The urological system and continence control issues in spina bifida

Chapter 4: Urological management of spina bifida (including management of urinary tract infections)

Active and ongoing surveillance for urological problems helps to minimise the impact of the major source of mortality and morbidity in spina bifida.

Key issues for clinicians

- Urological complications of spina bifida are a major source of morbidity and mortality.
- An understanding of the neurogenic bladder is central to the management of urological complications of spina bifida.
- Regular annual urological review helps prevent long term complications, especially renal failure.
- Recurrent urinary tract infections are a major source of long term morbidity and complications. Urinary tract infections demand close investigation and often require specialist follow up. Patients need to be educated for the early detection of urinary tract infections.

Understanding urological complications — the neurogenic bladder[1–2]

Effective control of urinary incontinence in spina bifida needs to acknowledge the special management issues related to the presence of abnormal neurological bladder function, specifically those related to the neurogenic bladder.

Effective bladder training depends upon the ability to sense the presence of urine in the bladder and the passage of urine through the urethra. Altered bladder sensation can cause decreased, altered or absent sensation, thereby interfering with effective continence control.

Urinary tract sensation may be decreased, and easily not noticed when the person is otherwise occupied such as while working, watching television or at other tasks. Abnormal anatomical distribution of the nerves may cause sensations to arise from inappropriate places. Absent sensation makes responding to a full bladder impossible, requiring other strategies to achieve continence control.

Faeces or flatus in the rectum can also alter bladder feeling, adding further confusion to sensation, which is important for continence control.

Neurogenic bladder and sphincter abnormal function patterns — the role of urodynamic studies

Not only is an understanding of bladder structure critical to successful continence control, but equally important is bladder and sphincter function. This is one of the main functions of urodynamic studies of the bladder. Fluoroscopic urodynamic studies help the urologist to:

- image the structure and function of the bladder and sphincter
- provide a prognosis for upper tract deterioration
- maintain surveillance for those at high risk of complications

- plan surgical intervention at the optimal time
- provide information for continence control.

Neurogenic bladder functional abnormalities

Neurogenic bladder functional abnormalities can be classified into three main types:
- hyper-reflexic bladders when the detrusor muscle is unstable or overactive
- areflexic when the bladder is lacking any muscle tone
- mixed picture where there are elements of the hyper-reflexic and areflexic patterns in the one bladder.

There are cases of normal bladder function in spina bifida, but this should only be determined after urodynamic studies in the symptomatic patient.

Sphincter functional abnormalities

Sphincter function can be classified as:
- nonfunctional when the sphincter does not work at all
- synergic when the sphincter control is coordinated with bladder emptying
- dysynergic when the bladder emptying is not coordinated with the sphincter relaxation. When the full bladder starts to contract, the sphincter remains closed.

Bladder function in spina bifida

The commonest neurogenic pattern in spina bifida is the areflexic bladder with a nonfunctional sphincter. This can lead to complications of the upper renal tract and can be a major source of morbidity and mortality. This often results in a high bladder pressure due to urinary retention. High bladder pressure can result in long term urinary sphincter damage.

The commonest urodynamic pattern in spina bifida is the areflexic bladder with a nonfunctional sphincter.

Neurogenic bladder complications

Most children born with spina bifida have normal renal function. However, the presence of the neurogenic bladder can lead to urinary retention with overflow, uretic reflux and subsequent deterioration of renal function.

Hydroureters and hydronephrosis

Ureteric reflux can result in distension of the ureters and the kidneys, affect renal function and predispose to urinary tract infections (UTIs).

Renal calculi

Renal calculi can complicate many spina bifida related renal tract abnormalities.

Renal failure

Renal failure is an endstage result of these complications that surveillance aims to prevent.

Aims of urological management

Urological complications are a major source of mortality and morbidity in spina bifida and their prevention and management is a large component of patient care. The main aims of urological management are to:
- preserve upper tract function
- restore low pressure storage
- ensure adequate emptying
- control continence
- minimise UTIs.

Managing UTIs and related urinary tract disorders — a critical issue

Urinary tract infections are common in the presence of abnormal urinary tract structure and function. Recurrent UTIs can seriously compromise renal function and cause permanent renal damage. Furthermore, they can be difficult to detect in the presence of abnormal sensation.

The need for referral

Recurrent UTIs are common in spina bifida and are a strong indication for referral. As hydronephrosis and hydroureter are common, those with recurrent urinary tract infections, or a person with spina bifida not receiving ongoing urological surveillance who develops a urinary tract infection, should be referred to a urologist.

Educating patients to increase their awareness of symptoms and signs of UTIs

While some patients with spina bifida will present with the classic symptoms of UTIs such as frequency, urgency and dysuria, all of these symptoms may be difficult to detect due to reduced or changed sensation resulting from decreased innervation secondary to the spina bifida lesion.

Altered sensation causes atypical presentation

Atypical presentations of UTIs may include any (or none) of the following symptoms:
- smelly, offensive urine odour; often like ‘old fish’
- cloudy or bloody urine
- dysuria, although pain sensation may be altered by innervation
- abdominal or loin pain
- fevers
- nausea

- anorexia
- vomiting
- headache
- confusion
- malaise.

Clinicians should also note that UTIs often develop in the presence of constipation.

Educating patients to detect UTIs early can improve quality of life

Late or delayed presentations of UTIs can result in severe complications — including renal failure — and patients need to be educated to be aware of the signs and symptoms of UTIs.

Management of urinary tract infections

Clinicians should have a low threshold of commencing antibiotics when treating demonstrated or suspected urinary tract infections in spina bifida.

> Routine antibiotic sensitivity tests ensure appropriate treatment.

Some patients may have had considerable or ongoing exposure to multiple antibiotics over time and, combined with the risks of repeated instrumentation such as during catheterisation, the risk of resistant organisms is high. For this reason, midstream urine (MSU) tests should always include sensitivity testing as well as culture to ensure appropriate antibiotic testing.

In view of reduced sensation, repeat MSUs are necessary to confirm that the infection has resolved.

Review of catheterisation techniques

Many patients presenting with urinary tract infection will be self catheterising to empty their bladders (see *Chapter 5 Controlling urinary incontinence*). Many of these patients will have reusable catheters and should be advised to use single use only catheters until the infection has resolved. Catheterisation techniques should always be reviewed after a urinary tract infection to ensure that the process is clean (see clean intermittent catheterisation in *Chapter 5)*. Assistance from specialist clinics may be appropriate at this point.

Urinary tract infections — should I refer this patient on?

The appearance of recurrent UTIs in someone with spina bifida needs to be taken very seriously, as treatment of any underlying abnormality may prevent the development of renal failure.

An MSU culture that demonstrates a low number of mixed organisms is likely to be due to contamination. In the absence of symptoms and presence of otherwise normal symptoms, signs and investigations, this is unlikely to represent a UTI. These patients may not necessarily require referral, but if any doubts exist, clinicians should feel free to refer patients for further assessment.

Case study: Learning to read the signs of UTIs

Michael is a 25 year old man with spina bifida who presents with recurrent septic shock secondary to UTIs requiring multiple hospital admissions. Due to his spina bifida lesion, he has reduced pelvic sensation relating to typical presentations of UTIs. For him, the early signs of UTI included cloudy, smelly urine, fever and tiredness. Management included referral to a specialist facility for urodynamic review, increasing oral intake of fluids and reviewing catheterisation techniques.

In addition, Michael was taught to present to his GP for dipstick and MSU testing if any of his characteristic symptoms of a UTI were present. Treatment is now started at an earlier stage and the number of his UTI related hospital admissions have decreased from 10 per year to three per year.

Routine urological assessment of spina bifida

As the complications of spina bifida are an ongoing process, regular monitoring of the urinary tract is necessary, especially in high risk patients, in order to prevent and treat potential urological complications. Urodynamic and renal tests are especially critical when determining the optimal time for surgery and prevention of complications.

Lifelong surveillance — the role of the GP

General practitioners may see patients with spina bifida who have not had regular urological monitoring or assessment. When reviewing patients with spina bifida, GPs need to ensure that urological monitoring appropriate for the age group has taken place. Clinicians can order basic monitoring tests for patients who are currently not receiving urological surveillance, followed by referral to a urological or spina bifida centre.

Urological monitoring — the basics

As part of routine medical care, GPs and other clinicians need to:

- assess current and past urological history
- review past renal investigations

- ensure that annual creatinine, urine microscopy and culture and renal ultrasound are ordered.

Key urological issues for each age

Assessment of newborn

After the closure of the spina bifida defect, the initial evaluation of the newborn urinary tract involves renal ultrasound, residual urine measure, voiding cystourethrogram and urodynamic studies. This is performed within the context of a specialised paediatric unit.

Children below five years of age

Children below the age of five years are at the highest incidence of renal damage. In addition to renal function monitoring, these children often require annual or biannual urodynamic studies.

School age

Social issues relating to incontinence are critical in school age children as they can interfere with educational opportunities, self esteem and social development. These children may require urodynamic investigations to assist with continence control.

Young people

The teenage and young adult years are often times of poor compliance, especially with continence regimens. There may be many psychological, educational and social issues (see *Chapter 2 Impact of hydrocephalus and other CNS conditions on care management*). Additional urodynamic studies may be required to sort out continence issues.

Adulthood

Urological surveillance does not stop just because a person with spina bifida reaches adulthood. Patients require baseline renal ultrasound, renal function tests, annual nuclear scans for measuring glomerular filtration rate and other investigations to assist diagnostic interpretation of complications in the event of change in symptoms.

Bladder and urinary tract management

Common approaches to the management of bladder and other urinary tract problems in spina bifida involve a combination of the following:

- conservative management including pharmacological agents
- surgical intervention.

The procedures are described within the context of urinary continence management in *Chapter 5 Controlling urinary incontinence.*

Urological surgery in spina bifida

Urological surgery for the management of incontinence is an important option for people with spina bifida where other procedures, such as intermittent catheterisation, are not feasible. In addition, there are many urological reasons for surgical interventions, all of which impact upon the control of incontinence.

The decision to proceed with surgical intervention for the control of urinary incontinence in spina bifida is a highly complex area. Indications for surgical intervention of the urinary tract in spina bifida include persistent high urinary storage pressure, upper urinary tract deterioration, vesicoureteric reflux and incontinence.

For an overview of the indications and techniques for each of these procedures, see *Chapter 5 Controlling urinary incontinence.*

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The urological system and continence control issues in spina bifida

Chapter 5: Controlling urinary incontinence

Achieving control of urinary continence is the key to achieving an independent lifestyle. A wide range of interventions and resources exist to assist in the successful management of urinary incontinence. This chapter provides the clinician with an introduction to management principles, some resources and also outlines the roles of specialist clinics in the management of incontinence (for issues of faecal incontinence, see Chapter 6).

Key issues for clinicians

- Incontinence impacts on all aspects of life. Successful management of incontinence overcomes a major barrier to personal and social independence.
- Incontinence is best managed in conjunction with a specialist continence clinic.
- Most young people and adults with spina bifida will have already established incontinence management. The role of the general practitioner is largely one of review and detection of management problems. These can then be referred to a specialist clinic if indicated.
- Conservative management is the first step to incontinence management.
- Clean intermittent catheterisation is a common and important component of incontinence management. Clinicians need to familiarise themselves with this procedure.
- Incontinence management procedures need to be reviewed after a urinary tract infection.
- Persistent changes in continence patterns should be referred to a specialist clinic.
- There are surgical interventions available to assist incontinence management if conservative measures fail. These are organised through specialist clinics.
- Clinicians need to be aware of the existence of possible latex allergies when treating patients with spina bifida.

Incontinence impacts on all aspects of daily living

Incontinence can prevent people with spina bifida from achieving full participation in all aspects of life, such as work, education, personal relationships and general activities of daily living. In addition, incontinence is almost a taboo subject, viewed by many in society as a weakness and a source of shame.

Impact on self esteem

Incontinence also brings many other daily problems, such as changing beds, and clothes, washing soiled linen and clothing, constant worry over possible episodes of incontinence, embarrassment, shame at soiling in public, accusing looks from teachers, work colleagues and the general public — all leading to difficulty in coping with daily life.

These issues can lead to poor self esteem, contributing to a sense of frustration, guilt, fear and isolation, making coping even more difficult.

Continence control — the incontinence management team

There are many resources and health care providers available for achieving successful continence control and GPs can help link people with spina bifida to these specialist clinics. Not only are there specialist teams to help children, but clinics for young people and adults with spina bifida also exist (see *Chapter 9 Organisations and further resources*).

Clinicians having problems identifying nearby adult treatment centres may be able to obtain their location by contacting a paediatric treatment centre.

Overview of bladder and urinary tract management[1–2]

Common approaches to the management of bladder and other urinary tract problems in spina bifida involve a combination of the following:

- conservative management including pharmacological agents
- surgical intervention.

All continence control needs to be managed under the direction of a urologist and continence clinic. Many adults with spina bifida are unaware of these resources, and GPs can greatly improve quality of life by referring patients to these specialist centres. The GP remains a key player in this team as the first point of contact for patients.

The following procedures are described to familiarise clinicians with common approaches to bladder management. Treatment should only be initiated under the direction of a urologist, or a spina bifida or continence clinic.

Conservative management

Conservative management of bladder problems usually involves a combination of clean intermittent catheterisation (CIC) and the use of pharmacological agents.

Establishing a routine

The key to successful incontinence control is to establish incontinence management procedures as part of every day living. When incontinence control becomes a problem, the aim is then to re-establish these routines.

Clinicians need to be aware of differences in incontinence control for those affected by spina bifida. For example, the experience of many GPs will be in children with normal bladders. Incontinence control issues in spina bifida are wider than this; incontinence occurs within the context of a neurogenic bladder, and is an ongoing issue for all ages for people with spina bifida.

Timing is the key

The key to successful control of urinary incontinence in spina bifida is bladder timing. Only a small number of people with spina bifida will be successfully bladder trained, but successful timing can be achieved in a majority of cases.

Successful bladder timing — that is, the regular emptying of the bladder — allows the person to have control and confidently participate in school, work and other areas of life.

Establishing routines and regular practice is the first step to achieving effective incontinence control. When routines become upset, this pattern can be used as a target to get habits back into line.

Intermittent catheterisation

Intermittent catheterisation of the bladder allows it to empty, in order to prevent retention, reflux and other complications, and to help control incontinence.

Practice points

In the presence of nerve damage, the person with spina bifida may have difficulty telling the difference between a full bladder and a full bowel.

Sensation from the muscle wall of overstretched bladders are weak or nonexistent.

Detruser sphincter dyssynergia can either cause a rush of urine flow when the sphincter does open, and usually occurs at inconvenient times, or may just cause a dribble when the urinary bladder pressure rises above a certain level.

Swimming and drinking will increase the urine output.

Anxiety, shocks and excitement can precipitate episodes of incontinence.

Sensations such as abdominal 'pain' can be confused with bladder or bowel fullness sensations.

Incontinence control routines can be upset by intermittent infections, procedures, illnesses and other precipitating factors.

This simple, clean (not sterile) procedure repeated a few times a day allows control of the timing of bladder emptying.

Clean intermittent catheterisation aims to achieve continence by emptying the bladder at scheduled intervals, as well as reducing residual urine volume in order to prevent infection and bladder overstretching.

Intermittent catheterisation gives the person with spina bifida a great deal of control over incontinence, and while achieving good technique may take some practice, the effort is well worthwhile and achievable. Self catheterisation requires good hand to eye coordination.

Self catheterisation gives a young person or adult increased self esteem through increased independence. Self catheterisation also means that the person can attend school or work unassisted.

Young people may have issues with compliance with self catheterisation and the clinician may need to check that techniques are being followed. This may involve referral to a specialist incontinence management team.

Reviewing catheterisation techniques

Many young people and adults with spina bifida will have already been using intermittent catheterisation for many years. In this case, the role of GPs seeing adult patients for the first time may not be to teach intermittent

catheterisation, but to review technique, check that the appropriate catheter type is being used and refer to specialist centres as needed.

Catheterisation techniques should be especially reviewed when there is a change in continence pattern or after a urinary tract infection. Reusable catheters should be replaced with single use catheters in the presence of a urinary tract infection.

Intermittent catheterisation

Catheterisation aims to empty the bladder to protect renal function and to achieve social independence through prevention of incontinence. Catheterisation is a simple, clean method of inserting a plastic catheter several times a day to drain urine *(Tables 5, 6).*

Catheterisation and the toilet

Catheterisation is usually performed in the toilet, as using this socially acceptable place helps to normalise the process of urination. That is, the toilet is the same place used for urination as that for continent people. Toilets are always available, even if they need to be cleaned afterwards. When the catheter is correctly inserted, the person can hear the urine fall into the water and knows that the catheter has been inserted sufficiently and into the correct orifice (for women). In addition, sitting upright gives better drainage and maximises the chances of using the correct method to withdraw the catheter — that is, downwards.

Catheterisation should be performed before emptying the bowels.

The self catheterisation routine — the role of specialist clinics

Self catheterisation is a complex technique and is best taught by specialist continence clinics. These clinics can adapt teaching to suit each individual according to their special needs and gender. However, clinicians can keep copies of any instructions issued to the patient to assist in reinforcing key messages directed by the specialist clinics.

While establishing catheterisation techniques is often done with the assistance of a specialist continence team, the GP can assist by going through the patient's technique to ensure that each step is performed correctly. A checklist has been prepared to help clinicians ensure the basic technique is adequate *(Table 7),* but more detailed information and assistance is available from the specialist continence team.

General practitioners with any questions can contact the continence nurses or other health professionals of continence clinics for further assistance.

Latex allergies

Be aware that allergies to latex are more common in people with spina bifida than for the general population. Reactions can vary between mild reactions to severe anaphylactic shock. Clinicians need to remain alert to this possibility and to refer to specialist clinics for advice if the situation arises.

Case study: Betty is a 26 year old woman with incontinence.

She has overflow incontinence, and although using a CIC routine — which she has been using for many years — needs continence pads. Wheelchair bound, she has oscillated between living at home and independently, the major issue being a constant smell of urine, although she is desensitised to the smell. She lacks confidence and seems to have given up any ambition of work. Apparently very disorganised, when you talk to her about the urine smell issue she becomes distraught and angry. You encourage her to attend an adult spina bifida clinic and a continence nurse.

The nurse reports that she is using inappropriate pads, wrong sized catheter, and is not catheterising frequently enough. With some planning assistance and assigning a friend who will tell her if she smells, her continence control is much improved. Establishing control involves keeping the catheterisation routine constant. Each time it is performed, it should be identical. Not only does this maximise effectiveness of the procedure, but decreases risk of urinary tract infection.

Pharmacological agents

Under the direction of a specialist, pharmacological agents can be an important adjunct to intermittent catheterisation. Common agents include anticholinergics such as propantheline bromide; musculotropics including oxybutinin, and antimuscarinics such as tolterodine.

Adverse effects

These agents can cause adverse central nervous system effects that can interfere with cognition, which may in turn complicate any deficits already present (see *Chapter 2 The impact of hydrocephalus and other CNS conditions on case management.*)

These agents may also contribute to constipation, which can contribute to faecal incontinence.

Surgical intervention

Surgical intervention for the management of incontinence is an important option for people with spina bifida where other procedures, such as CIC are not feasible.

Table 5: Clean intermittent catheterisation — instructions for males[3]

Equipment required:

Catheter, cleansing solution, lubricating gel, cottonwool balls or wipes.

Procedure:

1. Wash hands with soap and water.
2. Lubricate the catheter.
3. Retract foreskin if not circumcised and wash the tip of the penis using a cleansing solution.
4. Hold penis upright and gently insert the catheter into the urethra. If resistance is met part way, rotate the catheter or use gentle but firm pressure on the catheter until the muscle relaxes. It may also help to take some deep, slow breaths.
5. When the urine flow has stopped, advance the catheter one more inch to ensure the bladder is fully empty.
6. Slowly remove the catheter liberally.
7. Males with foreskins should always push the foreskin back again after the procedure.
8. Put on clean pad.
9. Wash hands with soap and water after washing and packing away equipment and cleaning toilet seat.

Table 6: Clean intermittent catheterisation — instructions for females[2,4]

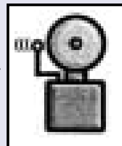
Equipment required:

Catheter, clean pad and clothing, lubricating gel, washer, cottonwool swabs or wipes.

Procedure:

1. Wash hands with soap and water.
2. Lubricate catheter liberally.
3. Sit well back on the toilet.
4. Clean the vulva with 3 swabs from front to back.
5. Wipe hands.
6. With one hand, hold the labia apart and see or feel for the clitoris.
7. With the other hand, place the tip of the catheter behind the clitoris. Insert gently until it enters the urethra. Gently push in until the urine flow begins.
8. When the urine has stopped flowing, slowly pull out the catheter.
9. Wash hands and put on clean pad.
10. Wash hands with soap and water after washing and packing away equipment.

Ensure that patients using reusable clean catheters for intermittent catheterisation move to single use disposable catheters during a urinary tract infection. After the infection has resolved, patients can then start using a new, reusable catheter.



In addition, there are many urological reasons for surgical interventions, all of which impact upon the control of continence.

The decision to proceed with surgical intervention for the control of urinary incontinence in spina bifida is a highly complex area. Indications for surgical intervention of the urinary tract in spina bifida include:

- persistent high urinary storage pressure
- upper urinary tract deterioration
- gross vesicoureteric reflux
- incontinence due to intrinsic sphincter deficiency (ISD).

High bladder pressure

As discussed previously, the most common neurogenic pattern in spina bifida is an areflexic bladder with a nonfunctional sphincter. This can lead to complications of the upper renal tract and can be a major source of morbidity and mortality. This often results in high bladder pressure due to urinary retention. High bladder pressure can result in long term urinary sphincter damage.

Surgical management of high bladder pressure may involve a variety of techniques

There is a wide range of surgical options to manage incontinence in spina bifida that can be tailored to each individual. These procedures are constantly improving and increasing in technical sophistication, emphasising the importance of patients receiving regular urological surveillance.

Overview of common surgical procedures

The following is a list of common urological procedures used in spina bifida, and their more common indications.

Vesicostomy

Vesicostomy is indicated in the presence of persistent hydronephrosis and recurrent urinary tract infection when the bladder continually fails to empty. This simple

Table 7: Checklist for reviewing self catheterisation technique

- Have copies of any patient instructions for procedures included in their medical history file to help check some of the following key issues.
- Patients may benefit from visual instruction using illustrations rather than verbal instruction if learning difficulties are present
- The routine must be kept the same each time.
- Ensure that hands are washed at each point indicated in the procedure.
- Ensure that the catheter is lubricated liberally.
- Instruct patients to be careful to prevent contamination from clothes. This may be done by folding the clothes upwards and using a peg to keep clothes fastened and away from genital area.
- The bladder must be fully drained as incomplete emptying is a common cause of urinary tract infections. To do this:
  - the full length of the catheter must be held below the level of the bladder throughout the entire procedure
  - gentle pressure is applied to the lower abdomen after the flow of urine has been stopped.
- The flow of urine is sometimes stopped if the sphincter closes on the catheter giving the impression of complete bladder emptying. This may be indicated by resistance when removing the catheter and by lower urine output than expected during drainage. In this case, repeat the procedure in 1/2–1 hour.
- Assess bowel habits: constipation may cause partial urethral obstruction.
- Associate bladder emptying to the daily routine, such as when getting up in the morning, after meal times and before going to bed.
- Also review techniques and any instructions given for cleaning catheters.

procedure which involves making a stoma from the bladder to the skin surface to allow drainage, has a low revision rate and allows normal growth and maturation. Vesicostomies are often performed as temporary procedures in children.

Urinary diversion

Urinary diversion can be used when augmentation procedures fail to work for many physical, personal and social reasons. Procedures include illeal and colon conduits and cutaneous ureterostomy.

Augmentation cystoplasty

Augmentation cystoplasty involves surgically configuring a segment of bowel to augment the bladder and correct vesicoureteric reflux. When deciding upon an augmentation cystoplasty, issues to consider include which part of the bowel to use, eg. illeum, stomach, sigmoid colon or other section. Complications can result from the mucosa of the segment of origin, such as haematuria when using gastric lining or mucus production when using sigmoid colon. Ureteric augmentation uses distended hydronephrotic ureters, if present, to augment the bladder. Other complications of augmentation can include perforation, infection, mucus production, calculi and the potential for malignancy, although this risk is small.

Catheterisable stomas

Catheterisable stomas may be useful in patients unable to perform intermittent catheterisation due to lack of dexterity or being wheelchair-bound. They also have a place when a urethra is unavailable, perhaps due to the presence of a stricture or a fistula.

The Mitrofanoff procedure is the formation of an abdominal stoma which is then connected to the bladder with a tubal structure such as the appendix. Urine is then drained by passing intermittent urinary catheters. For example, in the Mitrofanoff appendix procedure, the stoma is created from the appendix and part of the caecum with intact blood supply. The tip of the appendix is then buried through the bladder wall to create a passageway for urine. Other structures have also been used, including: gastric tissue; fallopian tubes; ureters; and other parts of the bowel.[6]

Transurethral injection

Transurethral injection therapy is used to treat intrinsic sphincter deficiency and involves the submucosal injection of a biocompatible substance such as collagen or

silicon. The efficacy of treatment depends largely upon selecting patients with suitable urodynamic patterns.[3] The advantage of submucosal injection is the low morbidity, but its main disadvantage is the lack of long term data on most of the substances.

Slings

Pubovaginal slings are the treatment of choice for females with intrinsic sphincter deficiency although there is also a role for the procedure in some males Suburethral slings use a variety of techniques and materials and many series have included long term follow up. Native tissue, such as the use of an autologous tendon, appears to be associated with less morbidity than using synthetic materials. Patients must be monitored postoperatively to ensure bladder emptying takes place and that there is no upper tract deterioration.

Artificial urinary sphincters

Artificial urinary sphincters are implanted silicon devices that close the urethra. The artificial sphincter may be placed at the bladder neck or bulbar urethra. The artificial sphincter is regarded as the main treatment option for male patients with intrinsic sphincter deficiency. Again, post-operative monitoring is essential to ensure that urinary tract complications due to the elevated bladder pressure associated with an artificial sphincter are prevented.

Circumcision

Circumcision may be indicated in males, especially when in the presence of recurrent urinary tract infections where circumcision can sometimes reduce their frequency.

Reversal of surgical procedures

Young people and adults with spina bifida may present having had a particular surgical technique for incontinence at some stage in the past but without a recent urological review. Many options are not permanent, and can be changed to suit the needs of the person at that time in their life.

In light of surgical advances there may now be further options for these patients to explore. Some of these patients may want to try alternate continence procedures and may want to have their surgery reversed. Referral to a specialist centre enables patients to explore the advantages and disadvantages of each of these procedures.

Reversal of urinary diversion (also called undiversion) may be an option in motivated patients when physical considerations allow. Patients may have had urinary diversion procedures in the past when these procedures were a more common first line treatment and may now wish to take advantage of more recently introduced augmentation procedures. Reversal of diversion allows the introduction of a clean intermittent catheterisation regimen that may be more beneficial for renal function and promotes independence. This process can offer significant benefits to a select group of patients, but motivation needs to be high as it involves considerable preoperative preparation and a high degree of postoperative compliance to ensure effective clean intermittent catheterisation

Case history: John is 22 years old

He is a highly motivated man who had a urinary diversion procedure when he was a toddler. He has managed with a bag for years, but is beginning to realise that he has missed out on many activities such as swimming and travelling and confides that he 'can't imagine a sexual relationship with the bag present'. He has also heard at a spina bifida meeting that his kidneys may be affected. He is amazed when you advise him that this procedure may be reversable, with him starting a clean intermittent catheterisation routine. Enthusiastic to find out more, he is eager to visit the adult spina bifida clinic.

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The urological system and continence control issues in spina bifida

Chapter 6: Controlling faecal incontinence (including constipation and bowel dysfunction)

Spina bifida may affect faecal continence in many ways. An understanding of the issues can assist general practitioners in supporting their patients through the process of learning to effectively manage faecal incontinence in conjunction with specialist centres.

Key issues for clinicians

- Faecal incontinence is a major source of poor quality of life for young people and adults with spina bifida.
- An understanding of the special faecal incontinence issues for people with spina bifida is necessary for successful incontinence management.
- Faecal incontinence control is best managed in conjunction with a specialist spina bifida clinic.
- Dietary management can help some people successfully manage diarrhoea, constipation and incontinence.
- Constipation should never be left untreated for longer than two days.
- Medication is useful for softening and loosening stools.
- Clinicians need to familiarise themselves with incontinence appliances such as anal plugs.
- Surgical procedures are an important method of incontinence control.

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Faecal incontinence — a major barrier to independence

Achieving and maintaining bowel continence is one of the most difficult challenges for people with spina bifida. Successful control of faecal incontinence is a key barrier that needs to be overcome in order to achieve full independence and free participation in activities of daily living. In addition, faecal incontinence has a major impact on issues of self esteem.

Reviewing bowel training and timing

Young people and adults with spina bifida visiting their GP will have already established their bowel habits, although some may not be benefiting from more recent developments in surgery and appliances. The GP's role will then be to review bowel training and timing techniques, and to ensure that the patient has full access to the latest management developments.

Special issues — physical and behavioural

Bowel training is usually started in childhood, but may become more difficult in the adolescent years. Growth may affect the nerves to the anus and rectum, causing a change in bowel habits and an increased risk of incontinence.

In addition, as adolescence marks a time of increasing independence, sense of invulnerability, experimentation and rebellion, many young people may pay less attention to health maintenance issues.

The role of GPs and specialist clinics

General practitioners managing adults and young people with spina bifida should assess faecal continence as part of any routine review. An awareness of factors that can cause intermittent problems may help the GP manage simple continence problems, but continuing incontinence should be referred to specialist clinics.

In addition, many adult patients may have lost ongoing contact with specialist treatment clinics and may not be aware of advances in the treatment of faecal

incontinence. By referring to specialist clinics, GPs can ensure that patients can benefit from latest management techniques.

Faecal incontinence control[1–2]

Spina bifida related nerve damage has a major impact on the ability to maintain faecal continence. While clinicians will be familiar with the general principles of incontinence management, the presence of spina bifida adds another set of issues that need to be addressed. The following is a list of areas that clinicians need to consider when assessing faecal incontinence.

Level of lesion

The presence of thoracic level spina bifida involvement may increase the difficulty in bearing down during defaecation, while patients with sacral lesions may have more difficulty obtaining appropriate and dependable stool consistency.

Stool consistency and frequency

Assessing the consistency and frequency of stools allows the clinician to formulate an appropriate management plan.

Constipation can occur very quickly in spina bifida, which can exacerbate incontinence. Poor diet and lack of exercise are common contributory factors. While young children may not eat foods that promote stool formation, dietary intervention can help some young people and adults prevent constipation. Medications to assist bladder relaxation may cause constipation. Constipation can also increase the risk of urinary tract infections.

Long term constipation requires long term treatment. When the bowel is overstretched from chronic constipation, faeces will reaccumulate quickly after treatment. Untreated constipation can lead to a toxic megacolon. After continuous treatment, the bowel may return to its normal size and if diet is adequate, the stool will return to its normal consistency. The length of treatment time depends upon how long the constipation has been present.

> Constipation should never be left untreated for longer than two days.

Diarrhoea can make the practical management of incontinence difficult, as well as increasing the risk of soiling.

Diet and meal times

Fluid and fibre intake influence stool consistency and frequency. In addition, eating can stimulate bowel function and timing bowel emptying after meals can facilitate incontinence control.

Anal and rectal canal

Moderate to severe neurological involvement in spina bifida can prevent the anus from fully closing. Nerve damage may cause a very lax anus resulting in a loss of ability to retain stools, especially during periods of heavy physical activity, such as swimming.

The presence of redundant tissue in the rectal canal may make correct insertion of a suppository more difficult.

Altered sensation from nerve damage

There may also be a reduced or absent rectal sensation, causing difficulty in recognising when the rectum is full or when bearing down occurs. This difficulty in detecting sensation changes may be greater when the person is busy or distracted, increasing the risk of soiling. Patients need to be encouraged to go to the toilet as soon as they feel the need, before any distraction can occur.

Difficulty detecting soiling

Some people will also have difficulties with genital skin sensation that may affect their ability to feel when their skin is wet after soiling. In addition, altered smell sensation may make it difficult to detect when soiling has occurred.

Slower bowel development

The bowels of children with spina bifida can be slow to develop. Some children may be as old as nine years before the bowel is mature. This may mean many years of faecal incontinence.

Mobility, activity and temperature changes

Increased physical activity and changes in temperature can cause the bowels to relax and empty, such as when having a bath or when swimming.

Medications

Anaesthesia and medications can cause constipation. Anticholinergic medications used for bladder incontinence control can also cause constipation. Antibiotics, often for urinary tract infections, may cause diarrhoea and increase the risk of faecal incontinence. Where indicated, the prophylactic use of probiotic yoghurt and increasing fibre intake may help to minimise these effects.

Intercurrent exacerbating factors

Lifestyle factors and life events may also cause changes in bowel habits. Common exacerbating factors include:

Table 8. Foods frequently associated with causing loose stools and faecal incontinence[3]

- Citrus fruit, fruit juice, passionfruit, pineapple
- Corn (fresh or tinned)
- Baked beans
- Chocolate/malt/chocolate powders used to flavour milks
- Nuts/dried fruit.

Table 9. Dietary control of stool consistency — foods that soften stools[3]

Food Group	Reduce	Increase
Bread/cereals	Highly refined (white) breads, biscuits	High fibre breads and natural whole grain cereal, eg. bran, oatmeal, rice, muesli
Vegetables/fruit	Tinned fruits, juice with high sugar content	Fresh, raw vegetables, raw fruit, sugar free juices
Meat/fish, poultry	Lean cuts such as veal, chicken, whiting	Fatty cuts (mince, sausage, mullet, tuna, mackerel)
Cheese/eggs		Matured or processed cheeses, yoghurt
Beverages	Soft drinks, cordials, skim milk.	Whole milk
Miscellaneous	Plain sugar, syrups, jellies, sweets	Herbs and spices, nuts, pizza, muesli bars, chocolate

- holidays
- disruption in usual daily routines
- a change in water, such as when travelling
- intercurrent illness, especially febrile illness
- anxiety, especially at school or at work
- hospital procedures or operations
- changes to family structure such as a new birth, separation, death of a relative, or
- starting a new school or job.

Effective continence control is best achieved within the context of a specialist clinic.

Access to facilities and aids

Difficulty in access to bathrooms, toilets at home, in educational institutions and the workplace can increase incontinence control problems. In addition, there may be difficulty in transferring to toilets if the person is in a wheelchair.

Overview of conservative management of faecal incontinence

Finding the balance — the role of the specialist clinic

As every patient is different, effective bowel control regimens need to be tailored to the needs of each individual.

Effective continence control is best achieved within the context of a specialist clinic. However, clinicians need to be aware of the general management principles and interventions used for faecal incontinence control.

Diet, fluids, diarrhoea and constipation[2,3]

While a healthy diet for people of all ages is a general health principle, diet can be used effectively by some people to alter stool consistency and frequency in order to facilitate bowel control.

Dietary control of stool consistency is not achievable by everyone, but some patients can benefit significantly if given appropriate information. Carers involved in food preparation may also benefit from dietary information.

Certain foods can cause diarrhoea, which may exacerbate soiling (*Table 8*). Also, stools can be softened by increasing foods high in fat, fibre and moderate in sugar intake (*Table 9*). Water intake should also be increased. Conversely, these foods should be decreased when trying to harden stools (*Table 10*).

Drug control of stool consistency[2]

Drugs can be used to control stool consistency, but should only be used for a limited period of time, as long term use of some drugs may have a deleterious effect on bowel function and increase the risk of drug interactions. Bulk forming agents, however, are not associated with long term adverse effects.

In general, expert advice should be consulted prior to initiating drugs to control stool consistency, especially in children. Also, some drugs, especially adsorbents, may interfere with the absorption of other drugs. Commonly used drugs are listed in *Table 11*.

Bowel emptying — overview of methods and techniques

There are many techniques and methods for emptying bowels ranging from normal toileting to sophisticated surgical techniques.

Most young people and adults with spina bifida presenting to their GP will have had extensive toileting program experience. The aim of the following overview is to familiarise clinicians with the principles of commonly used techniques in controlling faecal incontinence.

The prescription of individual bowel programs is best done within the context of a specialist spina bifida clinic. General practitioners should not hesitate to refer patients to these clinics if they detect continuing problems with incontinence.

Behavioural training

Effective bowel control involves creating a system for bowel emptying at regular intervals, at least every 24 hours. Due to the lack of rectal sensation common in spina bifida, developing a daily routine ensures regular evacuation. For example, associating the timing of bowel emptying with meals, baths, physical activities, particular times of day, helps establish predictable continence patterns. This will depend upon the person's physical, cognitive and functional level. Behavioural training will be more successful if lower motor function is intact.

When establishing new bowel emptying patterns, daily reinforcement of any bowel procedures with the assistance of regular home nursing visits, where available, can greatly expedite the adoption of new interventions.

Anal/rectal stimulation

Weakened nerves can sometimes be stimulated by wiping the anus firmly with toilet paper as soon as the person sits. If the stool is not being expelled, slight pressure can be applied with fingers to each side of the anus to replace the natural lift of the anus, lost due to neurological damage. Similarly, there are other techniques that can be learned to improve bowel emptying.

Digital stimulation involves inserting a gloved finger into the anal canal and internal sphincter, and massaging the mucosal wall to stimulate a contraction to eliminate a stool. This is more effective in the presence of lower motor neurons.

Suppositories and microenemas

Microenemas and suppositories can be used to establish timed bowel actions and treat constipation. Microenemas and suppositories can also fully empty the bowel, allowing a longer period between evacuation.

Patients and carers need to learn correct techniques, which can be taught in the specialist treatment clinic.

Table 10. Dietary control of stool consistency — foods that harden stools[3]

Food group	Reduce	Increase
Bread/cereals	High fibre breads and natural whole grain cereals eg bran, oatmeal, rice, muesli	Highly refined (white) breads, biscuits
Vegetables/fruit	Fresh fruits and vegetables, fruit juice.	Cooked vegetables with low fibre such as potatoes, pumpkins, carrots. Tinned fruits in small amounts
Meat/fish, poultry	Fatty cuts (mince, sausage, mullet, tuna, mackerel)	Lean meats such as veal, chicken, whiting
Cheese/eggs	Whole milk cheeses, fried eggs.	Cottage cheese, boiled or poached eggs.
Beverages	Whole milk, cream	Skim or low fat milk
Miscellaneous	Herbs, spices, pizza; minimise oil, butter and margarine	Honey, jelly

Table 11. Commonly used drugs to control stool consistency[2,3]

Laxatives	Bulk forming agents	Stool softeners	Drugs that harden stools
Milk of magnesia,	Normacol Parachoc)	Parrafin (Paralax, Methylcellulose,	*General absorbents*
Agarol	Psyllium or ispaghula husk	Poloxalkol (Coloxyl drops)	Kaolin, pectin, cholestyramine
Lactulose (Duphalac, Actilax)	(Metamucil or Fibogel	Dioctyl Na sulphosuccinate (Coloxyl tablets)	*Agents altering motility*
			Codeine, lomotil (not recommended in children), loperamide

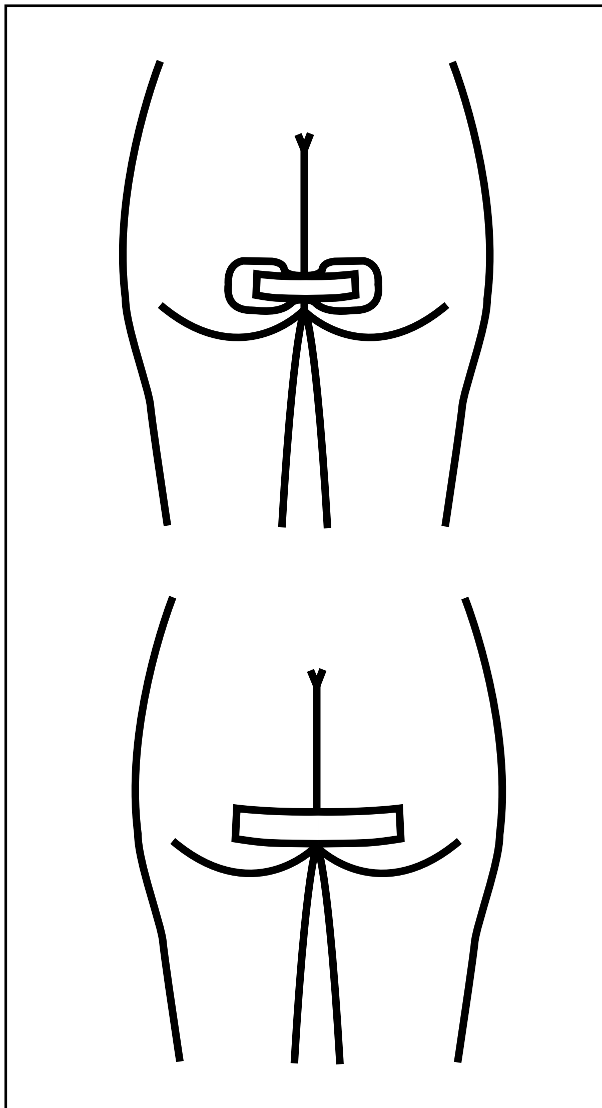

Figure 6. Buttock strapping

Strapping buttocks: procedure

- The tape is applied low on the buttocks to be under the person when they sit down.
- Cut the appropriate length of tape
- Look for the position of the anus
- Attach tape to one buttock. While holding buttocks together, attach the other end of the tape to the other buttock, ensuring that the tape passes over the anus.
- If the skin is sensitive, place some nonallergenic tape on each buttock. Stronger tape can then be applied on top of the nonallergenic tape.
- If the anus is very lax, a small piece of paper, such as half a piece of toilet paper, can be folded and placed over the anus. Females should check that the tape has not slipped down into the vagina.

Large volume fluid enemas[4]

If other methods have failed, large volume enemas also called colonic washouts, may be required to treat constipation. The amount and type of fluid is determined by the specialist clinic, but may include solutions of saline; water; soap and water; or other solutions. The enema is administered by using a 30 mL balloon catheter and a large syringe. These are available in specially designed colonic washout sets such as the Willis Washout System. Care must be taken not to use rubber catheters in those with latex allergies.

The volume of fluid required for the enema may increase the pressure on an already overstretched bowel, and there is an increased risk of soiling.

Enemas will clean the bowel for 2–3 days. While many patients find enemas a manageable way to control bowel emptying, large volume enemas may be difficult or virtually impossible for a person with limited mobility and can contribute to dependency.

Buttock strapping

This method can be used when prevention of soiling is important, but will not work when the stools are soft, or when there is diarrhoea. Strapping can be used when swimming, on special outings or as a regular approach to faecal incontinence *(Figure 6).*

Strapping should be removed from the buttocks when it is the regular time to empty the bowels or when the person feels the need to empty their bowels.

Try different types of tapes to ensure they are waterproof or that no adverse reactions occur. Typically used tapes include elastic adhesive tapes, nonallergenic tapes, waterproof adhesive tape and electrician's tape.

Anal plugs

Anal plugs are an important continence management tool and offer real independence for some people with spina bifida. The anal plug, made from foam, is lubricated with Vaseline and inserted into the anus. After coming into contact with the moisture of the bowel, it expands in about 30 seconds to form a mushroom like shape that prevents rectal leakage. The anal plug is made from slightly porous material so that air can pass through the plug. The plug is removed with an attached string, and is changed after each toilet visit. Removal of the plug does not stimulate the rectal muscles, and thus the plug may be removed slowly. The anal plug can be worn safely for up to 12 hours. Combined with diet and bowel regimens, anal plugs have significantly changed the lives of many people with spina bifida by increasing their independence.

Case study: Peter is 21 years and has faecal incontinence

Peter has ongoing faecal incontinence. He is a community walker with ankle–foot orthosis and the incontinence interferes significantly with his active lifestyle.
The problem has also caused a lack of confidence, causing him to quit two jobs and he remains housebound because of frequent bowel accidents.
Microenemas, routine training and diet management have not been fully successful. Anal plugs pop out.
He uses large volume washouts every two days or so and manual evacuation, both of which he needs assistance with. After a Malone procedure he is able to much more confidently self administer the enema, which seems to be working reliably. As a result, Peter is slowly becoming more outward looking.

Surgical procedures

When bowel emptying and faecal incontinence is not successfully controlled with the aforementioned methods, surgical procedures may be indicated. These procedures are only used after other more conservative methods have failed. Many patients express high degrees of satisfaction after this procedure is performed.[5–7]

These procedures are sometimes performed concomitantly with other urological surgical procedures.

Malone procedure and antegrade colonic enemas

The Malone procedure — also called continent appendicostomy — is used for the management of faecal incontinence and involves bringing the appendix to the surface of the skin and creating a stoma. Like similar procedures, the Malone procedure provides access to the proximal colon for the administration of enemas called antegrade colonic enemas (ACE). There is no unpleasant smell as the bowel contents are sterile at the level of the appendix. The stoma can be left in place longterm if necessary.

A major advantage of the Malone procedure is the ease of self administration, especially in people with poor mobility when compared to the more usual retrograde washouts.

One potential disadvantage of the Malone procedure is that the appendix is then unavailable to use when constructing catheterisable stomas, such as in the Mitrofanoff procedure (see *Chapter 5 Controlling urinary incontinence*, p.92).

Caecostomy catheters[8,9]

A caecostomy catheter is a nonlatex, flexible tube that is inserted into the patient's caecum through the skin in the right iliac fossa, providing a comfortable, convenient way to irrigate and empty the bowels with an enema solution. The enema is given through the tube and the faeces exits through the anus.

Caecostomy tubes can improve independence in those who experience faecal incontinence with troublesome soiling and in those patients that do not respond well to rectal enemas or other methods. For example, they may be unable to perform retrograde bowel washouts. Caecostomy tubes offer a chance for independence in patients who may have previously run out of treatment options.

The caecostomy tube is placed in a two part process. Firstly, a temporary tube is inserted into the caecum, which is followed about six weeks later by a long term tube, which is much less visible than the temporary tube.

There is a choice of washout fluids and many patients find that optimal function is achieved by varying the composition of these fluids. This is best discussed with a specialist continence clinic.

The caecostomy catheter provides a regular, predictable method for defaecation and, due to its position, can be used independently by wheelchair dependent people. Many people who previously wore pads are able to wear regular underwear after a caecostomy tube.

Caecostomy catheters may not be suitable in people who have had previous abdominal procedures.

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