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Childhood surgery for ambiguous genitalia: glimpses of practice changes or more of the same?

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The Chicago consensus statement of 2005 was created at the point of cumulative criticisms and debates around the clinical practice of childhood genital surgery. It was drawn up at a time when it had become clearer that medically non-essential paediatric genital operations were associated with poor adult cosmetic outcomes and sexual functioning. However, data were not available for non-intervention. Therefore, parents and clinicians had no reliable information on how a child growing up with atypical genitalia might fare. The most positive recommendation in the consensus statement was the strong recommendation for decisions to be reached by a multidisciplinary team in collaboration with affected families. Importantly, the value of user groups was likewise formally acknowledged. For many services, there has been a sea change in the way surgeons work. Whilst some surgeons may continue with the standard practice of childhood genital surgery, it is becoming clearer that with adequate support, more individuals and families choose to postpone elective interventions. However, these are our observations only. Authoritative evidence must be based on high-quality multi-centre multidisciplinary research to prospectively monitor the long-term multiple outcomes of surgery and no surgery. There is as yet no obvious move towards such an endeavour.

Keywords: DSD; intersex; ambiguous genitalia; childhood genital surgery

Introduction

The most controversial area of clinical management for Disorders of Sex Development (DSD) management is that relating to medically non-essential feminising genital surgery for infants and children with atypical genitalia. Standard medical practice has leaned towards early feminising surgery for all children assigned to the female gender. However, persistent concerns expressed by adults who had been the recipients of such interventions have prompted clinicians to examine outcomes.

Since the late 1990s, a flurry of publications has appeared in the medical literature to that effect. Whilst the reports were mainly retrospective or cross-sectional and varied in scientific merit, they have highlighted the lack of an evidence base to support early surgery. From about that time, a schism has developed between clinicians working in paediatric and in adolescent/adult services. This has led to the defence of standard surgical practices

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by some paediatric clinicians against increasing interrogation of the practice by clinicians looking after adolescent and adult patients.

The 2005 Chicago consensus group had a difficult task of addressing feminising genital surgery in the absence of authoritative data. The main consensual recommendation was that surgery should only be considered where the development of male-typical sex characteristics is significant in girls. It remains too soon to estimate the impact on clinical outcomes. The aim of this article is therefore to present the surgical situation for children and adolescents before and after 2005, and to explore whether there are signs of change in surgical practice. The focus is on girls and women as they are most likely to be recipients of infant surgery.

Genital surgery prior to Chicago

On the infant

Until recently, when atypical genitalia were identified at birth, a sex of rearing would be assigned as soon as possible, taking into consideration future potential for secure gender identity and sexual function. The determining factor in decision making was the size of the phallus. It was generally assumed that a boy with a small or absent penis would have poor developmental outcomes and, as phalloplasty was not particularly successful, the majority of children with a DSD were assigned as female – approximately in the ratio of 9:1 (Newman, Randolph, & Anderson, 1992). This included boys with a problem with enzymes that are needed to form androgens, the hormones that stimulate development of male-typical sex characteristics, and also boys born without a penis or with a very small penis who, with current assisted reproductive methods, are potentially fertile.

Feminising surgery was typically performed as early as possible to align the genital appearance. Normalised external genital appearance was deemed necessary in order to reinforce gender identity. The work of John Money and his colleagues in the 1960s and 1970s lent support for the practice. These workers suggested that the child is psychosexually neutral until the age of 2 years but from then on, extrinsic factors such as genital appearance, choice of clothing, parental attitude and social behaviour towards the child reinforced the assigned gender (Money, Hampson, & Hampson, 1955). Guidelines provided by the European and American paediatric endocrine societies are still in effect to suggest genital surgery between 3 and 6 months of age for patients with CAH (Endocrine Society, 2010; LWPES & ESPE, 2002). Surgeons have based their support on a number of unsubstantiated claims, which include better healing ability in an infant leading to less scar tissue and better cosmetic results, beneficial effects of oestrogen on tissue and avoidance of potential complications due to connection between the urinary tract and peritoneum via the fallopian tubes (Adzick et al., 1985).

Feminising surgery includes surgery to reduce the size of the clitoris and vaginoplasty to create a vagina or enlarge an existing one. Clitoral reduction surgery gradually evolved from clitoridectomy (amputation) to clitoral recession and then to clitoral reduction as an attempt to preserve sexual sensation. In clitoral recession surgery, a procedure now mostly abandoned, clitoral corpora are dissected, spared and anchored to the undersurface of the pubic bone (Newman et al., 1992). Whilst this may preserve vascularity and innervation, clitoral recession can lead to pain and discomfort secondary to tumesence during sexual arousal. Later surgical revision is often required. Clitoral reduction surgery involves the dissection and removal of part of the clitoral corpora, while retaining the neurovascular bundle at the dorsal aspect of the clitoris.

Paediatric surgeons used to recommend what they called 'one stage' genitoplasty, where a vaginal opening is created at the same time as clitoral surgery. The choice of a vaginoplasty depends on an individual's genital formation. In CAH for example, the operation can range from a relatively simple procedure to open the entrance of the vagina (introitoplasty) to a more complex procedure to separate the vagina from the urethra in patients born with a common channel (urogenital sinus mobilisation) (Rink & Cain, 2008). In women without a vaginal opening, an operation to create a vagina may involve the use of skin graft (McIndoe procedure), a segment of the gut (bowel or intestinal vaginoplasty) or lining from inside the abdomen (peritoneum) (Davydov procedure) These operations are based on the idea of creating a space between urethra and rectum and then lining it with skin, bowel or peritoneum, respectively. Depending on the procedure and the result, post-operatively, parents may be required to insert a mould of graduating sizes to maintain or increase vaginal size for their daughter.

There are some follow-up studies in the literature. These were mostly small case series of unsophisticated quality with a short-term follow-up period (Canty, 1977; Donahoe & Hendren, 1984; Newman et al., 1992; Sircili et al., 2006). For example, evaluation of cosmetic appearance was often carried out by the surgeons who did the operation. Since the rationale for childhood surgery was founded on normal development, the real test of the approach should be based on adolescent and adult wellness. For example, psycho-sexual outcomes by definition can only be studied when individuals have reached adolescence or adulthood – many years after the procedures are performed. However, longitudinal data are scant, and only a small select number of patients were unsystematically followed into puberty. As studies with adults emerged in the literature, a high rate of late onset complications were observed, such as painful intercourse, shrinking or narrowing of the vagina due perhaps to scarred tissue and poor sexual function (Alizai, Thomas, Lilford, Batchelor, & Johnson, 1999; Creighton, Minto, & Steele, 2001).

The widespread practice of childhood genital surgery has meant that there have been very few adults who have not been operated on to enable robust comparative studies to be carried out. Furthermore, until about 1990s, it was typical for doctors to withhold diagnostic and treatment information from girls and women with certain DSD diagnoses (Conn, Gillam, & Conway, 2005; Liao, Green, Creighton, Crouch, & Conway, 2010). When a large number of people operated on as children were unaware of their diagnosis and treatments, it made it impossible for them to be recruited for long-term follow-up studies (Creighton & Liao, 2004).

On the adolescent

Some children whose DSD was identified at birth would have continued to require medical input into adolescent and adult life. However, DSDs may also be diagnosed in early adolescence, perhaps due to atypical or absent pubertal development. In a girl, this may include absence of menstrual periods (primary amenorrhea) and/or vaginal, uterine or breast development or presence of sex characteristics more typical for males, such as enlargement of the clitoris, deepening of the voice and unwanted facial and body hair growth in a male-typical pattern distribution (Lee & Houk, 2008). In a boy, atypical or absent pubertal development may take the form of the presence of breast development (gynaecomastia) or monthly blood loss when passing urine (cyclic haematuria) (Lee & Houk, 2008). Surgery for newly presenting adolescents appears less controversial as the patient is able to be involved in discussions and in decision-making. The consensus document does not make recommendations about surgery in newly diagnosed adolescents.

Adolescent outcomes of infant surgery

The consensus document does recognise that further surgical treatment is likely to be required in adolescence, and surgical reconstruction in infancy will need to be refined at puberty. Long-term outcome data regarding sexual function amongst those with DSD assigned male and female are scanty and results are very variable. The contribution of genital surgery to overall outcome is not clear.

Sexual outcomes

With classical surgical techniques practiced in infancy, it is more a rule than an exception to identify narrowing of the vagina or at the vaginal entrance at puberty. Complementary dilation or surgery using grafts from bowel, skin or more recently lining from the inside of the mouth usually ensues, but the results can often be unsatisfactory (Auchus & Chang, 2010; Bailez, Gearhart, Migeon, & Rock, 1992; Creighton, 2004; Stikkelbroeck et al., 2003). Clitoral surgery in infancy was traditionally performed among those with marked ambiguity, with up to 44% undergoing clitoral revision surgery later in life (Newman et al., 1992).

Paediatric anatomical outcome in infancy is not synonymous with satisfactory cosmetic and sexual outcomes in later life. Indeed, studies have shown unsatisfactory cosmetic results, markedly impaired genital sensitivity and compromised sexual function in adult women following feminising surgery and clitoral surgery (Creighton et al., 2001; Crouch, Liao, Woodhouse, Conway, & Creighton, 2008; Minto, Liao, Woodhouse, Ransley, & Creighton, 2003). Interestingly, in genetic and assigned males presenting with undervirilisation, the genital and sexual outcomes vary according to the degree of hypospadias and the amount of erectile tissue. There are some long-term data to show that men with micropenis can have satisfactory sexual and relationship outcomes (Reilly & Woodhouse, 1989).

Implications of surgery for parenthood

As reproductive technology continues to develop and legislature continues to change, fertility potential for people with certain DSD diagnoses have improved over the last few years. This topic was not directly addressed in the consensus statement. However, fertility potential needs to be considered in all children where surgery could irrevocably remove possibilities in adult life.

Parenting in adults with DSD is affected by intrinsic and extrinsic factors, but may include adoption, surrogacy, assisted reproduction techniques such as *in vitro* fertilisation, ovarian tissue cryopreservation and ovum or sperm donation. Women without a uterus may consider surrogacy or adoption. Women with a uterus but without functioning gonads may consider egg donation. In men with Klinefelter's syndrome (KS) caused by an extra X chromosome, fertility is now possible using modern sperm extraction techniques (intracytoplasmic sperm injection, ICSI). In men diagnosed with KS who produce no sperm in the ejaculate, micro-testicular extracted sperm (TESE) is the only option besides sperm donation (De Sanctis & Ciccone, 2010). In people with both ovarian and testicular tissue (ovotesticular DSD), if gonadectomy is indicated, in the presence of functional tissue, ovarian or testicular tissue could be frozen in the attempt to preserve fertility potential (Nihoul-Fékété, 2004, 2008). Although still being researched, *in vitro* maturation of eggs have resulted in live births.

Genital surgery after Chicago

The multidisciplinary team (MDT)

The most fervent recommendation of the consensus document was to place the surgeon within the framework of a multidisciplinary team with an explicit mandate to collaborate with service users and families. This recognised the need for practice to be based upon a process of discussion and debate, including both the best clinical and scientific evidence alongside patient preferences and circumstances.

Integrated psychological management

Medical practice had centralised physical interventions with the assumption that if these be successful, psychological well-being would follow. The consensus statement formally acknowledged the need for integral psychological care provided by experienced practitioners within the MDT.

Adolescents with DSD can be especially vulnerable to the emotional impact of body differences (Kleinmeier et al., 2010). Delayed or absent puberty may render some individuals socially and emotionally vulnerable (La Rosa, Traggiai, & Stanhope, 2004). Issues that were of lesser importance before puberty, such as appearance of the genitals, sexuality, pair-bonding and infertility take on a new level of importance in life (Cohen-Kettenis & Pfaeffin, 2003). Security in gender identity, sexual adequacy and sexual preferences can become a source of preoccupation (Cohen-Kettenis, 2010). Individuals who have had a history of repeat intimate medical examinations, medical photography and exposure or who now require hormonal induction of puberty, may require consistent and high-quality psychological follow-up (Patton & Viner, 2007).

Has the practice of early childhood surgery changed?

There has been a trend towards a more child- and family-orientated approach in the past few years. Many surgeons have learned that ‘one size does not fit all’, and that each child and their family must be considered case-by-case. The MDT creates an environment for both the family and the professionals to draw on a collective expertise and to formulate the best treatment plan for the child. But, have these changes resulted in less infant genital surgery?

Despite many references to the consensus statement in the clinical literature, which may be mistaken for a tacit acceptance of its recommendations, there has been no audit of its implementation. Collection of even the most basic surgical data is poor and very variable. The UK National Health Services Hospital Episode Statistics in fact shows an increase in the number of operations on the clitoris in under-14s since 2006 (http//:www.hesonline.nhs.uk). It is not clear if this is secondary to an increase in the detection or incidence of DSD. It is also difficult to gauge the opinions and attitudes of surgeons. Some are dismissive of studies demonstrating poor long-term outcomes viewing the authors as ‘anti-surgery’ and the data based on inferior surgical technique (Karkazis, 2008, pp. 166–167). For those who profess a more conservative approach, it is not clear to what extent this translates into practice. Rather depressingly for those who advocate a more prudent orientation, recent publications in the medical literature tend to focus on surgical techniques with no reports on patient experiences.

Whilst the long-term outcomes of childhood genital surgery appear unsatisfactory, the absence of robust evidence to support non-intervention leaves clinicians and parents

uncertain about abandoning surgery, even though some institutions worldwide appear to have adopted this approach. The consensus statement suggests that there is inadequate evidence to abandon the practice of early separation of the urethra and vagina, i.e. vaginal reconstruction. In addition many parents of children with DSD continue to express deep concerns about the appearance of the genitalia and these concerns require appropriate professional responses.

Families who decide to bring up a child with atypical genitalia may require a significant amount of support from psychologists and from support groups. In most countries, there is no drive to develop funding models for these interventions, and no drive to resource user forums to provide consistent support to parents. There is currently no evidence to suggest that families are offered consistent expert social and psychological support should they decide to postpone surgery for their child.

Clitoral surgery

The size of the clitoris can vary significantly amongst women without a DSD and there is no defined normal range for children. The consensus statement recommends no surgery for girls with minor and moderate degrees of clitoral enlargement until they can decide for themselves. In girls with severe clitoral enlargement, clitoral reduction surgery may still be performed provided the family fully appreciate the potential risks and benefits. It is imperative that the family hear the arguments for and against clitoral surgery from both a paediatric urologist/surgeon and also from an adolescent gynaecologist whose experience with adolescents and adults are paramount. Parents should be given information about user groups to access additional perspectives.

Genital surgery for girls with DSD can involve surgery to the clitoris alone (clitoroplasty, clitoral reduction, clitorectomy), the vagina alone (vaginoplasty, total urogenital mobilisation) or both (feminising genitoplasty). A clitoroplasty and clitoral reduction involves removal of part of the erectile tissue of the clitoris with preservation of the glans and the nerve/blood supply to the clitoris. This technique is perhaps most favoured nowadays for clitoral surgery. Clitorectomy should no longer be practiced. Proponents of nerve sparing techniques for clitoroplasty, whereby the body of the clitoris is not completely removed, claim they achieve the desired outcomes of a feminising genitoplasty whilst avoiding the irreversibility of clitoral reduction (Pippi-Salle, Braga, Macedo, Rosito, & Bagli, 2007). Although an appealing proposition, the long-term effects on adults remain undetermined. Some surgeons also like to create labia minora from the elongated skin on the clitoris and this is termed ‘labioplasty’. A labioplasty is not routine practice in all centres as it can lead to unbalanced or unacceptable labial appearance as well as exposure of the glans of the clitoris, which some women find disturbing.

From their own practice, the authors observe an absence of linear relationships between clitoral size and parental anxiety. Some families cope well with more severe degrees of clitoral enlargement and are keen to postpone surgery. Other families are very distressed by what would appear to clinicians as very minor degrees of clitoral enlargement. It is important however to take their concerns seriously and guide them towards the best decision for their child.

Vaginal surgery

Anatomically, a wide spectrum of atypical presentations may be identified in girls with DSD. These include a flap of skin covering the vaginal opening, a fusion of the urethra and

the vagina into a single channel (urogenital sinus) or complete vaginal absence. Adolescent and adults may need a vaginal opening to allow for menstruation and penetrative vaginal intercourse.

Menstrual obstruction, where menstrual blood does not flow out but accumulates in the body, can occur but is rare. It has been suggested that it can in fact be beneficial to vaginal surgery as the distension of the vagina can provide more tissue for the vaginal surgery, although there is no evidence to support this. It is possible that menstrual obstruction could lead to endometriosis, whereby the lining of the womb (endometrium) grows outside and around the womb, causing pain, usually during periods. This should be closely monitored by the medical team.

Girls and women have different sexual preferences, but most would desire to have the choice of engaging in vaginal sex. For most girls and women, vaginal intervention is likely to be sought at some point. Based on our combined experiences, we suggest that doctors and patients consider the following factors when planning the timing of vaginal surgery:

Reasons for deferring treatment until adolescence

In some girls no surgery is needed and all that may be required is a short program of vaginal dilatation to stretch the vagina. This can be undertaken in adolescence or early adult life. Following an examination by and the recommendation of a gynaecologist, the individual can generally self-manage the regime with nursing input and sometimes psychological support. Girls with a small flap of skin covering the vaginal opening may require an introitoplasty and again this is best performed in adolescence to allow post-operative dilation. Vaginal replacement surgery, using either intestine or skin grafts should not be performed in infancy or childhood. The long-term outcomes are on the whole very unsatisfactory and for this very reason, this kind of surgery should only be carried out on consenting adults.

Reasons given for treatment in infancy

As opponents of infant surgery have pointed out, the vagina does not have a function before puberty and there is no evidence that creating a vaginal opening reinforces gender identity. It would seem appropriate therefore to defer any vaginal surgery until adolescence. However, some clinicians feel strongly that there are technical reasons to support the case for early vaginal surgery in some situations.

Girls with a urogenital sinus abnormality, where the vagina and urethra are joined together, require a major operation. The timing of the operation may depend on the level at which the vagina and urethra join to form a ‘common channel’. In children with a short common channel, where the vagina and urethra join up for a relatively short distance, surgery can be deferred until adolescence/adulthood. This approach has the potential advantage that pubertal oestrogens stimulate the vaginal tissue in ways that render surgery somewhat easier and potentially more successful. It also means that the individual can dilate the newly created vaginal opening herself should this be required. However surgery in girls with a long common channel is highly complex and some surgeons recommend that this should be undertaken at around 6–12 months of age. The reason given is that surgery is technically easier than in adolescence/adulthood, with faster healing, less post-operative pain and little memory of having had the surgery. The majority of these girls are likely to need to perform vaginal dilatation in adolescence for penetrative intercourse and a smaller number may require a further surgical procedure in adulthood (introitoplasty).

More modern techniques for this kind of surgery may offer better outcomes than those published so far, but these cannot be realised for many years to come. As yet there are no data to determine whether early or deferred vaginoplasty is technically easier or has better outcomes. Furthermore, just because a procedure is easier to perform in infancy does not necessarily mean the adult outcomes are better.

At present, parents have the right to decide if their child should or should not have genital surgery in infancy or childhood. It is important to adopt a respectful and non-blaming stance when considering parental decision to take care of their child as best they can, in uniquely difficult circumstances. Some surgeons are realising that one approach does not fit all. Opponents to surgery may equally consider the possibility that one criticism does not fit all. If we were to successfully lobby for and contribute to greater resources to educate and support parents, surgery may be better targeted for families for whom it is indicated.

Conclusion

The recommendations of the Chicago consensus statement on genital surgery were made at a time of increasing controversy about the need for infant and childhood genital surgery. As surgeons in paediatric, adolescent and adult medicine, we have experienced a sea change in the way we work. Having seen some poor outcomes of some childhood interventions, informing parents as fully as possible is a central focus for our consultation. Even with parental or adult consent, we would not consider requests for elective genital surgery without discussion and debate within a multidisciplinary team. On the whole there is much more discussion about any intervention.

On the other hand, we have also witnessed unabated enthusiasm for and confidence in childhood surgery. Whilst some units have embraced multidisciplinary working, other units have demonstrated little commitment to change. As yet the international medical community does not wholeheartedly conform to the care standards outlined in the consensus statement. Individuals and families may stumble upon a mature and progressive team of multidisciplinary experts, or an individual surgeon who defend their favourite interventions regardless.

A major weakness of the Chicago consensus statement then, is an absence of recommendations for implementation research. Whilst it cannot compel the global medical community to adopt its recommendations, it could have outlined performance indicators for DSD services with a high degree of specificity. The consensus statement encourages prospective multi-centre research focusing on multiple parameters; this would only be possible if care standards were comparable across sites. It has, perhaps not unreasonably, been left to the personal and professional ethics of individual surgeons to do what they believe to be best.

To answer the question that is the title of this article from a purely surgical perspective, there is presently no definitive evidence that the consensus statement has had a significant impact on the number of children undergoing genital surgery. There is also, as yet, no new evidence for significant improvements in long-term post-operative outcomes.

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