cutaneously administered terbutaline and epinephrine in patients with stable asthma; these reports are critically reviewed in the report in this issue of Chest (see page 129) by Smith et al, who have also extended these observations to patients with acute asthmatic attacks, who are more likely candidates for parenteral therapy with bronchodilator drugs. These studies clearly indicate that subcutaneously administered terbutaline has little therapeutic advantage over epinephrine; indeed, in man, terbutaline appears to have quantitatively greater hemodynamic effects than roughly equivalent bronchodilator doses of epinephrine. The discrepancy between these findings and the results of preclinical studies indicating a marked dissociation between the cardiac and bronchial effects of terbutaline is unexplained. In severe attacks of asthma not responsive to aerosol bronchodilator medication, subcutaneous administration of epinephrine is often helpful, possibly as a result of delivery of the bronchodilator drug via the circulation to airways peripheral to occlusive mucous plugs. A drug with clinically demonstrable bronchoselectivity administered by injection would undoubtedly have advantages over epinephrine, particularly for patients with both severe asthma and cardiovascular disease. Consequently, the search for newer synthetic agents with the nature of 

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Congenital and Acquired Syndrome of a Long Q-T Interval

The association of heritable prolonged Q-T interval, ventricular dysrhythmias, recurrent syncopal attacks, and sudden death in childhood constitutes a well-described clinical syndrome. In the presence of congenital deaf-mutism, it has been named the Jervell and Lange-Nielsen syndrome, after the authors who first described it. The mode of inheritance appeared to be autosomal recessive. Romano et al and, independently, Ward described a similar syndrome in children without deafness. The mode of inheritance of the latter seemed to be autosomal dominant. Both syndromes can be classified as congenital syndromes of the prolonged Q-T interval.

A recent review of the literature on the syndrome of prolonged Q-T interval reported 203 diagnosed cases. Only 30 percent of these cases were deaf. Of the 203 reported cases, 34 percent died. The mortality of patients with syncope and without treatment was found to be 73 percent. The effect of therapy on the natural history suggested that treatment with beta-adrenergic sympathetic blocking drugs significantly reduced mortality. The effectiveness of left sympathectomy is currently being evaluated and seems to be promising. The pathogenesis for the syndrome of a prolonged Q-T interval has not been completely elucidated. Several hypotheses have been proposed, two of these being (1) an abnormality in the vascular supply to the sinus node

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and specialized conduction system, and (2) a regional autonomic dysfunction. Results of experimental studies, clinical observations, and therapeutic intervention strongly suggest an association between unilateral sympathetic dysfunction and the congenital syndrome of a prolonged Q-T interval.

Prolongation of the Q-T interval associated with ventricular dysrhythmias and syncpe or sudden death can also be acquired and secondary to the use of drugs (quinidine, procaine amide, or phenothiazines) electrolyte imbalance (hypokalemia), or disturbances in conduction (atrioventricular block). Patients with acquired prolonged Q-T intervals are usually symptomatic later in life and have no family history of the syndrome. The attacks of ventricular dysrhythmia are often treated successfully by correction of the precipitating cause or causes.

In the case report by Schneider et al published in this issue (see page 210), the patient was in her fourth decade and had no previous history of syncope and no family history of sudden death. The episodes of ventricular fibrillation were probably precipitated by a combination of factors, including electrolyte imbalance (hypokalemia) and administration of drugs (procaine amide hydrochloride and perphenazine). The arrhythmia could be controlled with intravenous administration of potassium, discontinuation of therapy with drugs, insertion of a transvenous pacemaker, and rapid ventricular pacing. Electrocardiograms taken five years prior and 15 months after the attack showed persistent prolongation of the Q-T interval.

This case illustrates the difficulties in classification of the syndrome of a prolonged Q-T interval. I am not sure whether this case represented the congenital syndrome. Obviously, this was not a typical case of the congenital syndrome described by Romano et al and by Ward, however, it cannot be categorized as acquired, since the prolongation of the Q-T interval antedated the onset of symptoms. We have seen a few patients with syncope and ventricular tachyarrhythmias occurring late in life associated with prolonged Q-T intervals, but without obvious precipitating causes or a family history of the syndrome. Whether these cases represent part of a broader spectrum of the syndrome described by Romano et al and Ward or belong to a different group of cases with the syndrome of a prolonged Q-T interval is not known. In these patients, prolongation of the Q-T interval could also be acquired and reflect cardiac disease or acquired autonomic dysfunction. The natural history and response to therapy of these patients might differ from the typical patients with the congenital syndrome of a prolonged Q-T interval. Rather than lump together all patients with the syndrome of a prolonged Q-T interval, it is important to define subgroups based on the pathogenesis of the underlying abnormality. More rational therapy may be developed as understanding of these interesting diseases is improved.

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Treatment of Massive Pulmonary Embolism

In this issue of Chest (see page 213), there is a report by Scoggins and Greenfield describing transvenous pulmonary embolectomy. While this is a unique approach and perhaps the least traumatic described to date, many methods have been suggested for the removal of pulmonary emboli. The critical question still remains: Under what circumstances should the physician recommend pulmonary embolectomy?

It would be foolish not to recommend the removal of emboli if the patient certainly would die otherwise. The identification of an unequivocal set of circumstances or findings which indicate those patients who will die without embolectomy remains to be made. Therefore, the decision to recommend embolectomy is still one of judgment. In making this judgment, the physician should consider the following: (1) A positive diagnosis is essential and can only be obtained by pulmonary arteriographic