

CONDUCTION DISEASE IN SPECIFIC CONDITIONS

Recent Myocardial Infarction

In the reperfusion era the incidence of AV block has decreased to 7%,¹ whereas the incidence of bundle branch block remains at 5% (and additional 18% as transient).² They both predict higher mortality. Indications for permanent pacing (2-4 days after anterior and 1-2 weeks after inferior MI) are presented in the Chapter of ACS Therapy.

Pacing after surgery and heart transplantation

Patients who develop complete atrioventricular block within 24 hours after surgical valve replacement and which then persisted for > 48 hours, have been unlikely to recover.³ However, AV conduction block affects up to 15% of patients subjected to cardiac surgery, but block requiring pacing is seen in 4-6% following valve surgery and <1% following CABG because late AV conduction recovery is not uncommon after cardiac surgery in patients without preoperative AV conduction abnormalities.⁴ Thus, a period of clinical observation is necessary before permanent pacing is undertaken. Recommendations for the appropriate management of patients undergoing non-cardiac and cardiac surgery are presented in Tables 1 and 2. Specific recommendations for pacing after AS surgery and TAVR as well as after MV and TS surgery are also presented in relevant Chapters. Late-onset AV block occurs in 2.4% of patients with orthotopic heart transplant or heart-lung transplant. AV block is predominantly intermittent and, often, does not progress to permanent AV block.⁵ The bicaval surgical technique and young donor/recipient age are protective against a postoperative pacemaker requirement.⁶ Following orthotopic heart transplantation a degree of chronotropic incompetence is inevitable, but both sinus node and AV node functions improve during the first weeks after the operation. The optimal timing for permanent pacing should be individualized.

Congenital AV block

Congenital AV block may be immune (usually) or nonimmune. Transplacental penetration of ***anti-Ro/SSA*** and ***anti-La/SSB*** ribonucleoprotein antibodies from the mother, who may have ***systemic lupus erythematosus***, ***systemic sclerosis***, or ***Sjogren syndrome***, or may even be entirely asymptomatic, into the fetal circulation is associated with congenital conduction disturbances. Half of these asymptomatic women develop symptoms of a rheumatic disease,

most commonly arthralgias and xerophthalmia, but few develop lupus nephritis.⁷ Anti-Ro/SSA antibodies, specifically the anti-Ro/SSA-52kD subtype, may cross-react with T- and L-type calcium channels and the potassium channel hERG, and induce AV block.^{8,9} Congenital complete heart block is the more severe manifestation of so-called "Neonatal Lupus". It occurs in approximately 2% of neonates whose mothers are positive for anti-Ro/SSA and anti-La/SSB, suggesting that additional genetic and environmental and other factors, such as, possibly, vitamin D deficiency, may also play a role in the development of block.^{8 10 11} It is typically detected in utero or within the neonatal period (0-27 days after birth).¹² Of affected fetuses 17.5% die, 30% in utero, and the cumulative probability of survival at 10 years for a child born alive is 86%, with a higher case fatality rate in non-white patients.¹³ Twice-daily ambulatory home fetal heart rhythm monitoring with a commercial Doppler device in anti-Ro antibody positive pregnancies can successfully detect the irregularity of emerging second-degree AV block before progression to complete block, thus defining a window of time for successful therapy.¹⁴ The risk of recurrence of CHB in a subsequent child is 10-18%.¹⁵ Complete AV block is irreversible, while incomplete AV block may be potentially reversible after fluorinated steroid therapy.¹² Anti-Ro/SSA antibodies might also be pathogenic for the adult heart with second degree AV block, sinus bradycardia, QT prolongation, and ventricular arrhythmias seen in adults.¹² Anti-Ro Abs from patients with autoimmune diseases inhibit I_{Kr} by cross-reacting with the HERG channel.¹⁶ The adult AV node is generally thought to be resistant to the damaging effect of anti-Ro (SSA) and anti-La (SSB) autoantibodies. However, anecdotal case reports suggest that heart block developing in adult SS patients may be associated with such concurrent autoantibodies.

Maternal use of **hydroxychloroquine** is also associated with a reduced risk of recurrent cardiac manifestations.¹⁷ Congenital or childhood nonimmune AV block may also occur. ECG screening in parents of children affected by idiopathic AV block has revealed a high prevalence of conduction abnormalities, thus suggesting an inheritable trait.¹⁸ Patients with congenital complete heart block have a high incidence of late sudden death at any age, and permanent pacing should be considered in the asymptomatic individual.¹⁹ In a Danish Registry, AV block of unknown aetiology presenting before the age of 50 years and treated with pacemaker implantation was associated with a three- to four-fold higher rate of the composite endpoint of death or hospitalization for heart failure, ventricular tachyarrhythmia, or cardiac arrest with successful resuscitation.²⁰ Whether these outcomes can be attributed to the deleterious effects of pacing (pacing-induced cardiomyopathy) is not known. Theoretically, permanent pacing should avoid the right ventricular apex, and prefer a septal position.²¹ Indications for permanent pacing in congenital heart block are presented in Table 3.

Adults presenting with isolated AVB of unknown origin should undergo

comprehensive **anti-Ro/SSA-antibody** testing with different methods, among which iWB is mandatory to be included. In the case that at least one test is positive for circulating anti-Ro/SSA antibodies, specifically the anti-Ro/SSA-52kD subtype, the diagnosis of anti-Ro/SSA-associated AVB shall be made. In those that are positive, steroid administration (1mk/Kg per day) for one week may reveal responders who will benefit from long term immunotherapy without a pacemaker.⁹

Pacing in adult congenital heart disease

Indications for permanent pacing in adults with congenital conditions are presented in Table 3. Endocardial leads are generally avoided in adults with CHD and intracardiac shunts. Risk assessment regarding hemodynamic circumstances, concomitant anticoagulation, shunt closure prior to endocardial lead placement, or alternative approaches for lead access should be individualized.²² Devices that minimize ventricular pacing are preferred in SSS, and device with antitachycardia pacing properties should be considered if the underlying anatomic substrate carries a high likelihood of developing intra-atrial reentrant tachycardias.²³ AV block occurs in 1-3% after operations for **congenital heart disease**. Pacemaker implantation is recommended with persistent AV block for 7 days post-operatively. However, patients with post-operative second degree or complete heart left ventricular outflow tract surgery, atrioventricular valve replacement, or ventricular L-Looping could be considered for permanent pacing on the 4th post-operative day.²⁴ Endocardial leads, when feasible and in the absence of intracardiac shunts, are preferable to epicardial leads.²⁵

Sleep apnea

Sleep apnoea syndrome should be considered in the differential diagnosis of bradyarrhythmias, and the presence of sleep apnoea and reduced oxygen saturation may be considered as a risk factor for SCD in subjects with sleep disordered breathing.²⁶ Nasal continuous positive airway pressure (CPAP) therapy is effective in reducing sleep apnea episodes, whereas atrial overdrive pacing may be not.²⁷ Pacing may be needed in patients with symptomatic bradycardia despite CPAP, but its value is not established.

Neuromuscular disorders

The heart is commonly involved in inherited muscle and cardiac phenotypes, cardiomyopathies and rhythms disorders can be the first or predominant manifestations.²⁸ Several different types of muscular dystrophies, such as Emery-Dreifuss muscular dystrophy, limb girdle muscular dystrophy (Erb's muscular dystrophy), myotonic dystrophy type 1 (Steinert's disease), myotonic dystrophy type 2, and desmin-related myopathy, are associated

with cardiac conduction defects.²⁹⁻³¹ Patients typically present with sinus bradycardia, first or higher degree AV block or bundle branch block, and vulnerability for sudden cardiac death. The prevalence of critically prognostic conduction abnormalities in myotonic dystrophies I and II is >20%.³² Close follow-up of these patients is necessary and prophylactic permanent pacing or ICD may be indicated. Recommendations are provide in Table 4.

Pacing in pregnancy

For women who have a stable, narrow, complex junctional escape rhythm, PM implantation can be deferred until after delivery.³³ Complete heart block who exhibit with a slow, wide QRS complex escape rhythm needs permanent pacing. This can be performed safely, especially if the foetus is beyond 8 weeks' gestation using echo guidance (ESC 2013 GL on pacing, IIa-C) or, ideally, electro-anatomic navigation. REcommendatiosn are presented in Figure 1.

Other Issues

In patients who require permanent pacing therapy, before implantation, an assessment of the risk of future **ventricular arrhythmias** and need for an ICD should be performed (ACC/AHA/HRS 2018 GL on bradycardia, I-B-NR).

Although **antitachycardia** algorithms in pacemakers may be useful in certain patients, prevention and termination of atrial tachyarrhymias is not a stand-alone indication for pacing.

In patients who present for pacemaker pulse generator replacement, or for management of pacemaker related complications, in whom the original pacing indication has resolved or is in question, **discontinuation of pacemaker therapy** is reasonable after evaluation of symptoms during a period of monitoring while pacing therapy is off (ACC/AHA/HRS 2018 GL on bradycardia, IIa-C-LD).

Share decision making is essential for the appropriate managements of patients (Table 5).

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