873 Pediatric Electrophysiology

Wednesday, April 1, 1998, 8:30 a.m.–10:00 a.m.
Georgia World Congress Center, Room 365W

8:30

873-1 Clinical Arrhythmia and Sudden Death in Repaired Tetralogy of Fallot: A Multi-Centre Study

M.A. Gatzoulis, S. Balaji, C. Poile, S.A. Webber, J.S. Hokanson, S.C. Siu, T. Shinohara, J.H. Moller, M. Nakazawa, P.C. Gillette, G.D. Webb, A.N. Redington. *Coordinating Centre: Royal Brompton Hospital, London, UK*

Background: It has been suggested that QRS prolongation and QT dispersion (d) on the ECG predict sustained ventricular tachycardia (VT) and sudden death (SD) late after repair (r) of tetralogy of Fallot (TOF). In this study, we examined these markers of clinical arrhythmia and SD in a larger cohort of rTOF.

Methods: We reviewed the available 10-year records (1985–95) of all patients, who were alive in 1985 following previous rTOF.

Results: 737 rTOF patients (median 19, range 1.5–72 years in 1985) entered the study, allowing for over 7,000 patient-years follow-up.

	VT (n = 25)	SD (n = 14)	AF (n = 18)	Remainder
rAge	8.9 ± 7[#§] yrs	16.6 ± 15[*]	18.9 ± 12[*]	6 ± 7
FU	22.9 ± 7 yrs	23 ± 8	21.8 ± 7	24 ± 7 in 1995
QRS	196 ± 20[#§] ms	179 ± 24[*]	168 ± 20[*]	147 ± 24
QTd	97 ± 24[#§] ms	78 ± 28[*]	60 ± 26	54 ± 2
QRSp	5 ± 2[#§] ms/yr	3.5 ± 1.6[*]	2.9 ± 1[*]	1.4 ± 0.8

AF: atrial flutter, QRSp: QRS prolongation/year during the study period p < 0.01–0.0001 vs second (#), third (*) and last (§) column

4 cases with AF developed VT. Only 1 patient with previous VT died suddenly. A QRS ≥180 ms retained its sensitivity (VT-88% and SD-67%) and specificity (for both-84%).

Conclusion: While the extent and rate of QRS prolongation after rTOF clearly relate to clinical arrhythmia, the relationship between the development of VT and SD remains unresolved.

8:45

873-2 Very Long Term Follow-up (22–42 Years) After Correction of Tetralogy of Fallot: Transient Complete Heart Block Is Associated With Increased Risk of Sudden Death

J.S. Hokanson, J.H. Moller. *University of Minnesota, Minneapolis, MN, USA*

Background: Despite encouraging long term survival rates, sudden death continues to be a concern in patients who have undergone a corrective operation for Tetralogy of Fallot (TOF).

Methods: We evaluated the very long term outcome of all individuals who were discharged after surgical correction for TOF at the University of Minnesota, the first institution to perform this procedure, between 1954 and 1974.

Results: Of the 288 individuals discharged, current follow-up was available in 279 (96.9%). Surveys were received from 163/221 survivors (73.7%). A total of 7982 patient years of follow-up were evaluated. *Mortality:* There have been 58 late deaths, with a 40 year survival rate of 75.8%. Long term survival was less favorable in those patients who were older at the time of operation (P = 0.008) and when transient complete heart block was observed prior to discharge (P = 0.009). *Sudden Death:* 26 of the late deaths were sudden (44.8%). Of the variables evaluated, only the presence of transient complete heart block was related to the incidence of sudden death (P = 0.030). *Morbidity:* 95.1% of survey responders were in NYHA class I or II. The use of a transannular patch correlated with an increase in reoperations (P = 0.002) and a decline in NYHA class (P = 0.026).

Conclusions: Late sudden death after corrective operation for TOF occurs in a significant number of individuals and is correlated to transient complete heart block at the time of operation.

9:00

873-3 Repaired Tetralogy of Fallot: ECG Predictors of Death and Ventricular Tachycardia

M.S. Larson, C.A. Warnes. *Mayo Medical Center, Rochester MN, USA*

Background: QRS duration on resting ECG of ≥180 msec has been reported to be a sensitive predictor of life threatening ventricular tachycardia (VT) in patients (pts) with repaired Tetralogy of Fallot (TOF).

Methods: Predictors of death and/or malignant VT (VT and history of syncope or sustained VT) were analyzed using two-tailed Fisher's exact test.

Results: Between 1987–97, 100 (50 F, 50 M) pts with repaired TOF were seen. Mean age at repair was 16.8 ± 16 years. The mean follow-up after repair was 25.5 ± 10.3 years. Twelve pts (12%) had paced rhythm. Of the remaining 88 pts, 18 (20%) had a QRS ≥180 msec.

Six pts died during follow-up, 2 of whom had a history of VT. Age at death was 21–65 years (mean 44.0 ± 17.1 years). Five deaths were sudden. Of the 6 pts who died, 4 had paced rhythm. In the 2 pts without paced rhythm, 1 had QRS ≥180 (208 msec).

The mean age of the 94 survivors is 41.9 ± 12.3 years. Of these survivors, 6 pts had VT, 8 pts had paced rhythm, and 17 pts without paced rhythm had QRS ≥180 msec. In the 6 survivors with VT, 2 had paced rhythm and 1 without paced rhythm had QRS ≥180 msec. The presence of a paced rhythm was significantly associated with death (p = 0.0016). In the 88 pts without paced rhythm, QRS ≥180 msec was not associated with death (p = 0.3691), nor with malignant VT (p = 0.2686).

Conclusions: In these older pts with repaired TOF, QRS ≥180 msec did not predict VT or death, and risk stratification remains a challenge. The prevalence of death is relatively low (6%) despite the late age at repair. The presence of paced rhythm was significantly associated with death.

9:15

873-4 Usefulness of Invasive Cardiac Studies in Children With Monomorphic Ventricular Tachycardia and Normal Echocardiogram

P. Lachakunakorn, S. Balaji, H.B. Wiles, P.C. Gillette. *Medical University of South Carolina, Charleston, SC, USA*

The benefits of invasive cardiac studies in diagnosis and management of patients with ventricular tachycardia (VT) and normal echocardiogram is unknown. Between August 1988 and January 1996, 51 consecutive patients aged 1 to 21 years old (median 13) with VT underwent invasive cardiac study in the form of electrophysiologic study (EP) (n = 51), right ventricular (RV) angiogram (n = 45), and RV biopsy (n = 41). Symptoms were present in 21 (41%), including chest pain (10%), palpitation (24%), syncope or near syncope (10%), and cardiac arrest (4%).

There was no correlation between EP study outcome and clinical presentation. The RV angiogram showed a diverticulum in 7%. RV biopsy was abnormal in 17% (myocarditis 15% and cardiomyopathy 2%). None had fatty infiltration suggesting arrhythmogenic RV dysplasia. EP studies showed inducible sustained VT in 8 cases (16%). Based on the invasive studies, management was initiated in the form of drug therapy in 24 (47%), radiofrequency ablation in 9 (18%), surgical ablation in 1 (2%) and automatic implantable cardioverter-defibrillator (AICD) implantation in 1 (2%).

Invasive cardiac studies revealed abnormalities and influenced management in a significant proportion and are therefore recommended in patients with VT and normal echocardiogram.

9:30

873-5 Early Repair of Tetralogy of Fallot Is Associated With a Higher Incidence of Postoperative Junctional Ectopic Tachycardia

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Background: Postoperative junctional ectopic tachycardia (JET) is a known complication after surgery for congenital heart disease. The purpose of this study was to examine the relationship between age at the time of primary tetralogy of Fallot (TOF) repair and the development of postoperative JET.

Methods: Hospital records of 56 patients who underwent TOF repair from July 1995 to March 1997 at UCSF were reviewed. Diagnosis of JET was confirmed by noting narrow complex tachycardia and evidence of atrioventricular dissociation.

Results: Patients with JET had a lower mean age (69, range 2–305 days) as compared to patients without JET (396, range 1–3785 days). Using the nonparametric Mann-Whitney test, the ages of patients with and without JET were statistically different (P = 0.022). Patients under 4 months (N = 33) had 7.5-fold higher incidence of JET (30%) as compared to those older than 4 months (N = 23) (P = 0.019). Among patients with room air oxygen saturations ≥88%, who were electively scheduled for TOF repair (N = 47), there was a 4-fold higher incidence of JET (36%) in patients under 4 months as compared to those older than 4 months (9%), although this did not achieve statistical significance. JET onset was in the immediate postoperative period, lasting 1–3 days. One patient 42 days of age required cardioversion for hemodynamic instability associated with JET. Patients with JET did not have a longer ICU or hospital stay.

Conclusions: Early repair of TOF is associated with an increased incidence of JET, which is at least 30%. These results should be considered when determining the timing of elective TOF repair.