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Non-sustained ventricular tachycardia in patients with congenital heart disease: An important sign?

Christophe P. Teuwen [a,1], Tanwier T.T.K. Ramdjan [a,1], Marco Götte [b], Bianca J.J.M. Brundel [c], Reinder Evertz [d], Joris W.J. Vriend [b], Sander G. Molhoek [e], H.G. Reinhart Dorman [f], Jurren M. van Opstal [f], Thelma C. Konings [g], Pepijn van der Voort [h], Etienne Delacretaz [i], Nienke J. Wolfhagen [a], Virgilla van Gastel [a], Peter de Klerk [a], Dominic A. Theuns [a], Maarten Witsenburg [a], Jolien W. Roos-Hesselink [a], John K. Triedman [j], Ad J.J.C. Bogers [k], Natasja M.S. de Groot [a,*]

[a] Dept of Cardiology, Erasmus University Medical Center, Rotterdam, The Netherlands
[b] Dept of Cardiology, Haga Hospital, The Hague, The Netherlands
[c] Dept of Clinical Pharmacy and Pharmacology, University Medical Center, University of Groningen, The Netherlands
[d] Dept of Cardiology, Radboud University Medical Center, Nijmegen, The Netherlands
[e] Dept of Cardiology, Amphia Hospital, Breda, The Netherlands
[f] Dept of Cardiology, Medisch Spectrum Twente, Enschede, The Netherlands
[g] Dept of Cardiology, VU University Medical Center, Amsterdam, The Netherlands
[h] Dept of Cardiology, Catharina Hospital, Eindhoven, The Netherlands
[i] Dept of Cardiology, Inselspital, University of Bern, Switzerland
[j] Dept of Cardiology, Boston Children's Hospital, Boston, USA
[k] Dept of Cardio-Thoracic Surgery, Erasmus University Medical Center, Rotterdam, The Netherlands

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ABSTRACT

Background: Sustained ventricular tachycardia (susVT) and ventricular fibrillation (VF) are observed in adult patients with congenital heart disease (CHD). These dysrhythmias may be preceded by non-sustained ventricular tachycardia (NSVT). The aims of this study are to examine the 1] time course of ventricular tachyarrhythmia (VTA) in a large cohort of patients with various CHDs and 2] the development of susVT/VF after NSVT.
Methods: In this retrospective study, patients with VTA on ECG, 24-hour Holter or ICD-printout or an out-of-hospital-cardiac arrest due to VF were included. In patients with an ICD, the number of shocks was studied.
Results: Patients (N = 145 patients, 59% male) initially presented with NSVT (N = 103), susVT (N = 25) or VF (N = 17) at a mean age of 40 ± 14 years. Prior to VTA, 58 patients had intraventricular conduction delay, 14 an impaired ventricular dysfunction and 3 had coronary artery disease. susVT/VF rarely occurred in patients with NSVT (N = 5). Fifty-two (36%) patients received an ICD; appropriate and inappropriate shocks, mainly due to supraventricular tachycardia (SVT), occurred in respectively 15 (29%) (NSVT: N = 1, susVT: N = 9, VF: N = 5) and 12 (23%) (NSVT: N = 4, susVT: N = 5, VF: N = 3) patients.
Conclusions: VTA in patients with CHD appear on average at the age of 40 years. susVT/VF rarely developed in patients with only NSVT, whereas recurrent episodes of susVT/VF frequently developed in patients initially presenting with susVT/VF. Hence, a wait-and-see treatment strategy in patients with NSVT and aggressive therapy of both episodes of VTA and SVT in patients with susVT/VF seems justified.

1. Introduction

Sustained ventricular tachycardia (susVT) and ventricular fibrillation (VF) are recognized as late complications in adult patients with congenital heart defects (CHD) [1]. The reported prevalence of these dysrhythmias is up to 30% and increases with an older age [2–5]. susVT and VF have mainly been reported in patients with tetralogy of Fallot (ToF) and transposition of the great arteries (TGA).

Various factors, such as surgical incisions, patches, ventricular volume and/or pressure overload contribute to development of susVT/VF [6]. It has been suggested that scarring of ventricular tissue after cardiac surgery in ToF patients might give rise to enhanced automaticity or re-entry with susVT as a consequence [7–10]. These dysrhythmias increase both morbidity and mortality in CHD patients and are associated with

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* Corresponding author at: Translational Electrophysiology, Department of Cardiology, Erasmus Medical Center, Thorax Center, PO Box 616, 's Gravendijkwal 230, 3015CE Rotterdam, The Netherlands.
E-mail address: n.m.s.degroot@erasmusmc.nl (N.M.S. de Groot).
[1] Both authors contributed equally to this work.

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sudden cardiac death (SCD) in ToF and TGA patients [11–13]. It is therefore of utmost importance to determine whether the development of these life-threatening dysrhythmias in patients with CHD can be predicted [6,11]. So far, prolongation of the QRS duration complex in both ToF (≥180 ms) and TGA (≥140 ms) patients has been identified as a sensitive predictor for development of ventricular tachycardia [2,5,14]. The prognostic value of non-sustained VT (NSVT) detected during ambulatory monitoring in CHD patients has mainly been investigated in ToF patients and remains debatable. NSVT predicted implantable cardioverter defibrillator (ICD) shocks in these patient groups [15,16], although other studies did not find a correlation between NSVT or (asymptomatic) ventricular runs and development of susVT or sudden cardiac death [2,17].

The purpose of this multicenter study is to examine 1) the time course of ventricular tachyarrhythmia (VTA) including NSVT, susVT and VF and 2) the occurrence of susVT or VF after earlier NSVT in a large cohort of patients with a variety of CHDs.

2. Methods

This retrospective study is part of the "DysrhythmiA in patients with congeNitAl heaRt diseAse" (DANARA) project (MEC-2012-482), which was approved by the local medical ethical committee of the Erasmus University Medical Center Rotterdam. According to Dutch law, informed consent was not required for this project.

2.1. Study population

Patients were included in this study if they presented either at the emergency room or at the outpatient clinic with a VTA in the following centers: Erasmus University Medical Center, Rotterdam; Amphia Hospital, Breda; Medisch Spectrum Twente, Enschede; VU Medical Center, Amsterdam; Haga Hospital, The Hague; Catharina Hospital, Eindhoven; and Cardiology, Inselspital, University of Bern, Switzerland. A documented dysrhythmia was identified on a surface electrocardiogram (ECG), 24-hour Holter recording or pacemaker/implantable cardioverter defibrillator printout. For this study, CHD patients with documented VTA episode before January 2014 were included. The follow-up period is defined as the time between the initial VTA until the last visit to the outpatient clinic in June 2014. Patients who received an ICD as part of secondary prevention after an out-of-hospital cardiac arrest were also included.

2.2. Clinical characteristics

After inclusion, subsequent clinical and demographic information prior to VTA was retrospectively collected for the purpose of this study. Clinical data consisted of type of CHD, number and time-interval of corrective/palliative surgical procedures, ablative therapy, indications for an ICD and death. Patients with aortic valve disease (AVD), atrial septal defect (ASD), atrioventricular septal defect, coarctation of the aorta, mitral valve insufficiency, patent ductus arteriosus, pulmonary stenosis and ventricular septal defect (VSD) were considered as having a complete repaired/simple CHD. The rest of the patients were classified as complex CHD. In case of an ICD implantation after the VTA, the number of delivered appropriate and inappropriate shocks was also documented. Echocardiogram obtained before the first VTA were used to determine left and/or right ventricular function and classified according to the guidelines [18]. Classification of the ventricular function was based on ejection fraction; an ejection fraction ≤35% was considered as impaired.

2.3. Analysis of rhythm registrations

Surface ECG, 24-hour Holter registrations and ICD printouts were examined in order to assess the occurrence of episodes of VTA including NSVT, susVT or VF. Non-sustained VT was defined as ≥3 consecutive ventricular beats with a frequency >100 beats per minute and a duration ≤30 s and not interrupted by anti-tachycardia pacing or delivery of an electrical shock. The last available ECG within a year prior to onset of VTA was selected to assess mean QRS duration. QRS duration of ≥120 ms was considered as prolonged; QRS duration was not measured in ventricular paced rhythm.

2.4. Statistical analysis

Continuous variables were expressed as mean ± SD or median (range) depending on the distribution. Categorical data were denoted by percentages. Patient groups were compared with conventional group descriptive statistics. The Mann–Whitney U, t-test, X^2 test or Fisher's exact test was used to evaluate statistical significance of characteristics and frequencies where appropriate. Missing data are described in the text and excluded in the calculations. Statistical analysis was performed with SPSS, version 21 (IBM, Armonk, New York). A p-value of <0.05 was considered statistically significant.

3. Results

3.1. Study population

The study population consisted of 145 CHD patients with ToF (N = 42), TGA (N = 19), univentricular heart (UVH, N = 18), aortic valve disease (N = 18), atrial septal defect (ASD, N = 14), coarctation of the aorta (N = 6), congenitally corrected TGA (ccTGA, N = 6), pulmonary stenosis (PS, N = 6), ventricular septal defect (N = 6), truncus arteriosus (N = 3), mitral valve disease (N = 2), patent ductus arteriosus (N = 2), atrioventricular septal defect (N = 1), left ventricular aneurysm (N = 1) and Ebstein anomaly (N = 1); characteristics of the various CHD groups are summarized in Table 1.

Ninety-two percent of the patients (N = 134) underwent corrective/palliative cardiac surgery prior to onset of the VTA at a mean age of 12 ± 16 years. Mean age of the first surgical procedure differed among patients with a complex defect (median 2 years; range 0–55) compared to patients with a simple defect (median 21 years; range 0–70) ($p < 0.01$).

Only 11 patients (8%) did not have a history of corrective/palliative cardiac surgery at the time of the first presentation with a VTA (AVD N = 2; ASD N = 1; ccTGA N = 3; Ebstein anomaly N = 1, mitral valve disease N = 2 and VSD N = 2); 3 of them underwent cardiac surgery after revelation of the first VTA.

3.2. Presentation of ventricular tachyarrhythmia

Patients presented with either NSVT (N = 103, 71%), susVT (N = 25, 17%) or VF (N = 17, 12%); Fig. 1 illustrates the age at first presentation of the VTA for every CHD separately. The first episode of VTA occurred at a mean age of 40 ± 14 years (15–70); age of development of NSVT (40 ± 14 years), susVT (36 ± 13 years) and VF (44 ± 16 years) were comparable ($p > 0.05$). Clinical data regarding coronary artery disease was available in 114 patients; only 3 patients presenting with either NSVT (N = 2; ccTGA and CoA) or VF (N = 1; ToF) had undergone percutaneous coronary intervention (N = 2) or coronary artery bypass surgery (N = 1) for obstructive coronary artery disease. Information on ventricular function and QRS duration is summarized in Table 2.

In patients with UVH (N = 11, 30 ± 12 years) and complex CHD (N = 48, 38 ± 13 years) NSVT developed at a relative young age compared to patients with a simple CHD (N = 44, 45 ± 13 years; $p = 0.001$ and $p = 0.017$).

VTAs (susVT and VF) occurred at a relative young age in patients with UVH (N = 7, 26 ± 8 years) and complex CHD (N = 24, 36 ± 11 years), whereas patients with simple CHD appeared to be older at