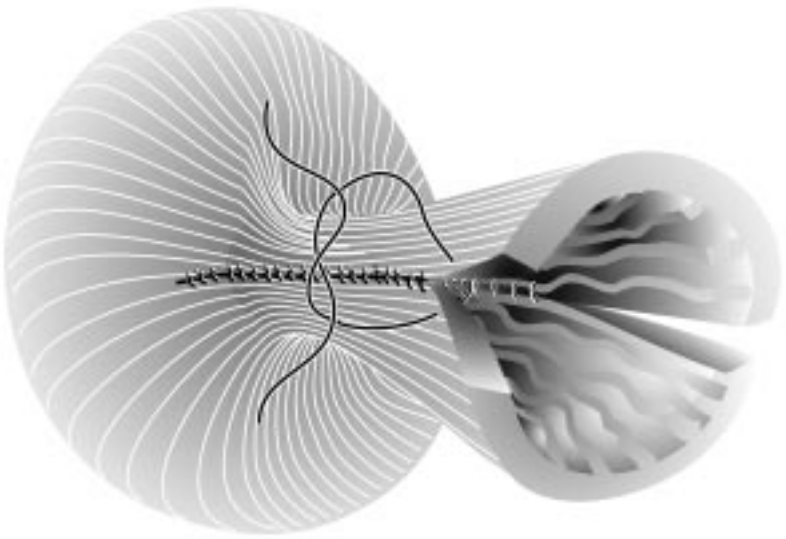
SURGICAL ASPECTS OF PEDIATRIC URINARY INCONTINENCE



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SURGICAL ASPECTS OF PEDIATRIC URINARY INCONTINENCE

(met een samenvatting in het Nederlands)

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Dit proefschrift is opgedragen aan de ouders van de geopereerde kinderen die steeds opnieuw het vertrouwen hebben moeten schenken om hun kind operaties te laten ondergaan waarvan het resultaat vooraf vaak onzeker is geweest. Zij zijn de helden in dit proefschrift.
Daarnaast vanzelfsprekend aan Anke, Jet, Jelle en Simone die dit alles met minimaal mopperen hebben laten gebeuren.

Contents

Introduction

Incontinence at pediatric age is a problem that can harm the psychological and physical development of children. Starting in 1986 we have searched for better solutions to treat incontinent children, by medication as well as by training and surgery. Important questions have been what surgical or non-surgical solutions do exist and what are the possibilities to improve the current techniques? This thesis is a synopsis of our publications on this theme.

The seven Chapters in this thesis deal with structural incontinence in children caused by congenital or acquired malformations of the urinary tract. Chapters 1, 2, 3 and 4 describe the sequelae of anomalous development

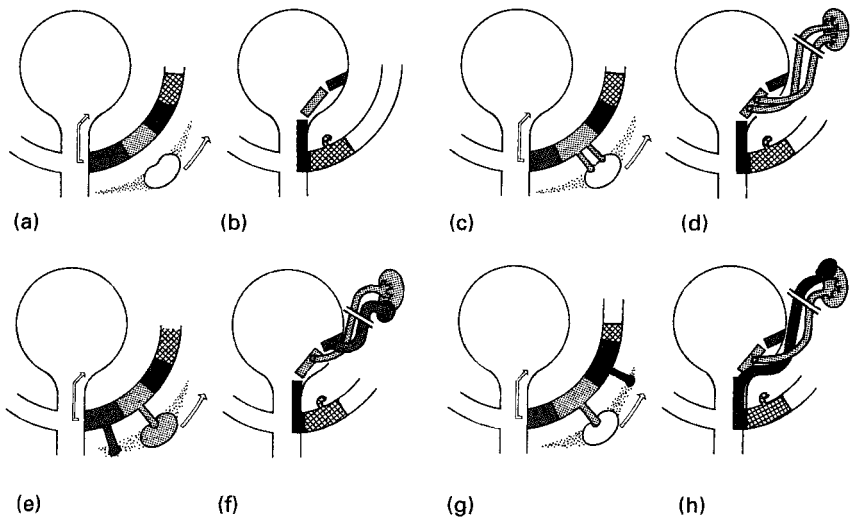

Figure 1. A. *Wolffian duct zones are shown in relation to nephrogenic area. Note direction of migration of kidney cranially and Wolffian duct into developing bladder.* ***B.*** *Migration of Wolffian duct zones into the bladder is completed.* ***C*** *and* ***D*** *show the development of a normal functioning double system with orthotopic ureteral orifices when double ureteral buds arising from normal Wolffian zone strike nephrogenic area and migration occurs in direction of arrows.* ***E.*** *Orthotopic bud arises from normal zone and strikes normal nephrogenic area. Ectopic bud strikes diminished tissue and migration occurs.* ***F.*** *After migration, the lower pole ureter is too lateral resulting in reflux and/or lower pole dysplasia.* ***G.*** *Ectopic bud arises too far laterally on Wolffian duct and strikes abnormal nephrogenic area.* ***H.*** *The ectopic upper pole ureter ends in the caudal zone and the upper pole of the kidney will be dysplastic. (with permission by the Williams and Wilkins Co.)*

of Wolffian ducts. Chapter 5 delineates the concept of structural incontinence in children born with ambiguous genitalia and a patent urogenital sinus. Chapters 6 and 7 present new insights in the treatment of children that have been born with bladder extrophy and epispadias.

Anomalous development of Wolffian and Müllerian ducts

During the first weeks of gestation, the interactions between the Wolffian and Müllerian ducts, the urogenital sinus and the metanephros define the functional and anatomical outcome of the urinary tract, from the top of the kidney to the tip of the urethra in females, and to the end of the prostatic urethra in males.

The Wolffian duct delivers material that develops into ureter, trigone, bladder neck and urethra. For a normally functioning urinary tract the Wolffian duct must deliver one or two ureteral buds at the right place to the core of the metanephros, in order to develop into a functioning kidney and collecting system. When the ureteral bud develops from a site on the Wolffian duct that prohibits contact with the optimal part of the metanephros, a dysplastic kidney or a dysplastic moiety of a double system will result. Moreover, ectopic delivery of the ureteral bud from the Wolffian duct results in an ectopic site for the ureteral orifice. This ectopic site can be located either cranially or caudally of the orthotopic position. A cranial ectopic position of the ureteric orifice results in vesicoureteral reflux, in severe cases combined with renal dysplasia. A caudal ectopic position of the ureteric orifice, below the level of the bladder neck, may result in dribbling incontinence and dysplasia of the corresponding renal moiety. An ectopic ureter may be accompanied by a congenital gap in the bladder neck, leading to bladder outlet insufficiency.

The clinical result of this combination of congenital malformations can be, simultaneously, dribbling incontinence, true congenital stress incontinence, and non-neuropathic bladder/sphincter dysfunction due to the constant presence of urine in the proximal urethra[1]. In conclusion, a small derailment in the embryological development of the ureteral bud and the Wolffian duct is able to cause dysplastic changes of the kidney as well as anatomical and functional anomalies of the trigone, bladder neck and urethra. This mechanism has been described superbly by Mackie and Douglas Stephens[2] (Figure 1).

chapter 1

In Chapter 1, the types of structural incontinence based on anomalous development of the ureteral bud and the Wolffian duct are described, juxtaposed with the standard-textbook-causes of structural incontinence in children. A complete list of structural causes is mandatory in the management of incontinent children, and true congenital stress incontinence should not be left out.

chapter 2

Patients with lower urinary tract symptoms based on anomalous development of the ureteral bud and the Wolffian duct will generally present with symptoms that fit into the category of non-neuropathic bladder/sphincter dysfunction, the clinical signs of which are symptoms of urge syndrome or dysfunctional voiding. In most patients, suspicion of structural anomalies arises only when treatment with a well-structured bladder training program has been proven a failure. Even in the case where a structural anomaly of bladder neck or urethral sphincter is the principal suspect, the first step in the treatment will often be a try-out with a training program, to prove that the condition is resistant to conservative treatment. This implies that the availability of a structured training program for the treatment of non-neuropathic bladder/sphincter dysfunction is a conditio sine qua non for departments dealing with structural pediatric incontinence. Without specific training experience the patients will be subjected to undue long conservative treatment plans that conceal the underlying surgical condition. In an editorial comment in the Journal of Urology the statement is made that a urotherapist needs to have the anatomical and physiological insight of a pediatric urologist, the patience of a pediatrician, and the tact of a psychologist[3]. Chapter 2 reviews non-neuropathic bladder/sphincter dysfunction of which urge syndrome and dysfunctional voiding are the main elements. The review is helpful to clarify the distinction between functional and structural incontinence. The review also delineates the therapeutical approaches that need to be followed in all patients, before deciding on surgery.

chapter 3
Patients with ectopic ureteroceles, not detected by prenatal ultrasonography, are presented with recurrent urinary tract infections, dysfunctional voiding and incontinence. Much discussion exists on two major items. Firstly, the question whether one should opt for primary complete reconstruction, or try and treat these patients with minimal surgery, implying a multi-stage approach with endoscopic incision of the ureterocele, with or without removal of the upper moiety of the affected kidney, possibly followed by lower urinary tract surgery. Secondly, the question whether major bladder surgery in neonates can be justified, in view of the long-standing (non-evidenced) belief that such surgery in the first year of life can seriously impair future bladder function. Backed up by urodynamic expertise and because of disappointing results with patients treated with a staged approach, we have opted for a one-stage complete reconstruction in all of our patients, regardless of age.

chapter 4
Historically, patients with vesicoureteral reflux and recurrent urinary tract infections have been treated by blind internal Otis-urethrotomy or urethral dilatation. This treatment was based on the assumption that the so-called spinning-top urethra, as visualised with the voiding cystourethrography used to detect reflux, represents a urethral obstruction. Urodynamics in children with urge syndrome or with dysfunctional voiding has taught us that the spinning-top urethra represents a functional anomaly, caused by simultaneous contraction of the detrusor muscle and the pelvic floor muscles during voiding or during filling of the bladder. As a result of a wide-spread urethrotomy policy we have been confronted with a series of girls with severe iatrogenic stress incontinence, combined with symptoms of the primary disorder: urge syndrome or dysfunctional voiding. In Chapter 4 we describe the difficult quest for a safe and definitive solution of the incontinence in these girls.

chapter 5
Both chromosomal anomalies and endrocrinological disorders can produce ambiguous genitalia with a patent urogenital sinus. Whenever female gender is assigned to these children, both a genitoplasty and a vaginal pull-

through operation needs to be done. Historically, staged procedures and delayed reconstructions have been performed, in many cases with lower urinary tract dysfunction as a result. These lower urinary tract symptoms are based on urinary retention in the vagina and on urethral insufficiency due to the shortness of urethra and/or a bladder neck defect. Clinically, the lower urinary tract symptoms will fit into the category of non-neuropathic bladder/sphincter dysfunction.
Next, the psychological impact of major genital surgery at a later age is an important factor in gender identity disturbances, as well as a possible confounder in the treatment of the bladder/sphincter dysfunction. In search for a way to avoid the lower urinary tract complications and to minimise psychological disturbances for the children and parents, we ended up with a one-stage procedure at neonatal age as the best possible option.

chapters 6 & 7
Patients with bladder extrophy/epispadias are generally presented as a neonatal emergency. Girls with epispadias are sometimes misdiagnosed as intersex patients. Major congenital anomalies such as the bladder extrophy/epispadias complex force us to constantly pursue an optimal result in terms of the number of operations, the time of hospitalisation, the urinary tract morbidity, the preservation of renal function, the rate of continence and the quality of life for the patient. The complex problem of female epispadias (girls born without a urethra and without a sphincteric mechanism) has also raised the question how to diminish morbidity and reduce the number of operative procedures. This question eventually prompted us to design a one-stage operative procedure that shows promise when compared with existing approaches. In Chapter 6, an overview of the treatment of bladder extrophy/epispadias is presented, and in Chapter 7 the results of our technique for female epispadias repair are described.

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Structural Incontinence

Structural incontinence in childhood

CHAPTER 1

Tom P.V.M. de Jong, William E. Kaplan, Miguel Podesta

In: Incontinence, Abrams P, Khoury S, Wein AJ (Eds), Paris, Health Publication; 1999

Introduction

Many structural anatomic causes for incontinence exist at pediatric age. Several, such as the exstrophy-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 derivates 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 ureteroceles-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 syringocele.[25] 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.[26-32]
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 occurr 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.[33, 34] We now know that this aspect is caused by a functional obstruction in the majority of cases.[35-47] 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

hymen.[48-51] 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 abdominal fissura 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.[52-58] 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.

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Functional Incontinence

Enuresis versus incontinence, diagnosis and treatment of the dysfunctional voiding sequence in children

CHAPTER 2

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Abstract

Incontinence in childhood, especially in girls, is predominantly based on non-neurogenic bladder/sphincter dysfunction, the so-called dysfunctional voiding sequence. Within this sequence three voiding patterns that follow each other closely can be recognised. The sequence starts with the urge syndrome and urge incontinence. The patients are frequent voiders, empty their bladder with a near normal flow on detrusor contraction and are wet on unstable detrusor contractions. The patients remain in this pattern or develop gradually the classical dysfunctional voiding pattern. These patients have a normal micturition frequency, empty their bladder straining with staccato or disrupted flow pattern and sustained detrusor action and are wet with unstable detrusor contractions or with overflow. Finally, patients can develop a true lazy bladder syndrome. These patients have a very low micturition frequency, empty their bladders straining with interrupted flow, without detrusor action, and are wet on overflow. The question whether one can be born with dysfunctional voiding or that it always follows some mishap such as a cystitis or urethritis due to infection or irritation is debatable. Both pathways appear to exist. The fact that the dysfunctional voiding sequence is accompanied by urinary tract infections in most cases and is complicated by vesico-ureteral reflux in 40% of cases, makes it into a serious disease that has to be recognised separately from urine loss due to behavioral and developmental problems that generally comes with a normal, uncompromised urinary tract.

Treating a child with dysfunctional voiding with behavioral therapy only is frustrating for the child and dangerous for the upper urinary tract. The authors have therefore adopted the strict rule to discriminate between enuresis, a complete emptying of the bladder at the wrong time by a child with a normal urinary tract, and incontinence that encapsules all other forms of involuntary urine loss. The English literature generally names all wetting children enuretics. Evidence from our clinical practice produces many arguments to discard this nomenclature and start to discriminate strictly between the diagnoses of incontinence and enuresis at pediatric age.

Introduction

In the treatment of children with involuntary urine loss we discriminate strictly between incontinence and enuresis. Enuresis is classified as a complete bladder emptying with normal urinary tract function due to behavioral immaturity. It occurs in 20% of our, selected, patient population and predominantly in boys. All other forms of urine loss in small quantities are called incontinence.
Functional incontinence due to the 'dysfunctional voiding sequence' in girls is the most common cause of involuntary daytime urine loss in the pediatric age group.[1,2,3,4,5,6,7] Rarely, dysfunctional voiding occurs in boys as well. Clinically dysfunctional voiding (DV) is characterised by complaints of urge and urge incontinence. The micturition pattern can develop from simple urge and urge incontinence into the dysfunctional voiding pattern, non-neurogenic bladder/sphincter dyssynergia, and eventually into the lazy bladder syndrome. The most important complications of DV are, in the majority of female cases, recurrent urinary tract infections (UTI), and in approximately 40% of all cases, vesicoureteral reflux (VUR). Thirty percent of the children have reflux nephropathy at referral.[8,9,10,11,12,13] Discussion exists whether DV can cause VUR or that DV can be the result of VUR. This question will probably never be resolved properly notwithstanding the fact that conservative treatment of reflux in a patient with DV will be unsuccessful as long as the DV persists.[14,15,16,17] Furthermore it is important to realise that several rare congenital and iatrogenic anomalies of the bladder neck and urethra can produce a pattern of incontinence that is clinically indistinguishable from classical DV.

Pathophysiology of dysfunctional voiding

The pathophysiology of DV has been clarified thanks to the pioneering work of Vincent, Allen, Koff, Hjalmas, van Gool, Vijverberg and few others.[9,13,18,19,20,21] The complex falls into three distinct micturition patterns that can progress from one onto the other. Firstly an urge syndrome and urge incontinence with normal micturition; secondly a bladder/sphincter

dyscoordination which begins with a staccato, and later a fractionated micturition. Finally, a lazy bladder syndrome with huge amounts of residual urine in an atonic bladder occurs. The *urge syndrome and urge-incontinence* arises when a child starts to postpone micturition, mostly during an episode of painful voiding due to UTI, vulvovaginitis, or urethritis. Simple problems like oxyuriasis or irritating detergents can be the primary reason for postponement of micturition in girls. The children hold back their micturition with active pelvic floor contractions, thus producing a functional urethral obstruction resulting in an overactive bladder with small capacity and high micturition frequency. Incontinence is based on high pressure detrusor instability. The patients have lost central inhibition of their bladder and use the pelvic floor as an emergency brake in the event of a detrusor contraction. In the typical urge syndrome urodynamic studies usually demonstrate normal pelvic floor relaxation while voiding. Small amounts of residual urine are the result of active pelvic floor contractions before the bladder is emptied. Some children maintain this voiding pattern for many years, others develop the *dysfunctional voiding pattern*. They learn over time to contract their pelvic floor effectively with more power, so that the bladder gives up most of its overactivity, and they have lost the ability to relax their pelvic floor properly when voiding. These children empty the bladder with near- normal frequency assisted by abdominal pressure with a typical staccato or fractionated flow. Incontinence is mostly based on overflow. Urodynamic studies in these children show a nearly normal or hypo-active detrusor with unstable contractions that can be inhibited easily, and a micturition phase with unsustained contractions. Non-neurogenic bladder/sphincter dyssynergia is remarkable. They have important amounts of residual urine after micturition. Eventually the dysfunctional micturition pattern can develop into the *lazy bladder syndrome* in which the detrusor muscle gives up completely. A decompensated, atonic bladder with very low micturition frequency is the result. The only driving force during micturition is the abdominal pressure. Incontinence is based on overflow with imperative urge at maximal capacity. Due to the pelvic floor overactivity these children develop enormous amounts of residual urine. Figures 1,2 and 3 give details on the urodynamic characteristics of these children.

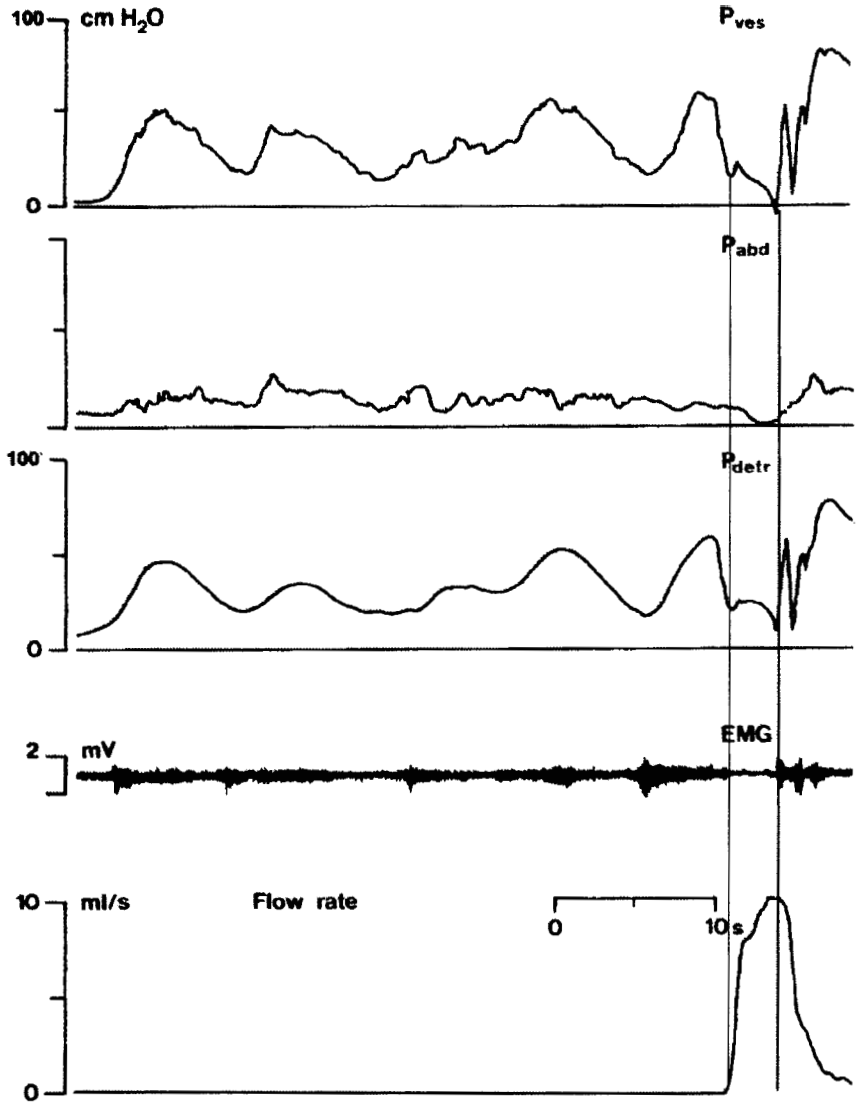

***Figure** 1: Typical urodynamic study of a child with urge syndrome and urge incontinence. Unstable detrusor contractions are countered with straining of the pelvic floor . When, during an unstable contraction the child gives up and relaxates the pelvic floor, the bladder is emptied with normal flow and normal relaxation of the pelvic floor. Note the drop in detrusor pressure that comes with opening the urethra.*

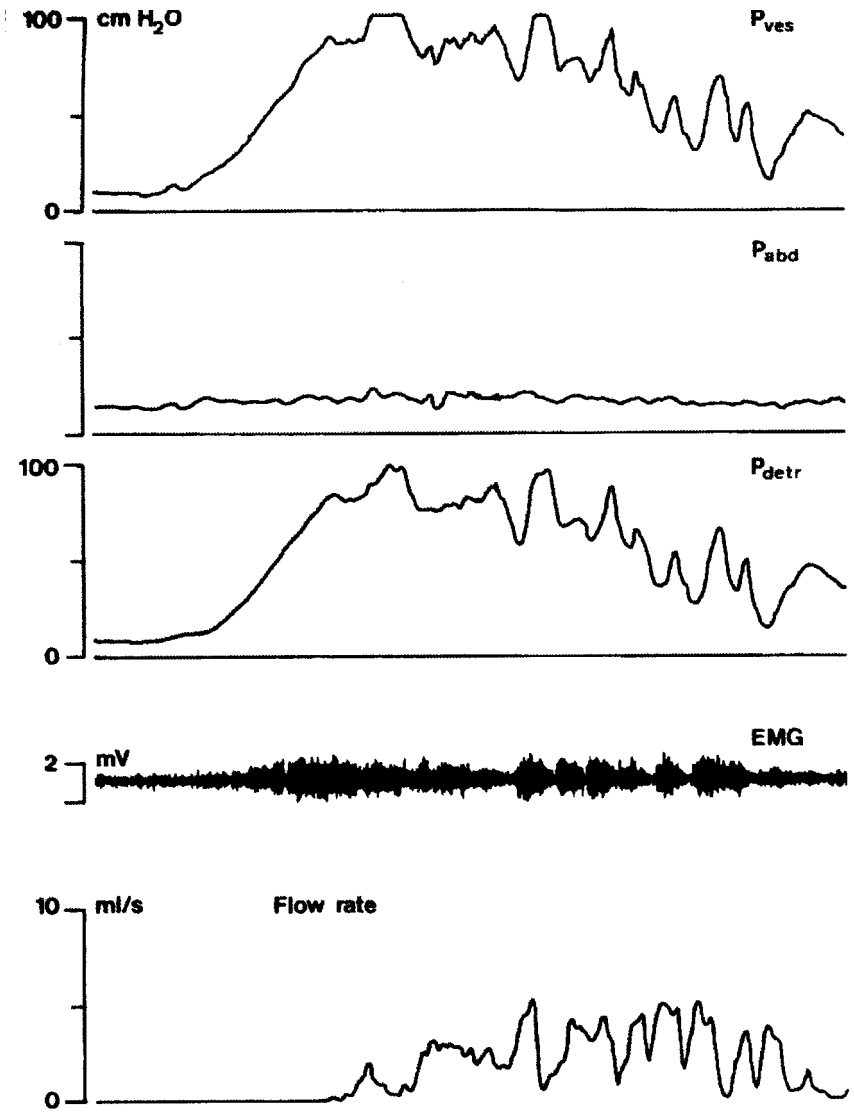

Figure 2: *Urodynamic study of a child with dysfunctional voiding. The bladder empties on a sustained detrusor contraction that is countered by the pelvic floor. With intermittent relaxation and contraction of the pelvic floor a staccato urinary stream is produced. Voiding will stop before the bladder is empty thus resulting in large residual volumes.*

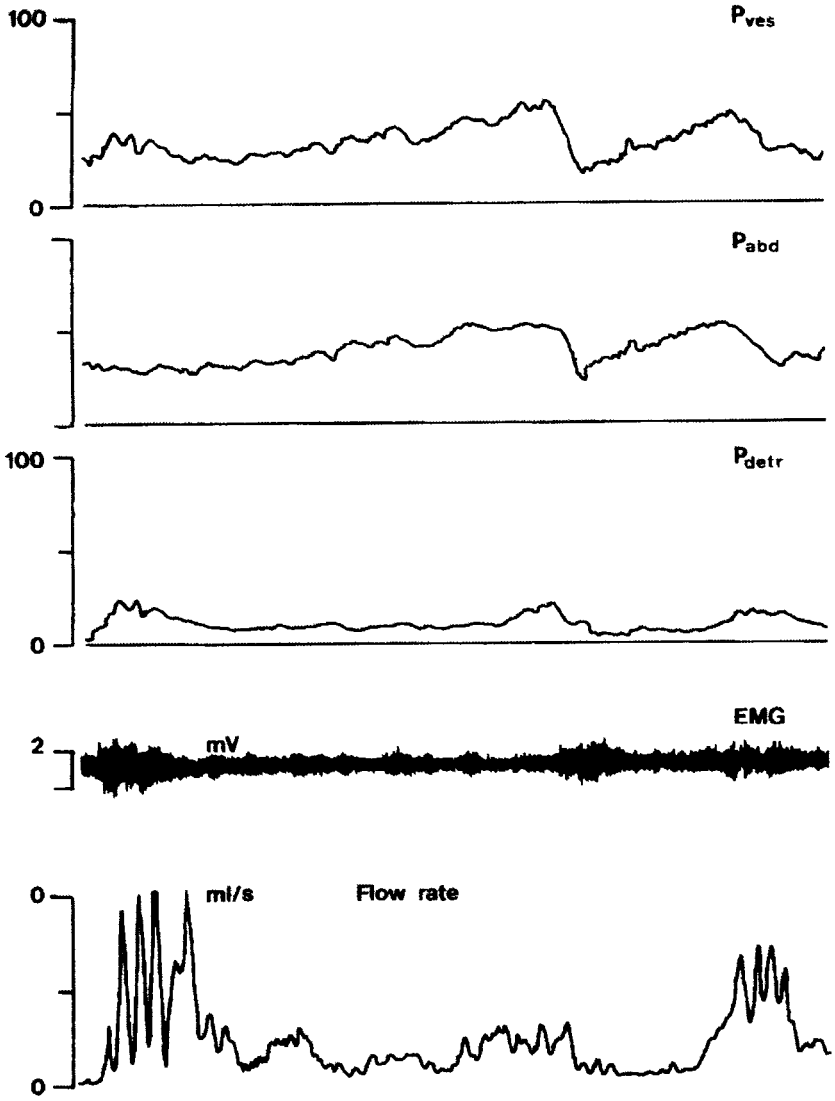

Figure 3: *Urodynamic study of a child with a lazy bladder syndrome. Note the absence of detrusor contraction. The only driving force to empty is the abdominal pressure. The pelvic floor does not relax. By straining the abdominal muscles the bladder is emptied in a fractionated flow with large residuals of urine.*

In addition to voiding dysfunction, nearly all children with micturition complaints due to the dysfunctional voiding complex have some form of fecal constipation. Clinically, in many cases, the fecal dysfunction only comes forward as skipping daily stools on a regular basis. The pelvic floor dysfunction hampers the rectum in the same manner as the bladder. Abdominal complaints connected to defecation are common in these girls. In addition, this problem often remains unrecognised by the parents in many cases. Only careful interview with specific questions will reveal the constipation

The patient's history

The characteristic patient with DV is a girl between age 3 and 12 years with frequency, urge and urge incontinence. In most cases this is combined with simple or complicated UTIs. Pathognomonic is some form of Vincent's curtsy sign: unstable detrusor contractions are felt as urge and suppressed by maximal contraction of the pelvic floor, when needed combined with compression of the perineum. This is accomplished by crossing their legs or by squatting or by sitting on a heel. When the unstable contraction has subsided and the interrupted daily action is continued they may have lost a small amount of urine in their pants.
Specific micturition history reveals a tendency to postpone voiding, to interrupt the stream (staccato stream or fractionated stream) and finally to strain when voiding. These patients are never enuretics: they never or nearly never empty their bladder completely in their pants. In nearly all cases they wet their pants more during the afternoon then in the morning, because after several hours of concentration on the bladder they tend to give up. Parents are often annoyed by the behaviour of their child: when squatting the children say they cannot go to the toilet, when the detrusor contraction has subsided children deny any urgency to go to the toilet. The child is right in both cases: standing up during the suppression of the unstable contraction would result in profuse wetting. When the contraction has subsided there is no further urgency to empty the bladder, because there is no relation between the amount of urine in the bladder

and unstable contractions. Clarifying this mechanism to the parents often works as an eye-opener. Finally, when a child has been successful in coping with unstable bladder contractions during a long period of time it can end with a decompensated bladder with low micturition frequency and large quantities of residual urine after voiding.
The majority of patients with dysfunctional voiding also wet during night. This does not always result in classical nocturnal enuresis. Often, it consists of wet spots in the underwear or bed during the night. Thus this should be classified as nocturnal incontinence instead of enuresis.
Apart from the micturition problems the majority of the patients are more or less constipated with or without fecal soiling.

The diagnosis

The preceding discussion automatically leads to the conclusion that the patient's history is the most important factor in coming to the proper diagnosis. Most of the efforts of the consulted urologist or pediatrician should therefore be spent on the micturition anamnesis. Extensive uroradiological and nuclear investigations are indicated when complicated UTI's have occurred. In most cases ultrasound of kidneys and bladder, uroflowmetry and determination of residual urine are sufficient at first. Urodynamic studies with good EMG recording are most typical in these patients (figure), but a thorough patient's history will allow the experienced investigator to draw the urodynamic study without actually doing it. The authors reserve urodynamic study for the patients that don't improve on a standard therapeutic scheme after 3 to 6 months. However, a major role for urodynamic studies in these patients remains. In rare cases only, an unsuspected urethral obstruction or a neurogenic component is detected by urodynamic study.

Psychology of incontinence

Children tend to cope with disability by ignoring the disabled part of their bodies. This natural defense mechanism is responsible for the fact that wet children are generally called enuretics, suggesting a behavioural ground for the wetting, thus resulting in incontinent children in educational circuits. The incontinent child will ignore the urgency of the bladder and wet pants, the enuretic child will deny these. The typical enuretic patient will answer no to a parent's question, shouldn't you empty your bladder, and then do a complete voiding within seconds or minutes afterwards. The incontinent patient will ignore the fact that pants gradually wet and smell and continue daily affairs as if nothing is wrong. The treating urologist or pediatrician must discriminate between these subtle differences. In general, one can state that incontinence as a sign of mental discomfort is very rare. A child in distress has many easier ways to express this discomfort besides wetting his/her pants. This makes the discrimination between true enuresis and incontinence a major priority in the treatment of wet children.
True daytime enuresis, complete emptying of the bladder in the pants in one run, occurs predominantly in boys. In girls there may be a behavioural factor in DV, but it remains indistinguishable in most cases. Thus treatment of DV should be straightforward along the following lines with an open eye for psychologic complications.

Treatment

Treatment of children with DV requires some form of cognitive bladder training after thorough education of the child and the parents on the underlying mechanism of the dysfunction. A comparison to a mouth full of water after tooth-brushing often elucidates the mechanism to the child. The water can be sprayed out with force by squeezing the cheeks and the lips both at the same time, this results in residual water in the mouth. The water in the mouth can also simply be evacuated by opening the lips so that it drops out. In the last case the evacuation will be complete.

In the out-patient clinic the child and parents are informed on measures to avoid urethral irritation: no soap or washing-glove for cleaning, shower bi-daily to rinse the genitalia, no irritating or perfumed bath-salts and no tight stretch-clothing. Avoid extra mechanical irritation by a narrow bicycle-saddle or improper gymnastic exercises. The child should be taught to maintain a voiding frequency of 6-7 times a day. Instruct the parents to monitor this, because many children detest voiding so much that they will stay in the bathroom for a few minutes, flush the toilet and return with the happy statement that they have emptied their bladder while in fact they have not even lowered their pants. Children with a large bladder capacity for age should learn that, lifelong, they have to empty their bladder when the time to do so has arrived instead of the urgency. The child should be taught to sit relaxed on the toilet with the thighs in a horizontal position, and when needed, the feet on a small stool or block. Make the child read one page of a comic-book when voiding and take care that the child does not strain during micturition. The easiest way to achieve this is by having them whistle a tune while voiding: it is very difficult to whistle and strain at the same time! Require a minimal daily fluid intake because many children with day-time incontinence stop drinking adequate volumes of fluid, thus producing urethral irritation by voiding concentrated urine. Dietary advice and adjustment to prevent constipation is essential. Finally have the child keep a calendar on micturition, incontinence and stools.

The above rules have to be supported by medication in the training period. Routinely prescribe chemoprophylaxis for several months to prevent infections. Once a day 2 mg/kg nitrofurantoin or trimethoprim will usually do the job. When frequency, urge and urge-incontinence are the most important symptoms and residual urine is minimal, spasmolytic support is needed in the form of oxybutinin 0,4 mg/kg up to three times a day. Oxybutinin works effectively for 6 hours. When major complaints occur only in the afternoon one dose of oxybutinin at noon will do. At the least suspicion of constipation, especially when spasmolytic therapy is needed, spasmolytics are postponed and laxatives are added to the treatment until daily stools are achieved. New spasmolytics have been developed, effect in these children has to be proven yet.

When supported and controlled properly this therapy will cure 30-50% of the patients in the out-patient clinic. The remaining hard-core group of patients will need a proper biofeedback training program to get rid of their symptoms.

Bio-feedback training

Biofeedback training can be done on an out-patient basis or during a period of hospitalisation.[21,22] Psychological determination of the maturity of the child and the motivation is important. In general, training before the age of 6 years, in girls, and 7 years, in boys, is disappointing because of lack of cognitive insight resulting in lack of motivation. The older the child the better the results. In our hospital we provide a training package of 10 days for the hard-core patients. Apart from psychological support they learn how, when and how often to void. They learn how to void by doing every micturition on a flowmeter with direct visualisation of the flow on a screen, thus eliminating interruptions of the stream with the pelvic floor. Bladder emptying is controlled by ultrasound. They learn when to void and to recognise bladder signals by wearing urine detector pants with an alarm, that rings when wet, during daytime. They must try to go to the toilet before the alarm rings. Finally they learn how often to void by making a chart during training. In general 80% of the hard-core patients obtain complete cure or significant improvement of incontinence and UTI's by this training schedule. This result remains permanent over 10 years follow-up in our patient group. After the training they have regained central inhibition of the bladder signals instead of using the pelvic floor as an 'urgency-brake'. After hospitalised training the children are followed up by the incontinence-therapists by weekly telephone contact for several months. When residual urine after micturition does not subside after training some children with a full-blown lazy bladder syndrome have to be treated by clean intermittent self-catheterisation.

The failed therapy

It is important to realize that dysfunctional voiding in a small group of patients can be a symptom of congenital bladder neck insufficiency as a result of embryological derangement of the Wolffian duct. Also, a prior generous internal urethrotomy at young age can produce a bladder neck insufficiency. In severe primary bilateral reflux, in single or bilateral ectopic ureters, in ectopic ureteroceles, in intersex cases with adrenogenital syndrome and a short urethra after reconstruction and in several other rare anomalies an insufficient bladder neck can force the child to try to withhold urine that leaks into the urethra with the pelvic floor. This can eventually result in a typical DV pattern that is resistant to training. Operative therapy is needed to cure these children. This operative therapy can consist of a simple colposuspension in uncomplicated cases. When severe reflux is coexisting with DV colposuspension must be considered during ureteral reimplantation. Needle suspension in the pediatric age group provides only temporary success. The authors sometimes use this procedure in difficult cases to judge the chances of a proper Burch type colposuspension. When a scarred bladder neck and urethra due to a previous internal urethrotomy appears to be the underlying cause of the incontinence simple colposuspension has been disappointing in our hands. Nowadays we excise the scar completely and reconstruct the urethra. Although very succesfull in a few cases it is too early to tell wether this is the best way to cure this problem.

Developments

An international prospectively controlled multicenter trial with financial support of the EEC has been conducted in Europe to determine which way of training for children with DV provides the best results without the exorbitant costs of hospitalisation. Surprisingly, the amount of attention given to the child appears to be the most important factor for success. We have started a new study to the treatment of DV with a controlled amount of attention and the addition of a new biofeedback device, a portable flowmeter and toilet that is used by the child at home every day.

Ultrasosonography of the pelvic floor and the rectum gives new insights in the pathophysiology and the treatment of DV. Recently we have proven that a direct correlation exists between the diameter of the rectum on ultrasound and the amount of fecal constipation. This has proven to be of help to convince both the parents and the children of the existence of constipation and it provides an easy, non-invasive control of therapy. Dynamic ultrasonography of the pelvic floor muscles has learned us that approximately 40% of the children with DV has a paradoxal movement of the musculature. They contract the puborectal muscles when ordered to relax and they relax the musculature when ordered to squeeze. Physical therapy to correct this paradoxal movement appears to have a 100% success rate. The effect of this correction on the results of DV therapy remains to be proven yet.

The authors do hope that after reading this paper on dysfunctional voiding therapists will recognize that the misused overactive pelvic floor is the primary problem in these patients and thus realise that the worst advice to give these patients is to do the 'stop-test' during micturition. This manoeuvre will only make the bladder derailment worse! Finally one must realise that these children are incontinent, they are not enuretics. Calling them enuretics delays cognitive treatment by inappropriate behavioural therapies and produces unneccesary trauma for the child.

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Ectopic Ureteroceles

Ectopic ureteroceles, results of open surgical therapy
in 40 patients

CHAPTER 3

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Cuno S.P.M. Uiterwaal and Jan D. van Gool

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Abstract

Purpose: The treatment policy of ectopic ureteroceles is controversial. Besides debate on the optimal treatment of ectopic ureteroceles, discussion exists whether there is an extra risk for deterioration of bladder function after extensive bladder surgery during the first year of life thus giving reason to postpone surgery. We have treated 40 patients with an ectopic ureterocele in a prospective, non-randomised trial, with complete surgical reconstruction of the lower urinary tract combined with upper pole resection of malfunctioning upper pole moieties at the time of referral, regardless of age. Three patients with only 1 affected renal moiety initially were excluded from the study.

Materials and methods: Forty patients, 31 female and 9 male, were treated for ectopic ureteroceles that extended into the bladder neck and urethra. Age at operation was between 0.0 and 8.8 years with a mean age of 2.17 years and 19 patients were under the age of 1 year at the time of operation. Of the patients, 37 underwent primary excision of the ureterocele with reconstruction of the urethra, bladder neck and trigone and with ureteral reimplants. In 3 patients, because of small size of the ureterocele, the ureterocele was left in situ, leading to secondary removal of the ureterocele in two, one because of obstructive voiding and one to treat urinary incontinence. In 5 neonates, a staged procedure was done with primary reconstruction of the lower urinary tract combined with upper pole cutaneous ureterostomies, followed by upper pole resection or ureteral reimplantation a few months later. In 16 patients, after bladder neck reconstruction, a colposuspension was performed as well to create a normal vesico-urethral angle. All patients had a clinical and urodynamic work-up at least 1.25 year after operation with a mean follow-up of 5.59 years. Patients who by Jan. 1999 were too young to have their continence clinically assessed were excluded from the study.

Results: All our patients are continent. Thirteen patients needed a secondary endoscopic procedure (cystoscopy only in 2, scar incision near ureteral orifice in 3, endoscopic reflux treatment in 4, resection of remnants of ureterocele in 2, bladder neck incision to treat obstructive voiding in 2), one patient needed a secondary open reconstruction of the bladder because of a diverticulum. Eleven patients had postoperative only one or

two uncomplicated urinary tract infection (UTI). Recurrent UTI's were seen in 4 patients, 1 complicated patient had severe infections leading to renal scarring. This patient had a pre-existing loss of renal function.
A normal voiding pattern is seen in 29 patients, 11 have a dysfunctional voiding type of micturition with recurrent UTI's in 5 cases. Urodynamic follow-up confirms these clinical findings. Bladder capacity in these patients is relatively high and averages 124% of expected capacity for age. There was no statistically significant difference in the follow-up parameters between children operated under the age of 1 year and those operated at a later age. The additional colposuspension in 16 patients did not produce any significant change in outcome compared to the patients that did not have this procedure.
Conclusion: Compared to the literature, complete primary reconstruction of the lower urinary tract in patients with ectopic ureteroceles appears to have better results then a staged approach with initial endoscopic treatment. Moreover, an important conclusion of this study is that no proof exists that extensive reconstructive surgery of the bladder in neonates and infants leads to deterioration of the bladder function at later age.

Introduction

In the treatment of ureteroceles, strict discrimination between intravesical and ectopic ureteroceles is mandatory. In contrast to ectopic ureteroceles, intravesical ureteroceles rarely have consequences for the urodynamic behaviour of the bladder and can be treated as seems best for each individual patient by endoscopic incision or by open removal with reimplantation of the affected ureters. The choice of therapy in intravesical ureteroceles is determined by the size of the ureterocele, by the function and degree of obstruction of the affected renal unit and by the presence or absence of vesicoureteral reflux (VUR).[1-6]
Ectopic ureteroceles, in general, can have consequences for the urodynamic properties of the bladder[7] and have a high morbidity for urinary tract infections (UTI), loss of renal function and incontinence. In contrast to intravesical ureteroceles, ectopic ureteroceles, in general, drain the

upper renal moiety of a double system, which shows a severely compromised function.[8] Literature review of ectopic ureteroceles reveals a tendency to start with upper pole removal or in the newborn with endoscopic incision of the affected unit , although in the last years more aggressive therapy for ectopic ureteroceles with reflux in 1 or more moieties is advocated.[5] In many reviews the authors were forced to do a complete removal of the ureterocele as a secondary procedure. VUR and recurrent UTI's formed the most important reasons to do so. As early as 1979 Hendren, for ectopic ureteroceles, proposed to do a complete reconstruction of the lower urinary tract with upper pole removal. He concluded that this type of surgery is not suitable for neonates and infants .[9] In addition, a non-evidenced concern exists that extensive lower tract surgery in infancy produces higher urodynamic morbidity compared to surgery at a later age. With advances in pediatric anesthesiology, nowadays duration of surgery hardly presents an extra risk or contraindication in deciding the time of reconstruction in neonatal cases. Because of our previous experiences with staged procedures that had a high morbidity for UTI's and incontinence, we began to search for an approach that is definitive and safe. Therefore, we have treated our patients with ectopic ureteroceles by primary, complete surgical reconstruction from 1989 on as a prospective, non-randomised study. All the patients were followed with respect to clinical outcome, urodynamic behaviour of the bladder, number of infections and renal function. In addition, we have compared the urodynamical and clinical outcome of patients operated before and after the age of one year.

Materials and methods

Between 1988 and 1998, 40 patients with 41, previously untreated ectopic ureteroceles presented to our hospital. Distribution of sexes was 31 female and 9 male patients.
In the 40 patients, 24 ureteroceles were on the left side, 15 on the right, 1 bilateral. Age at operation ranged from 0 to 8.84 years with a mean age of 2.17 years. Nineteen patients were less then 1 year old at the time of oper-

ation with a mean age of 0,22 years. Eleven cases were detected by prenatal ultrasound, 2 in evaluating incontinence and 27 by urosepsis or recurrent UTI's. Of these patients 10 were straining while voiding.
Nine patients presenting with life-threatening septicaemia were primarily treated by drainage of the affected renal units by endoscopic incision of the ureterocele and/or a nephrostomy drain in the distended upper pole. One infant with life-threatening urinary ascites and pleuritis in bilateral ectopic ureteroceles was first treated by a vesicostomy with intubation of the 4 ureters combined with evacuation of the urine from the abdominal and thoracic cavities. His lower urinary tract was reconstructed in a secondary procedure and included bladder augmentation with both upper pole ureters.
Excision of the ectopic ureteroceles with reconstruction of the lower urinary tract was performed as a primary procedure in 28 patients, as a secondary procedure, within a few weeks after presentation, in the aforementioned 10 patients. In five neonates reconstruction of the lower urinary tract was combined with the installation of upper pole ureterostomies followed after 2 months by upper pole resection in 3 cases, upper pole ureter reimplantation in 2, after establishing the function of the affected upper pole moieties. Reason for this delayed upper tract repair was the fact that no information on the function of the upper pole moieties existed at the time of the first procedure.
Reimplantation of all the refluxing contralateral ureters and of all the remaining lower pole ureters was performed after ureterocele resection. In cases of megaureters, tapering by plication was performed when the flat width of the ureter exceeded 10 mm during the first 6 years of the study, 6 mm from 1994 on. When, after reconstruction of the urethra, bladder neck and bladder base a vesico-urethral angle was considered non-existing, a Burch-type colposuspension was performed by the end of the procedure.
Preoperative diagnostics consisted of ultrasonography (US) and voiding cystourethrography (VCUG) in all the patients, DMSA-scanning in 23 patients and urodynamic studies in 10 patients. In all cases, the operative procedure started with cystoscopy and colposcopy. The differential diagnosis intravesical/ectopic ureterocele could not always be determined by US and VCUG. In approximately 25% of the cases only cystoscopy could

exactly determine the caudal extension of the ureterocele and discriminate between ectopic and intravesical ureterocele. A DMSA scan was not performed in neonatal cases and in cases that, on ultrasound, showed a very small and scarred upper pole moiety of the kidney in the presence of a normal looking lower pole.

Operative approach

Heminephroureterectomy was performed by lumbotomy with selective ligation of upper pole vessels. Nephropexia was done only when the whole of the lower pole needed to be mobilised. The reconstruction of the lower urinary tract was carried out by a low transverse Pfannenstiel incision with transverse opening of the fascia and splitting of the rectus muscles in the midline. The adventitial layer of the bladder is opened selectively, the bladder is opened in the midline, and the bladder dome is filled to capacity with 1 or more gauzes and stretched with the third blade of the wound retractor. The ureterocele is identified and grabbed in a clamp, the bladder mucosa around the ureterocele is incised with a diathermic needle and the ureterocele is peeled out, partly by blunt dissection, partly by cautery. The part of the ureterocele that runs through the bladder neck into the urethra is freed from mucosal lining as far as is necessary and possible. Surprisingly, even in a neonatal female urethra, one can reach down into the urethra for approximately 1.5 centimetres in cases of large cecoureteroceles. In cases of cecoureteroceles the whole urethral part of the ureterocele is incised endoscopically at the start of the procedure. Manipulation of the urethra is made easier when a small additional suction tube is placed retrograde into the distal urethra and the elevation of the anterior wall of the urethra can be optimised by putting an 8 Fr. silicon drain through the urethra that is subsequently tied around the symphysis. The upper pole ureter is excised, and, when necessary, the distal portion of the lower pole ureter. The lower pole ureter passes into the bladder cranially in the large bladder defect that is the result of this procedure. Starting as deeply as possible in the urethra, the dorsal urethral wall is closed with polyglycolic 5.0 interrupted sutures. Care is taken to grab

the urethral wall in such a way that the suture is positioned submucosally with the knot outside. In general, 3-5 sutures can be put into the urethra and the bladder neck. The defect of the bladder base is closed similarly with 4.0 sutures, leaving room in the cranial part of the defect for the passage of the lower pole ureter. The lower pole ureter ending is fixed in the physiological position by 2 5.0 polyglycolic sutures and covered with bladder mucosa.

When in cases of very large or bilateral ureteroceles this procedure appears to lead to a narrow, funnel-shaped trigonal part of the bladder, it may be necessary to bring the cranial rim of the defect down to the level of the bladder neck, resulting in a remodeling of the bladder. When after this procedure, macroscopically no vesico-urethral angle can be seen, a colposuspension is thought to be appropriate to restore a normal vesico-urethral angle. This is performed by lifting the anterior vaginal wall left and right of the bladder neck with a metal sound through the introitus and grabbing it into a clamp. Depending on the size and age of the child the anterior vaginal wall is lifted to Cooper's ligament with a 2.0 or 3.0 polyglycolic acid suture. The reimplanted ureters are splinted with 4-6 Fr. feeding tubes, a 12 Fr. silicon Malecot-type catheter is left as a suprapubic catheter. A 10 Fr. silicon drain is left transurethrally and fixed to the bladder wall by a 5.0 rapid soluble polyglycolic acid suture. The bladder is closed with 4.0 polyglycolic acid sutures, the adventitional layer is closed separately (figure)

The ureteral splints are taken out after 4 days; after 7 days in ureters that have been tapered. Suprapubic and transurethral drainage is maintained for 6-9 postoperative days. All patients were on chemoprophylaxis for at least 6 months after the operation.

A VCUG and a urodynamic study were done in all patients postoperatively. All patients repeatedly underwent US of the upper tracts and the bladder combined with uroflowmetry and US determination of post-void residual urine

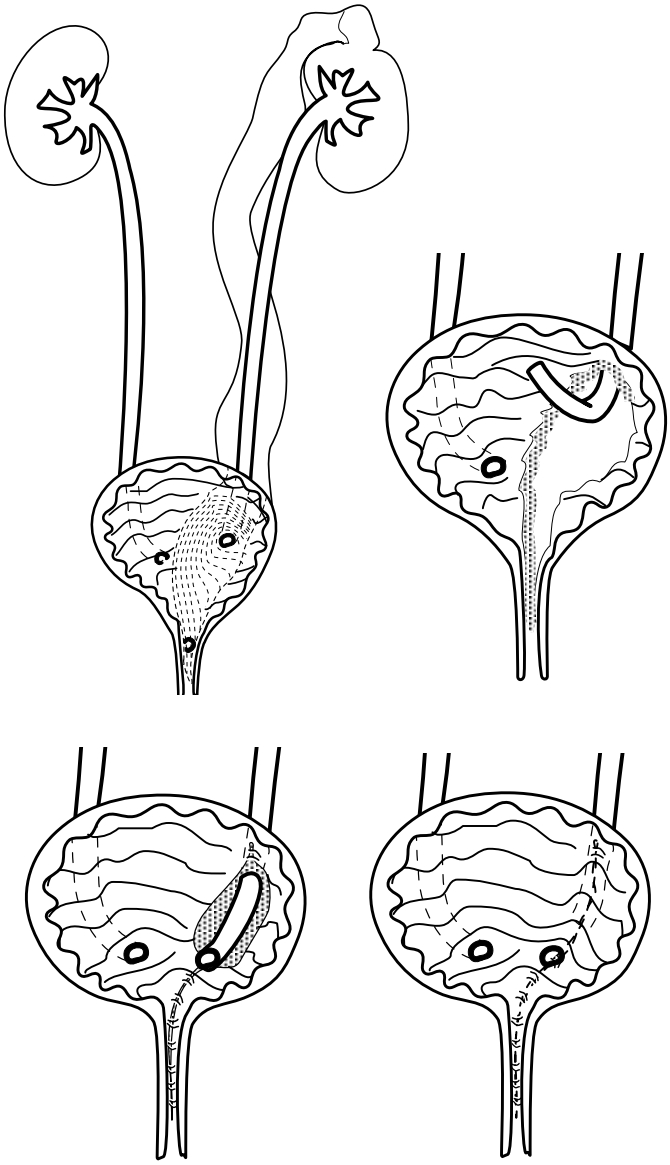

Figure: *Schematic drawing of a left double system with an ectopic ureterocele opening in the proximal urethra. The lower pole ureteric orifice is on the proximal part of the ureterocele. After upper pole nephrectomy and excision of the cele, a large defect runs from the bladder into the urethra. Closure of the defect results in a reconstruction of the urethra, the bladder neck and the trigone. After closure of the defect the lower pole ureter is fixed in the physiological position running over a part of the closed defect and covered by mucosa.*

Results

The opening of the ectopic ureteroceles in the female patients was situated in the bladder neck(5), proximal urethra (9), distal urethra (5), and vulva (11) and in the 9 males in the prostatic urethra. Ipsilateral reflux was present in 15 patients, ipsilateral and contralateral reflux in 9. A contralateral double system was present in 11 patients, 7 with ectopic ending of the upper pole ureter in the bladder neck, 1 ending in the urethra, 1 in a utricular cyst. Two patients had a single ectopic ureterocele draining a malfunctioning, non-doubled kidney. Two neonatal girls had a distal vaginal atresia that was corrected by transtrigonal approach in one and by perineal approach in the other during the same procedure. In 3 patients, first treatment consisted of upper pole resection alone, 2 because of the small size of the ureterocele in the absence of reflux, 1 because of the impression that the upper pole ureter ended in a huge Gartner's cyst and not in a ureterocele. Of these 3, 1 girl still has the ureterocele in situ and is free of complaints, 1 boy had the ureterocele removed 2 years later because of obstructed voiding with prolapse of the ureterocele into the urethra, 1 girl had the cele removed for treatment of incontinence after it became clear that she hadn't a Gartner's duct cyst but rather an ectopic ureterocele.
In one girl, the ureterocele was unroofed because of the wrong impression on US and during operation that sufficient bladder wall backing of the ureterocele was present. This patient developed a large bladder diverticulum that needed secondary open removal with bladder neck and bladder base reconstruction. In total, 39 patients had the ureterocele excised with reconstruction of the lower urinary tract.
Secondary procedures related to the reconstruction of the lower urinary tract were needed in 13 cases, 9 because of persisting reflux, and 4 because of obstructive voiding. These were all day care surgery endoscopic procedures and consisted of cystoscopy alone in 2 cases, incision of scar tissue at or near a ureteral orifice that pulled the orifice wide open in 3 cases, endoscopic treatment of persisting reflux in 4 cases. Obstruction therapy consisted of endoscopic resection of distal urethral remnants of the ureterocele in 2 cases and superficial bladder neck incision for treatment of obstructive voiding in 2 cases. Of the 5 infants, primarily treated by reconstruction of the lower urinary tract combined with upper pole

ureterostomies, 2 upper pole moieties surprisingly appeared to have a good function, so that secondary upper pole ureter reimplantation instead of upper pole resection was decided on.
Secondary nephrectomy of malfunctioning lower pole kidneys was performed in 2 cases, 1 because of hypertension, one because of the finding of nephroblastomatosis in the upper pole that, upon pathologic examination, appeared not to be present in the lower pole.
All patients were routinely followed at the outpatient clinic. Two patients were lost to follow-up 2,5 and 3 years after operation due to relocation abroad.
Follow-up ranges from 1,5 to 9,85 years with a mean of 5,59 years.
Clinically, all our patients have developed normal urinary continence. The vast majority has a tendency to void infrequently and many patients need to be reminded by the parents to maintain a normal micturition frequency. Five patients needed a biofeedback training programme for treatment of non-neurogenic bladder/sphincter dysfunction. Postoperative urinary tract infections did not occur in 24 patients and were a temporary problem without upper tract infections in 11 patients. Four patients are chronically on chemoprophylaxis because of recurrent infections. In only one patient with initially a very bad bladder and bilateral upper tract problems with preexisting loss of renal function, pyelonephritis 2 years postoperatively appears to have caused extra renal damage. Postoperative urodynamic studies show a normal bladder compliance in 100% of the patients. Bladder capacity as a percentage of normal bladder capacity for age (30+30 x age = ml)[10] ranges from 95% to 200% with a mean of 124%. Mild detrusor overactivity is seen in 5 patients. Micturition pressures are within normal ranges with a mean pressure of 39 cm water pressure. Only 7 patients have post-void residual urine from 5-40% of the capacity for age. Three patients temporarily have been on CIC for 10-48 months postoperatively because of residual urine and UTI's. Ten patients had a preoperative urodynamic study that showed a large average bladder capacity, elevated end-filling pressures in 4/10, detrusor instability in 8/10.
The results of the 16 patients that underwent a Burch type colposuspension as part of the reconstruction were compared to the other patients regarding number of UTI's, bladder capacity, micturition pressure, post-void residual volume, number of voidings per day and reoperation rate.

No significant difference between the 2 groups could be established.
A similar comparison was done between the 19 patients, operated under the age of 12 months (mean age 2,6 months) and the 21 patients operated at a later age (mean age 3,9 years). Again, no difference in outcome between the 2 groups could be established.
In 38 pathology reports, there was a diagnosis of kidney dysplasia in 22 cases, changes due to pyelonephritis in 11 and both dysplasia and pyelonephritis in 5.
Although many patients initially presented with compromised renal function based on septicemia, obstruction or both, at follow-up only one patient appears to have a permanent loss of renal function with diminished creatinin clearance 9 years after operation.
The results are summarised in the table.

Table: *Summary of the key-data from the results section.*

	Age at operation	
	Below 1 year	**Above 1 year**
Gender (% females/males)	79/21	76/24
Mean age at operation (years)	0.2	3.9
Follow-up (years)	5.7	5.4
Ureterocele (%left/right)	68.4/31.6	61.9/42.9
Ectopic ending of ureterocele (%)		
Bladder neck	15.8	14.3
Proximal urethra	21.1	19.0
Distal urethra	10.5	14.3
Vulva	31.6	28.6
Prostatic urethra	21.1	23.8
Ipsilateral VUR (%)	31.6	47.6
Contralateral VUR (%)	31.6	14.3
Contralateral double system (%)	42.1	14.3
Ending of contralateral upper ureter (%)		
Orthotopic	68.4	85.7
Bladder neck	21.1	14.3
Urethra	5.3	0
Utricular cyst	5.3	0
Burch colposuspension (%)	33.3	68.8
Contralateral reimplant (%)	52.0	48.0
Postoperative urodynamics		
Bladder capacity (% for age (SD))	140.3 (28.8)	124.2 (33.4)
Residual (%)	6.4 (12.6)	7.6 (18.1)
Micturition pressure (cm H_2O)	52.1 (35.1)	42.9 (18.1)

Discussion

The appropriate protocol for the management of ectopic ureteroceles is controversial. Especially endoscopic treatment alone but also upper pole nephrectomy alone carry a high reoperation rate, as reported in literature between 25% and 100%, certainly when reflux into 1 or more moieties is present.[1-6, 8,9]

These facts, combined with our own experiences concerning morbidity and reoperation rate in children with staged procedures, have determined our choice for primary and complete surgical reconstruction of both the upper and the lower urinary tract. Morbidity in our patient group is relatively low, compared to the literature. Thirty-two percent of our patients needed an endoscopic secondary procedure, only 1 of our patients needed a secondary open procedure due to our own misinterpretation of the bladder wall backing of the ureterocele during the first operation. Urodynamically, the results of our group of patients appears to be slightly better then reported series of patients that did not have bladder neck surgery. Abrahamsson e.a. report infrequent voiding in 59% of their patients, a bladder capacity of more then 150% of capacity for age in 55% of their patients and a significant residual after voiding in 56% . In their series of 34 patients 2 were on chronic clean intermittent catheterisation.[7] Unfortunately, most studies on ectopic ureteroceles do not include urinary continence as an outcome factor in the treatment of ectopic ureteroceles while, in most studies, patients with incontinence are mentioned.

One can discuss whether a standard policy of surgical intervention on both the upper and the lower urinary tracts should be considered as overtreatment in the small group of patients with no involvement of other renal moieties than the unit draining the ureterocele. Of the 3 patients that we primarily treated by upper pole nephrectomy alone, 2 needed additional surgery on the ureterocele, 1 for obstructed voiding, and 1 for treatment of incontinence. Our series proves that reconstructive surgery of the bladder performed on neonates and infants does not result in urodynamic deterioration of the bladder. There is no proof that the addition of a colposuspension to restore a normal vesico-urethral angle has any positive effect on the urodynamic outcome although no urodynamic harmful effect is seen. The reason to perform such a colposuspen-

sion has been the idea to create a platform for subsequent bladder neck injection with a bulking agent in cases of incontinence. Since no indication for bladder neck injection has come out of this group of patients one may conclude that the colposuspension is a superfluous extra and should be left out of the procedure. However, there are no means to predict whether more incontinence will occurr when a flat vesico-urethral angle is left untreated.

Conclusion

The primary surgical approach to the complex problem of the ectopic ureterocele seems to be a worthwhile choice. Extensive surgery of the neonatal bladder does not produce detectable anomalies in urodynamic or clinical behaviour of the bladder, compared to patients operated at a later age.

Editorial comment by dr. Ricardo Gonzalez

The authors are to be commended for presenting a detailed analysis of the outcome of complete excision and reconstruction in 40 children with extravesical ureteroceles. The documentation that the urodynamic outcomes and continence are not different when the operation is done before or after 1 year of age is important and agrees with my own experience. That the authors excised all ureteroceles in this group of children suggests that they do so regardless of the absence of preoperative reflux. Others have reported that when reflux is not present, partial nephrectomy alone is curative in 80% of patients (references 5 and 6 of the paper). Thus, a presumably large number of children in the present report may have been subjected to unnecessarily extensive bladder surgery. I also disagree with the policy of incising endoscopically ureteroceles extending into the urethra prior to excision to facilitate it. In my experience removal of an intact ureterocele is easier and more accurate and this is one of the reasons why I am opposed to the routine puncture or incision of ureteroceles when the procedure is not expected to be curative.

Perhaps, the need to re-operate to resect residual ureterocele tissue might have been avoided if the real extent of the ureterocele had not been masked by the preliminary incision. Also, the use of a preliminary upper pole ureterostomy seems unwarranted. Recovery of function is not to be expected as a rule and, removal of a poorly functioning upper pole does not cause a perceptible loss of function[1] I completely agree with the authors that a bladder neck suspension is not necessary and should not been done.

In summary, this paper adds valuable information to the existing literature on ureteroceles. I would urge the authors to simplify their approach and to reserve it only for children with proven reflux before surgery or for the rare cases with bladder outlet obstruction.

1. Vates T, Bukowski TP, Triest J, et al. Is there a best alternative in treating the obstructed upper pole? J Urol. 1996; 156:744

Answer by T.P.V.M. de Jong

We thank dr Gonzales for his kind remarks and will try to answer to the points of criticism.
First, the complete reconstruction in patients without vesicoureteral reflux instead of upper pole resection alone. Reference 6 reports a 85% cure rate in non-refluxing patients, incontinence is not taken into account. Reference 7 reports an 80% cure rate in 41 patients with incontinence occurring in 2 and clean intermittent catheterisation in 2 patients. Of the 16 non-refluxing patients in our group 13 were primarily operated. Indications have been: proven obstructive voiding in 2 patients, incontinence in 1, obvious bladder neck insufficiency at VCUG and/or cystoscopy in 9 with 5 of this group straining when voiding, no obvious indication in 2. Seven ureteroceles were considered huge in size. Of the 3 patients that primarily had their ureteroceles untouched secondary removal was neccessary in 2, one because of obstructive voiding, 1 because of incontinence. Only 1 patient still has her ureterocele. None of these 15 operated patients experienced infectious complications or incontinence at follow-up. This leaves a group of 10 patients that were operated without reflux and without proven obstruction. According to dr. Gonzales we have overtreated 80% of these 10 patients when our observation of bladder neck insufficiency is taken into doubt. But we have spared all these patients reasonable doubt on the outcome of treatment and have been able to end treatment relatively short after the operation. We have originally started this way of treating ectopic ureteroceles because of a large group of patients presenting with incontinence after prior surgery without removal of the ureterocele. Because of dr. Gonzales remarks we will even more carefully look to the selection of patients in the future.
Second point of criticism is the endoscopic incision of cecoureteroceles at the start of the procedure. This is not done to facilitate excision but to prevent the creation of a blind cul de sac in the distal urethra that can function as a valve. It simply is not possible to remove the whole lenght of the ureterocele without major perineal surgery when the ureterocele is prolapsing at the back of the urethral meatus. Of course, one can consider opening of the distal ending of the ureterocele only instead of opening

the whole lenght.
Third point of criticism is the upper pole ureterostomy combined with lower urinary tract reconstruction in a few neonates. Indeed, this issue has not been clarified adequately in the paper, part of the explanation was left out for shortness. It was done to create a safe situation for the patient with the smallest possible procedure in neonates that, in 3 cases had been treated for life-threatening septicemia shortly before this operation and in 2 cases were treated with urgence because of severity of clinical signs needing an indwelling catheter shortly after birth. The primary indication has not been to rescue possibly functioning renal moieties. To our amazement, few of these moieties actually appeared to have a well enough function to preserve them.

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Post-Otis Incontinence

The treatment of post-urethrotomy incontinence in pediatric and adolescent female subjects

CHAPTER 4

Tom P.V.M. de Jong, Jan D. van Gool, Pieter Dik,
Aart J. Klijn and Marianne A.W. Vijverberg

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Abstract

Purpose: Many urologists, until 1986, used a redundant internal urethrotomy as a standard therapy in the treatment of recurrent urinary tract infections in girls. We describe the results of therapy in patients that have become incontinent due to prior internal urethrotomy.

Patients and methods: Between 1986 and 1995, 21 female patients with post-Otis incontinence have been presented at our department. The girls presented with a mix of dysfunctional voiding, recurrent urinary tract infections and different types of urinary incontinence, partly based on bladder instability, often provoked by abdominal stress. All patients were diagnosed with repeated video-urodynamic evaluation, ultrasound of the open bladder neck and endoscopic proof of the scars in the bladder neck and the urethra. All patients but one had elaborate conservative treatment for at least two years. Conservative treatment consisted of pharmaceutical therapy, physical therapy and biofeedback training. Fourteen patients needed surgical therapy to cure their incontinence. A conventional Burch-type colposuspension was performed in 5 patients and a modified needle colposuspension in 4 patients. The last 5 patients were treated by complete endoscopic excision of the urethral scars followed by open reconstruction of both the bladder neck and the urethra in an abdominoperineal procedure.

Results: Surprisingly, conservative treatment has been completely successful in 7 patients. The results of primary open colposuspension or needle colposuspension are disappointing in 6 out of 9 patients leading to further surgery to become dry in several patients. Results of excision of the urethral scars with bladder neck and urethral reconstruction are good in 4 out of 5 patients with a follow-up of at least 4 years. Nowadays, this operation is our procedure of first choice when conservative treatment has failed.

Conclusions: When in the evaluation of incontinence an earlier internal urethrotomy appears to be an important factor, conservative therapy is the treatment of first choice. Conservative therapy should consist of biofeedback reeducation of the voiding pattern and physical therapy. When surgery is needed, excision of the urethral scars with reconstruction of bladder neck and urethra, added to a colposuspension is superior to colposuspension alone.

Introduction

For many years internal urethrotomy has routinely been used for the treatment of recurrent urinary tract infections (UTI's) and mild vesicoureteral reflux (VUR) in girls. The use of the internal urethrotomy was popularised in several publications[1,2], although the positive effect was doubted as early as 1974.[5] In 1986, Kessler and Constantinou[3] published data of post-Otis incontinence, and Allen, in an editorial comment, called the internal urethrotomy for treatment of recurrent UTI's a barbaric procedure.[4]
Nowadays we all realise that the spindle-top urethra, in general, is an expression of a functional non-neurogenic bladder/sphincter dyscoordination and that it is only rarely the result of a true meatal stenosis. Over the years, we have dealt with a group of girls with urinary incontinence that, by exclusion of other problems, had a prior urethrotomy as the most probable reason for their incontinence. Of course, this can be difficult to prove because most of the girls have been presented with some type of lower urinary tract dysfunction leading to recurrent UTI's and/or urge-incontinence prior to the internal urethrotomy. Specific publications on the therapy of post-urethrotomy incontinence cannot be found in the literature except for a few papers on female urethral reconstruction that include small numbers of OTIS victims.[6,7]

Materials and Methods

Between 1986 and 1995, we have treated 21 girls for urinary incontinence who beforehand, between ages of 1 and 11 years (mean 4.4 years), were subjected to a redundant internal urethrotomy as a part of the treatment of recurrent UTI's and/or VUR. In most cases, the original report of the urethrotomy could be traced. Most patients had an internal urethrotomy between 28F and 32F. Specific data on the patient's micturition pattern prior to the urethrotomy were not traceable in most cases. All the parents of girls that were toilet trained at the age of the internal urethrotomy reported that the procedure was the onset of their daughter's incontinence.

All the patients presented to us with urinary incontinence and, in most cases, a micturition pattern resembling a non-neurogenic bladder/sphincter dysfunction. All the patients had a complete urological work-up of the urinary tract including ultrasound of both the upper and the lower urinary tract, video urodynamic studies with coughing and straining and a cystoscopy with special attention to the bladder neck and the urethra. Urodynamic study was done with low filling rates and included recording of bladder and rectal pressures and recording of the pelvic floor EMG by external skin electrodes. Fluoroscopy of the bladder neck and urethra was done with transverse radiation. Endoscopy, in all these patients, revealed damage of the bladder neck and the urethra, ranging from a small lateral gap in the bladder neck or the urethra to a complete separation of the urethra into an anterior and a posterior halve separated by a plate of scar tissue (pictures 1 and 2).

All the patients except 1 have been treated conservative for at least 2 years. In this single patient the indication for operation was severe breakthrough infections with bilateral VUR, combined with stress-incontinence after internal urethrotomy. Conservative treatment consisted of antibiotic prophylaxis in cases of UTI's, antimuscarinic therapy when bladder instability was found either by urodynamic study or by clinical assessment and finally by physical therapy and a biofeedback training program. Biofeedback training was primarily done in an out-patient setting and eventually during a 10-day hospitalisation.[9,10]

After conservative treatment had proven to be insufficient, 14 patients were operated for their incontinence. Five patients were operated using a conventional Burch-type procedure, four patients by a modified needle suspension technique. The last five patients primarily underwent a complete excision of the urethral scar-tissue with reconstruction of the bladder neck and the urethra.

Age at operation of the colposuspension group ranged between 5 and 19 years (mean age 9.3 years), age of the reconstructed group ranged between 9 and 16 years (mean age 12.8 years). The Burch colposuspension was performed with 2 no 1 polyglycolic acid sutures placed in the anterior vaginal wall on both sides at the level of the bladder neck. The vaginal wall is lifted up with a metal sound that is placed in the vagina. The sutures are taken through Cooper's ligament and tied. The modified

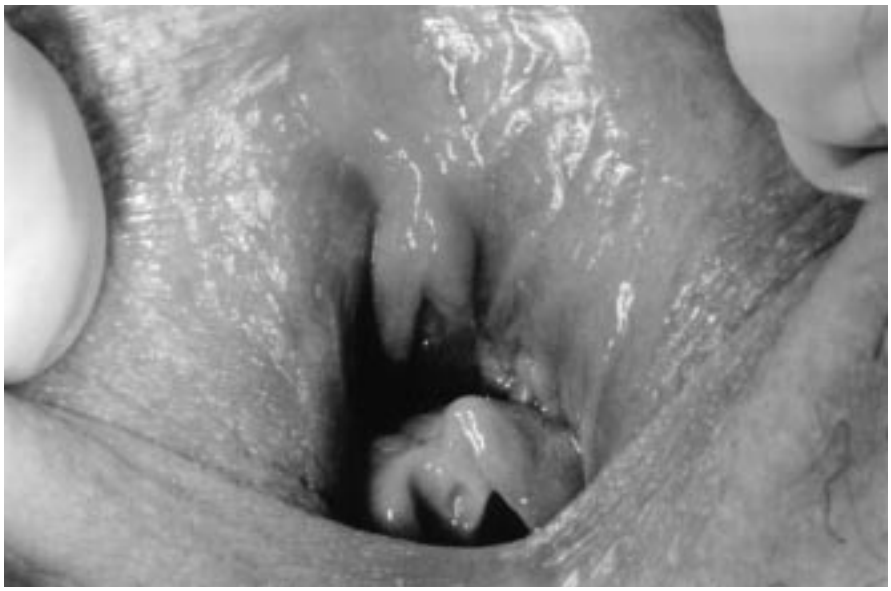

***Figure** 1: Typical outlook of the urethral meatus after early urethrotomy. This patient appeared to have a complete separation of the anterior and posterior halves of the urethra.*

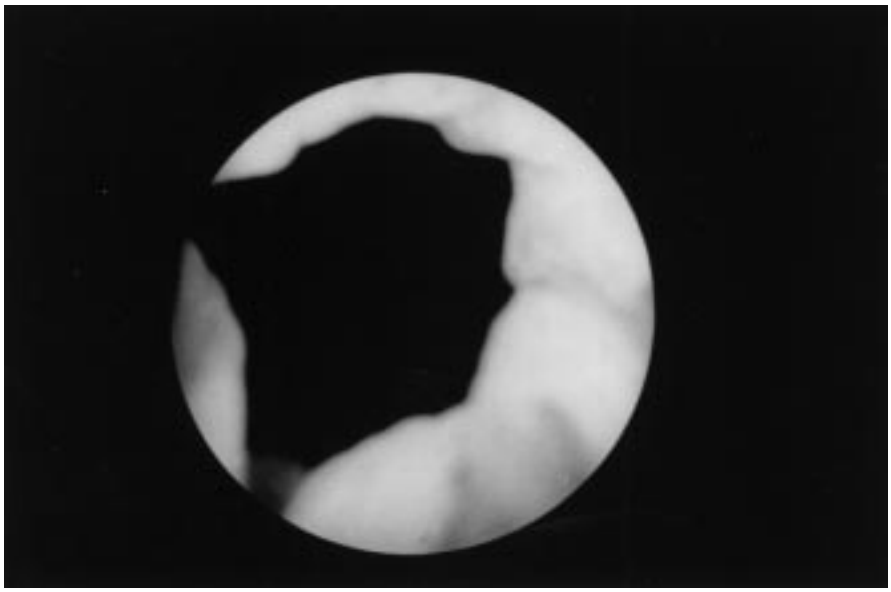

***Figure** 2: Endoscopic appearance of an insufficient bladder neck after internal urethrotomy.*

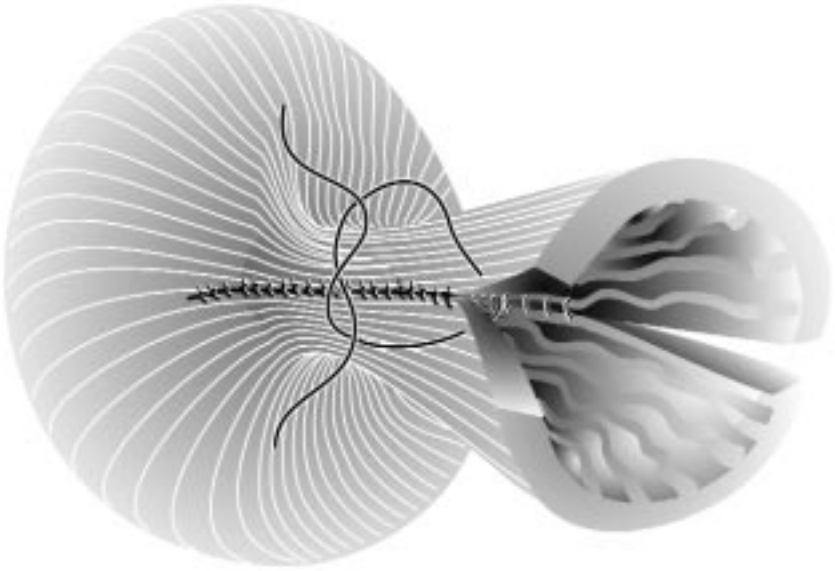

***Figure** 3: Artist's impression of bladder neck and urethral reconstruction.*

needle suspension was performed by putting 4 long 16 G infusion cannulas, 2 on each side, from a small suprapubic incision into the vagina at the level of the bladder neck. After endoscopic control of the bladder neck and the urethra for perforation by the cannulas, a no. 0 polypropylene suture is passed twice through the vaginal wall between the 2 cannulas and backfeeded through the cannulas. After withdrawal of the i.v. cannulas the sutures are tightened at the external abdominis fascia with endoscopic control of the closure of the bladder neck..

The reconstruction procedure starts with a cystoscopy and delineation of the scar tissue of the urethra and the bladder neck. With a standard 24 Fr. adult resectoscope, the scar tissue is excised completely into the surrounding tissues of the urethra. Through a lower abdominal transverse incision the bladder is opened and retracted. The proximal urethra and the bladder neck are reconstructed as deep down as possible with interrupted submucosal 5.0 polyglycolic acid sutures, thus restoring their anatomy. After the abdominal part of the reconstruction, the bladder retractor is released and the bladder neck is pushed downwards with a large sponge. The remainder of the urethra is reconstructed perineally, again with interrupted 5.0

sutures. With sufficient downward pressure on the bladder neck the whole length of the urethra can be reconstructed this way (figure 3). After the repair, a 10 Fr. silicon stent is left transurethrally and fixed to the trigone with a rapid-soluble polyglycolic acid suture. The bladder neck is elevated to Cooper's ligament with a Burch type colposuspension by lifting the anterior vaginal wall with a metal sound and grabbing it with a no 1 or 0 polyglycolic acid suture. Before closure of the bladder a suprapubic 12 Fr. silicon Malecot type catheter is left in the bladder. The patient is discharged with both catheters in situ. The transurethral catheter is removed after 12 days and the suprapubic catheter is clamped to restore voiding. The suprapubic catheter is removed when sufficient bladder emptying has been proven.

Results

Follow-up of the whole group of patients ranges between 4 and 14 years with a mean of 6,8 years. Conservative treatment, to our surprise, has been completely successful in 6 patients. Of course, we cannot predict whether these girls run a greater risk for stress-incontinence after having given birth in the future. Of the 9 patients that have been treated with a colposuspension alone, 3 are completely continent at follow-up, 6 still suffered from incontinence. Four of these had sufficient relief of complaints to refrain from further therapy, one opted for further surgical treatment and underwent a Young Deese bladder neck reconstruction with a silicon wrap around the reconstructed bladder neck in preparation for a possible future sphincter prosthesis. Unfortunately, the silicon wrapping eroded into the urethra and had to be removed transurethrally. Eleven years later, after injection of a silicon bulking agent in the bladder neck (Macroplastique^{R}) she is socially dry with a chronic dependence on oxybutinin and clean intermittent catheterisation. One patient had a subsequent injection into the bladder neck with a bulking agent and became socially dry, she needs a small pad when sporting.

Of the five patients treated by excision of the scar tissue and reconstruction of the urethra 3 have reached complete continence several months after the operation, one has reached complete continence after a second in-hospital biofeedback training programme. One patient, at the age of 15 years had, after initial success, a recurrence of stress incontinence that appeared to be related to a dehiscence of approximately 1 cm of the right-sided reconstruction of the urethra starting immediately distal from the bladder neck. Again, after filling the diverticulum with a bulking agent (Macroplastique^{R}) she was temporary continent but the silicon bulk migrated to the peripheral tissue and could be felt in the anterior vaginal wall. She refused further urethral reconstruction and is now dry and on clean intermittent catheterisation after a sling suspension of the bladder neck.

Eleven of the 14 patients had recurrent UTI's preoperative and needed chronic antibiotic prophylaxis, two patients had only an occasional infection, one patient had no infections at all. After treatment 10 patients are free of infections, 3 have had an occasional UTI, one patient is still on prophylaxis for recurrent UTI's.

Table 1: *Overview of the procedures and outcome.*

No.	age Otis	Otis related procedures	age op.	Operation	result	UTI's pre	UTI's post	special remarks
1	9		13	Burch	dry	no	no	dry after additional bladder neck injection
2	5		13	Burch+	dry	yes	sporadic	on cic after bladder neck reconstruction
3	2	Helmstein dilatation and ureteral reimplant	14	Needle s.	inc	yes	no	socially dry, refuses further therapy
4	3	ureteral reimplant and nefrectomy	12	Burch+	inc	yes	no	dry after additional bladder neck injection
5	4		15	Needle s.	inc	sporadic	no	socially dry, refuses further therapy
6	3		15	Burch	inc	yes	no	socially dry, refuses further therapy
7	3		12	Needle s.	inc	yes	no	socially dry, refuses further therapy
8	7		12	rec	dry	yes	no	reconstruction after earlier failed needle suspension
9	1	Ureteral reimplant	16	rec	dry	yes	no	
10	2		14	rec	dry	yes	sporadic	complete separation of the urethra in two halves
11	2		4	Burch	dry	yes	no	refluxnephropathy and reflux, Burch combined with ureteral reimplant
12	6	Ureteral reimplant	9	rec	dry	yes	yes	reconstruction after earlier failed needle suspension
13	11	ur. Reimplant, renal failure, transplanted kidney	19	Needle s.	dry	yes	no	
14	3		14	rec	inc	yes	sporadic	dry and on cic after additional sling suspension

Table 2: *Urodynamic data. Bladder capacity is given as the percentage of expected bladder capacity for age (30+30 x age = ml).*

uds preop

no	capacity	compliance	Overactive	Dyssynergia	bl.neck	stress inc	residual
1	*50*	*normal*	*no*	*yes*	*NA*	*no*	*20*
2	*60*	*normal*	*yes*	*yes*	*open*	*yes*	*0*
3	*50*	*normal*	*yes*	*yes*	*open*	*yes*	*0*
4	*90*	*normal*	*no*	*yes*	*open*	*no*	*0*
5	*45*	*normal*	*yes*	*yes*	*open*	*no*	*40*
6	*90*	*normal*	*no*	*yes*	*NA*	*no*	*25*
7	*60*	*normal*	*yes*	*yes*	*NA*	*no*	*90*
8	*18*	*normal*	*yes*	*no*	*open*	*no*	*10*
9	*100*	*normal*	*no*	*yes*	*open*	*yes*	*0*
10	*90*	*normal*	*no*	*yes*	*open*	*yes*	*0*
11	*180*	*normal*	*no*	*yes*	*open*	*no*	*35*
12	*60*	*normal*	*yes*	*no*	*NA*	*no*	*10*
13	*200*	*normal*	*no*	*no*	*open*	*yes*	*0*

uds postop

no	capacity	compliance	overactive	dyssynergia	bl.neck	residual	
1	*50*	*normal*	*no*	*no*	*closed*	*0*	
2	*90*	*normal*	*no*	*no*	*closed*	*0*	
3							
4	*75*	*normal*	*no*	*yes*	*closed*	*5*	
5	*NA*						
6	*100*	*normal*	*no*	*yes*	*open*	*60*	*stress-inc*
7	*120*	*normal*	*yes*	*yes*	*closed*	*15*	
8	*80*	*normal*	*no*	*yes*	*closed*	*0*	
9	*125*	*normal*	*no*	*no*	*closed*	*0*	
10	*120*	*normal*	*no*	*yes*	*closed*	*10*	
11	*NA*						
12	*120*	*normal*	*no*	*no*	*closed*	*0*	
13	*na*						
14	*85*	*normal*	*no*	*no*	*closed*	*0*	*stress-inc*

Discussion

It is difficult to prove that the internal urethrotomy has been the only reason for the therapy-resistant incontinence in these girls. Out of a cohort of approximately 750 girls, treated for dysfunctional voiding, UTI's and/or VUR, all these patients shared a scarring of the bladder neck and/or the urethra and they were all resistant to pharmacological therapy, biofeedback training and physical therapy. This makes it very probable that the OTIS-procedure has been the main cause of their incontinence. This theory is also supported by the fact that over the last years substantially less new pediatric cases have been presented. This seems to be a result of the publications in the Journal of Urology that have caused more caution in the use of the urethrotome knife. Clinically, most girls presented with a mix of symptoms. They all had a dysfunctional type of micturition, ranging from an urge syndrome and urge incontinence to true dysfunctional voiding.[8] In some of the girls the urge could be provoked by abdominal stress. Some had a dribbling incontinence as well, also provoked by stress. Finally, some girls suffered from true stress-incontinence. The pathophysiological pathway that explains the voiding pattern is that the insufficient bladder neck results in the constant presence of urine in the proximal urethra with constant contraction of the pelvic floor to withhold this urine. Of course, in patients with true dysfunctional voiding, a similar pathway often exists in a non-iatrogenic opened bladder neck. However, in general, these girls can be treated succesfully by biofeedback training.[9,10] whereas all the patients in the operated group were resistant to training or suffered a rapid relapse after initial success.

A conventional Burch or needle colposuspension has resulted in a disappointing 40% long-term cure rate in our hands. The procedure with scar excision and urethral reconstruction has been developed after a discussion with Dr. T. Allen on a few difficult cases with a scarred urethra. It was his remark, in 1990, that one should think about the possibility to excise the complete scar and reconstruct the urethra that has put our thought on this operation. It may be expected that, with the development of laparoscopic suturing techniques it will be possible in the near future to perform this operation as a perineal endoscopic procedure.

An important issue in this matter is the question whether we are looking

at a small group of patients or at the tip of an ice-berg that is coming to us. Allen, in 1986, expects this type of incontinence to become a national problem in the United States. A survey of the literature does not reveal any publications that support this theory.
For the pediatric population one may expect that this iatrogenic urethral scarring is a self-limiting disease with the understanding of the mechanism of the spindle-top urethra in dysfunctional voiding and the elimination of routine internal urethrotomy in the treatment of pediatric recurrent UTI's. But one cannot predict the numbers of adult females with a history of an internal urethrotomy. Keitzer advocated 30F, 38F , 45F and 48F full lenght incisions running through the bladder neck in 4 quadrants in 1963 and Halverstadt proposed full length incisions at the 5 and 7 o 'clock position without paying attention to the numbers on the urethrotome dial in 1968. This means that a large group of women between the age of 16 and 35 years must exist that has been subjected to a redundant internal urethrotomy.
It may be superfluous to report that we have encountered much psychological distress in our patients with an OTIS-induced incontinence.

Conclusion

In the treatment of female incontinence special attention must be paid to the bladder neck and the urethra of all the patients with an internal urethrotomy in their anamnesis. When a relationship between the internal urethrotomy and the incontinence is suspected, primary treatment should consist of biofeedback training of the micurition pattern and physical therapy. When surgery is needed reconstruction of the bladder neck and urethra after excision of the scar tissue added to a colposuspension appears to be superior to colposuspension alone.

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Surgery for Female Ambiguous Genitalia

Neonatal management of female intersex by clitorovaginoplasty

CHAPTER 5

Tom P.V.M. de Jong, Thomas M.L. Boemers

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Abstract

Historically, in female pseudohermaphrodites a staged procedure with early clitoral reduction and delayed vaginoplasty was often the treatment of choice. In recent years several authors have described 1-stage genitoplasty that is performed in the first year of life. The 1- stage procedure for clitoroplasty and vaginal exteriorization is preferable for an optimal functional and cosmetic result. Because of emotional aspects of the family in intersex cases, neonatal reconstruction offers advantages to the child and parents. Furthermore, the perinatal genital hyperstimulation by maternal and placental estrogens that persists into the first 3 to 4 weeks of life produces vaginal enlargement by mucous secretion and vaginal wall hypertrophy, facilitating identification of the cleavage planes and vaginal pull-through. These arguments have led us to treat these patients by neonatal 1 stage clitorovaginoplasty. Cosmetic and functional results in 5 consecutive cases have been excellent without serious complications.

Introduction

Many techniques for clitorovaginoplasty have been reported, mostly accompanied by advice concerning the timing of the operation. Historically, in moderate to high ending vaginas many advocated early clitoroplasty to ameliorate the cosmetic appearance of the external genitalia, followed by vaginoplasty at a later age. Recently, others proposed a 1-stage procedure with relatively early operation in the first year of life.[1-4] An argument for a 1-stage procedure is that early clitoroplasty can compromise the cosmetic result of later vaginoplasty. Furthermore, some patients have concomitant urinary tract problems based on the urogenital sinus that make early intervention preferable, especially in cases of hydrometrocolpos. In 5 consecutive cases that presented immediately after birth we opted for primary reconstruction in the first weeks of life.

Material and methods

Five virilized female neonates (4 with the adrenogenital syndrome and 1 true hermaphrodite) were treated by clitoroplasty and vaginal exteriorization. Patient age at operation was 1 to 3 weeks. All patients were referred with ambiguous genitalia immediately after birth. In 3 cases the diagnosis of a probable female gender and the adrenogenital syndrome was made at referral by ultrasound and retrograde genitography, and in 1 the diagnosis of the adrenogenital syndrome was confirmed by laparoscopy and endoscopy of the urogenital sinus with retrograde vaginogram. The diagnosis adrenogenital syndrome was confirmed by biochemical data in all cases. A normal 46XX karyotype was noted in these patients. Patient 5 had XO/XY mosaicism on karyotype, and an ovotestis and a streak ovary were removed by laparotomy while she was under anesthesia for genital reconstruction.

All patients underwent surgery using a perineal approach (fig.1). Clitoroplasty was started with circumcision and degloving of the clitoris, followed by the mid shaft lateral incision of Buck's fascia to create a cleavage plane with the tunica albuginea, thus permitting the dissection of Buck's fascia (containing the neurovascular bundles) and the glans clitoris off the corpora cavernosa (fig. 2). Subsequently resection of the corpora up to the caudal end of the symphysis past the bifurcation.[1,5,6] The spongiosum tissue of the remaining crura of the corpora was destroyed by dilatation with a 2 mm. metal sound to avoid later painful erections. The stumps of the corpora were closed by a running 5-zero polyglycolic acid suture. When necessary, the glans was reduced by wedge excision and/or partial de-epithelization and was sutured to the stumps of the corpora, providing a clitoris in the physiological position at the caudal end of the symphysis. Before vaginal exteriorization a Fogarty catheter was inserted endoscopically into the vagina and a balloon catheter was placed into the bladder. After opening of the perineum with a midline incision that continued into an inverted U-flap above the anus (fig. 1, A), the urogenital sinus was followed proximally until the Fogarty catheter balloon could be palpated (fig. 1, B). The vaginal tip was opened and the anterior vaginal wall freed from the urethra. The opening of the vagina into the urethra/urogenital sinus was closed with 6-zero polyglycolic acid sutures,

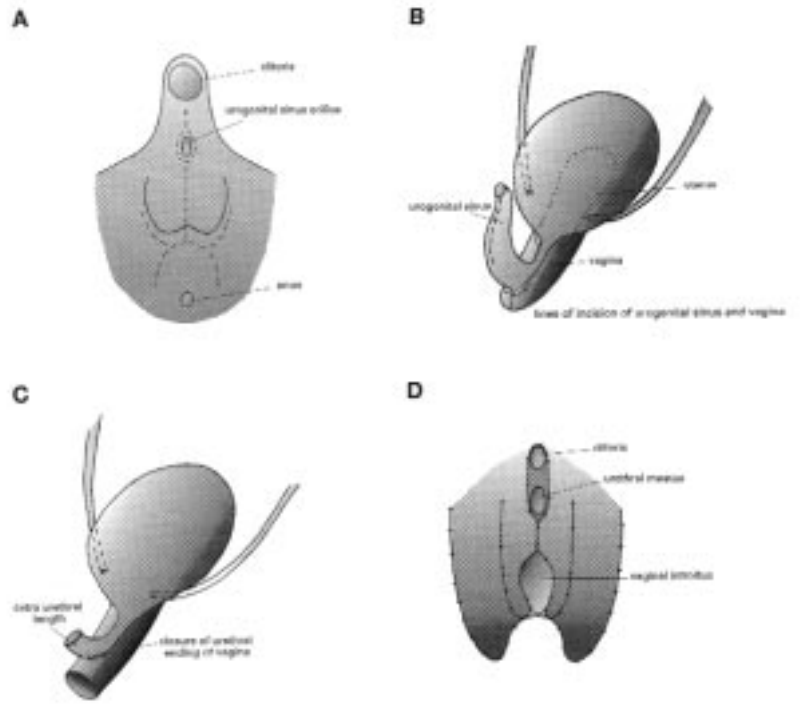

Figure 1: A. *skin incisions are made according to the dotted lines.* ***B.*** *urogenital sinus and vagina are incised according to dotted lines after inserting a small caliber balloon catheter into the vagina endoscopically.* ***C.*** *the situation after closure of the vaginal ending in the urethra/urogenital sinus.* ***D.*** *end-result: the anterior and lateral vagina is sutured to the minor labia consisting of phallic skin. The dorsal vagina is sutured to the inverted U-flap.*

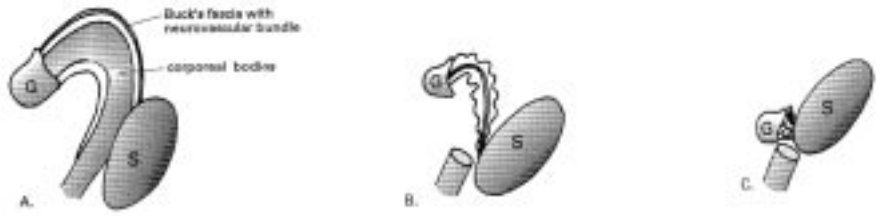

Figure 2: A. *longitudinal section of phallus and symphysis. After degloving of the phallus Buck's fascia with the neurovascular bundles is removed from the corporal bodies.* ***B*** *the corporal bodies are resected up till the bifurcation at the caudal end of the symphysis, the corporal stumps are closed after destroying the remaining erectile tissue with a metal sound.* ***C*** *after reduction of the glans this is sutured to the remaining stump of the corporal bodies.*

leaving a redundant length of urethra (figure 1. C). The urogenital sinus was partially opened until a urethra remained that was rather long for patient age. The open strip of the urogenital sinus epithelium was sutured cranially to the clitoris and caudally to the anterior vaginal wall. The phallic skin is remodeled to provide a preputial hood proximally as well as labia minora, which are distally sutured to the lateral vaginal wall. The inverted U-flap was then sutured to the dorsal vaginal wall. The labial folds are reduced and transposed posteriorly to create normal appearing labia majora.

Results

Convalescence was uneventful in all cases. With proper endocrinologic care no electrolyte or fluid complications developed in the post-operative period. Cosmetic appearance is considered excellent in all cases. Follow-up in the 5 cases is 28, 26, 12, 11 and 9 month, respectively. One patient underwent meatotomy of the urethral meatus at 6 and 12 months after reconstruction with an uneventful followup of 16 months after the last meatotomy. In the other patients, a 10F catheter passes the urethral meatus and a 14F catheter passes the vaginal orifice without resistance and any need for dilatation.

Discussion

Vaginal exteriorization in neonates seems to be relatively easy compared to reconstruction at a later age. A transtrigonal approach was not necessary in our patients although in 2 the vaginal ending into the urethra was considered to be high (Prader IV to V). Others who described this approach in these procedures indicate that reconstruction can be done early[1,3] but should not be done in high ending vaginas.[2]
However, to our knowlegde the results of neonatal procedures have not been previously published. The reason for easy vaginal exteriorization is

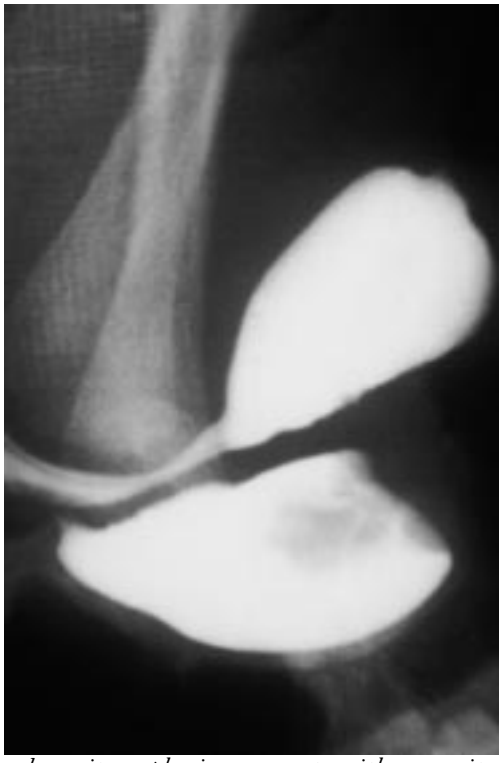

Figure 3: *retrograde genitography in a neonate with urogenital sinus showing a dilated vagina with a mucous plug.*

the neonatal hypertrophy of the external and internal genitalia at birth, based on maternal and placental hyperstimulation with estrogens. Indeed, all of our patients had a distended vagina filled with whitish mucous (fig. 3). This effect can probably be used to advantage during the first 2 weeks of life.[7,8] There is no sign whatsoever for less perfect results in neonatal reconstructions (fig. 4). The psychological advantage to the parents and children is enormous, while there are no signs of greater risk for these patients with current pediatric anesthesiological procedures. In contrast, in the same period we operated on 14 similar patients aged 4 years or older, and the patients and parents had severe psychological problems. Of these patients 10 experienced considerable urinary tract morbidity that has partly been based on the untreated urogenital sinus. Four patients had recurrent urinary tract infections, 2 complained of dysuria and urge

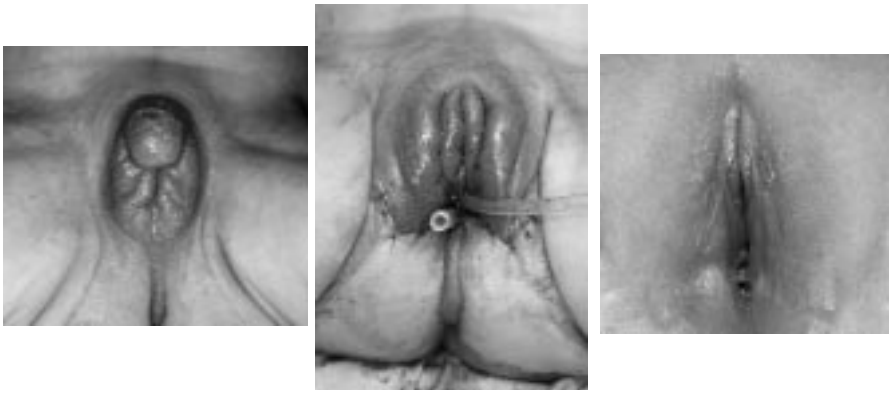

Figure 4: A. *pre-operative photograph of a patient with adrenogenital syndrome and high ending vagina.* ***B.*** *postoperative view of the same patient.* ***C.*** *the same patient 8 months after operation*

incontinence, and 2 had dribbling incontinence after voiding. These complaints subsided after reconstruction. Furthermore, vesicoureteral reflux probably unrelated to the urogenital sinus was present in 1 patient and infravesical obstruction resulting in a giant bladder diverticulum was present in 1. Both anomalies were treated during reconstruction without morbidity. The percent-age of urinary tract complications in this older group is high compared to the 35% described by Lobe et al.[9] In several older patients with adrenogenital syndrome and a high ending vagina proximal to the external sphincter who underwent surgery by other methods there has been stress incontinence due to an extremely short urethra after reconstruction. A review of these cases indicates the origin of incontinence: to create a normal vaginal introitus the short urethra was mobilized up to the level of the bladder neck to pull the orifice in level with the introitus. This procedure seems to have resulted in some damage of the sphincteric mechanism and lack of adequate functional urethral length after primary reconstruction. Our procedure as described provides a urethra with normal or redundant length for age without the need of mobilizing the proximal urethra. We hope that this adequately prevents incontinence based on a short functional urethra, even in high ending vaginas. In our first 2 patients normal daytime continence developed.

However, followup is too short to make definitive statements on this aspect. In conclusion, neonatal 1-stage reconstruction seems to be the most feasible approach to treat these patients. It will take many more years to prove definitely that the benefits of an early operation are maintained throughout puberty.

Postscript: The follow-up of the girls described in this article nowadays ranges between 7 and 10 years. They experienced not a single urinary tract complication over these years. Many other patients have been operated using this scheme with similar results. This paper has been the first publication worldwide that propagates neonatal reconstruction of female intersex patients. A survey on the treatment of female intersex patients was done by interactive panel discussion at the combined meeting of the European and the American Pediatric Urological meeting in Tours, France in june 2000. Twenty percent of the pediatric urologists and pediatric surgeons have adopted our policy and treat the children immediately after birth. Of six papers on this subject, published in leading journals since 1996, this article has been cited in 5.

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Bladder Extrophy and Epispadias

Treatment of bladder extrophy and epispadias

CHAPTER 6

Tom P.V.M. de Jong

European Journal of Urology 1999; 36/1 (curric Urol 1.3:1-9),
written by invitation

Abstract

Bladder extrophy and epispadias remain a challenging problem in pediatric urology. There are many therapeutic options ranging from different types of primary closure to the primary choice for diversion in extrophy patients. Further, there are several options for the treatment of male epispadias. In bladder extrophy, primary closure with meticulous reconstruction of the pelvic floor, combined with early clean intermittent catheterisation, seems to be the best first choice. In cases of female epispadias, a new technique will be described which provides a good chance to develop continence and voiding after one operation without the need for bladder neck reconstruction. The chapter tries to give a review of the contemporary treatment of extrophy and epispadias including some details of operative procedures.

Introduction

The exstrophy/epispadias complex represents, in most cases, dramatic congenital anomalies with lifelong consequences for both the child and the parents. It is rarely in mild epispadias only a cosmetic problem. The syndrome ranges from cloacal exstrophy with involvement of both the urinary tract and the bowel, through classic bladder exstrophy to epispadias.

Anomalies in the exstrophy/epispadias complex are rare. The prevalence of bladder exstrophy is approximately 1 in 30,000 live births. The prevalence of epispadias in boys is approximately 1 in 40,000, in girls 1 in 200,000. The incidence of classic bladder exstrophy in the Western world appears to be declining, possibly due to intrauterine detection with subsequent abortion of the fetus although specific numbers of prenatally detected cases cannot be found in the literature.

Many recent publications describe the results of surgical repair of exstrophy and epispadias without giving details of treatment techniques - in this chapter a more practical approach will be adopted.

Anatomy of Bladder Exstrophy and Epispadias

In classic bladder exstrophy the bladder (fig. 1) and the urethra are open structures lying on the anterior abdominal wall. The amount of the bladder that is visible can range from a large bulging bladder plate to a hardly visible small patch. In males, the colliculus protrudes at the beginning of the urethral plate. The urethral plate runs to the glans penis. The glans is flattened and is connected to diverging corpora cavernosa fixed to the always separated halves of the pubic bones. In females, the urethral plate runs between the separate halves of the clitoris to the anteriorly positioned hymenal ring that is present in nearly all cases. The umbilical cord is implanted at the cranial margin of the bladder, thus caudal to the normal position.

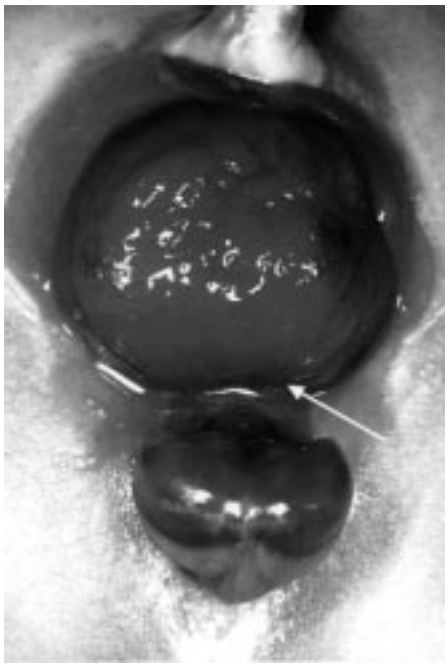

Figure 1: *Example of a new-born boy with bladder extrophy. The child has a relatively large bladder plate, a good candidate for successfull development of the bladder after primary closure. Notice the umbilical cord that has a typical, low implant adjacent to the bladder plate. The phallus is broad with a flattened glans. The arrow points at the left ureteric orifice.*

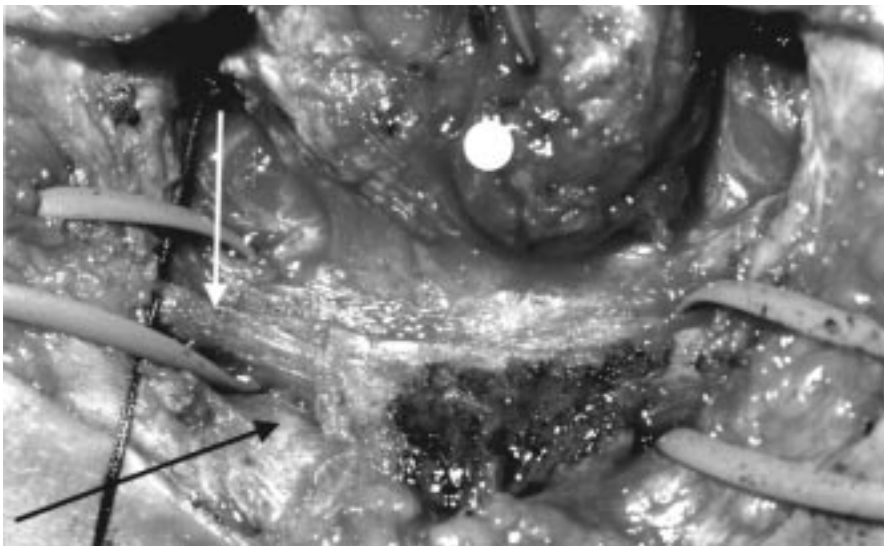

Figure 2: *The image of the anterior rim of the pelvic floor after mobilisation of the bladder and prostrate. The vessel loops are placed around the pelvic floor near the pubic bones. White circle = prostatic plate. White arrow = pelvic floor. Black arrow = corpus cavernosum with neurovascular bundle. Notice the close contact between corpus and pelvic floor and pubic bone. Care must be taken meticulously not to damage the phallus when closing the pelvic floor or the bony pelvis.*

In bladder exstrophy the pubic bones are always widely separated, ranging from 2 to 8 cm. The anterior rim of the pelvic floor can be palpated between the pubic bones (fig. 2).

In male epispadias the urethral orifice is situated flush with the abdominal wall at the level of the symphysis or more distally at the anterior side of the penis, thus classified as glanular, penile and subsymphyseal. It can come together with symphyseal diastasis. In proximal epispadias, with or without symphyseal diastasis, incontinence based on congenital sphincteric insufficiency is common.
In female epispadias presents, there is no urethra, the bladder neck opens at the level of the hymenal ring. The hymen is intact in most cases. These girls are often misdiagnosed as intersex cases or can present at later age with continuous dribbling incontinence.

Finally, a wide range of exstrophy variants exists. This can be a superior bladder fissure, an open bladder top with normal distal anatomy, a duplicate exstrophy with a normal bladder and urethra in the abdomen, a bladder plate and a dorsal chordee at the surface of the abdominal wall, a covered exstrophy with symphyseal diastasis in more or less normal anatomy of the lower urinary tract.[1-7] All these cases need individually tailored therapy.

Associated Anomalies and Diagnostics

In classic bladder exstrophy, in general, few associated anomalies occur. There can be vertebral anomalies in approximately 7% and hip dislocations, rarely there are associated upper tract anomalies. The uterus can be partially or completely duplicated. Syndromes that include bladder exstrophy/epispadias are not known, although inheritance of bladder exstrophy is apparent in the relatively few cases that have proven fertility. Most patients have an anteriorly positioned anus. Inguinal hernias are common, in both girls and boys with bladder exstrophy. In epispadias, the majority of children do not have other associated anomalies, apart from the diastasis of the pubic bones. The diastasis of the pubic bones, when untreated, seems to have little effect on the children other than external rotation of the hips with, sometimes, a waddling gait in patients with a wide separation of the pubic bones.[8-11]

In cloacal exstrophy many associated anomalies can occur, obviating the need of redundant diagnostics prior to the decision to start reconstructive operative treatment of the patient. Cloacal exstrophy comes with spinal dysraphism, upper tract anomalies, cardiac anomalies and omphalocele in a high percentage.[12-17]

Thus, in classic exstrophy and epispadias, ultrasound of the urinary tract and a plain X-ray of the spine will provide sufficient information in most cases. In cloacal exstrophy, additional investigations, including karyotyping and cardiac ultrasound are neccessary.

Embryology of the Exstrophy/Epispadias Complex

Several explanations for the embryogenesis of exstrophy have been proposed. The bladder is separated from the gastrointestinal tract by the 6th week of gestational age. The ureters and part of the trigone, bladder neck and prostate (males) or urethra (females) are of wolffian origin. The most accepted theory for the development of bladder exstrophy is that the persistence of a large cloacal membrane prohibits development of the mesoderm leading to abdominal wall closure and phallic formation as described by Marshall ans Muecke. Several alternative theories exist, ranging from the low origin of the umbilical cord as the principal anomaly to the suggestion that the first anomaly lies in a lack of rotation in the pelvic ring primordia, thus suggesting that the exstrophy is secondary to malformation of the pelvic bones.[14,18-20]

Timing of Treatment and Treatment Choice

Generally, for reconstructive surgery of the lower urinary tract it is preferable to finish treatment before the age of 18 months. Psychologically, surgery of the genitalia before the onset of mental coupling of gender and genitalia is a great advantage. Nursing complications such as the unwillingness to void after removal of catheters can be avoided by operation before the age of 18 months.

In male epispadias, treatment of the phallus can be scheduled similarly as in hypospadias. Penile growth in the first year is relatively important. When the stretched penile length is insufficient, testosterone treatment by intramuscular injection or by ointment can be a great help. Operation will take place around the age of 10-12 months. When necessary, bladder neck reconstruction takes place at the age of 4-5 years.

In female epispadias, phallic size is unimportant, thus timing of the correction depends on the preference of the pediatric urologist. A small bladder capacity can be a relative contraindication to early surgery, a tendency of the bladder to prolapse can force early surgery.

Classic bladder exstrophy is considered to be a newborn emergency by most pediatric urologists, although groups exist that choose for a primarily delayed closure after weeks or months. Several factors support the advantage of bladder closure within the first 48-72h of life. In the first days after birth the symphysial diastasis can be closed primarily, without osteotomies, due to the circulating maternal hormones that maintain a flexible pelvic ring. Moreover, bacterial ingrowth of the bladder increases by time and tissue quality of the bladder changes rapidly because of edema and dryness. After primary bladder closure, in general, correction of the epispadias takes place at age 12-14 months, followed by bladder neck reconstruction at the age af 4-5 years. When necessary, bladder neck reconstruction is combined with bladder augmentation by ileocystoplasty. A catheterizable, continent stoma of the bladder is also possible.
Both for epispadias and for exstrophy patients, nowadays, we try to achieve continence and satisfactory voiding in the majority of cases at primary closure, followed by temporary or permanent clean intermittent catheterization (CIC) if necessary.
The optimal treatment of classic bladder exstrophy, in Europe, was agreed on at the consensus meeting of the European Society for Pediatric Urology (ESPU) in 1992.

The following recommendations were made: (1) Early closure without osteotomies or delayed closure at 7-10 days with anterior (Salter type), osteotomies. (2) Epispadias repair at 12 months, when necessary after testosterone treatment. (3) Bladder neck reconstruction at the age of 4-5 years. Bladder capacity < 100 ml with ileocystoplasty, > 100 ml without cystoplasty. (4) When it is apparent that CIC is necessary, especially in girls, a continent catheterizable stoma of the bladder should be constructed.
For those who do not follow these recommendations there are a number of options.
Some opt for delayed closure with osteotomies after several months, some opt for primary deviation resulting in cystectomy with construction of a catheterizable continent urinary pouch constructed out of bowel.[21-24] The disadvantages of these approaches are many. Long-term safety of bowel reservoirs constructed in early life has yet to be proven. For patients in countries with restricted possibilities for health care a rectal bladder with

sigmoid pull-through through the anal sphincter can provide a safe solution to obtain urinary continence without endangering the upper tracts [personal communication with Prof. Dr. A. Pesamosca, Bucharest, Romania, with 30 years of follow-up].

The Surgical Treatment of Bladder Exstrophy

Technical Aspects of Primary Closure

In bladder exstrophy, treatment starts immediately after birth. Irritation to the bladder must be avoided, so plastic umbilical cord clamps are removed or fixed cranial to the bladder. We treat the bladder after admission to the hospital with an evaporator, normally used for upper airway problems, located at a few centimeters from the bladder. Operation is preferably sceduled within the first 48 h after birth to avoid the need of primary osteotomies and to preserve optimal quality of the bladder mucosa.

Because of the risk of developing latex allergy after multiple interventions, we treat our patients preferably in a latex-free environment with latex-free surgical gloves and catheters.[25,26]

Surgery takes place under general anesthesia, preferably with a caudal epidural catheter. We give amoxicillin/clavulic acid and gentamycin as prophylactic antibiotic treatment. The operation starts with identification of the ureteral orifices and the colliculus (in males). The umbilical cord is fixed at the cranial margin of the bladder plate. The umbilicus is freed from adjacent tissue, the umbilical arteries and vein are identified and freed. Cosmetically we prefer to 'transplant' the umbilicus to the normal position.[27] This is done by excision of a segment of skin, approximately 1.5 cm in diameter, at the normal site of the umbilicus through the rectus fascia into the preperitoneal space. After tunneling the plane between the freed umbilical cord and the newly prepared site the umbilical remnant is pulled into the new location. It is fixed to the skin with absorbable sutures. Cosmetically this procedure is better than a man-made umbilicus at the exit-site of a suprapubic catheter.

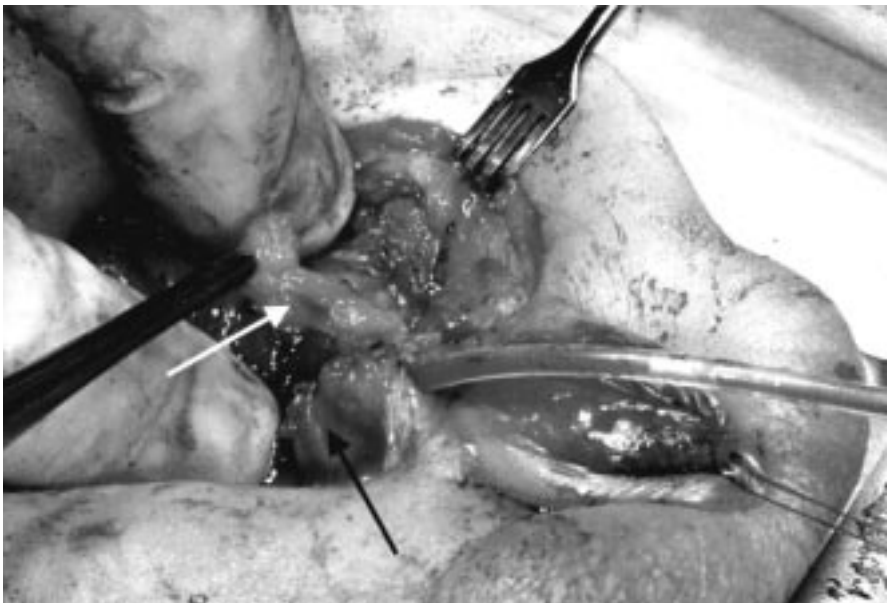

Figure 3: *Peroperative picture after closure of the bladder neck and the proximal part of the urethra. One half of the mobilised anterior pelvic floor (white arrow) is prepared to wrap around the proximal part of the neo-urethra. The apex of the reconstructed prostate is shown by the black arrow. In many cases, for optimal reconstruction of the pelvic floor, the pubic bones must be approximated partially to provide optimal wrapping around the urethra of the pelvic muscles.*

Next, the bladder is freed from the abdominal wall. It can be done in continuity with the urethral plate or disconnected with paraexstrophy flaps as proposed by Jeffs and Duckett, although many authors consider this procedure old fashioned. The advantage of leaving the urethral plate fixed at the bladder is a much easier primary procedure, the disadvantage is that certainly a secondary procedure to achieve continence will be needed. The theoretical possibility that the paraexstrophy flaps may prohibit easy intermittent catheterization has not been encountered in our patients, provided that they start CIC early after primary closure.
One advantage of the use of paraexstrophy flaps is the lengthening of the urethral plate, thus giving enough length to move the bladder neck into an intra-abdominal position, preferably with the proximal urethra cranial to the pelvic floor (fig. 3). The second advantage is that it provides the

opportunity to free the anterior pelvic floor from the rami of the pubic bones and and to close the muscle around the proximal urethra. Thus, in many cases, continence can be achieved after the primary closure. In the past, this procedure was criticized because of upper tract deterioration caused by urethral obstruction. By starting CIC shortly after removal of the urethral catheter this problem can be avoided. After several weeks or months most patients will start to void spontaneously.

Freeing the bladder from the abdominal wall is a meticulous procedure. The bladder plate is circumcised into the preperitoneal fat providing optimal mobility. Care must be taken to leave the vascular pedicles intact. At the level of the prostate, bleeding from spongiosal tissue can be difficult to cope with. By cutting straight up to the level of the pelvic floor bleeding can be stopped by suturing the margins of the spongious tissue. In girls, there is a serious risk of producing a urethrovaginal fistula, the plane between the bladder and the vagina can be difficult to find as both organs are very thin-walled.

Paraexstrophy flaps are continuous with the urethral plate. After mobilizing the bladder upwards, they can be brought together in the midline, thus lengthening the urethral plate.

Bladder closure is done with interrupted 5.0 polyglycolic acid sutures, paraexstrophy flaps and bladder neck are modelled into a urethra with 6.0 sutures around an 8- or 10- Fr silicone catheter. The catheter is fixed to the bladder with a rapidly reabsorbable suture.

When the bladder plate is large, ureteral splinting is not strictly needed. We perform a ureteral advancement into the midline, as described by Gil-Vernet, as a primary antireflux procedure at first closure of the exstrophy bladder. In our hands, this simple procedure prevents vesicoureteral reflux in approximately 60% of the cases. With a very small bladder plate, ureteral splinting with 4-Fr feeding tubes is carried out for 10-14 days.

The medial surface of the pubic bones is freed from muscular and fibrous tissue from the anterior pelvic floor. Distally, the pelvic floor has strong attachments to the corpora cavernosa. Care must be taken to identify and free the corpora cavernosa, thus avoiding penile necrosis when closing the pelvic ring. When the symphysial gap is wide, the pelvic floor remnants cannot be closed around the neo-urethra before partial approximation of the pelvic ring.

Pelvic ring closure can be done with a No. 1 polyglycolic acid mattress suture through the pubic bones. The suture is passed through the rami of the pubic bones and tightened slightly to give the opporunity to close the pelvic floor, immediately afterwards the bones are completely approximated. When separation of the pubis is wide, external pressure at the hips is needed to bring the pubic bones into close contact.
The abdominal wall is closed in layers. By closing the pelvic ring the rectus fascia can be easily closed in the midline, combined with a simple midline skin closure. Skin flaps are not needed in this procedure.
Whether epispadias closure is performed at the primary closure depends on phallic size and preference of the pediatric urologist. Strict contraindications for primary epispadias closure do not exist. Operating time in the neonate can be a relative contraindication.
Postoperatively, the patient is treated by Bryant's traction for the pelvic ring closure: both legs are separately fixed upwards with a weight that leaves the buttocks just free of the bed. It offers the child relatively free movement of the legs, while securing the pelvic ring. The child can be taken out of this traction for breast feeding and cuddling in a plastic shell that fixes the legs at a right angle. In the past, we have tried external fixation with some success. Bone quality in the neonate prohibits secure fixation for a longer period of time.

In general, children are taken care of in a neonatal intensive care unit for 24 h. Broadspectrum antibiotic prophylaxis is given for 24 h. Trimethoprim 2 mg/kg/day, once a day, is started on the 2nd day and continued for at least the first year of life. Ureteral stents are left for 5-15 days, depending on the size of the bladder plate. A transurethral catheter is left in situ for 3 weeks, after removal of the catheter CIC is started 5 times/day. All patients are kept on oxybutinin 0.5 mg/kg/day at 6-hour intervals for the first 6 weeks. The oxybutinin treatment obviates the need of extra care for bowel emptying. Sometimes, enema therapy is needed. During the first year, ultrasound examinations of the upper tract are carried out every 2 months after discharge from the hospital. Urinary cultures are performed every month.

Orthopedic Surgery in Exstrophy Patients

In delayed closure of the exstrophy bladder, osteotomies are mandatory to obtain closure of the pelvic ring. For continence, successful closure of the pelvic ring appears to be benificial. Moreover, it prevents gait problems at a later age. Many techniques have been described. In our hands, Salter-type osteotomies have resulted in temporary femoral nerve palsy in the majority of cases. Still, this technique provides optimal reconstruction of the bony defect.[8,10,11,19,28,29]

Surgical Treatment of Male Epispadias

Epispadias repair is subdivided into repair of primary, congenital epispadias or secondary epispadias after primary exstrophy closure. Preferably, surgery is timed around the age of 12 months, with or without treatment for phallic size with testosterone as a local ointment (2.5% testosterone in cream, twice daily for 3 weeks, to stop 3 weeks before operation, not to be used by the mother with bare hands) or systemic (2 injections of 25 mg of an androgen mix i.m, 60 and 30 days before operation). Many variations on dosage scedule exist. Preferably, before operation, information should be available on the quality of the external sphincter. This information can be obtained by observation of the infant when crying, does he lose urine with high abdominal pressure or not? Urodynamic study assisted by fluoroscopy can give an indication of the quality of the sphincter. Starting the procedure with a cystoscopy can also give additional information in most cases, although one should bear in mind that a caudal anesthetic block opens the sphincteric mechanism in boys under anesthesia.
The operation starts by placing a traction suture through the glans and opening of the skin alongside the urethral plate and around the urethral orifice. After degloving of the penis, the urethral plate is freed completely from the corpora and incised up to the top of the glans and finally rolled into a urethra with absorbable sutures. The tip of the glans is incised in the midline over a distance of a few millimeters and closed squarely to provide a better shape to the meatus (inverted MAGPI procedure, called

IPMAG). Cranial curvature of the penis is partly cured by excision of chordee-like tissue and finally by rotation of the corpora and fixation of the rotated corpora with or without opening and connecting the corporeal bodies in the midline. The corpora are connected anteriorly to the neourethra. Curvature is assessed by inducing an artificial erection using two needles and two syringes, one for each corpus because the connecting vessels existing in a normal penis are absent in epispadias. Finally, the glans is closed over the neourethra in two layers and the penile skin is repositioned.

When, in primary epispadias, sphincteric activity is thought to be ineffective, we have adopted a policy of trying to find the anterior side of the pelvic floor, free it from the pubic bone and wrap it around the urethra or taper it anteriorly to the urethra. This can be done even with a closed pubic symphysis. Success has been variable and the numbers are too small to draw conclusions.

When incontinence is apparent at a later age, physical therapy can be tried in mild cases, followed by injectional therapy with collagen, silicon particles, Teflon or other materials. Severe incontinence will end with formal bladder neck reconstruction, with or without clean intermittent catheterization and bladder augmentation.[30-32]

The Surgical Treatment of Female Epispadias

Female epispadias is a rare entity, series in the literature of more then a few cases do not exist. Straightforward operation techniques other then remodelling a urethra, combined with bladder neck reconstruction, have not been published. We have devised a technique that gives hopeful results with 3 out of 4 patients continent, 3 out of 4 voiding and 1 on CIC. One patient is too young to assess continence The operation starts with an incision alongside the urethral plate that runs between the halves of the clitoris, the incision is lengthened around the urethral orifice. The urethral plate is completely mobilized and tubularized. The bladder neck and trigone are freed completely from the vaginal wall and pelvic floor, permitting mobilization of the bladder outlet and the urethra into the

abdominal cavity. Long intravenous infusion canulas are passed from the inguinal region along the inner side of the pubic symphysis into the wound, two on each side. Nonabsorbable sutures are placed at the bladder neck and passed upwards through the cannulas. When the sutures are tightened and knotted at the external abdominis fascia, bladder neck and urethra slide into a physiologic position. The pelvic floor is identified and closed in the midline between bladder neck and vagina. Finally, the vulva is reconstructed by joining the clitoral halves and approximation of the labia majora anteriorly.[33,34]

The Treatment of Cloacal Exstrophy

Cloacal exstrophy is a heavy burden for the children, their parents and doctors. The bladder consists of two halves joined in the midline by the remnants of the extrophied colon and the protruding ileum. Primary urologic therapy is not very much different from classic exstrophy although a multidisciplinary approach combined with general pediatric and orthopedic surgeons is mandatory in most cases. Problems arise with related congenital anomalies that can be impressive. One should consider very thoroughly all the ins and outs for the baby and parents before starting therapy. The list of operations and complications is a very long one. Therefore, in our hospital, we think it is justified to advise the parents not to have the baby treated resulting in early death. The few cases that we have treated, according to the parents' wishes, have done relatively well although complications such as a short bowel syndrome, urinary tract infections, and, not to forget, major psychologic complications have been impressive.[13,16] Worldwide, this policy of nontreatment is not possible in most countries, forcing the medical teams to try and offer optimal therapy. In general, lifelong medical guidance is mandatory for these children.

General Care of the Patients

The exstrophy/epispadias complex patients are subject to a lot of medical technology resulting, in the best cases, in a relatively good situation of continency and voiding with the possibility of a relatively normal sexual life, in the worst cases with a continent catheterizable stoma and poorly functioning sexual organs. The hospitalization alone, even with a good end result, can produce psychologic distress. When surgery leads to a situation with a frustrated body image psychologic complications can be impressive, necessitating of thorough follow-up and treatment by a specialized team that takes the child through puberty into adult life.

CIC in these children produces significantly more aversion to the treatment then in spina bifida patients because of the normal sensibilty of the urethra.

Sexual life can be normal in female patients with normal fertility. However, prolapse of the uterus as a consequence of the absence of normal uterine suspension can occur very early, even in teenagers. Male patients mostly have a broad and relatively short penis with normal function and orgasm during intercourse. Ejaculation and fertility are hampered.

Psychosexual development in the exstrophy/epispadias patients is in need of professional support by psychologists, urologists and gynecologists with special knowledge in the field of gender identity and congenital malformations of the urogenital tract.[11,35-49]

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Female Epispadias Repair

Female epispadias repair: a new 1-stage technique

CHAPTER 7

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Abstract

Purpose: Female epispadias is a rare anomaly. According to the literature it is usually treated with staged procedures, including bladder neck reconstruction, to achieve continence.
We developed a 1-stage surgical technique that offers the possibility of achieving continence and a cosmetically normal appearance of the vulva.
Materials and Methods: We treated 4 patients 4 months to 8 years old. The main point of the technique is to free the urethral plate and bladder neck completely from surrounding tissues. After tubularizing the urethral plate into a urethra modified needle suspension brings the bladder neck and proximal urethra into the intra-abdominal position.
The pelvic floor is then reconstructed between the anterior vaginal wall and urethra. Thus, continence may be attained by intra-abdominal positioning of the bladder neck and proximal urethra as well as by pelvic floor reconstruction.
Results: Of our 4 consecutive cases of primary untreated epispadias, the technique proved successful in 3, while followup is too short in 1. One patient is completely dry and voids without a further procedure. Postoperatively 2 patients with 5 years or more of followup required injection of a bulking agent at the bladder neck level to achieve continence, including 1 who is damp during the day without the need to change clothes and 1 on clean intermittent catheterization twice daily because post-void residual urine volume causes recurrent uinary tract infection.
Conclusion: The described technique is promising for treating this disabling anomaly.

Introduction

Female epispadias is a rare anomaly, occurring in 1/150,000-300,000 live female births.[1] In the Dutch population this value predicts 1 newborn patient every 2 to 3 years. The anomaly is a type of exstrophy-epispadias complex. Generally patients present because of abnormal external genitalia with a wide anterior gap between the labia majora and a bifid clitoris.

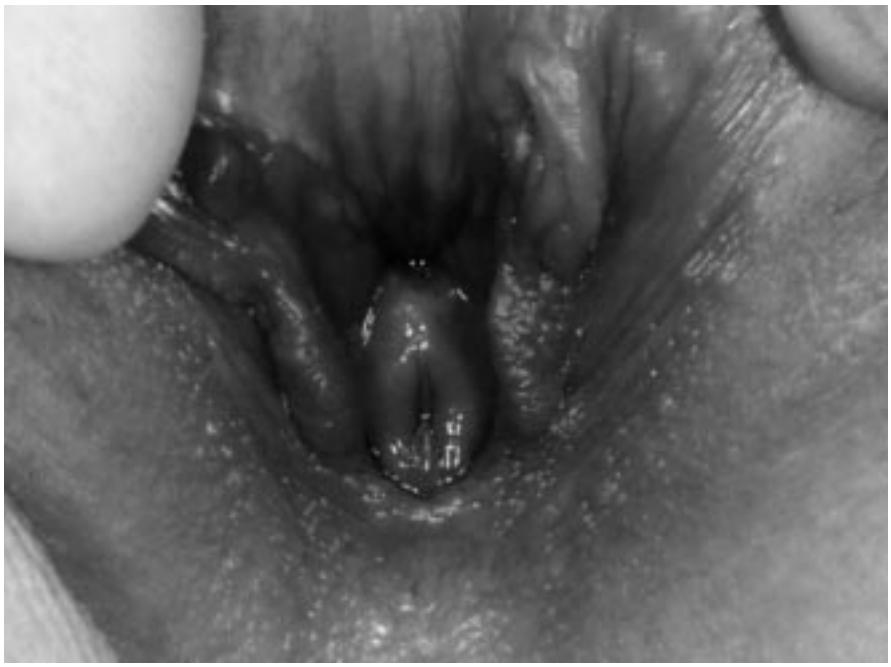

Figure 2: *Female epispadias. A normal hymenal ring, anteriorly the bladder neck, a bifid clitoris and the urethral plate spread out anteriorly between the halves of the clitoris.*

The urethral plate extends between the clitoral halves. In most cases the bladder neck is located at the level of the hymenal ring (fig 1). No urethra and no sphincteral mechanism is present, resulting in complete incontinence that is often accompanied by bladder capacity that is relatively small for age. When overlooked, after birth, female epispadias may present as severe incontinence. Sometimes the conditon is wrongfully considred a manifestation of intersex, resulting in a delayed diagnosis and unneccessary psychological distress for the family of the family.

Historically, treatment consists of staged repairs with urethral and vulvar reconstruction at stage 1, followed by bladder neck reconstruction at a later age. These procedures often do not have optimal results with respect to urinary continence. Moreover, clean intermittent catheterization and/or a continent catheterizable stoma on the often augmented bladder

is needed in many cases. We developed a 1-stage procedure that results in a competent urethra, functioning sphincteric mechanism and intra-abdominal bladder neck, providing an optimal chance for primary continence and normal voiding.

Patients and Methods

During an 8-year period corrective surgery was performed in 4 females 4 months to 8 years old with primary complete epispadias, including 3 who presented with intersex and 1 with therapy resistent incontinence. Preoperatively all 4 patients underwent ultrasound of the urinary tract, voiding cystourethrography and a urodynamic study. One patient had bilateral vesicoureteral reflux and 1 underwent emergency surgery at the age of 4 months because of recurrent bladder prolapse.[2] In 3 cases the timing of surgery was elective, while in 1 referred immediately after birth we chose to perform the operation at the age of 10 months. Two girls referred late underwent surgery after urinary tract evaluation. The procedure began with cystoscopy and colposcopy, followed by excision of the urethral plate between the clitoral halves, leaving the urethral plate attached to the bladder neck (fig 2, A). The bladder neck was then completely freed from attachments to the anterior vaginal wall and the symphysis up to the level of the trigone, resulting in complete mobility of the bladder outlet and attached urethral plate. The urethral plate was tubularized into a urethra (fig. 2, B and C). The bladder neck and proximal urethra were brought into the intra-abdominal position by a modified needle suspension technique. Four 16 gauge intravenous cannulas wee passed from 2 small inguinal incisions retropubically into the operating field. A polypropylene suture was fixed at the level of the bladder neck at the 3 and 9 o' clock positions. Each end of the suture was passed upward through the intravenous cannulas into the inguinal region. The intravenous cannulas were withdrawn, and the sutures tightened and knotted at the external rectus abdominis fascia, bringing the bladder neck and proximal urethra into the intra-abdominal position with the extended neourethral orifice at the anterior side of the hymenal ring. Subsequentely

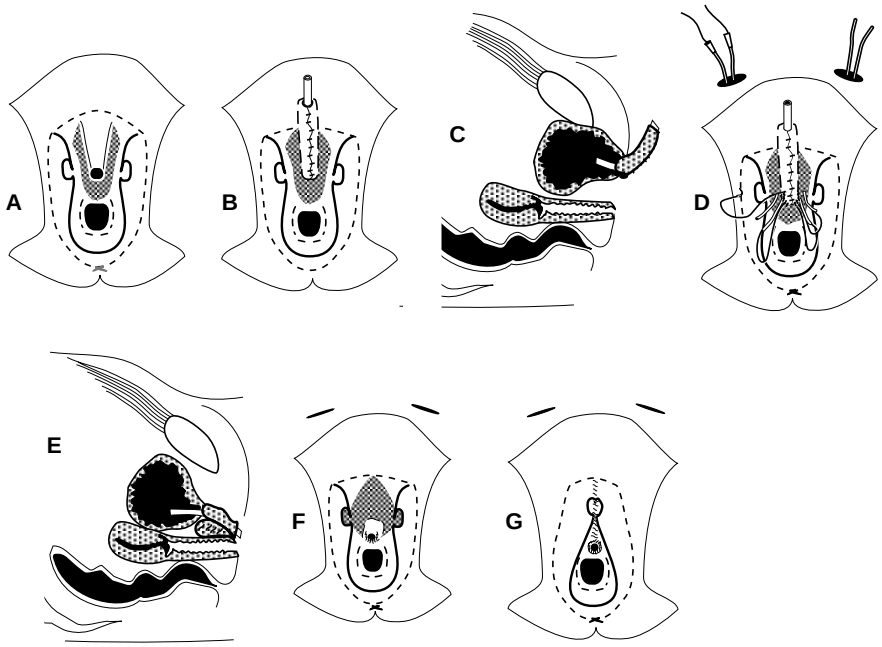

Figure 2: A. *The urethral plate is freed from surrounding skin. The plane between the anterior vaginal wall and the bladder is developed. The bladder neck is freed completely from surrounding tissues, especially the pelvic floor, to ensure free mobility.* ***B.*** *The urethral plate is tubularised around a 10 Fr. silastic tube with interrupted absorbable sutures.* ***C.*** *Sagittal view of the situation after tubularisation of the urethra and mobilisation of the bladder neck.* ***D.*** *Intravenous cannulas , 2 on each side (or 1 with adapted technique) are passed through 2 small suprapubic incisions to the perineal wound, taking care to pass through the gap made in the pelvic floor without penetrating the bladder. After endoscopic inspection to make sure that the bladder has not been perforated by the cannulas, a suture is passed through the bladder neck and both ends are back-fed through the cannulas. Subsequently the cannulas are removed. By pulling the sutures the bladder neck is brought into an intra-abdominal position. The sutures are knotted upon the fascia of the external abdominal muscle.* ***E.*** *Sagittal view. The pelvic floor is closed between the anterior vaginal wall and the urethra. When neccessary, in cases with a split symphysis, the pelvic floor is closed anteriorly as well. These sutures are very important. This is what makes the patient continent.* ***F en G*** *The urethra is sutured to the hymenal ring posteriorly, to the surrounding vulvar tissues medially and anteriorly. The clitoridal halves are denuded medially and fixed together with 7.0 absorbable sutures. The abundant tissue between the labia majora is excised, bringing the labia together in the mid-line.*

the pelvic floor was identified visually left and right of the urethra, and closed in the midline between the anterior vaginal wall and the urethra (fig 2, D and E). These sutures make continence possible. In 1 patient with a large gap between the pubic bones complete approximation of the pelvic floor was not possible in the midline behind the bladder neck, resulting in a 5 mm. gap between the muscles. The urethral end was sutured posteriorly at the level of the hymenal ring laterally and anteriorly at the labia minora. The procedure was completed with external genital reconstruction. The clitoral halves were fused by denuding them of medial epithelium and connecting them with 7 or 8-zeropolyglycolic acid sutures. The anterior distance of the labia majora was decreased by excising a lozenge of skin between the labia and suturing the defect longitudinally (fig. 2, F and G). Postoperatively bladder drainage was maintained for 10 days with a 10Fr transurethral single lumen catheter without a balloon. All 4 patients were on antibiotic prophylaxis during the 10-day period, consisting of 2 mg./kg. trimethoprim once daily and 0.4 mg./kg. oxybutinin daily given in 3 doses. One patient with bilateral grade 2 to 3 vesicoureteral reflux underwent simultaneous ureteral reimplantation through a separate transverse incision. Blood loss was negligible in all cases.

Results

Convalescence was unremarkable in all patients. When voiding, 2 patients had, temporarily had insufficient bladder capacity for age, producing urge, stress and overflow urinary incontinence. They eventually required bladder neck injection therapy with silicone granules in povidone. Technically bladder neck injection with 2 ml. of bulking agent at the 6 o'clock position was not more difficult than injection therapy for other indications. We routinely position the needle in the bladder neck by suprapubic puncture under direct vision via the transurethrally introduced cystoscope.
Each patient achieved sufficient bladder capacity for continence. One girl is on clean intermittent catheterizarion twice daily to prevent urinary tract infection and 1 suffers dampness during sports without the need to change clothes. One patient who underwent surgery at age 10 months is

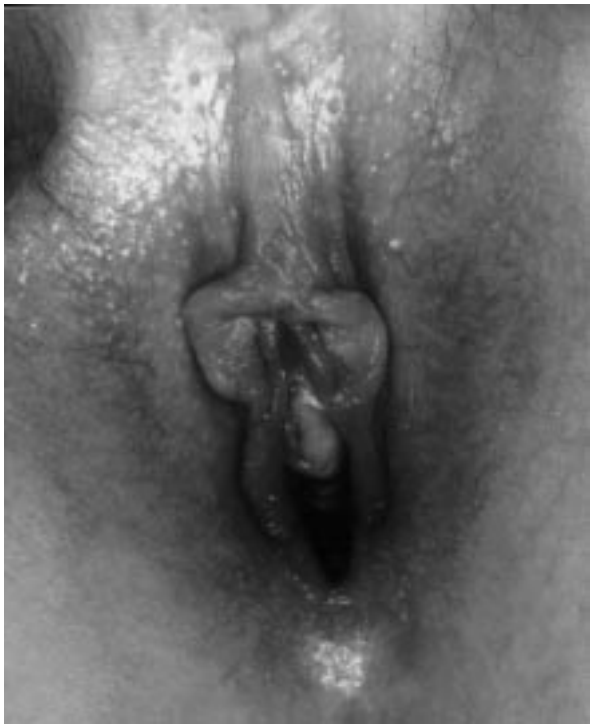

Figure 3: *Photograph 6 months after reconstruction. The result is a completely normal appearance of the external genitalia, a competent bladder neck and urethra in an intra-abdominal position and a functioning pelvic floor surrounding the proximal urethra.*

continent and voids normally since the operation. Urodynamic and radiological studies of the lower urinary tract were completely normal. The remaining patient has a follow-up too short to be conclusive at age 10 months. Three patients had no morbidity after surgery, while 1 had recurrent urinary tract infections. The tabel shows urodynamic results.
Vesicoureteral reflux was graded on a scale of 0 to 5, and compliance was considered normal when bladder capacity in ml. divided by bladder pressure in cm. water was greater than 10 at the end of the filling phase. Cosmesis is excellent in all cases (fig. 3). Followup ranges from 6 months to 6 years.

Discussion

Few reports exist in the literature of relatively large series of female epispadias with 6 to 12 patients collected for many years. Welch included 50 females in a review article.[3] Kramer and Kelalis described 12 patients, including 10 who were completely continent after bladder neck reconstruction.[4] Hendren reported on 6 patients treated with an abdominoperineal approach that included bladder neck reconstruction, of whom 3 were continent.[5] Gearhart et al described 11 females who underwernt staged reconstruction, including bladder neck reconstruction in the majority, in 3 of whom previous procedures failed.[6]

Five patients were completely dry, 3 were dry up to 3 hours during the day and 1 was dry for 1 to 3 hours, resulting in an overall daytime continence rate during of 87,5%. Mollard et al reported on 10 patients, including 8 with complete continence and some leakage in 2.[7] Four patients underwent staged repair and 6 an unstaged procedure consisting of genitoplasty combined with bladder neck reconstruction.

To achieve continence an intra-abdominal position of the bladder neck and proximal urethra is mandatory. The previously described techniques from the literature achieve this goal by bladder neck reconstruction that extends the urethra into the bladder. These methods do not use the complete length of the urethral plate to construct a competent urethra. The 1-stage technique that we developed has certain advantages. No major bladder neck surgery or ureteral reimplantation is needed in the absence of vesicoureteral reflux. The bladder neck and proximal urethra are brought into the intra-abdominal position by a relatively simple procedure that provides the optimal chance for primary continence without hampering possible subsequent bladder neck reconstruction, when needed. However, long-term results cannot be predicted in this extremely rare anomaly with severe consequences to the continence mechanism. The rarity of the abnormality prohibits evaluating the efficacy of surgical intervention in large groups of patients.

Conclusion

Our newly developed procedure involving 1-stage urethral reconstruction, intra-abdominal positioning of the bladder neck and proximal urethra with a modified needle suspension technique, and pelvic floor reconstruction appears to be promising for treating female epispadias. More experience is needed to prove the superiority of this technique for this rare anomaly. Recently this method was adopted in several pediatric urological centers, helping to prove its value for treating these difficult cases. Moreover, the technique has been tried by others as a secondary procedure in girls with epispadias after primary exstrophy closure. The results of these cases are not yet available.

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CHAPTER 8

Discussion

In **Chapter 1** we have shown that the list of structural causes for pediatric incontinence as described in current urological textbooks is incomplete. Especially the absence of post-urethrotomy incontinence and congenital stress-incontinence constitutes an unwanted bias in the management of these children.

A well-structured training program for pediatric incontinence as described in **Chapter 2** is an indispensable tool in dealing with pediatric incontinence based on structural anomalies. A team consisting of urotherapists and physical therapists experienced in pelvic floor therapy is needed, supported by child psychology, pediatrics, and pediatric urology. The training program is needed before deciding on surgery for continence. Finally, after a successful surgery, a final training course is often needed to adapt bladder and sphincter function to the newly created anatomy.

The primary complete reconstruction of ectopic ureteroceles, as described in **Chapter 3**, is superior to the multi-staged approach. The urodynamic outcome in neonates is completely comparable to the outcome in children operated after the age of 1 year. This is an important conclusion that brings to an end the long-standing discussion in pediatric urology about the risks of neonatal bladder surgery for subsequent deterioration of bladder function.

Awareness of post-urethrotomy incontinence, the topic of **Chapter 4**, does hardly exist, despite the fact that D. Innes Williams already in 1973 published a prospective study that pointed out the futility of trying to treat dysfunctional voiding with surgical procedures[1] .It will become clear in the coming years whether this Chapter describes a rare entity or the tip of an iceberg. After trial and error we have designed an operative procedure with scar excision, urethral reconstruction, and colposuspension that appears to be superior to other approaches, for this moment.

In **Chapter 5**, the one-stage neonatal reconstruction in intersex patients with an urogenital sinus, raised as girls, has been proven to reduce lower

urinary tract dysfunction in this patient group to zero. A plenary interactive panel discussion at the combined Annual AAP/ESPU Meeting at Tours, June 2000 has learned that more then 20% of pediatric urologists that deal with this pathology have adopted our policy to operate these children neonatally. Whether this approach is also favourable for the gender identity of these children is still under investigation.

The one-stage epispadias repair that we have developed for girls born without a urethra and without a sphincteric mechanism, described in **Chapter 7**, appears very promising in diminishing lower urinary tract morbidity and psychological distress. The true value of this procedure needs to be proven after long-time follow up, when other departments also have been able to obtain similar results. A few colleagues have started doing this procedure and, from personal communication, seem to be happy with the first results but follow-up is too short to draw final conclusions.

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Summary

Children with urinary incontinence and/or recurrent urinary tract infections can be devided into 3 groups. Firstly children with rare major congenital defects such as bladder extrophy and epispadias with complete absence of a sphincteric mechanism at birth. These children are completely dependant on surgery to reach urinary continence. The quality of life for these children is determined by the quality of the surgical management. Without optimal treatment urinary incontinence and impairment of sexual life with important health-care consumption will take place. The total number of new-borns in this group each year in our country is approximately 15. Besides surgical treatment these patients need multidisciplinary care from both the team of urotherapists to accomplish an optimal way of voiding and the team for ambiguous genitalia to guide the psychosexual development with deformed genitalia. The urotherapists teach the children a normal micturition pattern by explaining the mechanisms of voiding, by motivating of the child, by biofeedback training and by psychological guidance, both in dealing with incontinence and by dealing with fear for clean intermittent catheterisation, when needed.

The second group of patients consists of children with congenital or acquired anomalies of the urinary tract that compromise the patency of the bladder neck and the urethra. These children present with a mix of incontinence based on leakage and incontinence based on bladder/sphincter dysfunction. For these children the surgical procedure is not neccessarily always the one and only good solution. When surgery is performed in these children, it should be done with an open eye to bladder and sphincter function. When the indication for surgery is uncertain, a team approach with dedicated urotherapists and physical therapists is mandatory to try to reach urinary continence by biofeedback training and pelvic floor reeducation. The indication to operate on these children can only rise to the surface when thorough training of a motivated child has proven to be insufficient. Again, after surgery, the team approach will often be needed to reeducate a grinded bladder/sphincter dysfunction by training. The total numbers in this group are also relatively small and diffi-

cult to obtain because of the fact that the group consists of a protean mix of anomalies such as ectopic ureterocele reaching into the urethra, ureteral ectopy with congenital bladder neck insufficiency, true congenital stress incontinence, urogenital sinus in ambiguous genitalia etc.

The third group of patients is huge and consists of approximately 7% of all 7 year old girls suffering from non-neurogenic lower urinary tract dysfunction resulting in day-time wetting and/or urinary tract infections. The basic problem in the vast majority of this group is a pelvic floor dyscoordination, the children lack the ability to relax the puborectalis muscle when voiding and when passing stools. Instead of the puborectalis muscle they use all the surrounding muscle groups as an emergency break to retain urine when bladder overactivity occurs. The pertinent result of long-standing bladder/sphincter dysfunction is a lack of awareness of the filling state of the bladder and the rectum, leading to urinary incontinence and fecal soiling. The treatment of these children is straightforward and consists of motivating of the child to comply to the training program, explanation of the mechanism of the pelvic floor dysfunction and pharmacological treatment of bladder instability, urinary tract infections and fecal constipation. The treatment of this problem cannot be done by a single physician. A team with pediatric urologists, pediatricians, preferably with nephrologic experience, urotherapists, physical therapists and a psychologist is needed. Specific experience with pediatric urodynamic studies is mandatory for most cases.

This thesis tries to bridge between functional and structural non-neurogenic incontinence and to give insight in the surgical options. Children with anatomically based bladder neck and urethral insufficiency often present with the same symptoms as children with genuine non-neurogenic functional incontinence. Bladder neck insufficiency causes urine to leak into the proximal urethra and urine in the proximal urethra is one of the triggers of the micturition reflex with detrusor contraction and sphincteric relaxation as a result. Children with an insufficient bladder neck are constantly counteracting this micturition reflex by pelvic floor contraction and can present with typical symptoms of true dysfunctional voiding. Constant awareness of these facts is mandatory for all the members of

the team. The urologist should be very meticulous in the endoscopic and ultrasound judgement of the anatomy of the bladder, the bladder neck and the urethra. Video-urodynamic studies must be interpreted with the assumption that derailment of the Wolffian duct can be a cause of the incontinence. The urotherapist must know that some children cannot be cured by biofeedback treatment and eventually will need an operation to get rid of the urinary incontinence. The physical therapist is the only person that can specifically teach the child to use the puborectalis muscle instead of all the muscles around. This is an underestimated task because many physical therapists do not realise that this result can only be obtained by invasive treatment with rectal examination and biofeedback training by anal balloon expulsion. Meanwhile, the physical therapist must be aware of the rare neurogenic anomalies that sometimes can be overlooked by urodynamic studies. The psychologists and all the members of the team should realise the psychological impact of incontinence, sometimes combined with malformations of the external genitalia, on the behaviour of a child. Any child with a handicap copes with the disability by eleminating the concerning part of the body from the self-image. The resulting ignoring behaviour of the child that should be interpreted as a natural defence mechanism to cope with the disability is commonly misinterpreted as a behavioural disorder by parents and therapists. Finally one must realise that also children with structural anatomically based incontinence as in bladder extrophy and epispadias as well as children with ambiguous genitalia often need to be treated by the team of urotherapists and physical therapists to accomodate the function of the restored anatomy to the daily life situation.. Moreover, in many cases these children need specific guidance for gender identity and psychosexual development by a team that is specialised in dealing with ambiguous genitalia in childhood.

The chapters in this thesis describe functional incontinence and many aspects of structural incontinence in childhood. The main objective in the treatment of the different patient groups has been to minimise morbidity and health care consumption in patient groups that historically present with chronic lower urinary tract problems. Our approach to ectopic ureteroceles, in our opinion, reduces the morbidity as much as possible. More research in this field by other groups will be neccessary to prove this fact.

The Otis-urethrotomy victims, when untreated, are bound to life-long urinary incontinence with many frustrating events by failing therapies. Our approach for girls with ambiguous genitalia has been able to reduce the work-load of the urotherapists in this group of patients to zero, in contrast to their work load in the relatively large group of patients that has been treated by historic schedules resulting in non-neurogenic bladder/sphincter dysfunction and/or urinary tract infections in the majority of the patients. The treatment of female epispadias as presented in this thesis appears to reduce the morbidity and the number of surgical procedures for the patients.

The social significance of the treatment of pediatric pelvic floor dysfunction and structural pediatric urinary incontinence is high. Properly structured treatment of pelvic floor dysfunction in all the age groups should be able to reduce health care costs in the Netherlands by approximately 150.000.000 Euro a year on the long run and will enhance the quality of life for many patients. Investments to reach this goal in the pediatric age group are relatively small, approximately 5.000.000 Euro a year should be able to realise the needed structure. Several research programs in this field are conducted by the department of pediatric urology of the UMCU. The results of the multicenter EEC funded European Bladder Dysfunction study, initialised and guided in Utrecht by dr. J.D. van Gool will be pubished soon. This has been the first prospectively randomised study into this important group of patients with recurrent urinary tract infections and urinary incontinence. The second prospectively randomised study into these patients, using home- flowmeter biofeedback treatment is on the run. This study, done by the department of pediatric urology, is funded by Zorg Onderzoek Nederland (ZON). Other current research projects of the department are studies into the dynamic behaviour of the pelvic floor, especially the puborectalis muscle, by ultrasound imaging and studies into the relationship between hypermobility of the joints and pelvic floor dysfunction. Next to the non-neurogenic incontinence the department of pediatric urology is constantly working on the urological quality of life for patients with a neurogenic bladder/sphincter dysfunction.

The important conclusion of the thesis is that proper multidisciplinary surgical and behavioural care for pediatric incontinence pays off. The main advantage of proper care is an increase of quality of life for the patients. This includes quality of psychosexual development. Secondary advantages are a significant drop in health care costs, including costs for absorption of urine, costs of pharmacological treatment of the bladder and urinary tract infections, even costs of renal failure, dialysis and kidney transplantation.

Samenvatting

Kinderen met urine-incontinentie en/of urineweginfecties kunnen worden onderverdeeld in 3 groepen. (de 4de groep, kinderen met een neurologische stoornis en daardoor blaasfunctieproblemen wordt in dit proefschrift buiten beschouwing gelaten).
Groep 1 bestaat uit kinderen met zeldzame en indrukwekkende aangeboren afwijkingen zoals een open blaas en een epispadie, beiden met volledig afwezige kringspier en plasbuis bij de geboorte. Deze kinderen zijn volledig afhankelijk van operaties om continent te worden. De kwaliteit van leven voor deze groep staat of valt primair bij de kwaliteit van de chirurgische aanpak. Zonder optimale behandeling zijn deze kinderen incontinent en hebben zij een sexuele handicap door een onvolwaardig uitwendig genitaal met als gevolg een belangrijke consumptie van gezondheidszorg. Deze groep bestaat uit ongeveer 15 nieuwe pasgeborenen per jaar in Nederland. Naast de urologische behandeling moeten deze kinderen multidisciplinair begeleid worden door een team van incontinentietherapeuten om met een geconstrueerd blaas/kringspiermechanisme te kunnen plassen en door een team gespecialiseerd in omgang met kinderen met een onvolwaardig genitaal om een zo goed mogelijke psychosexuele ontwikkeling te garanderen.

Groep 2 bestaat uit kinderen met aangeboren of verworven afwijkingen aan de urinewegen die de kwaliteit van de blaashals en de urethra (plasbuis) aantasten. Deze kinderen presenteren zich met een mengelmoes van incontinentie door urinelekkage en door een gebrekkige samenwerking tussen blaasspier en kringspier. Blaashalsinsufficientie leidt tot het constant aanwezig zijn van urine in de plasbuis en daardoor constant de neiging om te gaan plassen, tegengewerkt door aanspannen van de bekkenbodemspieren. Dit mechanisme maakt het onderscheiden van deze kinderen van die in groep 3 vaak uitermate lastig. Operatieve ingrepen zijn bij deze kinderen niet altijd de beste optie. Als ze toch geopereerd moeten worden dient dit te gebeuren met veel inzicht in de functie van blaas en kringspier. Als de indicatie voor operatie onduidelijk is moet getracht worden de kinderen met training te behandelen binnen het team van incontinentietherapeuten en fysiotherapeuten. De indicatie tot chirurgisch

ingrijpen kan pas worden gesteld nadat zeer gedegen conservatieve therapie kansloos is gebleken. Na een operatie is dit team opnieuw noodzakelijk om de vaak diep gewortelde stoornis in blaas- en bekkenbodemfunctie door training aan te passen aan de nieuw gecreeerde anatomische situatie. Het aantal patienten in deze groep is relatief klijn en moeilijk exact te bepalen omdat de groep bestaat uit een mix van kinderen met ectope ureteroceles, met ectope ureteren en congenitale blaashalsinsufficientie, met echte aangeboren stress-incontinentie en met een sinus urogenitalis bij ambigue genitalien.

Groep 3 is enorm en bestaat uit ongeveer 7% van de 7-jarige meisjes met niet-neurogene blaas/sphincter dysfunctie met als gevolg urine-incontinentie overdag en recidiverende urineweginfecties. Bij jongens komt dit probleem in beperkte mate voor. Het basale probleem bij deze kinderen is een gestoorde coordinatie van de bekkenbodemspieren: de kinderen kunnen hun bekkenbodem niet ontspannen tijdens het plassen en bij de stoelgang. In plaats van de kringspier gebruiken zij alle omgevende spieren als noodrem bij heftige plasdrang die optreedt als de blaas krampt. Het obligate resultaat van deze situatie is verlies van het gevoel van blaasvulling en endeldarmvulling met als gevolg urine-incontinentie en verlies van vegen ontlasting. De behandeling van deze kinderen is goed omlijnd en bestaat uit motiveren van het kind om het probleem aan te pakken in een trainingsprogramma, uitleg van het mechanisme aan ouders en kind en tenslotte farmacologische behandeling van urineweginfecties, blaasinstabiliteit en obstipatie. Een team van kinderurologen, kinderartsen, liefst met nefrologische expertise, incontinentietherapeuten, fysiotherapeuten en een psycholoog is hiervoor nodig. Specifieke urodynamische ervaring is in veel gevallen onontbeerlijk.

Dit proefschrift probeert een brug te slaan tussen functionele en structurele, door anatomische afwijkingen veroorzaakte, incontinentie. Daarnaast beschrijft het een aantal operatieve mogelijkheden om structurele incontinentie te behandelen. Kinderen met anatomisch bepaalde incontinentie hebben vaak dezelfde symptomen als kinderen met functionele incontinentie doordat zij voortdurend trachten om urinelekkage tegen te gaan met de bekkenbodemspieren. Alle leden van het behandelteam moeten

goed op de hoogte zijn van dit fenomeen. De kinderuroloog moet nauwlettend echografisch en endoscopisch de anatomie van blaas, blaashals en plasbuis in kaart brengen. Video-urodynamisch onderzoek moet beoordeeld worden in de wetenschap dat anatomische problemen kunnen bestaan. De insontinentietherapeut moet beseffen dat sommige vormen van incontinentie therapieresistent zijn tegen training en uiteindelijk chirurgisch moeten worden behandeld. De fysiotherapeut is de enige die het kind kan leren om specifiek de musculus puborectalis te gebruiken in plaats van alle spieren daaromheen. Dit is een sterk onderschatte taak omdat weinigen zich realiseren dat dit alleen adequaat kan met rectaal onderzoek en met anale ballon training. Daarnaast is de fysiotherapeut degene die occulte neurologische oorzaken van incontinentie soms beter detecteert dan alle andere onderzoeken. De psycholoog en alle andere teamleden moeten besef hebben van de invloed van incontinentie op het gedrag van het kind zeker als dit ook nog gepaard gaat met afwijkingen aan de geslachtsdelen. Ieder kind met een handicap gaat hiermee om door het betreffende lichaamsdeel uit het zelfbeeld te snijden. Het ontwijkende en negerende gedrag dat daaruit resulteert is een natuurlijk verdedigingsmechanisme van het kind dat helaas veelvuldig ten onrechte als een gedragsprobleem wordt geduid met onzorgvuldig psychologisch en pedagogisch geweld door onervaren therapeuten en slecht geinformeerde ouders. Verder is van belang te beseffen dat kinderen met ernstige structurele anatomische afwijkingen van urinewegen en uitwendig genitaal veelal uitgebreide begeleiding nodig hebben van het gehele team om de nieuwgecreeerde anatomie aan te passen aan het dagelijks leven. Bovendien hebben deze kinderen extra begeleiding nodig om te leven met een gehandicapt genitaal, zowel ten aanzien van geslachtidentiteit als ten aanzien van hun psychosexuele ontwikkeling. Hiervoor is een multidisciplinair team nodig gespecialiseerd in het omgaan met ambigue genitalien.

De hoofdstukken in dit proefschrift beschrijven niet-neurogene functionele incontinentie en veel aspecten van structurele incontinentie op de kinderleeftijd. Voornaamste doel is steeds geweest om de morbiditeit van de urinewegen bij deze kinderen zo veel mogelijk te beperken en om consumptie van gezondheidszorg te beperken. De gepresenteerde aanpak van kinderen met ectope ureteroceles reduceert de morbiditeit zo veel als

mogelijk. Bovendien bewijzen we in deze groep dat neonatale chirugie van blaas, blaashals en plasbuis de blaasfunctie niet schaadt. De kinderen met incontinentie na urethrotomie lijden onbehandeld aan levenslange incontinentie met veel psychische frustratie door falende conservatieve therapie. De reconstructie van de plasbuis na excisie van littekens is veelbelovend voor deze moeilijke patientengroep. De neonatale reconstructie van uit-en inwendig genitaal van meisjes met ambigue genitalien met sinus urogenitalis heeft de behoefte aan zorg van het incontinentieteam tot vrijwel nul gereduceerd. De aanpak is internationaal veel nagevolgd. Het goede resultaat staat in schril contrast tot de grote hoeveelheid urinewegproblemen bij meisjes met ambigue genitalien die met gestageerde technieken zijn behandeld. De behandeling van meisjes met een epispadie, zoals gepresenteerd in dit proefschrift, blijkt het aantal operaties en de incontinentie optimaal te reduceren.

De maatschappelijke relevantie van goede behandeling van bekkenbodemdysfunctie en structurele incontinentie voor urine op de kinderleeftijd is hoog. Gestructureerde behandeling van bekkenbodemdysfunctie kan op termijn de kosten van de gezondheidszorg in Nederland met minimaal 150.000.000,- Euro per jaar beperken en bovendien de kwaliteit van leven voor veel mensen aanzienlijk verbeteren. De investering voor goede bekkenbodemzorg op de kinderleeftijd kan relatief klein blijven, 5.000.000 Euro per jaar is waarschijnlijk voldoende om de benodigde infrastructuur te realiseren. Het Kinderniercentrum van het WKZ/UMCU voert diverse onderzoeksprogramma's uit op het gebied van urine-incontinentie bij kinderen. De resultaten van de multicenter European Bladder Dysfunction Study, betaald door de EEG en geleid vanuit Utrecht door prof. dr. J.D. van Gool, zullen binnenkort worden gepubliceerd. Dit is de eerste prospectief gerandomiseerde studie naar behandelresultaten bij kinderen met recidiverende urineweginfecties en incontinentie. De tweede prospectief gerandomiseerde studie die het effect van homeflowmetrie training onderzoekt bij deze studie is een eind onderweg. Deze studie, gefinancierd door Zorg Onderzoek Nederland wordt uitgevoerd door de afdeling kinderurologie. Andere lopende projecten van de afdeling zijn een studie naar de dynamiek van de musculus puborectalis met behulp van perineale echografie en een studie naar de relatie tussen hypermobiliteit van gewrichts-

banden en bekkenbodemdysfunctie. Naast het werk aan niet-neurogene blaas/spincter dysfunctie is de afdeling kinderurologie voortdurend op zoek naar verbetering van de urologische kwaliteit van leven voor de grote groep kinderen met een neurogene blaas op basis van spina bifida.

De belangrijkste conclusie van dit proefschrift is dat een gedegen multidisciplinaire chirurgische en gedragsmatige aanpak van urine-incontinentie op de kinderleeftijd zijn geld ruim opbrengt. De grootste winst wordt behaald door een verbetering van de kwaliteit van leven van de kinderen. Daarnaast levert goede zorg een grote vermindering van consumptie van gezondheidszorg op, met besparing op urineopvangmateriaal, medicatie en zelfs op kosten van nierinsufficientie, dialyse en transplantatie.

Dankwoord

Dit boek had nooit geschreven kunnen worden zonder de inbreng van Marianne Vijverberg en Jan van Gool. De inbreng van Marianne is gekarakteriseerd in een editorial comment in The Journal of Urology (S. Shulman, september 2000)met de ultieme lof: zij heeft het inzicht van een kinderuroloog, het geduld van een kinderarts en de tact van een psycholoog. Jan van Gool is de internationale nestor op het gebied van de dysfunctional voiding en vertaalde samen met Marianne de inzichten in urodynamische parameters.
Mijn opleiders zijn belangrijk geweest. Hendrik van der Reijden voor het onnavolgbaar leren van de weg in een buik, daarnaast het leren omgaan met lastige Amsterdamse patienten. Hij heeft gezorgd voor een belangrijke vorming: doe maar gewoon als je staat te opereren, blijf ijzig kalm als alles fout gaat en zoek altijd naar de simpelste oplossing zonder dogmatisch te zijn. De Bossche urologen gaven de kans om dit om te zetten in een grote operatieve ervaring zonder de bestaande dogmata in de chirurgische gedachtenwereld af te dwingen. Rudi Janknegt, zelf uit de vrij strikte Boerema school, accepteerde de methodiek van van der Reijden, ongetwijfeld vaak met krullende tenen, een zeer effectieve methodiek die door sommigen JBF wordt genoemd. Adriaan Smans leerde mij om na te denken over effectiviteit van bewegingen tijdens operaties. Alan Perlmutter gaf alle inzichten in de genitale chirurgie, bij hypospadieen bij jongens en bij genitoplastieken bij meisjes. Bob Jeffs leverde de basisfilosofie voor extrophia vesicae correctie. Bernard Churchill leerde dat je iedere kinderurologische operatieindicatie kunt vangen in een systeem. Roel Scholtmeijer gaf de gelegenheid om ervaring op te doen en leverde veel steun in de beginperiode waarin nogal kritisch werd gekeken naar de Utrechtse kinderurologie door eerdere mislukkingen om de kinderurologie in het WKZ van de grond te krijgen.
De crux van de gecompliceerde incontinentieproblematiek bij kinderen is het team. Dit bestaat op de polikliniek, de functiekamer, de verpleegafdeling en op de operatiekamer. Het team bestaat uit polikliniekmedewerkers, secretariaatsmedewerkers, incontinentietherapeuten, fysiotherapeuten, functieassistenten, verpleegkundigen, operatieassistenten, anesthesieassistenten, anesthesiologen, psychologen en diverse specialisten binnen

en buiten de kinderheelkundige disciplines. Ik hoop dat alle betrokkenen zich realiseren hoe onmisbaar hun inbreng is en dank ze voor de sfeer die altijd positief is geweest.
Dank aan Raymond Donckerwolcke, Jan van Gool en Mark Lilien voor de broederlijk collegiale sfeer in het smeden van snode plannen door de jaren heen. Dank aan Tom Boon en Jan van Gool voor het organiseren van het leggen van het ei en dank aan Cock Schroder (voorzitter) en de leden van de leescommissie om hun tijd te besteden aan de beoordeling hiervan. Tevens dank aan alle medewerkers van de audiovisuele dienst van het UMCU voor hun altijd flitsende service. Met name het tekenwerk van Ingrid Janssen komt in dit proefschrift regelmatig terug. Grote delen van het manuscript zijn taalkundig gecorrigeerd door Andrea Gasten.
Verder een dankwoord in ongebruikelijke richting, de Raad van Bestuur van het UMCU, met name naar Rolf de Folter en Gerlach Cerfontaine. Zij doorbraken na de fusie van AZU en WKZ de hardnekkige patstelling in het WKZ waarin de kinderheelkundige disciplines nooit meedeelden in de eerste geldstroom voor onderzoek. Met name het aanstellen van Cuno Uiterwaal van het Julius Centrum ter ondersteuning van ons onderzoek heeft een onwaarschijnlijke boost gegeven aan onze mogelijkheden om onderzoeksprojecten te starten.
Tenslotte een dankwoord aan het thuisfront dat dit alles blijmoedig heeft laten gebeuren. Feitelijk is dit laatste dankwoord tevens een aanklacht richting Nederlandse overheid die in een tijd van maatschappelijke hoogconjunctuur persisteert in een te laag percentage van het BNP voor de gezondheidszorg waardoor het schrijven van een proefschrift door een clinicus nog steeds in de kleine uurtjes moet gebeuren.

Curriculum vitae

De auteur werd op 15 januari 1952 te Heerlen geboren. Na 5 klassen lagere school op de 'broederschool' en na 6 klassen Gymnasium B op het Bernardinus college werd in 1970 het Belgische staatsexamen Humaniora, afd. Wetenschappen –B en toelatingsexamen voor de universiteit gedaan. De medische studie is dat jaar aangevangen aan de Universiteit Utrecht en afgerond in 1978. Een jaar wachttijd voor de co-assistentschappen tijdens de studie is ingevuld als student-wetenschappelijk medewerker aan het fysiologisch laboratorium en door de Medische Faculteit gehonoreerd met een prijs voor excellent onderzoek.

De urologische opleiding is gestart in 1979 met 3 jaar vooropleiding algemene chirurgie in het Andreas ziekenhuis te Amsterdam, opleider H.J. van der Reijden, chirurg, gevolgd door bijna 3 jaar urologie in het Bosch Medicentrum, opleider prof. dr. R.A. Janknegt, uroloog. De afsluitende kinderurologie stage van januari tot mei 1985 is gevolgd in het St. Radboud ziekenhuis te Nijmegen, opleiders prof. dr. F.M.J. Debruyne en dr. J.D.M. de Vries. In 1985 en 1986 zijn kinderurologische stages van elk 2-4 maanden gevolgd in het Sofia Kinderziekenhuis te Rotterdam, prof.dr. R.J. Scholtmeijer, in het St. Radboud ziekenhuis te Nijmegen, dr. J.D. de Vries, in het Wayne State Children's Hospital te Detroit, USA, dr. A.D. Perlmutter, in het John's Hopkins Hospital te Baltimore, USA, dr. R.J. Jeffs, in het Hospital for Sick Children te Toronto, dr. B. Churchill en in het Great Ormond Street Hospital te Londen, UK, mr. P. Ransley.

De auteur is sinds 1 mei 1985 in dienst van de Medische Faculteit van de Utrechtse Universiteit cq het UMCU als hoofd van de afdeling kinderurologie. In 1986 is de kinderurologische kliniek in het WKZ opgericht en in 1988 is dit uitgebreid tot het Kinderniercentrum WKZ in nauwe samenwerking met de kinderarts-nefrologen dr. J.D. van Gool en prof.dr. R.A.M.G. Donckerwolcke. De afdeling kinderurologie is sinds die tijd uitgegroeid tot de grootste afdeling in Nederland en een van de groten in Europa. Het concept van het Kinderniercentrum als hecht samenwerkingsverband van kinderurologie en kindernefrologie is een Utrechtse uitvinding die nationaal en internationaal veel navolging heeft gekregen.

De afdeling kinderurologie in het WKZ/UMCU heeft internationaal een voortrekkersrol in de behandeling van incontinentie bij kinderen zowel

door neurogene oorzaken (spina bifida) als door niet-neurogene en structureel anatomische redenen en daarnaast op het gebied van de behandeling van kinderen met ambigue genitalien (intersex).

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STELLINGEN

1. De vagina doorhaal procedure en genitoplastiek van ambigue genitalien bij kinderen die als meisje worden opgevoed kan het beste gebeuren in de eerste 2 levensweken (dit proefschrift).
2. De doelstelling van de correctie van extrophia vesicae is het bereiken van continentie en spontane mictie door middel van de primaire correctie. Het intra-abdominaal brengen van de blaashals en de proximale urethra is een essentiele voorwaarde om urine-continentie te krijgen bij correctie van extrophia vesicae en epispadie (dit proefschrift).
3. Uitgebreide operatieve correcties van de blaas en de blaashals bij neonaten levert geen aantoonbare schade op aan de urodynamische eigenschappen van de lagere urinewegen (dit proefschrift).
4. Het gebruik van het OTIS-urethrotoom in de proximale urethra van meisjes met een bekkenbodemdysfunctie is obsoleet en dient als een kunstfout te worden beoordeeld (dit proefschrift)
5. Degelijke bekkenbodemzorg voor alle leeftijdsgroepen kan in de toekomst de gezondheidszorg op jaarbasis minstens 150.000.000 Euro besparen met minimale investering.
6. Het misbruik van de term enuresis bij incontinente meisjes en jongens is nog steeds verantwoordelijk voor veel psychische schade bij kinderen.
7. Het is onterecht dat er in Nederland wel wordt gediscussieerd over de religieuze circumcisie bij meisjes en niet over die bij jongens.
8. Medische faculteiten in Nederland claimen al jaren op papier dat excellente patientenzorg ondersteund en beloond moet worden. Uit niets blijkt dat dit voornemen ergens geimplementeerd wordt.
9. Diverse overheidsmaatregelen hebben ertoe geleid dat er een groeiende stroom patienten is naar kinderheelkundige centra. Dit betreft vooral Gezondheidsraadrapporten en de Bigwet. Daarnaast zijn jonge Nederlandse urologen slecht voorbereid op kinderurologische problematiek en werkt de druk van patientenverenigingen in de richting van een centrum. Dit leidt tot ernstige capaciteitsproblemen binnen de kinderurologische centra. Het dilemma voor de kinderurologie is nu: of de perifere collegae bijscholen of de kinderurologische capaciteit vergroten.
10. De in stelling 9 genoemde ontwikkelingen binnen de gezondheidszorg in de laatste 12 jaar hebben geleid tot een ernstig capaciteitsprobleem in de kinderheelkundige vakken. Binnen nu en 5 jaar is een verdubbeling van de capaciteit van de kinderheelkundige centra noodzakelijk om een basale kwaliteit van zorg te kunnen garanderen.
11. Het is triest om vast te moeten stellen dat in deze tijd van maatschappelijke hoogconjuctuur de kwaliteit van de zorg voor het zieke kind achteruit gaat.
12. Een ernstige omissie van de Nederlandse overheid is dat de fietshelm voor kinderen tot 12 jaar nog steeds niet wettelijk verplicht is. Eenzelfde plicht tot bescherming moet overwogen worden voor gebruikers van skeelers in alle leeftijden.
13. Sinds de massale overstap van ouders op z.g. hygienische doekjes bij het schoonmaken van anus en genitaal tijdens luierverwisseling is er in Nederland geen baby meer te vinden zonder geirriteerde hof in die regio. Daarnaast induceert het gebruik van deze doekjes specifieke mictieklachten bij meisjes door vulvovaginitis. Het gebruik van deze doekjes dient door de GGD's ontraden te worden. Water is beter en goedkoper.
14. De nieuwe honoreringsregeling voor Academisch medisch specialisten zal zonder aanpassing van het instroom-salaris voor jonge specialisten tot een markt-conform inkomen binnen 2 jaar resulteren in het faillissement van de 8 academische ziekenhuizen in Nederland.