COMPLETE DUPLICATION OF THE URINARY BLADDER AND THE URETHRA IN A WOMAN – A CASE REPORT WITH A REVIEW OF ARTICLES

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KEY WORDS

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ABSTRACT

A complete duplication of the urinary bladder and the urethra is a rare congenital anomaly which is often associated with other congenital malformations. There are only few articles in the literature on this rare topic. This anomaly is presented in early age. The authors present the case of a 52-year-old woman in whom this anomaly was detected by laparoscopy from gynaecological indication. After 10 years from the diagnosis of the anomaly she is referred to the Department of Urology.

ABBREVIATIONS USED

PID – pelvic inflammatory disease
IVP – intravenous urogram
USG – ultrasonography
ATB – antibiotic
CT – computer tomography

INTRODUCTION

A complete duplication of the urethra and the bladder (CDBU) is a rare congenital anomaly which is often associated with other congenital malformations [2, 4, 7, 8, 10, 11, 12]. There are few articles in the literature on this rare topic [2–13]. A child with such an anomaly should be followed up immediately after the diagnosis to prevent possible complications. Anatomical disorders of the bladder are also associated with functional disorders, which is the potential cause of complications related to this congenital anomaly.

CASE REPORT AND MANAGEMENT

A 52-year-old woman was referred to the Department of Urology from the regional Gynaecology Department where she had been an inpatient for colpitis and had undergone extraction of an intrauterine device (IUD).

Table 1

Data for the separate urodynamics of each of the bladders (Pura max – maximal urethral pressure, Qmax – maximal flow rate, Qave – average flow rate, EMG – electromyogram of anal sphincter, Pbla – bladder pressure, Pdet – detrusor pressure, Pabd – abdominal pressure)

	Left bladder	Right bladder
Normal sensation /Pbla	179 ml/18 cm H2O	228 ml/11 cm H2O
Maximal capacity /Pbla	260 ml/27 cm H2O	292 ml/15 cm H20
Compliance	11 ml/cm H20	15.7 ml/cm H20
Uninhibited detrusor contraction	No	No
Functional length of urethra	27 mm	28 mm
Pura max	51 cm H20	51 cm H20
Qmax	11 ml/s	34 ml/s
Qave	4 ml/s	9 ml/s
Pdet/Pabd during voiding	45 cm H20/174 cm H2O	17 cm H20/89 cm H2O
Voiding time	53s	33s
EMG	Physiological finding	Physiological finding
Post-voiding residuum (ml)	38 ml	0 ml
Note:	During voiding, a low Pdet and a high Pabd were seen, suggesting thea low detrusor contractility with an insufficient evacuation bladder ability. The curve of micturition was irregular and flat.	The voiding curve was continuous in comparison with left bladder.

In 1996, when she was 43 years old, a duplication of the bladder was diagnosed by laparoscopy for a gynaecological indication. No other malformations were detected. At that time she was experiencing pelvic pain syndrome (PID) manifested by chronic idiopathic abdominal and pelvic pain and recurrent colpitis. These problems led to the indication for laparoscopy. Apparently, the first manifestation of the anomaly was recurrent interstitial bilateral nephritis at the age of 20 years. In spite of the late detection of the anomaly, additional screening including urological examination was not performed at the time. In May 2006, ten years after the discovery of the double bladder, a urological examination was completed during hospitalisation at the Department of Gynaecology as a consequence of painful voiding.

Abdominal ultrasonography (USG) confirmed the duplication of the bladder and showed cystolithiasis in the left bladder. An accidental finding from the abdominal USG was liver haemangioma. A subsequent intravenous urogram (IVP) showed normal excretory function of both kidneys with filling of the two separate bladders, each with one normally located ureter, but on the right side with a saccular dilatation indicating ureterocele.

According to the IVP cystogram, the bladder appeared duplicated rather than as a septum bladder, and the differen-

tial diagnosis was focused on this aspect. The urine culture confirmed significant bacteriuria of Streptococcus alpha haemolyticus and Enterococcus species, and antibiotic therapy was started. In the results provided by the Regional Health Institute, there was no mention of a duplication of the urethra. This anomaly was diagnosed by physical examination (Figure 1). The renal scan did not show passage obstruction. The voiding cystourethrogram showed a complete duplication of the bladder in the sagittal plane, with two stones in the left bladder where post-voiding residuum was significant (Figures 2, 3, and 4). However, vesicoureteral reflux was not confirmed by x-ray examination.

After repeated therapy for bacteriuria, cystoscopy was performed under epidural anaesthesia, which showed trabeculisation of the left bladder and ureteric plica with diverticulum at its proximal end. At this time, three calculi of 1.5 – 2.5 cm were detected in the left bladder. Using the intravenous indigo carmine, the ureteric orifice was detected just inside the proximal region of the ureteric plica**.** The cystoscopic view from the right bladder was dominated by pseudodiverticula, and the ureteric orifice was seen in one of these. The bilateral retrograde pyelography indicated in terms of IVP findings and for clinical purpose showed free passage on the right side, but there was a hypotonic ureter with a cut in the pelviureteral junction on the left side and an image of hydronephrosis. The cystolithiasis was fragmented by electrohydraulic lithotripsy and completely evacuated. In the postoperative period, fever and a positive urine culture were observed. After repeated targeted antibiotic therapy and extraction of the catheters, the patient had urine retention in her left bladder. The patient was discharged from the hospital on the 4th postoperative day after therapy with a permanent catheter, which was extracted during the check-up. The post-voiding residuum was 10 ml in the right bladder and 45 ml in the left bladder. The positive urine culture from the urine of the catheterised left bladder persisted, although the patient did not report any symptoms. During the check-up, we performed a computed tomography (CT) scan of the kidneys and the pelvis, which showed small cysts of 6 mm to 8 mm in the kidneys, and a polycystosis of the liver of 10 mm to 60 mm. The CT of the pelvis confirmed the double bladder and showed thickening of the bladder wall due to a chronic cystitis. We also completed a separate urodynamic examination of both bladders (Figures 5 and 6), whose results are shown in Table 1. During voiding, a low detrusor pressure (Pdet) and a high abdominal pressure (Pabd) were seen in the left bladder, suggesting a low detrusor contractility with an insufficient evacuation bladder ability. The voiding curve was irregular and flat, but the bladder outlet obstruction (BOO) was not confirmed.

The residual urine caused by the evacuation failure increases the risk of recurrent infections and formation of bladder stones. The urodynamic parameters of the right bladder, in comparison with the left bladder, were acceptable, and full evacuation was well-kept.

Except for the liver cysts, no other malformations were detected in the abdominal cavity and in the pelvis. At present, the patient is without symptoms, the urine culture of the separately catheterised urethras is free of bacteria, and the post-voiding residuum of 50 ml in the left bladder is acceptable. The evacuation function of the right bladder is complete. The patient is periodically checked as an outpatient.

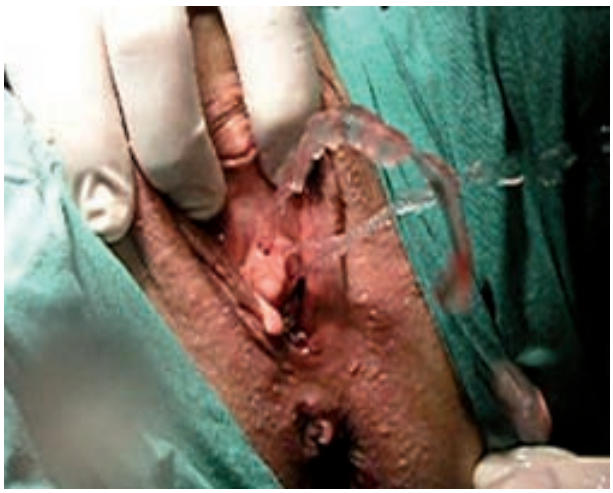

Figure 1
An examination of the vulva showed a double external orifice of the urethra and double flow during micturition

DISCUSSION

The urinary bladder and the urethra evolve from the endodermal element of the cloaca during embryogenesis. From the cloaca, the allantoideus duct (allantois) runs in the ventrocranial direction. Laterally in the cloaca, the Wolf's ducts end and later form the ureters. In the 2nd month of embryonic development, the urorectal septum divides the cloaca into a dorsal anorectal sinus and a ventral urogenital sinus. Later in embryonic development, the urinary bladder forms in the urogenital region, as well as the urethra and some of the genital organs. The caudal region of the urorectal septum later connects with the membrane of the cloaca in the 7th week of development. The membrane of the cloaca is divided into a ventral urogenital membrane and a dorsal anal membrane. The primitive urogenital orifice forms by the perforation of the ventral membrane. The membrane of the cloaca coalesces with the urorectal septum to create the perineum. The cranial region of the urogenital sinus forms the vesicourethral

Table 2
Reported cases of complete duplication of the urethra and the bladder (CDBU) with a review of their management (AU – accessory urethra, AB – accessory bladder)

Case No.	Reference	Year	Age	Sex	Associated anomaly	Treatment
2.	Voigt et al.	2005	1 d	M	Duplication of external genitalia; hypoplastic left kidney; bilateral bifid scrotums; imperforate anus; mass on perineum (teratoma)	Loop colostomy; removal of teratoma; left cystectomy and left penectomy; reconstruction of rectum
3.	Bae et al.	2005	7 yrs	M	None	Excision of AB and AU
4.	Gastol et al.	2000	1 yr 1 m 2 d	F F F	Duplication of vagina and uterus, symphysis diastasis; abdominal hernia Duplication of vagina and uterus; anal atresia with colon duplication Duplication of vagina and uterus; cloacal exstrophy with persistent urogenital sinus	Iliac osteotomy, approximation of symphysis and excision of hernia Excision of common wall of 2 descending colons and posterior sagittal anorectoplasty Reconstruction of cloacal exstrophy with anastomosis of bladders and vaginas
6.	Cheng et al.	1996	6 yrs	M	Accessory epispadiac meatal fossa	Excision of AU and AB, reconstruction of gland
7.	Ciftci et al.	1995	2 d	M	Incomplete diphallus, meningocele, imperforate anus	Transverse loop colostomy
8.	Goh et al.	1995	1st week	F	Duplication of vagina, uterus; stenosed anterior ectopic anus; double-outlet right ventricle; left uretrovesical junction obstruction, right vesicoureteral reflux, extrinsic duodenal obstruction	Vesicostomy; anorecto-urethro-vaginoplasty via the posterior approach; excision of bladder septum with reimplantation of right ureter; removal of vesicostomy; nephrectomy on the left
10.	Dajani et al.	1992	14 yrs	M	Chordee; congenital dislocation of right hip	Excision of AB and AU; correction of chordee
11.	Kapoor et al.	1986	1 d	F	Duplication of external genitalia, ventriculoseptal defect, malrotation of the gut, ectopic anal opening, maldescended left kidney, rachischisis of the lumbar spine and sacrum, umbilical hernia	Patient died of septicaemia
12.	Esham et al.	1980	7 yrs	F	Coarctation of aorta, ventral septal defect, duplication of colon with 2 appendices	Excision of diverticula from the left bladder, removal of the common septum
13.	Dunetz et al.	1985	14 d	M	Chordee	Excision of AB and AU, correction of chordee
14.	Varga et al.	2007	52 yrs	F	None	Electrohydraulic lithotripsy

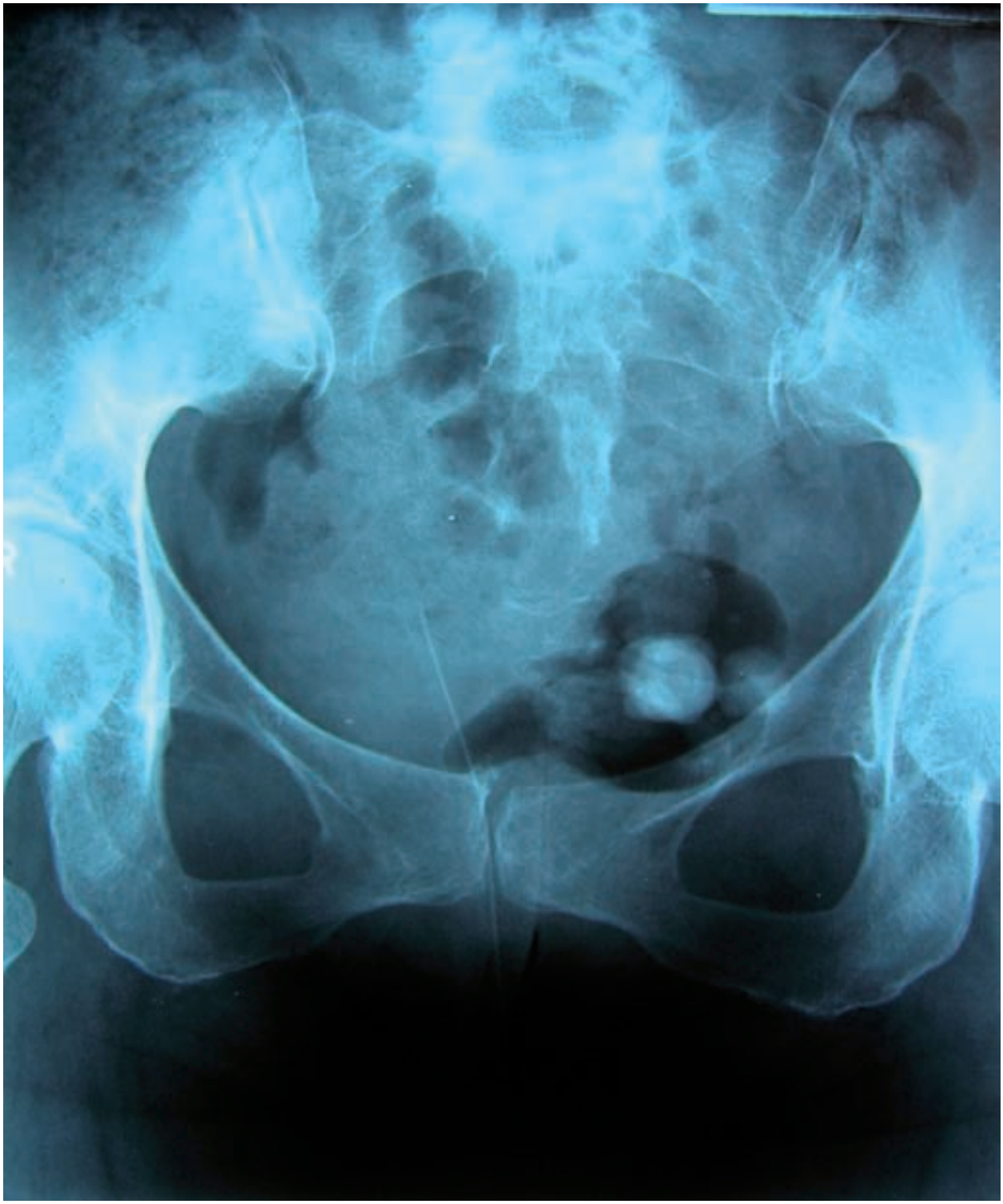

Figure 2
An X-ray examination of the pelvis with contrast calculi

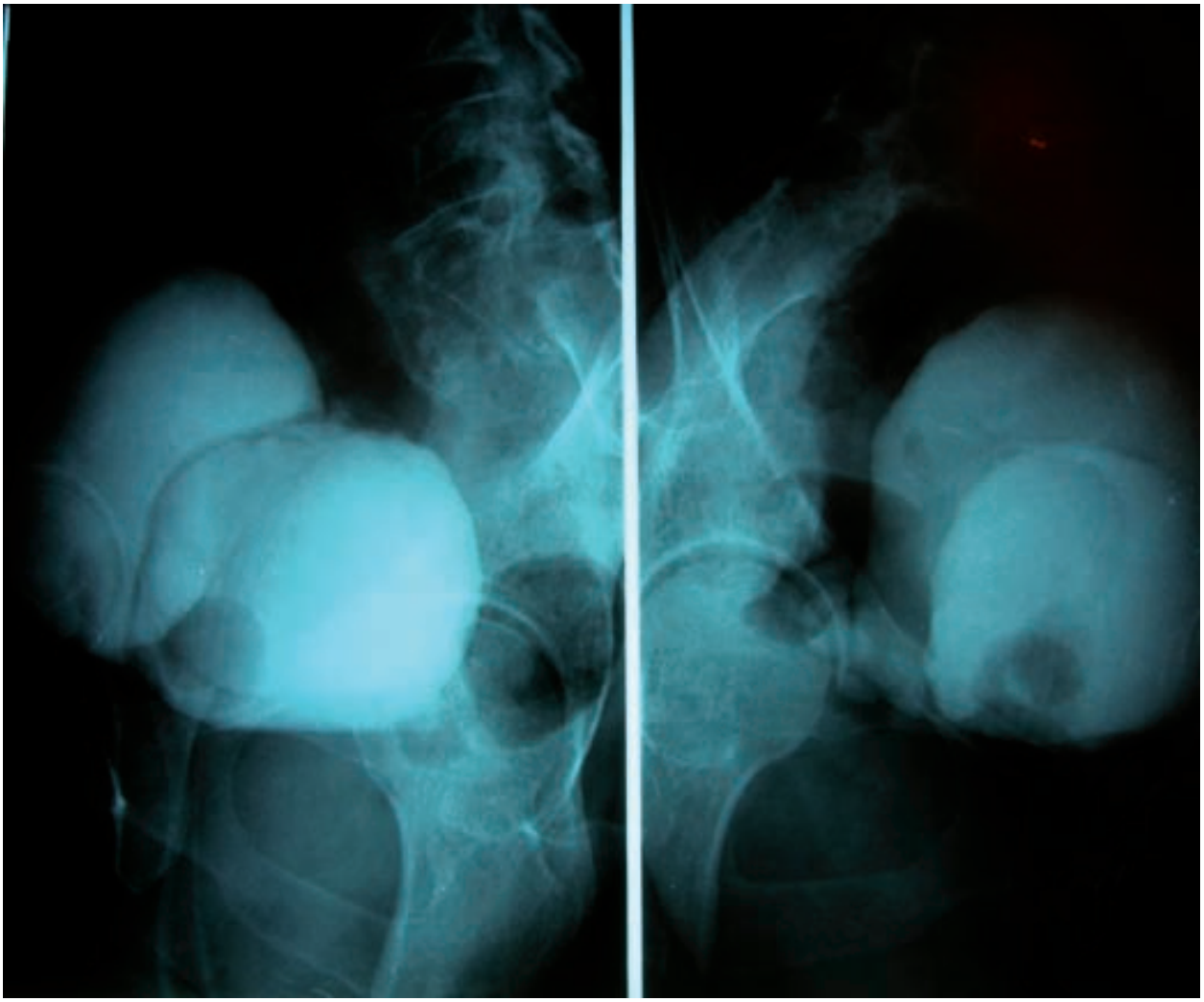

Figure 3
Cystography in the side projection, showing a complete duplication of the bladder

basement from which the urinary bladder evolves; a woman's urethra (or the prostate urethra in men) forms from the caudal region [1].

Failure of this developmental programme can lead to anatomical anomalies of the lower urinary tract [2, 8, 11]. The complete duplication of the urethra and the bladder in women is the result of a failure to form the vesicourethral basement of the urogenital sinus [1]. This anomaly, found more commonly in males than in females [12], is not only a latent "cosmetic" defect, but results in functional defects which may manifest clinically as pelvic pain disease (PID).

From the cases with CDBU reported in the literature, we illustrate the oldest patient in whom the anomaly was initially detected (Table 2).

CONCLUSION

This rare anomaly requires follow–up from the time of the first diagnosis because of the late complications that can result from bladder function failure, particularly evacuation of the bladder. In this time of neonatal ultrasound screening, this anomaly should not escape our notice in newborns.

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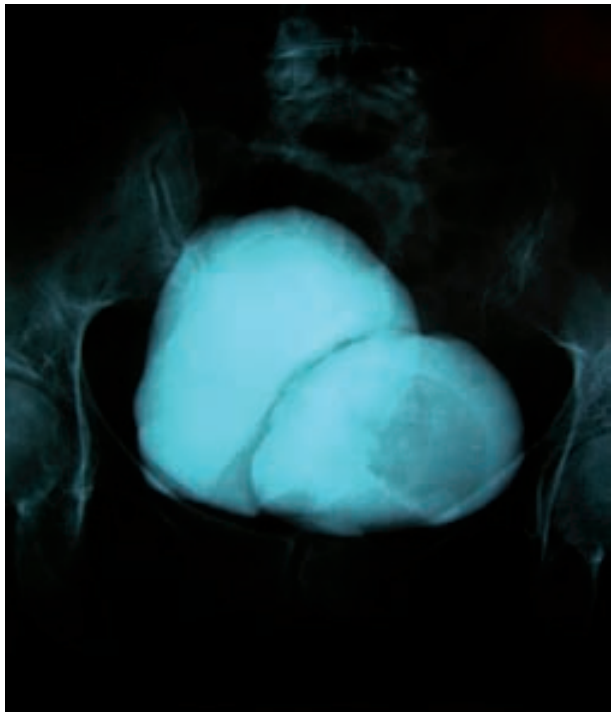

Figure 4
Cystography in the anteroposterioric projection, showing the complete duplication of the bladder

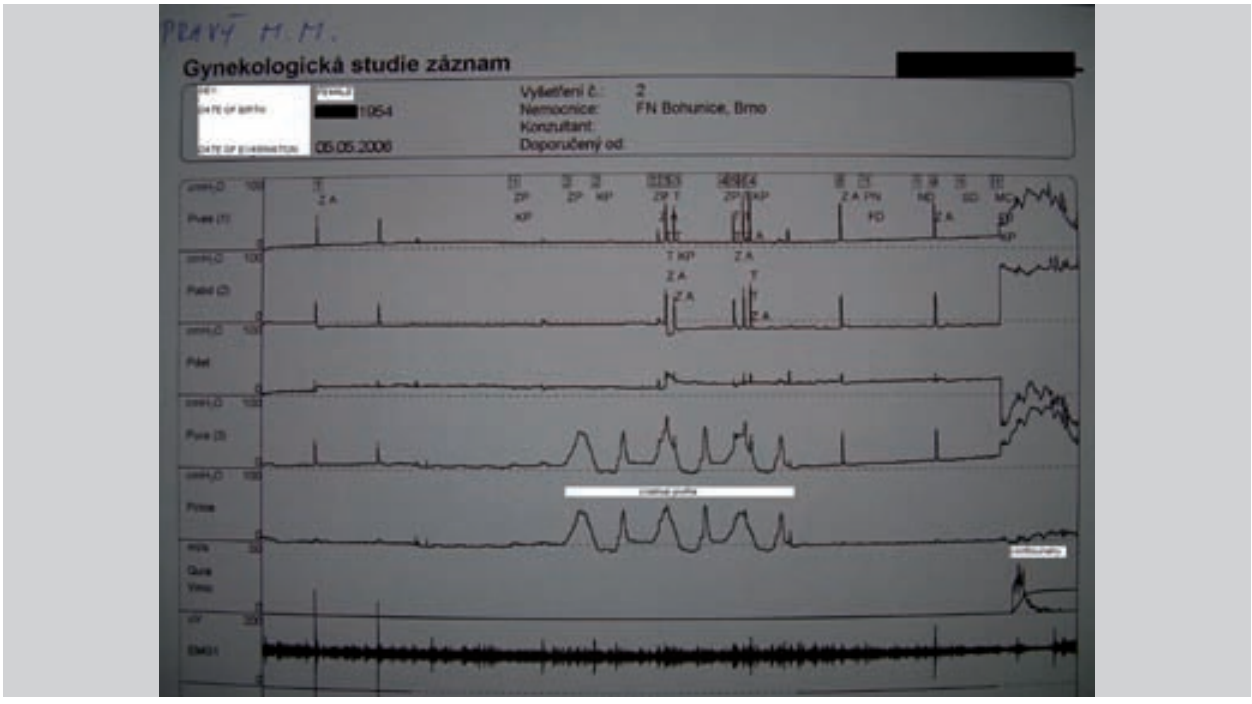

Figure 5
Image of the compresssed urodynamics curve of the right bladder

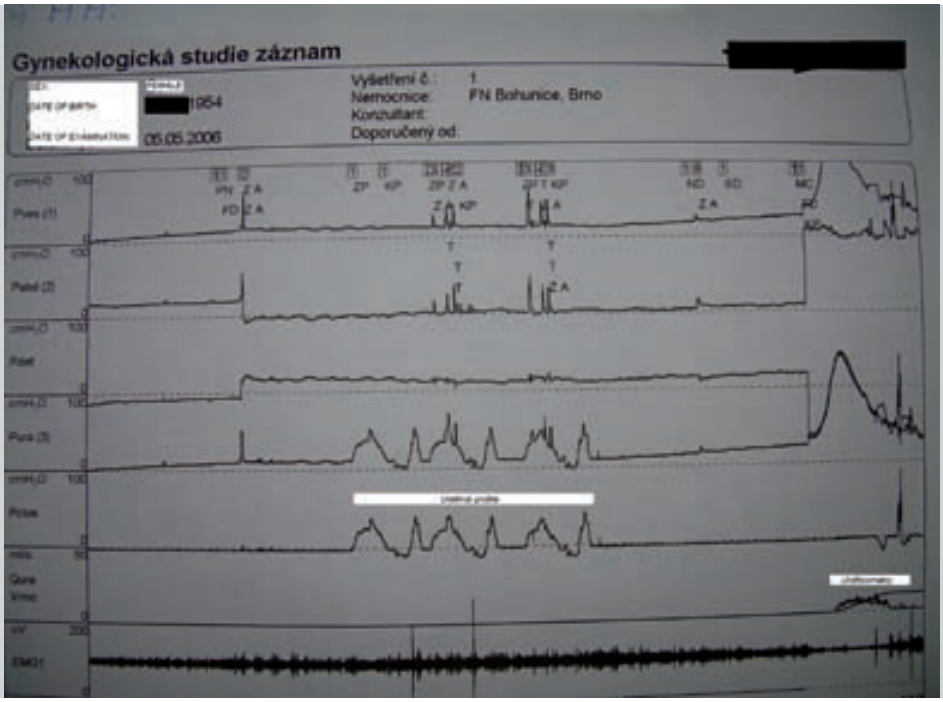

Figure 6
Image of the compressed urodynamics curve of the left bladder

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