INTERSEXUALITY

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Testicular Feminization†

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Historical Background

" Testicular feminization " is not well known, but is a clearly demarcated type of intersex which in its classical form is easily diagnosed purely on clinical grounds (Prader, 1957a). As a rarity and a freak of nature, the clinical picture has been recognized for a long time (Steglehner, 1817; Ricco, 1832; Fieux, 1871; Polaillon, 1891; Kochenburger, 1893; Pozzi, 1896; Kutz, 1898; Delagénière, 1899). De Quervain (1923) as the result of his own observations in 1923 was the first to write about this condition in a textbook of surgery: " in spite of plaits and a girl's name the structure in the inguinal hernial sac may even be a testicle ". There followed a series of single case reports which described this condition in a variety of ways (cf. Synonyms) without the fundamental and most important characteristics of this type of intersex being recognized. It was Schiller (1940) who delineated this syndrome from the confused mass of varieties of intersex; Goldberg and Maxwell (1948) recognized the typical picture, and Wachstein and Scorza (1951) reviewed 21 cases. However, it was not until 1953 that Morris coined the term " testicular feminization ", and made the clinical picture clear by collecting 80 cases from the literature and describing two of his own, thereby pointing out the importance, incidence and common features of this condition. The term " testicular feminization " is apt since the breast development and oestrogenization of the vaginal epithelium is a function of the testes, although the predominantly female appearance of the external genitalia develops *in spite* of the testes.

It is worth noting that the cases reported before Morris's paper in 1953 were nearly all discovered either at laparotomy performed for abdominal tumours involving the gonads (Abel, 1891; Marion, 1903; Menetrier *et al.*, 1922; Krückmann, 1937), or by finding the gonads (testes!) in the inguinal canal during herniotomy. There are very few records of cases found by accident at autopsy, because it is not a fatal condition (Neugebauer, 1908). This explains, firstly, the apparently high incidence of neoplastic change (cf. p. 272), and secondly, why the typical situation of the gonads in the abdomen was overlooked, because the peritoneal cavity is not usually explored during a herniotomy.

Until recently, therefore, there were only three ways of discovering this syndrome, namely, by finding a tumour (the most common), during the course of an operation to repair an inguinal hernia, and at post-mortem examinations. It is for this reason too that these were the outstanding signs of this condition. But as soon as Morris (1953) had described the true symptomatology, and particularly after the introduction of methods for determining nuclear sex, *more and more cases without either tumours or herniae were described*. Following the work of Morris (1953) publications have become so numerous that a complete review is almost

† Translated by Peter Huntingford.

exact diagnosis is not, however, necessary, since the children should in any case be brought up as girls. All the parents need be told is that the child will not menstruate; the chromosomal gonadal sex of the child should not be revealed.

An Abdominal Tumour

Recently cases of " testicular feminization " *presenting with a tumour* have become a rarity, the diagnosis is nowadays nearly always made before this occurs. However, both of Morris's (1953) cases had tumours, and one of them came from the Radiumhemmet, Stockholm. In a patient with a blind-ending vagina and primary amenorrhoea, determination of the nuclear sex should enable the correct diagnosis to be made prior to operation. If in addition to these signs, which should establish the diagnosis, the arrangement of the internal genitalia is also overlooked, and this may occur particularly if the tumour is large, the details of the case can be reconstructed afterwards, as we have been able to show with one of our cases (see Table 1, case 10). These unrecognized cases are usually labelled as " disgerminomata ". Because of this, the diagnosis in all previously reported cases of disgerminoma should be reviewed.

Differential Diagnosis

Any other condition that is associated with primary amenorrhoea may cause difficulty in the differential diagnosis, in particular, all types of gonadal dysgenesis, the Rokitansky-Küster syndrome, haematometra, occasionally patients with so-called arrhenoblastomata, and very rarely true hermaphrodites.

In contrast to patients with *gonadal dysgenesis* mammary development is normal, there is no cervix (or uterus), and at laparotomy testicular tissue is found. The nuclear sex in gonadal dysgenesis may be either XY and XO (both chromatin negative), or XX (chromatin positive).

In patients with an absent vagina, and therefore usually some pubic hair, it is very difficult to distinguish them from those with the Rokitansky-Küster syndrome (Hauser *et al.*, 1961; Hauser and Schreiner, 1961)† because in the latter, breast development is also normal; the distinguishing feature is the female nuclear sex. Exploratory laparotomy without taking a biopsy from the gonads may be misleading, since the large white ovaries lying near the inguinal canal may be confused with testes.

Cases of " testicular feminization " with a tumour may be wrongly thought to have an *haematometra* (Abel, 1891; Marion, 1905; Scharplatz, 1958; and one of our cases (see Table 1, case 10)). In such cases careful exploration of the vagina, and, if necessary, laparotomy will confirm the diagnosis.

A number of patients with so-called *arrhenoblastomata*, particularly those with no uterus, have subsequently been recognized as examples of " testicular feminization " (Dudman, 1939; Goldberg and Maxwell, 1948; Behrend, 1936; Javert and Finn, 1941).

True hermaphrodites (Tuffier and Lapointe, 1911) with similar symptoms and physical signs can only be distinguished by exploratory laparotomy and studying the histological structure of the gonads. In one case what was thought to be a myoma (Marion, 1905) proved to be a testicular tumour.

Natural History and Prognosis

" Testicular feminization " does not as a rule shorten the expectation of life, although the reproductive capacity is completely lacking. The primary amenorrhoea and infertility are permanent.

The *complications* include neoplastic changes in the atypical gonads (cf. treatment); and gonadal insufficiency that results in prolonged deficiency of the sex hormones, which in turn may occasionally cause osteoporosis, circulatory disturbances, etc.

Late complications hardly ever occur even in untreated patients. In castrated women, however, all the unpleasant symptoms that follow the menopause

† See footnote on p. 270.

may occur (Hauser and Wenner, 1961) particularly climacteric withdrawal symptoms, depression, adiposity (Cadiz and Lipschütz, 1933), hypertension, etc.

Treatment

1. Symptoms attributable to *inguinal herniae* must be relieved by herniorrhaphy, together with reposition of the gonads after a biopsy has been taken, or if necessary by castration (see below).

2. A question of fundamental importance is whether or not the ectopic and abnormally developed *gonads should be removed*. Under no circumstances should castration be performed before " puberty," otherwise growth of the breasts will not occur. Wachstein and Scorza (1951) found a tubular adenoma in 13 of 21 cases, and Morris (1953) found carcinomata in 7 of 82 cases. These figures have persuaded many surgeons that prophylactic castration during the course of an exploratory laparotomy should be performed, in case a neoplastic change should later occur. The school in Kiel (Schaumkell, 1956; Stange, 1958; Philipp, 1959) are the most ardent proponents of this point of view. But we energetically opposed this policy, as early as 1956, for the followinng reasons:

(a) *The risk of malignant change* is not as great as has been suggested. After a careful survey of the literature, we found that carcinomatous change occurred in only 10 of 128 cases; 6 of these were seminomata (see Table 1, case 5); 8 were cured; and only 2 patients died. If carcinoma does occur, it is generally relatively benign and radiosensitive. Jones and Scott (1958) also state that malignant change occurs in less than 5% of cases.

(b) The incidence of benign adenomata is much greater; Morris (1953) found 23 examples in 81 cases; and to date I have found 30 in 128 cases, but 6 of these adenomata were only discovered by using the microscope. Recently the number of cases without tumours has increased, because previously it was the discovery of a tumour that led to the diagnosis, today we are led to the same conclusion in patients with primary amenorrhoea. The earlier figures for the incidence of tumours are certainly too high.

(c) In " testicular feminization " we are not simply dealing with an ectopic testis, as is found in cryptorchidism (cf. Histology), and we therefore cannot assume that the rate of malignant change in these two conditions is the same.

(d) The castration of patients without a tumour converts symptomless individuals into invalids suffering from all the unpleasant consequences of castration. In addition to hot flushes, the increase in weight following castration may be considerable (Cadiz and Lipschütz, 1933), post-operative depression or psychoses may also occur (Pozzi, 1911; Blumreich, 1906). Prophylactic oestrogens, can, however, because of the absence of the uterus, be given with impunity. But this does not mean that the patient, like a diabetic, needs to take a drug for the rest of her life; this is psychologically undesirable, and also costly.

(e) Castration upsets a state of biological equilibrium that has been built up over a long period of time. Since most of these patients have normally or even very well developed breasts and sexual feelings, it is very difficult to restore this ideal state by replacement therapy.

(f) Other authors state that they remove the gonads, because *they do not correspond to the external, psychological and psychosexual sex of the patient*. This is a costly mistake, because these gonads produce oestrogens. Castration causes regression in growth of the breasts and an alteration in the structure of the vagina, which may lead to dyspareunia.

Naturally we are also of the opinion that tumours, both carcinomata and adenomata, should be removed. But we do not advise the prophylactic removal of gonads that are macroscopically normal testes,

or of the healthy gonad in patients with a unilateral adenoma. These women later have a " normal " climacteric (Hauser *et al.*, 1957). More recently, various authors, including von Miculicz-Radecki, 1959; Schreiner, 1959 and most recently Scharplatz, 1960 have all agreed with the opinions that we have expressed. Even the most enthusiastic proponents of prophylactic castration must wait until after the thelarche, otherwise the breasts do not develop. But we agree with Schaumkell and Stange (1956), however, that in all cases of primary amenorrhoea an exploratory laparotomy should be performed in order to make a diagnosis, and in order to be able to give a clear prognosis to the patient and her parents.

3. In border-line cases with hypertrophy of the clitoris, *amputation* of the clitoris may be necessary for cosmetic reasons (Witschi and Mengert, 1942; Novak, 1943).

4. If the *vagina* is *absent* or *very short* a plastic operation may be required in order to make sexual intercourse possible (Krückmann, 1937; Koller, 1943; Hauser *et al.*, 1957; Scharplatz, 1958). We do not recommend the complicated and dangerous procedure that involves transplantation of a loop of gut, but simply excavation of the space between the bladder and rectum, followed immediately by regular and continuous use of dilators together with the administration of large doses of oestrogens. The application of a prothesis covered with amnion or skin is not usually necessary.

Drug therapy is not usually needed in those patients who have not been castrated. Occasionally in patients with small breasts, small repeated doses of oestrogens given over a long period of time may promote their development. If the patient herself desires to increase the growth of her pubic hair, we advise a combined depot injection of androgens and oestrogens, e.g. Femandren. In castrated patients substitution therapy with oestrogens is desirable to maintain metabolic equilibrium.

Psychological and Psychosexual Upbringing

These patients should be brought up as females in spite of their male gonads and chromosomal sex. They should never be told they have testes, or that they are hermaphrodites, but we always do explain to them that they are sterile, and will remain so, and that they will never menstruate. Usually they accept this knowledge very well. Most married women either adopt children or take part in extra-household activities which they often pursue with great success because of their above average intelligence.

Summary

" Testicular feminization " is the most inconspicuous form of intersexuality. In spite of having testes the patients are of normal female physique. If they do not have inguinal herniae, the intersexual condition may remain unrecognized until puberty, when the condition first manifests itself, because of primary amenorrhoea and the sparseness of body hair. The diagnosis is usually easily made on clinical grounds when the patient reaches sexual maturity, by the presence of the following cardinal symptoms: primary and therapy-resistant amenorrhoea and sterility (absence of the uterus); a blind-ending vagina; a familial incidence on the maternal side; and in some cases absence of body hair, and inguinal herniae. The diagnosis is confirmed by demonstrating that the nuclear sex of such a person is male, and by histological examination of the gonads (biopsy).

After recognizing the nature of the intersexual disorder, it is the duty of the doctor to ensure that the patient's previous way of life is altered as little as possible. The gonads should be preserved, unless neoplastic change has occurred. It is particularly important that the patient's gonadal and chromosomal sex should remain a secret, although they should be informed that the amenorrhoea and sterility cannot be relieved.

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