Current Approaches to Evaluation and Management of Children With Myelomeningocele*

The past five years have brought considerable changes in the urologic management of children born with myelodysplasia. The most notable developments are clean intermittent catheterization (CIC) with and without adjunctive surgery; decreased use of suprapubic bladder expression; general disenchantment with the long-term results of the refluxing ureterointestinal conduit diversions; artificial urinary sphincter implantation; use of neuropharmacologic agents to control urinary incontinence; new modalities in the investigation of neurogenic bladder dysfunction; and a multidisciplinary approach. An expansion of these items is included in a presentation of current approaches to evaluation and management of children with myelomeningocele.

CURRENT APPROACHES TO UROLOGIC MANAGEMENT

Treatment of Choice

Ideal urologic management involves a minimum of therapeutic intervention. Such conservative therapy must insure the preservation of renal function, control of urinary infection, and be appropriate to age with regard to urinary continence; eg, use of diapers in infants and timed voiding in older children. Various chemical agents can be used to increase the percentage of children who can be managed by these simple measures.

Clean Intermittent Catheterization

CIC was introduced in 1972 by Lapides, soon was adapted for use in children by Rabinovitch and Lyon, and is now in use in most of the larger centers. Results over the first five years indicate that this is an excellent method of bladder drainage. Urinary infection and functional deterioration of the kidney can be treated, controlled, or prevented in most cases by this modality. Many children appear to fare better with this treatment regimen than with either no treatment or urinary diversion. CIC was introduced as a means of controlling infection and deteriorating function; it was soon found to be useful in achieving continence in patients with overflow incontinence. CIC can be started in the neonatal period and continued for prescribed periods of time or indefinitely.

Simple catheterization through the urethra is advocated in all girls and is the method of choice in boys. A perineal urethrostomy provides easy bladder access in boys when the penile urethra is very narrow or when catheterization is difficult. Routine construction of a perineal urethrostomy for intermittent catheterization is controversial because the infant male urethra can usually accommodate a No. 8 French plastic infant feeding tube. CIC in infancy must be carried out by an adult, but with normal intelligence most children are able to catheterize themselves from approximately 6 to 7 years of age, sometimes even earlier. Continence can be achieved in most children on CIC by use of drugs acting on the neuromuscular system, such as oxybutynin chloride, imipramine hydrochloride, ephedrine, and others. Adjunctive surgery also has been used to increase bladder outflow resistance and bladder capacity, thereby improving continence. Early results with CIC, plus adjunctive therapy, are encouraging.

Rhizotomy may be indicated in some children and may change the dynamics of small spastic bladders. Functional bladder capacity can sometimes be increased by rhizotomy, making management by CIC possible and obviating the need for urinary diversion.

Asymptomatic bacilluria is frequently present with CIC; it is probably of less clinical significance than bacteriuria present in a catheter specimen of urine from a normal child. Antibody-coated bacteria tests are usually negative, suggesting that bacilluria is confined to the lower urinary tract. The absence of either clinical or

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radiologic pyelonephritis tends to confirm this. CIC is not effective for all children with myelomeningocele and urinary incontinence and, thus, should not be considered a panacea in the urologic management of this condition. In the long run, CIC may not be an entirely simple solution; presently, children have only been followed for up to six years, and thus CIC has not yet passed the test of time. Indications for the use of CIC may change somewhat in the future, but it will doubtless retain an important place in the urologic management of myelodysplasia.

Suprapubic Bladder Expression

Suprapubic expression is useful in selected patients; however, as a routine method of bladder-emptying, it should be viewed with caution. Suprapubic expression is indicated only when the bladder can be easily emptied, when post-Credé residual is negligible, and in the absence of vesicoureteral reflux. Cystography and measurement of postexpression residual urine volumes may assure that the procedure will be safe. It is definitely dangerous in the presence of vesicoureteral reflux when high intravesical pressures are transmitted directly to the kidney during bladder expression. The possible development of a sagging perineum with growth could be accentuated by suprapubic bladder expression.

External Urinary Collecting Devices

No suitable external collecting device has been developed for females. In larger boys, a condom-type appliance attached to a leg bag affords urinary control if the phallus is of adequate size. Satisfactory light-weight pediatric urinals have been devised employing a condom-type appliance pressed against the suprapubic area. Some are modified with an interposed karaya gum seal. Such a urinal is suitable for the male with a small bladder-residual volume and a low bladder-outflow resistance.

Urinary Diversion

Urinary incontinence alone no longer constitutes an indication for urinary diversion. Overflow incontinence can now best be managed by CIC. Although urinary diversions have now assumed far less importance, they continue to have a limited but definite place in the urologic management of the child with neurogenic bladder dysfunction, especially for the small spastic bladder unresponsive to pharmacotherapy.

A temporary vesicostomy may be needed in children with bladder outflow obstruction in whom CIC cannot be carried out or in the presence of massive reflux with a relatively small bladder because antireflux surgery is technically difficult in these patients. Permanent vesicostomies per se are not satisfactory for bladder drainage, but, as a temporary expedient, can be excellent and may be maintained through the years of childhood when required. Continence-producing mechanisms at the bladder neck or external sphincter can be preserved for possible use at a later or more appropriate time.

Supravesical diversion may still be indicated in some patients, in which case permanent ureterostomy diversion is the procedure of choice in the child with permanently dilated ureters. In the patient with peristaltic ureters, vesicoureteral reflux, and a very small bladder with gaping outflow, or in patients in whom CIC or implantation of an artificial sphincter is not feasible, ureterointestinal urinary diversion is more appropriate. When indicated, a nonrefluxing conduit may be the diversion of choice—either a nonrefluxing colon conduit or an ileocecal conduit—because long-term results or refluxing ureteroileal cutaneous conduits at ten or 15 years are discouraging. The problems associated with refluxing intestinal conduits include pyelonephritis, calculous disease, hydroureronephrosis, progressive renal failure, and hypertension. The incidence and severity of these problems increase with the passage of time. Almost all patients have problems with odor, and many have psychological problems related to the stoma or the urinary diversion. But long-term results of nonrefluxing conduits have not yet been evaluated, nor will they be available for a further ten to 20 years.

Undiversion has been carried out successfully by some workers. It is suitable for some children, especially when overflow incontinence, reasonable bladder capacity, and adequate bladder-outflow resistance was present before diversion. Unfortunately, urodynamic studies of the defunctionalized bladder sometimes fail to predict function after undiversion. Preoperative evaluation by suprapubic filling of the defunctionalized bladder by trocar cystostomy is a useful diagnostic technique.

Vesicoureteric Reflux

In children with myelodysplasia, as in children with nonneurogenic bladder dysfunction, vesicoureteric reflux may be transient and disappear with successful CIC and suppression of infection. In cases of persistent reflux associated with pyelonephritis or progressive hydroureronephrosis in patients on CIC, ureteral reimplantation has been found helpful in preventing further kidney
damage. Antireflux surgery is also indicated before bladder neck reconstruction or artificial urinary sphincter implantation.

Artificial Urinary Sphincter Implantation

Development of an implantable artificial urinary sphincter by Scott has introduced a new dimension in the management of neurogenic bladder dysfunction. Other work has been carried out by Rosen and by Swenson and King with an implantable mechanical, rather than hydraulic, artificial sphincter.

An essential requirement for implantation of an artificial sphincter is complete bladder-emptying. Patients with significant postvoid or postemptying residual are not candidates unless they are rendered totally incontinent. This may require a preliminary transurethral resection of the bladder, continence mechanism, or an incontinence-producing cystourethroplasty. Artificial sphincter implantation, therefore, is suitable for patients with an adequate bladder capacity, an incompetent bladder outflow, and complete emptying.

Reflex neuropathic bladders and severely uninhibited bladders not modulated by parasympatholytic agents would exclude a patient as a candidate for an artificial sphincter. Also, caution is recommended before advocating destruction of any continence mechanism (ie, urethral hyperdilation, external sphincterotomy, or division of urethrovaginal septum as treatment for pyocystis) in patients with adequate bladder capacity and significant retention. Such management must be carefully considered by physician, patient, and parents, because once the forces causing urinary retention are destroyed, the alternative of CIC becomes inoperative.

The artificial sphincter is an appealing, apparently simple solution to a complex problem. Recent reports suggest that the Scott implanted sphincter is fairly successful in adults; however, caution and restraint should be exercised when recommending it for very young children who have to manipulate the device regularly. Growth of the child or damage to the prosthesis may necessitate revision or replacement of various components. The complication and failure rate in many centers is high and, apparently, increases with time.

Results with the electronic pelvic floor or bladder neck stimulators and the detrusor stimulators continue to be disappointing and should be considered entirely experimental. Such devices should be restricted to a few designated centers where carefully controlled studies can be carried out.

Neuropharmacology

Drugs may be used to improve bladder emptying or urinary control. The required sites of action of such drugs are primarily the detrusor muscle and the muscles contributing to the bladder-outflow resistance. Urodynamic evaluation can be used to determine pharmacologic agents of choice.

Drugs used to decrease detrusor muscle tone or eliminate hyperreflexia include propantheline bromide, imipramine hydrochloride, and oxybutynin chloride. Bethanechol chloride increases detrusor muscle tone and helps to reduce residual bladder urine volumes.

Bladder-outflow resistance can be increased by the use of ephedrine and/or imipramine hydrochloride by increasing muscle tone at the bladder neck. Bladder-outflow resistance may be reduced at the bladder neck by the use of phenoxybenzamine hydrochloride or at the level of the external striated muscle sphincter by the use of diazepam.

New Diagnostic Procedures

Radiology. Intravenous urography (or isotope renography) is mandatory, and cystography may be indicated before initial discharge of the child from hospital as a neonate. A 10% to 20% incidence of bladder outlet obstruction has been reported by Johnston and, if present, may require treatment before discharge from hospital. Such apparent outflow obstruction may be a temporary urinary retention following neurosurgical repair of the myelomeningocele.

Cystography will demonstrate the presence or absence of vesicoureteral reflux that, if present, demands closer monitoring of the child. The ability to confirm the presence of reflux outweighs the possible disadvantage of introducing infection. The introduction of such infection is only of significance when pyelonephritis supervenes; this is most likely in the presence of vesicoureteral reflux, which the study itself would demonstrate.

Radioisotopes. The use of radioisotopes for renography and cystography in the surveillance of children with neurogenic bladder dysfunction is suggested as a very useful diagnostic tool. Losses in imaging of the urinary tract are more than balanced by the minimal radiation doses required and the usefulness of comparative sequential studies. It appears to be particularly useful in following up children after urinary diversion. Providing that no changes in the perfusion and clearance of the radioisotopes from the kidney are
detected, repeated intravenous urograms are obviated.

**Urodynamics.** The field of urodynamics has expanded, particularly during the past five years, and detailed patient evaluation is now being carried out in many centers. The studies include electromyography of the anal sphincter and the external urinary sphincter either via the perineum or the urethra, urethral pressure profiles, cystometry, and uroflowmetry. Studies in children under 4 or 5 years of age are technically difficult, especially in the uncooperative patient.

Urodynamic evaluation is helpful during pharmacologic manipulation of bladder and sphincter function. The degree of value from urodynamic evaluations in all children, however, still remains to be substantiated. Urodynamic evaluation is indispensable when implantation of the artificial sphincter is considered or before urinary tract reconstruction. The predictive value of the studies, especially in the defunctionalized bladder, is unproven, and their use may still sensibly be questioned until more information is available.

**Antibody-Coated Bacteria Test.** This test, which is not yet generally available, may prove useful in accurately localizing the source of bacilluria. This may be of particular use in evaluating the significance of bacilluria in the presence of urinary diversion. Experience to date, however, is too limited for the value or accuracy of this investigation to be definitely substantiated.

**Multidisciplinary Approach**

When possible, children with myelodysplasia should be treated by a multidisciplinary team. Myelodysplasia is a complicated condition affecting many organ systems so that management should be coordinated when possible by physicians and allied health professionals who have an understanding of the basic goals and objectives outside and inside their specialty. When many specialists in isolation manage problems relating only to their specialties, fragmented and, therefore, suboptimal care is likely to supervene.

**Related Aspects**

**Circumcision.** Routine circumcision should be avoided in the incontinent boy because the prepuce protects the neurologically insensitive glans penis from ulceration by dermatitis or by trauma from continually wet diapers and later from external urine-collecting devices.

**Bowel Control.** Urologists are frequently called upon to give advice about bowel control. Control can be achieved by most children by use of the following, alone or in combination: substances to change the consistency of stool, laxative suppositories, timing bowel evacuations, and enemas.

Substances to change the consistency of stool are high fiber-containing food, such as bran; stool softeners, such as psyllium, psyllium hydrophilic muciloid, or diocetyl sodium sulfosuccinate; medication to slow down intestinal motility, such as diphenoxylate hydrochloride, which produces a more formed stool.

Bisacodyl suppositories in contact with rectal mucosa induce peristalsis throughout the large intestine. Insertion of suppositories can be timed to follow meals and thereby take advantage of the gastrocolic reflex to enhance colonic peristalsis.

Saline enemas may be required by some patients who have a poor response to stimulating suppositories. Enemas can best be retained if the child is prone and the buttocks are held together during and after the installation.

**Sexual Concerns.** Sex counselling is very important and should ideally be available for all neurologically impaired patients who do not have significant mental retardation. Some infant males and prepubertal boys have spontaneous erections; correlation with the degree of sexual impairment is often not predictable early. Many will have useful sexual function after puberty.

Sexual function in females is not dependent upon normal perineal sensation. Sexual concerns in females have a much stronger emotional basis than is found in males. Some girls have developed satisfactory marital relationships and have sustained normal pregnancies.

**Suggested Minimal Urologic Evaluation and Surveillance**

**Neonates and Infants.** An intravenous (IV) urogram, in addition to careful urologic and neurologic examination, urine culture, BUN, and serum creatinine estimations, is suggested. If these studies are abnormal, the appropriate therapy should be instituted. A persistently full bladder is abnormal and should be noted; such a child should be very carefully monitored for the development of upper tract changes. A cystogram, which can be performed by suprapubic puncture, is recommended in these patients. Intravenous urography and cystography or both can be replaced by radionuclide studies. Bladder outflow obstruction should be treated by CIC or a temporary vesicostomy; urethral dilatation is no longer recommended.

In the presence of normal upper tracts and in the absence of vesicoureteral reflux, routine urinalysis and culture should be obtained at
intervals of three to six months. (In remote areas, such urinalyses can be performed with a urine dip stick estimating urinary pH and presence or absence of proteinuria, hematuria, and/or baciluria.)

Follow-Up of “Normals.” In the absence of bladder distention and in the presence of normal urinalysis and negative urine culture and previously normal upper tracts, follow-up urography or isotope renography should be carried out at 1 and 3 years of age and, thereafter, at intervals of two or three years. This test can be replaced by radionuclide imaging.

Ideally, every child should be examined at regular intervals, along with a urinalysis and culture, through puberty. When the presence of a significant urinary residual is suspected, this should be confirmed by catheterization. If recurrent pyelonephritis or cystitis occurs, the disease should be evaluated and treated appropriately. Deterioration of the urinary tract is unlikely, providing the child is closely monitored and treated appropriately.

Follow-Up of Children with Reflux. When vesicoureteral reflux is demonstrated, close surveillance is necessary. Any modality may be used. Isotope cystography, if available, considerably reduces the child’s exposure to radiation. Children probably should be kept on long-term urinary antimicrobials in low doses, such as nitrofurantoin or trimethoprim sulfa.

Urinary Incontinence. In children with normal upper urinary tracts, satisfactory bladder-emptying, and freedom from urinary infection, urinary incontinence alone does not need any intervention before school age. At this age, urodynamic studies can be performed to determine appropriate management for achieving urinary continence. When incontinence is associated with upper tract deterioration or recurrent urinary tract infection, evaluation and treatment should be instituted regardless of age.

Urinary Conduits. Surveillance of urinary conduit diversions include regular inspection and evaluation of the stoma. Radiologic studies should include an IV urogram within six to 12 weeks of surgery. If abnormal, this should be repeated after a further six to 12 weeks to allow for additional resolution of the operative reaction unless changes are sufficient to require surgical correction. When postdiversion urography is normal, the child should be followed up indefinitely by isotope renography or pyelography at appropriate intervals. In the absence of reflux or in the presence of an abnormal radioisotope study, IV urography should be carried out. Urinary acidification is recommended to inhibit calculus formation. Long-term use of urinary antimicrobials is optional.

Great pains must be taken to impress upon both parent and patient the need for close and long-term surveillance of the urinary tract. The majority of severe problems develop in children in whom close surveillance has not been carried out.

Endoscopic Evaluation. Cystoscopy is not mandatory but may be helpful in the complete evaluation of children with urinary tract dysfunction. Endoscopy is mandatory when evaluating vesicoureteral reflux and before urinary tract reconstruction.

SUMMARY

The introduction of CIC and the artificial urinary sphincter have radically altered the management of the urinary tract in children with myelodysplasia. Supravesical urinary diversion is less often needed, although there is still a place for this treatment modality. When diversion is indicated, nonrefluxing intestinal conduits are suggested. The routine use of suprapubic bladder expression has only limited applicability. When possible, the child should undergo urodynamic study, and the family should be made aware of the treatment modalities available. Before proposing a urinary diversion or implantation of an artificial urinary sphincter, detailed explanations of the procedure and the alternatives must be given to parents and patients.

This report was reviewed by the present and former members of the committee who have contributed to its contents.

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