HEMODYNAMICS AND ANGIOGRAPHY AFTER THE MUSTARD OPERATION.
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Fifty children with d-transposition of the great arteries (TGA) underwent cardiac catheterization after a Mustard operation modified to reduce the baffle obstruction to the systemic and pulmonic veins. The data were analyzed to establish the new hemodynamic and angiographic standards in these reconstructed hearts with TGA and corrected physiology. The mean pressure in the systemic venous atrium (SVA) was 5±2mmHg and in the pulmonic venous atrium (PVA) 9±3mmHg. The pressure in the SVA in all patients indicated a low compliance atrial chamber with rapid negative Y descent. Only minimal pressure differences (mean 2mmHg) were found between the systemic veins and the SVA. The pressure tracings and angiography clearly define an anterior and posterior PVA segment due to interposition of the systemic venous conduit (mean gradient 2mmHg). Due to the persistence of ventriculo-arterial discordancy, the pressure-volume characteristics of the two ventricles remain basically unaltered after surgery. The degree of right ventricular (RV) dysfunction and posterior bulging of the septum was assessed by simultaneous biventricular angiographic contrast studies and correlated with the left ventricular (LV) outflow stenosis. No progression of LV outflow stenosis or occurence of tricuspid regurgitation was demonstrated postoperatively. The Mustard operation offers excellent physiologic correction of TGA. Potential hemodynamic problems may occur later due to: 1. Low compliance of SVA, 2. Obstructed flow between segments of PVA, 3. RV dysfunction.

PULMONARY VASCULAR DISEASE IN COMPLETE ATRIOVENTRICULAR CANAL DEFECT
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Because of the current success of corrective surgery for the complete form of atrioventricular canal (AVC), this study was undertaken to determine the incidence and time of occurrence of pulmonary vascular disease (PVD) in this lesion. Sixty patients (pts) with AVC were evaluated for PVD. All had a combined large atrial and ventricular septal defect. Thirty-nine lung specimens (3 obtained by open lung biopsy, 36 by post mortem) were evaluated microscopically for PVD and graded according to the Heath-Edwards classification. Forty-two children had cardiac catheterization (ages 4 days-7½ years, median 9 months), and pulmonary hypertension was the rule, with mean pulmonary artery pressures (PAP) from 30-85 mm Hg (median 55 mm Hg). Twenty-four pts had pulmonary artery banding (PAB). There were 12 deaths (50%). Ten children had excellent palliation from 1-9 years postoperatively, and 2 pts who were well palliated subsequently required shunts for too tight PAB. In unoperated children PAP increases with age, and by 2 years all pts studied had PAP of at least 50 mm Hg. None of the lung specimens were normal and 35/39 had grade 2 or above. In pts one year or older 8/12 without PAB had grade 3 PVD or above. This study indicates that severe pulmonary hypertension is present in all patients with complete AVC without pulmonic stenosis; advanced PVD develops at an early age, and is present in 2/3 of pts over one year of age. PAB is necessary to protect the lungs from the development of advanced PVD, if corrective surgery is to be delayed beyond 2 years of age.

SPECIFIC ECHOGRAPHIC FEATURES OF PAPILLARY MUSCLE DYSFUNCTION IN CORONARY HEART DISEASE: ULTRASOUND PROFILE OF DEPRESSED CARDIAC FUNCTION, DIFFUSE DYSSYNERGY AND MITRAL ACCELERATED CLOSURE WITH DISTINCTIVE DOUBLE CONFIGURATION
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Despite the common prevalence of mitral regurgitation (MR) and the several causes of this disorder, both MR detection and determination of etiology have been difficult by echography. To characterize the ultrasound features of MR due to ischemic heart disease, echograms of 15 patients (pts) with moderate to severe papillary muscle dysfunction (PMD) were compared to those of 15 pts without PMD (C) matched for age, sex and extent of coronary stenosis. As compared to C, pts with PMD manifested increases in mean left ventricular (LV) end-diastolic dimension (PMD: 6.5 vs C: 5.5 cm), LV end-systolic dimension (5.2 vs 4.1 cm), left atrial (LA) dimensions (3.7 vs 3.0 cm) and mitral valve (MV) early diastolic closing velocity (132 vs 106 mm/sec), while reductions occurred in ejection fraction (42 vs 60%), interventricular septal (IVS) contraction amplitude (4.5 vs 7.1 mm) and IVS contraction velocity (24 vs 38 mm/sec) (all $p<.05$). Importantly, the mitral echogram frequently displayed a distinctive double diamond shape configuration. No significant changes were noted in stroke volume (93 cc), posterior wall contraction amplitude (10.5 mm) or posterior wall velocity (35 mm/sec). Thus pts with significant PMD may be recognized by echographic profile of increased LV and LA dimensions, accelerated MV early diastolic closing velocity, double diamond mitral echogram, and reductions of ejection fraction, IVS contraction amplitude and IVS contraction velocity. These data indicate that MR due to coronary PMD occurs with diffuse LV dyssynergy and can be recognized by distinctive echographic constellation of abnormalities.

EFFICACY OF ORAL DISOPYRAMIDE PHOSPHATE FOR LONG-TERM TREATMENT OF VENTRICULAR ARRHYTHMIAS
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The efficacy and safety of oral disopyramide phosphate (DP) for long-term treatment of ventricular ectopic arrhythmias was assessed in 55 patients with frequent ventricular beats (VPBs) or recurrent ventricular tachycardia (VT). Each patient had more than 60 VPBs per hour during baseline ambulatory electrocardiographic monitoring, and 44 patients had been refractory to other antiarrhythmic agents or had experienced intolerable side effects from them. Ten hour Holter ambulatory electrocardiograms were recorded during a control period and at 3 month intervals during DP therapy. After the first 3 months of DP treatment, 52% of patients had greater than 90% reduction of VPBs and 64% of patients had more than 50% reduction ($p<.05$). This level of effectiveness was sustained for as long as 15 months. VT was abolished or reduced in frequency in 9 of 15 patients and unchanged or worse in 6 patients. Adverse effects included dry mouth (53%), blurred vision (9%), urinary hesitancy (5%), nausea (2%), and possible deterioration of ventricular function in 3 patients with cardiomyopathy. There was no evidence of hepatic, renal, or hematologic toxicity. Twenty patients unable to tolerate quinidine had only minor anticholinergic side effects from DP. DP is an effective and safe oral antiarrhythmic agent for long-term treatment of ventricular premature beats and ventricular tachycardia.