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An XYY Man

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SUMMARY This case report describes a young man with a 47, XYY karyotype who was convicted of arson. He suffered from a cardiac disorder which may well have been part of the XYY syndrome rather than a chance association. His abnormal karyotype was disclosed in court and used by the defence in a plea in mitigation.

Case Report

Although with hindsight it can be maintained that a full description of Mr S's medical history should start with his conception, the first documented abnormality occurred during delivery. The umbilical cord was twisted around his neck and he was born cyanosed.

His subsequent development was normal and there were no significant medical or psychiatric problems in the family. He was a warm affectionate child but his behaviour at school caused concern. He bullied other children and was physically aggressive towards his teachers. He truanted frequently. When he was nine years old he was referred to a child guidance clinic, but the family stopped treatment.

Mr S's aggressive behaviour continued into his adolescence and after he had left school, making it difficult for him to hold down jobs. He made reasonable peer relationships and also had a steady girlfriend who, it seemed, found his behaviour rather exciting.

In 1974, when he was nineteen, Mr S began to suffer from episodes of sudden loss of consciousness. These were usually preceded by dizzy feelings and abdominal pain. Alcohol sometimes acted as a trigger. The loss of consciousness was brief, never exceeding two or three minutes, but on recovery Mr S often behaved aggressively, though he seemed afterwards not to remember this.

Despite suffering at least two syncopal attacks a month Mr S did not seek medical advice until 1976 when he was referred to a neurologist. On examination he was 190 cms in height and heavily built. His pulse was 44 beats/minute but there was no other physical abnormality. Standard and sleep EEG recordings showed no abnormal activity. A standard 12 lead ECG was within normal limits with a PR interval of 0.18 secs. The exercise and 24 hour ECG's, however, showed multiple ectopic beats consistent with sinus node disease. It was concluded that Mr S's syncopal attacks were caused by this disorder of cardiac rhythm. His aggressive outbursts, it was suggested, might result from brain anoxia caused by poor cerebral perfusion.

Accordingly an endocardial pacing system was implanted but over the next year Mr S's course was complicated by pacemaker failure and bilateral axillary vein thromboses. Eventually, satisfactory pacing under anticoagulant cover was established. Despite this, Mr S continued to suffer from syncopal attacks and aggressive behaviour, though the frequency was reduced. During September, 1977, he was brought to King's College Hospital on three occasions because of his aggressive behaviour. The third time, having badly damaged an ambulance and thrown oxygen cylinders around the casualty department, he was referred to the duty psychiatrist. Mental state examination showed that Mr S was orientated and coherent. He was not deluded or hallucinated and there was no disturbance of short-term memory. He smelled of alcohol and claimed to have fainted prior to the aggressive episode. Further investigations were arranged and chromosome analysis using Giemsa banding revealed that Mr S had a chromosome complement of 47, XYY. Psychometry showed him to be of average intelligence.

Mr S's relationship with his girlfriend had been deteriorating and he was drinking more heavily. A few weeks later, after a drinking bout, he burgled an unlocked car and set it on fire. He attempted to put out the flames but was overcome by smoke. When questioned by the police he denied starting the fire.

In January, 1978, Mr S applied unsuccessfully for work in a factory. A few days later, after quarrelling with his girlfriend and again drinking heavily, he made his way back to the factory and started a fire. He called the police and when they arrived helped them extinguish the flames. Once again he denied causing the fire.

Later that month Mr S started work in another factory but was quickly dismissed. When informed of this he lost his temper and smashed some equipment before leaving. He was charged by the police with causing criminal damage. At the same time he confessed to the two offences of arson which he had previously denied.

The trial

Mr S. appeared in court charged with arson to which he pleaded guilty. A psychiatric report, made at the request of his solicitors, drew attention to his XYY karyotype and to the correlation between this abnormality and a propensity to irresponsible, poorly controlled

behaviour. The report pointed out, however, that such behaviour was not an inevitable consequence of the XYY karyotype, and that long-term psychiatric support could contribute to Mr S's ability to cope with his aggressive impulses.

Initially the Judge took the view that if the patient's behaviour could be accounted for, at least to some degree, by a pre-existing physical abnormality, then the best disposal would take full account of all the medical and psychiatric problems. At a subsequent hearing, however, a medical report was produced which was taken as indicating that the connection between the XYY karyotype and behavioural disturbance was conjecture and not relevant in sentencing. Mr S. was then sentenced to five years in prison.

Discussion

Price (1968) examined the ECG's of a series of XYY males. Compared to controls the XYY men had prolonged PR intervals and a high incidence of partial right bundle branch block. The PR interval in normal males is longer than in females and Price thus argued that the presence of an extra Y chromosome further prolongs atrioventricular conduction time. Not all Price's findings have been confirmed in other series (Steiness and Nielsen, 1970; Char and Borgaonkar, 1971) but Noel *et al* (1969) found a corresponding incidence of partial right bundle branch block. This abnormality has also been reported in a pair of 47, XYY monozygotic twins (Rainer *et al*, 1972). In addition Vianna *et al* (1972) had some success in finding XYY males through ECG screening.

From this it would appear that the disorders of cardiac conduction found in XYY males are bound up with their abnormal karyotype and are not a chance association. The disorders so far described have implicated atrioventricular and right bundle branch conduction, and this is the first report of an XYY man with sick sinus syndrome. Once again, however, a disorder of cardiac conduction is involved, the lesion in this case occurring at an earlier point in the conduction pathway. Why Mr S. continued to have syncopal attacks after pacemaker implantation is uncertain. It was difficult to exclude paroxysmal tachycardia, and clearly alcohol facilitated both the syncopal episodes and his aggressive behaviour.

The XYY syndrome can present difficulties for the psychiatrist preparing a court report. Should the abnormal karyotype be disclosed, and if so to what purpose? Pitcher has concluded that 'criminal responsibility is assessed on the evidence of a man's state of mind to the assessment of which a knowledge of his karyotype adds nothing' (Pitcher, 1971). Although the XYY syndrome does not affect fitness to plead, it may be more relevant when psychiatric evidence is given after conviction as part of a mitigation plea.

The incidence of the XYY karyotype among live male births has been between one and two per thousand in most studies (Pitcher, 1975; Sergovitch *et al*, 1969; Ratcliffe *et al*, 1970); whereas far higher levels, in the order of 20 per thousand, have been reported among groups of offenders (Hook, 1973). There would therefore appear to be a significant association between the XYY karyotype and criminal behaviour. There are, of course, many men with this karyotype who lead blameless lives, and what has been demonstrated is an association, and not a direct causal link. In the same way, personality change and a blunting of social sensibility and ethical standards may occur in the context of brain damage, but associated criminal behaviour is infrequent. Few psychiatrists, however, would hesitate to bring the possible relevance of such brain damage to the attention of a court despite the rarity of the association and the obscurity of the mechanisms involved.

The evidence suggests that an XYY karyotype predisposes an individual to develop a particular type of personality disorder. As in Mr S's case, associated congenital abnormalities may also be present. The crimes with which Mr S. was charged were one more event in a life-long propensity to explosive acts with trivial or obscure precipitants. It seems reasonable to propose that his abnormal karyotype may have some relevance to this situation.

When recommendations are made in psychiatric court reports a multiplicity of factors are

taken into account. The XYY abnormality seems to deserve its place among them.

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