

Surgery in infants and children with DSD

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There is no subject that creates more controversy and debate than that relating to ‘feminising’ genital surgery in infants and children with DSD. From the outset we need to be clear that the views expressed herein represent what would be considered to be best practice by a multidisciplinary team (MDT) dedicated to children with DSD. The views are not those of an individual surgeon but of the MDT as a whole and have been reached through the process of debate, critical appraisal and open-mindedness.

There is no doubt that there is no place for dogmatic views in DSD and one has to take into consideration what is best for the child, the family and listen to the views of other professionals before undertaking what is on the whole irreversible surgery.

Of paramount importance is to look at the long-term outcomes of early genital surgery, which on the whole are not very satisfactory. Research has shown that the final cosmetic appearances in adulthood may not be entirely satisfactory and there is a high reoperation rate in adulthood. The formation of some degree of scar tissue is a necessary part of any surgery, but this can be particularly relevant when one is dealing with the vaginal opening which has a stronger tendency to close down particularly when the surgery is performed in childhood. Unfortunately, abandoning genital surgery in childhood altogether is not an entirely satisfactory answer either, although many institutions worldwide have adopted this approach.

Many parents of children with DSD continue to express deep concerns about the appearance of the genitalia and these concerns need to be taken seriously and managed in an appropriate manner. Surgery to alter the genital appearances, to make them appear more ‘normal’ should not be considered to be the logical next step. We need to explore other ways to support these families in bringing up a child with DSD, with the help of psychologists, family therapists and the support groups. In the meantime, however, some individuals may find surgery the best option.

Another controversial area that needs discussion is the belief that genital surgery sometimes needs to be performed as a medical emergency. In over 10 years, we have never witnessed such a case. It is true that there are instances when the investigation of a child with a DSD is urgent, by means of laparoscopy for instance. In such cases, these investigations are necessary to help with determining the most appropriate sex of rearing and are therefore justified. It would be most unusual, however, to need to perform genital surgery to make the appearances congruent with the sex of rearing on an emergency basis.

Over the last 5 years, we have witnessed a changing trend in the surgical management of the genitalia in DSD. We are witnessing that more families are comfortable to defer decisions about surgery until adolescence or early adulthood, thereby allowing the individual to be involved in the decision-making process. In addition delaying surgery until after puberty may give a better outcome as the skin may be more supple and the individual can perform post-operative vaginal dilatation if this is required. This change has come about through healthy debate, adopting a more child and family focused approach and by moving away from generalisations. We have recognised that each

child and the associated family need to be treated on an individual case to case basis, promoting the philosophy of 'one size does not fit all'. What is the right approach for one child/ family may not necessarily be correct for another. This is where the benefits of the multidisciplinary approach to the child with DSD are most evident. It allows both the family and the professionals to draw upon the collective expertise of the MDT to formulate the best treatment plan for that individual child.

Genital surgery for girls with DSD can involve surgery to the clitoris alone (clitoroplasty, clitoral reduction, clitorrectomy), 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 generally favoured nowadays for clitoral surgery. Clitorrectomy refers to removal of the entire clitoris and should no longer be practiced. 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 unsightly labia as well as exposure of the glans of the clitoris, which some adult women find disturbing

1. Clitoral Surgery

Clitoral surgery is generally considered when the clitoris is larger than 'normal'. We know that the size of the clitoris in adult women can vary significantly and that there is no defined normal range for children. We have found that there is no direct correlation between clitoral size and the extent of family anxiety. Some families cope very well with quite severe degrees of clitoral enlargement and are keen to postpone surgery until the child has reached an age when that individual can make the decision for/against surgery herself. Other families are very distressed by what we would consider to be very minor degrees of clitoral enlargement. It is our duty, however, to take their concerns very seriously and guide them towards the right decision for their child.

Our current recommendation is that girls with minor and moderate degrees of clitoral enlargement should not have surgery in childhood. In girls with severe clitoral enlargement we remain happy to undertake clitoral reduction surgery, provided the family are fully informed and cognisant of 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 from an adolescent gynaecologist, whom will often have a much greater experience of the adolescent/ adult perspective of this kind of surgery.

2. Vaginal Surgery

There can be a wide spectrum of anomalies of the vagina in girls with DSD that require surgery. These can range from a small flap of skin covering the vaginal opening, a fusion of the urethra and the vagina into a single channel (urogenital sinus), to complete absence of the vagina.

Typically, girls need a patent vaginal opening to allow firstly for unobstructed menstruation, where this is possible, and secondly for penetrative intercourse. The former occurs at around 10 – 12 years of age and the latter usually in adulthood. Some would argue therefore that there is no need for vaginal surgery in childhood, and this is a very reasonable view to take. Obstructive menstruation can occur but is very rare. In some cases, it can in fact be beneficial to vaginal surgery as the distension and stretching of the vagina can provide more tissue for the vaginal surgery. It should, however, be closely monitored by the medical team.

If we take the view that most girls will want a patent vagina for sexual intercourse then some form of vaginal intervention will be required at some point in time:

- In some girls no surgery is needed and all that may be required is a short program of vaginal dilatation to stretch the vaginal opening. This would normally not be undertaken until adolescence or early adult life and the individual concerned would perform this herself.
- Girls with a small flap of skin covering the vaginal opening will require a small procedure to cut this skin and again this is best performed in adolescence.
- Girls with a urogenital sinus anomaly, where the vagina and urethra are joined together, require a more major operation. The determining factor in when that operation should be performed is the level at which the vagina and urethra join to form a 'common channel'. This is best determined by a simple daycase procedure called a 'cystovaginoscopy'.

In children with a *short common channel*, where the vagina and urethra join up for a relatively short distance, we recommend that surgery can be deferred until adolescence/adulthood. This approach has the advantage that the puberty softens the tissues of the vagina, making the 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.

Girls with a *long common channel*, where the vagina and urethra join up for a considerably longer distance are a major surgical challenge. Our current recommendation is that in this group of girls corrective surgery should be undertaken at around 6-12 months of age. The rationale behind this approach is that there is the impression that the surgery itself appears to be technically easier than in adolescence/ adulthood, with faster healing, less postoperative 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. More modern techniques for this kind of surgery may offer better outcomes than those published so far but these will not be realised for many years to come.

- 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 performed on the consenting adult.

Until such time as there is a change in the law, parents will continue to have the right to decide if their child should or should not have genital surgery in infancy or childhood. It is important that we maintain the flexibility for parents to be able to make decisions on behalf of their child, but seek to educate parents and society about the many variations that exist in the appearance of genitals. These very difficult decisions, which they have to make, should be supported with all the available resources, which are best delivered through a dedicated MDT.