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REVIEW

Ventricular arrhythmias and sudden death in tetralogy of Fallot

Arythmies ventriculaires et mort subite dans la tétralogie de Fallot

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Summary Malignant ventricular arrhythmias and sudden cardiac death may late happen in repaired tetralogy of Fallot, although probably less frequently than previously thought, especially with the advent of new surgical techniques/management. Ventricular tachycardias are caused by reentry around the surgical scars/patches and valves. Many predictive factors have been proposed, which suffer from poor accuracy. There is currently no recommended indication for prophylactic implantable cardioverter defibrillator implantation—except maybe in the case of multiple risk factors—while radiofrequncy ablation may be proposed in secondary prevention with or even without a back-up implantable cardioverter defibrillator in selected cases.

Abbreviations: ACHD, adult congenital heart disease; GUCH, grown-up congenital heart disease; ICD, implantable cardioverter defibrillator; RVOT, right ventricular outflow tract; SCD, sudden cardiac death; TOF, tetralogy of Fallot; VT, ventricular tachycardia.

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Repeated cardiological investigations and monitoring should be proposed for every operated patient.

MOTS CLÉS
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Mort subite ;
Arythmies ventriculaires ;
Cardiopathie congénitale de l'adulte ;
Ablation

Résumé Des arythmies ventriculaires malignes ou une mort subite peuvent survenir tardivement après correction chirurgicale de tétralogie de Fallot, quoique probablement moins fréquemment que précédemment supposé, surtout avec l'avènement de nouvelles techniques chirurgicales ou de prise en charge. Les tachycardies ventriculaires sont dues à des réentrées autour/entre les cicatrices de ventriculotomie ou patchs et les anneaux valvulaires. Un certain nombre de facteurs favorisants ont été proposés qui souffrent cependant de valeurs prédictives insuffisantes pour être utilisés seuls en pratique clinique. Il n'y a actuellement pas de recommandation pour l'implantation prophylactique de défibrillateur en prévention primaire — sauf peut-être en cas d'association de facteurs prédictifs multiples — alors que l'ablation percutanée par radiofréquence peut être proposée en prévention secondaire avec ou même sans défibrillateur dans certains cas sélectionnés. Des investigations cardiaques répétées et une surveillance au long cours doivent être effectués chez chaque patient opéré.

Background

Congenital heart disease is present in 0.9% of living births; currently, 90% of those affected will reach adulthood because of recent progress made in paediatrics, cardiology, surgery and resuscitation [1,2]. Among what are commonly called ''grown-up congenital heart diseases'' (GUCHs) or, more recently, ''adult congenital heart diseases'' (ACHDs), tetralogy of Fallot (TOF) has a preponderant place, because of its relatively high prevalence (7—10% of all congenital heart diseases; 1/3500 to 1/4300 in the adult population) [3,4], and because it is possible to have corrective surgery, leading to almost normal anatomy and physiology in adulthood. Indeed, very long-term follow-up has demonstrated that health status is excellent, with a mortality rate that is considered to be low (14% mortality for hospital survivors at 40-year follow-up, after surgery performed in the 1970s), even if it is still higher than in the general population, mainly because of heart failure and ventricular arrhythmias [5]. Thus, the third reason for making TOF one of the main GUCHs of concern is the late risk of malignant ventricular arrhythmias and sudden cardiac death (SCD).

The aim of this review is to highlight the mechanisms of ventricular arrhythmias in TOF, and to present current knowledge of secondary and primary prevention of SCD in this setting.

SCD in GUCH patients

The occurrence of SCD in patients with previous surgical repair of congenital heart defects is a tragic event, as many are usually considered to be ''cured'' of their congenital heart disease (even if this terminology may be a bit too optimistic), with rather low mortality rates and usually excellent quality of survival. Some of these SCDs are probably linked to paroxysmal high-degree atrioventricular block (e.g. in repaired TOF with relevant intraventricular conduction disturbances, but also in ventricular septal defect, cushion defect or congenitally corrected transposition of the great arteries). Some other SCDs are caused by ventricular fibrillation induced by fast ventricular rates during supraventricular tachycardia (e.g. atrial tachycardia with 1/1 atrioventricular conduction after atrial switch for transposition of the great arteries, or after Fontan procedures for single ventricle). Other causes probably include haemodynamic compromise, embolism, myocardial infarction or aneurism rupture, but it is now clear that most SCDs (a proportion estimated at around 75% [6]) are secondary to arrhythmias, and among these, malignant ventricular arrhythmias have been documented in 85% of cases at the time of the cardiac arrest [6].

Even if the culprit GUCH for SCD has changed in recent decades, TOF remains one of the main GUCHs carrying the risk of late SCD. In 1974, congenital aortic stenosis, Eisenmenger's syndrome, TOF and hypertrophic obstructive cardiomyopathy were responsible for more than half of SCDs in children (non-operated defects in most) [2]; whereas TOF, systemic to pulmonary anastomosis, pulmonary hypertension caused by left to right shunt and dilated cardiomyopathy were present in half of the SCDs in a report published 10 years later (postoperative in a significant number of cases) [7]. In 1998, Silka et al. found that in 3600 patients with GUCH, most late SCDs were linked to aortic stenosis, aortic coarctation, transposition of the great arteries and TOF, leading to a yearly SCD rate of 0.22% (50 to 200 times higher than in the general population); most were suspected to have an arrhythmic cause [6]. Similar causes of SCD have been found in more recent studies [8]. However, even if it is always a clinically relevant issue, SCD is not the major cause

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