Intersex among Africans in Rhodesia

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According to a Greek myth, preserved for us in Ovid and recorded by Rose (1953), Hermaphroditus was the son of Hermes and Aphrodite. A fountain nymph, Salmakis, who fell passionately in love with him, entreated the gods that she might be forever united with him, and her prayer was answered by the combination of lover and loved into a single being – a hermaphrodite.

By common usage the term hermaphrodite in man is loosely applied to individuals combining characteristics of both sexes. The 'true' hermaphrodite is one in whom both ovarian and testicular tissue are present. When both gonads are of one sex but anatomical characteristics of the other sex are present, the term pseudohermaphrodite is used. The number of schemes of classification of intersex that have been proposed is an index of the deficiencies in our understanding of the subject.

Hermaphroditism is fortunately relatively uncommon, and 'true' hermaphrodites are rare. In his book *Die Intersexualität* Overzier (1961) lists 146 hermaphrodites recorded in the world literature since 1900. It is therefore of interest that no fewer than 8 (and possibly 9) true hermaphrodites have been investigated in Rhodesia—all but one in Harari Hospital—during the past four years. We previously reported 4 of these cases (Hammar and Forbes, 1962; Forbes, Clayden, and Hammar, 1962), 2 are awaiting publication, and at least 2 more are reported in this paper. Furthermore it has been brought to our notice that yet another case has been encountered at Mpilo Hospital in Bulawayo (W. Houston, 1963, personal communication). Pseudohermaphrodites are more common, and among 31 cases of intersex investigated over a period of 11 years in Copenhagen by Winkel Smith (1960) there were 17 male and 14 female pseudohermaphrodites and no 'true' hermaphrodites. Female pseudohermaphroditism is usually due to the adrenogenital syndrome, and 13 of Winkel Smith's 14 cases were of this type. In Rhodesia only 4 cases of pseudohermaphroditism have been seen by us, and we have

yet to encounter the adrenogenital syndrome. It is possible that this discrepancy is related to an over-all infrequency of endocrine disorders among our African patients. A somewhat similar incidence in the South African Bantu was found recently by Klempman (1964) who recorded 7 true hermaphrodites, 3 male pseudohermaphrodites, and no female pseudohermaphrodites.

The purpose of this paper is to report 3 further cases of hermaphroditism and to analyse the features of all the cases we have encountered.

Case Reports

Case 1. Dominico was admitted to Ndola Hospital for investigation in May 1960. He had first been seen in the prison because, though he was classified as a male prisoner, he had the appearance of a female.

He had been brought up as a male and at school had been accepted by his fellows as a normal boy. He had taken part in boys' games and had not been interested in girls. At the age of about 14 years he began to develop breasts and female mannerisms. He and his relatives thought that he had been bewitched, but he was still accepted as a male. He obtained employment as a labourer and found that he could work as hard as any of his fellows. He still had no desire to mix with girls and he experienced no sexual feelings towards members of either sex. He was embarrassed by his large breasts and small penis. At the time of his admission to hospital he was in gaol serving a sentence for trespass, and he sought medical help partly on account of the unwanted attentions of other male prisoners.

On examination the general body configuration was female and the breasts well developed (Fig. 1). There was no facial hair and the pubic hair was fine and of female distribution. The voice and mannerisms were feminine in character. The penis was 1.5 in. (3.8 cm.) in length and hypospadiac, and the left side of the scrotum contained a tense hydrocele. No testicle was palpable in the right side of the scrotum nor in the right inguinal region, and no prostate was palpable on rectal examination.

Radiological examination of the pelvis showed it to have an anthropoid type of inlet. Buccal mucosal scrapings revealed the presence of sex chromatin—nuclear sex female.

Laparotomy was performed under spinal anaesthesia. A small uterus 1 in. (2.54 cm.) by 0.75 in. (1.9 cm.) was

FIG. 1.—The general body configuration of Dominico showing the well-developed breasts (Case 1).

FIG. 2.—The small uterus and normal-sized right tube and ovary removed at operation from Dominico (Case 1).

found, but there was no vagina. A normal fallopian tube extended from the right cornu of the uterus to a normal-sized ovary situated at the right internal inguinal ring. There was no intra-abdominal gonad on the left. The position was explained to the patient and, with his consent, the uterus, tube, and ovary were removed (Fig. 2). At a later date the left side of the scrotum was explored and a small testicle 0.75 in. (1.9 cm.) by 0.5 in. (1.3 cm.) was found in a tense hydrocele and was biopsied. Seven days after this operation, the day of the expiry of his prison sentence, the patient absconded from hospital.

Histological examination of the right gonad showed it to be a normal ovary with a number of ripening follicles, and the fallopian tube was also normal. The testicular biopsy showed seminal tubules lined by columnar cells lying in a loose fibrous stroma. Sertoli cells were also present but no spermatozoa. Interstitial cells were numerous.

Management. The patient's sex orientation and his age at the time of investigation clearly indicated that the sex designated by his parents should be maintained. The fact that his nuclear sex was female was of no importance in this respect, and the discovery of a small uterus, right tube, and ovary were of little significance. In retrospect it is unfortunate that bilateral mastectomy was not performed at the time of laparotomy, but it was not anticipated that the patient would abscond.

Case 2. Misheck, aged 6 years, was referred to Harari Hospital in September 1961, for repair of hypospadias. He had been brought up and was accepted as a boy, but his parents were aware that his genitalia were abnormal.

Examination revealed a normal-looking little boy apart from the genitalia. The penis was hypospadiac with the urethral opening at its base. There was no scrotum or vaginal opening, and no testes were palpable in the groins. Examination of blood and buccal mucosal cells showed the nuclear sex to be male, while 17-ketosteroid excretion was normal (1.1 mg. in 24 hours).

Laparotomy was performed on October 6. No uterus was present, but there were bilateral gonads in the position of normal ovaries, and each was distinctly divided into two portions with the macroscopical appearances of ovarian tissue and testicular tissue respectively. Fallopian tubes arose from the vicinity of the ovarian portions of the gonads and ran medially to unite in the midline. It did not seem feasible to bring the testicular portions of the gonads down, so both ovotestes and fallopian tubes were removed. At the same operation the first stage of an hypospadias repair was carried out.

Histological examination. Sections showed bilateral ovotestes. In the testicular portions the seminiferous tubules were immature and no interstitial cells were seen. In the ovarian portions there were numerous primordial follicles and three small follicle cysts.

Management. It was clearly desirable that Misheck should remain in his sex of rearing. Attention has been drawn to the increased incidence of malignancy in intra-abdominal testes, reported by Morris (1953) to be as high as 10% in male pseudohermaphrodites. In view of this and because it did not seem possible to bring down the testicular portions of the gonads, both ovotestes were

removed in entirety. Plastic procedures have been started to fashion as normal and functional a penis as possible. Hormonal therapy will be necessary as he approaches puberty.

Case 3. Possible true hermaphrodite. Maria, aged 2 months, was admitted for investigation in April 1963. The mother had been perfectly well throughout her pregnancy and had not been given hormone therapy. The delivery was normal and though the mother realized that the child's genitalia were abnormal she thought that she was probably a girl. She was afraid to take her child home because the grandmother believed that she (the mother) had been bewitched; she was convinced that the child would be killed at home.

Maria was the 8th child in the family. The 7th child had been born a year previously at the same hospital and at birth had been found to have a marked hypospadias with the urethral orifice at the base of the shaft of the

penis. Testes were not palpable. No investigations were carried out and the child died after a few months during an attack of proven malaria. The mother said that the 6 older sibs were normal, alive, and well.

On admission to Harari Hospital systemic examination was normal apart from the genitalia (Fig. 3). The phallus was large, about the size of a penis, and it had a ventral groove but no orifice. The urethral opening was in the perineum just posterior to the base of the phallus. No vaginal opening was present nor any corresponding perineal dimple. Prominent labioscrotal folds lay on either side, but no testes were palpable. On rectal examination there was a slight midline thickening with tubular structures extending out laterally from it on both sides, and a gonad was palpable on the left.

Examination of buccal smears revealed the presence of sex chromatin, nuclear sex female. The 24-hour urinary oxosteroid excretion was less than 1.0 mg./24 hours. A urethrogram was performed, confirming the presence of a bladder in front and a uterus and vagina behind, with a

TABCE
'True'

Name Age District	Lateral		Bilateral
	Ignatious 9 mth. Rusape	Dominico ± 18 yr. Ndola	Shamiso 1 yr. Fort Victoria
Rearing sex	Male	Male	Female
Appearance	Male	Female, prom. breasts	Female
Nuclear sex	Female	Female	Female
<i>External Genitalia</i>			
Phallus	Hypospadiac penis	Large clitoris	Hypospadiac penis
Urethra	Base of penis	Base of clitoris	Base of penis
Labia or scrotum	Labioscrotal folds	Scrotum	Labioscrotal folds
Testes	Nil	Left only—in hydrocele	Nil
Vagina	Nil	Nil	Present and entirely separate from urethra
Somatic sex	Female	Indefinite	Female
Diagram of external genitalia
<i>Internal Genitalia</i>			
Uterus	Small solid vestigial	Very small	Normal
Vagina	Blind ending below	Nil	Normal
Tubes	Bilaterally present	Right tube only present	Both present and normal
Gonads R.	Testis	Ovary, near R. int. ring	Ovotestis
L.	Ovary	Nil	Ovotestis
Diagram

FIG. 3.—The external genitalia of Maria (Case 3).

common urogenital sinus opening on to the perineum (Fig. 4).

A laparotomy was planned when the child's general condition had improved but before this could be carried out the mother took her home. We subsequently learned that the child died a few weeks later, having been readmitted to St. Theresa's Mission hospital in a state of advanced malnutrition and neglect. Unfortunately consent for necropsy could not be obtained.

It is possible that this child was a 'true' hermaphrodite. Male pseudohermaphroditism is excluded by the absence of palpable gonads, the presence of a uterus and vagina, and a chromatin-positive buccal smear. The adreno-genital type of female pseudohermaphroditism is ruled out by the normal 17-oxosteroid excretion, and the non-adrenal female pseudohermaphrodite, though a possibility, is the rarest type of intersex.

It is unfortunate that full investigation of both Maria and her sib, who was said also to have been abnormal, was not possible. A high family incidence has been noted in

Hermaphrodites

	Unilateral			
Misheck 6 yr. Fort Victoria	Mariani 12 yr. Umvuma	Peter 21 yr. Gatooma	Gift 7 yr. Salisbury	Faustina 24 yr. Mrewa
Male Male Male	Male Male, but has breasts Male	Male Female, prom. breasts Female	Male Male Female	Female Female, pend. breasts Female
Hypospadic penis— small Base of penis Nil definite Nil Nil Male	Hypospadic penis— small Base of penis Labioscrotal folds Nil Nil Female	Hypospadic penis Perineal, post. to base of penis Labioscrotal folds Right gonad at int. ring Nil Female	Almost normal penis but no urethra Perineal, post. to base of penis Labioscrotal folds Nil No external opening but dimple between anus and urethra Female	Normal sized hypo- spadic penis Perineal, at common urogen. sinus Labioscrotal folds Nil Small vagina, opening at common urogenital sinus Female

Nil Nil Both present, connected medially Ovotestis Ovotestis	Rudimentary Rudimentary Both present Ovary Ovotestis	Rudimentary, infiltrated by squamous carcinoma Nil Only right tube identified Ovotestis Ovary	Very small Blind ending below Both present Ovotestis Ovary	Moderately small Small; common urogen. sinus Left tube only present Ovotestis Ovary

FIG. 4.—Urethrogram demonstrating a vagina and uterus in Maria (Case 3).

female pseudohermaphrodites with the adrogenital syndrome, while among male pseudohermaphrodites, Rathbun, Plunkett, and Barr (1958) have pointed out a family incidence in cases with the 'testicular feminizing' syndrome. Among 'true' hermaphrodites, however, no such tendency exists, though Milner, Garlick, Fink, and Stein (1958) reported the condition in two brothers aged 13 years and 13 months, both of whom had bilateral ovotestes. Both also had male nuclear sex.

Discussion

The main features of 8 cases of 'true' hermaphroditism, coming from widely scattered areas of Rhodesia, are summarized in the Table. Many authorities have stressed the importance of early investigation in these cases in order that the most suitable sex may be designated and psychological trauma avoided, and Wilkins (1957) has stated that change of sex after the age of 18 months to 2 years should rarely if ever be considered. In African practice the ideal of early investigation is often impossible to achieve. Indeed, only 3 of our 13 cases presented in infancy. Berg, Nixon, and MacMahon (1963) have expressed the view that there is a place for recommending a sex reversal at a later age, where there is a possibility of achieving a

normal marital relationship; we are in agreement with this view in such cases, and particularly in our African patients who tend to present late. Dewhurst and Gordon (1963) recorded 19 patients in whom the sex of rearing was successfully changed after the age of 18 months, and while they emphasize the importance of early investigation, they feel that careful consideration should be given to changing the sex of rearing where the wisest decision has clearly not been made initially.

In 2 of the 8 true hermaphrodites we have seen, the general appearance of the patient contradicted the sex of rearing: however, both of these were adults, so clearly changes in body contour and development of secondary characteristics had taken place since the sex of rearing had been chosen. Investigation of these cases in infancy would have led us to advise that the child be brought up in the sex other than that designated by the parents, and a great deal of distress and embarrassment in later life would have been avoided.

The nuclear sex of 'true' hermaphrodites may be male or female. Murry Barr, quoted by Wilkins, Grumbach, van Wyk, Shepard, and Papadatos (1955), found that in 12 'true' hermaphrodites 8 had the female pattern and 4 the male. In our cases the ratio had been similar—6 female and 2 male.

The positive diagnosis of 'true' hermaphroditism requires a gonadal biopsy, and before the presence or absence of ovarian or testicular tissue in any gonad can be firmly established, serial sections may be necessary; so, unless both gonads are removed and serially examined, 'true' hermaphroditism cannot positively be excluded.

We have not had the facilities for obtaining chromosome analysis on our patients with intersex in the past, but it is hoped that in the future this investigation will be possible. Klempman and Wilton (1963) did chromosome analyses of 7 'true' hermaphrodites and found 6 to be 46 XX and the 7th to be a mosaic of 2 cell lines—46 XX and 46 XY.

Anatomical features. Three types of 'true' hermaphrodite have been described according to the sites of the gonadal tissue. (1) *Bilateral*, with an ovary and a testis on both sides or bilateral ovotestes; (2) *unilateral*, with testis and ovary (separate or united) on one side and either an ovary or a testis on the other; (3) *lateral*, with testis on one side and ovary on the other. 4 of the 8 'true' hermaphrodites we have seen were of the unilateral type, and in each case an ovary was on the 'normal' side; 2 were lateral, and 2 were bilateral. In the 6 who had ovotestes the ovarian and testicular portions were macroscopically distinguishable, though in juxta-

position, in 4. This is in contrast to the findings of Innes Williams (1952), who stated that in the majority of cases only histology revealed the true nature of the gonads. Ovaries may descend into the scrotum in the same way as normal testes; in none of our cases did this occur, though in one the right ovary lay at the internal inguinal ring. Histologically, maturing graafian follicles may be found in the ovarian portions of ovaries. The testicular portions contain seminiferous tubules, but spermatogenesis is nearly always absent or incomplete. Greene, Matthews, Hughesdon, and Howard (1952) described a Hindu aged 14 years who had an ovary on the left, and a testis on the right, showing all phases of germ cell lineage up to and including spermatozoa. This patient is perhaps the nearest recorded approach to a true human hermaphrodite, i.e. a being capable of self fertilization.

Great variation may be found in müllerian duct derivatives. The most fully developed of our cases had a normal uterus, vagina, and fallopian tubes, while at the other extreme was one with no uterus or vagina. In between these extremes the uterus may show all grades of development, while the vagina may be blind at one or other end, may open into the posterior urethra, or at a common urogenital orifice, or it may be vestigial. Cases of the lateral type sometimes show a vas deferens and epididymis on the side of the testis and a fallopian tube and uterine horn on the side of the ovary.

The phallus is nearly always hypospadiac and it varies in size from a large clitoris to a small penis. In all our 'true' hermaphrodites the urethra was perineal, though cases have been described with terminal phallic urethras.

Summary

The main features of 8 cases of 'true' hermaphroditism encountered over a four-year period are

tabulated and the histories of 3 previously unpublished cases are reported. Some of the problems of intersex are discussed with reference to diagnosis and anatomical features.

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