

Ectopic vas deferens as an incidental finding in a patient with VACTERL association

Conducto deferente ectópico como hallazgo incidental en paciente con asociación VACTERL

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Summary
Ectopia of the vas deferens is a rare congenital anomaly, usually associated with other congenital anomalies of the urinary tract and anorectum. It can have repercussions due to recurrent urogenital infections. From the knowledge of the embryology of the reproductive system and urinary tract, it is possible to understand the development of this entity. The ureteral bud arises from the mesonephric duct, the failure in the connection between these two primitive structures is what leads to the formation of an ectopic vas deferens. We present the case of a one-month-old male with VACTERL association and vas deferens ectopia.

Resumen
La ectopia del conducto deferente es una anomalía congénita poco común. Usualmente se encuentra asociada a otras anomalías congénitas anorrectales y del sistema urinario. Puede tener repercusiones como infecciones recurrentes urogenitales. Del conocimiento de la embriología del sistema reproductor y del tracto urinario se logra la comprensión del desarrollo de esta entidad. La yema ureteral surge del conducto mesonéfrico, la falla en la conexión entre estas dos estructuras primitivas lleva a la formación de un conducto deferente ectópico. Se presenta el caso de un paciente de un mes de nacido con antecedente de asociación de VACTERL y ectopia del conducto deferente.

Introduction
Ectopia of the vas deferens is an uncommon entity, one could even say that it is rare. Few cases have been reported in the literature, likewise, the description of the cases varies in the semiological characterization between them, it has been documented associated with other anomalies such as imperforate anus, undescended testicles, seminal vesicle agenesis and ureterovesical reflux.

The case described here is of a male infant patient with a history of association of VACTERL and urinary tract infection, in whom the finding is documented by voiding urethrocystography.

Case report
Male infant patient (1 month old) with a history of VACTERL association: imperforate anus, ostium secundum atrial septal defect without hemodynamic repercussions, agenesis of vertebral bodies, left cryptorchidism and left renal agenesis. Admitted to the emergency department for febrile syndrome, without other symptoms or findings of importance, normal paraclinical tests including hemogram, renal function, acute phase reactants, gastrointestinal and coproscopic panel. The urinalysis was altered and the urine culture was positive for multisensitive Escherichia coli, so intravenous antibiotic management with cefazolin was started. An ultrasound of the abdomen showed a single right kidney with dilatation of the central and peripheral collecting systems, associated with nephrolithiasis and reverberation artifacts in the calyces suspicious of air foci and urinary sediment.

He is evaluated by the pediatric nephrology and pediatric urology services who attempt, unsuccessfully, a ureteral shunt. Given the findings of bladder with thickened walls, intravesical detritus and dilatation of the upper urinary tract of the right single kidney, they suspected ureteral stricture. Renal scintigraphy with DMSA shows preserved cortical tubular function of the right single kidney, without signs of pyelonephritis or cortical scars.

Two days after the failed passage of the catheter, the patient was taken to transurethral cystoscopy + diagnostic retrograde ureteroscopy, finding a wide urethral

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meatus, an area of weakness towards the dorsal region near the bulbar urethra, and an area of anatomical alteration of the veru montanum due to anomalous insertion of the right ureter proximal to the bladder neck; the bladder showed moderate trabeculation. Distal ureteroscopy was performed with tortuous and dilated ureter and bladder catheter was passed. A control ultrasound of the urinary tract showed persistent central and right peripheral caliceal dilatation, associated with dilatation and tortuosity of the right ureter, with a slight decrease with respect to the previous study.

During the in-hospital evolution the patient did not present febrile peaks, had renal function within normal limits, received complete antibiotic management for nine days. After the negative control urine culture report, he was taken to voiding urethrocystography in which a tubular image is evidenced in the right posterior aspect of the bladder, which during the voiding phase shows a serpentiform configuration that extends towards the ipsilateral scrotal sac suggestive of ectopia of the right vas deferens; due to its disposition and given that this has communication with the bladder and urethra, it does not show alterations (Figures 1 and 2).

After the procedure, the urinary catheter is removed, the patient is discharged with recommendations, warning signs, a follow-up appointment by pediatric urology and prophylactic antibiotic therapy. In the subsequent outpatient course, he has had no new symptoms or associated infections and his kidney function has remained stable.

Discussion

The vas deferens are muscular canals located within the spermatic duct that form the continuation of the tail of the epididymis. Their function is the transport of spermatozoa, and they are usually between 50 and 60 cm long; at their distal end the ejaculatory duct starts (1, 2).

Ectopia of the vas deferens or also known as "persistent mesonephric duct" is a rare developmental anomaly, in which fusion of the ureter and vas deferens proximal to the bladder occurs; it is estimated to occur in less than 0.05% of the general population (3). Among the few documented cases, the first confirmed case was reported in the English literature around 1960 by Seitzman. It describes an age of presentation between three weeks of life and 40 years of age. Clinically, it may appear asymptomatic or associated with repeated urinary tract infections and epididymitis (4, 5). The different cases described in the literature vary in presentation and are usually associated with additional anatomical anomalies, the most frequent being seminal vesicle agenesis, vesicoureteral reflux, ectopia and renal abnormalities (5).

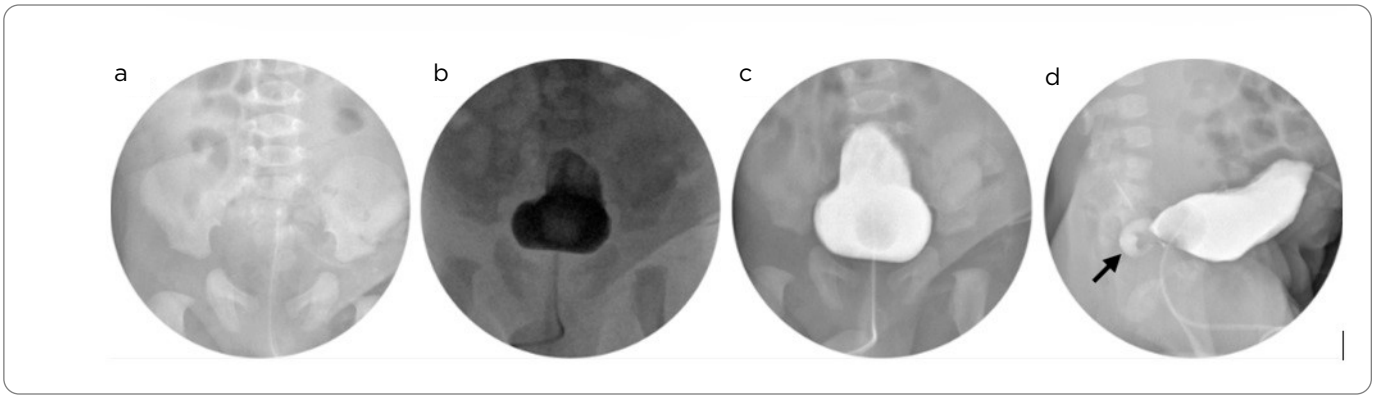


Figure 1. Urethrocystography. a) Preliminary anteroposterior image, without associated bone defects. b and c) Early (b) and late (c) bladder filling with 10 cm3 and 25 cm3 of water-soluble contrast medium, respectively, without evidence of reflux. d) Lateral projection showing complete bladder opacification with radiopaque tubular image in the posterior aspect of the bladder wall, with caudal trajectory, compatible with passage of the contrast medium to an ectopic insertion of the vas deferens (arrow).

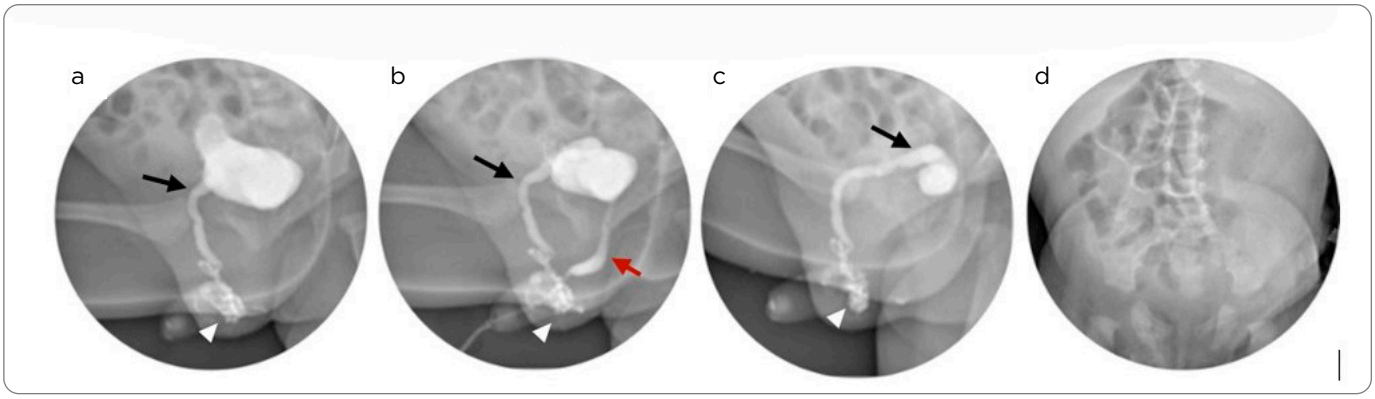


Figure 2. Urethrocystography. a, b and c). Oblique projections in voiding phase. A radiopaque, tubular and tortuous image can be seen, descending in a serpentiform manner (black arrows) towards the ipsilateral scrotum (white arrow head), compatible with anomalous trajectory, by ectopic vas deferens, with insertion in the posterior and right basal aspect of the bladder. The urethra is of usual characteristics (red arrow). d) No postvoid residue is observed.

Other less frequent associations of this entity include polyorchidism, with inguinal hernia and undescended third testicle associated with polydactyly (6), as well as tracheoesophageal fistulas and rectoureteral fistulas (7).

Ectopia of the vas deferens is usually caused by an alteration in the embryological formation of the vas deferens; the vas deferens originates from the Wolf's duct which, in turn, originates from the mesoderm; this will be the precursor not only of the vas deferens, but also of the ureter. Wolf's duct has three portions, a distal one, or common mesonephric duct, from which the ureteral bud originates; a medial portion, which gives rise to the juxtaureteral duct, and an upper portion, which gives rise to the vas deferens and the epididymis (8, 9). However, it is hypothesized that if the proximal portion of Wolf's duct invades the common mesonephric duct, it results in the vas deferens and ureter not achieving separate openings and sharing a common duct or ectopia of vessels of the ureter or bladder (10, 11).

This entity has a variable clinical presentation, in which it can also manifest with episodes of recurrent epididymitis, hematuria, unilateral testicular edema, infertility and as defects associated with recurrent urinary tract infections (6, 12). The diagnosis is confirmed by urethrocystography and cystourethroscopic catheterization. The role of cystography is key, since this study can demonstrate the passage of the refluxed contrast medium through the ureter and into the vas deferens. This was the method used in the present case, which allowed an anatomical determination of the site of insertion of the vas deferens, which can be left or right, or end in the bladder or in the ureter (6).

Other modalities, such as magnetic resonance imaging (MRI) or computed tomography (CT), can be used to confirm other abnormalities in the genitourinary tract, especially if a ductus deferens is found duplicated during surgery. Doppler ultrasound can also be useful to differentiate duplicated vas deferens from sperm vessels (6, 11, 12).

In terms of treatment, the aim is to preserve the renal function of the patients, preventing urinary tract infections and epididymitis, as well as preserving fertility. Surgical correction of ectopia has been described; however, there is still not enough medical evidence to recommend it (6, 7).

In this clinical case, an ectopia of the vas deferens was evidenced in a patient with a history of association of VACTERL, with imperforate anus, hypospadias and ectopic vas deferens, a triad recognized since 1989 (7, 12). It is believed that the association is secondary to the fact that the cloaca and the anorectal canal separate in the urogenital sinus around the fourth week of embryonic development, a period during which the absorption of the mesonephric duct into the urogenital sinus occurs at the same time (7, 13).

Conclusions

Because of its low prevalence, vas deferens ectopia is a diagnostic challenge. It is important to suspect, recognize and diagnose it in patients who present with recurrent urinary tract infections, hematuria and/or infertility, performing a multidisciplinary approach and understanding that it can be one of the manifestations of a syndromic presentation, usually associated with other urogenital or gastrointestinal malformations.

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