Hans-Werner Gessmann

Hermaphroditism

Hermaphroditism, also known as intersex, is a condition in which there is a discrepancy between the sexual and genital organs that are external and internal. It is grouped as a disorder of sex development along with other conditions (DSD). Even with the introduction of modern diagnostic methods, the cause of hermaphroditism is not able to be determined in many children. This is referred to as complex or idiopathic hermaphroditism. [1]

A hermaphrodite is an organism in reproductive biology that has complete or partial reproductive organs and generates gametes normally associated with both male and female sexes. Many animal taxonomic groups (mostly invertebrates) do not have separate sexes. In these groups, hermaphroditism is a normal condition, enabling a form of sexual reproduction in which either partner can act as the "female" or "male". [2]

There are four different types of hermaphroditism, as follows:

1. 46, XX hermaphroditism - An individual with 46, XX hermaphroditism has two XX chromosomes and the ovaries of a woman, but has external genitalia that appear to be male.

2. 46, XY hermaphroditism - An individual with 46, XY hermaphroditism has one X and one Y chromosome, as is usually seen in males, but the external genitalia are either not completely formed, or resemble those of females. The internal sexual organs may be normal, incomplete or absent, depending on the specific case.

3. True gonadal hermaphroditism - An individual with true gonadal hermaphroditism has both ovarian and testicular tissue, either in the same gonad (referred to as an ovotestis) or in one ovary and one testis. Some affected individuals have XX chromosomes, others have XY chromosomes, and others have a combination of both. Likewise, the external genitalia can vary in form, from male, or female, to ambiguous.

4. Complex hermaphroditism - Complex hermaphroditism involves other disorders of sexual development beyond simple 46, XX and 46, XY. These may include:

    - 45, XO
    - 47, XXY
    - 47, XXX

This type is not usually associated with a discrepancy between the internal and external genitalia. Instead, the individual shows abnormal levels of sex hormones and incomplete sexual development. [1]

Historically, in individuals of gonochoristic species particularly humans, the term hermaphrodite was also used to describe ambiguous genitals and gonadal mosaicism. As the term hermaphrodite is considered to be misleading and stigmatizing, as well as' scientifically specious and clinically problematic,' the word intersex has come into use for humans. [2]

The signs and symptoms of hermaphroditism depend on the person's condition. This may include:

- Labial fusion
- Ambiguous genitalia
- Micropenis
- Hypospadias
- Clitoromegaly
- Electrolyte abnormalities
- Undescended testes
- Delayed, absent or abnormal pubertal changes [1]

Intersex treatment in humans is dependent on the age at which the diagnosis is made. Historically, when diagnosed at birth, the choice of sex was made (typically by parents) on the basis of the condition of the external genitalia (i.e. the predominance of the sex organs), after which so-called intersex surgery was performed to remove the opposite sex gonads. To resemble those of the chosen sex, the remaining genitals were then reconstructed. Female genitalia reconstruction was done more readily than male genitalia reconstruction, so ambiguous individuals were often made to be female. However for affected individuals, intersex surgery has long-term consequences. For instance, later in life, the individual may not be satisfied with the results of surgery and may not identify with the gender assigned. [3]

In order to address the various needs presented, a child with hermaphroditism will usually require care from a multidisciplinary healthcare team.

There is also significant controversy and stigma associated with hermaphroditism treatment. More recently, however the complexity of gender and sexuality has been recognised. Consequently, the treatment of patients is becoming more individualized and less standardised. [1]

It has accumulated abundant evidence that the gender role and erotic orientation of an individual are established through the cumulative experiences of years of living as a boy or a girl. The child who has been steadfastly accepted as a girl or as a boy since early childhood, regardless of chromosomes, gonads or hormones, especially if the external genitals have been altered to

conform to this sex, will not question his or her own gender and will conform to the habits and behavior of the rearing sex. The child will be confused and perplexed when there is prolonged doubt and uncertainty on the part of the parents or when a change of sex is enforced after an early age and before late adolescence and psychological problems result. Consequently, in early childhood, every effort should be made to decide the sex of rearing and support, guidance and reassurance should be provided to the parents. Necessary corrective operations should be carried out as early as possible in life.

Instead of the form of gonads or the sex chromosomal pattern, it is advisable to select the sex of rearing according to the anatomical structure of the external genitalia. It is unwise and condemns the patient to a life of misery to attempt to make a boy of a person who does not have a fairly well-developed phallus. In puberty, male pseudohermaphrodites who have external female genitalia invariably feminize, so that orchidectomy is not required to prevent masculinization. Avoiding the possible risk of testicular malignancy could be the only indication. At puberty, male pseudohermaphrodites whose genitals resemble or are vague may either masculinize or feminize. If, because of the small size of the phallus, it is decided to raise such a child as a female, orchidectomy may be performed in infancy to avoid the risk of masculinization, or it may be delayed until masculinization begins. The former course often seems preferable. At puberty estrogen should be given in doses adequate to develop female sex characteristics. In these cases gonadectomy cannot be considered a mutilating operation or one which deprives the patient of fertility. On the contrary it is one which enables the patient to continue as a reasonably normal individual in the sex in which he has been reared and prevents the disastrous psychologic upheaval of a sex reversal. [4]

References

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