Case Report

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Aphallia: A case report and literature review

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ABSTRACT

Aphallia is a complex urogenital malformation. It is rarely described in literature. Treatment calls upon feminising genitoplasty in most cases. Authors describe a case in a neonate, insisting on the sociocultural realities which guided their indications.

Key words: Aphallia, phalloplasty, transposition

INTRODUCTION

Aphallia is a congenital malformation with dramatic psychological consequences.

About a hundred cases are reported in world literature.[1] Aphallia is an anomaly which occurs during the fourth week of embryonic development, which accounts for its frequent association with related malformations. It is related to result from the non-formation of the genital tubercle or its failure to develop. Its diagnostic is easy at birth, as there is an absence of penis. However, some cases were diagnosed late.[2] Treatment generally performed is feminising genitoplasty or in very few cases, phalloplasty. We are hereby reporting a case of aphallia which affected a neonate whose parents have decided to keep the male sex. This is the third case of aphallia described in Africa, after the two cases reported by Deschamps *et al*.[3] We insist on the sociocultural realities which guided our indication.

CASE REPORT

A neonate referred from a health centre 8 hours after birth due to an absence of penis. The pregnancy that led to his birth was at term. The child's mother has never had prenatal visit or undergone obstetrical echography.

Clinical examination showed a good general state, an absence of penis [Figure 1] and urinary meatus. Palpation revealed the presence of the gonad inside the scrotum. The anus was in normal position, permeable, with the presence of meconium mixed with urine.

Direct cystourethrogram showed a lower-end



Figure 1: Absence of penis

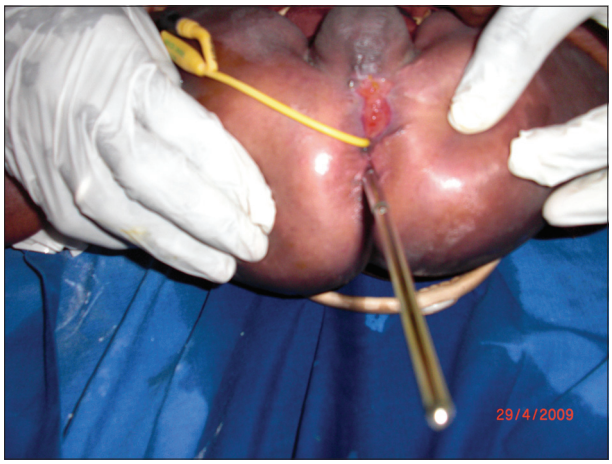


Figure 2: Anterior transposition of the urethra

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urethrorectal fistula and a grade III right vesicorenal reflux. The other imaging investigations (standard abdomen X-ray, abdomen and heart echography and the CT scan) were normal. Full blood count, blood ionogram, hepatic and renal state were normal. Cytobacterial examination of the urine was normal. The neonate was masculine, 46 karyotype, XY. Cystostomy was performed 4 days after birth followed by anterior transposition of the urethra at the 10th month [Figure 2].

He presented an evisceration at day 15 post-operatively, which was successfully repaired. At the age of 1 and a half years, the cosmetic aspect of the perineum was satisfactory with a well-anchored urethra at the level of the median raphe and the anus was at its normal position. The renal function was normal and no urinary infection was detected. However, the vesicorenal reflux persisted. It was planned that the child would undergo phalloplasty at the age of puberty because his parents decided to keep the male sex.

DISCUSSION

The birth of a child affected by aphallia puts parents in a highly delicate situation. It is the responsibility of the medical doctor to reassure them by proposing therapeutic options, that is, feminising genitoplasty and phalloplasty. When diagnosed early, feminising genitoplasty is often suggested, supplemented by bilateral orchiectomy before the age of 3 months and an oestrogen-therapy at the age of puberty,[4] along with psychological monitoring. When diagnostic is late, phalloplasty is recommended to minimise psychological trauma for both the child and parents.[2] The ultimate choice rests however upon the parents.

In Senegal, parents prefer to have boys rather than girls, even when the boy presents a complex genitourinary malformation. Consistent with that attitude, the parents of our neonate have decided to keep the male sex. That is why we have performed cystostomy first, followed by transposition of the urethra while waiting for the phalloplasty, which will be performed at the age of puberty.

As regards the two therapeutic methods, feminising genitoplasty is easier to perform than phalloplasty, which calls for a multidisciplinary team and presents in addition some disadvantages, such as functional and aesthetic sequelae and intolerance of the transplant.[2,5,6]

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