Original Article

Perspectives in Interventional Electrophysiology in Children and those with Congenital Heart Disease Electrophysiology in Children

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Recent developments in paediatric pacing and ablation of arrhythmia substrate have been characterised by adoption and modification of techniques used in adults. Infants, small children and those of all ages with congenital heart disease are a patient group with a higher risk profile needing a special approach.

Current success rates for catheter ablation are high and major complication rates are low. Important issues with respect to long-term outcome include questions about coronary injury, long-term effects of radiation exposure and late recurrence.

Non-fluoroscopic electro-anatomical mapping systems (3D systems), cryo-ablation and remote navigation are techniques recently improved such that it is possible to potentially reduce fluoroscopy and complications.

Pacing in young children and congenital heart disease often warrants an epicardial approach to avoid embolism, venous occlusion and lead failure related to growth. Defibrillator and resynchronisation therapy are increasingly important tools to reduce mortality, although the indications are not as clear as in adult patients without congenital heart disease.

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Introduction

The exploration of arrhythmias in childhood started shortly after the clinical introduction of the electrocardiography in the beginning of the 20th century. Medication, pacemakers and surgery for arrhythmias have been the only way to perform interventions in order to change the arrhythmia substrate permanently. The progress of cardiac catheterisation in combination with the developments in technology and insights of the cardiac conduction system finally triggered the exponential development of modern electrophysiology.

Catheter ablation of cardiac arrhythmias

Modern drug treatment is satisfactory for many patients but may be associated with significant side effects, especially in those undertaking long-term treatment. Cardiac surgery as definitive treatment of arrhythmias was introduced in 1968 although with the limitation of an extremely invasive approach. The first DC His ablation in children was performed in 1983 [1] as an emergency procedure for life-threatening junctional ectopic tachycardia. Soon after the introduction of radiofrequency ablation in adults the technique was adopted for paediatric patients [2].

Although treatment of arrhythmias in adults is directed by published guidelines, the lack of paediatric randomised studies means that the choice of therapy often involves experience and individual decision-making. The choice of therapy is therefore dependent on the nature of the symptoms, the potential risks of the tachycardia, the natural history and finally the risks of the therapy itself.

Infants and smaller children may only have subtle or non-specific symptoms. Slow incessant tachycardias that could eventually result in tachycardia-induced cardiomyopathy may go unappreciated for years. Lack of independence becomes an issue as the child grows and may become the trigger for therapeutic intervention.

The majority of tachycardias in children are paroxysmal supraventricular tachycardias (SVT) and tend to be more benign, although the risk of sudden death is present in WPW syndrome. Tachycardia-induced cardiomyopathy occurs in atrial ectopic tachycardia and the 'persistent form' of junctional reentry tachycardia (PJRT). Children with congenital heart disease are much more likely to have syncope or significant symptoms with SVT than children

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with normal anatomy. Age dependent phenomena like an accelerated ventricular rhythm might be physiologic and therefore not needing therapy at all.

The natural history of paediatric tachycardias is very variable—nevertheless there is a high rate of spontaneous regression in accessory pathways presenting in infancy as well as a spontaneous resolution of ectopy and ectopic tachycardias. Arrhythmias have an age-dependent incidence and therefore atrio-ventricular reentry tachycardia (AVRT) in young children is much more frequent than AV-nodal reentry tachycardia (AVNRT). Ventricular tachycardia (VT) is rare and often idiopathic although underlying heart disease has to be ruled out thoroughly.

Vagal manoeuvres are an option in children with paroxysmal reentry-tachycardia and pose no significant risk except ineffective treatment. Medication offers the chance of significant reduction of episodes or even complete suppression for the time of treatment, but no definitive cure. The side effects and risks of medical treatment have to be considered.

What are the current results of ablation in childhood?

The Paediatric Radiofrequency Catheter Ablation Registry [3] has collected data on 7600 ablations over 10 years (1991–2000). Although the character of this registry (without mandatory follow-up or inclusion of all procedures) will underestimate the recurrence rates and late complications it presents a good idea of the numbers of acute success and complications. The majority of ablation has been performed in accessory pathways (5383 AVRT (71%) followed by 1815 AVNRT (24%), and 402 focal or ectopic atrial tachycardia (5%)). Data from the early era and the late era were compared for the outcomes of success rate and complication rate. In the registry as a whole, ablation failure rates fell from 9.6% in the early era to 4.8% in the more recent era. Therefore the mean success rate for ablation in SVT in children with normal anatomy should be around 95%. Of course there is a certain variance depending on substrate and age, ranging from 81% in atrial ectopic tachycardia in children less than five years to 99% in AVNRT (all age groups).

Complications have been defined as significant and therefore included in this study in case they required emergency or ongoing treatment or follow-up or the residual effects interfered with normal function. Complications have been 9% for children less than five years of age compared with 3% in children older than five years of age.

What are the unknown/unaddressed risks?

Whereas AV block and short-term complications like bleeding, pericardial effusion and arrhythmias are well known and manageable there are potential long-term risks.

In the above-mentioned registry, fluoroscopy times have been documented. Although these do not precisely correlate with radiation dose, they do reflect the possible range of the dose. Fluoroscopy time is not dependent on age and decreased significantly in the more recent era from 50.9 min (±39.9 min) to 40.1 min (±35.1 min).

Clay et al. [4] calculated risk based on measurements with a mean fluoroscopy time of only 14.4 min, which is significantly shorter than the times above. Nevertheless the increased lifetime risk of fatal malignancy was 0.02% per single RFA procedure. Fluoroscopy should be considered a late risk factor, given the benign characteristics of SVT in childhood.

Although radiofrequency (RF) ablation has become established therapy for paediatric tachyarrhythmias, challenges remain in terms of the safety and of RF ablation in specific locations, especially in regards to the potential for injury to the coronary arteries. This risk seems to be highest when ablating within the coronary sinus and coronary veins. There have been reports of coronary stenosis in AVNRT ablation [5], atrial flutter [6] and for pathway ablation [7]. A systematic review in a paediatric group [8] showed that RF ablation of right postero-septal pathways may potentially cause coronary injury. As most groups do not routinely image coronary anatomy before and after ablation there might be a number of patients with subclinical narrowing due to ablations becoming symptomatic only later in life. More recently adult data showed cerebral ischaemic micro-lesions after ablation of atrial fibrillation in one third of the patients using diffusion weighted magnet resonance imaging [9]. The clinical relevance is unclear and the lesions seem to disappear in most patients [10]. So far no similar investigations have been performed in children after RF ablation in the left atrium or ventricle.

What is the role of the new technologies?

Cryo-ablation

Radiofrequency ablation creates a lesion at the tip of the catheter by conversion of electromagnetic energy into heat (resistive heating). The RF energy is concentrated at the catheter – tissue contact area disperses through the body and exits to a large surface electrode. The effects of the RF energy on the tissue depend on the size of the tip, quality of the electrode-tissue contact, the duration of the application, blood supply and proximity of blood vessels as well as the degree of heat dissipation due to blood flow around the tip. Macroscopically RF-lesions are covered by a thin fibrin layer although lacerations of the surface with adherent coagulated or charred material might be seen and stick to the catheter (Fig. 1). Heat transfer continues even after discontinuation of RF current delivery and might result in unpredicted lesion expansion. Therefore it is very difficult to exactly predict lesion size and depth with RF energy, increasing the risk of unwanted effects including coronary artery injury as mentioned earlier. The ongoing heat transfer also explains the higher risk of ablation-induced atrioventricular block in right septal locations with RF ablation despite a very careful approach [11]. In small children one should therefore strictly limit the energy applied, create only the lesions absolutely needed and limit ablation time in a given lesion to 20–30 s [12].

Cryo-ablation uses liquid nitrogen to create temperatures of down to negative 80 °C at the tip, forming a