TABLE OF CONTENTS

Spina bifida

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Cite as: *Can Urol Assoc J* 2018;12(4Suppl1):S3-9. http://dx.doi.org/10.5489/cuaj.5272

Case

You received the following referral from one of your pediatric colleagues:

Please see this 19-year-old G1P0 female who had been referred to your office approximately two years ago. Unfortunately, she did not attend her scheduled appointment. Currently, she has a singleton pregnancy at 30 weeks gestation. Past medical history includes: myelomeningocele (L5–S1) closed at birth, VP shunt (revision x 3), ileal augmentation, Mitrofanoff, Malone antegrade continence enema (MACE) procedure, and history of bladder stones. Please see for long-term urological management and assistance at time of delivery if C-section is required.

Background

Neural tube defects (NTDs) are a group of diverse anomalies of the central nervous system caused by faulty closure of the neural tube during embryological development. The most common NTDs are spina bifida, anencephaly, and encephalocele. Due to dietary folic acid supplementation, the prevalence of all NTDs (including spina bifida) in Canada declined after 1996. Increased prenatal screening and diagnosis may have also contributed to the decline of NTDs. In 2007, spina bifida accounted for 2.7 per 10 000 births (live and still births) in Canada.[1]

The goals for management of the urological system in children with spina bifida change with age. For the newborn, preservation of renal function is the primary goal. Urine and fecal continence are added to the list of objectives in school-aged children. For the adolescent, independence and sexual function become important goals.[2] Reconstructive procedures that may require ongoing urological followup include bladder augmentation, continent catheterizable channels, incontinent diversions, antegrade continence enema channels, or buttons.

Spina bifida results in a variable amount of insult to somatic, sympathetic, and parasympathetic systems. Bladder and bowel function are adversely affected and patients may have lifelong issues with urinary and bowel incontinence, constipation, urinary tract infections (UTIs), urinary stones, renal dysfunction, and sexual and reproductive function. Lifelong followup by a urologist is recommended.[3] Close to 90% of adult spina bifida patients report an active urological issue.[3,4] Of these, up to 80–97% require some form of diagnostic, medical, or surgical intervention.[3,4] The most common urological problems identified in adult patients with spina bifida include: incontinence (29–52%), recurrent UTI (24–34%), catheterization troubles (6–12%), and calculi (9–19%).[3,4] One of the most frequently unaddressed issues prior to transition is sexual and reproductive function.[5] Thus, active urological issues are common and frequently require intervention, mandating frequent followup.[3]

What are the challenges in transitioning these patients?

Pediatric perspective

The responsibility of the pediatric urologist is to identify an adult/transitional care provider and prepare the child for transition, that is to ensure that they can manage their own healthcare, such as being able to take and order medicines, use medical supplies, and communicate with healthcare providers.[6] It is estimated that less than one-quarter of adults with spina bifida will live independently in early adulthood.[7] Thus, it may be unrealistic for all young adults to achieve all of these markers of independence prior to transition.

Some studies have shown dismal rates of successful transition, as low as 40%.[8,9] Those that did not transition to adult

urology care had a higher rate of emergency department use.[8] Transitioned spina bifida patients have identified a lack of assessment of financial/employment issues and not visiting the adult clinic prior to transition as barriers in transitional care.[10]

The multisystem nature of spina bifida results in transition issues that are specific to this population that differ greatly from other adolescents transitioning to adult care, including increased rates of obesity, sexual dysfunction, impaired cognition, depression, decreased mobility, incontinence, aging caregivers, lack of insurance coverage, loss of the school support system, and loss of peer groups.[2]

An effective transition process can be supported by providing an opportunity to visit the adult clinic prior to transition[10] and ongoing communication between the pediatric and adult caregiver, even after transfer of care.[11] Trust and personal relationship with their pediatric urologist was reported by patients to be one of the most important factors in successful transition.[5]

Adult perspective

Over the last decades, there has been an enormous improvement in spina bifida patients' survival rates to adulthood, from around 25% in the 1960s[12] to as high as 82% in 2011.[13] These changes can be explained by the introduction of antibiotic improvements, along with ventriculoperitoneal shunts in the mid 20th century and clean intermittent catheterization (CIC) in the 1970s.[14]

The first step in order to properly transition a pediatric patient into an adult-oriented healthcare system is to state clear goals to be achieved. In this regard, it seems that the objectives of the health system, caregivers, and patients are not always aligned.[15,16]

Clinical urological challenges in spina bifida patients transitioning to adulthood

Urological outcomes of adults with spina bifida are greatly affected by several non-urological aspects of the disease, i.e., mental compromise, spinal deformities, decreased mobility, Charcot's arthropathy, eating disorders, obesity, and decreased respiratory and bowel function, all of which are beyond the scope of this article. There are also many critical psychological issues secondary to spina bifida and its treatments.[5] Therefore, the literature is consistent in expressing the importance of creating clinics led by multidisciplinary and transition-trained teams.

In regard to urological issues in transitioning patients with spina bifida, different authors have consistently stated specific goals or fields to be managed.[15,17-19]

Self-esteem and sexuality

In order to achieve satisfactory sexual development, individuals need neurological, functional, and anatomical features that work reasonably well. Decreased mobility and anatomical deformities may be incompatible with sexual activity, while sensory disturbances can make these experiences less satisfactory. Additionally, a highly dependent daily life often causes reduced privacy, and difficulties with both bowel and bladder control reduce self-esteem.[20] Ultimately, these issues become barriers to adolescents becoming capable of intimacy.[17]

If these barriers are overcome, women are much more likely to enage in sexual activity than men,[21] probably due to a multifactorial erectile dysfunction rate that can be as high as 86%.[22] This rate is dependent on the compromised spinal cord level.[23] Risk factors for sexual issues have been found to be low self-confidence, hydrocephalus, and urinary incontinence.[24] In addition to the obvious effects of erectile dysfunction and ejaculatory disorders on impaired fertility, some studies have also shown a high frequency of poor semen quality and azoospermia.[25,26]

Issues concerning female fertility and pregnancy will be discussed below.

Renal function

From an overall survival perspective, preservation of renal function is the mainstay of the management of spina bifida patients.[27-29] In one study, around one-third of all spina bifida patients who died by 35 years old did so due to renal failure.[29] In the same way that they affect other non-urological spina bifida-associated problems, both the severity (spina bifida occulta, meningocele, or myelomeningocele) and the spinal level at which the lesion is located, also affect risk to renal function. Higher lesions entail more risk of detrusor-external sphincter dyssynergia (DSD) and of an elevated detrusor leak point pressure (DLPP), which can ultimately impair the kidneys.[30] High bladder pressures due to neurogenic bladder, vesicoureteral reflux, and UTI have typically been described as main causes of renal damage in this setting. However, problems secondary to urinary diversions, like either bladder/reservoir, ureteral or kidney stones, ureteroileal anastomosis stricture, high reservoir pressure, poor compliance with self-catheterization and unknown causes, have been described as well.[31] Hypertension can be as frequent as 23% of patients >20 years old,[32] while other factors, such as sedentary lifestyle, can contribute to increased risk of hyperfiltration and subsequent renal damage progression.[18] In spite of the reservoir-related complications mentioned, it seems the intestinal reservoirs do not cause kidney impairment per se,[31] even more, their use in order to obtain a "high capacity and low pressure bladder" has been vital to

prevent or to treat secondary renal failure when conservative management has failed.[33]

Bladder management

A cornerstone issue to be managed in individuals with spina bifida is bladder function, namely filling and emptying phases in both native and reconstructed bladders.

Native bladders

As previously discussed, a "low-pressure bladder" is critical for renal function protection. Several factors can affect spina bifida patients' bladder at the same time: pelvic floor or sphincteric deficiency, impaired compliance, and detrusor hyperreflexia. Unfortunately, it seems that bladder pressure, along with outlet resistance, increases during puberty.[34,35] Continence is also a critical aim in patients transitioning to adulthood because it is highly associated with psychosocial outcomes and self-confidence. In neurogenic native bladders, conservative management should be initiated early, including anticholinergic agents and CIC as first-line treatment or botulinum toxin A,[36] depending on patient characteristics. However, adolescent adherence to any long-standing treatment is low, and it is even lower with regard to CIC and clinic appointments.[37] In addition, some complications also arise during adolescence, such as urethral strictures or injuries, UTIs,[18,38] and bladder stones.

Reconstructed bladders

When conservative management has failed, patients often undergo surgery to create a low-pressure reservoir and achieve continence. This can be accomplished by means of a bladder augmentation or a bladder substitution, which in turn can be continent or incontinent, usually constructed using ileum or colon. Bladder neck surgery and catheterizable channels may be needed as well. Although these surgeries are the best way to create low-pressure-continent reservoirs in failed patients, they can carry numerous long-term complications related to the bowel tissue used, i.e., bladder/resevoir stones in >50% of patients, permanent mucous production,[39] vitamin B12 deficiency,[18] hyperchloremic metabolic acidosis, recurrent UTI, and reservoir perforation. There is also higher malignancy risk between 1.2–5.5% in the reconstructed reservoir, depending on the gastrointestinal segment used,[40,41] although it remains unclear whether this risk is due to the reconstruction itself or other factors.[42]

In summary, the main goals for these individuals are: 1) to achieve a compliant patient with 2) a compliant/non-obstructed/continent bladder or reservoir; and 3) to minimize the associated complications.

Urolithiasis and chronic UTIs

In the setting of adult patients with neurogenic bladder, there are several situations resulting in increased incidence (between 9.2 and 20.8%) of urinary stones.[3,4,43] These include indwelling catheters, urinary diversion, immobilization, incomplete emptying, stasis, and UTIs.[44] When comparing spina bifida with non-spina bifida patients treated for urolithiasis, the former were younger, needed more complex treatment, and had more severe complications than the latter.[44] There is an estimated rate of UTIs as high as 34%,[4] while stones or UTIs were involved in about 50% of spina bifida patients who were hospitalized.[45]

The main aims in this regard are to 1) prevent formation and to make a timely diagnosis of urinary stones; 2) develop a cost-effective method to detect UTIs in spina bifida patients; and 3) create a protocol to determine who benefits from treatment and who does not.

Recommendations for management of these challenges

General recommendations

Although there are no currently available clinical guidelines for the ongoing management of transitioned adult spina bifida patients, we recommend close followup of the upper and lower urinary tract to anticipate and intervene prior to significant complications where possible.

In general, these patients are followed at our centre according to European Association of Urology (EAU) guidelines for followup of neurogenic lower urinary tract dysfunction (NLUTD).[46] This stipulates annual history, physical examination, blood biochemistry, and urine microbiology. We do not perform upper tract imaging every six months, but do so annually. We also lack the capacity to perform annual urodynamic testing, but will perform this whenever there is a change in baseline lower urinary tract function or upper tract deterioration (either radiological or biochemical).

Planning a transitional urology clinic

The health transition concept has been defined and widely standardized by the Society for Adolescent Medicine in the U.S. as, "a purposeful, planned process that addresses the medical, psychosocial, and educational/vocational needs of adolescents and young adults with chronic physical and medical conditions as they move from child-centred to adult-oriented healthcare systems."[47] This wide definition can be applied to a transition process of any kind. Having a poor transition system has consistently been shown to decrease health-related outcomes in adulthood.[48,49]

In 2008, Viner stated three mainstays when planning a transitioning process as, "Firstly, prepare young people and their families well in advance for moving from pediatric to adult services and ensure they have the necessary skill set to survive and thrive there. Secondly, prepare and nurture adult services to receive them. Thirdly, listen to young people's views."[50]

The transitional urology clinic should be part of a tertiary referral centre. We suggest the creation, to the extent that it is possible, of a comprehensive database including all spina bifida patients being seen in every family medicine and pediatric clinic that will potentially refer patients. Collaboration between adult and pediatric urologists, as well as primary care providers, is needed to define the future network system design. Some approaches may include: 1) the pediatric urologist continues caring for the patient until adulthood; 2) the joint clinics are housed in a pediatric or adult centre; and 3) the adult urologist periodically visits the pediatric clinic while the transition process begins.

After basic statistics and the burden of disease are properly determined, the main plan can be then drafted. Starting from the very clear and well-designed transition plan created by the REACH clinic[15] as a baseline, we suggest some minor modifications taken from one of the most widely accepted frameworks,[51] namely the "Six core elements of healthcare transition"[52] (Table 1).

What I tell my patient to prepare for transition (pediatric urologist)

I suggest introducing the topic of transition several years before it is planned and at multiple visits. We offer families an opportunity to visit the adult clinic prior to transition. Age at transfer is determined by the personal development and intellectual capacity of the child. It may take place as early as 16 years in a cognitively normal and independent child. In children with significant cognitive delay, the transition is often delayed until they have completed high school. Approximately one-half of patients feel the appropriate age for transfer is 18 years.[11]

At our centre, we are very fortunate to have a dedicated adult spina bifida clinic, thus, in preparing families for transition, I focus on the similarities between the two clinics (i.e., both are multi-disciplinary, have a similar structure, and a dedicated clinic nurse to contact if problems arise between visits). It is helpful to highlight the importance of attending scheduled appointments with adult care provider and the need for lifelong urological followup to avoid significant complications, including renal dysfunction.

As few young adults with spina bifida live independently, I reassure families it is acceptable and encouraged for parents to continue to attend medical appointments with the transitioned adolescent, if that is their preference. I encourage parents to gradually provide their child with opportunities to become more proactive in their healthcare, for example, having early school age children perform their own catheterizations or teenagers call the pharmacy to refill their own prescriptions.

I reassure families that there will be an ongoing dialogue between the pediatric centre and the adult care provider. I inform them that I provide all relevant medical records to the adult care provider (all clinic notes, reports of imaging studies, urodynamic reports, and operative notes) and will be available to the adult care provider for discussion should the need arise. Most patients feel that written information about the transition process would be helpful.[11] One-fifth of patients perceive the adult hospital to be an inappropriate environment for young adults.[11] This lends support to the concept of a transitional or adolescent clinic to meet the specific needs of this population straddling the pediatric and adult domains of healthcare.

Table 1. Six core elements of healthcare transition

Stage	Age	Providers involved	Setting	Description
T1	12–14	Pediatric only	Pediatric clinic	– Discuss office transitions policy with youth & parents – Literature and educational material provided
T2	14–15	Pediatric, introduction to adult providers	Pediatric clinic	– Initiate a jointly developed transition plan with youth & parents – Introduction of sexual questionnaires – Assessment of readiness
T3	16–17	Adult, pediatric providers available if needed	Adult clinic	– Update transition plan & prepare for adult care – Review young adult's health priorities – Share portable medical summary and emergency care plan – Continued assessment of readiness
T4	>18	Adult only	Adult clinic	– Implement adult care model – Assist to connect with adult specialists and other support services, as needed – Feedback for transition process

Case followup

If not already performed, the patient will require the following information to be obtained:

1. Renal function
    a. Serum creatinine, electrolytes, BUN
    b. Imaging of the upper urinary tract, ultrasound
2. Bladder management
    a. Clinical evaluation: lower urinary tract symptons, urine leakage, bowel function, voiding diary
    b. Imaging of the lower urinary tract, ultrasound

Challenges

There is no evidence supporting the contention that spina bifida, by itself, can significantly reduce fertility in women,[53] however, there are many factors producing a lower pregnancy rate. Social and psychological issues (low self-esteem), along with reduced mobility and obesity, are probably the main reasons. Since spina bifida patients are at 4.1% risk of having myelomeningocele (MMC) children,[54] special care must be considered as patients consider pregnancy. These patients require a higher dose of folic acid during the prenatal period. Obesity (a frequent condition in this setting) is also a risk factor for fetal MMC.

Pregnancy problems in SB women can be divided into:

***Due to underlying SB**:* Lumbosacral scoliosis and pelvic abnormalities secondary to muscular atrophy can occur,[55,56] increasing the risk of abnormal fetus presentation, and make vaginal delivery much more difficult.[57] Achieving adequate hip abduction may be imposible, owing to an ankylosed joint, ultimately forcing the decision to perform a cesarean.[58,59] An incidence of latex allergy as high as 60% has been found in MMC patients and the number of prior surgeries is associated with an even greater incidence.[60-62] As in the case presented, a considerable proportion of SB patients have undergone ventriculoperitoneal shunting for hydrocephalus during early childhood. Pregnancy entails an elevated intra-abdominal pressure, while an enlarged uterus can compress the peritoneal catheter so that a shunt malfunction syndrome can develop.[54,63,64] Some authors favour vaginal delivery and to perform extraperitoneal cesarean only for obstetric reasons among individuals with a ventriculoperitoneal shunt because of the risk of bacterial contamination of cerebrospinal fluid.[65] During labour, MMC patients may have normal uterine contractions, however, the neurological abnormality may cause unnoticed onset of labour, along with uncoordinated pelvic floor contractions, leading to an obstetric emergency.[55]

Due to urinary diversion/cystoplasty: Issues during pregnancy can be associated with the reconstruction previously performed and with the subsequent anatomic changes, i.e., abdominal adhesions, uterine retroflexion, fixation of the bowel segment used (at three different sites: ureteral anastomosis, mesentery, and efferent segment); compression of the ureters; and mesentery stretching, which may affect blood supply. However, all of these risk factors are mainly theoretical and without empirical support.[66] Stenosis or prolapse of the stoma,[53,67,68] along with catheterizable tube elongation making catheterization imposible, have been described, with 16–60% of individuals with continent diversions needing an indwelling catheter.[53,66] Leakage has been reported from stretched conduit stomas when appliances do not stick properly.[23,53] Hydronephrosis may occur in up to 75% of patients with continent diversions, 33% of them ultimately requiring a nephrostomy tuve.[53] Pyelonephritis occurs in approximately 30% of diverted pregnant patients,[53] which is higher than in the general population and strongly associated with hydronephrosis.[66]

Despite many surgeons favouring elective cesarean section, there is a lack of evidence supporting this, and it is generally true that patients with ventriculoperitoneal shunts, augmentations, and MACE reconstructions are best served by vaginal delivery when posible.[53,65] Our patient in particular is at risk of injury to both the vascular supply of her many orthotopic bowel segments and to the segments themselves in the event of cesarean section, and will require close urological support in this instance. We prefer to be in attendance for all of these procedures.

Conclusion: Spina bifida patients do not seem to have significant problems in regard to fertility. Nonetheless, there are many obstetric, quality of life, anatomical/surgical, renal, and infection-related problems that need to be assessed and followed up throughout the pregnancy. By taking the necessary precautions, encouraging rates of term or near-term birth of healthy newborns can be achieved in this population.

What I tell my patient now that they have transitioned (adult urologist)

In general terms, these patients are often quite individual in their approach to health management due to their extensive interactions with the healthcare system throughout their young lives. As such, a degree of negotiation is frequently required to reach agreement on management goals.

When patients are first seen in the clinic, all clinical information is reviewed and any deficits in knowledge are filled in with appropriate investigations, including history and physical examination, blood work (particularly looking at renal function and metabolic disturbance), urinalysis, imaging (particularly kidneys/ureters/bladder [KUB] ultrasound) and urodynamic testing, as appropriate.

If we are able to determine that the patient's upper tracts are not at risk of deterioration, then management of all other urological concerns is discussed in terms of degree of bother as it relates to risk of intervention. Frequently, the decision will be made to simply follow a concern, as it causes no immediate bother to the patient and no risk to their longevity.

Close collaboration with the pediatric urologist is desired to ensure that we are not missing any prior concerns that may be ongoing.

Competing interests: Dr. Gray has been a sponsored CME speaker for Allergan, Astellas, and Pfizer; and has participated in a clinical trial supported by Bioness. The remaining authors report no competing personal or financial conflicts related to this work.

This paper has been peer reviewed.

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