Diphalia

Torres-Medina E, Sánchez-Puente JC, Aragón-Tovar A, Camacho-Trejo V, Colorado-García A.

- ABSTRACT

Diphalia is a rare congenital abnormality that appears in 1 out of every 5 to 6 million male live births. It is usually accompanied by genitourinary, skeletal muscle, cardiac and hepatic alterations. Its treatment should be individualized and will depend on accompanying congenital abnormalities. The main purpose of treatment is to preserve continence and erectile function.

**Key words:** Diphalia, Congenital abnormalities.

- RESUMEN

La difalia es una anormalidad congénita rara, la cual aparece en 1 de cada 5 a 6 millones de nacidos vivos, suele acompañarse de alteraciones genitourinarias, musculoesqueléticas, cardiacas y hepáticas, donde su tratamiento deberá ser individualizado y dependerá de sus anormalidades congénitas acompañantes, con el fin de preservar la continencia y la función eréctil.

**Palabras clave:** Difalia, anormalidades congénitas.

- INTRODUCTION

Diphalia is a congenital alteration resulting from the lack of fusion of the genital tubercle and is accompanied by other congenital abnormalities. Diphalia was first described by Wecker in 1609 and less than 100 cases have been reported in the medical literature.

The case of a patient with a double penis, forked testicles, a double urinary bladder and double prostate is presented. The patient also presented with grade V right vesicoureteral reflux and skeletal and muscular alterations. He sought medical attention for his genital abnormalities. He had been evaluated in childhood but abandoned treatment.

- CLINICAL CASE

The patient is a 20-year-old male belonging to a low socioeconomic stratum. The patient’s brother had an unspecified congenital heart disease. Previous surgical
Excretory urography shows severe ureteropyelocaliceal dilatation of the right kidney at the moment of contrast material administration. The left kidney is shown to adequately concentrate and eliminate the contrast material.

Patient began his sexual activity at 18 years of age and stated that he had had sexual contact with both penises.

He sought medical evaluation for his double penis and forked scrotum. Study protocol revealed he presented with double urethra, double prostate, double urinary bladder, grade V right vesicoureteral reflux with secondary kidney damage and shortening of left inferior extremity and muscular hypotrophy, lack of right major gluteus and absence of ipsilateral Achilles tendon (Images 1, 2, 3, 4).

Patient presented with two penises in superior and inferior positions. The glans and foreskin of both presented with normal characteristics and urethral meatuses were in their normal position. Palpation revealed normal corpora cavernosa. Scrotal bag was forked and the testicles were also in superior and inferior positions. Size and consistency of the testicles was normal and the anus was permeable.

Laboratory tests reported the following: Leukocytes 5.7 1000/μL, Hb 15.2 g/dL, Htc 46.3%, Platelets 302 thousand/μL, Glucose 103 mg/dL, Urea 13 mg/dL, Creatinine 0.7 mg/dL Na 144 mEq/L, K 3.9 mEq/L, Cl 105 mEq/L, PT 11.3 sec, INR 0.99, PTT 29 sec. Urinalysis reported: Density 1010, pH 6.5, Acetone +, Hb ++, Proteins ++, Sediment: 35 leuk/c, 6 eryth/c, abundant bacteria. Urine culture: positive for *E. coli* with more than 100,000 CFU.
Treatment: Phalectomy of the superior penis was carried out leaving one aesthetic and functional penis (Images 5, 6, 7).

**DISCUSSION**

Diphalia is a rare congenital abnormality appearing in 1 in every 5 or 6 million live births (1,2). Fewer than 100 cases have been reported in the medical literature (2,3). It is usually accompanied by other congenital alterations such as urogenital, gastrointestinal, cardiac and skeletal muscle abnormalities (4,6,7). Double bladder is a rarer manifestation (8,9).

Diphalia is believed to take place during the embryonic development of the penis around the 3rd to 6th week of gestation (10). Its development begins with the coalescence of the bilateral cloacal tubercle from the anterior and final parts in the cephalic portion of the urogenital sinus. The columns of the mesoderm that develop around the lateral margin of the cloacal plaque form the genital tubercle. The probable cause of diphalia is a longitudinal duplication of the cloacal membrane including more than 3 columns of the primitive mesoderm that migrated centrally around the 2 cloacal membranes forming 2 genital tubercles. Other related abnormalities in the bladder, colon, anus and spinal column are also produced (4,7). It has been suggested that diphalia could be due to a failed twin development (11).

Diphalia can be orthotopic or ectopic depending on whether the division of the penis is sagittal or frontal and symmetric or asymmetric (2,3). The Aleem classification is the most accepted (5). It divides diphalia into true diphalia and forked phallus and subdivides it into partial and complete. Pseudodiphalia described by Villanova and Ranetos corresponds to partial diphalia.

Ultrasound is used for diagnosis. It detects the number of *corpora cavernosa* or *corpora spongiosa* and their accompanying abnormalities (12). Improved interpretation of anatomical structures has been made possible with the advent of magnetic resonance and better decisions can be made when carrying out surgical interventions (13).

Treatment will principally depend on the type of accompanying congenital abnormalities as well as preserving continence and erectile function (14), which means individualizing each case (12).
CONCLUSIONS

Treatment of these patients should be surgical and individualized. Corrections should be made during the first years of life, especially of the accompanying congenital abnormalities. Treatment must always attempt to preserve normal anatomy and result in good continence and erectile function (14).

The patient has had good postsurgical progression at 8 months. He conserves his sexual activity and has adequate continence and satisfactory erectile function. He is presently under surveillance by the urology department.

BIBLIOGRAPHY