1095-201 QRS Duration and Depolarization Heterogeneity in Tetralogy of Fallot Patients Before and After Surgical Repair of the Pulmonary Valve

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Background. QRS duration has a strong predictive value in Tetralogy of Fallot (ToF) patients, and is associated with arrhythmic death. One possible mechanism for increased arrhythmia vulnerability is that prolonged QRS duration contributes to repolarization heterogeneity. A positive association between QRS duration and QT dispersion has been reported in ToF patients, however, it is generally recognized that QT dispersion is unlikely to reflect repolarization heterogeneity. Here we investigate, in ToF patients, the relation between QRS duration and the second component of the singular value decomposition (SVD) of the T wave, which is mathematically related to repolarization heterogeneity (Van Oosterom, Int J Bioelectromagnetism 2002; 4: 59-60).
Methods. We analyzed two sets of ECGs of 23 ToF patients (13/10 M/F) recorded before and one year after pulmonary valve replacement (PVR) at age 31±8 years. QRS duration was measured by an interactive computer program (Intraval, author HRvE). The second component of the SVD of the T wave was measured by a Matlab program (authors CAS, HvdV, ACM).
Results. QRS duration after PVR (143 ± 31 ms) was significantly smaller than before PVR (148 ± 31 ms, $P<0.01$). The second SVD component of the T wave after PVR (134 ± 71 μV) was significantly smaller than before PVR (164 ± 93 μV, $P<0.05$). QRS duration correlated significantly with SVD before PVR ($r=0.48$, $P<0.05$) and after PVR ($r=0.43$, $P<0.05$).
Conclusions. QRS duration in ToF patients correlates significantly with the second SVD component of the T wave, before and after PVR. This finding suggests that impaired intraventricular conduction (expressed in QRS duration) in these patients causes part of the impaired repolarization heterogeneity (expressed in the second SVD component of the T wave), thus contributing to increased risk of arrhythmogenic death. PVR may reduce arrhythmia vulnerability by diminishing repolarization heterogeneity.

1095-202 Can We Predict Sudden Death After Atrial Repair for Transposition of the Great Arteries?

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Background: Sudden death (SD) is the commonest cause of late death in patients who undergo atrial repair of transposition (TGA) by Mustard's/Senning's technique. Little is known about the predictors of SD.
Methods: We performed a retrospective, multi-center, case-controlled study. TGA patients (n=47) who had a SD event (34 SD, 13 near miss SD) were matched (except one patient (who had only one control)) with two controls with TGA but without a SD event from the same center, era and age at surgery. Clinical reports, ECGs, echocardiograms, chest Xrays, Holter monitors, exercise tests, cardiac catheterization reports and electrophysiology (EP) studies were reviewed and compared with same information from controls at the same time frame.
Results: Presence of symptoms (of arrhythmia or heart failure) at most recent follow-up and history of documented arrhythmia (supraventricular tachycardia (SVT) in particular) were found to increase the risk of SD. QRS duration, QT and QTc on ECG were not predictive of SD. Neither sinus node dysfunction on Holter nor cardiomegaly on chest Xray were predictive of SD. There was inadequate data on echo, exercise testing, cardiac catheterization and EP findings for meaningful analysis. Neither medication use (mostly digoxin) nor pacing was found to be protective.
Conclusions: Presence of symptoms and documented SVT are the best predictors of SD in TGA patients. Patients with these features should be considered high risk and need close follow-up. Aggressive attempts to control SVT using techniques such as catheter ablation are justified in an attempt to prevent SD in these patients.

1095-203 Responsiveness to Inhaled Nitric Oxide Is Related to Mid-Term Survival in Patients With Congenital Heart Disease and Obstructive Pulmonary Hypertension

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Background: It was previously demonstrated that in several adults with congenital heart disease and obstructive pulmonary hypertension and/or the Eisenmenger syndrome, the pulmonary circulation remained responsive to inhaled nitric oxide (iNO). The purpose of our study was to evaluate whether the responsiveness to iNO could predict the mid-term outcome of these patients.
Methods: In 21 consecutive patients, the total pulmonary vascular resistance (TPR) was measured at baseline, after 5 minutes inhalation of NO (80 ppm), and after NO withdrawal. Patients were considered responders when TPR was reduced by at least 20 percent during NO inhalation or when TPR increased by more than 10 percent after NO withdrawal. This group of patients was followed prospectively and the primary endpoint of the study was defined as cardiopulmonary death. Kaplan Meier survival curves for responders and the non-responders were plotted and compared by using log rank testing. Statistical significance was defined as $P<0.05$.
Results: Ten patients were considered responders (4 male and 6 female, mean age 35.1 ± 22.7 years). Eleven patients did not respond (2 male and 9 female, mean age 32.7 ± 19.1 years). The median follow up time of the total group was 9.0 years, range 10.2 years. Four of the non-responders died during follow up (cardiovascular death); no events were found in the group of responders. The difference in survival between the responders and non-responders was statistically significant (log rank $P<0.05$).
Conclusions: The responsiveness to inhaled NO in adult patients with obstructive pulmonary hypertension or the Eisenmenger syndrome seems to be related to the mid-term outcome. These findings might be important issue for risk stratification and choice of treatment in this specific patient population.

1095-204 Ebstein's Anomaly: Incidence of Left Ventricular Noncompaction

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BACKGROUND Left ventricular noncompaction (NC) has been observed as an isolated anomaly or rarely in conjunction with congenital heart disease involving right or left ventricular outflow tract obstruction. In Ebstein's anomaly (EA), left ventricular anomalies resembling NC have recently been described in 3 of our patients (pt). The aim of the present study was to examine the incidence of left ventricular NC in pt with EA. - METHODS Between July 2001 and Feb 2003, 106 pt [64 females (60 %)], with EA underwent echocardiography at our institution. Data from all charts were collected. Videotapes of all pt were blindly reviewed by at least 2 reviewers looking for NC and other left-sided anomalies. RESULTS Mean age was 32±20 years, 31 pt (29 %) were <18 years at the time of index echo. In 84 pt, there was a history of atrial septal defect (ASD) or patent foramen ovale. A history of previous heart surgery was present in 57 pt (54 %) including 30 pt with tricuspid valve (TV) replacement, 16 pt with previous TV repair and/or 34 pt with closure of an ASD. Symptoms were present in 88 pt (83%), 19 pt (18%) were in NYHA class III or IV at the time of the index echo. Severe anatomic form of EA was described in 76 pt (72 %). The criteria of left ventricular NC were fulfilled in 19 EA pt (18 %). Additional findings included a bicuspid aortic valve in 7 pt (7 %), a ventricular septal defect (VSD) in 8 pt (8 %) and mitral valve prolapse in 16 pt (15 %).
Left ventricular ejection fraction was diminished in 7 pt (7 %), left ventricular dilation was found in 4 pt (4 %). Diastolic dysfunction was present in 34 of 95 pt (36 %). Accessory conduction pathways were present in 23 pt (22 %). The presence of left ventricular NC could not be predicted by any associated anomaly, but tended to be more common in pt with accessory conduction pathways ($p=0.11$).- CONCLUSIONS Anomalies of the left ventricle resembling left ventricular NC is noted in a high percentage of patients with EA (18 %). In addition, ventricular septal defect (8 %) and a bicuspid aortic valve (7 %) are observed in EA with increased frequency compared to non-EA patients. Careful assessment of the left-sided cardiac structures in patients with EA is important as cardiac anomalies are not confined to the right side of the heart.

1095-205 Factors Affecting Re-Replacement of the Pulmonary Valve in Children

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Background: Pulmonary valve failure (PVF) in children after pulmonary valve replacement (PVR) is a complex, multi -factorial process. Risk factors for PVF in children have not been systematically defined. If these are identified, this would translate into improved clinical outcomes.
Methods: An IRB approved chart reviewof children with congenital heart disease requiring PVR from January 1987-June 2003 was undertaken using our surgical database. PVF was defined as repeat replacement for valve dysfunction. Time to PVF was determined using actuarial methods. Variables analyzed included patient gender, initial diagnosis, age at surgery, type and size of valve, implantation time, and reason for reoperation. The type and size of second valve were also compared between patients with and without valve failure. Risk factors for failure were sought using univariate and multivariate analysis.
Results: A total of 127 patients underwent PVR, with a mean age of 15.6 +/- 9.3 years. There were 71% patients with TOF type anatomy and 29% with isolated pulmonary stenosis. Pulmonary valve insufficiency was the reason for reoperation in 79% and severe pulmonary stenosis in 21%. Fifty patients (39%) received a cryopreserved pulmonary homograft, 45 (36%) a Hancock porcine and 32 (25%) a Carpentier–Edwards pericardial bioprosthesis. PVF occurred in 19 (15%) patients. At 5 years follow up, PVF rate was 10 % for homografts vs. 10 % in bioprosthetic valves. However at 10 years follow up, 68 % of heterografts failed whereas only 28% of homografts failed. This was statistically significant ($p=0.04$). At 10 years follow up younger age, longer duration of hospital stay and use of bioprosthetic valves were all significantly associated with an increased risk of failure. When homograft vs bioprosthetic valve patient variables were analyzed, there was no difference in patient age, diagnosis, or reason for reoperation.
Conclusion: This is the first study in children which demonstrates that cryopreserved pulmonary homografts have improved durability in the pulmonary position when compared with bioprosthetic valves. It would thus appear to be prudent to preferentially use a cryopreserved homograft in the pulmonary position.

1095-206 Pregnancy in Women With Corrected Tetralogy of Fallot

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Background: Limited data are available regarding the potential complications of pregnancy, fertility and recurrence risk in corrected Tetralogy of Fallot (ToF) patients. We report the largest study so far.
Methods: In this multicenter study 79 patients with corrected ToF were enrolled. For all patients information regarding fertility and cardiologist's advise about pregnancy was obtained. For each pregnancy detailed information about haemodynamics, ultrasound, obstetric status, delivery and pediatric examination was collected. Questionnaires were