Hypospadias

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Abstract

Hypospadias is a relatively common abnormality of the external genitalia and the incidence seems to be increasing. Early recognition and appropriate referral to specialist centres allows for parents to be given timely and accurate information, although in general the surgical management of patients does not commence until one year of age.

It is vital that assessment of children with hypospadias includes the position of the testes at the time of recognition. When both testes are adequately descended and palpable within a well-developed scrotum, no further investigation is required urgently. However, abnormalities of testicular descent or scrotal development can point to more serious problems and should be investigated urgently. For these children, hypospadias may be a part of a more complex abnormality in sexual differentiation.

The operative management of hypospadias is undertaken by specialist surgeons. Results for distal hypospadias are excellent and few patients have long-term complications from surgery. Proximal hypospadias, particularly in association with other syndromes or disorders of sexual development have poorer long-term outcomes. There is no perfect operation for the correction of hypospadias, and long-term data outlining the advantages and disadvantages of different techniques is lacking in the literature.

Keywords hypospadias; paediatric surgery; paediatric urology

Introduction

Hypospadias is a relatively common abnormality of the external genitalia. Although female hypospadias can be appreciated at cystoscopy it is very infrequent and when we are talking about hypospadias we are usually talking about a series of abnormalities of the external genitalia of the male.

In the majority of cases the external signs of hypospadias are obvious at birth but can be missed if a hasty or incomplete examination of the external genitalia is made. It is important for children with hypospadias to be detected early as this allows for a detailed explanation of the condition to be made to the parents and appropriate surgical management to be initiated before the child is two years of age. This is important as it means the child can undergo reconstructive surgery prior to being continent.

History

There is a rich history in the medical management of hypospadias. There have been many attempts through antiquity to surgically treat hypospadias. The first recorded reports recognising the significance of hypospadias were made by Galen in the 2nd century AD, and his contemporary Antyl even recommended amputation of the penis distal to the ectopic urethral opening. Predictably, this was not a very effective treatment but little changed until the middle of the 19th century. There are now a very large number of operations described for the treatment of hypospadias and whilst a detailed description of the operations is beyond the scope of this article, the vast array of different techniques that have been described probably serves only to demonstrate that there is no ideal nor fool proof technique for the surgical correction of this condition.

Anatomy

The male external genitalia are a complex structure. The penis is composed of three tubes; dorsally the two corpora cavernosa sit above a third tube the corpus spongiosum which contains the urethra. Hypospadias is not a single distinct entity but rather a range of abnormalities involving the abnormal development of the corpora spongiosum. In the most severe forms of hypospadias, the corpora spongiosum is absent resulting a perineal urethral opening. The more profound variants of hypospadias are fortunately rare, but consideration of disorders of sexual differentiation must be given when they are seen at birth, as this can be a medical emergency. These boys require urgent assessment, and particular note should be made of presence or absence of adequately descending testicles. Profound hypospadias is also associated with a number of syndromes, so a complete and thorough clinical examination and detailed history taking are vital prior to deciding whether any further investigations are required. This can be a worrying time for families and prompt evaluation and referral to specialist centres is important.

Features of hypospadias

There are five distinct components to hypospadias, and these are all important in the assessment of the condition as well as indicating the likely underlying aetiology of the condition:

The cardinal feature of hypospadias is an ectopic urethral opening which can occur anywhere from the perineum through to glans. The normal or orthotopic urethral meatus is slit-like in appearance and located at the tip of the penis. Minor abnormalities of the position of the urethral meatus are quite common, but if the foreskin is intact at birth, these are often not appreciated until children are older. Fortunately, minor positional abnormalities of the urethral opening are not usually a cause for concern and parents should be reassured that functionally and aesthetically there is no need for surgical correction. Sometimes hypospadias is noted at the time of an elective circumcision for either religious or cultural reasons. In this instance it is important that the child is not circumcized until a thorough assessment of the hypospadias is made by a paediatric urologist, as circumcision is contraindicated in children with hypospadias.

Classification of hypospadias

Commonly hypospadias is classified according to the position of the meatus. Perhaps most usefully for the non-surgeon, it can be divided into *distal* (85%) or *proximal* (15%) hypospadias.

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The most distal form of hypospadias is ***glanular.*** Here the urethral meatus opens onto the glans itself. The most common form of hypospadias is where the opening is positioned at the junction of the glans and the shaft of the penis. This is ***subcoronal*** hypospadias. These two forms of distal hypospadias comprize 80–85% of all cases of hypospadias. They are not usually associated with other problems and remain the most straightforward forms of hypospadias to correct surgically.

As the opening becomes more proximal it can be described as ***distal-shaft*** or ***mid-shaft*** hypospadias. Although these forms of hypospadias are less common they are still often amenable to a single-stage operation to correct the abnormality. Care must be taken in assessing these infants, as the hypospadias can be more severe than it appears at casual inspection and assessing for other genital abnormalities is important.

Proximal hypospadias describes an opening close to the junction of the penile shaft and the scrotum, and where it occurs proximal to this it may be described as ***penoscrotal or scrotal.*** The most severe form of hypospadias is where the opening sits outside the scrotum within the perineum. This ***perineal*** hypospadias is often associated with scrotal abnormalities, most commonly a bifid scrotum. Fortunately such extreme forms of hypospadias are rare, and require specialist reconstruction and assessment. The different forms of hypospadias are shown below in Figure 1.

The second feature of hypospadias is not universal, however, in its absence it is not possible to detect the hypospadias at birth. The foreskin is usually incomplete on the ventral surface, giving rise to what is described as a ***hooded foreskin.*** Hypospadias in the absence of this defect is termed hypospadias with intact prepuce and is uncommon. Often, this is detected once a child has had the foreskin retracted for the first time. As previously mentioned if this is noticed at the time of circumcision it is imperative that this should not proceed until the child has been assessed by a paediatric urologist at the foreskin is often used in the operative repair of hypospadias.

The third feature of hypospadias is the most important functionally and that is the downward curve of the penis due to the replacement of the normal corpus spongiosum with scar tissue. This is termed ***chordee***, and the correction of this to allow the penis to become straight during erections is an essential component of any surgical repair. It is telling that even the earliest recorded account of hypospadias by Galen recognized this as the most important component of hypospadias resulting in infertility.

The fourth feature of hypospadias is often overlooked on superficial examination, but can be profound. There is often a degree of ***penoscrotal transposition***. The uppermost portion of the scrotum comes to lie above the dorsal aspect of the penis, surrounding it in scrotalized skin. This is an important physical finding as it points to a lack of virilisation of the external genitalia in the male. In the most severe case it is associated with a bifid scrotum.

The fifth and final component of hypospadias, ***urethral hypoplasia***, is also often overlooked as there are no obvious physical signs. The male urethra is often hypoplastic. This is often only obvious at the time of attempted hypospadias repair when a seemingly distal hypospadias is actually much more significant once repair is attempted. The distal urethra is often very shallow and small, and in such cases it is necessary to convert the hypospadias to a more proximal opening before attempting a staged repair.

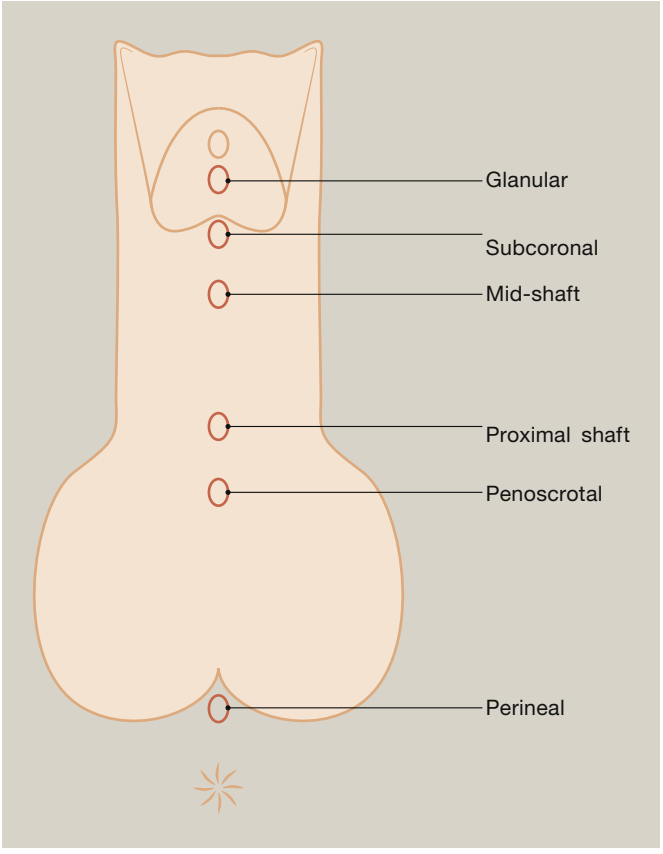

Figure 1 Classification of hypospadias.

Developmental biology

The internal and external genitalia of both sexes are genetically programmed to differentiate as female, however the presence of an active SRY gene on the Y chromosome causes development to move towards the male phenotype. This is a complex series of events, but essentially the primitive tests contains Leydig cells which produce testosterone, that is converted to the more active dihydrotestosterone by 5-alpha-reductase resulting in the normal development of the male external genitalia. Abnormalities in any of the number of complex steps in either production of dihydrotestosterone or inadequate response to this hormone result in inadequate virilisation of the male external genitalia.

From the seventh week of gestation, the urogenital sinus advances onto the developing phallus as the urethral groove. Ingrowth of the urethral groove is associated with the appearance of urethral plate tissue, which subsequently canalizes to form the anterior urethra. Closure of the urethra should be complete from around 15 weeks' gestation.

Assessment of the infant with hypospadias

Hypospadias is a relatively common abnormality, being seen in 1:200–300 live births in the United Kingdom. It appears to be increasing in frequency, although the reason for this is unclear.

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