weaker, so allowing for dilatation and dissection. 14

Adult polycystic kidney is a disease with family tendency transmitted through a simple autosomal gene of high penetrance. 7 Although the final cause of cyst formation is unclear, it is thought that the inadequate collagen support of the renal tubules may be the explanation. 8

Adult polycystic kidney is associated with cardiovascular anomalies. Aneurysms of cerebral or visceral arteries are well known to be frequent in this disease. 15 16 Recently, Leier et al. 17 have reported a high incidence of annuloaortic ectasia in patients with polycystic kidney. In their series, 18 percent of patients admitted in the hospital because of polycystic kidney had annuloaortic ectasia, and 20 percent of autopsied patients with polycystic kidney had associated aortic root anomalies. The cardiovascular lesions associated with APKD are varied, so that APKD patients should have a careful cardiovascular evaluation.

In our experience with 18 consecutive patients with annuloaortic ectasia who had IVP as part of the routine work-up prior to cardiac surgery, we have found four cases of adult polycystic kidney (22 percent). None of them had Marfan's syndrome and none of them had renal symptoms. The high rate of association of both diseases could be related to some genetic factor. Because of this high incidence of annuloaortic ectasia and adult polycystic kidney disease, the cardiac surgeon should be aware of the association and study the renal function and renal morphology in every patient with annuloaortic ectasia (Fig 1). Patients with annuloaortic ectasia manifest a hereditary disorder of connective tissue with the potential for involvement of other organs and should have a careful appraisal of family history and systemic evaluation, as well as the recommended renal evaluation.

REFERENCES
3 Comfort MV, Gray HK, Dahlin DC, Withsell FB. Polycystic disease of the liver: A study of 24 cases. Gastroenterology 1952; 20:60-68

Dilated Right Ventricular Cardiomyopathy: Uhl's Disease*

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Uhl's disease is a rare disorder originally described in 1952 in an infant with severe diffuse right ventricular dysfunction with total absence of the myocardium. Uhl considered the disease to be congenital in origin. We report a patient with severe dilated congestive cardiomyopathy limited to the right ventricle but apparently developing in adulthood.

Uhl's disease is a rare entity. 1 It is characterized by almost complete absence of the myocardium in the right ventricle and is thought to be congenital in origin. We report a patient with findings of severe and diffuse right ventricular

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dysfunction similar to Uhl's anomaly, but manifest and apparently developed only in adulthood.

**CASE REPORT**

A 37-year-old woman was referred to the Cardiac Investigation Center of the Victoria General Hospital for evaluation of a possible atrial septal defect. Her cardiac history began approximately seven years previously with onset of mild dyspnea on exertion. An M-mode echocardiogram at that time revealed normal right ventricular size (22 mm) and mitral valve systolic prolapse. She remained relatively well until two years prior to the present admission when her dyspnea began to worsen, progressing to functional class 3 (NYHA) at admission. She had no orthopnea, paroxysmal nocturnal dyspnea, edema of her extremities, or angina. She complained, however, of palpitations, described as sudden onset of rapid, regular and forceful heart action, usually lasting about 15 minutes. During an episode associated with weakness, the rhythm was found to be ventricular tachycardia and necessitated cardioversion. Her past history was otherwise unremarkable and included three normal pregnancies and deliveries. A brother had a "hole in his heart."

Cardiovascular examination revealed sinus rhythm with normal arterial pulse volume and contour. The maximal JVP height was at the sternal angle, had a normal waveform, and decreased with inspiration. Hepatojugular reflux was negative. The apical impulse was in the fifth left intercostal space and normal in intensity. There was a mild parasternal systolic impulse. The first heart sound was single; the second, widely and fixedly split. There was an early diastolic sound audible over the lower left sternal border which increased in intensity with inspiration. The rest of the cardiac examination was unremarkable.

The 12-lead ECG demonstrated sinus rhythm, right axis deviation and complete right bundle branch block. M-mode (Fig 1) and two-dimensional (Fig 2) echocardiographic images revealed a markedly dilated right ventricle with globally hypokinetic systolic motion. Left ventricular volume and systolic function were normal, and there was no evidence of any valvular abnormalities. Doppler study revealed no evidence of left-to-right shunt at the atrial or ventricular level and no tricuspid insufficiency. A chest x-ray film suggested right ventricular enlargement and no evidence of shunt vascularity.

Cardiac catheterization demonstrated normal hemodynamics apart from a right ventricular end-diastolic pressure of 12 mm Hg (post-right ventriculogram) and a left ventricular end-diastolic pressure of 14 mm Hg. Oxygen saturation studies revealed an insignificant (1.06 to 1.0) left-to-right shunt from an anomalous pulmonary vein draining into the left innominate vein and no shunt at the atrial or any other level. Angiography confirmed the anomalous pulmonary vein flow and also the presence of normal coronary arteries. Right ventriculography demonstrated a markedly dilated and diffusely hypokinetic right ventricle.

Re-implantation of the anomalous pulmonary vein to the left atrium was undertaken to decrease the volume load on the dilated right ventricle. At operation, the right ventricle was found to be grossly enlarged, and there was generalized thinning of the myocardium suggestive of Uhl's-type "parchment" myopathy. Because of the severe thinning, biopsy of the right ventricle was not attempted. Biopsy specimens of the left ventricle and left atrium revealed normal myocardium. Following postoperative recovery, the patient was discharged home receiving digitalis and a diuretic. Over the next year, she had, however, numerous episodes of ventricular tachycardia despite antiarrhythmic therapy that was selected on the basis of electrophysiologic stimulation testing.

**DISCUSSION**

Dilated, congestive cardiomyopathy limited to the right ventricle is an unusual and even rare disorder. The differential diagnosis includes atrial septal defect, partial anomalous pulmonary venous drainage, Ebstein's anomaly, tricuspid or pulmonary valvular insufficiency, right ventricular infarction, Uhl's disease, and right ventricular dysplasia. Our patient had a small left-to-right shunt as a consequence of a partial anomalous pulmonary vein connection. This was, however, too small to precipitate the patient's extreme right ventricular dilatation and dysfunction. There was no evidence of right ventricular volume overload from other congenital or valvular disease, and there was no evidence of ischemic cardiac disease. Thus, the most likely cause of her dilated and poorly functioning right ventricle was primary muscle disease.

**FIGURE 2.** Stop-frame four-chamber real-time echocardiogram (left panel) and diagnostic sketch (right panel) illustrating the large right ventricle of our patient. RA is right atrium; RV, right ventricle; LA, left atrium; and LV, left ventricle.
A major problem in dealing with a patient with right ventricular cardiomyopathy is the multitude of diagnostic labels and concomitant lack of meaningful etiologic data. In 1952, Uhl described a case of diffuse, severe right ventricular dysfunction with total absence of the myocardium in a seven-month-old infant. Uhl considered the disease to be congenital in origin, and since then, there have been numerous similar case reports of apparently congenital hypoplasia of the entire or near-entire right ventricle, with or without other associated congenital defects. As pointed out by Waller et al., congenital hypoplasia of portions of the right ventricle was first described by Osler in his textbook of medicine in 1905 and another case was reported in 1952, both prior to Uhl's communication. More recently, patients with largely segmental, but occasionally diffuse right ventricular hypoplasia and recurrent ventricular tachycardias have been described as having right ventricular dysplasia. These patients have a variable clinical presentation, including ventricular tachycardia, low output, right heart failure, or asymptomatic cardiomegaly. In addition, Fitchett et al. reported 14 cases of severe right ventricular dilated cardiomyopathy. Despite largely-preserved left ventricular function, these latter authors felt that the right ventricular cardiomyopathy was a manifestation of diffuse myocardial disease, with the left ventricle not yet affected. These same authors also considered that there was "almost certainly" some overlap among Uhl's anomaly, right ventricular dysplasia, and right ventricular dilated cardiomyopathy. We concur with the latter opinion that there is probably a continuum of the degree and timing of myocardial involvement in right ventricular cardiomyopathy and suggest that, until more is known about the pathophysiology and etiology of such patients, it is preferable to use generic descriptive terminology. Thus, a patient such as ours may best be described as having idiopathic dilated cardiomyopathy isolated to the right ventricle with ventricular dysrhythmia as a major manifestation and with clinical onset in adulthood.

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REFERENCES
1 Uhl NS. A previously undescribed congenital malformation of the heart: almost total absence of the myocardium of the right ventricle. Bull Johns Hopkins Hosp 1952; 91:197-205
4 Waller BF, Smith ER, Blackbourne BD, Arce FP, Sarkar NN, Roberts WC. Congenital hypoplasia of portions of both right and left ventricular myocardial walls. Am J Cardiol 1980; 46:885-91

Augmentation of Cardiac Function in End-Stage Heart Failure by Combined Use of Dobutamine and Amrinone*

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A patient with end-stage congestive cardiomyopathy had progressive hemodynamic deterioration while awaiting orthotopic heart transplantation. Attempts to support cardiovascular function by high-dose dobutamine infusions were complicated by life-threatening cardiac arrhythmias. The addition of the noncatecholamine inotropic agent, amrinone, improved ventricular performance, enabling reduction of the dose of dobutamine and resolution of the cardiac arrhythmias. Beta receptor stimulation by dobutamine combined with phosphodiesterase inhibition by amrinone may additively or synergistically augment cardiac function despite severe congestive heart failure and also have an adrenergic "sparring effect."

Orthotopic heart transplantation is an effective treatment of end-stage heart failure. However, many patients die while awaiting transplantation. In such patients, ventricular performance must be stabilized sufficiently to permit evaluation and organ procurement. Currently, inotropic support with dobutamine and intraaortic balloon counterpulsation are the principal modalities used for this purpose. However, there are no data regarding potential synergistic effects of adrenergic stimulation by catecholamines combined with phosphodiesterase inhibition by drugs such as amrinone to improve cardiac function in end-stage heart failure. We describe a patient with end-stage congestive cardiomyopathy and progressive clinical deterioration in whom such combined therapy improved cardiovascular function during intensive care and enabled subsequent successful transplantation.

CASE REPORT

A 47-year-old white man with a history of severe chronic congestive heart failure was admitted to the hospital because of repeated episodes of chest pain and dyspnea. He had had a myocardial infarction in 1977, three- vessel coronary artery bypass grafting in 1979, and a second infarction in 1982. Subsequently, he was hospitalized frequently for worsening congestive cardiomyopathy. One month before the most recent admission, he was admitted for treatment of progressive fatigue. Two-dimensional echocardiography disclosed a dilated left ventricular (LV) chamber. Radionuclide ventriculography showed biventricular dilation and a resting LV ejection fraction of 22 percent. He was treated with digitalis, diuretics, anticoagulants, nitrates, and captopril, and was

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Cardiac Function in End-stage Heart Failure (Guimond et al)