

Urology

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Introduction

The goals of urologic management and care of individuals with Spina Bifida focus on maintaining normal renal function during all ages, transitioning through stages of urinary continence, and achieving independence with personal care as aging continues through adulthood. Significant advancements in other specialties, particularly neurosurgery, have prolonged life and unmasked the importance of maintaining normal renal function and a healthy bladder. Over 90% of children at birth have a normal upper urinary tract (kidney and ureter). Historically, we know that if left unattended, 50% of those children will suffer upper urinary tract damage due to lower urinary tract (bladder and urethra) hostility.¹

During the first several years of life, the urologic focus on a child's health is based on maintaining normal kidney function at a time when the kidneys are most susceptible to kidney damage. As the child begins to approach school age, greater interest is directed toward gaining urinary continence. As a teenager, there is structured transition of care. Each of these urologic management milestones builds upon the last, potentially affecting their status in a positive or negative fashion.

Institutions create protocols based on their program's philosophy and available resources. Two general philosophies prevail: a proactive approach that attempts to identify children at risk for upper urinary tract deterioration and treat them before a problem occurs; and a reactive approach that follows a child closely and begins management at the first sign of any adverse change.²⁻⁴

Advocates of a proactive approach favor early identification of "at risk" children by assessing bladder function and managing hostile bladder parameters. This is done in an attempt to prevent adverse upper urinary tract changes and preserve normal renal function, thus limiting exposure to possible irreversible upper tract deterioration.

Institutions favoring a reactive approach rely on close evaluation of the upper urinary tract, renal function, and documentation of urinary infections. It is felt that adverse upper urinary tract changes can be detected early with minimally invasive assessment using ultrasonography. Renal function is typically assessed and followed with a serum creatinine. Adverse changes are assumed to be reversed with medical, pharmacologic, and operative management. Treating children reactively "as needed" allows for precise selective management limiting the stress and potential side effects of invasive procedures, medications, catheterization, and surgery.

The importance for maintaining normal renal function within this guideline cannot be overstated. It is also appreciated that while creatinine is a good screening tool of renal function, it is limited in the non-ambulatory child and adult with Spina Bifida with low muscle mass and thus provides a false sense of normality.⁵ Renal function may be more accurately measured with serum cystatin C or with a nuclear medicine glomerular filtration rate test (GFR).⁶ Currently, the best measure of renal function in children and adults with Spina Bifida is unknown and will require

ongoing investigation.

This guideline merges aspects of proactive and reactive philosophies based on a best practice methodology. The Centers for Disease Control and Prevention (CDC) are undertaking a prospective management protocol for newborns through age five developed by a team of pediatric specialists.⁷ It is anticipated that the outcome will positively impact the urologic care of children as well as the kidney health for individuals with Spina Bifida across the lifespan. Providing a strong foundation for pediatric care directly impacts the lifetime goals related to continence, self-management, and renal health. It is appreciated that urologic care is a dynamic, ever changing process.

Outcomes

Primary

1. Maintain normal renal function throughout the lifespan.
2. Achieve urinary continence as early as socially acceptable.
3. Maximize urologic independence.

Secondary

1. Eliminate hostile bladder dynamics through medical management.
2. Reduce or eliminate operative reconstruction of the bladder.
3. Maximize renal outcome while minimizing expense of studies, keeping watch over the timing and frequency of studies such as urodynamic testing, upper tract imaging, and lab studies.
4. Reduce impact of urinary tract infections (UTIs) and antibiotic overuse.
5. Establish a care program that allows for urologic independence, such as through clean intermittent self-catheterization (self-CIC).

Tertiary

1. Determine the best measure of renal function.
2. Minimize occurrence of urolithiasis.
3. Determine whether surgical interventions are effective in the long-term.

0-11 months

Clinical Questions

1. How do you define a symptomatic urinary tract infection and what is its long-term sequela?
2. Can diagnostic studies of the lower urinary tract (urodynamic) or upper urinary tract (ultrasonography) predict and prevent an adverse change in kidney function?
3. What is proactive management?
4. Is proactive management better than reactive to maintain normal upper tract?

Guidelines

1. Obtain the following baseline studies within three months of birth:
 - Renal/bladder ultrasound and repeat in six months
 - Urodynamic testing
 - Serum creatinine³ (clinical consensus)
2. Initiate CIC and antimuscarinic therapy for the treatment of mixed incontinence when indicated based on the above results.³ (clinical consensus)
3. Consider the presence of a UTI when there is a fever (100.4 F / 38.0 C). In neonates less than one month of age with failure to thrive and dehydration.

Define a UTI by:

4. a positive UA, and
5. a positive urine culture (UC) on a catheterized specimen, and
6. fever (100.4 F / 38.0 C).

Define a positive Urine Analysis (+ UA) as:

7. >trace nitrite or leukocyte esterase on dip UA, and
8. >10 white blood cells/high power field (WBCs/hpf), uncentrifuged specimen, or
9. >5 WBCs/hpf, centrifuged specimen.

Define a positive UC (+UC) as:

10. >50,000 colony forming units/milliliter (CFUs/mL) (sterile specimen obtained by catheter or suprapubic catheter aspirate).
11. >100,000 CFUs/mL in a clean voided specimen.⁸

1-2 years 11 months

Clinical Questions

1. How can providers account for neurologic bladder changes due to growth and/or tethering?
2. What diagnostic tools are reliable to assess renal function?
3. Are upper tract changes reversible once they occur?
4. How should symptomatic UTIs be defined? What is the sequela of symptomatic UTIs? What is the optimal upper and lower urinary tract surveillance?
5. Does the use of proactive CIC and antimuscarinic medication help to maintain a normal upper tract?

Guidelines

1. Obtain renal/bladder ultrasound every six months when the child is under the age of two. After that, obtain an ultrasound yearly if the child is stable, without UTIs or imaging changes. (clinical consensus)
2. Obtain a renal/bladder ultrasound, as needed if the child has recurring symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain urodynamic testing yearly through age three. Repeat as needed if the following are noted:^{1,2,7} (clinical consensus)
 - bladder hostility
 - upper urinary tract changes
 - recurrent symptomatic UTIs
4. Obtain a serum creatinine test if there is a change in the upper urinary tract. (clinical consensus)
5. Assess suspected UTIs with a urine specimen obtained by sterile catheterization technique. Repeat a positive bag urine specimen with a catheterized specimen. (clinical consensus)

Define a UTI by:

- a positive UA, and
- a positive urine culture (UC) on a catheterized specimen, and
- fever (100.4 F / 38.0 C).

Define a positive Urine Analysis (+ UA) as:

- >trace nitrite or leukocyte esterase on dip UA, and
- >10 white blood cells/high power field (WBCs/hpf), uncentrifuged specimen, or
- >5 WBCs/hpf, centrifuged specimen.

Define a positive UC (+UC) as:

- >50,000 colony forming units/milliliter (CFUs/mL) (sterile specimen obtained by catheter or suprapubic aspirate).
- >100,000 CFUs/mL in a clean voided specimen. ⁸

6. Initiate CIC and antimuscarinic therapy for the treatment of mixed incontinence when indicated by upper urinary tract changes, recurrent symptomatic UTIs, or bladder hostility noted on urodynamic testing.²⁻⁴ (clinical consensus)

3-5 years 11 months

Clinical Questions

1. How can providers account for neurologic bladder changes due to growth and/or tethering?
2. What diagnostic tools are reliable to assess renal function?
3. Are upper tract changes reversible once they occur?
4. How should symptomatic UTIs be defined? What is the sequela of symptomatic UTIs? What is the optimal upper and lower urinary tract surveillance?
5. Does the use of proactive CIC and antimuscarinic medication help to maintain a normal upper tract?
6. Are the caregivers compliant with CIC? Who is performing CIC – the caregivers and/or the child?

Guidelines

1. Obtain a renal/bladder ultrasound yearly, if the child is stable. (clinical consensus)
2. Obtain a renal/bladder ultrasound as needed, if the child has recurrent symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain urodynamic testing only if the following are present: (clinical consensus)
 - upper tract changes
 - recurring UTIs
 - interest in beginning a urinary continence program
4. If the child is on CIC, begin to involve the child in the process of self-catheterization. ⁹ (clinical consensus) (Self-Management and Independence Guidelines)
5. Obtain a serum creatinine test if there is a change in imaging of the upper urinary tract. (clinical consensus)
6. Obtain serum chemistries (includes serum creatinine) at age 5. Assess suspected UTIs with a catheterized urine specimen. Repeat a positive bag urine specimen with a catheterized specimen. (clinical consensus)

Define a UTI by:

- a positive UA, and
- a positive urine culture (UC) on a catheterized specimen, and
- leakage between CIC, and
- onset of pelvic or back pain, and
- fever (100.4 F / 38.0 C).

Define a positive UA (+ UA) as:

- >trace nitrite or leukocyte esterase on dip UA, and
- >10 white blood cells/high power field (WBCs/hpf), uncentrifuged specimen, or
- >5 WBCs/hpf, centrifuged specimen.

Define a positive UC (+UC) as:

- >50,000 colony forming units/milliliter (CFUs/mL) (sterile specimen obtained by catheter or suprapubic aspirate).
 - >100,000 CFUs/mL in a clean voided specimen.⁸
7. Initiate CIC and antimuscarinic therapy when indicated by upper urinary tract changes, recurring symptomatic UTIs, or bladder hostility noted on urodynamic testing.²⁻⁴ (clinical consensus)
 8. Introduce urinary continence and discuss interest in beginning the program and options at each visit.⁹⁻¹⁰ (clinical consensus) (Self-Management and Independence Guidelines)
 9. Introduce bowel management and discuss interest and options at each visit. (clinical consensus) (Bowel Function and Care Guidelines)

6-12 years 11 months

Clinical Questions

1. What is the best way to measure renal function in the child that is non-ambulatory?
2. What social, environmental, and economic limitations or hurdles are encountered when working to achieve urinary continence?
3. What is worse: stool or urinary incontinence?
4. How we define urologic continence? Is the definition of continence congruent with the perspective of the patient, family, and physician?

Guidelines

1. Obtain a renal/bladder ultrasound yearly, if the child is stable. (clinical consensus)
2. Obtain a renal/bladder ultrasound as needed if the child has recurrent symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain urodynamic testing when initiating a urinary continence program, if the following are present: (clinical consensus)
 - upper urinary tract changes such as hydronephrosis or renal scarring
 - recurring symptomatic UTIs
 - changes in urinary continence status
4. Obtain a serum creatinine test yearly. If the child has low muscle mass, consider an alternative measure of renal function.⁵ (clinical consensus)
5. Obtain serum chemistries yearly on any child who has had urinary reconstruction.
6. Obtain a serum B12 level test every year beginning two years after urinary reconstruction.¹¹⁻¹³ (clinical consensus)
7. Discuss a urinary continence program and interest in beginning the program and options at each visit.⁹⁻¹⁰ (clinical consensus) (Self-Management and Independence Guidelines)
8. Discuss a bowel management program and the interest and options at each visit. (clinical consensus) (Bowel Function and Care Guidelines)

13-17 years 11 months

Clinical Questions

1. How is continence affected by a shift in responsibility to self-care?
2. How is a normal upper urinary tract affected by a shift in responsibility to self-care?
3. What is optimal surveillance of the upper and lower urinary tract?
4. If reconstructive continent bladder surgery was undertaken, would you do it again?
5. If no reconstructive surgery was undertaken do you wish it had been?

Guidelines

1. Obtain a renal/bladder ultrasound yearly, if the child is stable. (clinical consensus)
2. Obtain a renal/bladder ultrasound as needed, if the child has recurring symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain a serum creatinine test yearly. If the child has low muscle mass, consider an alternative measure of renal function.⁵ (clinical consensus)
4. Obtain serum chemistries including B12 yearly on any child who has had urinary reconstruction.¹¹⁻¹³ (clinical consensus)
5. Transition urologic care to self-management, if doing so is developmentally appropriate for the child.¹⁴⁻¹⁵ (clinical consensus) (Self-Management and Independence Guidelines)
6. Transition bowel program to self-management, if doing so is developmentally appropriate for the child. (clinical consensus) (Bowel Function and Care Guidelines)

18+ years

Clinical Questions

1. What is optimal surveillance of the upper and lower urinary tract? What cancer screening is needed?
2. How do we define UTI in the adult and when do we treat?
3. How do we minimize sequelae of secondary incontinence in adulthood?

Guidelines

1. Obtain a renal/bladder ultrasound yearly. (clinical consensus)
2. Obtain a renal/bladder ultrasound, as needed if the adult has recurring symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain a serum creatinine test yearly. If the adult has low muscle mass, consider an alternative measure of renal function.⁵ (clinical consensus) (Self-Management and Independence Guidelines)
4. Obtain serum chemistries including B12 on anyone who has had urinary reconstruction.¹¹⁻¹³ (clinical consensus)
5. Undertake cystoscopy and appropriate upper tract imaging in adults who have had a bladder augmentation when the following are present:¹⁶⁻¹⁸ (clinical consensus)
 - clinically-noted change in upper or lower urinary tract status
 - gross hematuria
 - recurrent symptomatic UTIs
 - increasing incontinence
 - pelvic pain
 - the adult has had a renal transplant with the presence of BK/polyomavirus
6. Evaluate patterns of continence/incontinence and address issues collaboratively with the individual and family. Include assessment of amount (volume) of incontinence as the amount in adults may be more bothersome than frequency.¹⁹
7. Continue to support self-management and independent living. (Self-Management and Independence Guidelines)

Research Gaps

Proactive treatment: The foundation of management is based on the ability to predict individuals at risk for kidney deterioration and then influence management prior to an adverse event.

1. What is the ability of urodynamic testing to identify individuals at risk?

2. Does early medical (e.g. intermittent catheterization) and pharmacologic (antimuscarinic) management based on urodynamic testing prevent upper tract deterioration?

Renal Function: Renal function is assessed through serum studies and imaging. However, it is not known what the best assessment is in the population with Spina Bifida.

1. How is creatinine influenced by height, weight and mobility status of a patient with Spina Bifida?
2. Is cystatin C a more accurate indicator of renal function in the population with Spina Bifida?
3. What degree of renal dysfunction has occurred by the time changes are noted on imaging (i.e., renal scarring in ultrasonography or DMSA)?
4. Are changes on imaging reversible?
5. Is yearly serum and upper tract testing necessary?

Urinary Infections: Chronic bacteriuria is suspected to have less of an impact on adverse health and renal deterioration than symptomatic UTIs.

1. What is the definition of a symptomatic UTI?
2. Does the definition of symptomatic UTI change with aging?
3. Do symptomatic UTIs in children under the age of five have greater morbidity?

Continence: Continence of the bowel and bladder plays an important role in socialization. The following only relates to urinary continence. Continence from a medical perspective is absolute (i.e. dry or wet).

1. Does the medical definition of absolute continence translate into a patient's and their family's quality of life?
2. Is the perception of continence from the perspective of the medical care provider and patient and family congruent?
3. Does achieving "some" degree of continence become beneficial?
4. Is there a threshold of "social" continence that is critical?
5. Is the cost (e.g. change in patient and family lifestyle, need for increased supervision, risk of intermittent catheterization, risk of medicines, and both short- and long-term surgical risk) worth the benefit?
6. What are the long-term challenges of patients who have undergone surgical intervention?
7. Would patients who have chosen surgery as a management option, make the same decision if they had the opportunity?

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