HYPOSPADIAS AND CRYPTOSPADIAS

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From the literature one can learn that hypospadias arises from a defective closure of the urethral groove and the malposition of the meatus and the cleft prepuce can thus be simply explained. In contrast, it is much more difficult to explain the presence of the so-called chordee and impossible to explain the remarkable distribution of the skin with an excess on the dorsal side, a deficiency on the urethral side and two oblique raphes (Fig. 1).

When the anomalies which occur singly or in various combinations, and for which the literature offers no satisfactory explanation, are considered it seems certain that the development of the urethra is not so simple as the usual textbooks' description.

The investigations made by Glenister (1954) showed that this is so (Fig. 2). According to him, the differentiation of the urethral groove and the formation of the urethra take place as follows. The surface epithelium of the floor of the urethral groove reacts to contact with the ingrowing urethral plate, a lamellar structure which is placed perpendicular to the floor of the urethral groove. This reaction manifests itself first by proliferation and subsequently by regression. The groove, which is consequently deprived of its integument, is called the primary urethral groove. Later, in the 35 mm. stage, the secondary urethral groove is formed by disintegration of that portion of the urethral plate that is adjacent to the floor of the primary urethral groove and Glenister concludes that the ectoderm-lined edges of the urethral groove do not participate in the formation of that part of the urethra located proximal to the corona.

The differentiation of the urethral plate is induced by male hormones (Burns, 1942) and the severity of the hypospadias is dependent on the time at which this differentiation is disturbed (Jost, 1947).

On the basis of these data, it seems possible to explain not only the unusual morphology of hypospadias, but also the morphology of such other congenital malformations.
as hypospadias without hypospadias, chordee without hypospadias, congenital short urethra without hypospadias, congenital short urethra and congenital urethral fistulas. These malformations have been grouped together under the term "cryptospadias" thereby denoting their obvious relationship with hypospadias.

Hypospadias.—The dystopia of the meatus and the so-called chordee can be simply explained by a disturbance in the differentiation of the urethral plate, of which they are a direct consequence. The other characteristic features of hypospadias, such as the skin defect at the level of the urethral groove and the abnormal distribution of the skin over the surface of the penis, are indirect consequences of this defective differentiation; the skin defect persists because spontaneous closure over a defect in the urethra does not seem possible, while skin is abnormally distributed because growth and differentiation of the integument, in contrast to that of the urethral plate, proceed normally.

The resulting discrepancy in growth along the edges of the urethral groove must, in our opinion, result in a plication of the integument (van der Meulen, 1964). This plication, which is characterised by the excess of skin on the dorsal side of the penis, a shortage of skin on the urethral side and two oblique raphes, may be accompanied—also in the absence of a so-called chordee—by a curvature of the penis.

Cryptospadias.—The terms hypospadias without hypospadias, chordee without hypospadias, congenital short urethra without hypospadias, and congenital short urethra, are used in the literature for cases which show penile curvature of the penis with a normally situated meatus. An analysis of the infrequent descriptions of this anomaly by a small number of authors, however, makes it clear that this malformation can vary with respect to either the prepuce or the corpus spongiosum.
Prepuce.—In the cases reported by Sievers (1926), McIndoe (1948), Loughran (1948) and Fogh-Andersen (1964) the prepuce was intact while Ombredanne (1923) and Huffstadt and Bouman (1963) mentioned a defective prepuce (tablier de sapeur).

Corpus spongiosum.—Nesbit (1954) 1 case, Williams (1952) 1 case, Huffstadt (1963) 1 case, Moore (1965) 3 cases, Bergerhof and Gelbke (1962) 4 cases, reported absence of the corpus spongiosum but it was present in 2 patients treated by McIndoe and 2 patients treated by Bergerhof and Gelbke. On the basis of their findings in 6 patients, Bergerhof and Gelbke distinguish between hypospadias without hypospadias (when no corpus spongiosum is found) and congenital short urethra (when a corpus spongiosum is present.

The corpus spongiosum urethrae, however, does not start to develop until the end of the fourth month, in the peri-urethral tissue, i.e., after the urethra, whether defective or not, has formed. Consequently, it seems unlikely that there is any relationship between the nature of this defect and the presence of a corpus spongiosum. But, in my opinion, there is a relationship between the nature of the defect and the anomalous distribution of the skin over the surface of the penis or the absence or presence of chordee.

The following cases, seen by us, will illustrate this:

Seven patients (Fig. 3) showed a terminal meatus, a cleft prepuce and a plication of the integument without chordee. Correction in these cases could be obtained by skin rearrangement alone.

Three patients (Fig. 4) showed a terminal meatus, a cleft prepuce, plication of the integument and a short urethra (chordee). In such cases the treatment should consist of skin rearrangement and lengthening of the urethra.

Four patients (Fig. 5) showed a terminal meatus, an intact prepuce, plication of the integument and a short urethra (chordee).

Assuming that the urethra is formed by differentiation of the urethral plate and that
A. and B. A combination of an apical meatus, a cleft prepuce and a plication of the integument and chordee.

Fig. 4

Fig. 5
A. An apical meatus and intact prepuce.
B. A plication of the integument is indicated by ink lines following the course of the median and oblique raphe. In this case a slight torsion of the penis was found in combination with displacement of the median raphe towards the lateral aspect.
C. No corpus spongiosum was found in the distal part of the urethra. Chordee in this case was caused by a deficiency in growth of the urethra.
this process of formation can take place without being covered by skin, the following conclusions may be made:

1. The above-mentioned anomalies are, like hypospadias, the result of a disturbance in the differentiation of the urethral plate.
2. This disturbance is not always accompanied by a defect in the wall of the already formed urethra or malposition of the meatus, but can however affect the length of the urethra and result in chordee.
3. The discrepancy between the deficient formation of the urethra and a normal growth of skin is, as in hypospadias, accompanied by an abnormal distribution of the skin. Closure of the skin defect is possible because there is no defect in the wall of the urethra and in rare cases, even the formation of the prepuce can take place normally.
4. The curvature of the penis, can, as in hypospadias, be caused by an abnormal skin distribution, shortness of the urethra (chordee), or a combination of both.

There are few data in the literature concerning congenital fistulas of the urethra. In our experience, however, the anomaly occurs more frequently than would be expected from the published material.

Two forms can be distinguished. In the first, distal to the defect, there is a urethra that is not covered by skin. This urethra can end either open or blind. This is probably the type of case described by Voillemier (1868), Lippert (1886), Boissonnat (1954), Arnaud and Lacroix (1886); these cases illustrate the intrinsic capacity of the urethral plate to form a urethra without skin cover. Such cases can be differentiated from hypospadias only by the presence of an intact urethra distal to the defect.

In the second form, the urethra is covered by skin distal to the defect. Four examples of this type were found in the literature, Blandin (1868), Wallerstein (1904), Williams (1958) and Gupta (1962).

Five patients with a similar anomaly have been seen personally; in 2 the prepuce was cleft (Fig. 6) and in 3 intact (Fig. 7). Two of the latter 3 patients also had a dystopic meatus thus demonstrating the combination known as hypospadias sous prepuce complet (Barcat, 1947) (Fig. 8).
In 4 of the patients the urethra was too short and all 5 showed the plication of the integument described above.

On the basis of these data, the following conclusions seem justified:
1. Congenital urethral fistulas, like all the other malformations mentioned above, are caused by a disturbance in the differentiation of the urethral plate.
2. In these patients, this disturbance has led to a partial defect in the urethra. In some, the urethra is also too short.
3. Closure of the defect in the integument distal to the urethral defect is possible because of the presence of an intact urethra.
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DISCUSSION

A disturbance in the differentiation of the urethral plate can be accompanied by various changes in the structure of the urethra and in the form of the skin. Changes in the urethra can be characterised by dystopia of the meatus, chordee and a proximal defect (fistula). Changes in the skin can be characterised by a cleft prepuce, plication, and a proximal defect.

All these changes can occur separately or in combination. An attempt has been made to illustrate schematically the combinations apparently possible (Table). Exact diagnosis may be difficult however, since many transitional forms exist.

REFERENCES


