

Editorial

Current Situation of the Treatment of Arrhythmias in Children in Spain. Finding a Place of its Own

Estado actual del tratamiento de las arritmias en la edad pediátrica en España. Buscando su espacio

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Cardiac arrhythmias are relatively common in the pediatric age. The yearly estimated incidence of supraventricular tachycardia in persons younger than 19 years is 13 cases per 100 000 population.¹ Currently, the safety and effectiveness of percutaneous ablation treatment in this population is considered comparable to that of adults undergoing cardiac ablation, although the number reported is considerably smaller. Most literature on pediatric cardiac ablation is from multicenter registries in the United States,²⁻⁴ where these patients are treated in referral units with a large volume of cases, exceeding 100 per year. This factor has been associated with better clinical outcomes.⁵ Nonetheless, some recent reports from European countries have shown similar effectiveness and safety results, even though the number of cases treated is significantly smaller.^{6,7}

Regardless of the health care setting, ablation in pediatric patients, particularly those younger than 12 years, involves specific clinical characteristics, technical requisites, and management requirements. Hence, the professionals attending these patients should be well prepared to carry out the procedure with guaranteed safety and resolve any unforeseen complications that may arise.

TREATMENT INDICATIONS

The guidelines for the diagnosis and treatment of arrhythmias in pediatric patients and those with congenital heart disease (CHD) from AEPC/EHRA in 2013¹ and PACES/HRS in 2016⁸ provide specific information on the etiology, epidemiology, diagnosis, and treatment strategies for cardiac ablation in patients 0 to 18 years of age (Table).

Although these 2 guidelines are similar in general terms, there are some slight differences in the indications provided. For example, the class I indications for patients with Wolff-Parkinson-White syndrome: although both guidelines include as a class I indication the presence of this syndrome in patients with an

episode of resuscitated sudden cardiac death and in patients with syncope and an RR interval < 250 ms during atrial fibrillation (corresponding to a heart rate of 240 bpm) or an accessory conduction pathway effective refractory period < 250 ms, the American guidelines also include syncope with more than 1 accessory pathway as a risk factor. Furthermore, the guidelines differ regarding the treatment of paroxysmal supraventricular tachycardia (PSVT) without pre-excitation in patients with no heart disease. In these cases, the AEPC/EHRA guidelines only include PSVT associated with severe ventricular dysfunction as a class I indication. In the HRS/PACES guidelines, apart from ventricular dysfunction, a class I indication is also given to cases of recurrent or incessant PSVT in which medication is ineffective or poorly tolerated in patients weighing more than 15 kg, in patients with hemodynamic deterioration with syncope as a symptom associated with PSVT, and in cases with an electric cardioversion requirement for initial treatment in patients weighing more than 15 kg. Lastly, family preference is also included as a class I indication in patients with weight higher than 15 kg.

For the treatment of ventricular arrhythmias, the guidelines show no relevant differences. Both have a class I indication for patients with symptomatic ventricular tachycardia (VT) when drug therapy is ineffective in controlling the arrhythmia. The HRS/PACES guidelines also include a class I indication for intolerance to medication and for family preference as an alternative to medical therapy in patients weighing more than 15 kg.

Specific indications for arrhythmia ablation treatment in patients with CHD are only provided in the American guidelines. These include a class I indication for patients with PSVT related to specific CHDs, such as twin atrioventricular nodes or other CHD-related accessory pathways when medication is not effective or produces intolerable adverse effects. A class I indication is also assigned to ablation of multiple accessory pathways in patients with Ebstein anomaly, and to ablation of atrial arrhythmia outside the immediate postoperative period (3-6 months following surgery) when medical treatment is not effective and produces important adverse effects. Finally, a class I recommendation is included for VT in CHD patients with an implantable automatic defibrillator experiencing several episodes of tachycardia despite drug therapy and adequate device reprogramming, with the aim of avoiding activation of multiple shocks.

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Table

Comparison of the Class I Indications for Ablation Between the European and American guidelines

Patient group	AEPC/ESC 2013 class I recommendations	HRS/PACES 2016 class I recommendations
Indications for patients with WPWS	<ol style="list-style-type: none"> 1. Patients with WPWS and a resuscitated sudden death episode 2. Patients with WPWS and syncope with an RR interval < 250 ms during atrial fibrillation, or an accessory pathway effective refractory period < 250 ms 3. Patients with WPWS and palpitations with inducible and sustained PSVT on EPS 	<ol style="list-style-type: none"> 1. Patients with WPWS and a resuscitated sudden death episode 2. Patients with WPWS and syncope with an RR interval < 250 ms during atrial fibrillation, or an accessory pathway effective refractory period < 250 ms, or the presence of multiple pathways
Indications for PSVT	<ol style="list-style-type: none"> 1. PSVT associated with severe ventricular dysfunction 	<ol style="list-style-type: none"> 1. PSVT associated with severe ventricular dysfunction in patients > 15 kg 2. Recurrent or incessant PSVT when medication is not effective or poorly tolerated in patients > 15 kg 3. Documented recurrent or incessant PSVT when the family wishes to avoid chronic drug therapy in patients > 15 kg 4. Documented recurrent PSVT that poses an emergency for the patient or requires electric cardioversion in patients > 15 kg
Indications for VT	<ol style="list-style-type: none"> 1. Monomorphic VT with hemodynamic compromise treatable by catheter ablation 	<ol style="list-style-type: none"> 1. Focal VT or VE causing ventricular dysfunction when medication is not effective or causes intolerable adverse effects, as an alternative option in patients > 15 kg 2. PFVT, OTVT, or VT with syncope when medication is not effective or causes intolerable adverse effects, as an alternative option in patients > 15 kg
Indications for patients with CHD		<ol style="list-style-type: none"> 1. PSVT related to specific CHDs, such as twin atrioventricular nodes or other CHD-related accessory pathways when medication is not effective or causes intolerable adverse effects 2. A Wolff-Parkinson-White pattern and multiple risk pathways in patients with Ebstein anomaly weighing > 15 kg 3. Ablation of atrial arrhythmias outside the immediate postoperative period (3-6 months following surgery) if medical therapy is not effective and causes important adverse effects 4. VT in patients with CHD and an implantable automatic defibrillator with multiple VT episodes despite drug therapy and appropriate device reprogramming

AEPC/ESC, Association for European Paediatric and Congenital Cardiology/European Society of Cardiology; CHD, congenital heart disease; EPS, electrophysiology study; HRS/PACES, Heart Rhythm Society/Pediatric and Congenital Electrophysiology Society; OTVT, outflow tract ventricular tachycardia; PFVT, posterior fascicular ventricular tachycardia; PSVT, paroxysmal supraventricular tachycardia; VE, ventricular extrasystoles; VT, ventricular tachycardia; WPWS, Wolff-Parkinson-White syndrome

Both guidelines include a specific mention regarding cardiac ablation in lactating infants and children younger than 5 years, as there is sufficient evidence to consider the patient's weight as an independent risk factor for severe complications, including some reported deaths.⁹ Up to now, the AEPC/EHRA guidelines have defined these as patients weighing < 15 kg, but in the latest American guidelines this limit has been slightly modulated, with smaller patients being referred to as those weighing less than "approximately 15 kg". Despite this slight difference in the definition, there is consensus that professionals should be more restrictive with the indications in this age group, and attempt to optimize medical therapy by including several drug combinations to delay cardiac ablation. In addition, the following measures are recommended: 1) that these procedures should be carried out by electrophysiologists with experience in pediatric patients; 2) that ablation should be performed with a tailored strategy that minimizes the number of applications; and 3) that cryoablation should be used before radiofrequency in substrates carrying an elevated risk of atrioventricular block. Finally, special mention is made of the subgroup of smallest patients, weighing between 3 and 7 kg or younger than 6 months, in whom ablation should only be performed for life-threatening cardiac arrhythmia after failure of several combinations of antiarrhythmic drugs.

EFFECTIVENESS AND COMPLICATIONS OF PEDIATRIC CARDIAC ABLATION

Data on the effectiveness of ablation mainly come from retrospective and prospective registries in the United States,

which have shown a rise in the rate of effective ablations from 90.4% in the 1991 to 1996 period to 95.2% from 1996 to 1999.^{2,3} These results are very similar to those observed in a prospective registry including 2761 patients from 41 centers, reporting an overall effectiveness rate of 93%.⁴ To date, there are 2 population registries of pediatric ablation procedures in Europe, 1 in Finland⁶ and 1 in the Czech Republic,⁷ which show an overall final effectiveness rate similar to the values in the American registries, specifically 91% and 96%, respectively. Recent nonpopulation studies have reported very high effectiveness (> 98.5%) in both pediatric patients (< 12 years) and adolescents, although with a higher complication rate in the group younger than 12 years of age (5.4%) and a slightly higher recurrence rate (25.5% vs 17.6%).¹⁰ Furthermore, the effectiveness of ablation varied according to the substrate treated: highest in ablation of nodal reentrant tachycardia (effectiveness 95%-99%), followed by ablation of left lateral pathways (effectiveness > 95% in all series), and focal atrial tachycardias (effectiveness 93%). In contrast, the right lateral, right septal, and left septal pathways show lower effectiveness values ranging from 80% to 90%, similar to those obtained in VT ablation (effectiveness around 75%-80%).

Cryoablation use for the treatment of various substrates has been described in several studies in pediatric patients, with immediate effectiveness rates of 83% to 98% for nodal reentrant tachycardia, which is somewhat lower than the values obtained with radiofrequency (95%-100%). These procedures are also associated with a higher rate of recurrent arrhythmias than radiofrequency ablation: between 0% and 28%, depending on the series. These data concur with those reported in a recent meta-analysis comparing radiofrequency ablation and cryoablation for

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