The sexual consequences of congenital pelvic anomalies

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Congenital pelvic anomalies are relatively rare in males. Little is known about their impact on sexual function and activity as these boys grow into men. In this article the author looks at the data available and encourages health professionals who look after such patients not to be afraid to discuss sex.

There is a range of internal and external congenital anomalies in the male pelvis, which are linked by the impact they have on sexual and reproductive function in adults. In addition, they share, to a large extent, their innervation from the lower spinal segments so that neurological anomalies generate similar problems. Unfortunately, there are few reliable data on the sexual consequences of these anomalies and how to manage them.

UNDESCENDED TESTES

Although a testicular dysgenesis syndrome has been described to account for adverse trends in male reproductive health, in practice men who have had successful surgery for undescended testes (UDT) in infancy are normally masculinised and no specific sexual dysfunction has been reported.

Up to 17% of infertile men have a history of some UDT, which is much higher than the incidence of UDT at birth. Often the details of the original surgery are unrecorded.

However, where it has been possible to follow a cohort of children into adulthood, paternity is reported in 80–90% of men with unilateral and 33–65% with bilateral UDT. In unilateral cases at least, the trying time and the paternity rate are no different from unaffected controls.

Infertility in such men can be managed with the standard techniques. Most have low sperm counts rather than azoospermia, and even with unoperated bilateral UDT, some sperm have been retrieved for intra-cytoplasmic sperm injection (ICSI).

PENILE PROBLEMS

Hypospadias

This affects one in 200 to 300 boys. Unfortunately, evidence-based follow-up is only available for children and hence does not consider sexual outcomes. Where a protocol follow-up into adult life has been reported, about 15% of patients have late problems. Most are cosmetic and surgery is seldom required. In men who have had a good surgical result from an operation performed in the best childhood window (5 to 15 months of age), general psychological problems are no more common than in controls.

Figure 1. Clinical photograph of a man with a micropenis of unknown aetiology

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It is difficult to be certain about sexual function in men with well-corrected hypospadias. The milestones of masturbation, sexual kissing and intercourse are at the same ages as for controls. The most common problem is poor ejaculation, as the reconstructed urethra has no spongy erectile tissue around it to propel the seminal bolus. The more proximal the original hypospadias, the worse the problem. In meta-analysis, 26% of men reported the problem, but many were sexually satisfied nonetheless. Other problems include erectile dysfunction (19% versus 1% in controls) and sexual inhibition (19% versus 9%).

From the strictly 'physical' point of view, the penis with hypospadias works normally. If there are no associated anomalies, such as undescended testes or disorders of sex development (DSD), fertility is normal despite the low sperm counts reported. The risk of a baby having hypospadias may be as much as 20% higher than normal if a first-degree relative is affected.

Micropenis

Micropenis is defined as being more than two standard deviations below mean stretched length for age with otherwise normal anatomy. At birth, it may be a part of a specific endocrine problem, in which case it may become normal with treatment. Most cases are idiopathic and the man will grow up with a micropenis (Figure 1).

The number of affected men available for study is small. Voiding, erection and orgasm are normal. Several studies have shown normal psychosexual development, intimate relationships and satisfaction with genital function. Virtually all are content with their gender and there are no records of adult men seeking gender reassignment. Men report normal libido and sexual activity that is largely heterosexual, and form close and durable relationships.

There is no good way of enlarging the micropenis in adulthood. Phalloplasty, as for female-to-male gender reassignment, has been used with good results, although patients have been disappointed with poor erogenous sensation compared to their original penis.

THE PROSTATE

The prostate is an organ of little interest to paediatricians. The most common congenital anomalies are due to persistence of Müllerian structures, such as prostatic utricles, which generally cause no problems. The rarer but important condition of fetal posterior urethral valve (PUV) produces gross prostatic urethral dilatation as well as major upstream problems.

The prostate's sexual function is to make semen and create the initial force for ejaculation. Conditions that reduce the volume of prostatic parenchyma will reduce semen production and may contribute to infertility. Dilatation of the prostatic urethra reduces the force of ejaculation or may abolish it altogether (Figure 2). The first stage of ejaculation is to build up a bolus of semen in the prostatic urethra, which is then forcibly expelled. With a dilated prostatic urethra such force cannot be generated so that the expulsion is slow, analogous to trying to fire a .22 bullet from a .303 rifle. (a) is republished with permission from Adolescent Urology and Long Term Outcomes by CRJ Woodhouse, Wiley, Oxford, 2016; (b) is published courtesy of Dan Wood

Increased capacity of the prostatic urethra occurs with PUV and with Müllerian remnants that communicate directly with the urethra. In the former, the high pressure within the prostatic urethra during fetal life often causes poor prostatic development and thus reduced seminal volume. The semen that is produced may be abnormal in up to 75% of men. The impact of these abnormalities on fertility is uncertain, especially as at least one third of them develop end-stage renal failure by the age of 30. In those who have normal renal function, paternity is near normal, although the trying time may be prolonged.

Congenital abnormalities of the prostate or bladder neck may present in adult life with absent or reduced ejaculate volume. Investigation by transrectal ultrasound (TRUS) will often give the diagnosis. Sperm may be retrieved from the urine after orgasm and used for ICSI.

THE BLADDER

Exstrophy

About 12 babies a year are born in the UK with exstrophy, three quarters being male. In this condition, the bladder comprises only a portion of the posterior wall and is...
exposed due to divarication of the rectus muscles and absence of the overlying fat and skin. Bladder reconstruction is becoming progressively more sophisticated and, by one means or another, almost all those affected achieve continence by adult life, although many have to empty by clean intermittent catheterisation. Despite the severity of their anomaly and its treatment, they tend to be well motivated and high achievers.

The penis is short and broad. There is a tight dorsal chordee, which must be corrected to allow penetrative intercourse (Figure 3). The testes are normal, provided they have not been damaged by common childhood infections.

Erectile function and frequency are normal, and libido is high. Masturbation is universal and 75% have some seminal emission. The most common problem is fear of rejection by a potential partner due to the obvious penile anomaly and extensive abdominal scarring. Cohabiting partnerships are formed by about 75% of men and seem to include a satisfactory sexual component, although, as usual, there are no data on the partner’s satisfaction.

One third to half of men with exstrophy achieve paternity naturally. These figures can be improved by teaching them to collect their semen in a syringe and gently inject it into the vault of the vagina. Otherwise, standard management of infertility is needed.

Spina bifida
The incidence of spina bifida (SB) is decreasing. However, neonatal and childhood care has improved, so that more patients are surviving into adulthood. The critical issue is the management of bladder dysfunction, which, alongside the abnormal innervation of the genitalia, has a profound impact on sexual function.

The physical aspects of sexual function that depend on the brain, such as sexual identity and libido, are normal, but the emotional aspects may be impaired, especially when IQ is low. Difficult areas of community policy include the prevention of unwanted pregnancies and sexual abuse. A survey in 1999 showed that only 5% (of both sexes) of those with SB had adequate knowledge of sex.

Genital function is dependent on the level of neurological damage. Those with a level at L2 or below or with urinary continence are thought to have normal sexual sensations and responses, whereas only around 20% of those with higher-level damage or incontinence do so. All those with urinary continence and intact sacral reflexes have erections and ejaculation.

The extensive disabilities caused by SB may be more important than genital function in determining sexual activity. Fear of incontinence inhibits the initiation of intimate relationships. In a Dutch survey, although all males without hydrocephalus claimed to have had intercourse at least once, only 65% said that they were satisfied with their sexual function (only 45% of those with hydrocephalus).

Impotence responds to small doses of the standard medications. Men who do not have natural erections and ejaculation are often azoospermic. There is a high risk of a neural tube defect in the offspring of affected males at 1:23. The female partner must start prophylactic folic acid 5mg daily at least three months before attempting conception.

ANORECTAL MALFORMATION
Anorectal malformation (ARM) includes a very wide spectrum of anomalies. The simplest is a membranous obstruction at the anus and the most severe is an association with anomalies of the oesophagus, heart, duodenum, vertebrae, spinal cord, kidney and limbs (VACTERL). Survival and long-term outcome are dependent on the original severity of the anomalies, making assessment of sexual functioning difficult.

Studies of adults with ARM have small numbers, even when the original database from which they are identified lists several hundred patients. A consistent feature is that men feel they have had inadequate education on the sexual consequences of their anomaly. However, as usual with congenital anomalies, the sexual interests of the adolescent survive remarkably well. From the scarce data on adults, it seems that none have objectively normal anal function. Less than 20% have good continence and up to one third have frequent soiling or a colostomy. Almost nothing is recorded on the control of flatus, although the clinical impression is that it is usually poor and is a particular intercourse-limiting worry.

Up to one quarter of boys with ARM have an associated genital anomaly. It is often fairly minor, although hypospadias, cryptorchidism and DSD are recorded. Occasionally, the genital nerve supply is damaged at surgery in childhood. The most common associated malformations are in the spine, including hemivertebrae, but extending to absent segments and caudal regression syndrome.

A survey of those with ARM in the Netherlands showed that 24% had never had a durable relationship. As a ‘social’ issue, the high risk of leakage of faeces and flatus is certainly a bad start for a man wishing to share his life and bed with another. When spinal lesions are present, as
with SB, those who are continent and have intact sacral reflexes should be potent, as the neurological level of damage should be below D10. In the small number of patients studied, the rate of erectile dysfunction is between 6% and 41%. Minimal or dry ejaculation is reported by 10%. In a very careful review of 40 men, there was no relationship between the type of ARM and quality of life or sexual function. Poor anal control was the most limiting factor.

Some men born with ARM have fathered children. Failure to initiate a pregnancy may be due to several factors. Up to 70% live alone; the associated anomalies, particularly undescended testes and neurogenic impotence, may cause low sperm counts or azoospermia; and reconstructive surgery or recurrent urinary infections may obstruct the passage of sperm. In a series of 32 men, 13 were having intercourse, of whom three had initiated two or more pregnancies.

**COMMON THEMES**

A pelvic anomaly is a frequent cause of anxiety and depression, a significant part of which is related to sexual functioning. Most adolescent patients and their parents want to discuss these issues, but are afraid to ask – and most doctors are afraid that they will ask. In recordings of adolescent consultations when sex was discussed, it was for an average of 36 seconds. It is essential that all doctors charged with the care of such patients create an environment in which sex and fertility can be discussed as puberty develops. It should always be presumed that boys with even the most major anomalies will be interested in sex, able to have sex and be fertile. As some conditions have a hereditary link, appropriate genetic counselling should be a standard part of long-term care.

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**REFERENCES**


**KEY POINTS**

- Disability does not mean celibacy
- Assume that sex is desired and possible
- Assume fertility is normal until proved otherwise
- Facilitate sexual education
- Treat erectile dysfunction and infertility by standard protocols
- Arrange genetic counselling when appropriate

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