

Rosalia Misseri

The Spina Bifida Association reports that 7 of every 10,000 children born in the United States have myelomeningocele (MMC) and that there are more than 166,000 people currently living with MMC [145]. Today, survival into adulthood is reported as high 85 % (at age 34 years, 94 % of those without shunts, and 75 % of those with shunted hydrocephalus are alive) [32]. With decreases in birth prevalence of MMC after the introduction of prenatal diagnosis and mandatory folic acid fortification of grain products in the United States, there may be more adults currently living with MMC in the United States than children [23, 32, 59, 107, 136, 140, 161]. Despite a large number of people living with MMC, there are no randomized controlled trials regarding urologic care in this group, and there is a paucity of literature describing long-term urologic outcomes in adults with MMC [153].

Children born with MMC are very readily identified. Many children have been diagnosed antenatally and few have had closure of the spinal defect before birth. MMC results in varying degrees of disability including paraplegia, orthopedic and neurologic abnormalities, and bowel and bladder dysfunction. Cognitive function may

be affected in varying degrees as well. Once born, these children are promptly treated for their neurologic lesions by pediatric neurosurgeons and are quickly evaluated by the multiple subspecialists that will carefully monitor their progress and treat them over the course of their childhood. This group of medical specialists often includes developmental pediatricians, pediatric neurosurgeons, pediatric neurologists, pediatric physiatrists, pediatric orthopedists, and pediatric urologists. Many centers offer meticulous and successful multidisciplinary care for the child with MMC.

Such meticulous care has led to much advancement in the management of MMC. As survival of those affected has improved, concerns over transition of youth to adulthood and care for the adult with MMC have increased. Transition of care has been advocated by many individuals and multiple organizations. Each has identified variable important components and potential barriers to the ideal process, consequently a good model transition/transfer of care remains elusive [3, 4, 17, 49, 51, 52, 114, 121].

Earlier studies have shown that up to two-thirds of adults with MMC do not routinely seek regular urological follow-up [60]. Bladder function may change with aging and complications after reconstruction may occur into adulthood. Little is known of urologic outcomes after transfer of care and the majority of literature evaluating the neuropathic bladder in the MMC patient focuses primarily on pediatric patients. In a center

R. Misseri, M.D. (✉)
Riley Hospital for Children at Indiana University
Health, 705 Riley Hospital Drive, ROC 4230,
Indianapolis, IN 46202, USA
e-mail: rmisseri@iupui.edu

with an established adult MMC clinic, Cox et al. found that the experience at the adult MMC clinic and the patients' perspective on the transition process were independent of urological and neurosurgical medical outcomes. Additionally, caregivers preferred the care received at the pediatric center [29]. Regardless of patient/caregiver sentiment, concerns that arise with aging such as those associated with sexual function, fertility, benign urologic disease (e.g., BPH), and screening for urologic cancer may be better addressed by "adult" urologists. Successful transition/transfer programs will likely require formal exchange of medical records including past management strategies such as catheterization schedules, need for anticholinergics, attention to the upper urinary tract, prior surgeries, and surgical reconstruction. For successful transfer, the patient must be agreeable to the change and the accepting urologist must be interested and knowledgeable in the care of the aging MMC patient regardless of the patient's level of function or dependency on a caregiver.

Evaluation

Careful attention and preservation of renal function remain most important across the lifespan. It remains the responsibility of the urologist, adult or pediatric, that this is not forgotten. Those with untreated neuropathic bladders secondary to MMC are at risk for complications including urinary tract infections, urinary incontinence, and deterioration of the upper urinary tract with potential loss of renal function. Most urologists would agree that the management of the neuropathic bladder and the reconstructed urinary tract require yearly and lifelong follow-up [2, 63, 116, 147, 153, 164]. This would include physical examination, renal ultrasonography, abdominal radiograph, and serum creatinine measurement. Despite this general agreement, there are no standardized recommendations for the follow-up interval or what follow-up would involve [13, 116, 151, 155]. In a systematic review of people with MMC over 18 years old, Veenboer and colleagues found that 58.4 % performed CIC,

13.8 % voided normally, 10.5 % had incontinent diversions, 5.9 % had complete incontinence, 4.3 % had an indwelling catheter, and 2.9 % empty by Crede [153]. This heterogeneous group may contribute to the difficulties of standardizing follow-up protocols.

The European Association of Urology (EAU) Guidelines on Neurogenic Lower Urinary Tract Dysfunction recommends urodynamics (UDS) every 1–2 years [112]. In a recent study, Veenboer et al. reported that unfavorable urodynamic findings are very unlikely in asymptomatic individuals who are not wheelchair bound. They concluded that it is probably not necessary to perform UDS as frequently as is currently recommended by the EAU [155]. UDS may be most useful if new symptoms, e.g., urinary incontinence, increased leakage between catheterization, and recurrent urinary tract infection, occur. Unfortunately, as management protocols differ and follow-up differs, the effects of intervention and evaluation remain largely unclear.

Attention to Renal Function

Since the introduction of the ventriculoperitoneal shunt (VPS), renal failure and its consequences became a leading cause of mortality in children with MMC surviving infancy, accounting for 20–30 % of deaths [61, 93, 141, 164]. Renal function is typically normal at birth, but may deteriorate over the patients' lifespan due to the functional abnormalities of the neurogenic bladder and/or recurrent infections [44]. Untreated, up to 50 % would develop chronic kidney disease (CKD) [165]. Urodynamic risk factors for the development of CKD include detrusor sphincter dyssynergia, detrusor overactivity, high detrusor leak point pressure, and high intravesical pressure. UTIs in childhood were also linked to renal dysfunction later in life [63, 150]. Thirty to forty percent of children with myelomeningocele have been reported to have some renal impairment [18, 105]. In an analysis of 52 adults with lumbar myelomeningocele, Persun and colleagues found that only 38 % had a normal ultrasound and a serum creatinine level less than 1.5 mg/dl despite

performing CIC and being dry between catheterizations [116]. In a systematic review, 72 % of people with MMC were found to have well-preserved function, 25.7 % with CKD stages 1–5, 1.3 % with ESRD. Up to 20 % were found to have VUR [153]. Veenboer estimates that ESRD was the cause of death of 8.9–28.6 % of the patients included in the review [153]. In a cohort of children born in the modern medical era, i.e., after the introduction of CIC for bladder emptying and widespread use of anticholinergics, Misseri and colleagues found renal failure was a rare cause of death in people with MMC after bladder augmentation. Mortality from renal failure was 0.5 % at 10 years after augmentation, lower than historical cohorts. The most common cause of death was non-urologic infection [98, 99]. In a separate birth cohort of 160 children with MMC managed in the modern era, no deaths were observed related to renal failure by age 20. This supports the notion that death from renal failure in well-managed MMC is now rare [91].

Estimated GFR (eGFR) using serum creatinine has been most commonly used to determine renal dysfunction in the MMC population. However, because of low muscle mass and underdeveloped musculature of the legs, creatinine-based methods to evaluate renal function are of poor sensitivity in this group. Once an abnormal serum creatinine level is noted, a loss of more than two-thirds of nephrons may have already occurred [119]. In the general population, the American Society of Nephrology suggests referral to a nephrologist if the estimated glomerular filtration rate is 60 mL per minute per 1.73 m² or less (stage 3 chronic kidney disease) or if macroalbuminuria is present. However, no specific recommendations exist for patients with MMC. Cystatin C- or iothalamate-based eGFR may assess global renal function in this group of patients, but this is not widely available for clinical use [117]. Despite its ability to estimate GFR, these modalities have never been compared with each other or with creatinine-based methods for MMC patients. In addition, these methods do not provide any information on renal scarring [1]. This knowledge is best gained with dimercapto-succinic acid (DMSA) scan. Up to 25 % of people

with MMC have been reported to have scarring on long-term follow-up. A history of vesicoureteral reflux history and febrile urinary tract infections were most commonly associated with abnormal DMSA scan in follow-up of patients older than 10 years with MMC [137].

Careful attention to changes in continence, recurrent UTI, weight gain, hypertension, and urolithiasis is very important as they may independently affect renal function. In addition, some of these conditions may also indicate a change in bladder dynamics. Each may ultimately lead to upper tract changes and renal disease. With each episode of pyelonephritis, nephrons are lost and renal failure may ensue. Antibiotic prophylaxis may be helpful in carefully studied patients. Kidney stones are also a risk factor for CKD and progression to ESRD [127]. Screening for and treatment of hypertension is important as well. Treatment with angiotensin-converting enzyme inhibitors may prevent progression to CKD [65, 162].

Attention to Bladder Function and Continence

Changes in continence, recurrent UTI, and urolithiasis are important indicators of bladder dynamics. Worsening continence may be related to poor adherence to catheterization schedules or anticholinergic use, changes in bladder function, or tethering of the spinal cord (though less common after periods of rapid growth) [164]. Incontinence of urine affects many life domains including the physical and the emotional. Skin breakdown, infection, poor self-esteem, social isolation, and underemployment/unemployment are often a consequence of incontinence [104].

Anticholinergics

Since the 1980s, “proactive” management of the neurogenic bladder has included CIC and anticholinergic therapy starting at in infancy [36, 41, 44]. The necessity of long-term anticholinergics and the effects of long-term use of this class of

drugs have not been critically evaluated in adults. However, evidence suggests that management with CIC and anticholinergics through puberty may result in improved maximal bladder pressure, bladder capacity, and detrusor leak point pressure in adults [2]. The most commonly used anticholinergic in children is oxybutynin both for its efficacy in the treatment of detrusor overactivity and urinary incontinence and because it is the only anticholinergic to be approved for use in children by the U.S. Food and Drug Administration. Oxybutynin is a nonspecific antimuscarinic agent blocking M1, M2, and M3 receptors. In blocking efferent parasympathetic M3 receptors innervating the detrusor muscle, bladder contraction is inhibited. Many side effects encountered by patients are due to its non-specific nature. Due to its high lipophilicity, neutral charge and small molecular structure, oxybutynin crosses the blood–brain barrier. The side effects of anxiety, somnolence, hallucinations, and cognitive dysfunction are related to its action on the M1 receptors in the brain [154].

Concerns regarding cognition in adults using anticholinergics have recently been raised; however, little is known regarding long-term use in people with MMC whose use often begins in childhood. Only one study has examined behavior and long-term use of anticholinergics in children with MMC. Veenboer and colleagues explored possible associations between long-term antimuscarinic use (from birth to median age 10.6 years) and behavioral problems in children with MMC and neurogenic bladder. No significant differences in behavior between children with MMC with and without long-term use of antimuscarinics were found [154]. In a double blind, placebo controlled crossover study, the use of anticholinergics was found to impair cognition in older adults without MMC [70]. A subsequent systematic review and meta-analysis could not determine if antimuscarinics effect CNS function since standardized measurement of age-stratified CNS outcomes are lacking [113]. Two pediatric case–control trials have examined the effects of oxybutynin on cognition and short-term memory in neurologically intact

children and neither study has found any untoward effects on short-term memory of cognition [53, 143].

Recurrent Urinary Tract Infection

Despite improvements in the management of the neuropathic bladder with respect to renal preservation, the incidence of urinary tract infection in people with MMC remains very high and continues to be a common cause of morbidity and impaired quality of life for patients with MMC. Unfortunately, definitions for UTI are variable and are infrequently applied in studies resulting in difficulties assessing the true UTI rates in this group [90]. Filler and colleagues report that up to 50 % of children will experience a UTI by 15 months and 44 % will have more than 5 UTIs by age 15 [46]. Other studies have demonstrated that the annual incidence of UTI in patients with neurogenic bladder is as high as 20 % [160]. The high incidence of UTI may be associated to intermittent catheterization, incomplete emptying, constipation, anatomic abnormalities, and calculi. Due to altered sensation, people with MMC may not have typical symptoms of UTI [166]. By the time most patients reach adulthood they are well aware of individual, specific symptoms that indicate UTI, e.g., lethargy, headache, increased mucous production. Differentiating asymptomatic bacterial colonization from true infection is important particularly in people performing CIC [132]. Cautious therapy of positive cultures is important to help prevent multi-drug-resistant organisms in this patient population. Treatment with multiple courses of antibiotics may lead to more virulent bacteria and a requirement for more toxic antibiotic treatments.

Catheterization

Clean intermittent catheterization is often introduced very early in life and remains necessary throughout the entire lifetime of a patient with

MMC. It is generally well accepted and becomes routine both for the child and his/her caregivers [44, 85]. Hematuria, false passages, urethral stricture, and epididymitis may occur as a consequence of repeated and long-term catheterization. Reports of such complications range from 0 to 40 % [20, 167].

Over a median of 16 years, Lindehall and colleagues found 25 % of boys had major urethral complications including false passage, urethral stricture, and metal stenosis. To prevent complication, the authors recommend the use of as large a catheter as the urethra can accept and to start training children in self-catheterization early [84]. The use of lubrication may also decrease the complications associated with catheterizations. In a similar study reviewing the complications in young females, the authors found very few complications. These included gross hematuria, temporary difficulty catheterizing, and urethral polyps in two patients. Difficulty inserting the catheter and hematuria resolved spontaneously or by the use of lubricant [86]. With aging, progression to wheelchair-bound status, increasing independence, and weight gain associated with adulthood, some patients (particularly females) may have increasing difficulty accessing their urethra for CIC, which may require formation of a catheterizable channel or suprapubic diversion.

Mobility

Mobility is affected in most people with MM and is an important determinant of quality of life [134]. The ability to ambulate is affected by the level of the lesion and often deteriorates over time. Decreased ability to ambulate appears to be related to a deterioration of the neurologic level of the lesion, increased spasticity, contractures of the knee and hip flexors, back pain, and lack of motivation. Major medical events such as surgeries, stroke, and lower limb edema were also found to contribute to the decline in ambulation [9].

Weight

Limitations in mobility, orthopedic deformities, cognitive impairment, and psychosocial issues may lead to an inactive lifestyle and resultant obesity. Dosa and colleagues found the obesity rate among adults with MMC at 37 %. This is more than twice the rate for children and adolescents with MMC. Extreme obesity was found in 11 % of adult women and 4 % of adult men with MMC [42]. Obesity may lead to further deterioration in function due to a decline in ambulatory status, difficulties with transfers, wheelchair propulsion, increased risk of pressure sores and other obesity-associated complications [6]. Obesity is a strong and independent risk factor for urinary incontinence in women and may contribute to worsening continence in MMC [159]. Peristomal hernias may occur in those with urostomies. Revisionary surgery and recovery becomes increasingly difficult in the obese patient.

Dexterity

Upper limb function is impaired in about two-thirds of children with MMC and may worsen with aging. These motor deficits include weakness, hand and finger dexterity, motor speed, and bimanual coordination [37]. Difficulties with dexterity increase dependence on others for activities of daily living including catheterizations.

Memory

Functional independence in people with MMC may be diminished due to problems with memory and deficits in executive function, e.g., planning, initiating, problem solving, affects [37, 89]. These deficits may be related to hydrocephalus, shunt malfunction, Chiari Type II malformation, dysgenesis of the corpus callosum, hypoplasia of the cranial nerve nuclei, gray matter ectopia, and diffuse microstructural anomalies [24, 38]. Such

difficulties may lead to poor adherence to catheterization schedules, medication regimens, and follow-up.

Postsurgical Considerations

Bladder Augmentation and Diversions

Bladder augmentation is a widely used and successful surgical option for the neuropathic bladder when medical management has failed. With refinements in medical management and improved techniques for augmentation, urinary diversion is rarely used [100]. Augmentation leads to improvements in continence, bladder capacity, and upper tract changes such as reflux and hydronephrosis/hydroureteronephrosis secondary to the hostile neuropathic bladder [26, 76, 88, 120]. A reduction in the incidence and severity of symptomatic urinary tract infections has been reported, and even when reflux persists, Krishna and colleagues found no evidence of deterioration of the upper tracts or progressive renal scarring [75]. Urinary tract reconstruction has been shown to lead to improvements in quality of life, self-image, self-esteem, and the ability to cope in women with neurologic impairment [158].

Despite the benefits, augmentation is a major surgical procedure with significant long-term surgical and nonsurgical complications. The most common and serious surgical complications are at the bladder level and include chronic bacteriuria, mucus production, formation of bladder calculi, metabolic abnormalities from exposure of the bowel to urine, bladder perforation, bowel complications, and the potential risk of bladder or bowel patch malignancy. Despite improvements in technique and advances in tissue engineering, the ideal technique and tissue for bladder augmentation and the creation of a functional bladder remain unknown.

Given that potential complications may occur at any time after surgery great care is mandatory when identifying and counseling surgical candidates. Lifelong attention to the surgically altered urinary tract is imperative. In a review of the first

500 bladder augmentations performed at Indiana University, complications were identified in 169 patients (34 %) resulting in a total of 254 surgeries. The cumulative risk of further surgery at the bladder level was 0.04 operations per patient per year after augmentation [94, 95].

Stones

Bladder calculi are increasingly recognized as a complication of bladder augmentation, affecting 11–52 % of patients and usually requiring an operative procedure [14, 35, 92, 94, 95, 109]. Stones tend to form in bladders that are incompletely emptied via catheterizable channels, those augmented with ileal or colonic segments rather than stomach and those made using absorbable staples [67, 77]. Recurrent urinary tract infections and noncompliance with irrigations, catheterizations and routine follow-up are other potential predisposing factors [27, 71]. While most stones are considered to be infectious, up to 30 % of cultured bladder calculi have been found to be noninfectious [98, 99]. This may be related to metabolic abnormalities such as chronic acidosis related to the augmentation or to chronic kidney disease. Hypercalciuria from osteodystrophy related to wheelchair dependence and poor mobility may also contribute to the high rate of calculi. Once a bladder stone is treated, the risk of recurrence is high, ranging from 15 to 29 % in less than 2 years [111]. Although open cystolithotomy is often used in cases of large or multiple stones, endoscopic management has gained popularity. Endoscopic therapy offers high stone removal rates and low complication rates [129]. The approach avoids an abdominal incision and entry into the peritoneal cavity and may obviate suprapubic tube placement in some patients [123]. However, endoscopy via the urethra or a channel carries the risk of damaging the urethra, catheterizable channel or BN repair when performed to treat a large stone. In addition, residual fragments left in the bladder after stone fragmentation can become a nidus for bladder stone recurrence. This risk may be decreased by using ultrasonic lithotripsy inside an endoscopically

introduced entrapment bag [78] or by removing calculi intact by an open or percutaneous approach. Previous attempts at comparing recurrence rates by surgical technique did not detect a statistically significant difference but were performed in small series (up to 31 patients) and did not account for method of stone fragmentation [67, 77]. In a recent study, Szymanski and colleagues found that bladder stones recurred in nearly half of patients within 9 years after first stone surgery, independent of treatment technique and patient characteristics [148].

Malignancy

Several case studies of bladder cancer developing after bladder augmentation have been published [7, 8, 21, 47, 55, 79, 106, 142, 156]. Soergel et al. reported transitional cell carcinoma in 3/260 patients with neuropathic bladder who had undergone bladder augmentation [142]. Tumors in gastric segment appear to occur more frequently and sooner after augmentation when compared to ileal segments [21, 156]. In a review of 153 augmented patients with a minimum of 10-year follow-up after augmentation, Husmann and Rathbun found a 4.5 % incidence of bladder cancer [62]. Patients with coexisting carcinogenic stimuli (prolonged smoking/chronic immunosuppression), or the inherent risk of malignancy as in bladder exstrophy, had a higher risk of cancer in their series.

A full review of this topic can be found in Chap. 11.

Perforation

The spontaneous perforation of an augmented bladder is an uncommon but very serious complication. The prevalence of spontaneous bladder perforation is reported between 6 and 13 % [11, 34, 48, 76, 94, 95, 135]. Perforation can result in peritonitis, sepsis, and death. The diagnosis may not be straightforward due to impaired sensation and varying levels of developmental disability in many people who have had an enterocystoplasty.

Perforation may be due to traumatic catheterization, overdistension, chronic infection, ischemic necrosis of the intestinal segment, and increased intravesical pressure [5, 10, 30, 45, 125, 128]. Treatment usually requires emergent laparotomy, but conservative management has been successful in select cases. This approach has usually been reserved for patients without ventriculoperitoneal shunts [118]. In a review of 500 patients with augmentations, spontaneous bladder perforations were identified in 43 patients, for an overall risk of 8.6 %. The calculated risk was 0.0066 perforations per augmentation-year at risk. Approximately a third of the cases had perforated within 2 years of surgery, a third between 2 and 6 years postoperatively, and a third at more than 6 years after augmentation, highlighting the need for long-term follow-up. The use of sigmoid colon and bladder neck surgery were associated with an increased risk of perforation. The presence of a continent catheterizable channel was associated with decreased risk of perforation [94, 95].

Acid Base Disturbance

Due to the absorptive nature of bowel, metabolic changes may occur when enteric segments are introduced into the urinary tract. Given the small segments of bowel used, metabolic acidosis is rare in patients with normal renal function undergoing bladder augmentation [56, 96]. Hyperchloremic metabolic acidosis may result if ileal and/or colonic segments are used. When the ileum or the colon is exposed to urine ionized ammonium and chloride are reabsorbed. Ammonium absorption may also occur through substitution for sodium in the sodium–hydrogen antiport. The exchange of ammonium for a proton is coupled with the exchange of bicarbonate for chloride. Ionized ammonium may also be absorbed into the blood through potassium channels. Thus, bicarbonate and some potassium are lost [72]. Metabolic derangements may occur with the use of other enteric segment such as stomach or jejunum. Hypochloremic hypokalemic metabolic alkalosis may occur if gastric segments are used. The concentration of gastrin

seems to be important in this syndrome, as metabolic alkalosis becomes more severe with higher gastrin levels [149]. Though rare, when jejunum is used for urinary diversion, hyponatremic, hypochloremic, hyperkalemia, azotemia, and acidosis may develop [31]. Metabolic derangements are not isolated to augmented bladders and urinary reservoirs. Mild acidosis can be expected in up to 15 % of patients with an ileal conduit, of whom up to 10 % will require treatment [16, 22, 133].

Chronic acidosis may play a major role in the decrease in bone mineral density after bladder augmentation or urinary diversion. The majority of studies suggest that linear growth is not affected by bladder augmentation. Varying degrees of metabolic acidosis appear to resolve with no affect on linear growth [97]. However, prolonged acidosis could lead to osteomalacia and osteoporosis in adults [146]. In addition to the correction of the acidosis, dietary supplements with calcium, vitamin D and, in severe cases, bisphosphonates are recommended [115, 124, 138, 146].

In adulthood, progression of renal insufficiency teamed with worsening respiratory function (which can be restrictive airway disease, obstructive sleep apnea, and/or central apnea) can promote systemic acidosis. Baseline metabolic parameters should be carefully considered before any patient undergoes a surgery to change a patient from an incontinent, non-bowel-containing reservoir to a continent, bowel-containing reservoir because this may alter an important respiratory compensatory mechanism.

Vitamin B₁₂ Deficiency

A reported sequelae of the use of ileum for enterocystoplasty is vitamin B₁₂ (cobalamin) deficiency. A cobalamin concentration of 200 pg/mL is commonly regarded as the threshold below which supplementation should begin. The mechanism of the deficiency has been hypothesized to be secondary to removal of the distal ileum, which is the principal site of vitamin B₁₂ absorption. Vitamin B₁₂ plays an important role in DNA synthesis and

neurological functions. While most deficiency is asymptomatic, it can result in megaloblastic macrocytic anemia and potentially irreversible neurological changes, such as peripheral neuropathy, loss of positional and vibration sense, balance difficulty and dementia [57, 87]. Reduction in serum B₁₂ level may occur over a long period of time following ileocystoplasty and may require long-term serial vitamin B₁₂ measurements [12]. Replacement may be parenteral or oral. Oral vitamin B₁₂ replacement has been found to be well tolerated and highly effective in increasing serum levels to the normal range [152].

Attention to Conduit Urinary Diversions

Despite the popularity of bladder augmentation for those who have failed medical management, there are a myriad of surgeries performed with the goal of protecting the upper urinary tract from deterioration. These include sphincterotomy, detrusor injection of botulinum toxin, ileal chimney, and incontinent conduit diversion. In those with spinal cord injury, mortality from upper urinary tract disease after ileal conduit is historically reported at 25 % [28, 74, 102]. More recent studies show preservation of adequate renal function in almost all patients [25, 68].

People with MMC may be at increased risk of incisional and parastomal hernia. With long-term follow-up hernias may occur in up to 37 % of patients [66]. Risk factors for hernias include prior abdominal surgery, obesity, poor nutrition, chronic constipation, and poor abdominal musculature. Incisional hernias may include incarcerated viscera or enteric fistulization. Parastomal hernias are most common after ileal conduit/loop creation and may present with a poorly fitting stomal appliance or with bowel or urinary complaints [163]. Parastomal hernias are rarely encountered after catheterizable channels.

In addition to parastomal hernias, other complications of incontinent diversions include bleeding, skin irritation, prolapse, stenosis, ileoureteric anastomotic stricture, and retraction of stoma [25, 69, 74, 139]. Skin breakdown may be

related to poorly fitting appliances or retraction of the stoma. Stomal obstruction may lead to hydronephrosis, recurrent infections and/or renal failure from obstruction of the ileal conduit. Urinary tract infection may occur in up to 60 % of such patients [25, 68, 69, 139]. Pyocystis may also be encountered in those with a bladder left in situ. Periodic bladder irrigation, iatrogenic vesicovaginal fistula formation and cystectomy may become necessary when conservative methods fail. Marsupialization of the urethra and vagina in females as described by Spence with pyocystis may also prove effective and low risk for these patients [144]. Chronic metabolic acidosis has been reported in up to 20 % of patients after ileal conduit [73]. Renal, ureteral, and conduit calculi may occur in up to 40 % [69, 73, 74].

Attention to Mitrofanoff

The introduction of the catheterizable channel, whether utilizing the appendix or a channel constructed with tubularized ileum has significantly changed our approach to the care of neuropathic bladders [101, 103, 168]. Channels allow an alternative to catheterizing per urethra or, in the case of bladder neck reconstruction or closure, a substitute for the urethra. Placed in the lower abdomen or umbilicus, channels may increase independence or decrease the burden on caretakers, allowing the patient to be catheterized without transferring out of the wheelchair and removing clothes and undergarments.

Surgical revisions of catheterizable channels are not uncommon. With long-term follow-up in large series, up 39 % require revision [19, 82]. Complications may be relatively simple to correct as in stomal stenosis or may require extensive revision at a subfascial level due to channel incontinence, kinking, or angulation of the channel. Stomal stenosis rates as high as 50 % have been reported after long-term follow-up [82, 83]. Perforations and false passages may occur requiring creation of a new channel. Inflammatory or granulomatous polyps may occur due recurrent catheter trauma [43, 122]. These may be treated

endoscopically [126]. It has been hypothesized that early complications are related to the healing process, and that the later complications are related to wear and tear of the conduits and changes in body habitus [82]. Based on a retrospective review of over 500 patients, Cain and colleagues found that Monti channels were two times more likely than an appendicovesicostomy to undergo subfascial revision overall. The spiral Monti to the umbilicus, in particular, was five times more likely than the appendicovesicostomy to undergo revision [19].

Sexuality and Sexual Function

Sexual function is a complex phenomenon involving desire, motivation, arousal, and orgasm. All aspects are poorly understood in adults with MMC. In a person with MMC, all or none of these may be affected. Sexuality in patients with congenital disabilities, including MMC may be affected for reasons including impaired self-esteem, dependence on caregivers, and lack of privacy [54]. Despite this, Hirayama and colleagues found 95 % and 100 % of male MMC and 83 % and 75 % of female MMC to have interest in the opposite sex and sexual desire, respectively [58]. In a study of 76 young adults with MMC, Lassmann and colleagues found that 24 % were sexually active. Sexual activity was not related to gender, degree of urinary incontinence, or extent of physical disability, but it was more likely in patients with lower level lesions. Interestingly, sexual function was not found to affect health-related quality of life in these patients [80]. In addition to the level of the lesion, hydrocephalus has also been found to affect negatively sexuality. Verhoef and colleagues found that sexually active patients were significantly less likely to have hydrocephalus [157].

The response to sensory stimulation and the ability to orgasm may be affected as the pudendal nerve (S2–4) is often compromised in MMC. A distinct neurological level that favors better sexual function has not been defined; however, lower

and less severe lesions are more favorable [33, 80]. In a study of young women with MMC, Roberts and Sawyer found 80 % of women with MMC had some genital sensation and 37 % of them had experienced orgasm [131]. Vulvar sensation and orgasm are rare in women with lesion at or above L2 [39].

Sexuality in men with MMC and normal cognitive development is similar to that of healthy peers, with 80–100 % reporting desire, fantasy, and interest in sexual activity [58, 130]. However, sexual activity in men with MMC is often delayed and more common in the subset of older men who live away from their parents [50]. Erectile dysfunction affects approximately 75 % of adult men with MMC and is most dependent upon the level of neurologic lesion. Sixty-four percent of men with a lesion at T10 or lower are reported to have erections compared to only 14 % with a higher lesion [40, 50]. In a study of 22 men with MMC, 95 % achieved erection by visual stimulation and in 86 % by tactile stimulation. However, only 27 % of the patients with erections were satisfied with penile rigidity. Ejaculation and orgasm was noted in 67 %. Orgasm was more frequently seen in patients whose external sphincter activity was maintained [58]. Ejaculation has been reported to be dripping in nature and may not be perceived as orgasmic due to absent penile sensation [15, 33]. Sildenafil has been reported to improve erectile function by 80 % in men compared to baseline and placebos, with 50 mg providing greater improvement when compared to 25 mg [110]. Recently, anastomosis of the dorsal nerve of the penis to the intact ipsilateral ilioinguinal has been described to improve penile sensation due to neural tube defects and low spinal cord lesions [64, 108].

In men actively attempting fatherhood, paternity rates are reported to be between 56 and 73 % and are more likely in men with L5 or sacral lesions [33, 81]. Despite high a rate of ED and infertility, normal testosterone production has been demonstrated in 90 % of men with MMC [33].

A more in-depth review of these issues, including special considerations for pregnant women with MMC, can be found in Chaps. 5.

Summary

- The following best practices are based on a review of the literature. To date, most studies are observational.
- As bladder function may change with aging and complications after reconstruction may occur into adulthood, life-long, yearly urologic care is recommended.
- Urologic follow-up should include physical examination, renal ultrasonography, abdominal radiograph, and serum creatinine measurement.
- The use of routine urodynamics is controversial. The European Association of Urology Guidelines on Neurogenic Lower Urinary Tract Dysfunction recommends UDS every 1–2 years. However, UDS may be most useful if new symptoms, e.g., urinary incontinence, increased leakage between catheterization, changes in upper tract imaging, e.g., hydronephrosis and recurrent urinary tract infection, occur [155].
- In those with bladder augmentation, bladder substitution and urinary conduits, additional concerns include acid–base disturbances, vitamin B12 deficiency, calculi, malignancy, perforation, hernias, and obstruction.

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Transition and Lifelong Care in Congenital Urology

Wood, H.M.; Wood, D. (Eds.)

2015, X, 222 p. 33 illus., 30 illus. in color., Hardcover

ISBN: 978-3-319-14041-4

A product of Humana Press