# Observations On 69 Hermaphrodites In Kuwait

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In Kuwait, the 1.7 million cosmopolitan "mosaic" population show a unique profile with regard to structure characteristics and prevailing disorders. It is characterised by (1) A wide ethnic heterogeneity (2) A high average inbreeding coefficient (F=0.0227) (3) A large family size with overall population growth (4) The existence of genetic isolates and (5) The pre- sence of new and rare chromosomal and mendelizing gene disorders with a higher frequency of AR disorders compared to the Western World (Farag'& Al-Awadi, 1988). Among the 156 cases with different sex differentiation anomalies ascertained during a decade (from 1979 to 1988), the precise diagnosis and management of 49 cases (71%) was done in the prepubertal age.

Many interesting observations were detected among the 69 hermaphrodites with different etioloyy (Table 1) and will be discussed briefly:

(1) Forty one patients presented as females (59.5%) and 28 as males (40.5%). All cases were assessed by a multispeciality team. Histopathological studies of gonadal biopsies were done in 24 cases (34.8%) and receptor study was done in one case with Refeinstein syndrome (1.45%).

(2) 5/69 cases showed different sex chromosomal abnormalities including mosaic 45,X/46,XX; 46,XX/46,XY dispermic chimerism; 46,XX true hennaphroditism; mixed gonadal dysgenesis with 45,X/46,XY/46,XYq- and mos 45,X/46,XYq-.[1,2,3,4]

(3) 42/69 cases were detected with autoson.al recessive mode of inheritance (60.8%) including 18 female pseudohermaphro- dites (FPH) with congenital adrenal hyperplasia, seven male pseudohermaphrodites with persistent Mullerian duct syndrome and 17 (reductase deficiency cases.[5,6,7,8]

(4) 21/69 cases with X-linked recessive mode of inheritance (30.4%) were detected including 13 cases with testicular feminization syndrome, 6 cases with pure gonadal dysgenesis and 2 cases with Refeinstein syndrome.

(5) One patient with FPH (1.45%) was due to maternal intake of steroids during the first trimester.

(6)The prevalence of different forms of hermaphroditism is approximately 1 in 20,000 in Kuwait.

These observations emphasises the importance of clinlco- genetic studies of hermaphroditism among different ethnic populations for better understanding of prevalence, patho-genesis and effect of different transcultural factors in practical genetic counselling.

Table: I. Clinicogenetic data in the 69 hermaphrodites.

| Group | Diagnosis | Rearing sex | | No. of cases |
|---|---|---|---|---|
| | | M | F | |
| I | Male Pseudohermaphrodies: | 26 | 19 | 45 |
| | a. Testicular feminization syndrome (13) | | | |
| | b. Pure gonadal dysgenesis (6) | | | |
| | c. 5 α reductase deficiency (17) | | | |
| | d. Persistent mullerian duct syndrome (7) | | | |
| | e. Refeinstein syndrome (2) | | | |
| II | Female Pseudohermaphrodites: | 0 | 19 | 19 |
| | a. Congenital adrenal hyperplasia (18) | | | |
| | b. Iatrogenic (maternal teratogen) (1) | | | |
| III | True Hermaphrodites: | 2 | 1 | 3 |
| IV | Mixed Gonodal Dysgenesis: | 0 | 2 | 2 |
| | Total | 28 | 41 | 69 |

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