were six mosaics, all females, most with a 45, X cell line. No XYY males were included in the study. All subjects and controls have participated in a longitudinal study of growth and development as well as a psychiatric interview and standardised tests of general intelligence and functional academic skills. The findings make interesting reading. From the point of view of intellectual achievement, people with 47,XXY and 47,XXX karyotypes showed mean IQ scores 21 points and 26 points lower respectively than the control sibs. The group of girls with Turner's syndrome and variants seemed to split into two groups with one group functioning intellectually in the normal range and one group with significant intellectual difficulties. It is not clear whether IQ correlated with karyotype in these two groups. The mosaic group was indistinguishable from the control group with a mean IQ on the 50th centile. Seven of the 47,XXY boys had significant psychological problems on testing compared to 2/13 controls. Difficulties encountered included frustration, low self-esteem, depression, and conduct disorder. Four had had encounters with the police. Three boys had not encountered any significant problems and the authors commented that these three boys came from the most supportive families. The 47,XXX group had the most psychological problems. Depression was the most common feature. They also tended to be socially immature and easily led. Four of these girls became pregnant while at high school and several were involved in drug and alcohol abuse. Three girls experienced fewer problems. Unlike the Klinefelter males, these were not necessarily the girls with the most stable family backgrounds. The girls with Turner's

syndrome tended to be socially reticent, partly owing to their physical self-image. Psychiatric disturbance in this group was rare although the girls found difficulty in separating from their parents. They had fewer romantic relationships and were later in establishing these. Subjects from all groups described strong heterosexual orientation. This paper appears to go along with findings from the discredited early studies in suggesting that extra or missing sex chromosome material has a significant effect on intellectual development and psychological well being. If results of other prospective studies confirm this then there are considerable implications for genetic counselling of these families, especially in the situation where a SCA has been discovered fortuitously on antenatal screening. The outcome of 47,XYY subjects is perhaps even more interesting and remains to be addressed.

JILL CLAYTON-SMITH

Fibrosing colonopathy in cystic fibrosis: results of a case-control study

Smyth RL, Ashby D, O'Hea U, *et al. Lancet* 1995;346:1247-51.

The prognosis in cystic fibrosis (CF) has improved greatly with changes in therapy in the recent past. Along with improved life expectancy there has been the recognition of a previously unreported complication of cystic fibrosis. Fibrosing colonopathy was first described in 1994, with either fibrotic strictures or more extensive fibrosis of the colon. Smyth *et al* have undertaken a case control study of UK cases to investigate possible associations.

Contact was made with physicians responsible for patients with CF known to the UK CF Survey (an independent register of patients). Fourteen cases of fibrosing colonopathy were identified after examination of operative pathology samples (milder cases may have been missed as they may not have required surgery). Four matched controls for each case were chosen from the UK CF Survey. Clinical notes were examined and structured interviews carried out with the patients and their families. All the cases of fibrosing colonopathy occurred after April 1993. The condition occurred in 1.2% of boys (and 0.13% of girls) between 2 and 7 years. The presentation and course of the CF was similar in cases and controls. An association was shown with the use of certain high dose pancreatic supplements, predisposing to the development of the condition. The effect was dose related. Hypotheses to account for this effect are discussed, the most favoured being that the kinetics of constituent release and absorption from these preparations is impaired, and high doses of enzyme are delivered into the colon where they are not usually present. There may be other predisposing factors including other gastrointestinal disease. In a short report in the same edition of the *Lancet* (*Lancet* 1995;346:1265-7), Croft *et al* describe a method of assessing subclinical inflammation of the bowel in CF children. In two children taking high dose enzyme supplements there was evidence of inflammation. Both children had, in addition, evidence of distal intestinal obstruction syndrome. These preliminary results suggest the need for further assessment of the causes of this complication of CF.

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