Structural Incontinence

Structural incontinence in childhood

CHAPTER 1

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Introduction

Many structural anatomic causes for incontinence exist at pediatric age. Several, such as the extrophy-epispadias complex, are well known to everybody; many others are unknown to the majority of physicians dealing with children. The embryology of the urinary tract teaches us that the ureter, trigone, bladder neck and proximal urethra in the male, and the whole urethra in the female, are derivatives of the distal part of the Wolffian duct. This means that derailment of a small part of the distal Wolffian duct can lead to a cascade of anomalies in the urinary tract. In fact, embryology can provide a completely logical explanation for severe reflux, ipsilateral renal dysplasia and bladder neck insufficiency in one and the same patient. The same holds true for the combination of dribbling incontinence due to an ectopic ureter and stress incontinence based on bladder neck insufficiency. This is a subject with very sparse backing from published data.

Anatomic causes for incontinence in children

Primary stress-incontinence
In children, stress incontinence occurs as a stand-alone disease or as a symptom in children with hyperlaxity of the joints. Clinically, it can present as an urge syndrome, due to forced hold manoeuvres with the pelvic floor, resulting in squatting and urge incontinence, predominantly provoked by stress. Anatomically, the lower urinary tract is usually normal, except for a relatively flat vesico-urethral angle. It occurs more frequently in conjunction with a hypospadiac urethral orifice, inside the hymenal ring, in patients with a relatively short urethra. There is only sparse literature on this subject.1-9

Stress and/or urge incontinence due to bladder neck insufficiency
When dealing with children suffering from incontinence and recurrent UTI due to urge syndrome, an open bladder neck on cystography or ultrasound is a common finding. In the anatomically normal child the bladder
neck will present as a ring with a dilated urethra down to a mid-urethral ring at the level of the pelvic floor, with the bladder filled to capacity under anesthesia. However, a group of patients exists with a clear gap in the bladder neck, a gap that appears to be a congenital bladder neck defect. When such a gap leads to urinary leakage into the urethra, the child will try to cope with this leak by using the pelvic floor as emergency brake. Clinically, the child may present as a case of urge syndrome. A congenital bladder neck defect can be suspected when biofeedback training fails. Such a defect has been described in combination with an ectopic ureter, it is apparent with double ectopic ureters, and it can occur as a single anomaly leading to incontinence.

Bladder neck insufficiency also occurs in girls with ectopic ureterocele-ureteroceles running through the bladder neck into the urethra. However, a large group of these children appears not to suffer from incontinence. The literature rarely includes urinary continence as a factor in the follow-up of the treatment of ectopic ureteroceles.

Urogenital sinus with bilateral ectopic ureters is another candidate for bladder neck insufficiency. Also at risk for urethral insufficiency are patients with adrenogenital syndrome with a high ending vagina. We do not know how to classify the female hypospadias, with an urethral meatus placed inside the hymenal ring and a relatively short urethra, but these children can present clinically with either stress incontinence or urge syndrome.10-15

**Urethral obstruction in boys**

In boys, urethral obstruction leading to bladder overactivity and urge-incontinence is common. Urethral obstruction in boys can occur along the whole course of the urethra. Bladder neck obstruction can be primary, or secondary to a more distal obstruction. Utricle cysts or a cystic cap covering the colliculus can lead to obstruction.

Posterior urethral valves are the commonest cause of urethral obstruction in boys. Several types are recognised. By far the most common valves arise from the colliculus and present as a membrane that partially closes the urethra.16-19 A few millimeters past the sphincteric mechanism the passage of the urethra through the pelvic floor can cause obstruction, a constricting ring known as Cobb’s web or Moorman’s ring.20-24 Just distal of
the pelvic floor, cystic anomalies of Cowper’s gland duct can present as syringoceles. In the same area, anterior diverticula, that can be a direct result of a destroyed syringocele, can arise. More distally in the pars bulbosa or pars pendulans of the urethra, diverticula arise very rarely.

A rare and very nasty anomaly is congenital urethral stenosis, that occurs mostly starting from the penoscrotal angle.

In the penile urethra rare obstructions can be found based on obstruction of the duct of a urethral gland, a cyst of the duct of Littré’s gland. More commonly, obstruction is caused by the fossa naviculare, a stenosis of the last centimeter of the urethra. Common obstructions occur in boys at the urethral meatus, especially after circumcision, by friction of the meatus to the clothes. Rarely is an obstruction caused by debris in a fossa naviculare sinus, a sinus running parallel to the distal urethra. Relatively rare is true outlet obstruction based on phimosis in the uncircumcised boy.

It is important to state that voiding cystourethrography (VCUG) is an unreliable investigation in diagnosing urethral obstruction in boys. In our experience, comparing urodynamic results with VCUG, false-negative results of VCUG are found in 60%, while only 6% false negative results occur with urodynamics.

Urethral obstruction in girls

In girls, urethral obstruction is rare. In the past, it has been diagnosed and treated very frequently, based on the finding of a spinning-top urethra at VCUG. We now know that this aspect is caused by a functional obstruction in the majority of cases. Urodynamically, in our experience, approximately 2% of the spinning top urethras reflect a true urethral obstruction. In the cases with a proximal dilation of the urethra at VCUG, the obstruction can be found 5-10 mm proximal of the meatus.

Meatal stenosis does occur in girls. When inspecting the meatus, an anterior position is often present. The history often reveals a urinary stream that runs under the toilet seat on the floor in front of the toilet. Very rarely primary bladder neck obstruction can occur in girls. Urethral diverticula in girls are rare and can give obstruction. Also rarely, urethral obstruction and incontinence can be provoked by anomalies of the
A hymen annulare can completely cover the meatus and lead to micturition into the vagina with vulvovaginitis and post-void dribbling as a result. Finally, severe forms of synechia of the labia minora can lead to obstruction, infections and post-void dribbling.

Epispadias and exstrophy
Nobody doubts the incontinence problems in exstrophy patients. However, we tend to forget that epispadias in boys come with congenital sphincter insufficiency, with or without a gap between the pubic bones. Rare variants of bladder exstrophy such as covered exstrophy or abdominofissura can come with sphincteric incompetence. The very rare female epispadias present with complete absence of the urethra and the pelvic floor, with the bladder neck at the level of the hymenal ring. Rare congenital anomalies of the lower urinary tract in females can cause all kinds of urinary leakage. Impressive collections of cavities and sinuses can cause incontinence in combination with variations of cloacal anomalies and urogenital sinus.

Dribbling incontinence
In girls, true dribbling incontinence, continuously manifest, is mostly caused by ureteral ectopy. In general, an upper pole ureter from a duplex system is involved, rarely a single ectopic ureter can be seen. The ectopic ureter can end in bladder neck, urethra, vulva, vagina, uterus and even in the rectum. The ectopic ureter always will end in a remnant of the Wolffian system. In boys, all Wolffian remnants are proximal of the external sphincter, thus not resulting in incontinence except for urge complaints based on irritation by a ureter ending into the prostate, a seminal vesicle or a vas deferens.

Iatrogenic incontinence
Internal urethrotomy (Otis) has been a standard procedure for recurrent urinary tract infections and reflux until the late 80’s. Fortunately, nature has been mild in most cases. However, incontinence based on urethral scarring does occur and is difficult to treat, usually by complete reconstruction of the urethra after excision of the complete scar. Reimplantation of ectopic ureters can be rarely complicated by vesico-
vaginal fistulas in less experienced hands. Ectopic ureteroceles can be accompanied by incontinence: unfortunately, incontinence is seldom included as a criterium in assessing the results of ureterocele therapy, and because of that we do not know the prevalence. Incontinence can occur after intersex surgery, especially when a high ending vagina has been pulled through.
Literature

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