Modified Voiding Cystography in Intersexuality: Report of the Procedure in a Case of Intersex

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Summary


A two-and-half-year old patient with intersex secondary to adenogenital syndrome showed an intravenous urography, vaginal opacification. Conventional genitography also showed only vaginal opacification. A modified technique of voiding cystography is suggested as a possible genitographic screening test in intersexual disorders, which may lead to a reduction in cost of management and radiation risk to the patient.

Introduction

Genitography is of utmost importance in the investigation and management of intersexual states. By delineating the internal genitalia anatomy, the procedure facilitates the assignment of a practical sex, which is dependent on the structural capabilities of the patient. Thus, when a vagina is demonstrated, no matter how rudimentary, the practical sex of rearing of the patient is female. This is because it is easier to transform an individual with ambiguous genitalia into a cosmetically adequate female than into a male. The techniques of genitography include “flushing” of water-soluble contrast medium through the perineal opening with a blunt-nosed syringe—the flushing technique—and via a single or multiple catheters placed in the internal genitalia—the catheter technique. The success rate is variable. However, adequate genitography is often achieved by combining both techniques.

The purpose of the present report is to document a modified technique of voiding cystography that may reveal enough of the internal genitalia, particularly the vagina, thus obviating the need for conventional genitography and lead to a reduction in cost of management and radiation dose to the patient. The name, “voiding genitography” is being proposed for this new technique.

Case Report

AA, aged 2½ years, presented with an abnormal external genitalia. The mother felt that the patient was a male although his “penis” was curved...
downwards and he also had no scrotum. There was a growth of mature pubic hair which the mother had had to shave several times to save her from embarrassment should the child's condition be discovered. The pregnancy had been uneventful, but she had taken some non-specific herbal concoction late in her pregnancy to ensure a normal delivery. The patient was the last of four siblings, the others being a normal female and two males.

The significant findings included pubic hairs with a feminine distribution. The phallus was hypertrophied with severe chordee and a single tiny perineal opening beside the anus. The labial folds were prominent but contained no gonads (Fig 1). The clinical impression was that of female intersex, secondary to adrenogonadal syndrome.

Laboratory studies revealed normal electrolytes and urea. Buccal smear for sex chromatin and karyotype revealed a female complement of 46XX. Urinary 17-ketosteroids and serum cortisol could not be assayed.

Radiological Examinations and Techniques

Radiological examination of the bones of the hands and wrists revealed accelerated skeletal maturation of about 7-8 years, that is, 5 years in advance of the patient's chronologic age when compared with Greulich-Pyle standard charts. Preliminary abdominal radiograph revealed no evidence of an adrenal mass.

Technique of voiding urography

Ten millilitres of diatrizoate (60% Urografin) was injected intravenously. Radiographs of the upper renal tracts were obtained at 3 minutes, and the kidneys, ureters and bladder at 30 minutes, at which time the child was noticeably restless and told the mother that he wanted to void. Just before the radiographic exposure, the child was noticed to have voided some urine. The upper renal tracts were normal without evidence of renal displacement. However, a vescus was opacified and projected across the left side of the urinary bladder (Fig 2). This was thought to represent the vagina and was later confirmed at conventional genitography (Fig 3). No uterine or tubal opacification was seen at the latter examination.

On the basis of the genitographic findings, a female sex was assigned to the patient, with the consent of the parents. She is being treated with cortisone acetate and has also had vulvo-vaginoplasty. Follow-up management of the patient is being carried out at the children's out-patients clinic.

Fig 1. Perineum of patient at presentation. Note mature pubic hairs and hypertrophied phallus.

Comments

Excretory urography is indicated in intersexual states to show evidence of renal function and structure since dysplasias and other congenital malformations are commonly associated with abnormalities of the caudal end of the embryo. The present case shows that by modifying this procedure into a voiding cystographic examination with the patient in the supine recumbent position, a lot more information may be obtained at one examination. In applying this modified procedure, it is suggested that after the upper renal series of radiographs, the bladder
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factor in the assignment of a female sex. The vagina is also often the only opacified genital structure, as in the present case.

Shopfner has classified genitographic anatomy into six types, all representing various degrees of variation from the normal female, as in the foetal period all humans develop as females in the absence of testis. In type I, there is simple clitoral hypertrophy; the vagina is separate and well-developed. Types II to IV represent further masculinisation of the phallus and the urogenital sinus with the vagina opening into the posterior aspect of the urogenital sinus. A single perineal opening is thus present. Types V and VI represent various degrees of what is clinically called hypospadias; no vagina is present in either type. Thus, with the vagina opening into the posterior aspect of the urogenital sinus in Types II, III and IV, reflux filling of the vagina is most likely to occur in the supine recumbent position during voiding. Reflux filling of the vagina is also seen as a normal phenomenon in over 70% of females and may therefore, occur in Type I genitographic anatomy, in which a well-developed vagina is present.

The present proposed technique may fail in the immediate neonatal period due to failure of the immature kidney to excrete and concentrate the contrast medium. Some older children may also fail or refuse to void and they should be subjected to conventional genitography.

In conclusion, voiding cystography in the supine recumbent position may produce reflux opacification of the internal genitalia, particularly the vagina, in intersexual states and is hereby proposed as a screening procedure in intersexuality.

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References


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