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Agenesis of the Penis

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Summary

Odaibo SK. Agenesis of the Penis. *Nigerian Journal of Paediatrics* **1981; 8:81.** A case of complete absence of the penis with the presence of a normal scrotum is reported. As far as we are aware, only 42 cases of malformation of the penis have been reported in the literature. Penile agenesis involves an enormous amount of cultural, psychological, ethical and surgical problems to both the parents and the child. This case has been described to illustrate some aspects of these problems, their evaluation and management.

Introduction

AGENESIS of the penis was first reported in 1854[1] and by 1960, only 28 cases had been reported,[2] thus illustrating the extreme rarity of this anomaly. The incidence is estimated at one in every 30 million male births.[3] The condition results from incomplete development of the genital tubercle in the embryo, and the urogenital sinus thus opens immediately anterior to the anus.[4]

Case Report

BO was born on 4th March, 1977, to a 25-year old gravida 3, para 2, Nigerian woman. Delivery was in hospital and uneventful. There was no family history of congenital abnormality. Attendance at hospital for antenatal supervision was irregular. Other than Fergon and Multivite, no drugs or local herbs were ingested by the mother during the period of gestation. Examination of the newborn revealed absence of the penis and a normal scrotum with descended testes (Fig). There was no palpable penile tissue and no skin tag was seen around the anus. Both urine and meconium were passed from the anal orifice. The blood urea nitrogen was normal. Buccal smear revealed a male genotype. An excretory urogram (IVP) showed normal functioning kidneys. Anterior to the urinary bladder was a large cyst filled with contrast material. The urethra opened into the rectum. A barium enema was not done.

One week after birth, an exploratory laparotomy was performed. There was a thin-walled cyst lying to the right and slightly anterior to the urinary bladder and communicating with it. No female sex organs were seen. The cyst was excised and the bladder wall closed. A Foley's catheter was placed in the bladder and brought out in the suprapubic region.

Post-operative recovery was satisfactory and the wound healed well. At one month of age, the suprapubic catheter was changed. Repeated checks were made on the serum electrolytes which throughout, were within normal limits. Bilateral orchidectomy was planned, in conjunction with

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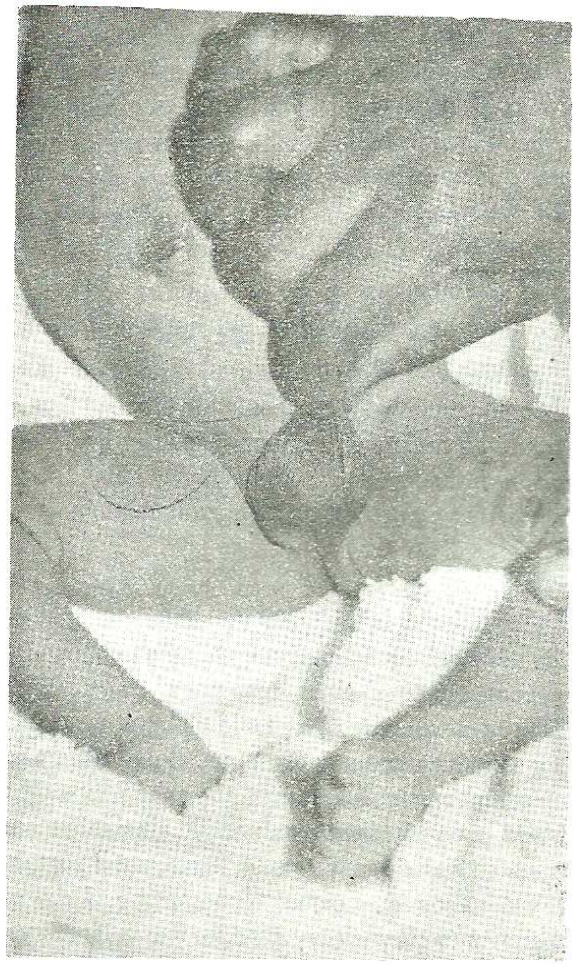

Fig. Photograph of a 5-day old infant, showing complete absence of the penis. Note the presence of a normal scrotum.

vaginoplasty, using scrotal skin. The presence of an ectopic urethra posed a major problem but the possibility of dilating this urethra which opened into the rectum and its ultimate anterior transposition, was considered. The parents of the child were approached and the full implications of the planned management explained to them. They had given an assurance to return within 24 hours with an answer, but 2 days later, they returned to request that the child be discharged home. They were made to understand that their request was being made against medical advice and at the risk of the child's life. Before discharge however, 2 weeks follow-up appointment was given. The parents turned up at the follow-up clinic to report that the child died, 2 days after discharge from hospital. The cause of his death remained unknown.

Discussion

Although penile agenesis is not incompatible with life, coexisting malformations have accounted for early deaths of approximately one-third of patients reported.[5] Sudden death occurred in one case of an adult with congenital absence of the penis[6] and in that particular case, suicide could not be ruled out. The psychological trauma from the feeling that he was neither masculine nor feminine might have been contributory.

In some Nigerian communities, elaborate preparations are made for the naming ceremony of a child on the 8th day of birth and the choice of names to reflect the sex of the child is made before that day. Appropriate family title is also conferred on the child on this day. Failure by parents to conform to this, often arouses family concern and community curiosity on the child and an unsatisfactory or cover-up explanation for the delay or postponement of this ceremony, becomes a source of embarrassment to the family. The psychological trauma on the parents of an infant with agenesis of the penis at this stage must be overwhelming. The reported sudden death of our patient which occurred under circumstances undisclosed by the parents was no surprise. For an infant with agenesis of the penis, the psychological trauma is borne directly by the parent who succumb to the pressure of cultural taboos rather than fight against it to raise a child who ultimately will become a social outcast in the community.

For those who survive infancy however, the importance of early sex assignment, within the shortest possible time has been emphasized.[7] Early sex assignment permits a timely orientation by parents, accurate birth records and an early development of the child's core gender identity. Infants born with agenesis of the penis should be raised as females as problems will be fewer and more easily manageable.[8]

Acknowledgements

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