FIG 2 Sequential Q- and C-banded metaphase showing enlarged secondary constriction (h) region of one chromosome 16 and complete inversion of one chromosome 9.

The variability of the h regions can not be determined precisely when G bands are present. Consequently, this defeats the object of the exercise.

In our experience, the Q-C sequence is the most satisfactory and highly reliable (fig 2).

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References

- de la Maza M, Sanchez O. Simultaneous G and C banding of human chromosomes. *J Med Genet* 1976;13:235-6.
- Coming DE, Avelino E, Okada TA, Wyandt HE. Mechanism of G and C banding of chromosomes. *Exp Cell Res* 1973;77:469-93.
- Lubs HA, McKenzie WH, Patil SR, Merrick S. New staining methods for chromosomes. In: Prescott D, ed. *Method in cell biology*. vol 6. New York: Academic Press, 1973;345-80.
- Rubenstein CT, Verma RS, Dosik H. Centromeric banding (C) of sequentially Q- and R-banded human chromosomes. *Hum Genet* 1978;40:279-83.

Familial X-linked mental retardation with an X chromosome abnormality and macro-orchidism

SIR,

Two forms of X-linked mental retardation have been described, one associated with an X chromosome which has a fragile site at Xq27 or 28,^{1 2} and the other associated with macro-orchidism.^{3 4 5} As a result of measuring the external genitals and calculating testicular volumes of retarded males with the fragile site at Xq27 or 28, Sutherland and Ashforth⁶ have suggested that these two forms of mental retardation are the same entity.

We have recently measured the genitals and calculated testicular volumes of some of the retarded males with fragile sites at Xq27 or 28 who were originally described by Harvey *et al.*¹ Of the seven males examined (figure, table), six had testicular volumes greater than the 90th centile of Prader.⁷

Turner *et al.*⁸ have independently re-examined the chromosomes, under conditions appropriate for demonstration of fragile sites,⁹ of the males originally described with mental retardation and macro-orchidism. They found that these males do have the fragile site at Xq27 or 28. That finding, in conjunction with the report of Sutherland and Ashforth⁶ and the data presented here, confirm that the two forms of X-linked mental retardation recorded in published

FIGURE Average (left and right) testicular volume of retarded males examined. The 90% line is the centile given by Prader¹ for ages 10 to 20 years. All males aged 25 years or more are shown as >25.

TABLE Genital measurements of males with mental retardation and fra (X) (q27 or 28).

Subject	Age	Penis		Testicular volume (ml)	
		Length	Circumference	Right	Left
Family A					
II.4	54	11.0	8.0	42	24
II.10	46	9.0	8.0	24	32
II.11	44	13.0	7.5	18	21
III.9	17	10.0	8.5	38	38
Family B					
III.1	23	10.0	9.0	39	58
Subject TK 15		11.0	8.5	37	29
Subject NK 18		12.0	10.0	30	29

Family identification as in Harvey *et al.*¹ Subjects TK and NK are brothers from another family mentioned in their addendum.

reports are the same. Hence, it is now possible to add large testes, as well as small ones, to the list of indications for chromosome studies.

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References

- Harvey J, Judge C, Wiener S. Familial X-linked mental retardation with an X chromosome abnormality. *J Med Genet* 1977;14:46-50.
- Sutherland GR. Heritable fragile sites on human chromosomes. II. Distribution, phenotypic effects and cytogenetics. *Am J Hum Genet* (in press).
- Turner G, Eastman C, Casey J, McLeay A, Procopis P, Turner B. X-linked mental retardation associated with macro-orchidism. *J Med Genet* 1975;12:367-71.
- Cantu JM, Scaglia HE, Medina M, *et al.* Inherited congenital normofunctional testicular hyperplasia and mental deficiency. *Hum Genet* 1976;33:23-33.
- Cantu JM, Scaglia HE, Gonzalez-Diddi M, *et al.* Inherited congenital normofunctional testicular hyperplasia and mental deficiency. *Hum Genet* 1978;41:331-8.
- Sutherland GR, Ashforth PLC. X-linked mental retardation with macro-orchidism and the fragile site at Xq27 or 28. *Hum Genet* (in press).
- Prader A. In: Labhart A, ed. *Clinical endocrinology, theory and practice*. Berlin: Springer-Verlag, 1974:1036.
- Turner G, Gill R, Daniel A. Marker X chromosomes, mental retardation and macro-orchidism. *N Engl J Med* 1979;299:1472.
- Sutherland GR. Heritable fragile sites on human chromosomes. I. Factors affecting expression in lymphocyte culture. *Am J Hum Genet* (in press).

Covesdem syndrome

SIR,

I am writing to point out the marked similarity between the patients reported by Wadia *et al.*¹ and subjects with the Robinow 'fetal face' syndrome.² The features described in the patients reported by Wadia *et al.*¹ namely hemivertebrae, rib defects, mesomelia, short stubby fingers, hypertelorism, depressed nasal bridge, and teeth anomalies are all features of the Robinow syndrome, as is the small penis reported in case 1. There also appears to be marked overall facial similarity to the Robinow syndrome, although the article does not contain good close-up photographs.

The x-ray changes in the spine, ribs, and forearms are all entirely consistent with the Robinow syndrome; in particular, the changes at the elbow in fig 6 are almost identical to the changes shown in fig 4 in an article by Wadlington *et al.*³ which describes four cases of the Robinow syndrome. Although an autosomal dominant inheritance was suggested in the original description of the Robinow syndrome⁴ on the basis of two affected generations, Wadlington *et al.*³ described a sibship containing two affected subjects with normal parents and postulated autosomal recessive inheritance with occasional manifestation in the heterozygote. The mode of inheritance in the family reported by Wadia *et al.*¹ would be consistent with this theory.