tant stream throughout diastole, sometimes actually increasing the diastolic murmur in later diastole.

Despite the intensity of the murmur of catheter-induced pulmonic valve rupture in our patient, he appeared to suffer no immediate untoward effects. Similar benign consequences of pulmonic valve regurgitation have been reported by others. In addition, one of the two other reported patients with catheter-induced pulmonic valve rupture survived and is doing well.

The mechanism by which the pulmonic valve cusp in our patient was torn by the catheter is unclear. There is no evidence that the tear was produced at the time of insertion of the catheter, but it must have occurred during the six days that the catheter was in place, since the murmur of pulmonic regurgitation was heard before the catheter was withdrawn. The patient had septicemia, but infective endocarditis clearly was not present at autopsy. Although our patient did have moderate pulmonary arterial hypertension, it is unlikely that the pulmonary arterial diastolic pressure of 30 to 40 mm Hg can cause rupture of a previously normal pulmonic valve cusp. Movement of the catheter while in place can presumably cause cuspal weakening and predispose the cusp to rupture, since others have reported damage to the pulmonic valve cusps by indwelling catheters in the form of focal cuspal hemorrhages and endocardial verrucae. However, these specific findings were not documented in this case.

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Aprindine in the Treatment of the Idiopathic QT Interval Prolongation Syndrome*

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Figure 3. Pulmonic valve at autopsy with probable catheter location and resulting cuspal tear.
A 19-year-old patient with idiopathic long QT interval syndrome (LQTS) was treated with diphenylhydantoin, propranolol, digoxin and bretylium tosylate. In view of continued refractoriness of ventricular arrhythmias, bretylium was discontinued and aprindine, 200 mg qd, was added. This drug regimen completely ablated the ventricular arrhythmias. Since the results of left stellatelectomy (the last therapeutic measure in LQTS) do vary, aprindine may be an important adjunct in the treatment of this syndrome.

The idiopathic long QT interval syndrome (LQTS) is known to be a lethal entity characterized by paroxysmal ventricular arrhythmias, syncopal episodes, and sudden death. This is the first case report in which aprindine was utilized to ablate completely a ventricular dysrhythmia in a patient who was refractory to all conventional medical therapy. Since the results of left stellate ganglionectomy, although encouraging, do vary, and since aprindine has not been demonstrated to prolong the QT interval significantly, it might be a promising drug in the treatment of refractory ventricular arrhythmias due to idiopathic QT interval prolongation.

Case Report

The 24-hour Holter monitor examination of a 19-year-old Caucasian woman revealed QT interval prolongation (QT = 0.60 sec) with R on T ventricular ectopic beats occurring immediately after the vulnerable period. The patient also manifested labile T wave changes (Fig 1). There was no evidence of previous cardiac arrhythmias; however, one year prior to hospitalization, the patient had complained of chest pain and dyspnea. No arrhythmias were diagnosed at that time. The patient’s father had an undefined history of heart disease. There was no family history of sudden death. Because of the severity of the arrhythmias, the patient was admitted to the hospital for continuous monitoring in August 1979. The physical examination was entirely normal; in particular, there was no evidence of cardiac murmurs or gallop rhythm. The chest x-ray film and the laboratory data were within normal limits. The echocardiogram demonstrated mitral valve prolapse.

The patient was treated with diphenylhydantoin (Dilan- tin), 300 mg qd, and with propranolol, 160 mg qd. Digoxin, 0.25 mg qd, was administered after an appropriate loading dose. The latter medication was initiated in the hope that it would shorten the QT interval. In view of the continued refractoriness of the arrhythmias, bretylium tosylate was added. The ventricular arrhythmias, however, persisted despite this combination of drug therapy. Bretylium tosylate was discontinued and aprindine, 200 mg qd, was initiated. This drug regimen resulted in complete ablation of the ventricular arrhythmias which was confirmed by treadmill stress testing and 24-hour Holter monitor examination.

Discussion

The case described represents an idiopathic long QT interval syndrome (LQTS) which is associated with severe ventricular dysrhythmias. The syndrome falls into various categories. The Jervell and Lange-Nielsen syndrome is associated with congenital deafness carrying an autosomal recessive inheritance. An autosomal dominant pattern of inheritance without congenital deafness is characteristic of the Romano-Ward syndrome. In view of the absence of a family history of sudden death or syncope, it is unlikely that our patient falls into the latter category.

A review of the literature indicates that the LQTS occurs far more frequently than previously assumed, and that increasing awareness has led to a sharp rise in the number of diagnosed patients. An association with mitral valve prolapse, as observed in our patient, has been described before, but is considered rare. The pathogenesis of the LQTS is unclear. An imbalance of the sympathetic activation of the heart, causing sympathetic hyperactivity, is considered an important factor. Studies in the experimental animal and in man have demonstrated that sympathetic overactivity, pro-

FIGURE 1. Holter monitor strips demonstrating labile T wave changes, a prolonged QT interval, and R on T ventricular premature contractions.
duced by stimulation of the left or by blockade of the right stellate ganglion, does produce prolongation of the QT interval and ventricular dysrhythmias.

Drugs which decrease cardiac sympathetic tone have been effective in the treatment of the LQTS. Propranolol and Dilantin successfully suppressed prolongation of the QT interval and diminished or abolished ventricular arrhythmias.5,6,11 Bretylium tosylate effectively abolished the LQTS, regardless of whether it was administered intravenously or per os.4,5 In contrast, drugs such as quinidine and procar clandest, which prolong the QT interval, are hazardous and have worsened the LQTS causing near lethal ventricular arrhythmias.5,12 Similar observations have been reported after the administration of disopyramide.13 Left stellate gangliectomy is considered in patients who have failed to respond to optimal drug therapy. A report of 20 patients who underwent this procedure is encouraging.2 Left stellectomy in combination with drug therapy sharply reduced the incidence of ventricular arrhythmias. Long term results, however, have to be evaluated.

It is apparent that the treatment of the LQTS remains a challenging problem. The observation that aprindine completely ablated ventricular dysrhythmias associated with the LQTS could be of great importance. Further clinical investigation is warranted for evaluation of its efficacy.

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IATROGENIC INTERNAL MAMMARY ARTERY-TO-INNOMINATE FISTULA

Percutaneous Nonsurgical Closure

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An internal mammary artery-to-innominate vein fistula, a rare complication of subclavian vein central venous pressure catheter insertion, was closed without surgery by percutaneous angiographic techniques. In this case and in similar cases of arteriovenous fistulas, percutaneous angiographic therapy can be both safe and effective and can lead to significant reductions in costs, hospitalization, and patient trauma.

Subclavian vein catheterization is frequently used for monitoring central venous pressure (CVP), extended administration of hyperosmolar parenteral alimentation, and rapid fluid infusion in hypovolemic patients.1 Various complications associated with insertion, long-term use, and removal of subclavian catheters have ranged from minor hematomas to perforation of major intrathoracic vascular structures.1,2 We report a case in which an internal mammary artery-to-innominate vein fistula following subclavian CVP catheter insertion was successfully treated without surgery by percutaneous angiographic catheter techniques.

CASE REPORT

A 32-year-old woman complained of a "pulse-like" noise and a sensation of pressure in her right clavicular area. Physical examination revealed a continuous bruit and palpable thrill in the right subclavicular region. Venous dis
tension of the right arm was present; however, there was no evidence of congestive heart failure. Her history in
cuded poliomyelitis during infancy with residual bilateral lower extremity weakness and severe scoliosis. Approximately two years earlier, a CVP catheter was inserted from the right infraclavicular approach for emergency treatment of severe hypotension and urticaria, secondary to an allergic reaction from a drug taken for a lower urinary tract infec
tion.

Selective right subclavian angiography showed an arterio-
venous fistula, 1 cm long, between the right internal mammary artery and the right innominate vein (Fig 1). After a thorough discussion of the available therapeutic alternatives for closure of the fistula, the patient chose to have percutaneous transcatheter occlusion. The right internal mammary artery was catheterized selectively by the femoral artery approach, and two Gianturco coil spring occluders were inserted; one was placed a few millimeters distal to the origin of the fistula, and the second just proximal to its origin (Fig 2). Within three minutes, the bruit and thrill had disappeared. A right subclavicular angiogram demonstrated proximal occlusion of the right internal mammary artery 2 cm from its origin, with no flow of contrast.

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