that she had thromb in the mesenteric vessels similar to those which had occurred in the skin. She was treated with heparin and phenindione, and given intravenous tetracycline. There was an immediate response to this therapy, but on 24.2.62 she developed atrial fibrillation, which was controlled by digoxin. She was discharged on 21.4.62 but a month later was readmitted with pain in the left cheek, and lumbar pain due to spinal osteoporosis. The latter symptom was relieved by a lumbar corset, anabolic hormones and a high calcium diet. She has now been discharged to an old people's home.

Comment

Reports of similar cases are uncommon. Swarts (1942) described two cases affecting the dorsum of the feet in young men. Both showed similarity to our cases in the sudden onset, the active phase when the purple area becomes vesicular, separation of a gangrenous plaque, and healing in six to eight weeks leaving a white scar. However, our cases both had haematological abnormalities at some stage, i.e. hemolytic anaemia in Case 1 and L.E. cells in Case 2.

The cause of the skin necrosis in these cases is obscure. That both manifested auto-immune processes in the blood might indicate that such a mechanism operated to cause the skin necrosis. Skin necrosis has been reported in disseminated lupus erythematosus (Dubois and Arterberry, 1962) and in rheumatoid arthritis without L.E. cells in the blood (Bywaters, 1957) but in these there was arteritis affecting the arteries supplying the necrotic skin and the gangrene was of the common distribution commencing at the tips of fingers and toes. Other members of the collagen diseases such as polyarteritis nodosa, thrombotic microangioopathy (Symmers, 1952, 1956) and thrombotic thrombocytopenic purpura seem unlikely as the disease process involved the venules and not the arterioles in Case 2 and in neither case was the platelet count lowered. Drugs might well be to blame, particularly in Case 2, for methyl thiouracil is known to cause both leucopenia and polyarteritis (Richardson, 1961).

Twenty years ago Swarts wrote of his cases: 'I can only speculate as to this peculiar type of cutaneous gangrene. It may be a trophoneurotic phenomenon . . . ' While auto-immunity may have played a part in our cases in neither is the cause clear; the features do not correspond completely with any known disease and the resemblance of the cases to each other is incomplete. For this reason we have reported them under the descriptive heading 'Acute idiopathic circumscribed gangrene.'

We would like to thank Drs. W. A. Bourne and R. Kemball-Price for permission to publish their cases and Dr. R. I. K. Elliott for his helpful guidance.

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TRANSVESTISM AND FERTILITY IN A CHROMOSOMAL MOSAIC

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Cross dressing or the wearing of clothes appropriate to the opposite sex was called transvestism in 1910 by Hirschfield. It is also known as eunon after Chevalier D'Eon de Beaumont, who was a diplomatic agent of Louis XIV and who lived most of his life as a woman. The problem of transvestism is probably a widespread one, particularly amongst men, as is borne out by Hamburger (1953), who received 1,117 letters from distressed patients throughout the western world with features of transvestism. This followed the publication, in the non-medical Press, of a dramatic case of 'change of sex' treated at his clinic.

Transvestism is known to be linked with fetishism, but the reported frequency of this association varies from author to author. Randell (1959) found only two cases of fetishism amongst 37 male transvestites, while Peabody, Rowe and Wall (1953) underlined the relationship between the two disturbances. Barr and Hobbs (1954) state that transvestism must be distinguished from fetishism. Epstein (1961) suggests the amalgamation of the
diagnoses, talking of fetishism/transvestism, and Lukianowicz (1959) supports this suggestion, stating that the various sexual perversions are closely interwoven. A case of sexual deviation is presented here where the prime disturbance seems to be fetishism, which later developed into transvestism.

Review of the literature reveals considerable variance in the importance attached by different writers to constitutional as opposed to environmental factors operative in such cases. The patient reported here has a mosaic sex chromosome pattern of the Klinefelter type and abnormal hormonal excretion levels: in addition he has experienced environmental difficulties, as will be discussed in the clinical history. Certainly the association between transvestism and Klinefelter’s syndrome as judged by reported cases is very rare.

Case Report

The patient, a 38-year-old man, was admitted to the psychiatric unit of a general hospital by his family doctor because of headaches, which had not responded to treatment with methaminodioxpoxide (Librium). He had also confessed to periodically wearing his wife’s clothing.

He was the third child in a family of five. Initially his mother was the preferred parent, intervening in frequent rows between the patient and his father. He describes his father as a strict, fierce-tempered man, who was often drunk, and when the patient was young he was frequently beaten by his father. In early childhood he displayed several neurotic traits, complaining of stammering when flustered or angry. He was frightened of the dark and had had nightmares with occasional sleepwalking.

After an unremarkable schooling, he worked for 18 months as an apprentice fitter before joining the Royal Navy in 1941 at the age of 16. Five years later he was discharged with C3 grading to a Service mental hospital allegedly, because of homosexual tendencies, although the Admiralty notes showed the diagnosis to be ‘reactive depression in a schizoid personality’. Whilst in the Navy the patient was conscious of the fact that he was attractive to the homosexual members of the ship’s company, and was constantly apprehensive that he would be the subject of their overtures. It was, in fact, an attempt at mutual masturbation initiated by such a homosexual which led the patient to complain to the ship’s medical officer and hence to his admission to the Service mental hospital. Since discharge from the Navy the patient has worked with various firms of painters and decorators.

Psychosexual Development and Practice. At the age of 9 or 10 the patient can remember wearing his sister’s pants and being aware of sexual desire at this time, which was alleviated by masturbation. The masturbation, with associated feelings of guilt and shame, continued, at times daily, until about the age of 18 and then spasmodically until his marriage at 26. There was also a history of self-induced sexual stimulation per rectum with such objects as pencils, pens, and rubber tubing. Puberty appears to have been somewhat delayed as judged by growth of pubic hair, which did not commence until about 18, and shaving began shortly after this. He was self-conscious about this retardation and took elaborate precautions to avoid being exposed in front of others. About this time he complained of swelling in the right nipple, a source of embarrassment to him when sunbathing.

His first heterosexual contact was at the age of 17, when he was introduced to his sister’s girl friend whilst home on leave. A courtship ensued, which extended intermittently (due to leave) over a two- to three-year period; the friendship was of great importance to the patient as he had hoped to marry this girl. However, on returning home, one of his mother informed him that the girl had gone to London on an anticipatory marriage to another sailor. The patient subsequently found out that the girl had not been pregnant at the time of marriage and during the interview he expressed marked hostility towards his mother and his girl friend regarding this episode. Shortly after this he was led into an affair lasting three months with a girl he describes as ‘over-sexed, very coarse and unpleasant’. He felt disgusted and depressed at this time and on several occasions tried unsuccessfully to stop the friendship before he was finally posted to Malta. There, on volunteering for blood donation two weeks later, he was found to have a positive Wassermann. Syphilis was confirmed and he received a full course of penicillin with adequate follow-up. Of that time the patient said: ‘I never wanted to have anything to do with women again; that really put me off them altogether’. This feeling of revulsion was reinforced approximately one year later when he was persuaded to go to a brothel in Cairo. He found the situation so sordid and disgusting that he was unable to consort with the prostitutes. For five years the patient ‘never bothered about women’, until six months before his marriage in 1951.

The marriage followed a six-months’ courtship and three months’ engagement. He regarded his wife essentially as a companion and because of her Christian outlook he considered her a suitable moral prop and practical friend rather than a sexual mate. Prior to the marriage there was some evidence of engyesis, as defined by Davies (1956). During the courtship he felt embarrassed in his wife’s company due to her marked strabismus. The patient had proposed marriage in a romantic setting and for two days afterwards he was unable to work due to anxiety, regretting his decision, but feeling he could not withdraw from his obligation. On one occasion he broke off the engagement because he felt apprehensive about marriage. One week before the ceremony he used unemployment as an excuse for postponing the wedding. While actually in the church he hoped that his wife would be unable to answer ‘yes’ at the appropriate moment. His marriage, however, was satisfactory for two years. Normal marital relations took place approximately three times per week and were mutually satisfactory, although his wife stated that during intercourse the patient always assumed the recumbent position. There are three apparently healthy children, all boys, aged 11, 7 and 3.

For a few months following his discharge from the Navy he began wearing men’s silk underwear and silk pyjamas, enjoying the feeling of the material against his skin. Two years after his marriage he again used silk underwear and nylon stockings, the texture of the material being all-important for the patient. This would occur once every two to three weeks, when he wore the clothes for half-hour periods. Donning the clothes usually spontaneously resulted in orgasm, and if not he resorted to masturbation. Such activity took place furtively, behind locked doors, and was associated with marked feelings of guilt. His wife commented that on shopping expeditions he would stare for long periods at lingerie shops. The need for female underwear became more frequent and persistent. He began wearing stockings, suspender belts, skirts and his wife’s shoes. Finally he used cosmetics, and wearing the above ensemble would admire himself
in the mirror. With progression of the disturbance the desire to become a woman strengthened. He was tempted to try tablets advertised in the gutter press, which suggested that breast development would be enhanced. When dressing in female garb he imagined himself as the passive member or recipient of the sex act, in a non-specific way.

Following a television documentary on homosexuality, the patient became distressed and for the first time confessed to his wife through the medium of a letter. He also requested her co-operation in the purchase of large-size female underclothing. This information caused his wife to become very distressed and bewildered, feeling quite inadequate to fulfil her proper role. She even felt that she could no longer wear her own underclothes! The resultant deterioration in the marriage relationship prompted the patient to seek help. Prior to admission he was wearing female underclothing almost continuously, even during the day under his working clothes. He frequently wore such underclothing in bed at night, although this was not a necessary adjunct to satisfactory sexual relations. At no time was there public display or exhibitionism; in fact fear of discovery by his children was another reason given by the patient for seeking help.

Examination. The outward appearance was of a mildly obese but otherwise apparently normal male. Facial appearance was also unremarkable apart from pre-auricular skin folds often seen in association with hypogonadism, and a rather scanty beard. The patient shaved on alternate days, although it is doubtful if this was truly necessary. The scalp hairline had a fairly normal male distribution. Body hair was almost totally absent with female distribution of pubic hair. His forearms were virtually hairless, although his legs were unremarkable in this aspect. There was no gynecomastia. The external genitalia were essentially normal and, while the testes were at the lower end of the range of normal size, they showed no external evidence of atrophy. Both pubic and axillary hair were easily palpatable. Careful examination failed to reveal any residue of syphilis apart from a faint penile scar.

The patient was given systematic, explorative and interpretative psychotherapy. In addition, to reinforce this, he had a modified course of aversion therapy, as described by James (1962). Apomorphine i/15 to i/10 gr. was administered by intramuscular injection three-hourly. This was coupled with handling of his wife's underclothes. Finally, at times of extreme sickness, he was made to wear these garments under firm and authoritative direction. The patient was unable to tolerate more than 24 hours' treatment and expressed the fear that aversion would extend to his wife when subsequently she wore similar underwear. It was accordingly stopped. Five months later the patient shows no return of symptoms and his various somatic complaints have disappeared.

Physical Assessment of Sex. Reference to Table 1 shows the scheme followed in this investigation. Results of urinary androgen-estrogen excretion levels are shown in Table 2. Buccal smear chromatin pattern showed 6% of cells positive for chromatin, the normal range being 0 to 15% for males. Chromosome pattern was studied on two separate blood cultures and visual analysis of the cells was as follows:

- No. of chromosomes . . . . <45 46 47 48>
- No. of cells (1st culture) . . 2 10 9 —
- (2nd culture) . . . . 6 24 17 —

7-year-old son of patient . . . . 12 2 —

Fertility Studies. Sperm count showed 75% of sperms actively motile within 1½ hours and less than 25% of abnormal forms. With a total number of active spermatozoa in the specimen at 84,000,000 the patient was considered relatively fertile. Blood-grouping studies (Table 3) are consistent with paternity.

EEG: A resting record with hyperventilation and photic stimulation was taken and found to be normal. There was no evidence of temporal lobe dysfunction as has been reported in transvestites and other sexual abnormalities by many writers (Epstein, 1961; Davies and Morgenstern, 1960).

Neither testicular biopsy nor laparotomy was considered justifiable or necessary. W.R. and Kahn: negative.

Table 2

<table>
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<tr>
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<tbody>
<tr>
<td>Androsterone . . .</td>
<td>. . . . 0.44 mg./24 hr.</td>
<td>. . . . 1.88</td>
<td>. . . . 0.77 mg./24 hr.</td>
<td>2.17 ± 1.26 mg./24 hr.</td>
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<tr>
<td>Testosterone . . .</td>
<td>. . . . 0.32 &quot;</td>
<td>. . . . 0.54</td>
<td>. . . . 1.32</td>
<td>2.31 ± 1.35 &quot;</td>
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<tr>
<td>Testosterone . . .</td>
<td>. . . . 1.45</td>
<td>. . . . 2.45</td>
<td>. . . . 0.54</td>
<td>2.31 ± 1.35 &quot;</td>
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<td>Estrogens . . . .</td>
<td>. . . . 238 μg./24 hr.</td>
<td>. . . . 118 μg./24 hr.</td>
<td>. . . . 140 μg./24 hr.</td>
<td>6.9 μg./24 hr.</td>
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<td>Estrone . . . . .</td>
<td>. . . . 57 &quot;</td>
<td>. . . . 61</td>
<td>. . . . 146</td>
<td>3.5 &quot;</td>
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<tr>
<td>Estradiol . . . .</td>
<td>. . . . 0.8-11</td>
<td>. . . . 0.8-11</td>
<td>. . . . 0.8-11</td>
<td>. . . . 0.8-11</td>
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Table 1

<table>
<thead>
<tr>
<th>Anatomical</th>
<th>Functional</th>
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<tr>
<td>1. Nuclear sex (a) Buccal smear (b) Leucocyte 'drumsticks'</td>
<td>1. Gametogenic function (a) Sperm count (b) Testicular biopsy</td>
</tr>
<tr>
<td>2. Chromosomal sex</td>
<td>2. Hormonal function (a) F.S.H. (b) 17-hydroxycorticoids</td>
</tr>
<tr>
<td>3. Gonadal sex</td>
<td>(c) Estrone-estradiol</td>
</tr>
<tr>
<td>4. Gonadal ducts</td>
<td>(d) Estradiol</td>
</tr>
<tr>
<td>5. External genitalia</td>
<td>(e) Androgen</td>
</tr>
</tbody>
</table>
**Table 3**

**Results of Blood Grouping to Support Paternity**

<table>
<thead>
<tr>
<th>Patient</th>
<th>ABO Phenotype</th>
<th>RH Phenotype</th>
<th>Probable Genotype</th>
<th>MN</th>
<th>S</th>
<th>P</th>
<th>Le&lt;sup&gt;a&lt;/sup&gt;</th>
<th>K</th>
<th>Fy&lt;sup&gt;a&lt;/sup&gt;</th>
<th>Jk&lt;sup&gt;a&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>38</td>
<td>O</td>
<td>CCDe</td>
<td>R&lt;sub&gt;r&lt;/sub&gt;R&lt;sub&gt;1&lt;/sub&gt; CDe/CDe</td>
<td>MN</td>
<td>-</td>
<td>-</td>
<td>(++)</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>36</td>
<td>A&lt;sub&gt;1&lt;/sub&gt;</td>
<td></td>
<td>r&lt;sub&gt;r&lt;/sub&gt; cde/cde</td>
<td>M</td>
<td>+</td>
<td>+</td>
<td>++</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>11</td>
<td>A&lt;sub&gt;1&lt;/sub&gt;</td>
<td>CcDe</td>
<td>R&lt;sub&gt;r&lt;/sub&gt; CDe/cde</td>
<td>M</td>
<td>+</td>
<td>+</td>
<td>+</td>
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<td>-</td>
<td>-</td>
</tr>
<tr>
<td>7</td>
<td>A&lt;sub&gt;1&lt;/sub&gt;</td>
<td>CcDe</td>
<td>R&lt;sub&gt;r&lt;/sub&gt; CDe/cde</td>
<td>MN</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>O</td>
<td>CcDe</td>
<td>R&lt;sub&gt;r&lt;/sub&gt; CDe/cde</td>
<td>MN</td>
<td>+</td>
<td>+</td>
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</table>

**Discussion**

There are thus three main features of interest in this case.

(i) The role of chromosomal and endocrinological features as opposed to dynamic factors in the genesis of this 'perversion'.

(ii) The association between chromosomal abnormalities and transvestism.

(iii) The fertility of patients showing mosaic patterns of sex chromosomes of the Klinefelter type.

Raboch, Sipova and Maly (1961), in a study of 600 men with sexual disturbances, found a much higher percentage came from homes with marital discord and they stress the importance of harmonious environment, which was obviously lacking in the early background of this patient. Benjamin (1954) divides transvestites into three groups: (a) primarily psychogenic; (b) intermediate type or psychic hermaphrodite; and (c) somato-psychic transsexualism, where constitutional disturbance predominates. This balanced view is supported by Worden and Marsh (1953), who describe 'a complex psycho-biological project', rather than a simple function of some biological or endocrine factor in the sense of being 'male' or 'female'. Illchmann-Christ (1959) emphasized that the psycho-sexual role is not determined by the chromosome sex. Walter and Brautigam (1958) described for the first time transvestism occurring in a patient who was phenotypically male, but with a female sex chromatin pattern and endocrinological pattern consistent with Klinefelter's syndrome. They point out that in their case and in reviewing the literature no direct connection can be assumed between the sex chromatin pattern and the actual sex role. They suggest that sexual behaviour is decisively formed by educational interests. Such was the viewpoint taken in the management of our case, as it was felt that the contribution of constitutional factors could not be basically altered, but, through psychotherapy, and reinforcement of this by aversion therapy, the patient's attitude might be modified. Over the short follow-up period of five months this viewpoint seems justified. However, this sexual readjustment along more conventional and acceptable lines may well be precarious as the disturbance has been a long-standing one. Besides elements of fetishism and transvestism, there is evidence of trans-sexualism, i.e. the wish to change anatomical sex (Benjamin, 1954), engnosis and perhaps narcissistic tendencies. There was, however, no exhibitionism or definite evidence of homosexual tendencies.

The significance of the androgen-oestrone excretion levels is a matter for speculation. Measurement of urinary oestrogens is notoriously difficult and a biological fluctuation of excretion levels is often found. However, fractionation of urinary steroids in this patient consistently revealed very low levels of androgens and the oestone-oestradiol level of 238 µg./24 hr. is in the range normally found only in adult females. The relationship between the chromosomal pattern and the hormonal disturbance is obviously conjectural.

There have been only three reports of transvestism occurring in cases of Klinefelter's syndrome (Overzier, 1958; Walter and Brautigam, 1958; Money, 1963). This appears to be the first case reported of such a perversion in association with chromosomal mosaic pattern. Barr and Hobbs (1954) studied skin biopsies in five transvestites which showed typical male morphology and they inferred from this that such patients bore the male XY sex chromosome complex. Worden and Marsh (1953) also failed to discover abnormal gonadal status in their series of transvestites. Randell's cases (1959) showed no abnormality of buccal smear chromosome structure. He also failed to demonstrate ketosteroid abnormality. It will be noted that the sex chromatin pattern obtained from buccal smears in our patient was normal and we would make the plea that buccal smear examinations and skin biopsies are inadequate and that full chromosomal studies should be made in suspect cases.

The final unique feature in this case is the fairly conclusive proof of the patient's fertility. As recently as March of this year, Kaplan, Aspillaga, Shelley and Gardener (1963) state that previously no man with Klinefelter's syndrome had been reported as a father of children. They report the case of a 70-year-old man who claimed to have seven children. Two weeks later Lennox (1963), in a letter to the *Lancet*, repudiated this claim of paternity as it had not been validated by blood typing. He reported four cases of Klinefelter's syndrome where fertility had been disproven. However, Froiland and Ulrich (1963) claim a case of 'apparent fertility' in a man with XXY sex chromosome constitution, and Warburg (1962) has a similar case. Reference to Table 3 can leave little doubt that our patient was the father of the three boys, who incidentally bore a strong facial re-
emblance to the patient. The second child, aged 7, whose blood group bears close resemblance to the patient, has shown a preference for playing with dolls, wearing his mother's apron and displays several other features more compatible with the play of a female child. Study of his buccal smear pattern was chromatin-negative, and his blood chromosome pattern was inconclusive, 12 cells showing 46 chromosomes and two cells containing an added X chromosome, giving a total of 47 chromosomes. We would stress, however, that our patient is a chromosomal mosaic and does not fulfill completely the criteria for the diagnosis of Klinefelter's syndrome.

Summary

1. A case of fetishism/transvestism is presented, in whom the psychodynamics and constitutional factors have been considered in some detail with reference to current literature on this problem.

2. The patient showed abnormally low androgen excretion levels and, though fluctuant, high levels of oestrogen excretion.

3. His sex chromosomes showed a mosaic pattern with approximately 45% of cells showing the XXY pattern found in Klinefelter's syndrome.

4. This appears to be the first case of transvestism in such a chromosomal mosaic.

5. The case is also unusual in that it is the first time fertility has been proven in such a patient.

6. The authors support the view of others that environmental influences play a prominent role in the genesis of such cases and, apart from the modification of anatomical sex by surgery and hormone administration which raises difficult problems of medical ethics, it is the most important aspect of treatment.

7. A plea is made that full chromosomal examination should be made in suspect cases and that examinations of buccal smears are not adequate for the exclusion of chromosomal abnormalities.

Thanks are due to Professor J. G. Gibson for permission to publish this case. We are grateful to Mr. D. W. Neill, Department of Biochemistry, Royal Victoria Hospital, Belfast, for his assistance with steroid excretion studies, to Dr. W. R. M. Morton, Department of Anatomy, Queen's University, Belfast, for his assistance with chromosomal studies, and to Mr. J. Graham White, Clinical Psychology Section, Department of Mental Health, Queen's University, Belfast, for his assistance with German translation. We would also like to thank Dr. M. C. Huth for the Northern Ireland Blood Transfusion Service for her assistance with blood group studies.

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Transvestism and Fertility in a Chromosomal Mosaic

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