Reconstructive surgery for major sexual congenital anomalies in adults

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Summary

Objective. — Major sexual congenital anomalies can be traumatic as they are associated with dissatisfaction with body image, low self-esteem, depression and anxiety and as they can damage psychosexual development, deteriorate sexual function, long-term relationships and reproduction. The purpose of this article is to describe the surgical solutions offered to overcome these anatomical anomalies and describe the results of these interventions.

Method. — A PubMed review of the literature on micropenis, bladder extrophy, hypospadias and epispadias was performed along with a collection of our surgical experience at the Hospices Civils de Lyon (France).

Results. — Empirical studies along with our surgical experience show that corrective or reconstructive surgery often improves patient satisfaction with the appearance of their genitals, improves body image and self-esteem and remedies sexual dysfunctions related to penetration and resulting from a small or deviated penis. However, surgery or congenital anomalies can have negative impacts on ejaculation and fertility.

Discussion. — The results from studies and from our experience advocate surgical intervention to correct various types of congenital anomalies as they result in psychological, social and sexual improvements for many if not most of these patients, even if these procedures are difficult and associated with complications.

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The consequences of major congenital sexual malformations can be sexually traumatic as they cause poor body-image, delayed or immature psychosexual development and sometimes have functional or painful consequences that can have a detrimental effect on sexual activity, relationships with the partner and fertility. These consequences can be divided into main ‘syndromes’ as follows:
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- late onset of sexuality due to feelings of abnormality and fear of mockery from others;
- difficulty in changing partner because any change implies a phase of explanation of the disease plunging the patient back deep into his original pain;
- increased frequency of functional disorders and sometimes sexual orientation.

In addition to hormone treatment, which can be offered for development anomalies and pharmacological and/or mechanical treatments for sexual dysfunctions, surgical solutions are available to correct genital anomalies and improve sexual functions. The main difficulty in treatment stems from the patient's expectations for "normality" from these surgical interventions which are extensive and which do not always give satisfactory results from a functional standpoint.

**Isolated micropenis, congenital or the consequence of circumcision or other trauma**

Isolated micropenis can take the form of a "buried" penis (Cromie et al., 1998; Ghanem et al., 2007; Perović, 1995), deformed penis, or in the form of a stump (Lumen et al., 2008; Perović, 1995; Zenaty et al., 2006) (Fig. 1). The size of the penis is generally insufficient for vaginal penetration or to allow the patient to urinate standing up. Isolated micropenis, and micropenis as the consequence of circumcision or other trauma are different from micropenis associated with other pathologies: micropenis, urinary anomalies and undervirilization as in the case of gonadal dysgenesis (Nihoul-Fékété et al., 2006; Sharma and Gupta, 2008; Wu et al., 2002), androgen insensitivity syndromes (Mazur, 2005; Wisniewski and Migeon, 2002), and pseudohermaphroditism or true hermaphroditism (Nihoul-Fékété et al., 2006; Sharma and Gupta, 2008), where treatment is more complex. Isolated micropenis is a hypoplasia of the phallus with a normal urinary meatus where the genitals are definitely male beyond doubt. From the embryologic standpoint, development is normal at least until the 12th week of development, then a deficiency appears, preventing further normal development of the penis. Patients suffering from micropenis are unhappy with the appearance of their penis, the distribution of hair and its juvenile appearance (Wisniewski and Migeon, 2002; Wisniewski et al., 2001), which can in itself cause low self-esteem and sexual and relationship problems (Lumen et al., 2008) requiring psychosexual help (Ravish et al., 2007). An isolated micropenis does not always cause major sexual dysfunctions, with some patients reporting good erections (Wisniewski et al., 2001), in some cases normal and quite satisfactory sex lives, and relationships within a couple in 75% of cases (Woodhouse, 1994). Patients with micropenis do not suffer from gender identity disorders (Mazur, 2005; Nihoul-Fékété et al., 2006; Wisniewski and Migeon, 2002; Wisniewski et al., 2001), their sexual orientation is comparable to that of the general population, with most of them considering themselves to be heterosexual with a few patients homosexual or bisexual (Wisniewski et al., 2001).

**Surgical treatment of micropenis**

In addition to hormone treatments (Ludwig, 1999; Zenaty et al., 2006) and psychosexual support (Ravish et al., 2007; Wisniewski and Migeon, 2002), there are surgical solutions on offer to correct micropenis (Byun et al., 1994; Cromie et al., 1998; De Fontaine et al., 2001; Ghanem et al., 2007; Gilbert et al., 1993; Lumen et al., 2008; Perović, 1995). As patients mainly complain of the size of the penis, the main surgical treatments focus on penile enlargement.

**Cutaneous plasty with release of the suspensory ligament**

Cutaneous plasty is technically simple with an inverted V-shaped cutaneous incision made in the pubis, then dividing of the suspensory ligament up to the pubis and sectioning of the ligament close to the pubic bone (Fig. 2). After the surgery, inverted Y sutures are made, extending and...
dropping the penis downwards (Fig. 3). The lengthening of the penis remains moderate, between 1.3 and 2.4 cm; the results are variable according to literature (Li et al., 2006; Spyropoulos et al., 2005; Panfilov, 2006).

Dropping of the penis with skin graft and soft implants

Dropping of the penis consists in releasing the corpora cavernosa and the corpora spongiosa from their attaches. Sometimes this is accompanied by a lipectomy of the mons pubis so as to reduce the amount of prepubic fat and better enable the penis to stand out. Then plasty is conducted with skin graft (full thickness or split skin), and sometimes a soft implant is used in one of the corpora cavernosa to maintain the lengthening effect.

“Apollo” tissue expander

This technique is not used very often. It consists in fitting a telescopic prosthesis that enables the penis to be gradually lengthened. This technique enables the penis to be lengthened by between 1 and 5 cm if there is not too much scarring. This technique has been used successfully by our team, but presents certain drawbacks, notably the risk of erosion and the need for a prosthesis to obtain erection.

Phalloplasty

There are a large number of phalloplasty techniques. Two of the most common techniques are antebrachial phalloplasty and abdominal phalloplasty.

Antebrachial phalloplasty (radial forearm phalloplasty)

Antebrachial phalloplasty remains the main golden standard technique, despite the fact that it is technically difficult and extensive (Bodlund and Kullgren, 1996; Leriche et al., 2008; Lumen et al., 2008; Timsit, 2004). It consists in taking a flap of skin from the forearm which is vascularized by the radial artery and the vena cephalica and innervated by the nervus peroneus superficialis. The flap is microanastomosed with the epigastric artery, the saphenous vein and the dorsal nerve of the penis. A neo-urethra is formed using skin from the flap.

This technique enables the penis to be lengthened by 12 to 15 cm. Rigidity is achieved by the use of an inflatable prosthesis. The results (Bodlund and Kullgren, 1996; Leriche et al., 2008; Timsit, 2004) are generally good in terms of lengthening of the penis with only 4 to 6% of flap loss, but there are often complications (15—50%) with the urethra and the prostheses. The appearance of the penis is satisfactory to the patient in 90 to 100% of cases, but there is an unattractive scar on the forearm.

Abdominal phalloplasty

Abdominal phalloplasty is transfer of skin to the sub-pubic part of the body in several stages. The first stage consists in placing subcutaneous sub-umbilical expansion balloons, the second involves the fabrication of a tube which will become the penis and which is left in place several months. The third stage involves the release of the upper part of the tube. A prosthesis is fitted at a later stage. The drawbacks are the length of time necessary between each stage, unattractive scarring on the abdomen, a penis located slightly higher that normal, and the impossibility at the current time of making a satisfactory urethra. In our series of patients, we have 100% of existing flaps, but nearly 10% of the patients are not entirely satisfied with the results from an appearance point of view.

Exstrophy of the bladder

Exstrophy of the bladder is a congenital malformation of the bladder and the urethra associated with a malformation in the genital organs. The consequences of exstrophy of the bladder include dorsal curvature of the penis and a small penis (Fig. 4), with the corpora cavernosa often set rather
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far apart, giving an overall impression of the penis having a pyramid shape.

Studies show that exstrophy of the bladder causes anxiety (Reiner et al., 1999), depression (Mukherjee et al., 2007; Reiner et al., 1999), social difficulties for 25% of adolescents, relationship problems for 59% (Ebert et al., 2005). At the adult age, 28% of patients are in a long-term relationship (Baird et al., 2004).

Satisfaction with appearance and functions of the genital organs is low for these patients (Ebert et al., 2005; Reiner et al., 1999). In terms of sexual activity, few adolescents and young adults want to have sexual relations, and only 14% of them have masturbated (Reiner et al., 1999). Sexual dysfunctions (difficulties with penetration, ejaculation difficulties), relative insensitivity to touch of the genitals, and fertility disorders have all been noted (Reiner et al., 1999) and only 18 to 50% of patients according to the most optimistic results (Woodhouse, 1994) can have children. Some earlier studies suggested a higher level of sexual satisfaction, with 77 to 83% satisfied with their sex-life with or without surgery (Woodhouse, 1994; Ben-Chaim et al., 1996; Mollard, 1990). In our experience, we have noted only occasional sexual activity.

The consequences of exstrophy of the bladder are such that early psychological support and surgical intervention are recommended (El Khader et al., 2003). Surgery, notably penis enlargement, improves sexual relations with penetration (Audry et al., 1991; Baird et al., 2004). It is associated with a high level of satisfaction in the appearance (94%) (Stein et al., 1996, 1997) and improves the body image and the psychosocial integration of the adolescents (Baird et al., 2004; Meyer et al., 2004). Surgery for bladder exstrophy improves penetration but is associated with ejaculation disorders, leading some authors to suggest that the high level of satisfaction in appearance is achieved to the detriment of fertility (Audry et al., 1991; Stein et al., 1996, 1997). The fertility disorders are due to epididymitis and orchitis (Stein et al., 1996, 1997).

Surgical treatments for exstrophy of the bladder

Exstrophy of the bladder entails complex problems with the length, diameter and the curvature of the penis, in addition to erectile disorders and urinary incontinence. Urinary incontinence can be treated by transileal ureterostomy (e.g. Bricker type), but this is not without psychosexual effects. The sexual problems are as much related to the inability to meet a partner as to a sexual dysfunction.

Repair and reconstructive surgical techniques give good results from an aesthetic point of view (Kramer et al., 1986; Meyer et al., 2004) and from a functional standpoint too, with improvements in the levels of sexual satisfaction (Kramer et al., 1986), but the patients retain a certain level of anxiety about their sexual relations (Meyer et al., 2004).

Treating deviated penis

Classic surgical treatment involves techniques of plication and plication-excision or incision (such as Nesbit or Yachia procedures) on the convex side. Other techniques involve incision with graft on the concave side of the penis so as to reduce the curvature.

For young patients, plication-excision techniques are preferable to graft-based, because the size of the penis is reduced and the dissection of the urethra is complex and risky in terms of secondary fistulae.

Treating pyramidal inconspicuous penis

The penis drops by fully releasing the corpora cavernosa and spongiosum from their attaches and their retractile scars and bringing the corpora cavernosa closer together so as to lengthen the penis and make it more functional. Sometimes soft implants are positioned to maintain the lengthening effect. This technique is unfortunately not often very effective for patients suffering from exstrophy of the bladder.

Phalloplasies

Antebrachial phalloplasty is the procedure most generally used, enabling a neo-urethra to be formed, and the flap to be re-innervated (Fig. 5) (De Fontaine et al., 2001). The corpora cavernosa are used to insert the future inflatable prosthesis, enabling the penis to become rigid. The ablation of the glans results in significant loss of sensitivity on the penis and sometimes difficulties in reaching orgasm.

Treating reproductive disorders

Stenosis of the urethra is generally treated by laser to facilitate ejaculation. It is important to remember to take and freeze samples of sperm as quickly as possible after puberty before infections and surgery might compromise the patient’s fertility.

Hypospadias

Hypospadias is an abnormally placed urinary meatus, on the underside of the penis. The local anomalies associated with hypospadias are deviated penis, flattening of the glans, stenosis of the meatus, urethral fistulae, difficulties in urinating standing up and prepuce open at the front (Anikwe

![Figure 5](Grafted phalloplasty on the penis of a patient with exstrophy and dysfunctional micropenis.)
Patients with hypospadias have a normal psychosexual development. (Bracka, 1999; Mureau et al., 1995a,b; Moriya et al., 2006; Schönbucher et al., 2008). They have a satisfactory sex life (Aho et al., 2000; Bubanj et al., 2004; Nelson et al., 2005), although severe cases of hypospadias have a more negative image of their genitals, they can be dissatisfied with the emptiness of their bladder and with the size of their penis. They have more inhibitions than others in having sexual relations (Aho et al., 2000; Bracka, 1999; Dodds et al., 2008; Moriya et al., 2006; Mureau et al., 1995a,b, 1996). Appearance is important for 72% of them and 44% would like corrective surgery to improve this aspect. (Bracka, 1999). A ventral curvature greater than 30° is found in 18% of cases (Bracka, 1999) and can hinder sexual relations. Seventy-nine percent of patients can have sexual relations, even if they suffer from sever hypospadias (Miller and Grant, 1997). Their first sexual relations take place later, (Moriya et al., 2006; Schönbucher et al., 2008) and intercourse is generally less frequent, or involves fewer sexual partners, there are fewer married men (Aho et al., 2000; Bracka, 1999; Bubanj et al., 2004; Miller and Grant, 1997). Men suffering from hypospadias rarely have erection or orgasm disorders (Bubanj et al., 2004; Nelson et al., 2005), although there are some cases of anejaculation, premature or asthenic ejaculation (Bubanj et al., 2004; Nelson et al., 2005). Men suffering from hypospadias are more masculine (Sandberg et al., 1995; Schönbucher et al., 2008). They demonstrate emotional instability in 33% of cases (Bracka, 1999).

Raising awareness with patients and providing them with information fosters a more positive perception of their penis (Schönbucher et al., 2008). These patients are often reticent in trying to obtain help in overcoming their difficulties (Mureau et al., 1995a,b) and they do not all want repair surgery (Anikwe et al., 2000; Dodds et al., 2008).

Surgical treatment for hypospadias

Correcting the curvature and urethroplasty

Straightening of the penis by excision of the pre-urethral fibrous tissue, with artificial erection test to judge the quality of the surgery. At this point a decision is taken concerning the urethroplasty technique, which are too numerous to cover them all here.

Classical techniques of plication and excision are proposed, despite the fact that they reduce the size of the penis. Sometimes a complex and risky dissection of the urethra is necessary for severe curvatures with small penises.

Improvement of appearance

Classical lengthening plasty techniques are used depending on "reasonable" requests on the part of the patient.

Epispadias

Patients suffering from epispadias complain of the pyramidal shape and small size of the penis, the short urethra and the opening on the dorsal side of the penis, the curvature which can cause dissatisfaction with the patient’s image of his body and self-esteem issues, and anxiety about sexual relations (Ebert et al., 2005; Gearhart et al., 1995; Meyer et al., 2004; Stein et al., 1996, 1997). Post-surgical complications are urinary with fistula, stenosis, recurrent epididymitis and orchitis (Audry et al., 1991; Gearhart et al., 1995; Stein et al., 1996, 1997). Surgery can improve the patient’s satisfaction with the appearance of their penis, their body image and self-esteem (Meyer et al., 2004). It can leave sequelae in terms of ejaculation and reproduction (Audry et al., 1991; Stein et al., 1996, 1997). Treatment of urinary disorders must take place before or during the treatment of the small penis, but should always be taken into account.

Surgical treatment of epispadias

Treating the small penis

Dropping of the penis or phalloplasty (see exstrophy).

Improvement of appearance

The treatment consists of glanuloplasty with displacement of the urinary meatus to the ventral side of the penis, and repair to the skin if necessary.

Conclusion

Major sexual congenital malformations can cause sexual trauma by damaging the patient’s image of his body, his psychosexual development, and sometimes his sexual functions. Surgical solutions can be used to correct these malformations and restore some functionality, although the morbidity is high. The results of empirical studies and our own clinical experience show that repair surgery improves the psychological, social and sexual functioning of the patients, although the expectations of normality from these extensive forms of surgery are not always as satisfactory as hoped.

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