The question-Who is the right person to take decision regarding choice & time of surgery- Parents or Patients?- continues to stare us,\textsuperscript{1} the birth prevalence of genital anomalies remains as high as one in 300 births.\textsuperscript{2} However, the extent of genital ambiguity may depend on the expertise of the observer, and prior to presentation to a clinical expert, the label of ambiguous genitalia is often assigned to newborns where the most appropriate sex of rearing is not immediately clear to those present at the child's birth.

What defines DSD and how it is managed and treated is a political and controversial area with individuals, families, clinical practitioners, mental health professionals, ethicists, lawyers, advocates and activists often having differing views.

The sex of a newborn baby is usually the first question asked by & from the parents. There is enormous pressure for people to conform to cultural and social expectations.

In the past, most medical experts believed the birth of an infant born with an DSD condition is a medical and social emergency usually requiring surgical intervention and that living with ambiguous genitalia is psychologically traumatic for the child. Early normalization was, therefore, sought so that children could be given a successful gender identity and bonding with parents would become easy.

More recently, the medical sense of urgency to correct the situation has decreased. Babies born with DSD conditions are regarded as having a significant medical condition, but more emphasis is placed on getting an accurate diagnosis of the cause of the ambiguous genitalia and making a careful decision about assigning gender before making any decisions about surgery.

**Age & Timing for Surgery**

*Three age periods can be distinguished in the problem of sex assignment:*
1. The newborn period, extended to early infancy, when parents decide
2. From 2 to 10 years of age, when sex re-assignment is generally not recommended
3. From 11 years of age up to late adolescence or young adulthood, when patients decide. When sex assignment becomes necessary, particularly in newborns, the aim is to choose that sex that will allow the best future functional adaptation that better goes along with the biological sex. Decisions might be difficult and controversial because in some cases there is no "good" solution.

*In general recommendations in infants are based on:*
1. Etiologic diagnosis (if available, molecular diagnosis), diagnosis of type of DSD is useful because disorder evolution partially depends on etiology, known from both personal experiences and scientific publications
2. Development of external genitalia and potential future sex function
3. Possibility of surgical correction
4. Development of internal genitalia, and fertility potential
5. Parental acceptance
6. Psychological evaluation of parents and family, including social environment

Understanding the Social and Cultural Background of the Family is Crucial for a Fruitful Relationship Between Parents and Doctors

However, it is sometimes difficult to synchronize delivery of information to parents by different members of the team. Special efforts should be made by the informing professional to ensure appropriate understanding by the family of the situation, including chromosomal constitution, gene function, heredity, gonadal development, as well as external and internal genitalia differentiation. During follow up patients will go through all stages of growth and development and information has to be delivered according to patient's mental development and improved understanding. The medical team should constantly update itself with new developments that might arise along many years, and to give answers to questions, which are frequently, influenced by a high emotional impact.

Several pioneering research teams have studied the wide variety of sexual development disorders with a view to changing the way they are treated.

One example is Congenital Adrenal Hyperplasia (CAH). In this condition, due to enzymatic deficiency, there is a surfeit of the precursors of testosterone. As a result there are distinct changes in the female genitalia. Most notable is the enlargement of clitoris which resembles the penis. The most frequently done surgery is to reduce the size of the clitoris. However, some researchers found, that, these patients may have reduced sensuality during sex.\[3,4,5\] It led to a growing trend towards a more conservative approach with increasing emphasis being placed on the preservation of sexual function later in life. 'Endocrine Society' guidelines of 2010 state "Surgical guidelines emphasize early single-stage genital repair for severely virilized girls, performed by experienced surgeons.\[6\]

The international Consensus statement on management of DSD cites gender dissatisfaction occurring more frequently with individuals with a DSD condition. However Crawford et al's long term outcome study, found that early intervention yielded acceptable cosmetic results while producing minimal impairment in quality of life.\[7\]

Type of Surgery

Among the most contentious issue in regard to those with CAH involves XX females. Females with CAH are generally born with an enlarged clitoris and often a vagina that is atypically formed. Questions arise as to whether they should have genital surgery or not-it is often recommended. Sometimes, dependent upon the degree of masculinization of the genitals, there is question of whether they should be reared as males or females.

Not infrequently, for various reasons such as the disease's late onset, some will be reared as males and some will request such assignment. Recommendations as to the management of severely masculinized infants with CAH are controversial. When it was first suggested that severely masculinized females be raised as males the idea was generally rejected. Lately, however, the idea is being revived.\[8,9\]

Regarding the management of infants with DSD conditions and ambiguous genitalia in a survey\[10,11\] the Paediatric Urologists overwhelmingly favored female gender assignment for females with CAH even if they were extensively masculinized. They recommended feminizing surgery-reducing the size of an enlarged clitoris-and considered that preservation of female fertility was of foremost importance and the masculinization of behaviors or inclinations, was of lesser importance. There was a great difference of opinion as to the age it would be best to do the surgery.

- There are case reports where patients have reported dissatisfaction with early genital surgery. However, the magnitude & extent of this satisfaction needs further verification\[12\]
- On reaching adulthood some individuals see themselves as distinctly non-male/ non-male.\[13\]
- This has given an advocacy for another category to be recognized as the 'third gender'. some authors advocate the consideration of a third gender or of even more gender categories.\[14,15\]
Australia has become the first industrialized country known to have removed the legal barrier to such gender assignment by allowing an X, signifying unspecified sex or intersex, in the sex field of passports with the State of Victoria has issued a corresponding birth certificate that lists sex as "indeterminate-also known as intersex."[16]

**Ethical Considerations**

Assigning sex to children born with DSD condition raises a number of important ethical issues,[17] such as:

- Raises questions about the authority of parents and others to make irretrievable decisions for young children
- Raises questions about what constitutes disease or malfunction
- Poses questions about treatment decisions versus evidence of outcome
- Highlights the need for evidence in the form of systematic long term outcome studies

The practical guiding ethical principles are as follows:[18]

- Minimise physical risk to child
- Minimise psycho-social risk to child
- Preserve potential for fertility
- Preserve or increase capacity to have satisfying sexual relations
- Leave options open for the future to provide for the future autonomy and self determination of the young person
- Respecting the wishes and beliefs of the parents because they are guardians of the child who have primary responsibility for protecting their child's best interests and will raise and care for the child

Early genital surgery for psychosocial reasons, e.g., feminizing surgery for gender confirmation or vaginoplasty to ease later penovaginal intercourse has also led to controversies involving side effects of surgery, erotic sensitivity or orgasmic capacity.[19]

Therefore, there is an urgent need for a surgical policy based upon clinical data.[20] *(To be continued)*

**References**


18. Gillam, L; Ethical principles to guide discussion about the issues related to the management of disorders of sex development, submitted by Professor Garry Warne, 25 March 2009.
