The Diagnosis and Management of Micropenis

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The term micropenis describes a penis which is abnormally small but otherwise perfectly formed with the urethra opening at the tip of the glans (Fig. 1). This should be distinguished from a small but hypospadiac penis, for which the term microphallus is sometimes used. The immediate importance of micropenis is that it may be the only outward manifestation of multiple pituitary hormone deficiences which, if unrecognized and untreated, may be lethal. In other cases, it may indicate the presence of dysgenetic testicular tissue which may have a malignant potential. Irrespective of the underlying cause, the question will always arise as to whether the penis will enlarge sufficiently with hormonal stimulation for the infant to be raised as a boy, or whether sex reassignment should be considered.

In addition to the small hypospadiac penis, micropenis must also be distinguished from a webbed scrotum anomaly in which the scrotal sac extends well up onto the ventral aspect of the shaft of the penis, and a buried penis in which the shaft lies hidden within an abundant mass of prepubic fat.

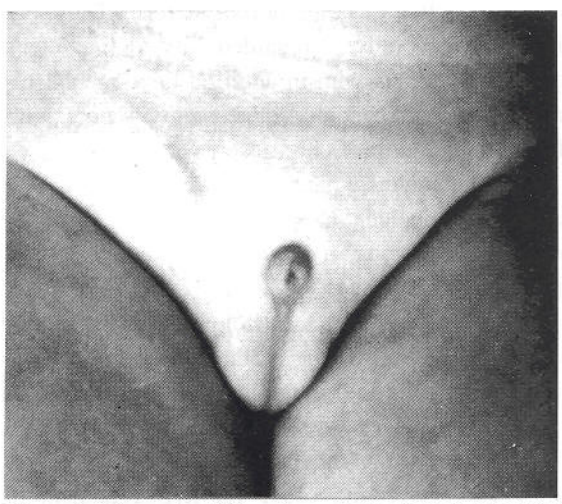

Figure 1. Micropenis. Although completely formed, the penis is minute and obscured by the prepuce. The testes in this case are impalpable.

Measurements of the penis

Measurement should be made using a rigid ruler, and gripping the glans firmly in order to stretch the penis to its full length, the distance is recorded along its dorsal aspect from the symphysis pubis to the tip of the glans, disregarding the prepuce. Micropenis defined as a penis which has a stretched length less than 2 1/2 standard deviations below the mean, thus lying outside the range found in 99.4 % of a normally distributed population. Table 1, based on data compiled by Lee et al [16] records the stretched penile length of normal males from pre-term neonates through to puberty. A penis in a full-term baby having a stretched length of less than 1.9 cm therefore qualifies for the diagnosis.

Normal penile development

Development of the penis takes place in three phases. In the genital tubercle phase when the embryo measures between 8 and 15 mm, the phallus appears as a hillock in the perineum. In the phallic phase, which lasts from the 16 to 38 mm stages, the organ becomes progressively elongated and cylindrical with the urethral groove extending to its tip. The labioscrotal folds are now also apparent. The definitive phase begins during the third month, or approximately 38 mm, when the urethral tube closes and the glans becomes demarcated by the formation of the coronal sulcus. The scrotum also becomes prominent at this stage. Development of the penis is therefore complete by the end of the third month or 45 mm, except for the formation of the prepuce which come to cover the glans during the following few weeks. These events are brought by the placental secretion of hCG which stimulates the testicular Leydig cells to produce testosterone. From the fourth month onwards the fetal pituitary gland takes over and begins to secrete LH and FSH in re-

Table 1. Stretched penile length in centimeters: normal males

Age	Mean±SD	Mean--2 1/2 SD
Newborn 30 wks	2.5±0.4	1.5
Newborn 34 wks	3.0±0.4	2.0
0-5 months	3.9±0.8	1.9
6-12 months	4.3±0.8	2.3
1-2 years	4.7±0.8	2.6
2-3 years	5.1±0.9	2.9
3-4 years	5.5±0.9	3.3
4-5 years	5.7±0.9	3.5
5-6 years	6.0±0.9	3.8
6-7 years	6.1±0.9	3.9
7-8 years	6.2±1.0	3.7
8-9 years	6.3±1.0	3.8
9-10 years	6.3±1.0	3.8
10-11 years	6.4±1.1	3.7
Adult	13.3±1.6	9.3

sponse to GnRH produced by the hypothalamus, thus continuing growth of the penis which, by this stage, is already fully formed. Micropenis, therefore, results from a hormonal defect which arises after the fourteenth week of embryonic development.

Causes of micropenis

The principal causes of micropenis are summarized in Table 2. In the majority of cases, the primary fault lies in the hypothalamus which fails to produce an adequate amount of GnRH, although in some cases the anterior pituitary gland itself may be deficient. Circulating testosterone levels are correspondingly low and growth of the penis during the second and third trimesters is inadequate. Collectively, these disorders are known as hypogonadotropic hypogonadism.

Somewhat less commonly, the primary fault lies in the testes themselves which although initially capable of producing sufficient testosterone to completely masculinize the external genitalia, undergo degeneration from the mid-trimester onwards with a fall of in testosterone production. The negative feedback to the hypothalamus and anterior pituitary gland is thus impaired resulting in elevated concentrations of circulating LH and FSH. This group of disorders is therefore sometimes referred to as hypergonadotropic hypogonadism.

Major chromosomal defects

Genital anomalies are frequently found among infants with major chromosomal defects. These usu-

Table 2. The main causes of micropenis

MAJOR CHROMOSOME ANOMALIES

Klinefelter's syndrome
Down's syndrome
Other defects

HYPOTHALAMIC-PITUITARY DYSFUNCTION

Structural brain defect
Hypogonadal syndromes
- Kallmann
- Prader-Willi
- Laurence-Moon-Biedl
- Other syndromes

Isolated hormone deficiencies
- GnRH deficiency
- LH deficiency
- Multiple hormone deficiencies

GONADAL DEFECTS

Testicular dysgenesis
Vanishing testis syndrome
LH receptor defect
Other intersex states

IDIOPATHIC

ally take the form of cryptorchidism, hypospadias or simply a small penis, although only rarely will it be sufficently small to qualify as a micropenis. Klinefelter's syndrome (47 XXY), the commonest major chromosomal anomaly associated with hypogonadism, occasionally presents in the newborn period with a small penis associated with small testicles [21].

A small penis is also a feature of other X polysomy syndromes, e.g. 48XXXY, 49XXXXY, which may occur either in a pure or mosaic form. These patients are generally regarded as Klinefelter's syndrome variants who in addition have mental retardation which becomes increasingly severe with the number of extra X chromosomes.

Other anomalies which may be associated with a small penis include Down's syndrome [11], translocations, deletions and trisomy involving particularly chromosomes 8,13, and 18. A small penis is also a feature of 69XXY triploidy. Many of these aneuploid infants will be stillborn, but among live births a major chromosomal defect will be suspected by an unusual facies, intrauterine growth retardation and the presence of other major congenital anomalies. The underlying cause for hypogonadism in most of these cases is unclear.

Hypothalamic-pituitary dysfunction

Structural brain defects

The most striking and perhaps the most easily understood malformation associated with micropenis is anencephaly in which the exposed neural tissue including the midbrain undergoes hemorrhagic disintegration with loss of hypothalamic function. Congenital pituitary aplasia, in which the adenohypophysis, derived from the foregut, fails to develop, may also present at birth with cryptorchidism and a micropenis. Although even less common than anencephaly, it is of much greater clinical significance as it is readily treatable. The diagnosis should be made from the serum electrolytes and pituitary hormone levels rather than radiographically as the pituitary fossa may be unremarkable on a lateral x-ray view of the skull.

Agenesis of the corpus callosum is the commonest of a group of developmental midline brain defects which may be associated with micropenis. Although sometimes occurring as an isolated lesion causing minimal neurological disturbance, it is more often encountered as part of a wider field defect in which the hypothalamus is frequently involved. The diagnosis should be suspected clinically when the facial appearance is unusual, particularly if the forehead is broad and the eyes are widely spaced (hypertelorism).

Other midline brain defects associated with hypogonadism include septo-optic dysplasia, occipital encephalocele, Dandy-Walker cystic malformation of the fourth ventricle and cerebellar malformations with ataxia.

Hypogonadal syndromes

The commonest single underlying cause of micropenis is congenital GnRH deficiency without any major structural defect of the brain. Although sometimes occurring in children who are otherwise entirely normal, more often it is associated with other anomalies, some of which have been recognized as discreet syndromes.

Kallmann syndrome

This condition, also known as genito-olfactory dysplasia, is the commonest of the syndromes of gonadotropin deficiency. It is characterized by a defect in the sense of smell which may be either absent (anosmia) or impaired (hyposmia) [13] and an inheritance pattern which may be autosomal recessive or dominant, or X linked [24]. GnRH production is very low with corresponding low serum levels of LH, FSH and testosterone. The secretion of the other pituitary hormones is usually normal [17].

This curious association between anosmia and hypogonadism is not simply fortuitous but is derived from the fact that embryologically, the GnRH producing cells originate in the olfactory placode. A lesion at this site early in gestation is therefore likely to affect both the developing olfactory pathways as well as the gonadotrophs. The latter normally migrate along the vermo-nasal and terminalis nerves to the septal-preoptic area and the hypothalamus, but in Kallmann syndrome, this migration is arrested [20] with the GnRH producing cells ending in a tangle within the meninges just above the cribriform plate. Grossly, the hypothalamus and pituitary gland are normal but the olfactory sulci are usually hypoplastic.

Other defects which may be associated with Kallmann syndrome include short stature, mental retardation, skin lesions and unilateral renal agenesis, the genes for which appear to lie close to the Kallmann gene at the tip of the short arm of the X chromosome [2]. The precise function of the Kallmann gene itself, now localized to within 210 kilobases, seems to be the elaboration of proteins necessary for neural cell adhesion and axonal pathfinding, both of which are vital for cell migration to occur [6].

Prader-Willi syndrome

This condition, first described nearly 40 years ago among Swiss children, will be suspected when a baby is noted to be floppy at birth and have a small penis with a flat, empty scrotum. In early childhood, the muscular hypotonia becomes less noticeable and the clinical picture is dominated by short stature and obesity, which is often gross (Fig. 2), and mild mental retardation [9]. At puberty, diabetes mellitus may develop and behavorial problems are very common. The cause of the hypogonadism is not entirely clear, for although in most cases it appears to be central in origin in others a primary testicular defect may be

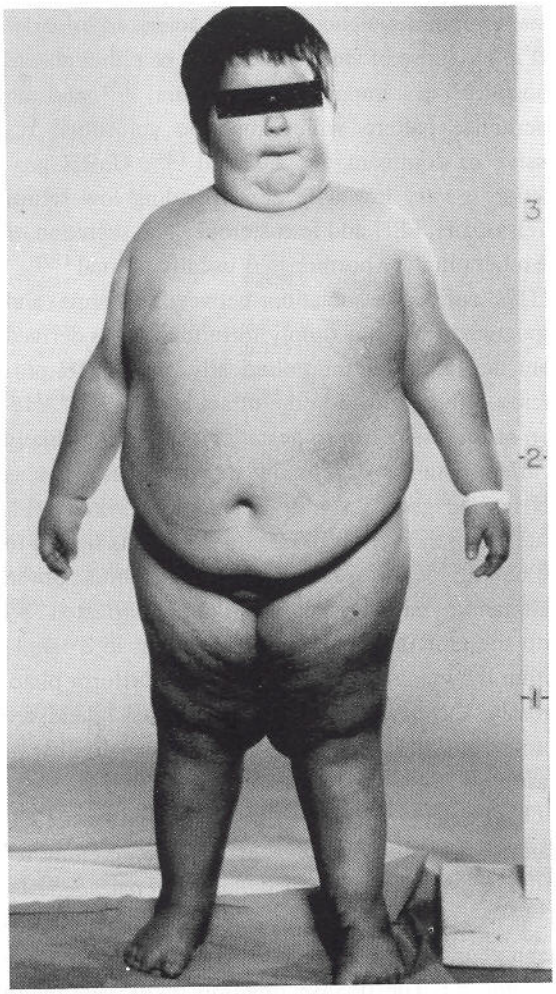

Figure 2. Prader-Willi syndrome showing short stature, gross obesity, small hands and feet and diminutive genitalia in an eight-year old boy.

present.

Recent advances in genetics, which include the wider availability of high resolution chromosome banding techniques, having demonstrated an interstitial microdeletion of bands 11 nd 12 in the proximal long arm of chromosome 15 in approximately 70 % of cases [14]. Among those without microdeletion of bands 15q 11-12, disomy for chromosome 15 may sometimes be found in the mother. A clinical scoring system has also recently been compiled to assist in the diagnosis of the Prader-Willi syndrome [10].

Laurence-Moon-Biedl syndrome

This less clearly defined hypogonadal syndrome shares with the Prader-Willi syndrome the features of obesity and mental retardation, but is characterized by a pigmented retinopathy leading to blindness in early adult life, and polydactyly. Reports of endocrine studies in these patients are few and whilst in some the hypogonadism appears to be central in origin, in others the testes seem to be inadequate [22].

Other syndromes

A small penis has been described as a feature of a large number of rare syndromes which are often familial. These include the Rud, Martsolf, Robinow, Smith-Lemli-Opitz, Boucher-Neuhauser and Mobius syndromes, in many of which an atypical facial appearance and mental retardation are striking features.

Isolated hormone defects

Rarely, a deficiency of GnRH or LH may occur as an isolated abnormality. More often though other pituitary hormones, notably growth hormone, thyroid hormone or cortisol may be deficient and thus be life threatening. Micropenis in a newborn infant should therefore act as a red flag and mandate the exclusion of these conditions as soon after birth as possible.

Gonadal defects

Testicular dysgenesis

In this condition, the Leydig cells are initially sufficiently abundant to produce some testosterone but their function subsequently seems to fall away, leaving either the development or subsequent growth of the external genitalia incomplete [5]. The final phenotypic appearance, which will range from normal female to micropenis will depend on the exact timing of this deterioration (Fig. 3).

The surgical importance of this group of infants lies in the danger of leaving dysgenetic testicular tissue within the abdominal cavity where it may possibly become malignant. Such testes may not be recognized on any imaging study and may even remain undetected by an hCG stimulation test [3]. Laparoscopy or laparotomy therefore are mandatory in suspected cases, at which time an excision biopsy

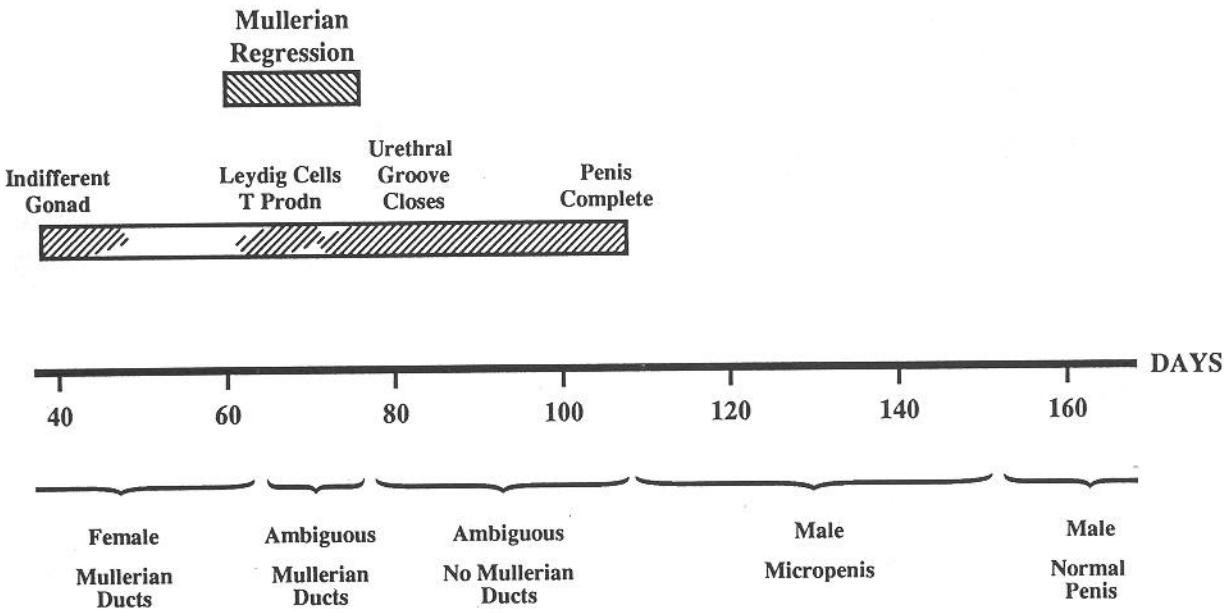

Figure 3. Development of the penis. Temporal relationship between the maturation of embryonic testicular function and the appearence of the internal and external genitalia.

should be carried out of the tissue surrounding the termination of the gonadal vessels. Testicular dysgenesis occurs as either a sporadic phenomenon or may be familal. Among the latter are two siblings reported by Josso and Briard [12], in whom the testes appeared to degenerate at different times, resulting in one being raised as a girl and the other as a boy.

Vanishing testis syndrome

This term describes children with cryptorchidism and a normal penis in whom exploration reveals one or both gonadal vessels to end blindly [1]. What causes the testicles to disappear remains uncertain but as the penis is usually well developed, a catastrophic event such as torsion occuring late in gestation seems to be the most likely explanation.

Other intersex states

In the vast majority of intersex disorders, the penis although often small is also incompletely formed with some degree of hypospadias. In addition to testicular dysgenesis, a micropenis with the urethra opening on the tip of the glans may very rarely be found in true hermaphroditism and the androgen insensitivity syndrome. Among girls with severely virilizing congenital adrenal hyperplasia, the urethra may also open on the glans but the phallus in these patients is usually sufficiently enlarged to take it out of the category of micropenis.

Idiopathic micropenis

Finally, there remains a small group of children with micropenis in whom the underlying cause cannot be clearly determined but who, at puberty, spontaneously virilize with adequate growth of the penis. Among 45 children studied by Lee et al [15], six were placed in this category. Hormonally these children appear to be normal and the cause of the micropenis remains a matter of speculation.

Determining the cause of micropenis

History and examination

The occurrence of a previous stillbirth or first-degree relatives with hypospadias, cryptorchidism, infertility or major congenital anomalies will suggest the presence of a transmitted genetic defect. Anosmia is frequently found in families of infants with Kallmann syndrome although it may be only partial and therefore go unreported. The development of deafness or deteriorating eyesight among relatives at an early age should also be noted.

An obstetric history of reduced fetal movements or floppiness at birth will raise the suspicion of the Prader-Willi syndrome.

Examination may reveal an unusual facial appearance which will suggest a chromosomal or midline brain defect. The hands and feet may reveal an unusually small size, syndactly or polydactly. In early childhood, short stature or obesity may be apparent, whilst the skin may show multiple pigmented nevi or ichthyosis. In older children, formal testing of hearing and smell should be carried out and the fundi examined for pigmentary changes in the retina.

Local examination of the genitalia should include, in addition to an accurate measurement of the stretched penile length, an assessment of the bulk of the corpora upon which the potential for satisfactory erection depends. The size, symmetry and rugosity of the scrotal sacs should be noted, and if palpable, the size and consistency of the testes.

Investigations

The principal task in investigating infants and children with micropenis is to determine whether the underlying cause is central in origin or arises in the gonads. The former may be associated with multiple hormone deficiences which must be promptly recognized and treated, or other defects which may affect the long term outlook. When micropenis is the result of primary testicular dysfunction, the location of any testicular tissue must be identified and the risk for malignant degeneration assessed. Among those cases in which it is decided to continue raising the infant as a boy, the establishment of the underlying cause will allow the most appropriate method of hormone replacement to be chosen, as well as indicate how the testes, which may be impalpable or incompletely descended, should be managed. The need for genetic counseling will also become apparent.

Anterior pituitary secreening tests

Infants with micropenis may present shortly after birth with hypoglycemic convulsions as a result of hypopituitarism with low levels of ACTH and growth hormone. Although relatively uncommon, the life-threatening nature of these defects requires the routine serial measurements of serum glucose, sodium, and potassium concentrations during the first few days of life. Serum cortisol levels should also be measured routinely, together with an estimate of growth hormone activity. This can be measured directly, or indirectly from the concentration of insulin-like growth factor binding protein-3 generated in peripheral tissues in response to growth hormone action. Thyroid function should also be measured.

Karyotype

Standard karyotyping should be carried out routinely in all infants and children with micropenis to rule out Klinefelter's syndrome or a major chromosomal defect. Among infants who are hypotonic at birth or have other features suggestive of the Prader-Willi syndrome, high resolution cytogenic or molecular studies should be carried out looking specifically for a deletion in the proximal region of the long arm of chromosome 15. The ability to identify interstitial deletions of the tip of the short arm of the X chromosome in suspected Kallmann syndrome is currently confined to research centers.

Magnetic Resonance Imaging of the skull

This should form part of the routine workup in all infants and children with micropenis to determine the anatomical integrity of the hypothalamus and anterior pituitary gland. In children with cranio-facial anomalies, attention should also be directed to other midline structures particularly the optic chiasm, fourth ventricle and corpus callosum. When Kallmann syndrome is suspected, the size of the olfactory sulci, which are best seen on transverse images of the forebrain, should be noted.

Gonadotropin studies

These studies should be carried out by a pediatric endocrinologist and are designed to determine, as far as possible, the functional integrity of the hypothalamic-pituitary-testicular pathway, in order to distinguish hypogonadotropic hypogonadism from primary testicular dysfunction.

Normal male infants show a brisk but transient rise in serum LH, FSH and testosterone levels

peaking at about eight weeks of age, which subsides by six months. Serial measurements of these hormones at intervals during the window of opportunity presented by this "mini-puberty" are all that is necessary to determine whether the hypothalamus, anterior pituitary gland and testes are functionally intact. A peak serum testosterone concentration of over 100 ng/dl can be regarded as normal. By contrast, a low serum testosterone associated with relatively raised serum LH and FSH concentrations, will suggest the diagnosis of primary testicular failure. This can be confirmed by a formal hCG stimulation test. Among infants in whom the peak concentration of all three hormones remains low, hypogonadotropic hypogonadism will be suspected.

An hCG stimulation test should first be carried out to confirm that the testes are functionally adequate. This can be followed by a formal GnRH stimulation test to confirm the integrity of the anterior pituitary gland, but because of the practical difficulty this entails, it can be assumed that if the anterior pituitary is anatomically and in other respects functionally normal, the underlying cause of the micropenis lies in the hypothalamus.

Between the ages of approximately six months and the onset of puberty, the serum concentrations of LH, FSH and testosterone remains very low. Testing therefore begins by challenging the testes with hCG in order to exclude them as a cause for the micropenis. It is important to remember however that a negative response reflects only Leydig cell function and does not automatically exclude the presence of rudimentary testicular tissue which may have a malignant potential. When the testes are shown to be functionally intact, the next step is to determine anterior pituitary function by measuring plasma cortisol, growth hormone and T4, TBC or TSH levels. A GnRH stimulation test may also be carried out although the same limitations apply as in infants as to its clinical usefulness.

Beyond the age of expected puberty, the measurement of the basal concentrations of circulating LH, FSH and testosterone again forms the basis for diagnosis. In cases in which all three are low and the anterior pituitary gland and testes appear to be intact, primary hypothalamic dysfunction will need to be distinguished from delayed puberty.

Diagnosis of androgen insensitivity

Although androgen insensitivity is a very rare cause of micropenis, it is of great clinical importance and must be excluded when the above endocrine studies point to normal hypothalamic, anterior pituitary and testicular function. It is most simply excluded by demonstrating growth of the penis in response to a course of hCG or testosterone.

Laparoscopy

When endocrine studies point to a testicular cause for the micropenis and the testes are impalpable, laparoscopy should be performed to determine the precise nature of any testicular tissue which may be lying within the abdominal cavity. When the spermatic vessels are clearly indentified and appear to end blindly, the diagnosis of vanishing testis is confirmed but the region of the termination must be closely examined and any nubbin of tissue, however small, should be removed either laparoscopically or by formal laparotomy for histological examination (Fig. 4). Müllerian duct structures are very rarely found in patients with micropenis.

Laparoscopy should also be carried out in children in whom the diagnosis of hypogonadotropic hypogonadism has been made, but in whom the testes remain impalpable in spite of a course of hCG, in order to determine their position. When the testes

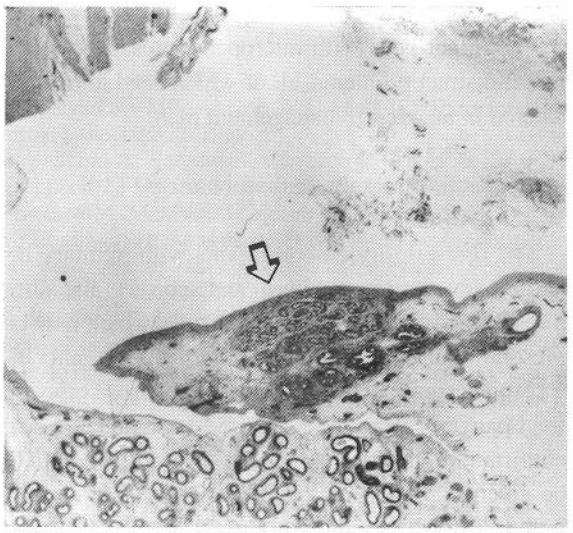

Figure 4. Testicular dysgenesis in an infant with micropenis and cryptorchidism. Photomicrograph of tissue excised at the termination of the gonadal vessels showing clusters of disordered tubules (arrow). Hematoxylin and Eosin. Original magnification x 80.

are lying well above the internal ring but appear otherwise normal, the gonadal vessels are clipped in preparation for a Fowler-Stephens orchidopexy six months later. Abnormal testes should be biopsied, and those showing disordered histology should be removed. Substitution therapy is started between 12 and 14 years of age.

Renal imaging studies

A renal ultrasound scan should be carried out in all cases of hypogonadotropic hypogonadism because of its known association with unilateral renal agenesis. When only one kidney is identified, a 99mTc DMSA renal scan will exclude an ectopic location of the missing kidney and usually identify any perfused nubbin of dysplastic renal tissue which may be too small to see on ultrasound.

Genitogram

A genitogram is not indicated in the initial work-up of micropenis. However, this should be carried out when dysgenetic gonads, an ovotestis or Müllerian duct structures have been identified at laparoscopy, or when the possibility of partial androgen insensitivity has been raised.

Management

The infant born with micropenis presents a number of distinct problems, all of which need to be considered in an overall management plan.

Correction of metabolic disturbances

The first priority among infants with pituitary insufficiency is to stabilize and correct the life-threatening biochemical abnormalities which may be present. This will usually entail the urgent intravenous administration of a 10 % dextrose infusion for hypoglycemia. Following the rapid biochemical confirmation of the diagnosis, hydrocortisone should be given at 50 mg/m^2 together with half normal saline to correct hyponatremia if present. Infants with hypopituitarism may require long-term replacement of cortisol, growth hormone and thyroxine.

Stimulation of penile growth

The next priority, particularly in the newborn or very young infant, is to determine whether sufficient growth of the penis can be achieved with androgen stimulation in order for the baby to continue to be raised as boy. A good idea of this can be obtained simply by observing the response of the penis to the course of hCG given as part of the initial diagnostic studies. If this is unimpressive, a second course of 500 IU i.m. should be given every five days for up to three months and the outcome assessed at six weeks and again at 12 weeks. If there is no significant growth of the phallus, sex reassignment should be considered.

When the underlying cause of the micropenis is testicular in origin, Testosterone enanthate or proprionate 25 mg should be given i.m. at three weekly intervals for a total of four doses and the penis reassessed after a further two weeks. Because of its erratic absorption, topical testosterone should be avoided. The above protocol will usually result in a significant increase in both the length and girth of the penis, together with increased rugosity of the scrotum. A light but transient growth of fine pubic hair may sometimes also be seen. Some regression however in penile size must be anticipated in the months following treatment, although precisely how much will vary from case to case [8]. Any lost ground can sometimes be made up by a second course given around four or five years of age.

The most important single predictor of adult penile size appears to be the initial length of the penis at birth [4]. A penis which at the outset is minute, i.e. smaller than 3 SD below the mean, is likely to cause lifelong problems and such infants, particularly those in whom the penis is little more than a skin tag, are best considered for early sex reassignment.

Surgical reconstruction of the penis has been undertaken in a few children with micropenis. The technique involves the creation of a double lumen flap from the radial aspect of the forearm which is transferred to the perineum by direct anastomosis of vessels and nerves using microsurgical techniques [7]. Erectile function is subsequently achieved by the insertion of a penile prosthesis. Although this is without doubt an invaluable contribution in cases of traumatic amputation of the penis, its adaptation to children with micropenis, in whom sex reassignment

can generally be achieved without difficulty, must be questioned.

Management of the undescended testes

Among children with hypogonadotropic hypogonadism in whom the testes are incompletely descended, an expectant policy is justified as spontaneous late descent sometimes occurs [23]. This however must be offset against the possible benefits of early orchidopexy. The author's practice therefore is to give a course of hCG injections between four and five years of age which, as well as possibly further enhancing penile size, may bring the testicles down. A final assessment is made three weeks after the last injection when, if descent remains incomplete, orchidopexy is performed.

Induction of puberty

It can be assumed that all patients, with the exception of those with an idiopathic micropenis, will not go through a normal puberty without the help of exogenous hormones. Testosterone will be necessary when the testes are congenitally absent, poorly functioning or have been removed because of dysgenesis. Treatment is usually begun around 12 years of age with Testosterone enanthate or proprionate, 100 mg i.m. every six to eight weeks. After one to two years, this is increased to four weekly. Among children who are also growth hormone deficient, final height should be achieved before beginning testosterone treatment.

Children with idiopathic micropenis who undergo puberty spontaneously should have serial measurements of their serum LH, FSH and testosterone concentrations, as well as semen analysis to be sure that testicular function remains adequate.

Family screening and genetic counseling

Genetic counseling is indicated in the Kallmann, Prader-Willi and Laurence-Moon-Biedl syndromes, and also when micropenis is associated with the other familial syndromes listed above.

Psychological sequelae

Behavioral and psychosexual problems must be anticipated in children with micropenis as they grow up, particularly among those in whom penile growth during childhood has been disappointing. Among a small group of such children followed into adult life by Money et al [18], most adopted avoiding strategies during urination or in the locker room. More disturbingly, a higher than anticipated number had adopted a homosexual lifestyle, whilst some had expressed a desire for sex-change surgery or had even contemplated suicide. It should be noted, however, that androgen replacement therapy in these children had been delayed or given erratically and that the stretched length of the penis in all cases had remained less than 2.5 SD below the mean.

By contrast, Reilly and Woodhouse [19], following a larger but heterogenous group of children diagnosed as having micropenis in infancy, found that among those who were post-pubertal, all were heterosexual and happy with their sexual identity. Most were sexually active and although often needing to employ special techniques, expressed themselves as sexually satisfied and enjoyed stable relationships. Close medical follow up, compliance with androgen replacement therapy and understanding and supportive parental attitudes were identified by these authors as being more important than the actual size of the penis. Nonetheless, when the penis at birth is minute or clearly lacks the potential for erectile function, early sex reassignment will invariably be the best course.

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