Letter to the Editor

Short P–R intervals and tachyarrhythmias in Fabry's disease

Sir,
The new report (Efthimiou et al., 1986) on brothers with Anderson-Fabry disease first published in 1966 (Johnston et al., 1966, family B), describes some interesting features of the disease. Although paroxysmal tachycardia is not unknown even in affected females (Erdmann et al., 1980) it is certainly less common than atrial fibrillation. A short P–R interval occurs in about 15% of adult males and intracardiac pacing has shown rapid conduction through the A–V node (Matsui et al., 1977). We published the ECG of an affected female whose rhythm abnormalities included a short P–R interval (Wise et al., 1962) and have since seen a woman with Anderson-Fabry disease and intermittent Wolff-Parkinson-White syndrome.

Curved turtle-back shaped nails with pale nail beds are not uncommon in this disease and were present in the brothers reported by Dr Efthimiou and his colleagues when they were seen by me over twenty years ago. The fingers in Anderson-Fabry syndrome are sometimes described as clubbed (Dempsey et al., 1965; Francois et al., 1968; Spence et al., 1978) but the subungual tissues usually lack the thick springy feel commonly found in clubbing; similar nail changes develop in fucosidosis with angiokeratomata. Peripheral circulatory studies in Anderson-Fabry disease show abnormal prolongation of vasoconstriction following minor stimuli (Dempsey et al., 1965; Seino et al., 1983). Some of our patients have limited finger extension and the appearance suggests scleroderma; rarely, the terminal phalanges are shortened (Flynn et al., 1972; Gosling, 1979).

The manifestations of Anderson-Fabry disease seem infinite!

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References


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