——, RISCH, N., HAMBURGER, R., *et al* (1987) Genetic linkage between X-chromosome markers and bipolar affective illness. *Nature*, **326**, 289–292.

BERRETTINI, W. H., GOLDIN, L. R., GELERNTER, J., *et al* (1990) X-chromosome markers and manic depressive illness. *Archives of General Psychiatry*, **47**, 366–373.

GERSHON, E. S., TARGUM, S. D., MATTHYSSE, S., *et al* (1979) Color blindness not closely linked to bipolar illness. *Archives of General Psychiatry*, **36**, 1423–1430.

GIANNELLI, F., ANSON, D. S., CHOO, K. H., *et al* (1984) Characterisation and use of an intragenic polymorphic marker for detection of carriers of haemophilia B (factor IX deficiency). *Lancet*, *i*, 239–241.

MCGUFFIN, P. (1988) Major genes for major affective disorder? *British Journal of Psychiatry*, **153**, 591–596.

MENDLEWICZ, J. & FLEISS, J. L. (1974) Linkage studies with X-chromosome markers in bipolar (manic–depressive) and unipolar (depressive) illness. *Biological Psychiatry*, **9**, 261–294.

——, LINKOWSKI, P., GUROFF, J. J., *et al* (1979) Color blindness linkage to manic–depressive illness. *Archives of General Psychiatry*, **36**, 1442–1447.

——, SIMON, P., SEVY, S., *et al* (1987) Polymorphic DNA marker on X-chromosome and manic–depression. *Lancet*, *i*, 1230–1232.

RISCH, N., BARON, M. & MENDLEWICZ, J. (1986) Assessing the role of X-linked inheritance in bipolar-related major affective disorder. *Journal of Psychiatric Research*, **20**, 275–288.

SPITZER, R. L., ENDICOTT, J. & ROBINS, E. (1978) *Research Diagnostic Criteria (RDC) for a Selected Group of Functional Disorders* (3rd edn). New York: New York State Psychiatric Institute.

*Michael Gill, MD, MRCPsych, *Research Fellow and Lecturer, Institute of Psychiatry, DeCrespigny Park, London SE5 8AF*; David Castle, MB, ChB, *Research Fellow, Institute of Psychiatry, London*; Conor Duggan, MB, BSc, MRCPsych, *Lecturer, Institute of Psychiatry, London*

*Correspondence

# Anorexia Nervosa in a Patient with XY Gonadal Dysgenesis

SARA E. McCLUSKEY and J. HUBERT LACEY

**This is the first report of a case of anorexia nervosa in a woman with XY gonadal dysgenesis. Anorexia nervosa is a potential complication of gonadal dysgenesis, stemming not only from the disorder itself but from its investigation and treatment.**

*British Journal of Psychiatry* (1992), **160**, 114–116

We report the first case of XY gonadal dysgenesis and anorexia nervosa. There have been 27 case reports of coincident anorexia nervosa and gonadal dysgenesis; the majority of these cases have had XO chromosomes, although mosaic XO/XX chromosomes have been reported.

## Case report

At 17 years, the patient was investigated for primary amenorrhoea. She was below normal weight for her height, felt self-conscious about her 'boyish' figure, and had received, anonymously, a letter calling her a 'freak'. Although the amenorrhoea was initially suspected to be weight-related, on investigation a diagnosis of gonadal dysgenesis with XY chromosomes was made.

The patient was given oestrogen therapy which resulted in significant breast development and weight gain to a maximum of 66 kg at 20 years. She was teased and became preoccupied with her weight and concerned about her prominent 'masculine' jaw, high hips, and big feet. The patient was never told directly that she had a Y chromosome but discovered this on reading her own notes at 18 years while in hospital for the removal of her streak ovaries.

These events coincided with the break-up of her parents' marriage, work stress and, later, sexual harassment by a male flatmate. What began as minor dieting had, by 21 years, become anorexia nervosa. She was 41 kg and, although her weight fluctuated, rising to 47 kg at the time of her only, but short-lived, sexual relationship at 23 years, she lost weight and at the time of presentation she again weighed 41 kg (height 1.74 m; body mass index (BMI) 13.6). She fulfilled the DSM-III-R criteria for anorexia nervosa (American Psychiatric Association, 1987) and had a phobia of normal body weight. She severely restricted her diet, avoiding carbohydrates, and exercised vigorously to maintain her low weight.

During a period of in-patient treatment she was able to discuss in psychotherapy her fears of sexuality. She dreaded appearing masculine and was unsure of her feminine identity. The patient felt that she should have been allowed to discuss these feelings earlier, at the time of the initial diagnosis.

## Discussion

To the authors' knowledge this is the first reported case of concurrent XY gonadal dysgenesis and

anorexia nervosa. There have been a total of 27 case reports of concurrent Turner syndrome and anorexia nervosa. Of these, 15 had XO chromosomes (e.g. Darby *et al*, 1981; Ohzeki *et al*, 1988), six had mosaic XX/XO chromosomes (e.g. Halmi & DeBaulb, 1974; Brinch & Manthorpe, 1987), and in six the exact chromosome analysis was unknown, the diagnosis being made on clinical features (e.g. Kron *et al*, 1977).

Turner syndrome and XY gonadal dysgenesis share a failure of secondary sexual development and primary amenorrhoea. There are, however, some features which are particular to each. Patients with XY gonadal dysgenesis are above average height, with a eunuchoid habitus, while patients with Turner syndrome have slow growth and sometimes a webbed neck, a low hairline, and cubitus vulgaris.

Authors of reports of concurrent anorexia nervosa and Turner syndrome have emphasised the role that these characteristic physical abnormalities may play in the genesis of anorexia nervosa. Our case suggests some of these should be looked at with caution. Short stature did not act as a predisposing factor for anorexia nervosa, as described by Brinch & Manthorpe (1987). Similarly, the suggestions that gonadal dysgenesis results in a 'psychoinfantile' personality (Kihlbom, 1969) and abnormal visuospatial perception (Darby *et al*, 1981) which predispose to the anorexia nervosa must be questioned.

Consideration of the features common to our patient and women with Turner syndrome clarifies how gonadal dysgenesis may predispose to anorexia nervosa. It has been suggested that the exogenous sex steroids used to induce development of secondary sexual characteristics may precipitate anorexia nervosa (Dougherty *et al*, 1983) and psychological preparation for this artificial induction of puberty has been stressed. Certainly, in most cases, including our own, the steroid therapy and anorexia nervosa are temporally associated (Kron *et al*, 1977; Darby *et al*, 1981; Brinch & Manthorpe, 1987; Ohezki *et al*, 1988) and in one case withdrawal of sex steroids resulted in an improvement of anorexia nervosa (Kauli *et al*, 1982).

Our patient appeared immature and different from her peers. She was teased and conscious of her heavy jaw and boyish figure. The discovery of her Y chromosome confirmed her fear that she was a freak and unfeminine. The sudden development of fat tissue on her breasts, bottom and thighs, with weight gain, heightened her anxiety and sexual confusion. She retreated from sexual maturity by dieting, but it was direct sexual confrontation by a flatmate which precipitated the clinical anorexia nervosa. The patient's struggle to come to terms with her sexuality is made graphic by the rise in weight at the time of her first sexual relationship and the drop in weight when this proved unfulfilling. Her ambivalence towards a mature female form is illustrated by her request for silicon breast implants when she was severely emaciated, at 41 kg.

She started to explore some of these issues during in-patient psychotherapy. It emerged that she had always felt different within her family. Her physical problems were superimposed on feelings of insecurity and isolation within her family throughout her childhood. Because of her parents' long-standing marital problems, her mother used her as a confidante during her adolescence, to the extent that the patient viewed her mother as a friend rather than a parent. These psychological factors acted synergistically with the gonadal dysgenesis to nurture feelings of insecurity concerning adult identity and sexuality.

Adverse psychological sequelae in young women with gonadal dysgenesis may be reduced by sensitive management. Oestrogen replacement therapy should keep development of secondary sexual characteristics on a par with that of the peer group (Brook, 1986) to aid psychosexual development and prevent feelings of alienation.

Our patient wished she had been given more information about the chromosomal abnormality so that she would have had the chance to discuss her fears openly. It is difficult for a clinician to decide when, or if, a patient should be told she has a male genotype. Factors which need to be considered include education, curiosity, personality, and family relationships. To minimise psychological morbidity, all patients with gonadal dysgenesis require careful early assessment, and appropriate education about the disorder. Both the patient and her parents need continuing support, with encouragement to join self-help contact groups as outlined by Nielsen (1989) and provided in the UK by the Turner Syndrome Society (c/o The Child Growth Foundation, 2 Mayfield Avenue, Chiswick, London W4 1PW).

Anorexia nervosa is a potential complication of gonadal dysgenesis. The anorexia stems not only from the biological and psychological confusion inherent in it, but also from its investigation and treatment.

**Acknowledgement**

Dr Sara McCluskey is supported by a fellowship awarded by the Priory Hospital, Roehampton, Surrey.

**References**

AMERICAN PSYCHIATRIC ASSOCIATION (1987) *Diagnostic and Statistical Manual of Mental Disorder* (3rd edn, revised) (DSM-III-R). Washington, DC: APA.

BRINCH, M. & MANTHORPE, T. (1987) Short stature as a possible etiological factor in anorexia nervosa. *Acta Psychiatrica Scandinavica*, **76**, 328–332.

BROOK, C. (1986) Turner syndrome. *Archives of Disease in Childhood*, **61**, 305–309.

DARBY, P., GARFINKEL, P., VALE, J., *et al* (1981) Anorexia nervosa and "Turner syndrome", cause or coincidence? *Psychological Medicine*, **11**, 141–145.

DOUGHERTY, G., ROCKWEL, W., SUTTON, G., *et al* (1983) Anorexia nervosa in treated gonadal dysgenesis. *Journal of Clinical Psychiatry*, **44**, 219–221.

HALMI, K. & DEBAULT, L. (1974) Gonosomol aneuploidy in anorexia nervosa. *American Journal Human Genetics*, **26**, 195–198.

KAULI, R., GUREWITZ, R., GALAZER, A., *et al* (1982) Effect of gonadotrophin excretion in a patient with gonadal dysgenesis. *Acta Endocrinologica*, **100**, 363–368.

KIHLBOM, M. (1969) Psychopathology of Turner's syndrome. *Acta Paedopsychiatrica*, **36**, 75–81.

KRON, L., KATZ, J., GORZYNSKI, G., *et al* (1977) Anorexia and gonadal dysgenesis. *Archives General Psychiatry*, **4**, 332–335.

NIELSEN, J. (1989) What more can be done for girls and women with Turner syndrome and their parents? *Acta Paediatrica Scandinavica*, **356** (suppl.), 93–100.

OHZEKI, T., IGARASHI, Y., EGI, S., *et al* (1988) Turner syndrome with anorexia nervosa. *American Journal of Medicine*, **84**, 792–793.

PITTS, F. & GUZE, S. (1965) Anorexia nervosa and gonadal dysgenesis (Turner syndrome). *American Journal of Psychiatry*, **119**, 1100–1102.

Sara E. McCluskey, MRCPsych, *Lecturer, Department of Adult Psychiatry, St George's Hospital Medical School*; *J. Hubert Lacey, FRCPsych, MD, *Reader and Head of Adult Psychiatry Section, Department of Adult Psychiatry, St George's Hospital Medical School, Tooting, London SW17 0RE*

*Correspondence

# Milieu and Mutilation – A Case for 'Special' Treatment?

EMMET P. LARKIN

**Special hospitals are principally concerned with the treatment of patients of dangerous, violent or criminal propensities referred from the courts and prisons. However, patients can be transferred from local NHS hospitals. The case of one such patient illustrates the potential benefits of such a transfer, even for patients who are not of immediate danger to others.**
*British Journal of Psychiatry* (1992), **160**, 116–119

The provision of special hospitals is required by the Secretary of State for Social Services under section 4 of the National Health Service Act 1977. There are three such hospitals in England and Wales – Broadmoor, Rampton, and Moss Side and Park Lane (now one hospital). They provide the same range of therapeutic services as ordinary psychiatric hospitals, but are intended for the treatment of detained patients who, in the opinion of the Secretary of State, require such treatment under conditions of special security because of their dangerous, violent, or criminal propensities.

The majority of patients admitted to special hospitals come from the courts (via a hospital order under section 37 of the Mental Health Act 1983) or on transfer from prison (under sections 47 or 48 of the Mental Health Act), usually after the commission of a serious offence (e.g. murder or manslaughter). Approximately 20% of patients are transferred from National Health Service (NHS) hospitals under section 3 of the Mental Health Act 1983 (civil patients). Many of these patients will not have been convicted of a criminal offence but will have been transferred because of persistently dangerous and unmanageable behaviour in a local hospital.

The following case highlights the difficulties such civil patients can present before transfer to a special hospital; the patient was unusual in that his primary difficulties lay in self-destructive behaviour rather than dangerousness towards others.

## Case report

J was born following a full-term breech delivery. No abnormalities were noted in early development. At school he was noted to be slow at reading, and although he made friends he was teased and bullied as a young teenager. He left school at 15 with no qualifications or ambition.

His leisure activities were mainly solitary in nature (e.g. stamp and coin collecting). He occasionally drank, did not smoke, and experimented with LSD twice in his early teens. After leaving school he worked as a trainee chef for almost two years. Further employment was interrupted by illness.

The patient's father, in his late 60s, is a retired county council clerk. He was a lay preacher with fundamentalist