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Update on the surgical approach for reconstruction of the male genitalia

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ABSTRACT

The majority of patients with DSD will be found to carry an XY karyotype and be assigned male gender. From a phenotypical standpoint, most will present with proximal hypospadias ± cryptorchidism. In this review article, the authors present the current status of reconstruction of the male genitalia in this setting. The report addresses the following topics: surgical input in the evaluation of the newborn with an undervirilized external genitalia, including gender assignment considerations; controversies surrounding timing and indication for hypospadias surgery in proximal cases as well as use of testosterone; surgical techniques and decision-making process for one- vs. two-stage repairs; complications of hypospadias surgery based on technique used for repair; and long-term follow-up. The high complication rates observed in the treatment of proximal hypospadias attest to its challenging nature. Concentration of experience, tracking carefully identified patient-centered outcomes and long-term follow-up of this patient population are recommended.

Most patients identified as having ambiguous genitalia at birth will eventually be categorized as undervirilized males with an XY disorder of sex development (DSD).[1] Given that most of these babies will be raised as males (see discussion in detail below about gender assignment), specific findings pertaining to the external genitalia that require surgical attention will include the presence of hypospadias (which almost invariably will be of the proximal variant) and uni- or bi-lateral undescended testicles. This report will focus on the surgical management of this subgroup of patients, specifically the decision-making process and surgical approach to reconstruction of the male genitalia.

Surgical evaluation of the newborn with an undervirilized external genitalia

Although no surgical intervention is required in the neonatal period, pediatric urologists/surgeons will frequently be consulted in cases of babies born with ambiguous genitalia. Most of these patients will present with hypospadias that tends to belong to the more severe end of the spectrum (penoscrotal or perineal—Fig. 1). Nonetheless, the presence of a proximal urethral opening does not affect micturition and functional issues are not to be expected in the early stages of life.

Assessment of the position of the gonads is a key aspect of the initial evaluation as it heavily influences the direction of further investigation. We have documented in the past that in patients with severe hypospadias and palpable, bilateral scrotal gonads (testes) on physical examination, 100% were found to bear an XY karyotype, received male gender assignment and underwent hypospadias repairs.[2] Nonetheless, up to one-third of patients born with proximal hypospadias and at least one gonad in an non-scrotal position can have an abnormal karyotype,[3] with the yield being even higher when such gonad is non-palpable.[4]

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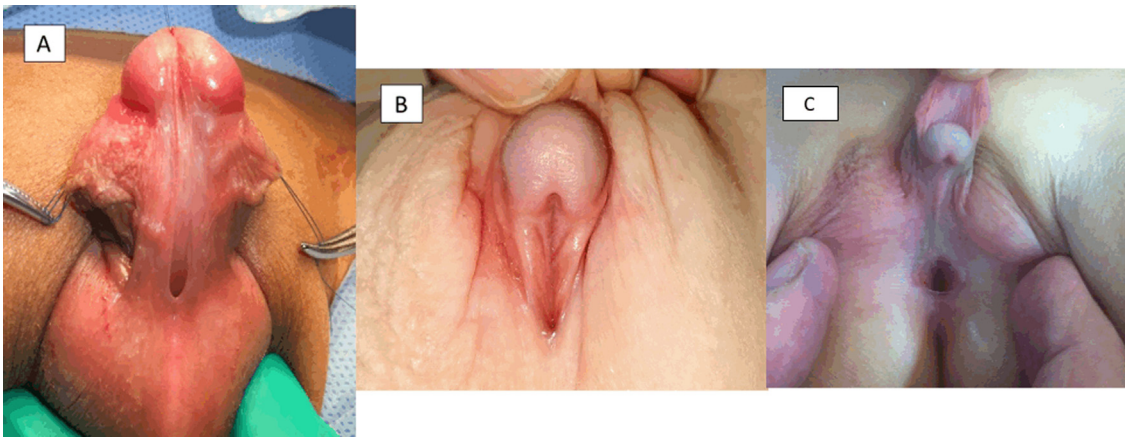

Fig. 1 – Examples of proximal hypospadias.

Gender assignment considerations

In severe cases of undervirilization and XY DSD, the surgical team may be called upon to weigh in on gender assignment decisions based on the feasibility of surgical reconstruction. The "nurture vs. nature" approach used in the past, where patients in this situation would be assigned female gender due to ease of reconstruction, has shifted significantly in the last decades. In undervirilized XY males with the presence of testicular tissue and some amount of phallic tissue, the current trend is for male gender assignment, particularly if adequate response to stimulation of the pituitary-gonadal axis is demonstrated (i.e., rise in testosterone and increase in phallic size after stimulation test with gonadotropins).[5–7]

Based on registry data, patients with disorders of androgen synthesis or a partial androgen insensitivity syndrome (PAIS) profile have seen substantial increase in the rates of male gender assignment over the last 3 decades.[8] Hence, from a surgical standpoint, most of these patients will be seen initially as part of a multidisciplinary consultation geared at reaching an accurate diagnosis and offering advice on gender assignment. After that, they will be referred for repair of their proximal hypospadias as well as orchidopexy(ies) for their undescended testis(es). Less often, penoscrotal transposition may also warrant surgical intervention.

Timing of re-evaluation and surgery

A surgical clinic visit around the age of 3–6 months allows the parents to process the overwhelming amount of information received in the perinatal period related to the DSD investigation, establish a strong bond with their child and have a more focused discussion about future surgical plans. The age of 3–6 months is appropriate because a significant proportion of these patients will have undescended testes and the ideal age for orchidopexy is 6–18 months based on the most current guidelines.[9,10]

Historically, a similar age has been recommended for hypospadias repair predicated on the fact that infants tolerate the procedure well and benefit from superior wound healing and hence decreased complications compared to older children and adults.[11,12] Nonetheless, the discussion around the ideal timing for hypospadias repair has increased in complexity in the last decade from an ethical perspective.

On one hand, the indications for surgery in proximal hypospadias are almost undisputable due to the functional issues associated with the condition. Sexual intercourse may be hindered both in terms of penetration and adequate ejaculation, the former due to ventral curvature ("chordee") often associated with the condition and the latter due to the actual position of the urethral opening. The inability to void standing up can also be listed as a social limitation associated with this type of hypospadias. On the other hand, some have argued that sexual function will not be established until adulthood and that only the patient himself has the ability to consent to a surgical intervention.[13]

As with any DSD/intersex condition the issue of timing and need for surgical intervention is a delicate one and certainly not clear-cut. The position of the authors is that the two sides of the controversy must be clearly presented to the family in the most unbiased way possible to assist in their decision to proceed with surgery or not.

Preoperative administration of testosterone

Historically, hypospadias surgeons have used testosterone before the repair of severe cases with the goal of increasing the size of the penis and making the tissues more robust to tolerate surgical handling. No standardized criteria for testosterone administration, dose or route (intramuscular vs. topical) have been established. In recent years, some authors have raised concerns about the use of testosterone with the argument that wound healing might be compromised.[14] Such a hypothesis is hard to prove since testosterone administration has never been studied in the context of a well-designed trial and surgeons are naturally biased to offer testosterone to cases perceived as being more challenging.

A recent systematic review and meta-analysis of 11 studies on this topic concluded that the quality of the literature is poor and the exact effect of testosterone on wound healing and complications after hypospadias surgery is currently