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Sustained Ventricular Tachycardia in Children after Repair of Congenital Heart Disease

To investigate an association between surface electrocardiographic (ECG) parameters and sustained ventricular tachycardia (VT) in children after repair of congenital heart disease (CHD), data were obtained and analyzed in three groups (group I, 7 postoperative patients with episode of sustained VT (4 tetralogy of Fallot (TOF), 2 double outlet right ventricle (DORV), 1 truncus arteriosus); group II, 14 children with postoperative TOF not associated with VT; group III, 14 normal children). Mean age at the onset of sustained VT was 129±77 months (range 60-232); mean age at corrective surgery, 44±33 months (range 10-102); mean follow-up period after surgery, 84±74 months (range 20-185); the duration from repair to the onset of sustained VT, range 1-185 months. Compared to group II and III, group I showed longer QRS duration (group I, 137±10 msec; group II, 114±22 msec; group III, 65±12 msec) and shorter corrected J to T_{max} interval (group I, 209±24 msec; group II, 272±44 msec; group III, 249±18 msec). QT and corrected QT, J to T_{max} interval, and their dispersions in group I and II are significantly different from those of group III. In conclusion, QRS duration and corrected J to T_{max} interval could be helpful to predict ventricular tachycardia in postoperative CHD.

Key Words: *Tachycardia, Ventricular; Heart Defects, Congenital*

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INTRODUCTION

After repair of congenital heart disease (CHD), sudden death is a major concern. The terminal event causing sudden death is presumably ventricular arrhythmia in postoperative patient with tetralogy of Fallot (TOF) (1).

Potential risk factors for ventricular arrhythmia include older age at intracardiac repair, longer interval after surgery, increased right ventricular systolic pressure, moderate or severe pulmonary regurgitation, and ventricular dysfunction (1-4). An increasing amount of evidence suggests that ventricular arrhythmia originating from the right ventricular outflow area is a cause of late sudden death in patients after repair of TOF (5, 6).

QRS prolongation on the electrocardiogram (ECG) caused by bundle branch block and ventricular outflow damage may provide the ventricular inhomogeneity as a mechanism of ventricular arrhythmia. It has recently been suggested that QRS prolongation with QRS duration ≥180 msec could predict ventricular arrhythmia and sudden death with 100% sensitivity and 94.7% specificity (7). It is also reported that prolonged QRS duration on the ECG could be associated with induced sustained monomorphic ventricular tachycardia (VT) on electrophysiologic study (8). The study of Daliento et al. (9) showed QT dispersion (QTd) and end-diastolic volume of right ventricle (EDVRV) could be predictive indices in the stratification of patients at risk of life threatening arrhythmia.

The aim of this study was to describe our experience of sustained VT in children after repair of CHD, and to investigate the association between the 12-leads surface ECG parameters and VT especially from the standpoint of ventricular inhomogeneity.

PATIENTS AND METHODS

At the Department of Pediatrics of Seoul National University Children's Hospital, seven patients for this study were identified by the presence of spontaneous, sustained VT during the follow-up period after repair of CHD.

Patients (group I) presented with either 1) sustained symptomatic VT documented by a 12-lead ECG, requiring either medical therapy or electrical cardioversion

to terminate the arrhythmia or 2) palpitations usually associated with syncope or near syncope. Sustained VT is defined as persistent VT longer than 30 sec in duration. Patients with recent myocardial infarction, the long QT syndrome, hypertrophic cardiomyopathy or arrhythmias due to transient or reversible disorder were excluded. CHD were confined to whom had a pattern of right bundle branch blocks in surface ECG with or without right ventriculotomy. Seven patients satisfied above inclusion criteria.

Fourteen postoperative TOF children in good hemodynamic state and without ventricular arrhythmia were selected as an another control (group II). All patients were age- and sex-matched. Total correction was performed with the right ventriculotomy in all TOF control patients. Normal control group matched for age and sex consisted of 14 healthy children without cardiovascular problems and disease affecting surface ECG parameters (group III).

The ECG recordings were taken with a paper speed of 50 mm/sec at normal filtering. Several parameters of ECG were measured manually. QRS duration was defined as the maximum QRS duration in any lead from the first to the final sharp vector crossing the isoelectric line. QT interval was measured from the lead II using calipers. QT interval was defined as the interval between the beginning of QRS complex and the end of T wave. The onset and offset of T wave were defined as the intersections of the isoelectric line and the tangent of the maximal slope on the up and down limbs of T wave, respectively. Care was taken to avoid U waves in any measurement, and when U waves were present, the end of T wave was taken as the nadir between T and U waves. Three consecutive cycles were measured in each of the standard 12 leads, and a mean value was calculated from the three values. The JT interval was then calculated by subtracting QRS form QT in individual leads. The J to T max interval (JTM) was measured from J point to the maximum of T wave. The T max to T end interval (TME) was measured from the maximum of T wave to the end of T wave. Bazett's formula was used to obtain corrected QT/ JT/ JTM/ TME intervals and represented as QTc/ JTc/ JTMc/ TMEc, respectively. The QT/ JT/ JTM/ TME dispersions were defined as the difference between the maximum and minimum of QT/ JT/ JTM/ TME intervals occurring in any of the 12 ECG leads and represented as QTd/ JTd/ JTMd/ TMEd, respectively.

The data are expressed as means plus standard deviations (SD). The data were analyzed using Kruskall-Wallis test. A value of $p<0.05$ was considered as statistically significant.

RESULTS

Patients profile

A total of seven children presented with sustained VT after repair of CHD. Mean age at the onset of sustained VT was 129±77 months (range 60-232). Mean age at corrective surgery was 44±33 months (range 10-102). Mean follow-up period after surgery was 84±74 months

Table 1. Clinical data of patients with sustained ventricular tachycardia after repair of congenital heart disease

Case	Sex	Diagnosis	Op age (month)	VT onset (month)	Treatment for VT	Studies	F/U duration (month)
1	M	TOF	10	29	(-)	Reoperation for severe PR and LPA stenosis	26
2	F	DORV, PDA	18	185	(-)	(-)	169
3	M	Truncus arteriosus	22	60	Flecainide → (-)	Significant TR, AR	87
4	M	TOF	43	93	Beta blocker → Sotalol → Mexiletine	Reoperation for severe PRCryoablation	81
5	M	TOF	46	232	(-)	VT on treadmill test	186
6	M	TOF	71	200	Mexiletine → beta blocker → (-)	Tilt test with Isoproterenol and exercise test induced VT	182
7	M	DORV, remote VSD	102	103	Beta blocker	Not induced in exercise test and isoproterenol	21
Mean±SD			44±33	129±77			108±71

Op, operation; VT, ventricular tachycardia; F/U, follow-up; TOF, tetralogy of Fallot; PR, pulmonary regurgitation; LPA, left pulmonary artery; DORV, double outlet right ventricle; PDA, patent ductus arteriosus; BB, beta-blocker; TR, tricuspid regurgitation; AR, aortic regurgitation; VSD, ventricular septal defect; SD, standard deviation

Table 2. Echocardiographic findings of patients with sustained ventricular tachycardia after repair of congenital heart disease

Case	TR	PR	AR	MR	RV dimension	Overall ventricular function
1	Mild	Severe	Mild	No	Severely dilated	Normal (FS 32%)
2	Mild	Minimal	No	No	Normal	Ventricular function improved to normal range during follow-up tolerable ventricular function
3	Mild	Moderate	Mild to moderate	No	Moderately dilated	Mildly to moderately dilated LV
4	Mild	Moderate to severe	No	No	Moderately dilated	Normal
5	Mild	Mild	No	No	Normal	Normal
6	Minimal	Mild	No	No	Normal	Normal
7	Mild	Mild	No	No	Mildly dilated	Normal

TR, tricuspid regurgitation; PR, pulmonary regurgitation; AR, aortic regurgitation; MR, mitral regurgitation; RV, right ventricle; FS, fractional shortening; LV, left ventricle

Table 3. Comparison of electrocardiographic parameters among three groups

Parameters	Sustained VT (Group I)	TOF control (Group II)	Normal control (Group III)	*p* value
QRS (msec)	137±10*†	114±22†	65±12	$p<0.05$
QT (msec)	475±73†	431±48†	371±33	$p<0.05$
QTc (msec)	547±68†	531±57†	433±22	$p<0.05$
JT (msec)	338±66	317±32	306±31	NS
JTc (msec)	388±58	386±34	376±63	NS
JTM (msec)	182±27†	179±31†	214±23	$p<0.05$
JTMc (msec)	209±24*†	272±44	249±18	$p<0.05$
TME (msec)	156±71	139±35	158±16	NS
TMEc (msec)	179±72	222±52	171±70	NS
QTd (msec)	95±63†	67±33†	39±17	$p<0.05$
JTd (msec)	92±66†	67±33†	33±15	$p<0.05$
JTMd (msec)	25±13†	29±21†	10±6	$p<0.05$
TMEd (msec)	80±75†	60±34†	32±9	$p<0.05$

Values are mean±standard deviation.
QTc, corrected QT; QTd, QT dispersion; JTc, corrected JT; JTd, JT dispersion; JTM, J to maximum T; JTMc, corrected JTM; JTMd, JTM dispersion; TME, maximum T to the end of T wave; TMEc, corrected TME; TMEd, TME dispersion; msec, millisecond; NS, not significant; *$p<0.05$ vs. postoperative TOF control; †$p<0.05$ vs. normal control

(range 20-185). The duration from the cardiac repair to the onset of VT varied with the range of one month to 15 yr and 5 months. All patients were in New York Heart Association functional class 1 (Table 1).

The seven children with congenital heart disease were diagnosed as: four with TOF; one with double outlet right ventricle (DORV) and patent ductus arteriosus (PDA); one with double outlet right ventricle, persistent left superior vena cava, unroofed coronary sinus and remote ventricular septal defect (VSD); one with truncus arteriosus type 1, ventricular septal defect, partial anomalous venous return of right upper pulmonary vein into superior vena cava.

In three patient with TOF, right infundibulectomy was undertaken with transannular patch and widening of right ventricular outflow tract (case 1, 5, and 6). One with TOF had taken REV procedure (case 4). In a patient with DORV and remote VSD, VSD extension, intraventricular baffling, and widening of hypertrophied right ventricular outflow tract was performed (case 7).

After total correction of TOF, hemodynamic problems that were caused by a residual lesion or a newly developed lesion made it necessary to perform re-operation in three patients (case 1, 3, and 4). All three patients had pulmonary regurgitation due to the failure of own valve or inserted valve in pulmonary pathway. In two patients (case 1, 4), after suffering from sustained VT, pulmonary homograft interposition for severe pulmonary regurgitation was performed to prevent right ventricular volume overload, which might precipitate ventricular arrhythmia. Aortic regurgitation was significant in postoperative truncus arteriosus (case 3), but overall ventricular function

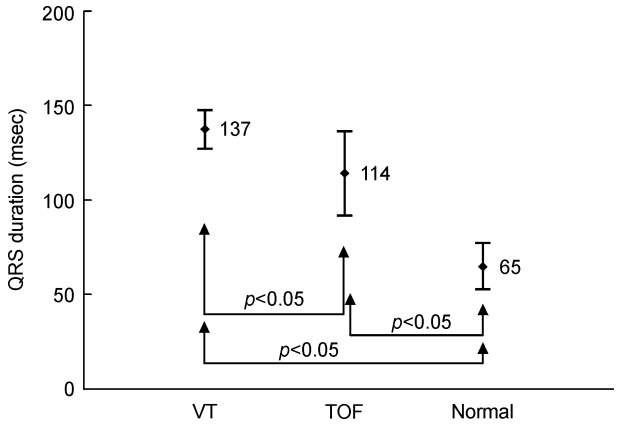

Fig. 1. Comparison of QRS duration among three groups (group I, ventricular tachycardia (VT); group II, postoperative tetralogy of Fallot (TOF) without ventricular arrhythmia; group III, normal control). QRS duration of group I is significantly prolonged compared to the others ($p<0.05$).

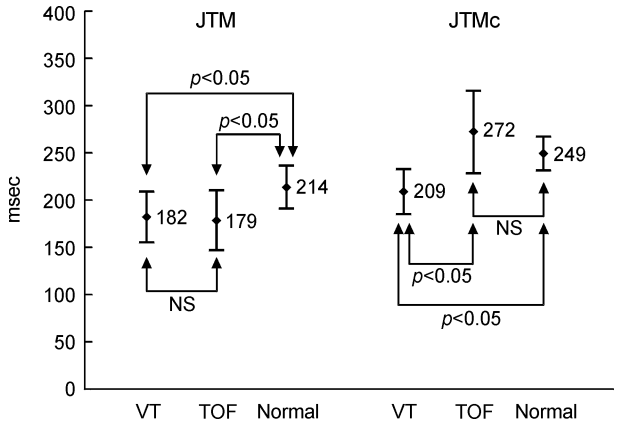

Fig. 2. Comparison of J to T maximum (JTM) and corrected JTM (JTMc) duration among three groups (group I, ventricular tachycardia (VT); group II, postoperative tetralogy of Fallot (TOF) without ventricular arrhythmia; group III, normal control). While JTM interval is not significantly different between group I and II, JTMc interval in group I is significantly shorter than that of group II and III. NS, not significant.

was good without hemodynamic compromise.

VT occurred in various situations. Exercise played a role of triggering VT in three patients (case 4, 5, and 6). Voiding and wake-up in the morning were the triggering factors in one case (case 7). The others had no associated particular situations. Antiarrhythmic agents were not prescribed in two cases (case 2, 5), that have showed no more symptomatic attacks until now. In the rest, several medications were used to control ventricular arrhythmia. Digoxin, beta-blocker such as Atenolol and Nadolol, class Ic antiarrhythmic drugs (Flecainide), class Ib drugs (Mexiletine), and class III drugs (Sotalol) were given.

Electrocardiographic parameters

In group I who presented with sustained VT, QRS duration were prolonged significantly compared to those in group II and III (group I, 137±10 msec; group II, 114±22 msec; group III, 65±12 msec) ($p<0.05$) (Table 3) (Fig. 1). QT and corrected QT, J to T maximum (JTM) interval, and dispersions of QT, JT, JTM, TME in group I and II (VT & TOF control) were significantly different from those of group III (normal control) ($p<0.05$). While JTM interval was not significantly different between group I and II, corrected JTM interval in sustained VT (group I) was significantly shorter than that of TOF and normal control (group II and III) (Fig. 2).

DISCUSSION

It appears that ventricular arrhythmias are present after cardiac surgery and that a number of factors are associated with these arrhythmias that might lead to sudden death. The electrical instability of postoperative CHD may result from anatomical modifications caused by surgery or mechanical events such as ventricular dilatation or stretch (7).

The sustained VT in postoperative CHD, especially in TOF, results from reentry, which requires areas of slow conduction circuit. When intracardiac electrophysiologic studies are performed on postoperative TOF patients, the induced VT is mapped to areas adjacent to surgical scar (6). The ventriculotomy scar, ventricular septal defect, and outflow patch have each been implicated as areas of potential block within the reentry circuit (5, 6). An animal study reported that the scar length grew by approximately 200% in the newborn and young puppies, whereas in the adult dogs the scar shrank by 20% (10). These investigators speculated that a critical mass of scar tissue provided the substrate for ventricular arrhythmias. The fate of operation scar in ventricle has not been fully elucidated in children with CHD. The concern about ventricular arrhythmia makes surgeon leave as small scars as possible on open-heart surgery, but we cannot abolish those problems. In unavoidable cases, we should be alert to the occurrence of ventricular arrhythmia, of which the malignant form needs prompt identification and treatment with appropriate drugs. Therefore, the guideline is necessary for the detection of malignant ventricular arrhythmia in children with postoperative CHD.

The cause of QRS prolongation after TOF repair is unclear. Right bundle branch block was present in all patients and we were unable to distinguish QRS prolongation secondary to postoperative damage of the con-

ducting tissue from QRS prolongation due to right ventricular dilatation. It is suggested that QRS prolongation be related to right ventricular dilatation from long-standing pulmonary incompetence (7). The combination of QRS ≥180 msec plus QTd >60 msec or QRSd >35 msec, or JTd >60 msec has been reported to be highly sensitive and specific for identification of patients with VT (98.3% of sensitivity and 100% of specificity) (11). In this study, the mean age of the 10 patients with VT was 30.5±6.8 yr. In another study whose mean age was 12±6 yr, the combination of QRS ≥170 msec and JTd ≥80 msec was reported to have a good positive predictive value (100%), negative predictive value (89%), and specificity (100%), but have a low sensitivity (21%) (12). The prolongation of QRS, an ECG reflection of abnormal and delayed depolarization, may play a role in the creation of a reentry circuit for VT in these patients.

In our study, the mean age at repair and follow-up duration were shorter compared to those in the previous reports of Gatzoulis et al. (7) and Berul et al. (12). In children with postoperative CHD, repolarization and depolarization parameters on surface ECG were supposed to be different from normal control. However, the clinical implication of those parameters in malignant ventricular arrhythmia has not been fully elucidated. Prolongation of QRS duration was expected in both postoperative groups (VT and TOF control). Ventricular dilatation was noted in 4 cases (case 1, 2, 3, and 4). Ventricular dilatation and dysfunction was thought to be associated with valvular insufficiency as well as myocardial damage. The prolonged QRS duration is thought to be partly due to right bundle branch block after ventriculotomy or due to ventricular dilatation caused by valvular insufficiency (pulmonary regurgitation). Although QRS duration (137 ±10 msec) in patients group was prolonged compared to those in TOF and normal control, the duration is far less than those of 170-180 msec in previous reports. Therefore, we think that QRS duration over 180 msec, could not be a good predictor of life threatening ventricular arrhythmias in children with short follow-up duration. Daliento et al. (9) reported that all six patients with sustained VT or ventricular fibrillation had QRS duration of less than 180 msec. In that study, the mean age at repair and follow-up duration was 8.7 yr and 13.3 yr, respectively. Thus the prolongation of QRS duration should be assessed in association with the patient's age and follow-up duration.

The measurement of QRS duration is partially redundant compared with the information provided by QTd (9). QTd was unrelated to the presence of right bundle branch block or to the functional conditions of the right and left ventricles, particularly ventricular dilatation. QTd proved to be a better screening method than QRS duration or QRSd because QTd makes it possible to discriminate different probabilities for nonsustained VT as opposed to sustained VT (9). T peak to T end interval, total T wave area and late T wave area have been reported as new indices in the ECG assessment of dispersion of repolarization (13).

This study has a limitation in interpretation due to the small number of patients. QT, QTc, QTd, JTd, JTM, JTMc, JTMd, and TMEd in normal control are significantly different from those of the other two postoperative groups. However, all repolarization parameters except JTMc are not different between VT group and TOF control. The QT duration and QRS duration were significantly prolonged in both groups of sustained VT and TOF control compared to normal control but JT interval was not different among three groups. In sustained VT group, QRS duration was significantly prolonged compared to TOF control but QT and QTc intervals were not significantly different. So the main cause of QT prolongation is thought to be a prolongation of ventricular depolarization. Our data showed that the change of ventricular depolarization as well as ventricular repolarization inhomogeneity could be involved in the development of ventricular electrical instability for malignant arrhythmia. Ventricular dilatation may lead to the induction of VT even in postoperative children with short QRS duration.

The dispersions of ventricular repolarization such as QTd, JTd, JTMd, and TMEd were prolonged in both sustained VT group and TOF control compared to normal control. These findings suggest that the open-heart surgery with right ventricular procedure increase ventricular inhomogeneity throughout the repolarization period. While the early portion of ventricular repolarization represented as JTM interval shortened, total and late repolarization in both postoperative groups (VT and TOF) showed prolonged duration and increased dispersions than those of normal control. The total changes of ventricular inhomogeneity on surface ECG are thought as an electrical summation of the increment of late repolarization period (Tmax to Tend) and the decrement of early repolarization period (J to Tmax). Unlike previous report of Zabel et al. (13) the late repolarization period (Tmax to Tend) did not show statistically significant differences among three groups. However the early repolarization period corrected by heart rate showed significant shortening in VT group compared to TOF control group. This suggests that the early portion of ventricular repolarization tend to be affected profoundly after ventricular injury and play an important role in ventricular arrhythmogenesis. So the early portion of ventricular repolarization could be used as an another predictive index to malignant ventricular arrhythmia in children with postoperative CHD.

The interval from the repair to documentation of VT was so variable that we could not easily predict the risk of ventricular arrhythmia based on follow-up duration. Frequent and regular checks for the risk of life-threatening arrhythmia should be introduced earlier in postoperative state of CHD patients.

In conclusion, QRS duration and JTMc interval may be helpful to predict the occurrence of malignant ventricular arrhythmia in children with postoperative CHD. Also ventricular dilatation and dysfunction may be an important factor. In addition, it should be reminded that even though QRS duration is not so much prolonged over 180 msec in these patients, malignant ventricular arrhythmia could occur.

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