LETTERS TO THE EDITOR

Genes for intelligence on the X chromosome

The delightful letter by Turner and Partington\(^1\) proposed that (1) genes associated with non-specific mental retardation determine human intelligence, (2) they are located primarily on the X chromosome, and (3) they were selected in polygynous males. A strict interpretation of their first point would require that all loci influencing intelligence have at least one allele causing non-specific mental retardation. They define non-specificity to mean “no somatic changes, no recognisable metabolic abnormalities, no other neurological signs, and no progression with age”, which is therefore a failure to recognise an undiscovered abnormality. This point of their proposal cannot be confirmed nor disproven.

The number of autosomal recessive loci causing mental retardation has been estimated by two independent methods to be about 325, most, but not all, of them meeting the necessarily imprecise criteria for non-specificity.\(^2\) The ratio of DNA in the X chromosome and autosomes is known from cytometry to be 0.054,\(^3\) and so by chance there should be about 17 loci on the X chromosome at which mutations for retardation may arise. With a Poisson distribution the standard error of this estimate is about \(\sqrt{17}\), giving an upper limit of at least 25 X linked loci for retardation. Although new loci are still being discovered, it is not clear that the number of X linked loci (at present estimated at three) will exceed chance. The astonishingly high frequency of the fra(X) syndrome has no implication for the number of X linked loci that can cause retardation.

With respect to the third point, the model of a harem won by “exhaustive combat between adult males” has its charms. However, if these hypothetical conquests and rewards depended on what is now recognised as intelligence, sexual dimorphism for intelligence should be as great as for size. This is not the case in man or other mammals, raising children must have required as much intelligence as bashing other chaps or species.

Although Turner and Partington must be thanked for an attractive hypothesis, a special relation between the X chromosome and genes for intelligence, in the past or at present, is not supported.

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Another human homologue for the mouse mutant disorganisation

I read with interest the paper by Lowry and Yong\(^4\) in which two brothers with cleft lip and palate, sensorineural deafness, and sacral lipoma were described. In addition, aberrant digits and an anterior sacral meningocele were found.

Recently I had the opportunity to investigate a male newborn with similar features. He was the first child born to non-consanguineous Caucasian parents. The father had unilateral postaxial polydactyly, which was removed in the neonatal period. The family history was otherwise unremarkable. The mother denied any exposure to teratogenic agents during pregnancy. The boy was born prematurely (36 weeks), weighing 2500 g (25th centile). He had an occipital aplasia cutis, 2 cm in diameter. The underlying calvarium was absent, but there was no bulging encephalocele. Furthermore, he had a right sided cleft lip and palate, an absence of the right ear, aplasia of the abdominal musculature, slight flexion contracture of the right knee, and a right club foot. On the medial side of the left upper arm a 2 cm long, digit-like appendage without a nail was present.

The skull defect was closed on the first day of life and the appendage was removed. Histologically it consisted of normal cutis and subcutis without cartilage or bone. Further investigations, including radiography of the whole body, CT scanning of the brain, renal sonography, ophthalmological investigations, and karyotyping of lymphocytes showed no other anomalies. Auditory hypolimniasis evoked potentials were normal. He had no somatic problems, especially no obstipation. Subsequent growth and development were normal.

This unusual combination of symptoms resembles those described in the sibs by Lowry and Yong,\(^1\) and is similar to the manifestations in the mouse mutant disorganisation.\(^1\) Another possible case of a human homologue of this mouse gene was recently described by Viljoen and Kidson,\(^2\) as pointed out by Winter and Donnai,\(^3\) and by Naguib et al.\(^4\) Several other cases can be found in older publications.\(^5\)

The postaxial polydactyly in the father of the present boy may be coincidental, but may also be a clue to an autosomal dominant pattern of inheritance with variable expression. The presence of anomalies in two sibs without visible symptoms in the parents, as described by Lowry and Yong,\(^4\) may be explained by germinal mosaicism or non-variability, as was reported in the mouse mutant.\(^6\)

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9 Licetus F. De monstris. Padua, 1668.