Prolonged P-R interval in a male with 47,XYY karyotype

SUMMARY A 47,XYY male with an extremely prolonged P-R interval (0.42 sec) on the electrocardiograph is described. There was also a secondary R wave in lead V₁. He had no past history of heart disease and no cardiac abnormality on physical examination.

In 1968, Price reported a prolonged P-R interval in electrocardiograms of 47,XYY males. Later, he and his co-workers confirmed this finding in a larger sample (Price and Wilson, 1971; Price *et al.*, 1974). However, several authors failed to support this finding (Noël *et al.*, 1969; Steiness and Nielsen, 1970; Char and Borgaonkar, 1971; Borgaonkar, 1972). Thus, the length of the P-R interval in 47,XYY males remains controversial. We wish to report a 47,XYY male whose P-R interval was extremely prolonged.

Case report

The case was one of five 47,XYY males identified in an incidence study among juvenile delinquents (Nanko *et al.*, 1979). The parents were both 29 years old when he was born and were cousins. The patient, aged 18 years when identified, was the sixth in a sibship of six; he was a healthy man, 183 cm tall, and weighed 57 kg. He had no past history or symptoms of heart disease. Physical examination including pulse, blood pressure, cardiac impulse, and heart sounds were all normal.

A conventional 12 lead electrocardiogram (leads I, II, III, aVR, aVL, aVF, V₁₋₆) was carried out. The P-R interval was prolonged (P-R = 0.42 sec). The normal range for Japanese adults is 0.14 to 0.18 sec. There was a small secondary R wave in lead V₁, but no notching or reduction in size of the S wave in lead V₁. The electrocardiogram is shown in the Figure.

Discussion

Since the normal range of the P-R interval in

Fig. ECG record of case with 47,XYY karyotype.

Japanese adults is 0.14 to 0.18 sec, 0.42 sec is thought to be exceedingly rare in this population. Price (1968) suggested that a prolongation of the P-R interval, secondary R in lead V_1 , and a reduction in size with notching of S in V_1 were characteristic features of the electrocardiogram in 47,XXX males. The first two of these features were observed in the present case. There was no evidence of a cardiac abnormality to account for the electrocardiographic findings. More study is needed to clarify whether prolongation of the P-R interval in 47,XXX males is more than a coincidence.

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A case of complete testicular feminisation and 47,XXX karyotype

SUMMARY A very rare case of complete testicular feminisation with a 47,XXX sex chromosome complement is described. The X-chromatin is positive. The subject studied, who belongs to a family in which four other members have Morris's syndrome and have a 46,XY karyotype, is a perfect phenotypic female. The endocrine situation is unique and resembles, in part, that of subjects with Klinefelter's syndrome.

While there are many published reports on complete testicular feminisation with a 46,XY karyotype, it is rare to find the same condition with a 47,XXX complement. The report of German and Vesell (1966) describes Morris's syndrome and a 47,XXX karyotype in monozygotic twins, and Bartsch-Sandhoff *et al.* (1976) reported a case of incomplete testicular feminisation with the same chromosome complement. Subjects with the same syndrome and mosaicism of the sex chromosomes are very rare (Uozumi *et al.*, 1967; Gordon *et al.*, 1969). We report the clinical, endocrine, and cytogenetic findings of a subject with a rare coexistence of Morris's syndrome and 47,XXX karyotype.

Case report

The proband, 35 years old, the eighth of nine children, is one of five subjects with complete testicular feminisation in two successive generations of the same large family (Fig. 1). At the time of her birth her non-consanguineous parents were 40 years old (mother) and 48 years old (father). She is a phenotypic female with primary amenorrhoea, attractive, and married. At 14 years of age she was operated on for a right inguinal hernia, a frequent finding in this condition. Height (167 cm) and body proportions are normal and similar to those in other subjects with Morris's syndrome. Facial, pubic

Fig. 1 Pedigree of family

● Complete testicular feminisation (46,XY)

○ Complete testicular feminisation (47,XXX)