Neurourology and Urodynamics 26:978–980 (2007)

REVIEW ARTICLE

Urological Follow-Up of Adult Spina Bifida Patients

Imran Ahmad* **and Paraskeve Granitsiotis**†
Department of Urology, Southern General Hospital, Glasgow, Scotland, United Kingdom

Aims: The vast majority of the current urological literature understandably has concentrated on the management of children with spina bifida, because in the past the majority did not survive into adulthood. With improvements in the understanding and multidisciplinary care of spina bifida patients it has become a disease of adults. Our aim was to evaluate the current literature to attempt to formulate evidence based guidelines for the management of this difficult group of patients. **Methods:** We reviewed the literature on adult urological management of spina bifida, all relevant articles which concentrated on adults and long-term management were studied in full. **Results:** Renal function may begin/continue to deteriorate into adulthood, becoming the leading cause of adult death. This is thought to occur because of changes in the adult bladder, with increases in storage pressure. Medical and surgical management should aim to preserve renal function as well as the maintenance of continence in the face of the growing and changing urinary tract. Follow-up should be regular and in the context of a specialist multidisciplinary clinic. Despite being unvalidated in the follow-up of adult spina bifida patient's annual serum creatinine, ultrasound and urodynamics are currently the best tools available. **Conclusions:** There is no reason why the majority of spina bifida sufferers cannot use their own kidneys for the rest of their lives. This however relies on urological treatment being instigated soon after birth and continuing into adulthood. *Neurourol. Urodynam. 26:978–980, 2007.* © 2007 Wiley-Liss, Inc.

Key words: follow-up; spina bifida; urodynamics

INTRODUCTION

The majority of urological literature has concentrated on the management of children with spina bifida, because in the past the majority did not survive into adulthood. With improvements in the understanding and multidisciplinary care of spina bifida patients it has become a disease of adults. There is the trend that children who are continent with no signs of upper tract damage or other urological problems are discharged from their urologists' care to be haphazardly followed up by their general practitioners. However it is becoming increasingly clear that a proportion of these adults continue to be at risk of further upper urinary tract deterioration. Often when these patients are referred to the urology services it is too late for any meaningful interventions since the lasting damage has already occurred.

Our aim was to evaluate the current literature to attempt to formulate evidence based guidelines for the management of this difficult group of patients.

MATERIALS AND METHODS

We reviewed the literature on adult urological management of spina bifida, searching using the keywords "spina bifida, meningocele and myelomeningocele." A Medline search of the English literature from January 1966 to December 2006 inclusive was done on OVID. All relevant urological articles which concentrated on adults and long-term management were studied.

RESULTS

Background

The incidence of spina bifida in the United Kingdom is approximately 0.50–0.70/1,000, having fallen dramatically with the recognition of the effects of folic acid as well as improvements in prenatal care.[1]

Over 90% of infants with spina bifida have normal renal function at birth. If unattended, however about 50% will have renal deterioration.

Urological management of spinal bifida has undergone multiple changes over the past 20–25 years. Initially treatment was aimed at urinary diversion, with no attempt to alter bladder behavior. It was soon recognized this procedure created its own problems. Effective shunting devices to treat the co-existent hydrocephalus have meant that an increasing number of patients with myelomeningocele have survived and thus it has become the commonest condition that presents urologists with neurogenic bladder dysfunction to

No conflict of interest reported by the author(s).
Chris Winters led the review process.
†Consultant Urological Surgeon.
*Correspondence to: Imran Ahmad, Clinical Research Fellow, Flat 2/2, 80 Beith Street, Glasgow G11 6DQ, Scotland, UK. E-mail: imranahmad@doctors.net.uk
Received 17 January 2007; Accepted 27 March 2007
Published online 11 July 2007 in Wiley InterScience
(www.interscience.wiley.com)
DOI 10.1002/nau.20447

urologists. In the 1970s and 1980s a greater understanding of functional derangement of the bladder and urethra was achieved with by urodynamic evaluation, allowing a more rational treatment approach to evolve.

Lapides introduced the concept of clean intermittent catheterization (CIC) to empty the bladder.[2] This method is now universally accepted as the most popular option for patients with myelodysplasia to achieve continence and prevent upper tract deterioration. Our improved understanding of bladder dysfunction has allowed us to use pharmacological agents to alter this abnormal detrusor behavior.

Current surgical techniques focus on improving functional bladder capacity by promoting continence, lowering bladder pressure, and protecting upper tracts from further damage.

Urodynamics

Urodynamic patterns can range from normal to severe bladder and sphincter dysfunction. Some degree of bladder atonicity in conjunction with a fixed open sphincter is found in most children with myelodysplasia.[3,4] Low storage pressures or high and unsafe intravesical pressures may also occur. In these children the greatest concern is detrusor sphincter dyssynergia (DESD). This type of bladder occurs in approximately 30% of newborns and is associated with high rates of upper urinary tract deterioration.[4] It is now well known that urodynamic variables that seem to be linked to upper tract deterioration in children include detrusor leak point pressure,[3] detrusor-sphincter dyssynergia,[5] the maximum urethral closure pressure,[6] neurogenic detrusor overactivity,[5] and vesical compliance.[7] Due to this it is accepted that children need to be evaluated regularly because of the frequent changes during the first few years of life.[8]

Renal

Lawrenson et al.[9] showed that adults with spina bifida have an eight times age standardized risk of renal failure as compared to the adult population. Figures also suggest that renal failure is the leading cause of adult spina bifida death, especially in those with sensory levels above T11.[10,11]

Lewis reported that the chance of renal failure increased with age.[12] Cortical damage was of the order of 13.3% in children under two with spina bifida, but increased to 27.3% in those older than 10 years.

Muller[13] reported evidence of renal damage in up to 40% of patients, but other recent studies have not provided accurate data on progression.

Capitanucci et al.[14] followed up a cohort of patients who had a variety of treatments including CIC, anticholinergic drugs, and surgery. Long-term follow-up (2–14 years) revealed that socially acceptable continence was achieved in 78% of 58 children. Upper urinary tract deterioration occurred in 15% and renal failure in 7.5%.

Rickwood et al.[15] assessed 181 adolescents and young adults with spina bifida. At the age of 16 years, abnormalities were present in 41%: the risk of these complications was related to the extent of neurological deficit and the presence of urinary diversion. During follow-up, fresh or further deterioration of the upper tracts occurred in 27%. This was influenced by whether or not previous urinary diversion had been performed. However, even without diversion, the incidence of renal damage is nearly doubles after puberty.

The incidence of hypertension also increases after puberty, both in those with scarred and normal kidneys. Even in those with normal kidneys, the incidence rises in the third decade. This may be due to an autonomic neuropathy or unrecognized renal disease.[16] The likelihood is that kidney damage leads to hypertension in these patients.

When urine is stored in intestinal reservoirs the incidence of renal deterioration is approximately 20% at 10-year follow-up, with complications such as stones, infection, and failure to catheterize the main causes of damage.

Persun et al.[17] evaluated 40 adult spina bifida patients with no history of renal failure or predisposing conditions to renal failure. They followed these patients up with renal ultrasounds and serum creatinine. Of the 20 patients with abnormal renal ultrasounds and/or elevated serum creatinines, 6 (30%) had elevated storage pressures at their average catheterized volumes. The causal relationship between elevated storage pressure and upper urinary tract deterioration was defined by McGuire et al.[3] He demonstrated that an intravesical pressure exceeding 40 cm H_2O at the time of urethral leakage in myelodysplastic patients predicted eventual renal decline. Thus Persun et al.[17] identified that stable appearing adults that would normally have been discharged by their urologist may still develop renal tract damage by silently generating high bladder pressures. The etiology of these high pressures is still unknown. Explanations include DESD, atrophy, and fibrosis of the striated muscle in the sphincter, decreased bladder compliance or a non-compliant urethral closure mechanism.[18]

Tethered Spinal Cord

The tethered spinal cord may develop years after surgery, but its incidence remains unknown. Clinical manifestations of a tethered spinal cord are the result of ischemic changes from tension on the cord. The areas most likely affected influence bladder and sphincter function.

Tarcan et al.[19] followed up 25 newborns with normal urodynamic studies at birth that had had early repair of their spinal cord defect. He found that during the follow-up (mean 9.1 years) a neurourological deterioration occurred in 8 (32%) of children. Subsequent MRI revealed a tethered spinal cord in all these children, which subsequently was repaired with mixed results.

Bladder

Yamamoto et al.[20] followed 228 patients into adulthood. He studied parameters which included upper urinary tract deterioration (hydroureter and/or hydronephrosis), vesicoureteral reflux (VUR), and bladder deformity. He found these parameters deteriorated/developed in 9.3%, 8.0%, and 29.3% respectively in adulthood. This at least indicated that the adult urinary tract continues to change and further analysis of the urinary tract in these adult patients is warranted.

There is the distinct impression that bladder function deteriorates at puberty. Whether this is just a teenage rebellion we are witnessing remains to be shown.[16] The adolescent may develop increased bladder outlet resistance, presumably from growth of the prostate in males and oestrogenization of the urethra in females. Thus a patient who becomes dry at puberty may actually have developed a raised leak point pressure and thus be at risk of renal failure.[16]

Management

There is little data on the outcomes in the management of patients with renal failure. The stigmata of spina bifida means

that many of these patients were not considered for dialysis or renal transplant. Those that are treated have results that are as good as those for the normal population and much better than the diabetic subgroup of patients.[21,22]

The graft survival rates appear to be similar to that for other groups of patients with abnormal but non-neuropathic bladders (70% at 5 years).[22]

Follow-Up

Hunt revealed that up to two thirds of adult spina bifida sufferers had no regular urological follow-up.[10]

Urological follow-up is essential, even well into adult life, since renal or bladder function frequently deteriorates into adult life. There is a wealth of data suggesting regimes for follow-up of spina bifida children. There however are no such studies looking at the effectiveness of screening tests in the adult spina bifida population.

A recent study shows serum creatinine as a good choice for frequent evaluation of renal function.[23] It gives a good baseline and the trend will be useful in detecting deterioration. However serum creatinine will be normal as long as the contralateral kidney function is intact (even down to 20% of function). This also makes creatinine clearance a poor measure for renal function assessment in spina bifida patients.[24]

Follow-up with ultrasounds of the kidney is a reliable monitoring method, however permanent deterioration in renal function may have occurred by the time changes are seen in the upper tracts.[25,26] Patients do not have to be subjected to radioisotope scans if serum creatinine and ultrasound scans are normal, especially if urodynamics do not indicate high intravesical pressures. Regular urodynamics might be one way of analyzing the changes in the adult urinary tract as well as ensuring that CIC and antimuscarinic treatments are being used to their full potential. Late changes in bladder behavior usually indicate a change in neurological status, such as a tethered cord, and is an absolute indication for neurological opinion.

As important as the above are, it is essential that the patient is regularly seen by the urologist to ensure compliance with CIC and their antimuscarinic medications.

Specialist clinics such as those of Dik et al.'s[24] have excellent results. At these "one-stop" clinics patients can have all their investigations and a chance to see all the specialists within one day. They may not see them all the at same check up, and some specialties may even discharge them, but it is essential that some form of formal follow-up exists to allow early detection of deterioration before it is too late for any meaningful interventions. Meticulous attention to every aspect of care is essential to allow these children to achieve their full potential.

CONCLUSION

There is no reason why the majority of spina bifida sufferers cannot use their own kidneys for the rest of their lives. This however relies on urological treatment being instigated soon after birth and continuing into adulthood. Follow-up should be regular urological assessment in the setting of a specialist multidisciplinary clinic with the aims of preservation of renal function as well as the maintenance of continence in the face of the growing and changing urinary tract.

REFERENCES

1. Poppas DP, Bauer SB. Urological evaluation of the myelodysplastic child. AUA Update Ser 1997;16:36.
2. Lapides J, Diokno AC, Silber SJ, et al. Intermittent clean self catheterisation in the treatment of urinary tract disease. J Urol 1972;129:1120–2.
3. McGuire EJ, Woodside JR, Borden TA, et al. Prognostic value of urodynamic testing in myelodysplastic children. J Urol 1981;126:205–9.
4. Sidi AA, Dykstra DD, Gonzalez R. The value of urodynamic testing in the management of neonates with myelodysplasia. J Urol 1986;135:90–3.
5. Bauer SB, Hallet M, Khoshbin S, et al. The predictive value of urodynamic evaluation in the newborn born with myelodysplasia. JAMA 1984;252: 650–2.
6. Kobayashi S, Shinno Y, Kakizaki H, et al. Relevance of detrusor hyper reflexia, vesical compliance and urethral pressure to the occurrence of vesicourethral reflux in myelodysplastic patients. J Urol 1992;147:413–5.
7. Gonhiem GM, Roach MB, Lewis VH, et al. The value of leak point pressure and bladder compliance in the urodynamic evaluation of meningomyelocele patients. J Urol 1990;144:1440–2.
8. Spindel MR, Bauer SB, Dyro FM, et al. The changing neurourologic lesion in myelodysplasia. JAMA 1987;258:1630–3.
9. Lawrenson R, Wyndaele JJ, Vlachonikolis I, et al. Renal failure in patients with neurogenic lower urinary tract dysfunction. Neuroepidemiology 2001; 20:138–43.
10. Hunt GM. Open spina bifida: Outcome for a complete cohort treated unselectively and followed into adulthood. Dev Med Childhood Neurol 2000;32:108–18.
11. Singhal B, Mathew KM. Factors affecting mortality and morbidity in adult spina bifida. Eur J Pediatr Surg 1999;9:31–2.
12. Lewis MA, Webb NJ, Stellman-Ward GR, et al. Investigative techniques and renal parenchymal damage in children with spina bifida. Eur J Pediatr Surg 1994;4(S1):29–31.
13. Muller T, Arbeiter K, Aufricht C. Renal function in myelomeningocele: Risk factors, chronic renal failure, renal replacement therapy and transplantation. Curr Opin Urol 2002;12:479–84.
14. Capitanucci ML, Lacobelli BD, Silveri M, et al. Long term urological follow-up of occult spinal dysraphism in children. Eur J Pediatr Surg 1996;6S1:25–6.
15. Rickwood AMK, Hodgson J, Lonton AP, et al. Medical and surgical complications in adolescent and young adult patients with spina bifida. Health Trends 1984;16:91–5.
16. Woodhouse CRJ. Myelomeningocele in young adults. BJU Int 2005;95: 223–30.
17. Persun ML, Ginsberg PC, Harmon JD, et al. Role of urological evaluation in adult spina bifida patient. Urol Int 1999;62:205–8.
18. Mandell J, Lebwitz R, Hallet M, et al. Urethral narrowing in the region of external sphincter: Radiologic-urodynamic correlations in boys with myelodysplasia. Am J Radiol 1980;134:731–5.
19. Tarcan T, Bauer SB, Olmedo E, et al. Long-term followup of newborns with myelodysplasia and normal urodynamic findings: Is followup necessary. J Urol 2001;165:564–7.
20. Yamamoto M, Kashiwai H, Tanaka Y, et al. Long term follow up of patients with spina bifida—a review of 228 cases. Jpn J Urol 1997;88:820–5.
21. Muralikrishna GS, Rodger RS, MacDougall AI. Renal replacement treatment in patients with spina bifida or spinal cord injury. Br Med J 1989; 299: 1506–7.
22. Patrick GM, Mahony JF, Disney AP. The prognosis for end stage renal failure in spinal cord injury and spina bifida—Australia and New Zealand 1970–91. Aust NZ Med J 1994;24:36–40.
23. Abrahamsson K, Arnell Vu-Minh M, Jodal U, et al. Evaluation of renal function in children with myelodysplasia. Br J Urol 2004;93:26–7.
24. Dik P, Klijn AJ, von Gool JD, et al. Early start to therapy preserves kidney function in spina bidida patients. Eur Urol 2006;49:908–13.
25. Barry BP, Hall N, Cornford E, et al. Improved ultrasound detection of renal scarring in children following urinary tract infection. Clin Radiol 1998; 53:747–51.
26. Levart TJ, Kenig A, Fettich JJ, et al. Sensitivity of ultrasonography in detecting renal parenchymal defects in children. Pediatr Nephrol 2003;17: 1059–62.