THE
Journal of Obstetrics & Gynaecology of India

VOLUME XVII, NO. 6 DECEMBER 1967

# CERTAIN ASPECTS OF INTERSEXUALITY*

by

PREMA M. NAIDU,** M.D., F.R.C.S., F.R.C.O.G., F.A.M.S., F.A.C.S.

Mr. President, may I express my thanks to you and the members of your Society for the unexpected honour of being invited to deliver the eleventh Sir Kedarnath Das Memorial Oration. It is a humbling experience, made more so, because the preceeding orators have been recognised as men and women of superb intellectual and professional attainments. My ability to do justice is doubtful, especially on an occasion which marks the centenary of his birth.

Sir Kedarnath, whom we honour to-day, tirelessly taught and wrote in terms well in advance of his day. Born here in 1867, he qualified in 1892 and obtained his M.D. degree from the Madras University even before the turn of the century.

He was an outstanding teacher and a distinguished professor for over 30 years. And what is more he preferred the term 'Obstetrics' to 'Midwifery' anticipating all its scientific advances of to-day. This was indeed prophetic! His book on the 'Obstetric Forceps' is a monumental work covering his arduous journeys across the seas to meet men of common interest, and to collect intimate knowledge on the evolution of this sacred implement. He addressed the American Gynaecological Society at Washington in 1922 on "Midwifery in India" showing that the impact of his teaching was felt far and wide.

If we are permitted to turn over in our minds the events of his day and accept the paucity of antenatal care, with the hazards in childbirth, which was prevelant in 1930, we would readily express our admiration for Sir Kedarnath and his work in no uncertain terms. Even in England, at about that period, the maternal mortality was 4 or 5 per every 1000 live births, higher than in any other

---

*Sir Kedarnath Das Memorial Oration delivered in July 1967.

**Professor Emeritus Dept. of Obstetrics and Gynaecology Osmania Medical College Hyderabad.

Received for publication on 4-8-67.

country in the western world. Every expectant mother secretly or openly feared that she may not survive her childbirth. The term, perinatal mortality was then unknown. Deaths from eclampsia were as great as ours of to-day. Puerperal sepsis was the greatest single killer from epidemics which raged in the lying-in wards. Malnutrition was rampant and it was just being recognised as a cause of pelvic deformities even in Glasgow. The problem of faulty judgment and technique in the conduct of labour was a serious one. The large maternity hospitals were all too familiar with deaths which followed cases of 'failed forceps.' The past President of the Royal College of Obstetricians and Gynaecologists recalls 36 such cases being admitted in one year. Post-partum haemorrhage was dreaded since there was no organized blood transfusion service. It was in 1929 that the Royal College of Obstetricians and Gynaecologists was founded for the training of specialists and confidential enquiries into the causes of maternal deaths were instituted. And in this self-same year was published this great book on Forceps, by Sir Kedarnath. It would not be impertinent to add here that Domagk was to discover the sulphanidomides in 1937, i.e. one year after the passing away of our distinguished obstetrician.

In the later phases of his life, we see him fostering medical education after having been an inspired teacher all his life, undaunted by his courage and singleness of purpose. These were the days when criticism and factions raged in the west. We can well picture a faculty member waiting, outside the hospital gates in Edinburgh, horsewhip in hand, (as told by Prof. Chassar Moir) for a colleague whose views are at variance with his own!

However, we are aware that Sir Kedarnath's attention was not limited only to the birth of a healthy infant from the undamaged birth canal. Sir Kedarnath must have faced problems of twin pregnancies, the mono- and dizygotic, and mused over the possibilities of the 'female born co-twin with a male' being sterile. In fact this had posed a serious problem to Sir James Y. Simpson, an illustrious Scottish predecessor, till the latter disproved that free-martins are unusual in the human.

We are sure Sir Kedarnath had many and varied interests; not merely in the gynaecoid female enjoying her motherhood, but even in her less happy counterpart, the infertile and the hermaphrodite. He had published a paper on pseudo-hermaphroditism in 1909.

Nevertheless, Sir Kedarnath being a visionary, would have readily accepted the part played today by genetics in the world of health and disease. With increasing control of infant mortality and infectious diseases, inherited abnormalities are assuming a proportionately greater importance in medical practice. Needless for me to add, that genes carried by chromosomes contain the information, encoded in their chemical structure, which is translated into the developmental pattern of the individual in his embryonic, pre-adult and adult life.

Therefore I would like you to bear with me if I confine myself to a dis-

cussion on the inherited aberrations of the sex chromosomes leading to various problems of intersexuality as met with in gynaecological practice. The possibilities of adequate therapy, in such cases, if diagnosed correctly, will be mentioned in passing.

The study of sex determination and sex differentiation has made considerable advance in recent years. Women have 2 X chromosomes while men possess X and Y chromosomes.

Though sex determination takes place at the time of fertilization, the primary germ cells play an important part in the development of the gonads, XY chromosomes promoting the medulla to become the testes, and XX the cortex to become an ovary. Further differentiation depends on the medullary and cortical constituents of the primordia as shown in the charts 1 a and b. (Stewart, 1965).

Moreover, concepts on sex differen-

CHART 1 a

*Normal female sex differentiation*

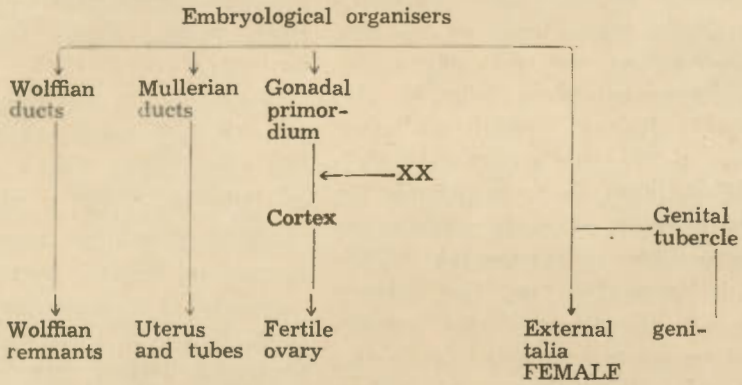
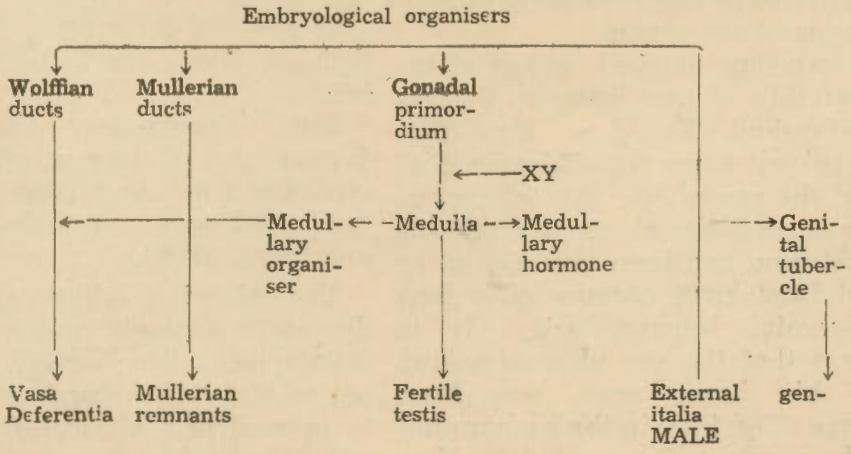



CHART 1 b

*Normal male sex differentiation*



tiation and its relation to the hormonal secretion of the embryonic gonads, have been largely elaborated by Professor Jost of Paris who has been able to prove experimentally, that gonadectomy in early foetal stage leads to an individual with a female phenotype, irrespective of the genetic sex. Jost was also able to show that masculinization of gonaducts is dependant upon the action of hormones of the foetal testis.

Usually sex chromatin studies (which show that Barr bodies are less than the total number of X-chromosomes by one) and chromosome analysis when applied to sex anomalies of man, in certain persons show findings inappropriate to the phenotype. Here I refer to such syndromes as Turner's and Klinefelter's. Mosaicism is a mixture, in the same person, of cells with more than one variety of karyotype. This finding is common in the above syndromes. Further these conditions are suspected to arise from the accident of non-disjunction, either during one of the two meiotic divisions during gametogenesis in one of the parents, or from the early mitotic divisions of the zygote.

Introducing clinical aspect of intersexuality—I now present to you Ramalakshmi (Fig. 1 a. Case 1). This patient came for assignment of sex at the age of 18. My interest in intersex problems stemmed from this patient who had been brought up as a girl from birth because of a deep peno-scrotal hypospadiasis. It is obvious that the sex of rearing had held this young man, now Ram Sharma (Fig. 1 b), in the background till the gender role asserted itself.

Classical mythology provides us with legends and innumerable pieces of sculpture showing the then existing concepts of intersexuality.

Ardanarishwar at Madurai temple (Fig. 2 a) is a classical Indian equivalent of hermaphrodite which represents the concept that man and woman are equally necessary and important. One cannot do without the other while neither sex is superior to the other!

In Bengal, the river Yamuna is often confounded with the ancient mythological she-twin Yami whose he-twin Yama was believed to be the first man, Yami being the first woman.

Even Iran's national epic, Firowzi's Shah Namah, refers possibly to the same King Yama who was severed into two, like a piece of wood.

This idea also resembles the account, in Plato's symposium, on the primordial hermaphrodite who was feared by the gods as being too perfect and hence was cut in half and thrown apart so that they may consume their energy in searching for each other.

In Hebrew mythology we are told that Eve was created from Adam's rib.

Eros, (Synonym: Cupid), the winged god of love is often represented as a hermaphrodite (Fig. 2 b). in Greco-Roman mythology (Jones and Scott, 1958).

The following tables will indicate diagnostic methods, and order of sex development. They would depict experimental and natural intersexuality as well as a clinical classification with illustrative cases.

TABLE I

*Diagnostic Methods*

| | |
|---|---|
| HISTORY | Sex of rearing<br>Gender role |
| EXAMINATION | Genitalia<br>Secondary sex characters |
| LABORATORY TESTS | Gonad histology<br>Hormone excretion |
| GENETIC INVESTIGATIONS | Nuclear sex<br>Chromosomal sex |

*Comment:* Diagnostic methods begin with careful history taking.

TABLE II

*Sex Development*

| | | |
|---|---|---|
| DETERMINATION | Chromosomes | → Nuclear sex |
| DIFFERENTIATION (GONAD) | Gonads | → Adult hormones → Gametes and secondary sex characters |
| | Foetal secretions | → Internal genitalia |
| DIFFERENTIATION (Genital tubercle) | External genitalia | |
| ORIENTATION | Sex or rearing | → Legal sex |
| | Gender role | |

*Comment:* Foetal secretions control development of internal genitalia.

TABLE III

*Experimental and natural intersexuality*

| Cause | Gonads | | Genitalia | |
|---|---|---|---|---|
| | Chromosomes | Histology | Wolffian | External Mullerian |
| Gonadectomy | — | — | — | + |
| Testicular Graft | XX/XY | Variable | + | — |
| Exogenous androgen | XX | Ovary | — | + |
| Free-martin | XX/XY | Variable | + | — |

*Comment:* Exogenous androgens may introduce bisexual characters but chromosomes remain XX.

TABLE IV

*Clinical classification*

| Abnormality of sex development | Clinical group | Clinical criteria |
|---|---|---|
| Sex determination | Chromosomal | Chromosomal sex, Nuclear sex |
| Sex differentiation Gonad | Gonadal | Gonad histology |
| Sex differentiation Genital tubercle | Genital | External genitalia |
| Psycological orientation | Psychological | Sex of rearing Gender role |

*Comments*: Note that this order is in the reverse of the order used in diagnostic methods. The central column shows the broad clinical groups. A few typical examples of the clinical groups will be depicted here.

TABLE V

*Chromosomal intersexuality*

| Clinical syndromes | |
|---|---|
| Medullary Dysgenesis (Klinefelter's syndrome) | (case 2) |
| Cortical Dysgenesis (Turner's syndrome) | (case 3) |
| Metafemales | |

*Comments:* Klinefelter's syndrome (incidence 1:400) Fig. 2 a, b. (Karyotype 47/XXY, 46/XX). Turner's syndrome (Incidence—1:2500) Fig. 3 a, b. (Naidu and Mehdi 1959) (Karyotyping yet to be done).

A specimen of hernia uteri inguinalis (case 4) suspected to be showing characteristics of true hermaphrodite is included (Figs. 4, a, b, c.). Next, in this series, are two sisters (cases 5 a and b) who are examples of pure gonadal dysgenesis with streak gonads (Moszkowski and Taubert 1965). Karyotyping in case 6 shows 2 cell lines, 47/XXY and 46/XX (Figs. 5 a, b, c, d.).

The third case in this group (case 6) is one of testicular dysgenesis. This case was reported in detail, elsewhere (Naidu *et al* 1965) Figs. 6 a, b, c, d, e, f, show follow-up changes and the karyotypes). The above article may be referred to for details of histopathology of the gonadoblastoma found growing in the dysgenetic gonad.

TABLE VI

*Gonadal intersexuality*

AMBISEXUALITY
- True hermaphroditism (case 4)
- Pure gonadal dysgenesis (cases 5 a and b)

DISCORDANT DYSGENESIS (Sex reversal)
- Testicular dysgenesis (case 6)
- Ovarian dysgenesis

PARTIAL MEDULLARY FAILURE
- Anorchia
- Agonadism
- Paradoxical syndrome

TABLE VII

*Genital intersexuality*

(Pseudohermaphroditism

Ho monal
- Congenital adrenal hyperplasia
- latrogenic pseudohermaphroditism
- Testicular feminization.
- Congenital adrenal insufficiency.

Dysontogenetic
- Hypospadias — (case 1)
- Vaginal atresia
- Ano-rectal agenesis
- Sirenomelia

Deformities
- Labial fusion
- Avulsion.

TABLE VIII

*Psychological intersexuality*

Eonism

Symptomatic transvestism
- Homosexuality
- Fetishism

TABLE IX

*Clinical management*

Psychological care
- Explanation
- Reassurance

Surgical treatment
- Correction of malformation
- Prevention of malignancy

Hormone therapy
- Sex hormones
- Corticosteroids

TABLE X

*Surgical Treatment*

Extra-genital malformations
- Congenital heart disease
- Neck webbing

Genital malformations
- Hypospadias
- Vaginal atresia

Dysgenetic gonads
- Excision
- Orchidopexy

TABLE XI

*Gender role of the individual*

a) What a person says or does to disclose himself or herself as having a status of boy or girl, man or woman.
b) Spontaneous — topics of talk, casual conversation.
c) Replies to oblique or direct inquiry.

TABLE XII
*Assignment of sex to an individual*

| | |
|---|---|
| Pitfalls:- | 1. Uncertainty too long Confusion and protective secrecy. |
| | 2. Unwise emphasis on genetic, gonadal and hormonal data. |
| | 3. Premature decision from external genitalia Vs. congenital adrenal hyperplasia. |

*Summary*

I would like to summarise that within the last 10-15 years great strides have been made, though not all problems of intersexuality have been solved. Experimental embryology has contributed some insight into the pathogenesis of various forms of hermaphroditism. Cytology has contributed ability to establish sex by chromosomal arrangement. This is a tremendous diagnostic aid to the clinician. Endocrinology again has offered the means of treatment to achieve and maintain a relatively normal growth and development in the abnormal forms of sex differentiation. Finally, based on sympathetic understanding of sex of rearing and gender role, gynaecological and urological surgery, by removal of contradictory organs and construction of useful ones, have helped to establish more physiological and psychological relationships. Procedures should not ordinarily be based on estimation of genetic and gonadal sex or else these patients would stumble through life with major social and psychological handicaps.

*Acknowledgements*

I am extremely grateful to Dr. G. Sadasivan, Additional Professor of Anatomy, Osmania Medical College, Hyderabad, and his team for their in valuable help in preparing the karyotypes for two of the cases.

My thanks are also due to Dr. Suresh Rao, Research Assistant, Institute of Obstetrics and Gynaecology, Hyderabad, for his immense help in preparing this presentation.

Due to the good work of Mr. V. Swamy, the Photoartist, Government Maternity Hospital, the slides were made available. My sincere thanks are also to my Secretary Miss Shantha Narasimham for her ever-ready co-operation.

*References*

1. Jones, Jr., Howard, W. and Scott, William Wallace: Hermaphroditism, Genital Anomalies and Related Endocrine Disorders, Baltimore, 1958, Wilkins and Williams.
2. Moszkowski, E F., et al: Obst. & Gynec. **25**: 329, 1965.
3. McKusick, U. A.: Human genetics, p. New Jersey, 1965, Prentice — Hall. Inc.
4. Naidu, Prema M. and Mehdi, Z.: J. Obst. & Gynec. Brit. Emp. **66**: 122, 1959.
4. Naidu, Prema M., Ramaswamy, S. and Srinivas Rao, K.: J. Obst. & Gynec. Brit. Comm. **72**: 437, 1965.
5. Stewart, J. S. S.: Annals Roy. Col. Surg. Eng. **37**: 374, 1965.

*Figs. on Art Paper I to VI*