Over the past eighteen months we have underwritten four life insurance applicants with a prolonged QT interval on their electrocardiogram. Three of these cases are the basis for this paper. The fourth, a young female with multiple cardiac arrests and subsequent sympathectomy, could not be retrieved.

The congenitally prolonged QT interval had a total mortality of 73% in one series of 203 patients. Unfortunately, the study group was heterogenous with regard to symptoms, QT duration and length of follow up which varied from several years to greater than ten. The average age at death was 9.3 years with the youngest being 44 days and the oldest 40 years. Since it was first described in 1957, the incidence in the general population has not been ascertained: however, it appears far more unrecognized than rare. The frequency of the prolonged QT interval among deaf-mute children is about 0.25%; conversely, cases with deafness represent 30% of all patients affected by the syndrome.1 Due to its high potential for premature mortality, medical directors and underwriters should be aware of this electrocardiographic abnormality.

The first applicant was a 56 year old female applying for 5 million dollars of term insurance. She had no history of a prolonged QT interval, although review of her electrocardiogram from June 1984 showed the same QT interval as the current tracing. Her past medical history included hypothyroidism and mild idiopathic edema for which she was taking synthroid and aldactazide daily. Family history was negative for sudden death with three living siblings ages 60, 58 and 52. Her father died at age 84 from a heart attack, her mother at age 80 from old age. Cardiac and neurologic symptoms were absent. Physical examination revealed no abnormalities. Electrolytes, blood chemistries and a treadmill were all normal. Current electrocardiogram is shown in Figure 1.

Our second applicant was a 28 year old asymptomatic male with a family history of sudden death associated with prolongation of the QT interval on the electrocardiogram. The application was for 10,000 dollars of whole life. He was initially evaluated in 1977 after his younger sister experienced a spontaneous cardiac arrest during a violent electrical storm. She was documented to be in ventricular fibrillation and was successfully resuscitated. Subsequent evaluation revealed her to have a prolonged QT interval. An older sister had previously suffered a fatal cardiac arrest. There was no family history of deafness or other premature deaths. The applicant's initial medical evaluation revealed a normal physical examination, echocardiogram, treadmill and blood chemistries. The resting electrocardiogram had a sinus rate of 70 with a QT interval of 0.40 seconds, the upper limit of normal being 0.396 seconds. The T waves were described as abnormal (that tracing is unavailable). He was begun on propanolol. Figure 2 shows the electrocardiogram of July 1979, after the initiation of beta blocker therapy. The T waves are identical to those described prior to treatment and reveal the typical bifid configuration. His last medical follow-up in September 1980 noted that he had discontinued his medication.

The final applicant was a 24 year old female applying for $100,000 of whole life. Her family history included sudden deaths of her mother, at age 27, a cousin and an aunt. Her brother, at age 25, was found to have a prolonged QT interval. Historically, she had several syncopal episodes prior to her initial evaluation in 1982. Examination at that time revealed a grade 2/6 systolic ejection murmur at the apex without a click. Subsequent examinations did not confirm the presence of a cardiac murmur. No hearing impairment was noted. The QT interval was 0.56 seconds. She was begun on beta blockade therapy; this, however, was poorly tolerated due to fatigue and discontinued. In February 1983 she had a cardiac arrest; ventricular fibrillation was documented. Subsequent to successful resuscitation, the QT interval was noted to be 0.55 seconds. She was again begun on a beta blocker and remains asymptomatic. Her current electrocardiogram is shown in figure 3. The QT interval is 0.50 seconds while on therapy.

Discussion
Prolonged QT interval with sudden death secondary to ventricular fibrillation is associated with two congenital syndromes, 1) Jervell and Lange-Neilsen and 2) Romano-Ward. The former includes deafness and shows an autosomal recessive inheritance; the latter has a
autosomal dominant inheritance. The familial disorders exhibit a 2:1 female preponderance. Of those affected, 88% had relatives with a prolonged QT. In both, sudden cardiac death can be precipitated by fright (auditory startle stimulus), anger, emotion and exercise. Of note is the second applicant’s sister fibrillation during a violent electrical storm.

Acquired prolongation of the QT interval can be produced by drugs (quinidine, tricyclics, phenothiazines, procaine amide), electrolyte abnormalities (hypocalcemia and hypomagnesemia), myocardial ischemia, acute neurologic abnormalities with secondary sympathetic discharge, severe weight reduction, mitral valve prolapse syndrome and acute myocarditis. Hypokalemia prolongs the QU not the true QT interval. As with those of a congenital etiology, these can predispose the individual to ventricular fibrillation of sudden death.

The QT interval represents the depolarization and subsequent repolarization of the ventricular musculature. Systole begins at the peak of the R wave and terminates near the end of the T wave.

Correct measurement of the QT interval can be difficult. Measurement should be taken in a lead where there is an initial Q wave. The termination of the T wave can be indistinct; therefore care must be taken to separate the terminal part of the T wave from a U wave or, in sinus tachycardia, from a P wave. In sinus rhythm, one should average the QT interval for at least three successive cardiac cycles; in arrhythmias such as atrial fibrillation, at least ten cycles should be measured.

Rate, age and sex affect the QT interval with females having slightly longer normal limits than males and children. The interval varies inversely with rate. Tables are found in nearly all electrocardiographic texts for determining the rate corrected QT interval (QTc). As a general rule, at normal sinus rates, the QT should be less than one-half the preceding R-R interval. Characteristically, the T waves associated with a prolonged QT interval are described as being either notched or bifid in configuration as in figure 2.

The etiology of ventricular fibrillation with a prolonged QT interval is unclear. Most investigators consider it to be secondary to an imbalance of sympathetic stimulation or innervation of the heart. Often, those affected have sinus bradycardia. During exercise, they prolong their QT but do not appropriately increase their rate, thus increasing the duration of the vulnerable period for the initiation of ventricular arrhythmias. Torsades de pointes is frequently associated with a prolonged QT. It is prefibrillatory and represents a transition between ventricular tachycardia and fibrillation. The rhythm is a multiformed, polymorphous tachycardia in which the axis and amplitude of the QRS complexes vary from beat to beat at a rate of 100 to 300. Almost invariably, it is initiated by a premature ventricular contraction superimposed on a long, broad T wave (R - on - T phenomenon). The rhythm can either spontaneously cease or degenerate into fatal ventricular fibrillation.

Treatment includes high thoracic left stellate ganglionectomy and administration of beta blockers, both of which shorten the QT interval. Beta blockade must be aggressive and complete, often to the point where cardiac pacing is required to maintain an adequate rate due to the secondary bradycardia. Missing a single dose has been described as precipitating sudden death in those with a history of syncope.

Our applicants present a diverse spectrum of the prolonged QT interval. A middle aged female, QT interval moderately prolonged with an excellent family history; a young male, QT interval at the maximal normal limit with a dreadful family history; and a young female, QT interval markedly prolonged, who has a family history of sudden death and who, herself, has survived a cardiac arrest. The prolongation of the QT interval in each appears to be congenital. The first applicant appears to be a standard risk on the basis of her age, asymptomatic state and family history. The second is highly questionable as an acceptable insurable risk. If he remains asymptomatic, he might become insurable at some undefinable time in the future. Although both drug and surgically induced reduction of the QT interval do appear to decrease mortality, I would consider the third applicant an uninsurable risk.

References
Figure 1. The sinus rate is 84 beats per minute with the QT interval 0.44 seconds (upper limit of normal 0.376).
Figure 1. (continued)
Figure 2. There is sinus arrhythmia and bradycardia at an average rate of 45. The QT interval is 0.47 seconds (upper limit of normal 0.47 seconds). The T waves are typically bifid in the precordial leads.
Figure 3. Sinus rhythm is present at a rate of 63 beats per minute. The QT interval is 0.50 seconds (upper limit of normal 0.423). The T waves are slightly biphasic in the lateral precordial leads.