the same form of intersexualization as described above. Since a familial tendency to this type of male pseudohermaphroditism has been reported (Schneider *et al.*, 1952) one may suppose that the postulated oestrogen hypersensitivity is genetically determined. This explanation will not suffice for the new type, where the oestrogen levels seem to be higher and yet there is little or no mammary development and the vaginal smear appears to be hypo-oestrogenic. In the absence of factual knowledge of the gonadal structure in these patients it is probably better to refrain from speculation on the mechanism of intersexualization.

Summary

Two examples of a new and apparently undescribed type of male pseudohermaphroditism are reported. Both patients presented as eunuchoidal "females" complaining of primary amenorrhoea. They had normal axillary and pubic hair, little or no mammary development, and normal external genitalia, except for marked enlargement of the clitoris in one. The cervix was about normal, but the uterus was rudimentary and the vaginal smear was markedly hypo-oestrogenic. The urinary 17-ketosteroids were normal, and the oestrogen levels seemed to be definitely higher than those found in other cases of primary amenorrhoea. The gonadotrophins were low in one and normal in the other. The chromosomal sex, as determined by blood-film studies, and confirmed by skin biopsies, was male.

The differentiation of this type of male pseudohermaphroditism from the generally recognized variety of "intersex male with purely feminine external genitalia and bodily habitus" and from other relevant forms of hypogonadism is discussed.

I am indebted to Dr. W. M. Davidson for examining and reporting on the blood films, and to Dr. Sylvia Dawkins, who referred Case 1, and Dr. E. E. Pochin, who referred Case 2, for permission to include these cases in the report.

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ADDENDUM.—Since this paper was written R. B. Greenblatt and N. Carmona (*J. clin. Endocr.*, 1955, **15**, 877) have described the case of a 25-year-old tall "female" of eunuchoidal appearance with primary amenorrhoea, failure of breast development, infantile uterus, scanty pubic hair, and markedly enlarged clitoris. The chromosomal sex was male, the vaginal smear was atrophic, the urinary 17-ketosteroids were decreased, and the gonadotrophins were within normal limits. This case resembles very closely Case 1 described above. Laparotomy revealed rudimentary gonads, histological examination of which showed a cortex composed of stroma without follicles and a medulla and hilus containing many "Leydig cells."

The Haemophilia Society, whose president is Sir LIONEL WHITBY, has produced a pamphlet describing its purposes. *The Haemophilia Society: What it is and How it can Help* may be obtained from the hon. secretary, 94, Southwark Bridge Road, London, S.E.1. The society's aims include the provision of fellowship, advice, and help for the individual haemophilic and promotion of the study of the disease. Membership is open to all haemophilics, their families, and friends. The minimum subscription is 5s. a year.

EUNUCHOIDISM IN IDENTICAL TWINS WITH CONGENITAL FUSION OF THE THORACIC VERTEBRAE

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The occurrence of congenital developmental abnormalities in twins, uniovular or binovular, is not unusual, and numerous instances have been reported in the literature. The abnormality may be single, or multiple abnormalities may coexist, and any of the systems may be involved. Endocrine disturbances may occur, and infantilism and pituitary dwarfism in identical male twins have been reported by van Braam Houckgeest and Sanders (1940) and Guillain and Rouzard (1943), in the latter instance accompanied by retardation of intellectual development and the presence of major epilepsy in one of the twins. A careful search of the literature, however, has not disclosed any report of eunuchoidism in identical twins, and the following is therefore of interest.

Case 1

A dustman aged 40 complained of having had painless swelling of the feet and ankles for a fortnight. There was obvious endocrine disturbance, and he was admitted to hospital for further investigation.

He was one of twin brothers. He had always been of large physique, and the brothers were the biggest pupils in their class at school, but were poor scholars and did not reach a high standard before leaving school at the age of 14. He had never shaved. He was unmarried, and had shown no interest in the female sex, but admitted to masturbating about twice monthly with ejaculation, although spontaneous nocturnal emissions had not occurred. He said he drank 12 pints (6.8 litres) of fluid a day and passed large quantities of urine. Tibial osteotomy for genu valgum had been performed when he was aged 16.

On examination he was found to be mentally backward, of placid temperament, very lethargic, unruffled, and easy to please, and was indifferent to his physical condition apart from the recent ankle oedema. He looked younger than his years, and the voice was high-pitched and unbroken. The skin of the face was smooth and devoid of hair, and the axillary and pubic hair was sparse, the latter being of female distribution. There was considerable obesity, the fat being of "buffalo" distribution, with gynaecomastia and colourless striae of the abdomen. The limbs were excessively long and genu valgum was present, but his height was diminished by a gross dorsal kyphosis. The genitalia were of infantile proportions, and the testes, the size of small beans, were fully descended into the scrotum (Fig. 1). The prostate could not be felt on rectal examination.

Examination of the systems revealed no gross abnormality. The heart sounds were normal and the blood pressure was 160/100. There was no optic atrophy and the visual fields were full. Pitting oedema was present over the lower third of both legs.

The following investigations were carried out: haemoglobin, 92% (13.6 g.%); Wassermann reaction negative; blood urea, 24 mg. per 100 ml. Urinalysis: albumin present; pus cells present in moderate numbers; red cells present in large numbers; casts absent; moderate numbers of calcium oxalate crystals seen; culture, no growth obtained. Serum

calcium, 10 mg. per 100 ml.; blood cholesterol, 160 mg. per 100 ml. B.M.R.—14% (normal). Fasting blood sugar, 86 mg. per 100 ml. Blood sugar after ingestion of 50 g. of glucose: $\frac{1}{2}$ hour, 166 mg.; 1 hour, 114 mg.; $1\frac{1}{2}$ hours, 100 mg.; 2 hours, 64 mg. per 100 ml. Urine was free from sugar throughout. E.C.G. was normal. Urinary excretion of 17-ketosteroids, 7.2 mg. in 24 hours. Blood group A₁ Rh positive (see Table I for detailed analysis).

The maximum fluid intake never exceeded 70 fl. oz. (1,980 ml.) in 24 hours, when the output was 90 fl. oz. (2,550 ml.). A seminal specimen could not be obtained, and biopsy of the breast or testis was not performed.

TABLE I.—Blood Grouping

Case	ABO	Rh	MN	S	P	Lu	Kell	Le ^a	Le ^b	Fy ^a
1	A ₁	R ₁ r (CDe/cde)	M	+	++	—	—	—	—	—
2	A ₁	R ₁ r (CDe/cde)	M	+	++	—	—	—	—	—

TABLE II.—Anthropometrical Data

	Case 1	Case 2
Weight	238½ lb. (108.2 kg.)	225 lb. (102 kg.)
Height	66½ in. (168.2 cm.)	67½ in. (171.4 cm.)
Span	71½ in. (181 cm.)	73 in. (185.4 cm.)
Vertex to pubis ..	31 in. (78.7 cm.)	32½ in. (82.5 cm.)
Pubis to ground ..	35 in. (88.9 cm.)	35 in. (88.9 cm.)

Radiography of the skull showed no bone change or evidence of intracranial calcification. The pelvis was large and flattened. The femora were rather massive and the epiphyses had united normally. No abnormality was shown in the hands, apart from increased length of the metacarpals and phalanges. Multiple calculi were present in the left kidney. The report on the vertebral column states: "Well-marked dorsi-lumbar kyphoscoliosis. There is fusion of the

bodies in the mid-dorsal region anteriorly—this is obviously congenital. Osteoarthritic changes also present in the lower dorsal region" (Fig. 2). The anthropometrical data are shown in Table II.

Case 2

The twin brother of Case 1 was then seen: the striking similarity in appearance and bodily configuration is shown

FIG. 2.—Case 1. Radiograph of dorsal spine.

FIG. 4.—Case 2. Radiograph of dorsal spine.

FIG. 1.—Case 1.

FIG. 3.—Case 2.

in Fig. 3. Obesity of the pituitary type was present with gynaecomastia, and there was genu valgum and considerable dorsal kyphosis, though less severe than in the brother. The genitalia were infantile and a left inguinal hernia was present in the scrotum. He did not shave, was unmarried, and displayed no interest in the opposite sex. He, too, was employed as a dustman, and his temperament and mental development closely resembled those of his brother.

Radiography of the dorsal spine showed the same congenital abnormality to be present (Fig. 4). There were no opaque renal calculi.

His blood group was A₁, Rh positive, and is identical with that of his brother (Table I). No other investigations were carried out. The anthropometrical data are seen in Table II.

Family History

The twins are the youngest of a family of 12 children, comprising five males and seven females, and were born when their mother was aged 46. The eldest son had been killed while serving in the first world war, and, so far as could be ascertained, was physically and mentally normal. The remaining two brothers had died, one from meningitis in childhood and the other from convulsions in infancy, and no further medical details are available of them. A sister had died of pneumonia in childhood and another had died in childbirth. The five remaining sisters are healthy and married, with families of from three to six children, of both sexes. They are thus quite fertile. When interviewed they appeared to be of normal intelligence, and each asserted that the twins had always been of a different mental and physical development from the rest of the family. The mother stated that the twins had been abnormal since birth, and the family practitioner had, in fact, told her that they "shared the brains of one." She confirmed that they had fared badly at school, and had been of large physique even in their youth.

The mother and daughters were unaware of any abnormalities resembling the present among cousins, uncles, or any of the 23 grandchildren, and the abnormalities therefore appear to have arisen sporadically rather than have been inherited as a familial trait.

Discussion

In their study of organic nervous disease in identical twins Kinnier Wilson and Wolfsohn (1929) declared "the extreme similarity of certain human twins to each other is strongly in favour of their uniovular origin," and quoted Abt (1924) that "homologous twins usually show marked similarity physically as well as mentally. Not rarely they show the same anomalies and faults of development." They concluded, *inter alia*, that (1) in health homologous twins show a striking similarity of structural, functional, and mental equipment; (2) when anomalies and deformities are present in identical twins they are most probably the result of inherent defects and are always present in both twins; (3) an anomaly of development is always similar and equivalent in both homologous twins; and (4) biologically considered, homologous twins are only one individual; physically they are two (this last is of interest in view of the comment of the family practitioner to the mother).

Among the reasons advanced by Guillaud and Rouzaud (1943) for assuming their twins to be monozygous were their resemblance, the same disturbance in general development and the same abnormalities present in the brothers, and the similarity in their blood group. In view of these statements there can be no doubt that the patients here presented are identical or uniovular twins. They are of the same sex, and the striking similarity in facial appearance and body configuration is obvious from the accompanying photographs. The same skeletal deformity is present in both, and both exhibit gynaecomastia and infantile genitalia. They

are also of the same blood group. The colour of their eyes is the same, as is also the shape of their ears and hands. Their hair is similar in colour and texture, and the hair pattern is the same, the whorl turning in the same direction (so that they are not mirror images of one another). Thus the criteria for the recognition of identical twins formulated by van Braam Houckgeest and Sanders (1940) are also fulfilled. They are not colour-blind (Ishihara method) and do not suffer from anosmia. Their temperaments are similar and both are intellectually retarded. They share the same interests or lack of interests, and are both employed as dustmen, and even work on the same dustcart. Their fingerprints are not identical, but it is known that this is not found in identical twins. We feel justified in concluding that our patients are identical twins.

The nature of the endocrine disturbance presents a difficult problem. In eunuchoidism the primary defect is in the gonads and not in the pituitary. In infantilism the defect is in the pituitary, and the patient is usually thin and subnormal in stature, as the secretion of pituitary growth hormone is also defective. In the two brothers with gynaecomastia reported by Ferriman (1954) there were some features suggestive of eunuchoidism, but the dwarfism and low output of urinary gonadotrophins indicated that their testicular hypoplasia was secondary to a pituitary defect. The differentiation from Fröhlich's syndrome is more difficult, but in this condition the patient is usually subnormal in height and the syndrome is due to a lesion affecting the pituitary-hypothalamic mechanism, which may itself give rise to symptoms, although it does not invariably do so. ("Fröhlich's syndrome" is often loosely applied to a condition Simpson (1948) has renamed "adipose gynandrium," and, clearly, our patients were not suffering from this.)

The presence of diabetes insipidus, at first suspected, was not confirmed, and the excretion of 17-ketosteroids suggests that the pituitary is intact. An estimation of the gonadotrophic hormone in the serum and urine would have been valuable, as this is normal or above normal in eunuchoidism and deficient or absent in pituitary deficiency or Fröhlich's syndrome. Unfortunately, we were unable to do this. Eunuchs, though usually tall, may be of average height. The height of our patients, though average, would have been greater but for the extent of the kyphosis. The span of their outstretched arms is greater than their height, the pubis-to-ground measurement is greater than the pubis to vertex, and the excretion of 17-ketosteroids in Case 1 is on the low level of normal. All these features are in keeping with a diagnosis of eunuchoidism. Their sexual behaviour and outlook, their mentality and passive and accommodating demeanour, also point to this diagnosis.

The nature of the breast changes is worthy of comment. There was no enlargement of the areola or nipple, and the appearance of the breasts suggested a local disposition of fat comparable to the excess of fat elsewhere. In the youth with gynaecomastia and testicular atrophy described by Ferriman (1950) enlargement of the areola and nipple was present, and biopsy of the breast showed hyperplasia of the duct elements and fibrous tissue. He discussed the mechanism whereby gynaecomastia may appear with aplasia of the testes. As no breast biopsy was performed on our patients we are unable to confirm that the breast enlargement is simply due to a local acquisition of fat, or whether hyperplastic changes are present, but, clinically, we believe the former to be the more likely.

The aetiology of the condition is open to speculation. Eunuchoidism is due to a primary defect of the gonads which may follow an infection, such as mumps, or result from trauma or interference with the blood supply to the testes; but the commonest cause of eunuchoidism appears to be genetic (Simpson, 1948). Ferriman (1950) expressed the opinion of Dr. Raymond Greene, that the testicular aplasia was very likely due to a genetic disturbance, though in his later report he thought the dwarfism and low secretion

of gonadotrophin pointed to a pituitary origin for the testicular hypoplasia. In the cases under review, not only has the eunuchoidism to be accounted for, but also the identical skeletal deformity and the mental retardation, which is uncommon in eunuchs, who are usually of normal or above normal intelligence. The three abnormalities may be explained on a single basis if the concept of a degeneration of unknown aetiology in the parental germ plasm is accepted. If the cause of the defect be a matter of heredity in the germ cells, the monovular twins should be alike—either both normal or both defective (Macklin, 1936). It is probably significant that the twins were the last of 11 pregnancies, and at the time of their birth the ages of the father and mother were 44 and 46 years respectively.

The absence of colour-blindness and anosmia from our patients strengthens the belief that the condition has arisen sporadically, as Kallman *et al.* (1944) concluded that these tended to be associated with eunuchoidism only when hypogonadism appeared as a familial trait.

Summary

Eunuchoidism in identical twins is described, together with mental retardation and congenital fusion of the thoracic vertebrae.

Reasons are advanced for the belief that the twins are uniovular and that the condition has arisen sporadically, and the opinion is also expressed that the primary cause for the abnormalities lies in an unknown degeneration of the parental germ plasm.

Our thanks are due to Dr. D. Lehane, Director of the Regional Blood Transfusion Centre, and Mr. W. Donohoe, senior technician, for the investigation and report on the blood groups.

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The National Old People's Welfare Committee, now to be called the National Old People's Welfare Council, "is now established as the accepted focus and centre to deal with the diverse and often difficult problems relating to the welfare of old people," said Mr. HAROLD MACMILLAN last year when he was Minister of Housing and Local Government. The *Progress Report* for 1954 shows that, while the main work of the council in co-ordinating the efforts of both voluntary and government bodies working for old people has continued satisfactorily, other schemes have also advanced. Training courses for voluntary and paid workers, including one for matrons of homes, have been developed. In Birmingham alone 310 voluntary visitors visited 1,735 old people regularly. The King George VI Foundation gift of £80,000 has been used to build new clubs or improve those existing. By September, 1954, 5,000 social clubs for the elderly were reported to exist. A building society has opened a home for the mentally frail in London, and many accommodation problems have been dealt with; interest in boarding-out old people as pioneered in Plymouth and Exeter in 1952 has revived. The council has sponsored several publications to educate the public on the problems of the elderly and how they can best be tackled.

PROLONGED PREGNANCY

BY

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A prolonged pregnancy may be defined as a pregnancy which continues for more than 14 days after the expected date of confinement provided that the estimation of this date is accurate. It occurs with relative frequency in lower animals, and most obstetricians would agree that it also occurs in human species. Many authentic cases have been published, and as examples the cases of Hendry (1921), Wells (1948), and Smith (1951) may be mentioned.

By definition the diagnosis depends upon an accurate estimate of the expected date of confinement. This is obtained by calculation based on Naegele's rule, and the fallacies of this calculation are such that it is difficult to decide in any given case whether pregnancy is really prolonged or whether the calculation is in error. The intelligence of the patient, her accuracy of observation, and the regularity of her menstrual cycle all help in reaching a decision. The size of the uterus in early pregnancy and the date of quickening are also useful aids. In retrospect the weight of the baby at birth may be used as a check on the accuracy of diagnosis. An investigation was undertaken, utilizing these aids to diagnosis, in order to discover the incidence of prolonged pregnancy and the pathological significance of the condition.

The Investigation

A clinical investigation is more satisfactory if all cases are supervised personally, but an investigation into prolonged pregnancy requires such a large series that personal supervision is impossible. Most published reports deal with cases which have been collected from hospital records, a method which has obvious disadvantages. In an attempt to overcome these disadvantages an investigation based on personal interview was undertaken. Approximately 6,000 women were interviewed by me during 1953 and the early part of 1954. These women were patients in the Jubilee Maternity Hospital or the Royal Maternity Hospital, Belfast, and the interviews took place during the first week of the puerperium. The series was consecutive, excluding holidays, and the interviews were continued until 5,000 case records had been obtained.

It will be noted that approximately one in six cases were excluded from the series, but it must be emphasized that these exclusions were made before the fate of the child had been ascertained. The reasons for exclusion were: (1) A fabrication of the "dates" to agree with a marriage early in pregnancy. (2) An uncertain estimate of the date of confinement due to (a) pregnancy commencing during lactation, or (b) prolonged or irregular menstrual cycles, or (c) lack of ordinary intelligence on the part of the patient.

If the duration of pregnancy is being investigated an interview during the first week of the puerperium has many advantages. Firstly, the patient retains a clear knowledge of the date of her last menstrual period, the date of quickening, and her menstrual history prior to pregnancy. Secondly, her case records are available to furnish information about the size of the uterus at various stages in pregnancy. Lastly, the date of confinement, the weight of the child, and the outcome are known. Information which has been obtained in this way may be criticized on the grounds that, as delivery has occurred, the patient may tell her story accordingly. It