

P3-P343 Genital abnormalities as seen in the University of Port Harcourt Teaching Hospital, Nigeria.

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OBJECTIVES

Introduction: Genital abnormalities are a source of concern and anxiety to parents and patients and in some cases, for the physicians who may have difficulty making pathological and eventually genetic diagnosis. They range from simple small penis and labial adhesions to the complex genital ambiguity and disorders of sex development.

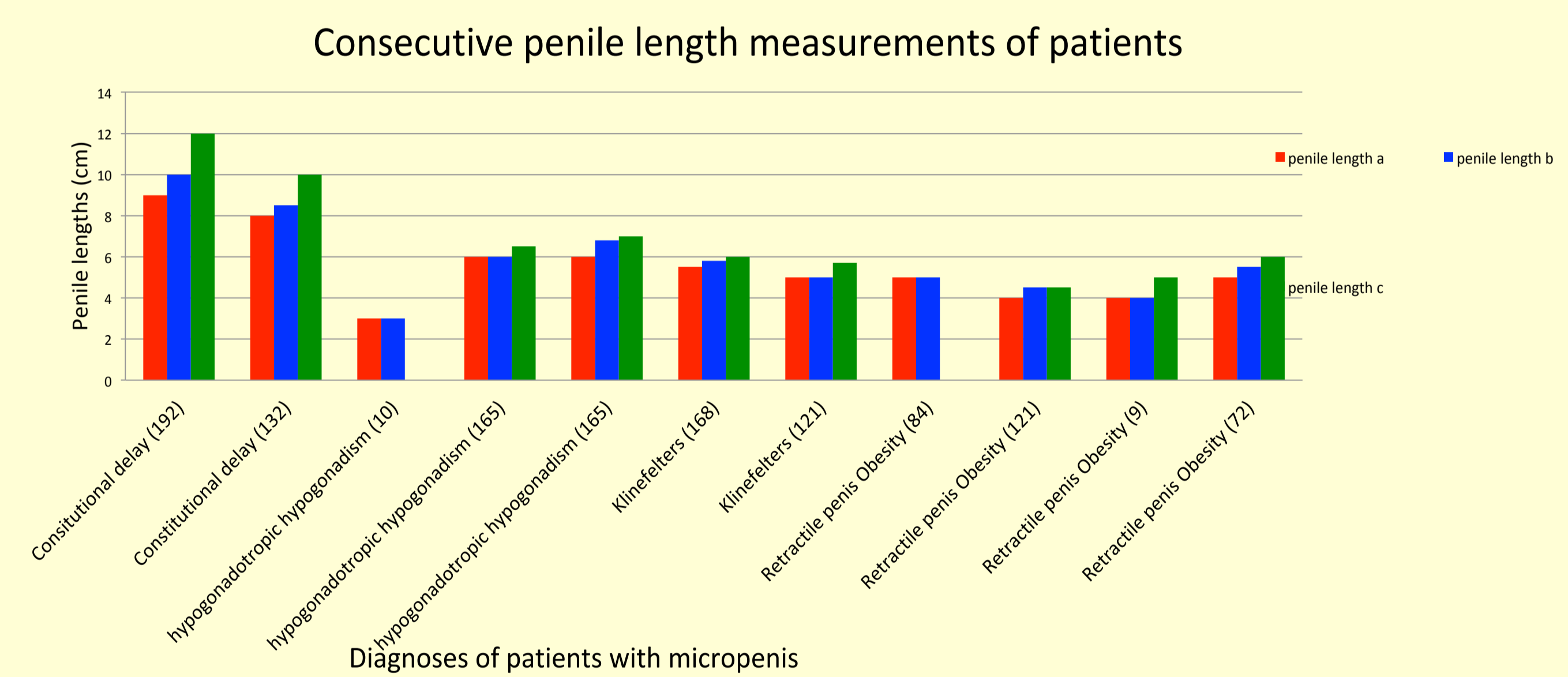
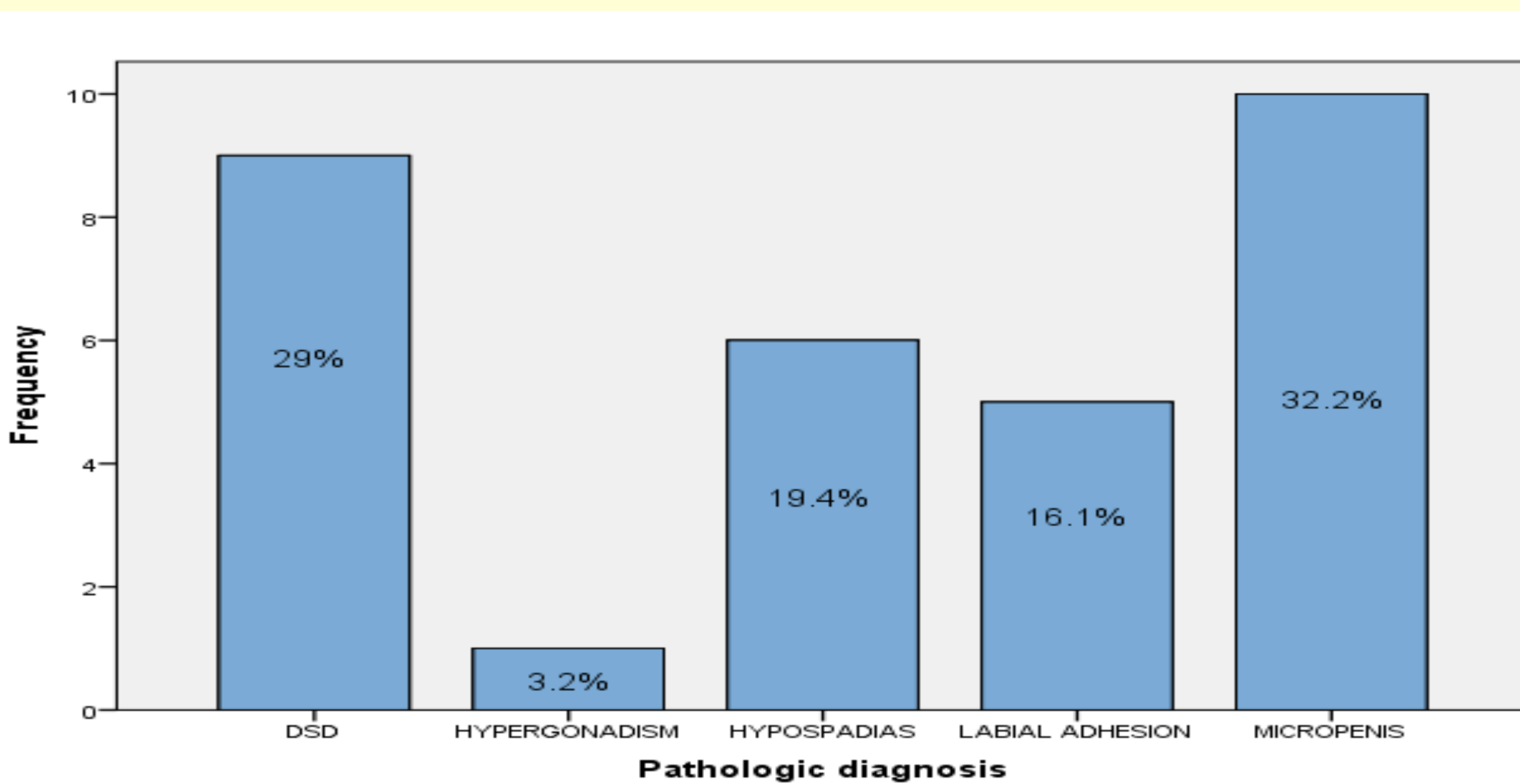
Objectives: To determine the types of genital abnormalities presenting in the Paediatric endocrinology unit of the University of Port Harcourt Teaching Hospital and discuss the management and challenges faced during this period

PATIENTS AND METHODS

A retrospective cohort review of all children presenting to the Endocrinology unit of the Department of Paediatrics, UPTH with genital abnormalities was undertaken from 1st of January 2013 to 31st of December 2017. The evaluation of the children included detailed history, physical examination, age and sex of rearing at presentation, clinical presentation, investigations, management and outcome of treatment /follow up. Stretched penile lengths were measured with a non stretchable calibrated tape, using a wooden spatula placed at the dorsal base of the penis with the penis fully stretched minding the comfort of the patient. Human chorionic gonadotropin test was done for 7 children with micropenis and their penile lengths measured at 3 months and 6 months after. Karyotype of patients was done in the genetic laboratory of the Obafemi Awolowo University Teaching Hospital to determine chromosomal sex.

RESULTS

There were 31 children presenting with genital abnormalities of various kinds. Sex assigned to these children was 8 females and 23 males irrespective of complete pathological diagnosis. The median age of presentation was 13 months with a range of 0.1 – 168 months. The commonest diagnosis was micropenis (32.2%) with various forms of DSD being the second commonest (29%) and most females had labial fusion (16.1%). Females with labial fusion had complete resolution following oestrogen cream application, and 4 of the 7 children with DSD died during the period under review. hCG stimulation increased penile length of males with constitutional delay in puberty, who were reassured and only marginally in those with hypogonadotropic hypogonadism and Klinefelters syndrome.



DISCUSSION AND CONCLUSIONS

Making diagnosis and managing complex genital abnormalities like DSD in UPTH remain challenging because of lack of diagnostic equipment and drugs. Collaborating with European and other national laboratories help our team reach some pathologic diagnosis however genetic diagnosis is still difficult. Though some conditions seem simple to diagnose, managing the psychological consequences may not be as straight forward. Parents of children with DSD always ask for surgical intervention and with counselling, many reluctantly agree to postpone the surgeries. Hydrocortisone tablets for children with CAH is usually difficult to obtain so many have to use prednisolone. All parents were concerned about their children's chances of fertility and some who were counselled on fertility preservation, were willing to assist their children undergo this process if needed.

References

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