Incontinence In The Girl Child-Approach To Diagnosis And Treatment

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The approach to girls with incontinence differs from that in adults because the spectrum of problem is different, the urinary tract is in the process of maturing, a variety of environmental and household influences affect lower urinary tract function at this impressionable age, and because certain treatment modalities are difficult to institute in a small child. Urinary incontinence is a common problem encountered in practice. This discussion will deal with the etiology, presentation, diagnostic approach and treatment principles for incontinence in the girl child. A brief outline of the normal process of development of lower urinary tract function is necessary to understand the pathophysiology.

Normal Continence

'Incontinence' is the norm in all newborn babies. The bladder empties about 20 times in a day without volitional control by means of a local spinal reflex with coordinated sphincters. At this age unstable detrusor contractions are normal and often initiate micturition. Around 2 years of age, the child develops the ability to sense fullness of the bladder although voluntary postponement may still not be possible. In this transition phase, the child tries to become socially continent by a voluntary contraction of the pelvic floor and sphincter. This 'dyssynergic' continence is however soon replaced by a central inhibition of the bladder as the nervous system matures. The usual sequence of continence is-nighttime bowel control, daytime bowel control, daytime urinary control and finally nighttime urinary control. Girls tend to achieve continence somewhat earlier than boys. Most children achieve all these steps before the age of 4 years (Rushton, 1995). It is important to understand these pathogenic mechanisms since abnormalities in the development phase can lead to functional voiding disorders.

Etiology and Presentation of Urinary Incontinence in Girls

Incontinence is not a disease but a symptom of underlying disease the cause of which may or may not be obvious. Incontinence in girls can occur due to a variety of reasons (Table 1). These problems can conveniently be classified into 3 groups: anatomic, neurogenic and functional problems in neurologically normal children (Himsl and Hurwitz, 1991).

Table 1 Classification of Urinary Incontinence in girls.

Anatomic

1. Bypass of the sphincteric mechanism-ectopic ureter
2. Abnormal storage or sphincteric mechanisum-exstrophy, epispadias, congenital short urethra
3. Outlet obstruction with overflow-ectopic ureterocele, distal urethral stenosis

Neurogenic- myelodysplasia, lipomeningocele, intradural lipoma, tight filum terminale, dermoid cyst or sinus, sacral agenesis, spinal trauma, cerebral palsy.

Functional- nocturnal enuresis, detrusor instability, non neurogenic neurogenic bladder, lazy bladder, giggle incontinence

Anatomic causes

Anatomic causes are far less common than functional problems but they need to be excluded in every child who comes with incontinence. Anatomic causes occur because of a structural problem in the storage mechanism. This may affect the bladder such as exstrophy, or the sphincteric mechanism, such as in epispadias and congenital short urethra. The normal continence mechanism is bypassed by the ectopic ureter. Anatomic

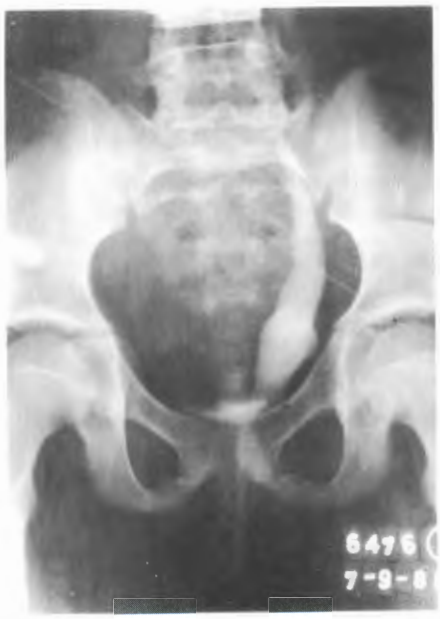

Fig 1.: 6 - year old with urinary incontinence due to left ectopic ureter opening at the vaginal vestibule. Poorly functioning unit not seen on IVP. Antegrade pyelography performed. Underwent left nephrectomy. (Courtesy Dr. Rama Subba Rayudu, Radiologist, Medwin Hospital)

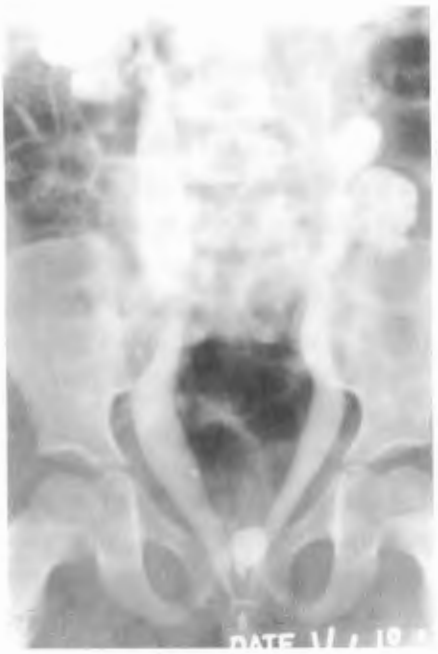

Fig 2. 7 - year old girl with urinary incontinence due to bilateral single system ectopic ureters, a rare form of ectopia. Right ureter opening at the vulva, left in the distal urethra. The bladder is rudimentary and filled by retrograde trickle of contrast. Marked bilateral hydronephrosis is noted. This requires creation of a neobladder with bilateral ureteric reimplantation. *(Courtesy Dr Rama Subba Rayudu, Radiologist, Medwin Hospital)*

incontinence may also occur due to bladder outlet obstruction with overflow such as ectopic ureterocele obstructing the urethra. Some of the important conditions are described.

Ectopic Ureter

A ureter that opens anywhere except on the trigone of the bladder is termed as ectopic (Figures 1 &2). In girls, the usual sites are bladder neck, proximal or distal urethra, vagina, uterus or cervix. The opening may be within or outside the sphincteric mechanism and thus is an important cause of incontinence. It occurs 4 times more often in girls than boys, is more common on the left and is most often associated with duplication of the ureter. In the duplication it is the upper pole moiety which has an ectopic ureter. The classical presentation is continuous incontinence in a girl who otherwise passes urine normally. The condition is most often detected after bladder control has been achieved when the incontinence is noticed. If the orifice is in the vagina, it may present as a vaginal discharge and be mistaken for infection. The child may have recurrent febrile episodes with failure to thrive. If the opening is in the proximal urethra, there may be urge incontinence. (Retik and Peters, 1992)

Exstrophy and epispadias

These are a group of related abnormalities in which there is failure of the normal development of the infraumbilical abdominal wall in varying degrees. In exstrophy, the diagnosis is obvious- there is a lower abdominal defect, exposed bladder, open urethra, bifid clitoris, often a short and stenotic vagina and the labia and mons pubis are divergent. Epispadias may be missed on a cursory examination unless specifically sought for (Wojcik and Kaplan, 1998). Mangement of these conditions requires referral to a specialized pediatric urology unit.

Ectopic ureterocele

Ureterocele is a congential cystic dilatation of the terminal ureter. Ectopic ureterocele implies that the ureterocele opens outside the trigone. When such a ureterocele opens in the urethra it may obstruct it and lead to chronic urinary retention and overflow of urine. The ureterocele may be seen as a systic mass in the perineum. There may be recurrent urinary infection, failure to thrive or intermittent

prolapse of the ureterocele though the urethral orifice (Mandell et al, 1980)

Pseudoincontinence or vaginal voiding may occur in some girls who keep their legs tightly closed together while passing urine such that there is retrograde filling of the vagina(Wojcik and Kaplan, 1998). In this situation urine trickles for sometimes after a void. If suspected, the child should be asked to pass urine supervision with the legs spread apart in the squatting position.

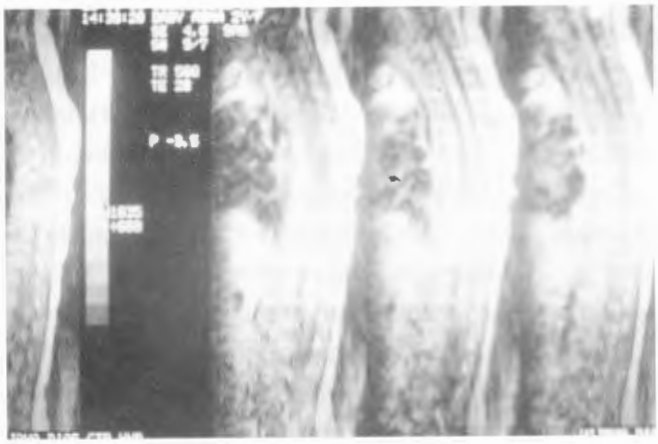

Fig 3a. MRI spine in a 2-year-old girl with dorsal midline sinus going upto spinal cord. The girl underwent excision of the sinus tract. The urinary tract was normal at that time.

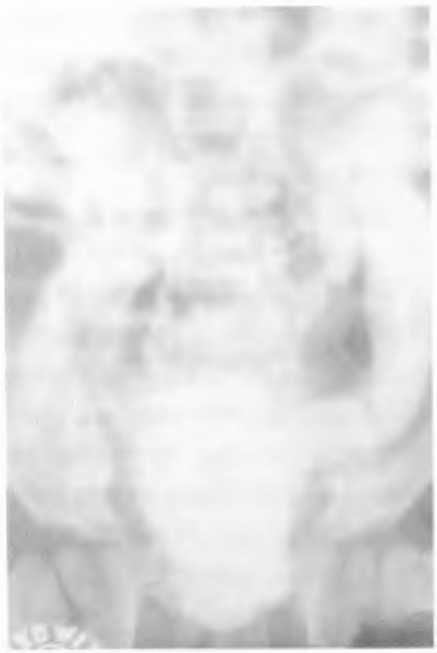

Fig 3b. The same girl presented recently at 4 years age with failure to thrive. Micturating cystourethrogram shows classical changes seconday to detrusor sphincter dyssynergia with gross bilateral hydronephrosis and a trabeculated pinetree-like bladder. The girl was placed on CISC and anticholinergic therapy. She may require augmentation cystoplasty to reduce the intravesical pressures.

Neurogenic causes

Neurogenic causes occur because of an abnormality of bladder innervation such as in girls with spinal cord abnormalities. The commonest cause is myelodysplasia, which occurs in 1 in 1000 children(Bauer, 1988). Myelomeningocele is obvious on examination. However, occult spina bifida may not be immediately apparent and these girls may present during adolescence. This may be suspected clinically by presence of a tuft of hair, lipoma, abnormal pigmentation or sinus in the lower midline over the spine(Figure 3A and 3B). Such girl have incontinence due to a variety of causes such as detrusor sphincter dyssynergia, acontractile bladder or hyperreflexia. The level of lesion is not a good guide to the type of bladder lesion and these children require a formal urodynamic evaluation.

Sacral agenesis may present as urinary incontinence. This is the absence of part or whole of 2 or more sacral vertebrae and is more common in children born to insulin-dependent diabetic mothers. Such children have flattened buttocks and a short, low gluteal cleft that one needs to look for to arrive at the diagnosis(Guzman et al, 1983)

Functional Disorders

The commonest functional disorder causing involuntary loss of urine in all children is nocturnal enuresis. Pure nocturnal enuresis is a benign condition that occurs in 15% of children at the age of 5 years and spontaneously disappears in 15% per year. Most children and parents require only reassurance unless it persists beyond 6 to 7 years when the child may demand a solution. Nocturnal enuresis does not cause any day time symptoms and children with day time symptoms need evaluation for an organic cause. In fact any of the functional or neurogenic disorders can produce nocturnal incontinence and one must be careful not to lable them as nocturnal enuresis. Details regarding nocturnal enuresis and its management

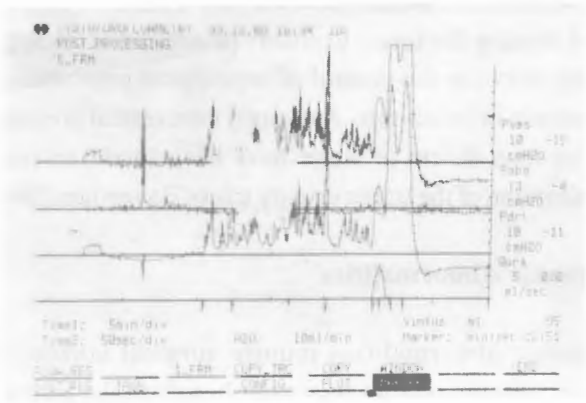

Fig. 4. Urodynamic sudy in a 5-year-old girl with urge incontinence. Marked detrusor instability throughout the filling phase. The girl was placed on probanthine with subsidence of symptoms.

can be obtained from standard textbooks.

A common cause of urinary incontinence in girls is detrusor instability(Figure4). This usually presents as urgency and frequency. The incontinence often but not always manifests as urge incontinence. Children may adopt the classic 'Vincent's curtsy' position (squatting with the heel of one foot compression the perineum) to aid in urinary control (Himsl and Hurwitz, 1991). Detrusor instability can be precipitated in girls with urinary infection and it has been shown that such instability could persist for several months after the infection has cleared.

Children with non-neurogenic neurogenic bladder present like a neurogenic bladder except that there is no neurologic abnormality noted. They show dyssynergic voiding with high pressures and associated instability. This is thought to be a continued usage of the dyssynergic method of achieving continence noted during the transitional phase of maturation of the urinary tract alluded to earlier. These children have often been subjected to forcible toilet training or have a disharmonious household environment. There may be fecal incontinence, or severe constipation (Hinman, 1980). Some girls present with 'lazy bladders' which have become acontractile due to overdistension giving rise to overflow incontinence, or severe constipation (Hinman, 1980). Some girls present with 'lazy bladders' which have become acontractile due to overdistension giving rise to overflow incontinence. These are children who persistently tend to delay micturition either due to fear of soiling themselves or after an adverse event such as dysuria. They may pass urine only once or twice in a day leading to an ever-increasing bladder capacity and finally overflow incontinence(DeLuca et al, 1962).

Giggle incontinence is precipitated by an alteration of the muscle tone during laughter or emotion. The exact mechanism is unclear but it is not a form of stress incontinence. These children in fact may often viod to completion at the time of giggling unlike a patient with stress incontinence and are perfectly continent on coughing or sneezing. This disorder may have a relation to other narcoleptic syndromes (Sher and Reinberg, 1996).

Diagnostic Evaluation

Careful clinical history taking and examination often give a clue to the diagnosis. This must include a detailed voiding history and a voiding diary. Urine examination for infection and a non-invasive ultrasonographic examination are the initial step. In girls with ectopic ureter, most often there is duplication of the ureters and pelvis and this can be detected by an experienced sonologist. Also, ectopic ureters often drain dysplastic, small kidneys and have stenotic openings giving rise to small hydronephrotic kidneys on the USG. Ureteroceles can be identified as a dilatation of the terminal ureter, which can be seen well against the bladder window. These abnormalities can almost almost always be diagnosed by the subsequent IVP that follows such an ultrasonographic appearance. On the IVP the findings in a girl with ectopic ureter are classical. There may be the drooping flower appearance of the kidney due to a poorly functioning upper moiety in the duplication that almost always accompanies. The uppermost calyces appear pushed away from the spine resulting in a change in the axis of the kidney. The pelvis and upper ureter may also be pushed away from the spine by the hydronephrotic non-visualized moiety. A nuclear scan, usually Tc-99m labeled DMSA, is useful to quantify renal function especially if ablative surgery is planned, or to localize small ectopic dysplastic kidneys.

Neurogenic causes almost always produce other sensorimotor deficits. Perineal sensation, reflexes, sphincter tone, gait and spine must be examined. If a neurogenic cause is suspected then a micturating cystourethrogram should be obtained. This may show the typical findings of detrusor sphincter dyssynergia-pine tree appearance of the bladder with irregularity of the walls due to hypertrophy with or without secondary vesicoureteric reflux. Similar findings would be noted in a non-neurogenic neurogenic bladder except that there would be no neurologic basis to explain the pathology. In girls with an acontracile bladder(secondary to sacral agenesis or 'Lazy bladder') the bladder is huge in capacity with a smooth outine. Plain X-ray of the spine may be obtained to confirm a diagnosis of occult spina bifida or sacral agenesis. MRI of the spine may be necessary to search for intraspinal lesions.

Urodynamic evaluation is mandatory for girls with neurogenic dysfunction. The findings of detrusor instability, acontractile bladder or detrusor sphincter dyssynergia can be confirmed. It is important to duplicate the patient's symptoms while doing the test otherwise one may miss the diagnosis. Performing urodynamics in children requires tremendous patience, skill and persuasiveness and the author recollects spending over 3 hours in obtaining one reliable tracing! Slow fill cystometry is recommended to avoid misleading unstable contractions that may occur at fast filling. Due to the problems of urodynamic evaluation in children, they are not recommended as a first line investigation for functional disorders. Functional disorders may be treated on an empiric basis initially unless the patient has very severe symptoms or fails therapy. Similarly micturating cystourethrogram and cystoscopy should be avoided in these children unless an anatomic abnormality is suspected(Himsl and Hurwitz,1991)

Treatment if Urinary Incontinence in Girls

The goals of management for urinary incontinence in children are upper urinary tract preservation, adequate storage and emptying of the bladder at low intravesical pressure, absence of infection, continence, avoidance of a catheter or stoma and social acceptability. Thus although continence is desirable and is the problem that bothers both the parents and the child, the first concern of the treating doctors is to ensure protection of the upper urinary tracts by the control of intravesical pressures and prevention of infections. Sustained intravesical pressures greater than 40 cm of water have been shown to cause deterioration of the upper urinary tracts(Bavendam,1996)

Anatomic Abnormalities

Anatomic abnormalities require surgical correction. Ectopic ureters require treatment based on the functior of the kidney they are draining, and the presence of duplication. Nephrectomy or partial (upper) pole nephrectomy is the treatment of choice if the unit is not functioning or has poor function. It is usually not necessary to remove the complete ureter unless there is accompanying reflux. In a minority of cases when there is acceptable renal function, a pyelopyelostomy can be done to anastomose the two ureters or the ureters may be reimplanted back into the bladder either as a common sheath implantation along with the duplicated unit or separately. The latter is chosen when there is reflux in one of the two moieties. Ectopic ureteroceles are managed along similar lines. Endoscopic incision of the ectopic ureterocele can be performed and many children may require no further treatment.

Exstrophy and epispadias require complex and staged reconstruction best handled by specialized units doing sufficient number of these procedures. In confirmed urethral stenosis urethral dilatation is preformed.

Neurogenic and Functional Abnormalities

These two entities are grouped together since the approach to treatment is similar and based on the urodynamic abnormality found (neurogenic) or suspected (functional, as intial empiric therapy). The abnormality is classified into problems of storage or problems of emptying and each of these are subclassified into bladder and outlet problems (Wein,1984). For instance, a patient with detrusor instability has a problem of storage with the bladder. A patient with detrusor sphincter dyssynergia has a problem of emptying, with the outlet as well as often, a storage problem with the bladder due to secondary instability or hyperreflexia. In patients with an

Table 2 Commonly Used Drugs in the Treatment of Childhood Incontinence

Drug	Minimum Dose	Maximum dose
Propantheline	0.5mg/kg bid	0.5mg/kgqid
Oxybutynin	0.2mg/kg bid	o.2mg/kgqid
Imipramine	0.7mg/kg od	1.2mg/kgtid

acontractile bladder there is an emptying with the bladder.

It is important to understand that an individual patient may show features belonging to multiple categories and treatment must then be directed primarily at the predominant urodynamic finding followed by less important findings in order of priority. Thus the treatment protocol below should be viewed as a trouble-shooting cookbook; the correct recipe for the complete meal has to be selected for the individual young guest!

Therapy to improve storage by the bladder- The first line agent is oxybutynin which has recently been introduced into the Indian market(Table2). This drug has anticholinergic activity, direct smooth muscle relaxant effect and a local anaesthetic effect. There is inhibition of unstable bladder contractions which can be confirmed on urodynamics if necessary. Propantheline bromide is a pure anticholinergic drug which was being used in India prior to the availability of oxybutynin and can still be used since it is probably as effective. Both these drugs may have anticholinergic side effects such as dryness of mouth or constipation. If there is blurring of vision or dizziness then the drug may have to be discontinued. Imipramine is another drug used to improve stroge. The exact mechanism is unclear. It reduces unstable contractions and additionally improves outlet resistance (Wein, 1991). In children with giggle incontinence the centrally acting drug methylphenidate has been found to be effective(Wojcik and Kaplan, 1998)

Calcium antagonists such as nifedipine and terodiline , potassium channel openers such as minoxidil, glipizide, and pinacidil, beta adrenergic agonists, alpha adrenergic antagonists and prostaglandin inhibitors are other potential options being assessed for the future (Wein, 1991)

Therapy to increase outlet resistance- Primary outlet weakness (sress incontinence) in girls without anatomic abnormalities is very rare. In outlet weakness due to anatomic abnormalities such as epispadias, the treatment is surgical correction of the abnormality.

Therapy to improve bladder emptying- Several drugs have been tried for improving the detrusor contractility in patients with acontractile bladders such as bethanechol or perinorm but none of them have been shown to have any objective effect. PGE_2, F_2alpha and GABA antagonists are other agents being tried (Wein, 1991). Certainly, any child with an overdistended bladder and overflow incontinence must have a period of bladder drainage by a catheter to allow the bladder musculature to regain whatever lost tone that it is capable of regaining. But if after this rest, the bladder remains acontractile, medicines do not help and such children may require to be taught CISC or clean intermittent self catheterization. Valsalva or Crede's manouver, or trigger voiding may be used in selected patients with neurogenic bladder.

Therapy to decrease outlet resistance- In children with detrusor sphincter dyssynergia, there is a need to reduce outlet resistance at the time of voiding. There are however no reliable drugs that can do that. The external sphincter(which is involved most often in the dyssynergia) is a striated muscle. All striated muscle relaxants tend to cause unacceptable side effects at doses that relax the sphincter. These are drugs such as baclofen or diazepam (Wein, 1991). Injection of botulinum toxin into the external sphincter area has been tried with mixed success(Steinhardt et al, 1997)

Circumventing the problem- In patients with intractable acontractile bladders or dyssynergic sphincters the problem has to be 'bypassed' by placing the child on clean intermittent self catheterztion (CISC). Several studies have shown the feasibility of (CISC)in small children and the most important factors in success have been shown to be the motivation of the instructor, the initial experience and the motivation of the family. The frist of these is most crucial. The author has taught CISC even to a 4 years old girl who developed neurogenic

bladder after surgery for a midline dorsal sinus which happened to be going into the spine (Figure 3). CISC is a major conceptual advance in urology in this century and has saved the lives of numerous patients with neurogenic bladder(Kass, 1979; Plunkett and Braren, 1979). In children with associated detrusor hyperreflexia, intravesical instillation of oxybutynin, capsaicin and other agents have been used to reduce bladder pressures (Kaplinsky et al, 1996; Dasgupta et al, 1997)

Other modalities- Several non-pharmacologic approaches continue to be pursued for the management of these patients. How many of these will find widespread acceptance is difficult to judge at this stage. The authors have no personal experience with them but some are promising. Pelvic floor relaxation biofeedback was found to cure 82% of girls with urodynamically proven functional voiding problems at a follow up of 6 months (Bozorgi et al, 1996). Maximal electrical strimulation of the anal canal was found to reduce the unstable contractions and improve the cystometric capacity and bladder compliance in 75% of 73 girls with unstable bladders (mean age 9.7 years) at a mean follow up of 14.5 months after the end of maximal electrical stimulation (Trsinar and Kraij, 1996). Behaviour modification with biofeedback was used on 95 children (86 girls) for 10 days with improvement in 81% at 6 months, (Vijverberg et at, 1997)

The treatment of constipation has an independent strong beneficial effect in those girls in whom it is found. In a study of urinary incontinence in 234 children with chronic constipation it was noted that 29% of children has daytime incontinence of daytime incontinence and 34% has nighttime incontinence at presentation and there was disappearance of daytime incontinence in 89% and night time incontinence in 63% following relief of constipation. Children may need initial enemas to empty the colon. Subsequently, a high fiber diet, plenty of fluids and education of parents and children are important in keeping the colon empty. Constipation must always be actively searched for and treated aggressively in these children (Loening-Baucke, 1997).

Timed voiding is a useful adjunctive tool in girls with poorly conractile bladders or those with detrusor instability at large volumes but a stable bladder at small volumes of bladder filling (Wojcik and Kaplan, 1998).

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