SBA National Resource Center:
800-621-3141

SPINA BIFIDA ASSOCIATION

Urologic Care and Management

The urologic care of children with Spina Bifida has undergone several important changes in the last decade. The emphasis is now on early catherization of the child's lower urinary tract and preventive treatment to preserve both kidney and bladder function in an attempt to maximize the child's long-term urologic functioning and overall health.

Newborn assessment

As soon as the neurosurgical condition is stable after birth, an assessment is made of the newborn's kidneys with a serum BUN, creatinine and renal ultrasound (ECHO); the bladder is evaluated with urodynamic studies. Children whose bladder will fill and empty with low pressure are managed expectantly.

Management of the neurogenic bladder

If the child's bladder pressure is elevated above 40 cm H20 during filling and/or the urethral sphincter is dyssynergic (not coordinated during a bladder contraction), then the child should begin intermittent catheterization (CIC) and possibly anticholinergic drugs to ensure complete emptying and low filling pressure. Antibiotics are usually prescribed for a short time until the family is comfortable with the technique of catheterization.

If the kidneys are dilated (hydronephrotic) or the urodynamic studies reveal high bladder filling pressure or a dyssynergic sphincter with high voiding pressure, a voiding cystogram (VCUG) is performed. When reflux is present, most children will need to be started on CIC and antibiotics to achieve adequate drainage of the upper urinary tract. A high-pressure bladder in the presence of reflux and/or hydronephrosis warrants the additional use of anticholinergic medication. Children with lesser grades of reflux or mild hydronephrosis, whose bladders empty with low pressure, are managed without CIC because it is unlikely they will develop any damage to their kidneys. These children are followed very carefully though to see if their condition changes.

Surgical options

If the infant with impaired upper urinary tract drainage fails to respond to CIC and anticholinergic medication; or the parents are unable to perform the procedure easily and routinely, a vesicostomy may be performed. This procedure consists of bringing the dome of the bladder to the skin as a stoma midway between the umbilicus and the pubic bone. The advantage of this temporary form of diversion is that it is easily reversible when the child is older and the family is amenable to CIC, and it does not alter the child's future continence mechanism.

Surveillance

Throughout infancy and early childhood the children are followed with routine urine cultures, evaluation of post-void residual urine volumes (in those not on CIC), and kidney and bladder ECHOs. Urodynamic studies are repeated on a regular basis in those children with normal or minimal denervation in the sphincter to see if their neurologic lesion has changed and/or spinal cord tethering has occurred and to determine if the children who are on expectant therapy, have altered their bladder and sphincter function such that prophylactic intervention with CIC and/or anticholinergic drugs is warranted.

Urinary tract infections

When a child develops recurrent urinary infection, a VCUG or radionuclide cystogram (RNC) is performed (even if it has been done previously) to evaluate for reflux. Children not on catheterization are given appropriate antibiotics. In children on CIC but without reflux, only symptomatic infections are treated with short courses of antibiotics. Children on CIC with reflux are managed similar to those with normal bladder function, i.e., antibiotics as indicated, frequent urine culture, regular ECHOs and, if indicated, RNCs or VCUGs to monitor the progress of their reflux. If a child with mild reflux who is not on CIC develops symptomatic UTIs, he/she is begun on regular catheterizations.

The use of CIC, antibiotics and anticholinergics has resulted in a spontaneous resolution of reflux in more than 50% of children, with a reduction in its severity in another 30 to 40%. Antireflux surgery (required in 10 to 20%) is undertaken when children have repeated infection despite adequate antibiotics and a good bladder- emptying regimen. The success rate for surgery is almost as high as it is for children with normal bladder function, i.e., 96 to 98%.

Continence-medical therapy

Because the emphasis has shifted to mainstreaming children with Spina Bifida into normal school and social situations, it is very important for bladder and bowel function to be controlled before the start of school. If the child is not on CIC in the months preceding this milestone, catheterization is started and urodynamic studies performed to determine how best to modulate the incontinence. Medications are given to a) lower bladder filling pressure and increase capacity, and b) raise urethral resistance to prevent leaking between each emptying. Success rates for achieving continence range as high as 65%.

There are several alternatives within each category of drugs that may be given so that if one medication does not achieve the desired effect or produces side effects, another may be substituted. Even the method of administration has changed, for example, Oxybutynin

(Ditropan) tablets may be dissolved in normal saline and instilled directly into the bladder through the catheter used for CIC.

When these measures fail to lower bladder pressure and increase capacity, several alternatives exist. Transurethral electrical bladder stimulation (TEBS) has been conducted at a number of centers around the United States; this consists of delivering an electrical stimulus to the bladder through a specially designed catheter for 90 minutes, 5 days a week for 5 weeks. In one study, this treatment has been shown to attain continence in 15 to 25% of children with another 25 to 30% requiring less medication to achieve a low-pressure large capacity bladder. In addition, a significant majority report improved bowel continence after completing one or two courses of treatment. Several research centers in North America have started to directly stimulate the nerves that control bladder and urethral sphincter function in an attempt to improve both storage capabilities and emptying efficiency of the bladder. Assessment of preliminary results from these centers has been encouraging. It is not known how long lasting the effects of these programs will be.

Injection therapy

Over the last several years, advances have been made in the endoscopic management of incontinence. This has resulted in less invasive procedures being performed prior to major open reconstructive surgery. Botulinum toxin (Botox) injected directly into multiple sites within the bladder muscle can paralyze it leading to increased capacity and compliance, resulting in improved continence and kidney drainage. The botox may be effective for upwards of 6–9 months but repeated injections are needed to maintain its effect.

Recently, bulking agents (primarily Deflux) injected into the ureter adjacent to its orifice at cystoscopy has been promoted, with a success rate ranging as high as 80 – 90%, for the milder grades of reflux. It is imperative that the bladder needs to be optimally managed even in these cases of endoscopic treatment.

Dextranomer hyaluronic acid polymers (Deflux) injected directly into the bladder neck or continent stoma conduit (where it enters the bladder) is currently also being employed to enhance bladder outlet resistance and improve continence when drug therapy or prior surgery fails to achieve total continence. The effect seems to be stable but long-term studies are needed to judge it efficacy.

Surgical treatments

When these measures fail to achieve continence, various formal surgical procedures have proven useful. Augmentation cystoplasty in which a segment of the gastrointestinal (GI) tract is isolated and added onto the bladder has been performed on numerous occasions. All portions of the GI tract have been utilized – stomach, intestines, colon and sigmoid. The bowel must be detubularized to prevent physiologic contractions that occur in the normal bowel segment from producing unwarranted incontinence. Mucus and urinary infection may be persistent problems, even years later. Perforation of the intestinal segment from overdistension has occurred, especially if the individual's bladder is not emptied routinely. A disturbing feature relating to these intestinal segments has been the slowly but steadily increasing incidence of tumor formation within the bladders of these individuals beginning 15 to 20 years after the augments were fashioned. Further investigative studies are under way to determine the true incidence and susceptibility to this potentially devastating complication. The need for long-term surveillance is paramount.

Autologous tissue engineered constructs of bladder tissue grown from an individual's own bladder biopsy material has produced favorable results in phase 1 trials. This may hold promise as a viable alternative to intestinal sources for augmentation, thus eliminating the potential complication noted above.

Procedures designed to increase bladder outlet resistance include bladder neck reconstruction, fascial sling or bladder neck suspension, artificial sphincter implantation and the Kropp procedure. The artificial sphincter consists of an occlusive cuff fitted around the bladder neck or bulbar urethra, a pressure regulating balloon reservoir and a deflate pump placed in the scrotum or labia. When the child wants to void, he or she squeezes the pump, opening the cuff and allowing the bladder to empty. The cuff automatically refills with fluid from the reservoir over the next five minutes occluding the bladder neck once again. If the child can empty the bladder spontaneously with straining before the sphincter is implanted, then he/she should be able to do so postoperatively. The long-term success rate for the artificial sphincter at least five years after implantation ranges from 65 – 80%.

All other operations designed to increase bladder outlet resistance require that the child catheterize him/herself afterwards to empty the bladder. In the fascial sling procedure, a strip of fascia (or fibrous tissue surrounding the rectus muscle) is isolated and wrapped around the bladder neck compressing and buttressing it against the undersurface of the pubic bone. The added resistance and the repositioning of the urethra prevent leakage between catheterizations in more than 80% of children. The Kropp procedure isolates a strip of anterior bladder wall, tubularizes and tunnels it below the surface of the posterior bladder wall in order to elongate the urethra. This creates a very effective continence mechanism (95%) but it requires reimplantation of both ureters in a slightly more lateral position to accommodate the neourethra, and augmentation cystoplasty because so much of the bladder is used in the reconstruction.

Continent diversion

When the child cannot be catheterized transurethrally or requires a urinary diversion, most urologists are now performing an operation to create a continent catheterizable stoma. This conduit can be fashioned from ureter, appendix (preferred), fallopian tube (in girls), intestine or stomach—anything that can be easily catheterized. One end is brought to the skin and the opposite end implanted into a urinary storage reservoir if the bladder cannot be salvaged.

When the bladder remains as the urinary reservoir, its neck might need to be obliterated, especially if the outlet resistance is very low, in order to eliminate the possibility of urinary leakage from the urethra. The continent stoma is small and inconspicuous and is easily covered with a band-aid or small gauze pad. Continence is achievable in over 90% of children. The children with this anatomic arrangement have done well on a reasonable long-term basis but more time is needed to be certain about late follow-up. The only consistent yet easily manageable complication has been stenosis at the skin stoma site. Most families report the child's quality of life and care giving are vastly improved, with many children becoming more independent in managing their urinary tract than previously achievable.

Incontinent diversion

Urinary diversion, commonly performed in the 1960's to the mid 1980's, consists of isolating a segment of small intestine, attaching one end of it to the ureters after they have been detached from the bladder, and bringing the opposite end to the abdominal wall skin as a wet stoma. The urine passes through this system continuously to a bag placed over the abdominal wall stoma. This operation is rarely performed anymore unless it is deemed difficult or impossible to get the child or adolescent to catheterize their urinary reservoir on a regular basis. Individuals who currently have these incontinent stomas are not necessarily candidates for either an undiversion or a continent stomas operation, especially if they have not had any adverse reactions from their original conduit surgery.

The development of complications, e.g., pyelonephritis (kidney infection), urinary tract stones, obstruction in the urinary conduit, or stomal stenosis requiring a revision, may warrant conversion of the conduit to a continent stoma.

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