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Review

The pathophysiologic aspects and clinical implications of electrocardiographic parameters of ventricular conduction delay in repaired tetralogy of Fallot

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Abstract The 12-lead surface electrocardiogram is a valuable and feasible clinical tool in the management of patients following tetralogy of Fallot (TOF) repair. The importance of QRS duration in TOF patients has long been acknowledged. A prolonged QRS complex has been associated with increased risk for subsequent life-threatening ventricular arrhythmia and sudden cardiac death. Our current ability to risk-stratify TOF patients for malignant arrhythmogenic events primarily on the basis of QRS duration is rather limited. Nevertheless, increasing evidence suggests that QRS morphology and duration may be useful as surrogate markers of infundibular and regional right ventricular myocardial disease. The aim of this review is to provide a critical appraisal of the clinical implications of established and new electrocardiographic markers of ventricular conduction delay in TOF patients following surgical correction with a particular focus on QRS duration, lengthening, and fragmentation. In addition, the pathophysiological background of these parameters is addressed.

Keywords: Tetralogy of Fallot; Sudden cardiac death; Ventricular arrhythmia; QRS duration; QRS fragmentation; Myocardial fibrosis

Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic heart defect, affecting nearly 10% of neonates with congenital heart disease [1]. Advances in surgical strategies and perioperative management have dramatically improved outcome, with more than 90% of patients reaching adulthood [1,2]. Nevertheless, long-term survival remains lower than in the general population. The difference in survival is commonly attributed to sudden cardiac death caused by sustained macroreentrant ventricular tachyarrhythmias commonly arising from the right ventricular outflow tract (RVOT), the region of the heart most exposed during cardiac surgery [2]. The overall incidence of sudden death has been estimated to be 1.2% at 10 years of follow-up [3]. Although the arrhythmogenic burden (ventricular and supraventricular arrhythmias) is substantial, affecting approximately 43% of TOF patients during long-term follow-up, heart failure and exercise intolerance also have an important impact on morbidity and mortality [4–6]. Therefore, identifying TOF patients at highest risk for major adverse events (sudden cardiac death, ventricular arrhythmias, heart failure) is a high priority research topic [4,5].

The 12-lead electrocardiogram (ECG) remains a valuable and feasible clinical tool in the management of patients following TOF repair. It has been used for risk stratification of future arrhythmogenic events, and for prognostic purposes such as identifying a possible subgroup of TOF patients in whom an invasive intervention such as insertion of an implantable cardioverter defibrillator or replacement of the pulmonary valve may improve overall outcome [4–7]. Although several electrocardiographic markers, including interlead QT dispersion, JT dispersion, QTc, JTc, and presence of left anterior hemiblock, have been investigated for their predictive value of clinical outcome in TOF patients, QRS duration only has evolved as a simple, reliable and highly reproducible parameter for risk stratification and prognostication [5,6,8–11]. A recent promising new electrocardiographic marker, although in its infancy, seems QRS fragmentation (fQRS) [12,13].

The interpretation of these electrocardiographic markers in clinical practice remains challenging. In this report we give a brief overview of the pathophysiologic aspects and clinical implications of QRS prolongation, lengthening and

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fragmentation in TOF patient management and clinical decision making.

Effects of corrective surgery on QRS morphology and duration

What exactly is the pathophysiologic background of QRS prolongation in repaired TOF patients? To answer this question we first have to describe the surgical procedure used to repair TOF and its effect on ventricular conduction.

TOF is characterized by obstruction of the right ventricular outflow tract (RVOT), at the subvalvular, valvular, and/or supravalvular level, secondary RV hypertrophy, a large ventricular septal defect, and an aorta that overrides both ventricles (Figs. 1 and 2) [1]. During corrective surgery, the ventricular septal defect is closed with a patch, and the RVOT stenosis is relieved by excising obstructing myocardial bundles, and, in many cases, by implanting a transannular patch after RVOT incision. Although the main goal of surgery is to completely relieve the RVOT obstruction, a less aggressive surgical strategy has been recently adopted, in which the use of a transannular patch and rigorous excision of muscle bundles in the RVOT is limited [5–7]. Studies have shown that a mild RVOT obstruction after corrective surgery might protect TOF patients, to a certain degree, against the detrimental adverse effects of long-standing pulmonary valve regurgitation (PVR), present in almost all TOF patients following repair [14]. However, long-term comparisons of development of adverse effects and patient outcome between different surgical techniques are still missing [6].

Widening of the QRS complex occurs in most TOF patients following surgical repair [5–9]. The His–Purkinje conduction system may be damaged at different levels along its ventricular course during cardiac surgery. The sites of damage have been extensively described elsewhere [15,16]. In brief, conduction disturbances may be located in the proximal right bundle branch (region of the ventricular septal defect), distally (level of the moderator band) or at the peripheral ramifications of the right bundle branch. A delay in conduction of normal electrical activity along the specialized Purkinje system, typically gives rise to a postoperative right bundle branch block (RBBB) pattern on the 12-lead surface ECG. With RBBB, the left ventricle is first activated across the fast conducting left bundle branches. Then, the electrical impulses are spread to the RV, thereby prolonging the QRS complex. The delayed electrical activation of the RV usually produces a late additional R wave (R') in the anterior leads (V1 and/or V2), and 'slurring' of the negative S waves in leads I and V6 (Fig. 3).

Depending on the location of the conduction disturbance along the RBB, important differences in the ventricular activation sequence can be appreciated [16]. A proximally located RBBB is associated with more pronounced conduction abnormalities than if a block is present in the terminal ramifications of the RBB. With this type of block, the entire RV is activated via the left bundle branch, resulting in significant delays among the septal, apical, RVOT, and RV free wall myocardial segments [15,16]. If the conduction delay is located to the terminal part of the RBB, then the activation delay is located only to the anterior segment of the

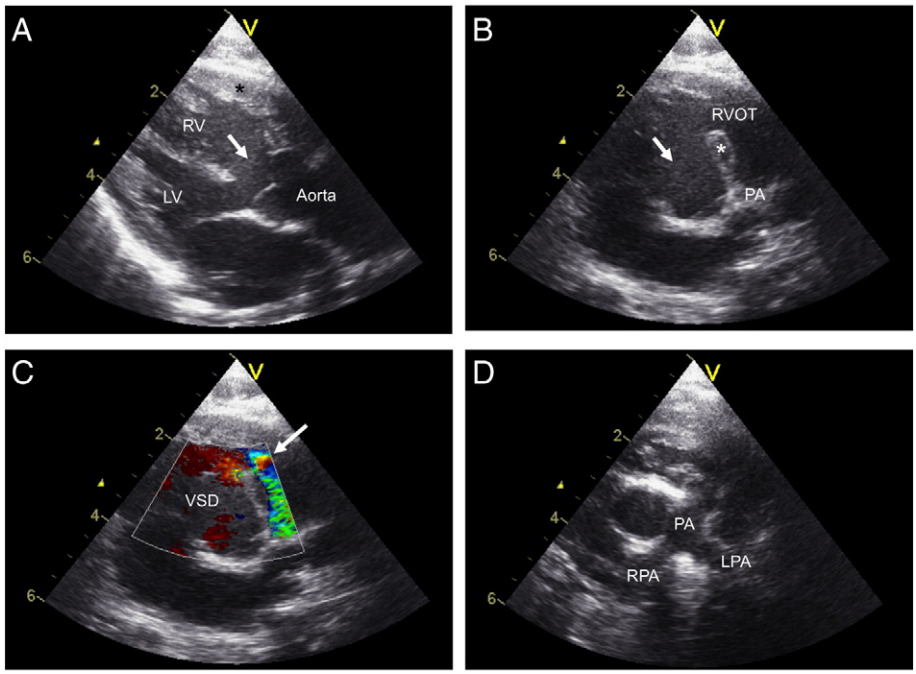

Fig. 1. Echocardiographic views of the morphological features of tetralogy of Fallot. (A) Parasternal long-axis view showing the overriding aorta and the ventricular septal defect (VSD, arrow). Right ventricular hypertrophy can also be appreciated in this image (*). (B) The anatomy of the right ventricular outflow tract (RVOT). The essence of TOF is an antero-superior deviation of the infundibular septum (*), giving rise to the VSD (arrow) and narrowing of the RVOT. (C) Parasternal short-axis view. Color Doppler image visualizing the subpulmonary obstruction (arrow). (D) View of the main pulmonary artery (PA) and its branches, showing mild hypoplasia of the pulmonary arteries. LPA = left pulmonary artery; LV = left ventricle; RPA = right pulmonary artery; RV = right ventricle.