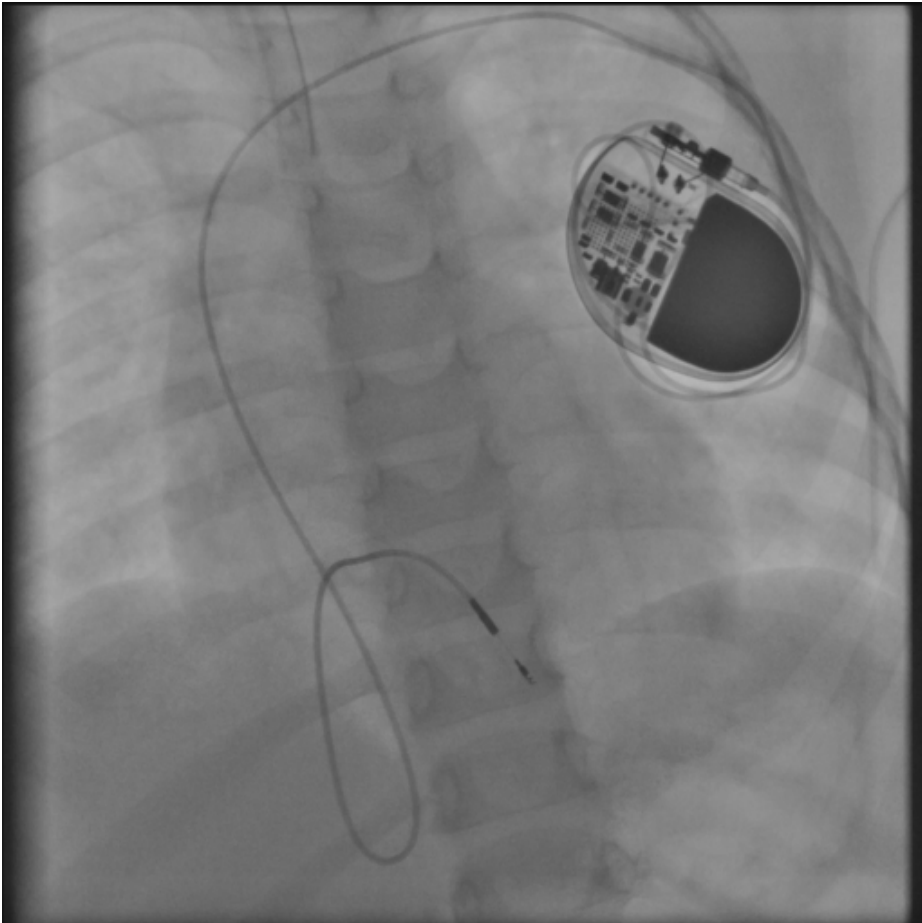
Fig. 3. Posteroanterior chest X ray of a child with congenital complete atrioventricular block who received a single chamber transvenous pacing system after developing bradycardia-related symptoms. The child's weight was 12 kg. A redundant lead loop for future somatic growth was placed in the inferior vena cava.

resulted in a tremendous reduction of the need for lead repositioning during follow-up (Furman et al., 1979). Passive fixation leads can be used for lead placement in the right ventricular apex. These leads can be easily positioned within the trabeculae of the right ventricle. The chronic pacing thresholds of passive fixation leads tend to be slightly better than those of active fixation leads (Hidden-Lucet et al., 2000). A disadvantage of passive fixation leads is that future lead extraction and replacement are more difficult, and this is an important issue in children (Hidden-Lucet et al., 2000).

Active fixation leads, usually equipped with a screw-in helix mechanism can be fixed more easily at almost any position in the atrium and ventricle, making this lead type ideal for selective site pacing (Chakrabarti et al., 2009). Active fixation may also prevent lead displacement when a redundant atrial loop is used, which is again of particular relevance to

young children (Rosenthal & Bostock, 1997). In specific anatomic situations, such as transvenous pacing after the Mustard or Senning procedure, where the leads have to be securely positioned at unusual anatomic sites, active fixation is preferable.
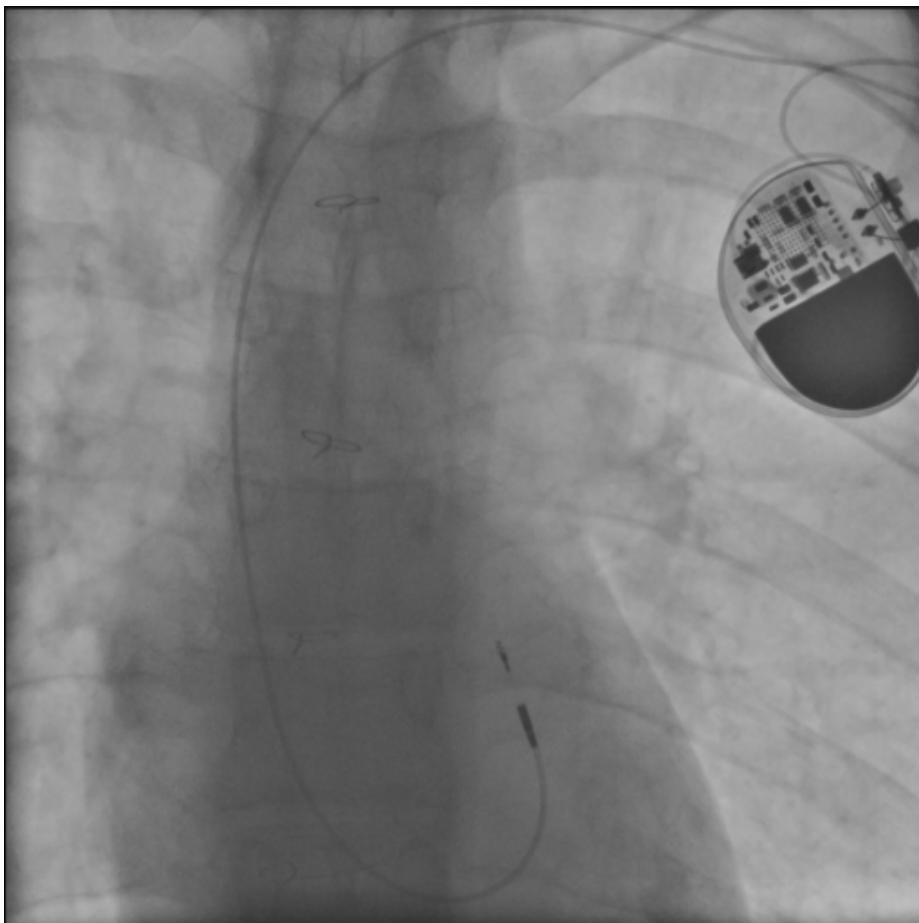

Fig. 4. The chest x ray shows an active fixation lead implanted in the roof of the native left atrium in a young patient who had undergone a Mustard operation for palliation of transposition of the great arteries.

5.6 Unipolar and bipolar leads
Epicardial and endocardial leads are available in unipolar or bipolar electrical configurations (Breivik et al., 1982; Breivik, et al., 1983; Mond, 1991). In general, the external diameter of a unipolar lead is usually smaller, because each coil is separated using insulating material. Although unipolar epicardial leads are still being used in paediatric pacing practice, most modern endocardial leads have a bipolar electrical configuration (Udink ten Cate et al., 2002).

The advantages of the unipolar lead configuration are lower pacing impedance, smaller size, and better lead survival and probably pacemaker longevity. With recent advances in lead design, the differences in pacing longevity between unipolar and bipolar lead systems have become marginal (Breivik et al., 1983; Mond, 1991). In addition, the lead diameter of bipolar leads has significantly decreased. Pectoral muscle stimulation, which may occur in unipolar pacing systems, is not often encountered in bipolar pacing systems. Bipolar leads are also less likely to detect far-field signals and electromagnetic interference (Mond, 1991).

6. Pacing and lead complications in children: implications for follow-up

6.1 Lead fractures, dislodgements, non-capture and insulation breaks

Possible mechanisms of epicardial and endocardial lead failure include lead fracture, dislodgement, high thresholds and insulation break. A recent retrospective study of 1007 implanted epicardial and endocardial leads in children with and without congenital heart disease requiring permanent pacing therapy found that lead fracture and exit-block were common complications seen with epicardial leads, whereas insulation breaks and lead dislodgements occurred more frequently in transvenous pacing systems (Fortescue et al., 2004). In this study, 15% of the leads failed during follow-up, affecting 23% of the patients. Of these patients, 28% experienced multiple lead failures. The most common lead failure types were lead fracture and insulation break. The rates of lead failure are in accordance with those noticed in several other studies, reporting lead failure rates ranging between 2 – 28% of implanted leads (Sachweh et al., 2000; Cohen et al., 2001b; Udink ten Cate et al., 2002; Bakhtiary et al., 2007; Welisch et al., 2010). Lead longevity of epicardial pacing systems were lower in most studies (Sachweh et al., 2000; Udink ten Cate et al., 2002; Fortescue et al., 2004; Welisch et al., 2010).

Risk factors for lead failures in children may include younger age at implant and presence of congenital heart disease (Fortesue et al., 2004). Epicardial pacing leads are associated with a higher risk of lead failure. An explanation for these findings is that children undergo rapid linear growth and are more physically active than adults, thereby placing additional stress on the pacing lead system and increasing the risk for lead failure.

Interestingly, it has been observed that only a minority of children (8%) report symptoms of pacing lead failure (Fortescue et al., 2004). Symptoms included skeletal muscle stimulation, palpitations, dizziness, or syncope. The remainder of patients were found to have pacemaker lead failure during regular pacemaker interrogation, routine chest radiography, or intraoperative lead examination. Routine and regular pacemaker follows up and interrogation are of upmost importance for optimal patient care.

6.2 Pacemaker pocket infections and endocarditis

Although pacemaker infections are uncommon in the paediatric population, deep pocket infection and pacemaker endocarditis are serious and life-threatening complications. When pacemaker-related infections occur, removal of the infected pulse generator and pacing leads are often required for infection control (Klug et al., 1997). Infections may be divided into superficial cellulitis, pocket infection, and endocarditis (defined as positive blood cultures). Pacemaker-related infections may develop in children before hospital discharge, or during early follow-up. Early pacemaker infections probably result from wound contamination during implantation (Cohen et al., 2002). In one paediatric retrospective study, the incidence of pacemaker-related infection was 7.8% for the whole cohort. The incidences of superficial

cellulitis, deep pocket infection and pacing lead infection were 4.9%, 2.3%, and 0.5%, respectively (Cohen et al., 2002). The authors concluded that the incidence of deep pocket infections was similar to those seen in adult studies (Kiviniemi et al., 1999; Klug et al., 1997), while more superficial infections occurred in their paediatric cohort. Moreover, Down syndrome was identified as an independent risk factor for pacemaker related infections. Patients with Down syndrome were 4 times as likely to develop an infection. The more severe pacemaker-related infections are most often seen following revision of a pulse generator with or without pacemaker lead exchange (Cohen et al., 2002; Fortescue et al., 2004).

Most superficial infections can be managed with (intravenous) antibiotic therapy alone. Appropriate management of deep pocket infections and pacemaker lead infection, or endocarditis, involves complete removal of the entire pacemaker system and long-term intravenous antibiotic therapy. With aggressive and appropriate therapy, mortality rates in children with pacemaker infection are minor. A subpectoral pocket may be preferable for transvenous pacing systems in children, because infection seems to be less likely (Gillette et al., 1991; Cohen et al, 2002).

6.3 Postpericardiotomy syndrome after pacemaker implantation

Postpericardiotomy syndrome (PSS) is a well known complication after cardiac surgery. It has been observed in 10 – 50% of patients following open heart procedures (Miller et al., 1988). PPS may also occur after pacemaker implantation (Polin et al., 2006). PPS occurs in 2 – 6% of children following initial pacemaker implantation of both epicardial and transvenous lead systems (Udink ten Cate et al., 2002; Zeltser et al., 2004). Acute PPS may also develop after a subsequent lead revision in a small subset of children (Zeltser et al., 2004). Patients typically present with respiratory distress, chest pain, fever, and lethargy. Cardiovascular compromise was often seen. Most PPS episodes occurred within 14 days after pacemaker placement (Zeltser et al, 2004). In some instances, PPS may have a late onset (Spindler et al., 2001).

PPS can be successfully managed with medical therapy that includes both anti-inflammatory agents (NSAIDs) and diuretics. Pericardiocentesis is an option when cardiovascular compromise is present. The clinical outcome of PPS in children after permanent pacemaker implantation is excellent.

6.4 Pacing induced ventricular remodelling and dysfunction

The right ventricular (RV) apex has been the conventional site for endocardial ventricular pacing in children and adults (Karpawich, 2004). Recent studies have demonstrated that pacing from the RV apex may induce left ventricular dyssynchrony (Thambo et al., 2004; Tops et al., 2007; Cheng et al., 2009). It has long been recognized that ventricular dyssynchrony caused by a variety of cardiovascular diseases, including heart failure, ventricular preexcitation, and left bundle branch block mediate ventricular remodelling and subsequently myocardial dysfunction (Prinzen et al., 1995; Kass, 2008; Udink ten Cate et al., 2010a; Udink ten Cate et al., 2010b). Evidence is now emerging that pacing-induced ventricular dyssynchrony disturbs myocardial regional workload and wall stress, which may result in wall motion abnormalities, myocardial perfusion defects, changes in coronary blood flow, increased left ventricular cavity volume, and asymmetrical changes in left ventricular wall thickness (Karpawich, 2004; Kass, 2008; Cheng et al., 2009). Moreover, interstitial and cellular histopathological alterations have been demonstrated in the hearts of animals and patients after long-term right ventricular apical pacing (Karpawich et al., 1999; Cheng et al., 2009).

The concept of pacemaker-induced heart failure is particularly important when permanent pacing is applied to young children, who often need life-long pacing. Dilated cardiomyopathy may develop in 6 – 11% of paced children with autoimmune CCAVB during follow-up (Moak et al., 2001; Udink ten Cate et al., 2001; Kim et al., 2007). In addition, heart failure associated with prolonged pacemaker therapy was found in 46% of adolescents with repaired congenital heart disease (Nothroff et al., 2006). Although other risk factors may play an additional role in the pathogenesis of cardiomyopathy in these patients, pacing-induced LV dyssynchrony is an important but under-recognised contributor.

6.4.1 Strategies to minimize ventricular pacing

Identification of risk factors for pacing-induced cardiomyopathy, unravelling its pathogenesis, and developing pacing strategies to avoid the adverse effects of right ventricular apical and lateral wall pacing are crucial. The detrimental effects of pacing on right ventricular function are in part related to the cumulative percentage of ventricular pacing (Sweeney et al., 2005). An infant in whom the right ventricle is paced for 100% of the time is at a higher risk for developing ventricular dysfunction compared to an infant who is only paced for 50% of the time. This has prompted the search for novel pacing strategies that reduce unnecessary ventricular pacing, and thereby reducing the cumulative percentage of ventricular pacing (Kaltman et al., 2008; Sweeney et al., 2004).

Managed ventricular pacing (MVP) and AAISafeR modes are new pacing algorithms designed for reducing cumulative percentage of ventricular pacing, with established benefits in the adult population (Mansour et al., 2006; van Mechelen & Schoonderwoerd, 2006). This pacing strategy requires the implantation of a dual chamber device, programmed in DDD or DDDR mode. Patients who have sinus node disease, second-degree AV block, or high-degree AV block with intermittent complete AV block are thought to benefit most of this pacing algorithm (Kaltman et al., 2008).

MVP is an atrial based pacing mode (AAI or AAIR). It primarily paces the atrium and monitors the ventricle for loss of AV conduction. When no ventricular beats are sensed between two consecutive atrial sensed or paced beats, MVP automatically mode switches to DDD or DDDR backup pacing (Kaltman et al., 2008; Mansour et al., 2006). Adverse effects have been described in the adult population, including tachyarrhythmias, ventricular fibrillation and failure of mode switch to ventricular backup pacing (Mansour et al., 2006; van Mechelen & Schoonderwoerd, 2006). The experience with MVP in children is limited. There is one retrospective study evaluating the safety and effectiveness of MVP in reducing unnecessary ventricular pacing in a large group of children with various diseases, including congenital and acquired AV block (Kaltman et al., 2008). Congenital heart disease was present in 64% of the children in the study group. MVP was effective in reducing the cumulative percentage of ventricular pacing, and only 1 MVP-related adverse effect was reported. Frequent nonconducted atrial depolarizations were noted in a patient with intermittent AV block. No proarrhythmic effect of MVP was noted in this study. Although there are no large randomized prospective trials in paediatric patients, MVP seems to be a promising pacing strategy for a subgroup of children of AV block.

6.4.2 Should right ventricular apical pacing be avoided in children at all cost?

The important question which has arisen following recent reports of pacing-induced cardiomyopathy is: should permanent pacing from the RV apex (for transvenous systems)

or RV anterior free wall (for epicardial systems) be abandoned in children with complete AV block? This is not an easy question to answer. First, a large proportion of children are at risk of developing ventricular dysfunction due to pacing, not all of them develop heart failure. Several studies have been undertaken to assess the incidence of ventricular dysfunction in paced children with CCAVB, and to identify potential risk factors (Udink ten Cate et al., 2001; Moak et al., 2006; Beaufort-Krol et al., 2007; Kim et al., 2007; Vatasescu et al., 2007; Gebauer et al., 2009). Most studies found an incidence of dilated cardiomyopathy and LV dilation in 6.0 to 13.4% of children after long-term pacing. Risk factors for development of dilated cardiomyopathy and LV dilation with ventricular dysfunction may include presence of anti-SSA/Ro – SSB/La antibodies, increased heart size at initial evaluation and the absence of pacemaker-associated normalization of left ventricular size during follow-up, prolonged QRS duration, and transvenous pacing location (RV apex and RV free wall worse compared to RV septum). The interpretation of these results is troublesome, because most studies involved small groups of patients. Interestingly, recent studies have demonstrated physiologically benefits of alternative site pacing, including RV septal, outflow tract, or Hisbundle pacing (Karpawich, 2004). The degree of pacing-induced LV dyssynchrony is lower with these pacing strategies. However, large clinical paediatric studies are needed to determine if selective RV pacing sites are superior to pacing the RV apex, which is still most frequently used in clinical practice.

6.4.3 Echocardiographic follow-up of children with permanent pacemaker therapy

Serial echocardiographic investigations are important in the management of children with permanent pacemaker therapy. Although shorthening fraction, left heart size and ejection fraction can be easily measured, these conventional echocardiographic parameters are not sensitive enough to identify paced children at risk for development of dilated cardiomyopathy at an early stage in the disease process. However, this should be the ultimate goal for selection of children who may benefit from upgrading their pacing system to DDD pacing, biventricular pacing or selective site pacing. Tissue Doppler echocardiography and speckle tracking imaging may be used for measuring LV dyssynchrony, regional and global ventricular function (Figure 5). When LV dyssynchrony, regional and global myocardial function deteriorates, we might be able to identify patients who are at the highest risk for development of dilated cardiomyopathy. Although more research work needs to be done, these new techniques may guide crucial improvements in the management of children with permanent pacing devices.

7. Conclusions

Pacing in children with congenital or acquired AV block is a safe and feasible therapy. Selecting an appropriate pacemaker system for a child with complete AV block remains challenging. Several patient-related and pacemaker-related issues should be considered, including patient size, cardiac anatomy, pacing indication, pulse generator capabilities and pacing lead characteristics.

A single chamber pacemaker is the preferably initial pacing mode in young children with complete AV block and normal ventricular function. Early establishment of AV synchrony in the young patient with a structurally normal heart and normal ventricular function is not thought to be beneficial. Conversely, single chamber devices should not be implanted in children following the Fontan operation or with ventricular dysfunction.

Fig. 5. This image shows a parasternal short-axis view at the mid-ventricular level of a child with permanent right ventricular pacing for congenital complete AV block. The ventricular walls are divided into six myocardial segments (farb-coded). During one cardiac cycle, the time to systolic peak radial strain can be measured. The time delay between the septal and posterior wall (yellow and purple arrows, respectively), a parameter of LV dyssynchrony is estimated. There is significant LV dyssynchrony in this patient (184 ms, normally the time delay is < 130 ms). In addition, there is an inhomogeneous pattern of radial strain, with severely reduced peak radial strain values for all myocardial segments. (Radial peak strain values are presented as positive values).

Excluding children weighing less than 15 kg, transvenous pacing systems may be safely implanted in children with structurally normal hearts. Active fixation endocardial leads are preferred in most children. Epicardial pacing is preferred in neonates and small infants, or when coexisting congenital heart disease exists with difficult venous access of the heart.
Pacemaker-induced ventricular dysfunction and adverse remodelling are of concern in children who need life-long pacing. Definitive risk factors are to be determined. The concept of pacemaker-induced heart failure will have important implications in the way we treat and manage children with permanent pacemakers, with selective site pacing being likely to play a more important role.

8. References

Adwani, SS.; Sreeram, N. & DeGiovanni, JV. (1997). Percutaneous transhepatic dual chamber pacing in children with Fontan circulation. *Heart*, 77:574-575.

Alexander, ME. (2004). Transvenous pacing in infants: a faith based initiative? *Pacing Clin Electrophysiol*, 27:1463-1465.

Anderson, RH.; Becker, AE.; Arnold, R. & Wilkinson, JL. (1974). The conducting tissues in congenitally corrected transposition. *Circulation*, 50:911-923.

Anderson, RH.; Weninck, ACG.; Losekoot, TG. & Becker, AE. (1977). Congenitally complete heart block: developmental aspects. *Circulation,* 56:90-101.

Bar-Cohen, Y.; Berul, CI.; Alexander, ME.; et al. (2006). Age, size, and lead factors alone do not predict venous obstruction in children and young adults with transvenous lead systems. *J Cardiovasc Electrophysiol,* 17:754-759.

Bakhtiary, F.; Dzemali, O.; Bastanier, CK.; Moritz, A. & Kleine, P. (2007) Medium-term follow-up and modes of failure following epicardial pacemaker implantation in young children. *Europace,* 9:94-97.

Balmer, C.; Fasnacht, M.; Rahn, M.; Molinari, L. & Bauersfeld, U. (2002). Long-term follow-up of children with congenital complete atrioventricular block and impact of pacemaker therapy. *Europace,* 4:345-349.

Balmer, C. & Bauersfeld U. (2003). Do all children with congenital complete atrioventricular block require permanent pacing? *Indian Pacing Electrophysiol J,* 3:178-183.

Barakat, K.; Robinson, NM. & Spurrell, RA. (2000). Transvenous pacing lead-induced thrombosis: a series of cases with a review of the literature. *Cardiology,* 93:142-148.

Barber, BJ.; Batra, AS.; Burch, GH.; et al. (2005). Acute hemodynamic effects of pacing in patients with Fontan physiology: a prospective study. *J Am Coll Cardiol,* 46:1937-1942.

Batra, AS.; Wells, WJ.; Hinoki, KW.; Stanton, RA. & Silka, MJ. (2003). Late recovery of atrioventricular conduction after pacemaker implantation for complete heart block associated with surgery for congenital heart disease. *J Thorac Cardiovasc Surg*, 125:1291-1293.

Batra, AS. & Balaji, S. (2006). Pacing in adults with congenital heart disease. *Expert Rev Cardiovasc Ther,* 4:663-670.

Bauersfeld, U.; Nowak, B.; Molinari, L.; et al. (1999). Low energy epicardial pacing in children: the benefit of autocapture. *Ann Thorac Surg,* 68:1380-1383.

Beaufort-Krol, G.; Schasfoort-Van Leeuwen, MJM.; Stienstra, Y. & Bink-Boelkens MThE. (2007). Longitudinal echocardiographic follow-up in children with congenital complete atrioventricular block. *Pacing Clin Electrophysiol,* 30:1339-1343.

Benditt, DG.; Milstein, S.; Buetikofer.; et al. (1987). Sensor-triggered, rate-variable cardiac pacing. *Ann Intern Med,* 107:714-724.

Boutjdir, M.; Chen, L.; Zhang, ZH.; Tseng, CE.; El-Sherif, N. & Buyon, JP. (1998). Serum and immunoglobulin G from the mother of a child with congenital heart block induce conduction abnormalities and inhibit L-type calcium channels in a rat heart model. *Pediatr Res,* 44:11-19.

Bracke, F.; Meijer, A. & van Gelder, B. (2003). Venous occlusion of the access vein in patients referred for lead extraction: influence of patient and lead characteristics. *Pacing Clin Electrophysiol,* 26:1649-1652.

Breivik, K.; Engedal, H. & Ohm, OJ. (1982). Electrophysiological properties of a new permanent endocardial lead for uni- and bipolar pacing. *Pacing Clin Electrophysiol*, 5:268-274.

Breivik, K.; Ohm, OJ. & Engedal, H. (1983). Long-term comparison of unipolar and bipolar pacing and sensing, using a new multiprogrammable pacemaker system. *Pacing Clin Electrophysiol,* 6:592-600.

Breur, JM.; Udink ten Cate, FE.; Kapusta, L.; et al. (2002). Pacemaker therapy in isolated congenital atrioventricular block. *Pacing Clin Electrophysiol,* 25:1685-1691.

Breur, JM.; Udink ten Cate, FE.; Kapusta, L.; et al. (2006). Potential additional indicators for pacemaker requirement in isolated congenital atrioventricular block. *Pediatr Cardiol,* 27:564-568.

Brucato, A.; Jonzon, A.; Friedman, D.; et al. (2003). Proposal for a new definition of congenital complete atrioventricular block. *Lupus,* 12:427-435.

Buckingham, TA.; Janosik, DL. & Pearson, AC. (1992). Pacemaker hemodynamics: clinical implications. *Prog Cardiovasc Dis,* 34:347-366.

Buyon, JP. & Winchester, R. (1990). Congenital complete heart block: a human model of passively acquired autoimmune injury. *Arthritis Rheum,* 33:609-614.

Buyon, JP.; Waltuck, J.; Kleinman, C. & Copel, J. (1995). In utero identification and therapy of congenital heart block. *Lupus,* 4:116-21.

Buyon, JP.; Hiebert, R.; Copel, J.; et al. (1998). Autoimmune-associated congenital heart block: demographics, mortality, morbidity, and recurrence rates obtained from a national neonatal lupus registry. *J Am Coll Cardiol,* 31:1658-1666.

Buyon, JP. & Clancy, RM. (2005). Autoantibody-associated congenital heart block: TGFbeta and the road to scar. *Autoimmun Rev,* 4:1-7.

Cabrera, ME.; Portzline, G.; Aach, S.; Condie, C.; Dorostkar, P. & Mianulli, M. (2002). Can current minute ventilation rate adaptive pacemakers provide appropriate chronotropic response in pediatric patients? *Pacing Clin Electrophysiol,* 25:907-914.

Chakrabarti, S.; Morgan, GJ.; Kenny, D.; et al. (2009). Initial experience of pacing with a lumenless lead system in patients with congenital heart disease. *Pacing Clin Electrophysiol, 32:*1428-1433.

Chamberlain, DA.; Leinbach, RC.; Vassaux, CE.; et al. (1970). Sequential atrioventricular pacing in heart block complicating acute myocardial infarction. *N Engl J Med,* 282:577-582.

Chen, CA.; Chang, CI.; Wang JK.; et al. (2008) Restoration of cardiac function by setting the ventricular pacing at a lower range in an infant with congenital complete atrioventricular block and dilated cardiomyopathy. *Int J Cardiol*, 131:e38-e40.

Cheng, A.; Helm, RH. & Abraham, TP. (2009). Pathophysiological mechanisms underlying ventricular dyssynchrony. *Europace,* 11:v10-v14.

Cohen, MI.; Vetter, VL.; Wernovsky, G.; et al. (2001a). Epicardial pacemaker implantation and follow-up in patients with a single ventricle after the Fontan operation. *J Thorac Cardiovasc Surg,* 121:804-811.

Cohen, MI.; Bush, DM.; Vetter, VL.; et al. (2001b). Permanent epicardial pacing in pediatric patients: seventeen years of experience and 1200 outpatient visits. *Circulation*, 103:2585-2590.

Cohen, MI.; Bush, DM.; Gaynor, JW.; Vetter, VL.; Tanel, RE. & Rhodes, LA. (2002). Pediatric pacemaker infections: Twenty years of experience. *J Thorac Cardiovasc Surg,* 124:821-827.

Cohen, MI.; Buck, K.; Tanel, RE.; et al. (2004). Capture management efficacy in children and young adults with endocardial and unipolar epicardial systems. *Europace*, 6:248-255.

Cruz, RB.; Viana, VS.; Nishioka, SA.; Martinelli, FM. & Bonfa, E. (2004). Is isolated congenital heart block associated to neonatal lupus requiring pacemaker a distinct cardiac syndrome? *Pacing Clin Electrophysiol*, 27:615-620.

Dewey, RC.; Capeless, MA. & Levy, AM. (1987). Use of ambulatory electrocardiographic monitoring to identify high risk patients with congenital complete heart block. *N Eng J Med*, 316:835-839.

Driscoll, DJ.; Gilette, PC.; Hallman, GL.; Cooley, DA. & McNamara, DG. (1979). Management of surgical complete atrioventricular block in children. *Am J Cardiol*, 43:1175-1180.

Emmel, M.; Sreeram, N.; Brockmeier, K. & Bennink, B. (2007). Superior vena cava stenting and transvenous pacemaker implantation (stent and pace) after the Mustard operation. *Clin Res Cardiol*, 96:17-22.

Epstein, AE.; DiMarco, JP.; Ellenbogen, KA.; et al. (2008). ACC/AHA/HRS 2008 guidelines for devicebased therapy of cardiac rhythm abnormalities. *Heart Rhythm*, 5:e1-e62.

Esscher, E. & Michaelsson, M. (1983). QT interval in congenital complete heart block. *Pediatr Cardiol*, 4:121-124.

Figa, FH.; McCrindle, BW.; Bigras, JL.; Hamilton, RM. & Gow, RM. (1997). Risk factors for venous obstruction in children with transvenous pacing leads. *Pacing Clin Electrophysiol*, 20:1902-1909.

Fortescue, E.; Berul, CI.; Cecchin, F.; et al. (2004). Patient procedural and hardware factors associated with pacemaker lead failures in pediatrics and congenital heart disease. *Heart Rhythm*, 1:150-159.

Fryda, R. & Kaplan, S. (1970). Postoperative heart block in children. *Am J Cardiol* 25:96.

Furman, S.; Pannizzo, F. & Campo, I. (1979). Comparison of active and passive adhering leads for endocardial pacing. *Pacing Clin Electrophysiol*, 2:417-427.

Furman, S. (1994). Pacemaker syndrome. *Pacing Clin Electrophysiol*, 17:1-5.

Gasparini, M.; Mantica, M.; Galimberti, P.; Coltorti, F.; Ceriotti, C. & Priori, S. (2000). Inferior vena cava loop of the implantable cardioverter defibrillator endocardial lead: A possible solution to the growth problem in pediatric implantation. *Pacing Clin Electrophysiol*, 23:2108-2112.

Gebauer, RA.; Tomek, V.; Salameh, A.; et al. (2009). Predictors of left ventricular remodelling and failure in right ventricular pacing in the young. *Eur Heart J*, 30:1097-1104.

Gheissari, A.; Hordof, AJ. & Spotnitz, HM. (1991). Transvenous pacemakers in children: relation of lead length to anticipated growth. *Ann Thorac Surg*, 52:118-121.

Gillette, PC.; Wampler, DG.; Shannon, C. & Ott, D. (1986). Use of cardiac pacing after the Mustard operation for transposition of the great arteries. *J Am Coll Cardiol*, 7:138-141.

Gillette, PC.; Zeigler, V.; Bradham, GB. & Kinsella, P. (1988). Pediatric transvenous pacing: a concern for venous thrombosis? *Pacing Clin Electrophysiol*, 11:1935-1939.

Gillette, PC.; Edgerton, J.; Kratz, J. & Zeigler, V. (1991). The subpectoral pocket: the preferred implant site for pediatric pacemakers. *Pacing Clin Electrophysiol*, 14:1089-1092.

Goldman, BS.; Williams, WG.; Hill, T.; et al. (1985). Permanent cardiac pacing after open heart surgery: congenital heart disease. *Pacing Clin Electrophysiol*, 8:732-739.
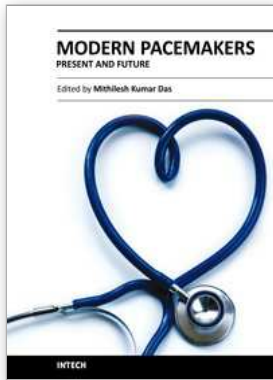
Goldman-Cutler, N.; Karpawich, PP.; Cavitt, D.; et al. (1997). Steroid-eluting epicardial pacing electrodes: six year experience of pacing thresholds in a growing pediatric population. *Pacing Clin Electrophysiol*, 20:2943-2948.

Graham, TP Jr.; Bernhard, YD.; Mellen, BG.; et al. (2000). Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. *J Am Coll Cardiol*, 36:255-261.

Gross, GJ.; Chiu, CC.; Hamilton, RM.; Kirsh, JA. & Stephenson, EA. (2006). Natural history of postoperative heart block in congenital heart disease: implications for pacing intervention. *Heart Rhythm*, 3:601-604.

Hartzler, GO.; Maloney, JD.; Curtis, JJ.; et al. (1977). Hemodynamic benefits of atrioventricular sequential pacing after cardiac surgery. *Am J Cardiol*, 40:232-236.

Hayes, CJ. & Gersony, WM. (1986). Arrhythmias after the Mustard operation for transposition of the great arteries: A long-term study. *J Am Coll Cardiol*, 7:133-137.

Hidden-Lucet, F.; Halimi, F.; Gallais, Y.; Petitot, JC.; Fontaine, G. & Frank, R. (2000). Low chronic pacing thresholds of steroid-eluting active fixation ventricular pacemaker leads: a useful alternative to passive fixation leads. *Pacing Clin Electrophysiol*, 23:1798-1800.

Hoorntje, T.; Kammeraad, J.; Bennink, G. & Sreeram, N. (2000). Transvenous permanent pacemaker implantation in neonates. *Int J Cardiol*, 75:103-104.

Horenstein, MS. & Karpawich, PP. (2004). Pacemaker syndrome in the young: Do children need dual chamber pacing as the initial pacing mode? *Pacing Clin Electrophysiol*, 27:600-605.

Horenstein, MS.; Karpawich, PP. & Tantengco, MVT. (2003). Single versus dual chamber pacing in the young: noninvasive comparative eveluation of cardiac function. *Pacing Clin Electrophysiol*, 26:1208-1211.

Inoue, S.; Mizobuchi, M.; Yoshimura, N.; Yamaguchi, M. & Nakao, H. (2005). Succesful perinatal management of a very low birthweight infant with congenital complete atrioventricular block. *Am J Perinat*, 22:387-390.

Jaeggi, E.; Laskin, C.; Hamilton, R.; Kingdom, J. & Silverman, E. (2010). The importance of the level of maternal anti-Ro/SSA antibodies as a prognostic marker of the development of cardiac neonatal lupus erythematosus a prospective study of 186 antibody-exposed fetuses and infants. *J Am Coll Cardiol*, 55:2778-2784.

Janousek, J.; Tomek, V.; Chaloupecky, V. & Gebauer, RA. (2004). Dilated cardiomyopathy associated with dual-chamber pacing in infants: improvement through either left ventricular cardiac resynchronization or programming the pacemaker off allowing intrinsic normal conduction. *J Cardiovasc Electrophysiol*, 15:470-474.

Johns, JA.; Fish, FA.; Burger, JD.; et al. (1992). Steroid-eluting epicardial pacing leads in pediatric patients: encouraging early results. *J Am Coll Cardiol*, 20:395-401.

Johnson, C. & Galindez, L. (1998). Multiple systemic emboli complicating the course of a patient with an atrial septal defect, an atrial septal aneurysm and an endocardial right atrial pacemaker lead. *P R Health Sci J*, 17:281-284.

Julkunen, H. & Eronen, M. (2001). Long-term outcome of mothers of children with isolated heart block in Finland. *Arthritis Rheum*, 44:647-652.

Kaltman, J.; Ro, P.; Zimmerman, F.; et al. (2008). Managed ventricular pacing in pediatric patients and patients with congenital heart disease. *Am J Cardiol*, 102:875-878.

Kammeraad, J.; Rosenthal, E.; Bostock, J.; et al. (2004). Endocardial pacemaker implantation in infants weighing < 10 kg. *Pacing Clin Electrophysiol,* 27:1466-1474.

Karpawich, PP. (2004). Chronic right ventricular pacing and cardiac performance: the pediatric perspective. *Pacing Clin Electrophysiol,* 27:844-849.

Karpawich, PP.; Rabah, R. & Haas, JE. (1999). Altered cardiac histology following apical right ventricular pacing in patients with congenital atrioventricular block. *Pacing Clin Electrophysiol,* 22:1372-1377.

Kass, DA. (2008). An epidemic of dyssynchrony. But what does it mean? *J Am Coll Cardiol,* 51:12-17.

Khairy, P.; Landzberg, MJ.; Lambert, J. & O'Donnell, CP. (2004). Long-term outcomes after the atrial switch for surgical correction of transposition: a meta-analysis comparing the Mustard and Senning procedures. *Cardiol Young,* 14:284-292.

Khairy, P.; Landzberg, MJ.; Gatzoulis, M.; et al. (2006). Transvenous pacing leads and systemic thromboemboli in patients with intracardiac shunts: a multicenter study. *Circulation,* 113:2391-2397.

Kim, JJ.; Friedman, RA.; Eidem, BW.; et al. (2007). Ventricular function and long-term pacing in children with congenital complete atrioventricular block. *J Cardiovasc Electrophysiol,* 18:373-377.

Kiviniemi, MS.; Pirnes, MA.; Eranen, HJK.; Kettunen, RVJ. & Hartikainen, JEK. (1999). Complications related to permanent pacemaker therapy. *Pacing Clin Electrophysiol,* 22:711-720.

Klug, D.; Lacroix, D.; Savoye, C.; et al. (1997). Systemic infection related to endocarditis on pacemaker leads: Clinical presentation and management. *Circulation,* 95:2098-2107.

Lev, M.; Silverman, J.; Fitzmaurice, FM.; Paul, MH.; Cassells, DE. & Miller, RA. (1971). Lack of connection between the atria and the more peripheral conduction system in congenital atrioventricular block. *Am J Cardiol,* 27:481-490.

Llanos, C.; Izmirly, PM.; Katholi, M.; et al. (2009). Recurrence rates of cardiac manifestations associated with neonatal lupus and maternal/fetal risk factors. *Arthritis Rheum,* 60:3091-3097.

Liberman, L.; Pass, RH.; Hordof, AJ. & Spotnitz, HM. (2008). Late onset of heart block after open heart surgery for congenital heart disease. *Pediatr Cardiol,* 29:56-59.

Maginot, KR.; Mathewson, JW.; Bichell, DP. & Perry, JC. (2000). Applications of pacing strategies in neonates and infants. *Progr Pediatr Cardiol,* 11:65-75.

Mansour, F.; Talajic, M.; Thibault, B. & Khairy, P. (2006). Pacemaker troubleshooting: when MVP is not the most valuable parameter. *Heart Rhythm,* 5:612-614.

Martin, MV.; Lime, AB.; Almeida, CS; et al. (1966). Implantation of Chardack-Greatbatch adjustable rate and current pacemaker in a 4 month old infant. *Pediatrics,* 37:323-328.

McCue, CM.; Mantakas, ME.; Tingelstad, JB. & Ruddy, S. (1977). Congenital heart block in newborns of mothers with connective tissue disease. *Circulation,* 56:82-90.

van Mechelen, R. & Schoonderwoerd, R. (2006). Risk of managed ventricular pacing in a pacing with heart block. *Heart Rhythm,* 11:1384-1385.

Michaelsson, M. & Engle, MA. (1972). Congenital complete heart block: an international study of the natural history. *Cardiovasc Clin,* 4:86-101.

Michaelsson, M.; Jonzon, A. & Riesenfeld, T. (1995). Isolated congenital complete atrioventricular block in adult life: a prospective study. *Circulation,* 92:442-449.

Michaelsson, M.; Riesenfeld, A. & Jonzon, A. (1997). Natural history of congenital complete atrioventricular block. *Pacing Clin Electrophysiol,* 20:2098-2101.

Miller, GL.; Horneffer, PJ.; Gardner, TJ.; Rykiel, MF. & Pearson, TA. (1988). The epidemiology of the postpericardiotomy syndrome: a common complication of cardiac surgery. *Am Heart J*, 116:1323-1329.

Moak, JP.; Barron, KS.; Hougen, TJ.; et al. (2001). Congenital heart block: development of late-onsetcardiomyopathy, a previously underappreciated sequela. *J Am Coll Cardiol,* 37:238-242.

Moak, JP.; Hasbani, K.; Ramwell, C.; et al. (2006). Dilated cardiomyopathy following right ventricular pacing for AV block in young patients: resolution after upgrading to biventricular pacing systems. *J Cardiovasc Electrophysiol,* 17:1068-1071.

Mond, HG. (1991). Unipolar versus bipolar pacing – poles apart. *Pacing Clin Electrophysiol,* 14:1411-1424.

Mond, HG. & Stokes KB. (1992). The electrode-tissue interface: the revolutionary role of steroid elution. *Pacing Clin Electrophysiol*, 15:95-107.

Nield, LE.; Silverman, ED.; Taylor, GP.; et al. (2002). Maternal anti-Ro and anti-La antibodyassociated endocardial fibroelastosis. *Circulation,* 105:843-848.

Nothroff, J.; Norozi, K.; Alpers, V.; et al. (2006). Pacemaker implantation as a risk factor for heart failure in young adults with congenital heart disease. *Pacing Clin Electrophysiol,* 29:386-392.

Ohmi, M.; Tofukuji, M.; Sato, K.; et al. (1992). Permanent pacemaker implantation in premature infants less than 2,000 grams of body weight. *Ann Thorac Surg,* 54:1223-1225.

Oldroyd, KG.; Rae, AP.; Carter, R.; Wingate, C. & Cobbe, SM. (1991). Double blind cross over comparison of the effects of dual chamber pacing (DDD) and ventricular rate adaptive (VVIR) pacing on neuroendocrine variables, exercise performance and symptoms in complete heart block. *Br Heart J,* 65:188-193.

Polin, GM.; Zado, E.; Nayak, H.; et al. (2006). Proper management of pericardial tamponade as a late complication of implantable cardiac device placement. *Am J Cardiol*, 98:223-225.

Prinzen, FW.; Cheriex, EM.; Delhaas, T.; et al. (1995). Asymmetric thickness of the left ventricular wall resulting from asynchronous electrical activation. A study in patients with left bundle branch block in dogs with ventricular pacing. *Am Heart J,* 130:1045-1053.

Ragonese, P.; Guccione, P.; Drago, F.; Turchetta, A.; Calzolari, A. & Formigari, R. (1994). Efficacy and safety of ventricular rate responsive pacing in children with complete atrioventricular block. *Pacing Clin Electrophysiol,* 17:603-610.

Rosenthal, E.; Qureshi, SA. & Crick, JC. (1995). Successful long-term ventricular pacing via the coronary sinus after the Fontan operation. *Pacing Clin Electrophysiol,* 18:2103-2105.

Rosenthal, E.; Bostock, J.; Qureshi, SA.; Baker, EJ. & Tynan, M. (1997a). Single pass VDD pacing in children and adolescents. *Pacing Clin Electrophysiol,* 20:1975-1982.

Rosenthal, E. & Bostock, J. (1997b). Use of an atrial loop to extend the duration of endocardial pacing in a neonate. *Pacing Clin Electrophysiol,* 20:2489-2491.

Sachweh, JS.; Vazquez-Jiminez, JF.; Schondube, FA.; et al. (2000). Twenty years experience with pediatric pacing: epicardial and transvenous stimulation. *Eur J Cardiothorac Surg,* 17:455-461.

Sanjeev, S. & Karpawich P. (2006). Superior vena cava and innominate vein dimensions in growing children: An aid for interventional devices and transvenous leads. *Pediatr Cardiol*, 27:414-419.

Seiden, HS.; Camunas, JL.; Fishburger, SB.; et al. (1997). Use of single lead VDD pacing in children. *Pacing Clin Electrophysiol,* 20:1967-1974.

Simon, A.; Dick, M 2nd.; Stern, A.; Behrendt, D. & Sloan, H. (1982). Ventricular pacing in children. *Pacing Clin Electrophysiol*, 5:836-844.

Silka, MJ. & Rice, MJ. (1991). Paradoxic embolism due to altered hemodynamic sequencing following transvenous pacing. *Pacing Clin Electrophysiol,* 14:499-503.

Silvetti, MS.; De Santis, A.; Grovale, N.; Grutter, G.; Baccarini, A. & Drago, F. (2007a). Ventricular pacing threshold variations in the young. *Pacing Clin Electrophysiol,* 30:175-181.

Silvetti, MS.; Drago, F.; De Santis, A.; et al. (2007b). Single-centre experience on endocardial and epicardial pacemaker system function in neonates and infants. *Europace,* 9:426-431.

Sholler, GF. & Walsh EP. (1989). Congenital complete heart block in patients without anatomic cardiac defects. *Am Heart J,* 118:1193-1198.

Spindler, M.; Burrows, G.; Kowallik, P.; Ertl, G. & Voelker, W. (2001). Postpericardiotomy syndrome and cardiac tamponade as a late complication after pacemaker implantation. *Pacing Clin Electrophysiol*, 24:1433-1444.

Stephenson, EA. & Kaltman, JR. (2006). Current state of the art for use of pacemakers and defibrillators in patients with congenital cardiac malformations. *Cardiol Young,* 16:151-156.

Sweeney, MO.; Shea, J.; Fox, V.; et al. (2004). Randomized pilot study of a new atrial-based minimal ventricular pacing mode in dual-chamber implantable-cardioverter-defibrillators. *Heart Rhythm*, 1:160-167.

Sweeney, MO.; Ellenbogen, KA.; Casavant, D.; et al. (2005). Multicenter, prospective, randomized safety and efficacy study of a new atrial-based managed ventricular pacing mode (MVP) in dual chamber ICDs. *J Cardiovasc Electrophysiol*, 16:811-817.

Thambo, JB.; Bordachar, P.; Garrigue, S.; et al. (2004). Detrimental ventricular remodelling in patients with congenital complete heart block and chronic right ventricular apical pacing. *Circulation,* 110:3766-3772.

Tomaske, M.; Harpes, P.; Pretre, E.; Dodge-Khatami, A. & Bauersfeld, U. (2007). Long-term experience with AutoCapture-controlled epicardial pacing in children. *Europace,* 9:645-650.

Tops, LF.; Schalij, MJ.; Holman, ER.; van Erven, L.; van der Wall, EE. & Bax, JJ. (2006). Right ventricular pacing can induce ventricular dyssynchrony in patients with atrial fibrillation after atrioventricular node ablation. *J Am Coll Cardiol,* 48:1642-1648.

Turley, K.; Hanley, FL.; Verrier, ED.; et al. (1988). The Mustard procedure in infants (less than 100 days of age). Ten year follow-up. *J Thorac Cardiovasc Surg*, 96:849-853.

Vardas, PE.; Auricchio, A.; Blanc, JJ.; et al. (2007). Guidelines for cardiac pacing and cardiac resynchronization therapy: the task force for cardiac pacing and cardiac resynchronization therapy of the European Society of Cardiology. Developed in

collaboration with the European Heart Rhythm Association. *Eur Heart J,* 28:2256-2295.

Vatasescu, R.; Shalganov, T.; Paprika, D.; et al. (2007). Evolution of left ventricular function in paediatric patients with permanent right ventricular pacing for isolated congenital heart block: a medium term follow-up. *Europace,* 9:228-232.

Vetter, VL.; Tanner, CS. & Horowitz, LN. (1987). Electrophysiologic consequences of the Mustard repair of D-transposition of the great arteries. *J Am Coll Cardiol,* 10:1265-1273.

Villain, E.; Coastedoat-Chalumeau, N.; Marijon, E.; et al. (2006). Presentation and prognosis of complete atrioventricular block in childhood, according to maternal antibody status. *J Am Coll Cardiol,* 48:1682-1687.

Vogt, P.; Goy, JJ.; Kuhn, M.; Leuenberger, P. & Kappenberger, L. (1988). Single versus double chamber rate responsive cardiac pacing: comparison by cardiopulmonary noninvasive exercise testing. *Pacing Clin Electrophysiol,* 11:1896-1901.

Warfield, DA.; Hayes, DL.; Hyberger, LK.; Warnes, CA. & Danielson, GK. (1999). Permanent pacing in patients with univentricular heart. *Pacing Clin Electrophysiol,* 22:1193-1201.

Welisch, E.; Cherlet, E.; Crespo-Martiney, E. & Hansky, B. (2010). A single institution experience with pacemaker implantation in a pediatric population over 25 years. *Pacing Clin Electrophysiol*, 33:1112-1118.

Udink ten Cate, FE.; Breur, JM.; Cohen, MI.; et al. (2001). Dilated cardiomyopathy in isolated congenital complete atrioventricular block: early and long-term risk in children. *J Am Coll Cardiol,* 37:1129-1134.

Udink ten Cate, FE.; Breur, JM.; Boramanand, N.; et al. (2002). Endocardial and epicardial steroid lead pacing in the neonatal and paediatric age group. *Heart*, 88:392-396.

Udink ten Cate, FE.; Kruessell MA. ; Wagner, K. ; et al. (2010a). Dilated cardiomyopathy in children with ventricular preexcitation : the location of the accessory pathway is predictive of this association. *J Electrocardiol,* 43:146-154.

Udink ten Cate, FE.; Wiesner, N. ; Trieschmann, U. ; Khalil, M. & Sreeram, N. (2010b). Dyssynchronous ventricular activation in asymptomatic Wolff_parkinson-White: a risk factor for development of dilated cardiomyopathy. *Indian Pacing Electrophysiol J*, 10:248-256.

Zeltser, I.; Rhodes, LA.; Tanel, RE.; et al. (2004). Postpericardiotomy syndrome after permanent pacemaker implantation in children and young adults. *Ann Thorac Surg*, 78:1684-1687.

Modern Pacemakers - Present and Future

Edited by Prof. Mithilesh R Das

The book focuses upon clinical as well as engineering aspects of modern cardiac pacemakers. Modern pacemaker functions, implant techniques, various complications related to implant and complications during follow-up are covered. The issue of interaction between magnetic resonance imaging and pacemakers are well discussed. Chapters are also included discussing the role of pacemakers in congenital and acquired conduction disease. Apart from pacing for bradycardia, the role of pacemakers in cardiac resynchronization therapy has been an important aspect of management of advanced heart failure. The book provides an excellent overview of implantation techniques as well as benefits and limitations of cardiac resynchronization therapy. Pacemaker follow-up with remote monitoring is getting more and more acceptance in clinical practice; therefore, chapters related to various aspects of remote monitoring are also incorporated in the book. The current aspect of cardiac pacemaker physiology and role of cardiac ion channels, as well as the present and future of biopacemakers are included to glimpse into the future management of conductions system diseases. We have also included chapters regarding gut pacemakers as well as pacemaker mechanisms of neural networks. Therefore, the book covers the entire spectrum of modern pacemaker therapy including implant techniques, device related complications, interactions, limitations, and benefits (including the role of pacing role in heart failure), as well as future prospects of cardiac pacing.

How to reference

In order to correctly reference this scholarly work, feel free to copy and paste the following:

Floris E.A. Udink ten Cate and Narayanswami Sreeram (2011). Pacing Therapy in Infants and Children with Congenital and Acquired Complete Atrioventricular Block: Optimal Pacing Strategies, Management, and Follow-up, Modern Pacemakers - Present and Future, Prof. Mithilesh R Das (Ed.), ISBN: 978-953-307-214-2, InTech, Available from: http://www.intechopen.com/books/modern-pacemakers-present-and-future/pacing-therapy-in-infants-and-children-with-congenital-and-acquired-complete-atrioventricular-block-

INTECH
open science | open minds