942-40 Late Results of Balloon Pulmonary Valvuloplasty in Adults

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Late results of balloon pulmonary valvuloplasty (BPV) in adults are not well known. That is, the results are either early or medium-term ones, or they consist of limited cases. For this purpose we followed up 127 patients (pts.) after successful BPV for a period of 5–7 years. Of these pts. 86 were male and 41 female with an average age of 29.7 (16–54) years. Average pulmonary valve gradient (PVG) was 108.4 + 40.3 (55–240) mm HG before and 22.3 + 8.5 (80–50) mm HG immediately after BPV ($P < 0.0001$). Mild to moderate pulmonary insufficiency was detected in 35 pts. All patients were followed-up every six months by doppler echocardiography for a period of 61–86 months. As they did not refer at the last months of follow up, 16 pts. were excluded from the study. After this period, the average PVG in the remaining pts. was 20.5 + 10.4 (8–60) mm HG ($P < 0.0001$). Of these Pts., in 27 cases, the average PVG was less than its value just after BPV; in 29 cases it was the same and in the remaining 10 cases, the average PVG increased to a value of 44.6 + 10.6 mm HG. After this period of follow up, pulmonary insufficiency was increased slightly in 4 pts. One Pt. showed moderate right ventricular dysfunction due to severe PI. Average PVG was increased mainly in those pts., who showed a PVG of more than 35 mm HG shortly after BPV. Right ventricular hypertrophy in ECG was resolved in 78 Pts. after this period of follow up.

In conclusion, PBV in adults is an effective and safe procedure. Late results of this procedure are as good as their immediate ones.

942-41 Dysplasia of the Atrioventricular Nodal Artery in Patients with Mitral Valve Prolapse and Sudden Cardiac Death

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Patients with isolated mitral valve prolapse (MVP) without significant mitral regurgitation are at a slightly increased risk of sudden cardiac death compared to the normal population. The mechanism(s) of sudden cardiac death in isolated MVP are uncertain. We studied gross hearts and histologic sections of the atrioventricular (AV) node in 21 patients with isolated MVP dying suddenly without apparent cause other than MVP (Group 1, mean age 36 ± 8 years, mean heart weight 420 ± 89 grams). No patient in Group 1 had a history of or treatment for mitral regurgitation, and no left atrial dilatation was present at autopsy. Sudden cardiac death MVP cases were compared to 15 control hearts from trauma victims without cardiac disease (Group 2, mean age 30 ± 7 years, mean heart weight 350 ± 96 grams). Sections of the artery to the AV node within the atrial septum (mean 7 levels in each group) were stained for elastic tissue and proteoglycans. Dysplasia was defined as focal deposition of proteoglycans within the media and partial disruption of the internal elastic lamina. The ratio of lumen area to arterial area was determined by computerized planimetry at the level of greatest luminal narrowing and measured (mean ± SEM) 0.33 ± 0.04 for Group 1 and 0.52 ± 0.04 for Group 2 ($p = 0.001$); this ratio was independent of heart weight, age, or sex of patient by stepwise regression. Dysplasia of the AV nodal artery was present in 9/21 (43%) hearts in Group 1 versus 1/15 (7%) hearts in group 2 ($p < 0.02$). We conclude that narrowing of the AV nodal artery and arterial dysplasia are more prevalent in patients with MVP and sudden cardiac death than controls and may represent a more generalized disorder of proteoglycan deposition. The relationship between these findings and sudden cardiac death needs to be further studied.

942-42 Is Mitral Valve Prolapse with Significant Mitral Regurgitation a Different Condition from Uncomplicated Mitral Prolapse? Results of Family Studies

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Mild instances of mitral valve prolapse (MVP) have been suggested to represent variants of normal, whereas individuals with complicated forms of MVP have a distinct medical condition. This hypothesis would predict different phenotypic features and patterns of inheritance in relatives of index cases with complicated or uncomplicated MVP. Accordingly, we performed clinical and echocardiographic assessment of 16 MVP patients with and 76 without moderate to severe mitral regurgitation (MR+ and MR– probands) and 60 and 256, respectively, first-degree relatives (MR+ and MR– relatives). MR+ probands were older ($p = 0.01$), more likely to be male ($p = 0.002$), were more overweight ($p = 0.004$) and had higher systolic blood pressures ($p = 0.05$) and larger aortic roots ($p = 0.034$) after the effects of age and body size were taken into account. MR+ and MR– relatives had similar prevalences (27 and 32%) and age distribution of MVP, but affected MR+ relatives were younger (expected because more children and fewer parents of MR+ probands could be evaluated), and more likely to be male. MR+ and MR– relatives were virtually identical in regard to body habitus, blood pressure, the prevalence of auscultatory findings, thoracic bony abnormalities and palpitations and all echo measurements including anterior mitral leaflet thickness. Four instances of significant MR and two MVP-related complications (infective endocarditis and transient ischemic attack) occurred in the 82 relatives of MR– probands as opposed to none among relatives of MR+ probands. In 20 families, one proband or relative with MVP had severe MR and at least one other with MVP (presumably due to the same gene) was free of MR or complications. Thus, MVP with severe MR does not represent a heritable phenotype and commonly coexists with mild forms of MVP in the same family, making their classification as separate conditions illogical and potentially misleading.

942-43 Papillary Fibroelastoma: A Review of Echocardiographic Criteria for Diagnosis with Pathologic Correlation

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Papillary fibroelastoma (PFE) is a rare, benign cardiac tumor, diagnosis (dx) usually made at autopsy. Consistent experience with echo analysis is not available. With availability of high resolution echocardiography (echo), dx is being made more frequently clinically. From 1988–1994 13 pts had pathologic confirmation (path) of PFE and an echo performed. Echo made the dx of PFE in 11/13 (85%) pts. Echo analysis of these 11 pts showed the PFE characteristics to be: relatively small tumors (mean 12.2 x 8.6 mm), homogeneous frond-like echodensity, usually pedunculated, attached to the endocardial surface, mobile, with a characteristic vibration or shimmer in the blood flow.

A total of 47 pts had a diagnosis of PFE made by echo found in the absence of endocarditis, with a total of 54 PFE (5 pts had multiple PFE). In 25 the diagnosis was possible by transthoracic echo (TTE) and confirmed by transesophageal (TEE) in 9. In 15 the diagnosis was uncertain by TTE and made by TEE. In 7 only the TEE was available for diagnosis. The location of the PFE was valvular endocardium in 48/55 (87%) (mitral 21, aortic 21, tricuspid 5, and pulmonary 1) and nonvalvular endocardium in 6. The stalk of attachment was seen in 36/54 (67%). Concomitant valvular disease was seen in 17/47 pts (31%) but usually was mild and not due to the PFE. Embolic phenomena occurred in 8/47 pts (17%), and neurologic in 6/8.

We conclude that: (1) Echo can make a dx of PFE in a high percentage of pts with PFE; (2) the incidence of embolism is relatively high, suggewting PFE may be a source of embolism, (3) there is an incremental value of TEE over TTE in diagnosis; (4) most cases were incidental findings without valvular dysfunction.

942-44 Aortic Stenosis — Noninvasive Assessment of Prognosis

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During the last 10 years (y) 97 patients (pts), mean age 52.5 y, with aortic stenosis (AS) were regularly controlled in the out-patients clinic (mean follow up 4.5 y). Throughout the follow up 16 cardiac death occured (9/16 pts declined the operation) and 39 pts were operated. According to the Doppler echocardiography (DE) at the beginning AS was mild in 43, moderate in 34 and severe in 20 pts. Initially 30/43 (70%) pts with mild, 12/34 (35%) with moderate and 7/20 (35%) with severe AS were in NYHA class I. The great majority (41/49, 84%) of NYHA class I pts had normal exercise capacity, regardless of AS severity. At the last visit 16/43 (37%) with initially mild had moderate and 10/43 (23%) severe AS. 4/10 pts developed severe AS in the time of 5 y. 19/34 (54%) pts with initially moderate developed severe AS. Progression in DE was in 25/45 (56%) followed by an increase in NYHA class but in 8/33 (24%) pts worsening of NYHA class was not related to an increase in DE. Progression to severe AS was followed by an increase in NYHA class in 20/29 (69%) and decrease in exercise capacity in 17/29 (59%) pts; systolic pressure did not increase or decreased with exercise in 10/29 (35%) pts. All 16 pts who died developed symptoms before death but in 2 physical activity was not limited. 1/16 had mild and 3/16 moderate AS. These 4 pts had angina and died sudden; coronary artery disease (CAD) was proven in 3/4 (1 pt without angiography). 37/39 (95%) of pts admitted to surgery were symptomatic but 8/39 (20%) had only moderate AS.

Conclusions: 1) within 5 y 10% of pts progress from mild to severe AS, 2) 30% of pts with severe AS were asymptomatic with a normal exercise capacity, 3) increase in symptoms, decrease in work capacity and absent increase in systolic pressure at ergometry are the best signs of stenosis progression to severe AS, 4) the risk of sudden death in asymptomatic pts is very low regardless of AS severity, 5) pts with angina in even mild or moderate AS are