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EDITORIAL

Differences (Disorders) of Sex Development (DSD): The Quintessence of Perennial Controversies-IV: Genitoplasty for CAH- Management, Socio-cultural and Legal Issues

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In children with differences in sex development (DSD), by and large, it is the parents who make decisions regarding the surgical options. Socio-cultural influences play a significant role in gender assignment. While gender assignment has traditionally been influenced by sex of rearing, the treatment was dependent upon the size of the phallus [1]. The latter still holds, including the developed countries [2]. It is now being increasingly realized that gender identity is a result of complex interaction between genes and environment and it is impossible to predict what gender any child will come to identify with.

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The general trend in belief remains that surgery benefits the patients physically and psychosocially. However, sometimes complications linked to surgical intervention have led to the emergence of a platform for intersex advocacy groups.

Thus, the following 4 groups of caregivers justify the part played by them in the child's welfare at its center. However, a lack of sufficient evidence relegates them to a surrogate role rather than an exemplar:

A. Role of parents

a) Parents have the strong wish to surgically "normalize" their child's sexual anatomy. They view genital surgery as "obvious" and "necessary" to assure their child's positive psychosocial and psychosexual adaptation. The follow-up studies suggest predominantly favorable attitudes toward early feminizing procedures.

b) Parents and the role of Health Care Professionals (HCP): In the initial gender assignment, the HCP interprets the test results, anatomy & physiology (e.g., hormone production, hormone receptors, gross anatomy) & informs the parents. Individual cases are referred to by the institute's ethics committee (IEC)—clinicians, ethicists, members of the law, as well as the community. When IEC finds it difficult to negotiate, the matter is referred to a family court to decide upon the best interest of the child.

B. Role of surgeons

Surgery benefits the patients physically and psychosocially. Atypically developed genitalia affect:

- Physical appearance
- Body image
- Function of the urinary tract
- Gonads &
- Psychological and psychosexual development

Hence, the suggestion that therapeutic management of these patients is limited to 'cosmetic' surgery is not universally true.

Surgery in CAH involves clitoroplasty. This could be in the form of urgent surgery in order to create unobstructed outlets for urine.

- While early clitoral reduction surgery has been categorized as cosmetic and may carry the risk to genital tissue sensitivity, there is less disagreement for clitoroplasty for severe clitoromegaly. The consensus statement and clinical practice guidelines

C. Role of DSD advocacy groups

Their concerns stem from the following beliefs:

a) Parents seek to act before becoming fully informed about all options.
b) Legal and ethical questions are still unclear, and patient's autonomy is not taken into consideration. Therefore, the decision for any intervention should be deferred.
c) Comparative outcomes associated with performing surgery later in life should be known first.

D. Legal issues

These arise primarily around consent for medical interventions, such as genital surgeries, which raise ethical questions about parental authority and the rights of the individuals affected, particularly when considering their capacity to consent as they grow older. This area involves complex intersections of medical ethics and legal standards that often result in intense debates. To ensure that the consent is truly informed, it has to be "qualified and persistent:"

- The consent must be in writing.
- The information provided must be complete.
- The parents must be informed about the dangers of current treatments,
- The possibility of delaying surgeries and giving adequate psychological support to the child.
- The authorization must be given on several occasions over a reasonable time period [3].

The decision to perform surgery early or late would depend upon evidence—Whether later surgery has: Better, Poorer, or Comparable—Physical, Psychosocial, and Psychosexual Outcomes.

It is equally questionable if a general moratorium on all surgeries is justified. It is a common belief that nothing is 100% in medicine, and decisions on individual cases should be taken up on their merit.

In this context, the study by Dsd-LIFE is a laudable effort [4]. This is a multicentre cross-sectional, Clinical evaluation study with:

16 partners and 14 recruiting centres in Germany, France, the Netherlands, Poland, Sweden, and the United Kingdom, of whom 14 were active recruiting sites.

The study was carried out in adolescents (≥ 16 years) and adults with DSD (conforming to the Chicago Consensus). Interview, retrospective chart, and medical examination were carried out, and the following patient-reported outcome questionnaire was filled out and results published:

Clitoroplasty—Effect of timing of Sx on Outcomes (n. 415)

Questionnaire:

Q: General Postponement of Surgery until legal age

i. 51.2% disagreed
ii. 27% agreed
iii. 22% do not know

Q. Appropriate Time for Genital Surgery

i. 46%: infancy
ii. 20%: 4 years to 12 years

Q. Clitoral Reduction is necessary in girls (n. 314)

i. 38.2% agreed
ii. 14% disagreed
iii. 18.8% undecided
iv. 29% don't know

Q. Vaginoplasty: n. 415 Adolescence/Adulthood or Infancy (n. 323)

66% approved of surgery in infancy or childhood

These data suggest that:

- CAH persons predominantly favor interventions in childhood.
- A moratorium —one-for-all solution is not justified
- Case-by-case decision making is better suited [5].
- Efforts in improving information on long-term outcomes, informed consent, and assent.
- Contact between support groups should be strengthened.

If considerable uncertainty exists, parents should be motivated to postpone elective genital surgery.

These data are also in consonance with the ESPU & SPU-stand point (2014), viz., the medical and surgical management aims at:

i. Avoiding potential health hazards—anatomy and function of the urogenital tracts
ii. Meeting parents' expectations
iii. Helping an individual's future satisfactory sexual function.
iv. Consistent with their gender identity

Recent years have seen a shift from calling for shared decision making (SDM) between parents and the young child's healthcare providers, e.g., [6] to appeals for protecting the child's right to bodily autonomy and for the "right to an open future" (interpreted as a deferral of decisions regarding elective gonadal or genital surgery "until the patient himself/herself can participate meaningfully in decision making") [7-10].

Shared decision making: It comprises 3 essential elements:

- Explicit acknowledgment that a decision is required
- Evidence concerning the risks and benefits of each option
- Process takes into account the patient's/family's values and preferences.

Shared decision making: Six steps KON & KARZAKIS, Journal of Endocrinology & Metabolism

- Inclusion of sub-specialists
- Involve parents in decision making
- Parents need help addressing the emotional feelings
- Avoidance of terms referring to genitals
- Providers should strive for objectivity-evidence-based
- Parents should have received unbiased information… hopefully leading to a consensus based on trust and understanding.

Conflicts of interest

The authors declare that they do not have conflict of interest.

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