SPINA BIFIDA ASSOCIATION

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Urologic Care and Management

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Urologic care from childhood to the transitional years

The goals of urologic care and management of a child with Spina Bifida center on maintaining normal renal function, transitioning through stages of urinary continence, and achieving independence with personal care as aging continues through adulthood. The significant achievements in other specialties particularly neurosurgery have unmasked the importance of maintaining normal renal function and a healthy bladder environment for urinary storage and elimination. Over 90% of children with Spina Bifida will have a normal upper urinary tract (kidney and ureter) at the time of birth. With no follow up, half of the children will experience considerable upper urinary tract damage due to lower urinary tract (bladder and urethra) hostility.

With appropriate urologic management, children at risk for renal damage can be identified early and intervention undertaken to prevent long term compromise of kidney function. During the first several years of life, the urologic focus on a child's health is based on maintaining normal kidney function. As the child begins to approach school age, interest will be directed toward gaining urinary continence. As a teenager, there is a structured transition of care. Each of these urologic management milestones builds upon the last, potentially affecting their status in a positive or negative fashion. There are overlapping factors that can affect both kidney function and urinary control.

Several principles must be recognized that are not unique to urology or to children with Spina Bifida. The most important is that no two children are alike. As you meet other families and discuss issues related to their child's problems and successes, you must remember that what is appropriate and effective for one child may or may not be appropriate for your child. From a medical point of view, we are unable to simply look at a child's back and determine what type of bladder or kidney function they will have. It is for that reason, children with Spina Bifida require very close urologic evaluation.

The following description of urologic care is based on a protocol used at our Children's Hospital. It works well for our children because of our particular resources. The philosophy presented here can be carried out elsewhere. However, there are multiple approaches taken when caring for neurogenic bladder dysfunction and if your child is managed differently that should not be considered incorrect. Institutions create a protocol based on their program's philosophy and resources. The critical factor is that you continue with consistent urologic follow up and actively participate in the urologic management that has been recommended for your child.

Children will be managed based on the general philosophy of their institution. The two traditional management options are proactive—attempting to identify the child at risk and treating him/her before a problem occurs, or retrospective—following a child closely and at the first sign of any adverse change begin management. Pediatric urologists have had good results treating children in either fashion. The protocol discussed below is based on a proactive philosophy.

Newborn children and infants

The infant bladder functions as a reservoir for urinary storage and elimination. Babies with normal bladder function void strictly by reflex. The fact that a baby's diaper is wet does not equate with normal voiding. Studies need to be performed to determine at what pressure urine is stored and eliminated. It is also helpful to learn whether unusual bladder contractions are occurring and whether the external urinary sphincter (muscle) is relaxing appropriately when a bladder contraction occurs. This is called bladder sphincter synergy. Children with

neurogenic bladder dysfunction often contract their external urinary sphincter at the same time their bladder contracts. This results in abnormally high voiding pressure and is called bladder sphincter dyssynergy. A special bladder study (urodynamics) is required to understand how an infant is voiding. A baseline assessment of the kidneys should be established with a renal ultrasound. Blood work is obtained to assess kidney function in children with abnormal kidney changes.

We begin intermittent urethral catheterization on all newborns with Spina Bifida. This helps eliminate urine when abnormal bladder function is present. We quickly taper catheterization if the volume of urine obtained is consistently low. We have found it beneficial to begin catheterization early, in order to identify the small group of children who carry a significant amount of urine and are at risk of injuring their kidneys. Catheterization may be re-initiated after the first urodynamic study. In addition, all families become familiar with the technique of catheterization which is helpful if it is recommended at a later date. Approximately 20% of the infants discharged from our hospital, will be maintained on intermittent catheterization. And all neonates will be placed on a low dose of an antibiotic. This is continued at least through their first urodynamic study.

In order to determine whether a child is at risk for injuring their kidneys, bladder function must be assessed. This can be undertaken with urodynamic testing. This study assesses the bladder's ability to store and eliminate urine. The urodynamic test is complicated and uncomfortable particularly when the child has normal sensation. The test can be combined with x-ray imaging of the bladder in order to evaluate any structural bladder changes due to nerve damage, the presence of vesicoureteral reflux (urine backing up, into the kidneys), the bladder neck, the urethra, and residual urine after voiding. During the urodynamic study, the bladder is checked for changes that are considered hostile to kidney growth and development (e.g., elevated storage pressure, abnormal bladder contractions, bladder sphincter dyssynergia). Vesicoureteral reflux has been noted in 20% of our children and they are continued on prophylactic antibiotics.

The results of the urodynamic study will determine what follow up is required. When the urodynamic test is normal, or shows favorable bladder function, the child will be scheduled for a return visit in appropriately six months with a sonogram to reassess their kidneys and a urine culture. The urodynamic study will be repeated yearly in order to identify bladder changes that cannot be recognized by simply visualizing the baby's daily voiding routine. Significant bladder changes noted on multiple urodynamic studies may reflect the effects of a chronic urinary infection, progressive nerve damage, or the development of a tethered spinal cord. Often the first indication of spinal cord tethering is manifested by changes in bladder and bowel function. In infants, it is difficult to determine any adverse changes unless urodynamic analysis has been performed. In some children, it is difficult for the neurosurgeon to determine if symptoms and spinal MRI findings are consistent with a tethered cord. They may rely on urodynamic changes to help with their management.

When a hostile bladder environment is found on urodynamic testing, treatment will be recommended based on the specific abnormality. Treatment will follow various progressive steps, beginning with the easiest to perform and the least involved. Some children have a large residual of urine. That can be eliminated by the Crede´ technique (pushing on the bladder with your fingertips above the pubic bone). This technique is very helpful but not appropriate for all children especially if vesicoureteral reflux is present or if the child has been shown to have a Crede´ reflex (compression of the bladder results in a reflex contraction of the external urinary sphincter and very high-pressure elimination of urine). Using the Crede´ technique over a long period of time when these problems are present can be detrimental to the kidneys. Some children will have excessive overactive neurogenic bladder contractions. They will be started on medication in an attempt to control the contractions and

decrease pressure on the kidneys. The commonly used medications are oxybutynin (Ditropan), tolterodine (Detrol), hyoscyamine (Levsin) and solifenacin (VESIcare). Medication is first given in an oral fashion but if poorly tolerated, some medications can be given directly into the bladder if the child is catheterized.

Intermittent catheterization is the most important urologic advancement over the past 30 years in the treatment of the neurogenic bladder. Intermittent catheterization successfully eliminates urine from the bladder and mimics normal voiding by inserting a tube into the bladder. This can decrease urinary infections and reduce the effects of hostile bladder pressure on the kidneys. Intermittent catheterization is performed in a clean but not sterile fashion. All children will eventually have bacteria in their bladder but only about 50% will have symptoms related to the bacteria resulting in a clinical urinary tract infection (UTI). Asymptomatic bacteria in the urine do not always need to be treated. Symptomatic infections are usually easily controlled. Approximately 50% of the children who catheterize will require a low dose of an antibiotic in order to remain symptom free.

There is a small group of infants with significant bladder hostility who do not respond well to medication or intermittent catheterization. When that occurs, operative intervention may be required. The most effective operative procedure to eliminate urine and decrease pressure in an infant is creating a vesicostomy. This is a small opening made in the bladder connecting it to the abdominal wall between the pubic bone and the umbilicus. A vesicostomy is well tolerated and reversible. Normal bathing is possible. It is common for the infant to develop a rash around the opening because of continuous urine dribbling. This will improve with time but may intermittently require topical treatment. The vesicostomy provides excellent bladder decompression and usually improves the pressure on the kidneys due to high-pressure bladder storage and high-pressure voiding.

Children

As the child ages the upper urinary tract should remain normal. Interest can be shifted to focus on gaining urinary continence. The age at which a child begins to work toward urinary continence is individualized, based on their physical capabilities and social situation. It is practical to consider urinary continence when a child enters school. While this is a realistic goal, it may not be appropriate for all children. In order to gain urinary continence, it will be important to adhere to a consistent continence program. This program may include medications, intermittent catheterization, and possibly operative reconstruction. A urinary continence program must be continued even when the child is in a structured school environment.

Some children with Spina Bifida can void on their own and achieve normal urinary control and continence. The majority of children, however, will require a combination of treatment measures similar to what was described above. Urodynamic bladder testing will help assist the pediatric urologist in developing a program that is specific for your child's problem and needs. Most often, conservative, reversible treatment is initiated first. This includes medications and/or intermittent catheterization. Only when this management has been maximized will operative intervention be recommended.

In order to achieve urinary continence, three important factors of bladder function need to be assessed. The first is the ability of the bladder to store an adequate volume of urine at low pressure. The second is the ability of the bladder to contract or eliminate urine. And the third is the ability of the bladder neck and/or external urinary sphincter to remain closed when the child is not voiding in order to remain dry. Any one or all three of these factors may be dysfunctional. Treatment will be dependent on what abnormality is present.

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When the bladder capacity is small and/or the resting bladder pressure is elevated, medications can be tried to enlarge the capacity and decrease the pressure. The commonly used medications are oxybutynin (Ditropan), tolterodine (Detrol), hyoscyamine (Levsin) and solifenacin (VESIcare). When medications have been unsuccessful the bladder may need to be enlarged or augmented. Various tissues have been used to augment the bladder, most often the small or large bowel is selected. There are advantages and disadvantages of using any segment of bowel. The large bowel is in close proximity to the bladder and its large diameter results in a smaller segment that needs to be isolated. But there is a greater discharge of mucus and there can be an effect on bowel control for some children. For those reasons, small bowel (ileum) is the common choice. When bowel is used, it is reconfigured into a "cup patch" which is added to the bladder. Reconfiguring the bowel creates a reservoir of greater capacity and also decreases elevated pressure that can occur when the bowel segment contracts. Some centers are beginning to explore the use of injecting Botox into the bladder to reduce unwanted high-pressure contractions. This injection is easy to preform but the benefit is limited by time and is considered a temporary when effective. Alternatives to using bowel are under investigation. Growing additional bladder tissue in the laboratory using a child's own cells, and then placing that tissue on the bladder may be shown to have a clinical benefit. That treatment is several years away from becoming the standard of care.

When any segment of bowel is used in urinary reconstruction, there is a long-term risk for a bowel obstruction. This risk persists throughout life but remains at a low percent. The bowel naturally secrets mucus and the mucus needs to be intermittently irrigated from the child's bladder. Failure to do so will result in excessive accumulation of mucus, poor urinary drainage, increased risk of urinary infections, odor, and the formation of bladder calculi. Draining the bladder completely and irrigating the bladder on a routine basis helps diminish those risks.

Augmenting the bladder with a portion or bowel will likely result in the child requiring intermittent catheterization. Therefore, it is beneficial for all children and parents to feel comfortable with the technique of catheterization for several weeks prior to reconstructive surgery. After the augmentation, it is essential that catheterization is maintained on a very strict schedule. Often times when children are left to catheterizing themselves, poor technique will lead to either incomplete emptying or excessive stretching of the bladder and augmented segment. Over a long period of time a bladder rupture can occur and become a life-threatening problem. Teenagers often are complacent regarding the importance for maintaining a strict catheterization schedule. They need to be supervised when they become responsible for their own care.

Unfortunately, oral medication has not been effective when the urinary sphincter and/or bladder neck are not strong enough to hold the urine within the bladder. Various operative techniques are used to tighten the sphincter and bladder neck. Multiple procedures are available because there is no single procedure that is effective for all of the different types of outlet problems. Some operative techniques use natural bladder tissue which is fashioned into a "one way valve". This allows a catheter to be passed for drainage but prevents urine from flowing out. Another technique uses a strip of fascia (strong tissue that surrounds a muscle) which is placed around the bladder neck and tightened like a belt. An artificial (mechanical) sphincter can be placed and is helpful for selected children. Most bladder neck repairs will require the child to catheterize. The specific bladder neck operation that is recommended for your child will be dependent on the problem that is identified.

It should be appreciated that intermittent catheterization is a common theme for maintaining normal kidney function and urinary continence. Most often catheterization is begun through the normal position of the urethra

(the tube that connects the bladder to the vaginal opening in girls, and travels through the penis in boys). As the child begins to mature, it may become more difficult to catheterize through the natural urethra. In addition, some children who use a wheelchair have difficulty reaching their genital area for self-catheterization. It is possible to make a small catheterizable tube that enters the bladder from the body wall above the pubic region. The stoma (opening) of the tube can be hidden within the umbilicus or positioned very low on the abdominal wall. The tube is commonly created by using the appendix (Mitrofanoff) or a portion of the small bowel (Monte/Yang).

Bowel Control

It is practical to consider bowel continence at the same time a child is working toward urinary continence. It is essential for a child to maintain appropriate stool consistency which can be achieved by a diet that includes plenty of fluid and fiber. Some children benefit from fiber supplementation. Even when the stool is of normal consistency, some children need to have assistance in eliminating the stool from their rectum and colon. There are various techniques recommended including digital manipulation, rectal suppositories, and enemas.

For more information on bowel management, please refer to the Spina Bifida Association Lifespan Bowel Management Protocol.

Transitional Years

When the child becomes a teenager, their urologic care should slowly make a transition from parent or caretaker to the teenager. This transition can be difficult for both the teenager and parent or caretaker. Teenagers should be educated about their particular problem(s) and the reasons they need to perform certain activities. It is important that the adult allow the teenager to assume the responsibility for their bladder and bowel needs. While the parent or caretaker provides independence, it is necessary for them to continue close monitoring assuring that the teenager is accomplishing the goals of the expected treatment program.

The teen years are a struggle for children with Spina Bifida just as they are for other children. However, the medical consequences resulting from the lack of attention paid to specific personal needs is much greater. Parents may need to refocus their child on what is required to maintain healthy kidneys and urinary continence.

With appropriate urologic care, most children with Spina Bifida will transition through life with normal renal function and achieve urinary continence.

Summary

- Providers will gather information to assess baseline urologic function by:
  - Imaging and functional studies (urodynamic testing and ultrasound)
  - Blood tests (creatinine and electrolytes)

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- Using the information, they will recommend appropriate next steps that may include:
  - Crede´ technique: pushing on the bladder to help eliminate urine
  - Intermittent catheterization to eliminate urine by inserting a tube into the bladder
  - Medications to decrease bladder activity (Ditropan, Detrol, Levsin, VESIcare)
  - Antibiotics to help prevent urinary tract infections (UTIs)
  - Surgery (vesicostomy) to create an opening in the bladder for urine to drain
    - Can use the small bowel (Monte/Yang) or appendix (Mitrofanoff)
  - ACE (anterograde colonic enema) procedure to assist with stool elimination

- Imaging and functional studies are repeated to monitor urologic function
  - Ultrasounds are completed every 6 months until 2 years old then completed yearly
  - Urodynamic testing is completed yearly until 3 years old and then as needed
  - Both repeated sooner with recurrent UTIs, imaging changes, or new symptoms

- Each care plan is individualized to the specific needs of the child

- Families are educated on how to perform any necessary procedures and care

- It is important to promote independence in self-care as much as possible as the child ages

Please reference the information sheet, "*Frequently Asked Questions about Pediatric Urinary Tract Infections and Catheterization in Children with Neurogenic Bladder and Bowel*" for more information on urinary tract infections (UTIs), vesicoureteral reflux (VUR), and voiding cystourethrograms (VCUG).

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Revised by Nancy Moran, MD Candidate 2023

This information does not constitute medical advice for any individual. As specific cases may vary from the general information presented here, SBA advises readers to consult a qualified medical or other professional on an individual basis.