Inter-Sex

Newer Concepts, Surgical and Nursing Management

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The birth of a child with ambiguous genitalia constitutes a crisis situation, quite different from any other congenital anomalies. Even though inter-sex is not a physical emergency, it can produce serious long-term social tragedy for the affected children and their families. The health professionals, especially Nurses, can do much in encouraging and supporting these children and families.

Disturbances in the normal order of events in sex determination will produce abnormal sexual development, with the presence of ambiguous genitalia at birth. In some instances, however, the external sexual structures represent those of a perfectly normal male or female, whereas the genetic sex is the direct opposite. A situation in which the phenotypic sex differs from the chromosomal sex is termed as inter-sex.

From conception through the first 6 weeks of intrauterine life, the embryo is morphologically neither male nor female. For normal sexual differentiation to occur, a specific sequence of events must take place. The bipotential gonad requires at least two 'x' chromosomes to develop into an ovary. If any portion of an 'x' chromosome is deleted, streak gonads result. In the presence of the testicular determining factor (TDF) gene located on the 'y' chromosome, a testis develops. Between the 7th and 14th weeks of gestation, testosterone produced by the foetal Leydig cells transforms Wolffian ducts into the male genital tract.

Growth of the penis occurs mainly in the late second and third trimesters, requiring stimulation of the testis by foetal pituitary gonadotropins. Mullarian duct inhibing factor, a glycoprotein produced by the sertoli cells, causes regression of the internal female duct structures during the first 8 weeks of gestation. In the absence of this factor, the uterus and fallopian tubes develop and mature. In general abnormalities in testosterone synthesis or action cause male pseudohermaphroditism, frequently with ambiguous genitalia and presence of excessive androgens in a female foetus cause female pseudohermaphroditism.

The four conditions producing ambiguous genitalia in the newborn that require prompt and accurate evaluation are: (1) Female pseudohermaphroditism (the musculinized female), (2) Male pseudohermophroditism (the incompletely musculinized male), (3) True hermaphroditism and (4) Mixed gonadial dysgenesis.

Diagnostic Evaluation

- History
- Physical examination
- Buccal smear – detects presence or absence of sex chromatin.
- Chromosomal studies – detects chromosomal abnormalities and precise genetic sex.
- Endoscopy and radiographic contrast studies – reveal presence, absence or nature of internal genital structures.

Management

The assignment of gender sex to the infant whose sex is doubtful constitutes a social emergency. Thorough studies should be carried out early to assist in gender selection before a final sex is assigned. Accordingly appropriate, surgical reconstruction techniques that provide normal appearing external structures are carried out. Removal of inappropriate internal structure and dysgenic gonads is recommended.

Nursing Management

The management of a child with inter-sex is a team approach, consisting of Paediatric Surgeon, Plastic Surgeon, Endocrinologist, Urologist, Geneticist, Clinical Psychologist, Paediatric Nurse and Social Worker. The main aim of Nursing Management is the acceptance of the affected child by the parents and the society.

If a baby with defective genitalia is born, the news should be communicated to the parents at the earliest in a compassionate manner. The Nurse should encourage parent-infant bonding as in the case of any other normal child. Parents should be allowed time to grieve and reorient them to the reality gradually. The family support system is to be encouraged to reassure the parents. Encourage parents to provide a unisex name to the child and instruct them to call the child as 'baby' or 'infant' instead of 'he' or 'she'. Since sexual identity in children occurs around 1.5 years, it is said that the surgical correction should ideally be carried out before the age because any reassignment of sex after that age may cause serious psychological trauma to the child.

The parents' initial impression of sex may be different from the assigned sex of the child. Nurses have to explain about the defect, surgical correction, fertility after treatment, etc. If the parents are not accepting assigned sex, they should be referred to a clinical

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psychologist and if even after that the parents are not willing for correction it means their feelings should be respected.

Genetic Counselling is essential for parents who have had an infant with ambiguous genitalia, is also needed for affected children if they are capable of becoming potential parents. The Counselling team should consist of the Genecticist, Paediatric Surgeon and the Nurse expert. In counselling, we are helping the family to adjust with the defect and also helping the family to alleviate negative emotions like fear, guilt or anxiety.

Community Health Nurse

The Community Health Nurse has a vital role in identifying the cases and making appropriate referrals. As she becomes a member among them, through frequent visits, the family will be more comfortable in discussing the fears, worries, etc. She can mobilise the community resources to alleviate the social stigma associated with the condition.

Surgical Correction, Preoperative Care : The important points are:

- Admission of child.
- Preparation for laboratory investigation.
- Obtain informed consent.
- Psychological reassurance.
- Preoperative teaching about usual post-operative period, care, hygiene, follow-up, etc.

Post-Operative Care: Post-operative Care of the child who had undergone corrective surgery is similar to that of any child who had undergone abdominal surgery. Since the incision has a close proximity to rectum, care must be taken to avoid soiling of the surgical wound by faecal matter. Usually, the sutures are removed on the 8th post-operative day.

The family may be anxious about the prognosis. So, realistic explanation and support are to be given to the family. Avoid false reassurances.

Long-Term Care: The family and child should be encouraged to come for follow-up visits or a Community Health Nurse should visit them occasionally. Counselling is to be continued and gender identity is to be reinforced. The hospitals can conduct outreach programmes to identify cases, the mass media also helps in health education. Self-help groups are to be encouraged among the victims to support and reassure them. Research activities are to be encouraged among Medical and Nursing professionals to get more details about the problem. The Government can also protect the victims by enforcing some favourable laws.

Agenda Items : TNAI Meetings

Members are requested to send their agenda items separately for the TNAI meetings to be held at Imphal during October 23-29, 1999 latest by August 31, 1999 as follows:

- TNAI Executive Committee Meeting – October 23
- TNAI Council Meeting – October 24-25
- House of Delegates Meeting – October 26-27
- Concurrent Sectional Meetings : Nursing Service Section, Nursing Education Section, Public Health Nursing Section and Nursing Research Section – October 29

Each Agenda item should be either self-contained or accompanied with explanatory notes. Only those items may please be included which require national level discussions/decisions.

Secretary-General, TNAI