

**EFFECTIVE MANAGEMENT OF  
INCONTINENCE FOR YOUNG PEOPLE  
AND ADULTS WITH SPINA BIFIDA**

MANUAL

# spina bifida

## Introduction

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‘Effective management of incontinence for young people and adults with spina bifida is more than just learning to deal with a socially difficult situation.

Effective management enables people with spina bifida to achieve physical, social and financial independence. General practitioners can develop the skills required to help young people and adults with spina bifida to manage their own continence. This is why this manual exists.’

*Spina Bifida Foundation of Victoria*

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# spina bifida

## Introduction

### Using this manual

The aim of this manual is to provide quick access for general practitioners and other clinicians into the general primary care management of young people and adults with spina bifida, focusing on the complex issues of continence management.

While children are managed in paediatric specialist centres, upon reaching maturity, many young people and adults with spina bifida lose contact with specialist treatment centres and consequently may benefit from the latest treatment developments.

While many general practitioners may not have patients with spina bifida, *Taking control: effective continence management in spina bifida* enables clinicians to quickly familiarise themselves with key, essential management issues as they arise. For example, consider the following case study.

#### **Case study: New adult patient with spin bifida**

Geoffrey is a 25 year old man with spina bifida living in a remote rural area. He presents to you, his general practitioner, with increasing urinary and faecal incontinence. He has been stable on a regimen of clean intermittent catheterisation and daily bowel washouts, but has had increasing difficulty in maintaining continence over the past 3 months. With reluctance, he admits to increasing difficulty in achieving and maintaining erections.

How would you manage him?

In addition, the manual provides a link to specialist centres that manage spina bifida in young people and adults, enabling easy referral.

A companion patient information manual called *Passport to success* has been produced in association with this clinician's manual that focuses on incontinence issues in plain language and is available through spina bifida associations and foundations listed in the resource section of this manual. Both manuals can be downloaded from [www.racgp.org.au](http://www.racgp.org.au). They will be continually updated and available on this site.

The Spina Bifida Foundation hopes that these manuals become a valuable resource for clinicians managing young people and adults with spina bifida.



Rhys Williams  
A/President

Spina Bifida Foundation of Victoria

### *Spina bifida: key primary care issues for clinicians*

*<sup>1</sup>Spina bifida is a static congenital neural tube disorder resulting in continuing evolving disease involving multiple organ systems. Those affected require lifelong surveillance with coordinated management involving the patient, parents, general practitioners, neurologists, physiotherapists, occupational therapists, social workers, nurses, neurosurgeons, urologists, orthopaedic surgeons, physicians and other health professionals.*

#### **Key issues for clinicians**

- Spina bifida is one of the most severe congenital abnormalities compatible with a full and active life.
- Active surveillance to diagnose problems early is more likely to keep a person with spina bifida active and independent.
- Routine surveillance must involve regular review of the neurological, urological and musculoskeletal systems.
- Changes in neurological signs can indicate severe, treatable underlying pathology. Clinicians need to have a record of past neurological signs for a clear comparison in any ongoing neurological review.
- All new neurological signs require immediate referral to a specialist centre.
- Urological complications of spina bifida are a major cause of morbidity and mortality in spina bifida.
- Urological assessment is an integral part of ongoing management of spina bifida at all ages.
- Progressive loss of mobility profoundly decreases the quality of life for people with spina bifida. Attending to orthopaedic issues and encouraging appropriate physical activity helps to minimise the impact of progressive loss of mobility.
- Incontinence management is not just an aesthetic issue: it is a key barrier to achieving independence for young people and adults with spina bifida.
- Dealing with sexuality and reproductive issues is a routine part of managing spina bifida.
- Paediatric centres can help general practitioners locate adult spina bifida treatment centres. In the absence of adult centres, paediatric centres routinely offer support for adults with spina bifida.
- Cognitive dysfunction often adversely affects perception of symptoms and the ability of the patient to follow medical management instructions.

<sup>1</sup>The term spina bifida is used throughout this manual to refer to all myelodysplasias unless specifically stated. Similarly, issues specific to hydrocephalus alone will be highlighted, although many of the issues relating to spina bifida are relevant.

### Spina bifida: a severe congenital condition compatible with independent life

Spina bifida is a term used to describe a group of multiple complex congenital anomalies and abnormalities affecting development of the neural tube and related structures during the third and fourth week of pregnancy (see *Chapter 1 The spinal cord and brain in myelodysplasias* for a more detailed overview of spina bifida and neural tube defects).

One of the most serious medical conditions compatible with full life.

Spina bifida has been described as one of the most serious congenital conditions compatible with life. Prior to antibiotic and surgical advances of the last half century, life expectancy was often less than a year. Improvements in the management of children, young people and adults with spina bifida, related myelodysplasias and hydrocephalus has meant that many are now not only surviving into adulthood, but live active and happy lives, which may involve working and having children.

### Independent living for young people and adults with spina bifida: overcoming barriers

As more children with spina bifida reach adulthood, the special needs required to achieve and maintain independent living in the community are becoming more obvious and pressing.

Generalist clinicians who take a proactive role in detecting and overcoming the multiple problems facing young people and adults with spina bifida maximise their patient's chances of achieving physical and social independence. The clinician achieves this by becoming familiar with the physical, psychological, sexual and social barriers that impact daily upon the life of those with spina bifida, as well as their affected partners, families and friends.

### Assess now — don't wait until problems occur

*I live in the country and hadn't been to a specialist clinic for a check up for 10 years. I didn't know that spina bifida could change as I got older or that I needed to be seen regularly.  
Ben, 25 years*

Because the medical consequences of spina bifida are continually evolving, regular follow up is essential to prevent major problems before they occur.

Many people with spina bifida may have not attended a specialist centre for some time, and should be encouraged to attend for a routine assessment.

Familiarity with the primary care management of young people and adults with spina bifida enables treating doctors to ensure that regular, thorough and appropriate health maintenance occurs.

### The role of general practitioners and generalist health care providers

General practitioners can take a central role in the daily management of the young person or adult with spina bifida. As well as day to day treatment issues, the GP plays a major role in the early detection and referral of spina bifida related complications to treatment centres.

### Locating referral clinics for young people and adults with spina bifida

Paediatric centres can help general practitioners locate adult spina bifida treatment centres and also see *Chapter 9*.

Specialist clinics managing adults with spina bifida are a relatively recent development due to the increasing survival of children with spina bifida.

Children with spina bifida are managed in paediatric centres until adulthood, but upon reaching maturity may then lose contact with specialist treatment centres.

Paediatric centres can identify the location of adult spina bifida treatment centres, but many paediatric centres will treat adults with spina bifida if no other services are available.

### Critical areas of primary care of spina bifida: physical and psychosocial

The management of young people and adults with spina bifida and related disabilities depends upon many factors, but is largely determined by the level of the spina bifida lesion and the presence of hydrocephalus, related central nervous system problems and urological dysfunction. The nature and extent of complications can profoundly affect many body systems.

There are many psychosocial implications of spina bifida including personal, psychological, social and sexual issues.

### Monitoring of physical conditions<sup>1,2</sup>

While spina bifida is a multisystem condition, there are three main areas of functioning that require special ongoing surveillance:

- Neurological system
- Urological system
- Musculoskeletal system.

### Neurological system

Ongoing assessment of the neurological system is mandatory. Any changes in neurological signs require

immediate referral to a specialist centre.

While this may appear to be obvious, many individuals with spina bifida lose contact with specialist services on reaching adolescence and adulthood and may accept changing neurological function as an inevitable part of spina bifida. Always examine the nervous system as part of the general patient review and look for changes in neurological signs.

Changes in neurological signs can indicate severe, treatable, underlying pathology. The clinician's task is easier when they have a record of past neurological signs for a clear comparison in ongoing neurological review. All new neurological signs require immediate referral to a specialist centre.

Changes in lower limb neurological signs or continence can indicate the presence of a serious condition called tethered cord which can occur at any time of life (see *Chapter 3 Spinal cord tethering*). Clinicians managing patients with spina bifida need to be familiar with this condition.

Clinicians managing individuals with spina bifida need to be familiar with the tethered spinal cord syndrome.

The manifestations of a blocked ventriculoperitoneal shunt extends beyond headache and includes visual symptoms, confusion, somnolence, behavioural disturbances and loss of continence.

Clinicians managing patients with spina bifida need to be familiar with the symptoms and signs of a blocked ventriculoperitoneal shunt.

## Urological system

Although neural tube defects are neurological disorders, their impact on the urological system is responsible for a significant proportion of life threatening complications. As a consequence, an understanding of the detection and screening for urological abnormalities is central to an understanding of the management of spina bifida and related disorders.

In line with the urological management of spina bifida, attention to incontinence issues is central to the quality of life for all those with spina bifida.

Urological complications of spina bifida are a major cause of morbidity and mortality in spina bifida.

Due to neurological involvement of the bladder, bladder

control is commonly impaired. Not only does this greatly impact upon quality of life, but lack of bladder control can lead to major kidney damage through reflux and recurrent infection.

Urological complications are responsible for a large part of morbidity and mortality from spina bifida. These issues are fully covered in *Chapter 4 Urological management of spina bifida*.

Urological assessment is an integral part of ongoing management of spina bifida at all ages.

## Musculoskeletal system<sup>3</sup>

Normal nerve development and function do not occur at or below the spinal cord level of the spina bifida lesion. Therefore, the higher the lesion, the higher the level of the paralysis. However, the pattern of muscle weakness will vary with each individual, depending upon the site and extent of the spinal defect.

Muscle groups on opposite sides of the body may not be equally affected leading to orthopaedic abnormalities, ranging from scoliosis, dislocations, leg and foot problems. Lack of weight bearing can also result in an increased risk of fractures and progressive deformity.

Ongoing orthopaedic assessment and review helps to maximise function and minimise the impact of the disability.

Progressive loss of mobility profoundly decreases the quality of life of people with spina bifida. Attention to orthopaedic issues and encouraging appropriate physical activity helps to minimise the impact of progressive loss of mobility.

## Alarms bells for clinicians: some conditions require urgent referral

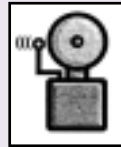
Clinicians also need to be aware of a number of important conditions that require more urgent referral to specialist centres. These conditions include:

- Spinal cord tethering (back pain and loss of lower limb, sexual and/or sphincter function (see *Chapter 3*)
- Headaches or cognitive impairment, especially as a sign of suspected ventriculoperitoneal shunt problems (see *Chapter 2*)
- Any change in neurological function or signs, regardless of how slow or minor
- Recurrent urinary tract infections (see *Chapter 4*)
- Renal impairment (see *Chapter 4*)
- Renal calculi.

These conditions are more fully explained in each section

as indicated, and clinicians involved in the care of people with spina bifida should familiarise themselves with these possible serious complications.

When you see the 'alarm bell' icon, this means that this point is important and may require urgent referral to a specialist centre.



### **Incontinence: a major barrier to independence**

A major barrier to total independence is urinary and faecal incontinence.

Effective management of incontinence enables access to education, work and financial independence, socialisation and fulfilling relationships.

#### Self esteem and incontinence

As well as preventing independence, poor incontinence management contributes to poor self esteem, which can discourage people from being active and social. Commonly reported feelings illustrate the personal impact of incontinence on self esteem: wondering if the person can go out; odour; worry; dismay; embarrassment; accusing looks; guilt; fear; isolation; sadness; and frustration.

### **Monitoring psychosocial impact of spina bifida**

Spina bifida can have a profound impact on body image and self esteem, and this can adversely affect the ability to form healthy relationships with family, friends, and partners. In addition, the common issue of incontinence can cause a further loss of self esteem.

This loss of self esteem not only interferes with a person's independence, but can erode their ability to participate fully in education, employment and other aspects of daily life that come with living a modern, active life.

A supportive primary care clinician aware of these issues can help a patient overcome these issues and gain full participation in society.

### **Dealing with sexuality and reproductive issues is a routine part of managing spina bifida**

Concern about sexuality and reproductive issues may be symbolic of the increasing need for independence in a person who has been, and often still is, highly dependent upon others in their daily living needs.

Spina bifida affects the spinal cord and inevitably affects sexual functioning. Dealing with these issues is a routine part of the daily management of spina bifida.

As for those without spina bifida, sexuality and reproductive issues become increasingly important for young people as they reach adolescence and adulthood. However, children with spina bifida have often grown up being especially dependent upon their parents and the emergence of sexuality issues can be a source of conflict for the person, their family and carers. This is dealt with in *Chapter 8 Sexuality and reproductive issues*.

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# Overview of spina bifida and the nervous system

## Chapter 1: The spinal cord and brain in myelodysplasias

*This chapter defines various neurological abnormalities and terms specific to the management of spina bifida and related disorders.*

### Key issues for clinicians

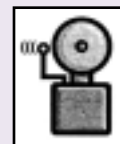
- While spina bifida is a congenital condition, complications may occur in young people or adults.
- Neural tube defects can affect the entire length of the central nervous system. This includes the brain as well as the spinal cord.
- Hydrocephalus affects most people with spina bifida.
- All those with spina bifida and their relatives require genetic counselling prior to conception.
- High dose folate supplementation is given to all high risk cases.

Spina bifida is a complex birth defect that has been recognised for thousands of years. Until the middle of the last century, most babies born with spina bifida did not survive more than a year, but due to medical advances — especially in neurosurgery and urology — most infants will survive into adulthood.

### Definitions

Spina bifida is the collective term used to describe a group of multiple, complex congenital abnormalities and anomalies of the neural tube known as myelodysplasias<sup>1,2</sup>. As spina bifida affects the entire length of the neural tube, additional central nervous system abnormalities are very common, especially hydrocephalus and Arnold-Chiari malformations (see below).

While spina bifida is a congenital condition, complications may occur in later life. The underlying pathology provides an understanding for the ongoing clinical management of those affected.



The commonest types of myelodysplasias are:

- Spina Bifida Occulta
- Meningocele
- Myelomeningocele
- Lipomatous malformations of the spinal cord and central nervous system
- Other related conditions: sacral agenesis, Vater's Syndrome.

**Spina bifida** occurs when the spinal column does not form correctly at some point along its length. In reality, spina bifida refers to a complex of conditions. Of the 260 000 children born in Australia each year, 400 are affected by this condition<sup>3</sup>.

**Spina bifida occulta** refers to incomplete development of the vertebrae, but the spinal cord is intact with no obvious skin defects. Spina bifida occulta occurs in 5% of live births. The overlying skin may also be normal, but may also be





Figure 1. Herald mark. Hairy patch over repaired lumbosacral spina defect.

associated with a dimple, hair patch or red discoloration — the so called herald or signature mark (see Figure 1)<sup>4</sup>. Signature marks may be associated with significant spinal abnormalities.

Abnormalities in function can emerge at any time later in life.

A **meningocoele** is a cerebrospinal fluid (CSF) filled cystic lesion where the meninges protrude into an external sac, usually located in the lumbosacral region, due to failure of closure of the vertebral arches. The amount of skin covering the lesion varies.

However, there is no nerve involvement. The lesion can be associated with hydrocephalus and central nervous system abnormalities.

A **myelomeningocoele** is one of the most important and severe types of spina bifida that occurs about 1 in 1000 live births. It occurs when part of the spinal column is undeveloped, with incomplete formation of the overlying vertebrae and no overlying skin. Most frequently located in the lumbosacral region, myelomeningocoeles are generally cystic and contain CSF that drains when the thin sac is disrupted. Both the meninges and spinal cord protrude into the sac, and the spinal cord is often abnormal. The level and severity of the lesion affects malformations and patterns of functional loss. Myelomeningocoeles are often associated with Arnold-Chiari Type II malformations. Approximately 80% of children with this lesion develop hydrocephalus.

### Other lesions

**Diastematomyelia** describes how the spinal cord is split into two hemicords with each having a set of dorsal and ventral nerve roots, each contained in a dural sheath. This belongs to the group of lesions called split cord malformations. Vertebral body abnormalities are often present, leading to scoliosis.

**Syringohydromyelia** occurs when a syrinx (a cystic cavity) forms within the spinal cord and may be found in the medulla or located anywhere from the cervical to the lumbar areas. Syringohydromyelia is associated with neural tube defects. The cystic cavity may or may not communicate with cerebrospinal fluid (CSF) flow pathways. The syrinx may progressively enlarge until

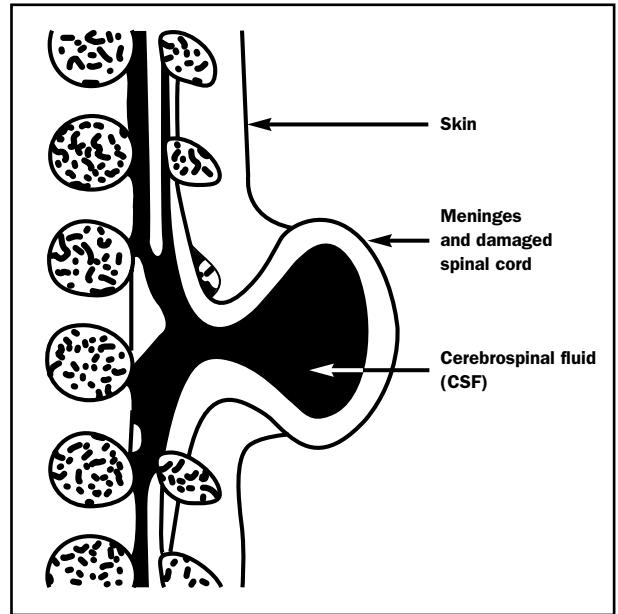


Figure 2. Myelomeningocoele. Myelomeningocoeles are the commonest cause of serious spina bifida related disability. The extent of the disability largely depends on the level of the lesion.

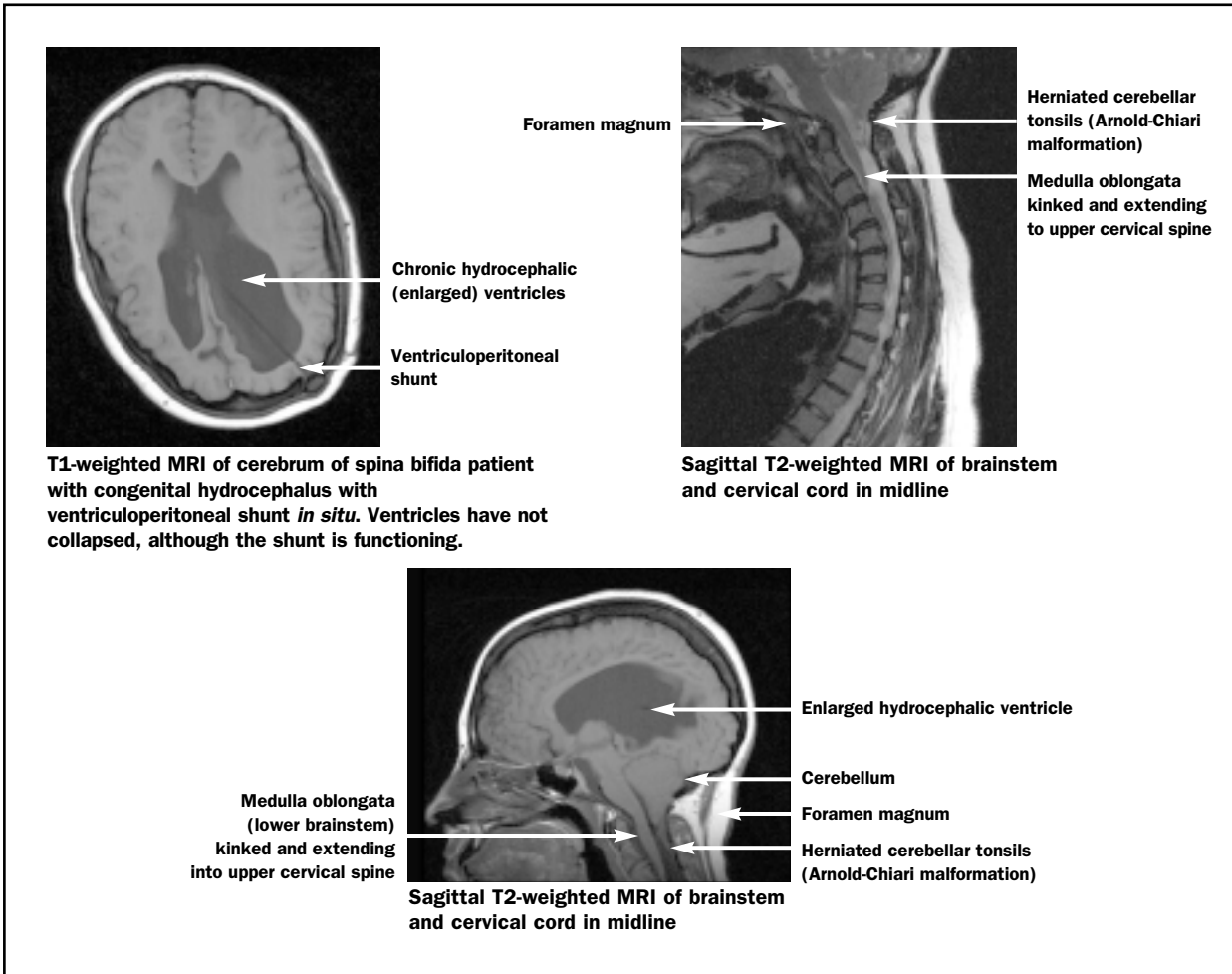
pressure on the nervous tissue leads to serious, variable complications.

**Lipomatous malformations** — commonly called lipomeningocoele — occur when excessive lipomatous tissue is within or attached to the spinal cord or filum terminale. This group of malformations are by far the most common form of closed spinal neural tube defects and vary from an enlarged filum terminale containing adipose tissue to a huge fatty mass occupying much of the dorsal lumbosacral region and contains the spinal cord and CSF. This group of lesions includes the lipomeningocoele, lipomyelocoele, leptomyelolipoma, lumbosacral lipoma and lipoma of the filum terminale. They represent a continuum of embryologic maldevelopments with similar clinical findings and prognosis to the open neural tube defects.

### Spina bifida and the brain

As neural tube abnormalities affect the entire length of the spine and central nervous system, most individuals with myelodysplasia will have associated brain abnormalities. This often results in hydrocephalus.

The **Arnold-Chiari malformation (Type II)** is the commonest brain abnormality in spina bifida. Affecting almost all people with spina bifida, this is an anatomical defect of the lower brain and cerebellar structures. This causes herniation of the cerebellar tonsils through the foramen magnum and the medulla to kink and move downwards into the cervical spinal canal, also



**Figure 3. Hydrocephalus and Arnold-Chiari malformation.** (Courtesy of Department of Diagnostic Imaging, Monash Medical Centre, Victoria.)

displacing cranial nerves. Around a quarter of patients with Arnold-Chiari malformations develop brainstem dysfunction, with symptoms often appearing in the first months of life.

Other central nervous system malformations can occur, including encephalocoeles, syrinxes above the level of the lesion, callosal agenesis and other brain stem abnormalities.

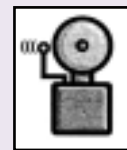
Symptomatic decompensation secondary to anatomical abnormalities can also occur in adults.

### Hydrocephalus

Hydrocephalus is found in most infants during prenatal ultrasonography. Most individuals with spina bifida and hydrocephalus require a ventriculoperitoneal shunt to relieve intracranial pressure.

If symptomatic hydrocephalus is present at birth, back surgery and shunt insertion may be performed at the same time.

Clinicians need to be aware of the signs and symptoms of hydrocephalus and the related signs and symptoms of ventriculoperitoneal shunts dysfunction (see *Chapter 2* for further information).



### Skin abnormalities may herald serious signs

Skin abnormalities can be associated with neural tube defects. As the skin and nervous system share a common ectodermal origin, this may explain the simultaneous presence of malformation of the skin and nervous system (*Figure 1*).

Skin lesions can be indicators of occult spina bifida, and are important as they may connect to the spine and be a source of infection.

The presence of a 'herald mark' is an important sign in young people with lower lumbar pain or sphincter dysfunction.

Commonly associated lesions include: dimples, tufts and patches of hair, pigmented areas, achordhons (pseudotails), lipomas, hemangiomas, dermoid cysts or sinuses<sup>3</sup>.

## Development of myelodysplasias: mechanisms and risk factors

Myelodysplasias are also called neural tube defects, which refer to their origin as developmental abnormalities of the central nervous system. These developmental abnormalities can occur at any site along the length of the brain and spinal cord.

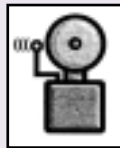
The critical development of the nervous system occurs in the first month after conception. If this process is disrupted, spina bifida may occur. When the brain does not develop, this results in anencephaly.

### Spina bifida occurs early in pregnancy

As spina bifida occurs during the first month of pregnancy, most cases will develop before most women know that they are pregnant.

### Role of low folate

Low folate at the time of conception is one factor with an increased risk of myelodysplasias. Prevalence has decreased in areas where folate had been added to food supplies, such as bread.



### Genetic factors

There is a strong familial tendency for myelodysplasias, although the exact mechanisms are unknown. A family with one child with spina bifida has a 4% chance of a second child being born with spina bifida and a 10% risk after two children with the condition. Siblings of a person with spina bifida and adults with spina bifida have a 2% chance of having a child affected with spina bifida. All relatives of people with spina bifida require genetic counselling.

### Genetic counselling is recommended:

- For mothers older than 35 years
- For all mothers with previously affected children
- When either prospective parent has myelodysplasia  
NB This includes all potential fathers with spina bifida
- When any family history of myelodysplasia is present.

### Drug induced myelodysplasias

The antiepileptic drug valproic acid (Epilim, Valpro) causes spina bifida. Alcohol and some antipsychotic drugs have also been implicated as causes.

**Valproic acid can cause spina bifida.**

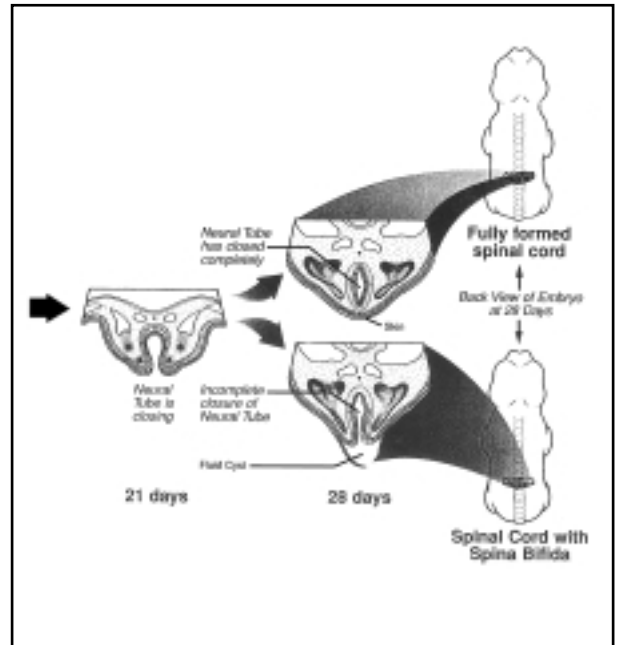


Figure 4. Development of spina bifida compared to normal development of the spinal vertebrae and central nervous system.

## Prenatal detection and management of spina bifida

Currently about 1 in 1000 pregnancies are affected with myelodysplasia although the number of new myelodysplasias is decreasing due to the increased use of routine folate and early detection of myelodysplasia through ultrasound scanning and serum alphafoeto-protein testing during pregnancy.

Current management practice consists of:

- **Preconception genetic counselling** usually by a geneticist, neurologist and obstetrician on the risks and benefits to enable patients to make informed decisions.
- **Preconception folate**
- **Antenatal diagnosis.**

### Preconception genetic counselling<sup>4-7</sup>

Those at increased risk of having children with spina bifida are offered genetic counselling (see *Chapter 9 Organisations and further resources*). Some young people and adults may not be aware of these increased risks and clinicians should always assess whether their patients are familiar with the risks of pregnancy and refer accordingly.

### Preconception folate

High dose preconception folate supplementation is the cornerstone of reducing the risk of spina bifida in families at risk.

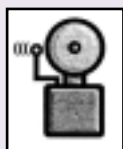
**Table 1. Preconception genetic counselling issues for those at increased risk of children with spina bifida**

- Discussion of risks of newly affected child
  - Approximately 2% risk where one parent affected
  - Approximately 4% risk where previous child is affected
- Attitudes of couples towards having a baby with neural tube defects including exploring their personal experience and understanding of neural tube defects.
- Discussion of possibility, risks and impact of anencephaly and spina bifida
- Role of preconception folic acid supplementation in reducing risk (see below)
- Availability and use of screening tests during pregnancy (see below)

While routine folate administration has been demonstrated to reduce the risk of neural tube defects on a population basis (such as 0.4–0.5 mg folate once daily), higher dosages of folate are routinely used in high risk patients (such as 5 mg once daily).

High folate supplementation is given in ALL high risk cases — not only just to high risk mothers, but may also be given to fathers in a group at high risk of developing spina bifida.

High dose folate supplementation is given to ALL high risk cases.



### Antenatal diagnosis

Ultrasonography can detect around 95 percent of cases of spina bifida by the age of 18 to 20 weeks. High risk pregnancies need to be screened by ultrasonographers experienced in the detection of neural tube defects. The first child with spina bifida in a family is often missed on ultrasound, as the pregnancy may be considered at low risk due to lack of previous family history of spina bifida.

Expert ultrasound can provide an approximate guide regarding the severity and level of the neural tube defect to assist in determining the prognosis for the foetus.

Alphafetoprotein serum levels can be raised at the

16th week of pregnancy. Spina bifida is not the only cause of a raised serum alphafetoprotein, but this is an indication for further testing. This especially applies to at risk pregnancies, such as in older women and individuals with, and relatives of those with, spina bifida.

A typical screening schedule would include:

- 12 week expert ultrasound examination
- 18 week expert ultrasound examination
- option of maternal serum screening (for trisomy 21 and neural tube defects).

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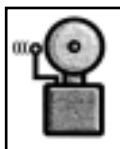
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### Chapter 2: The impact of hydrocephalus and other CNS conditions on case management

*Hydrocephalus affects most people with spina bifida and can impact upon continence management adversely in two main areas. Firstly, increased intracranial pressure can cause a deterioration of continence and mental status, which sometimes can be insidious and at other times dramatic. Secondly, the effect of cognitive deficits secondary to central nervous system abnormalities can be subtle, but can be a profound barrier to achieving successful case management, including continence control and independent living.*

#### Key issues for clinicians

- If there is any suspicion of raised intracranial pressure at all, patients with spina bifida need immediate referral to specialist centres for a full assessment. Any suspicion, no matter how small, is an indication for referral.
- Cognitive deficits secondary to hydrocephalus and other central nervous system abnormalities have a major impact on compliance with diagnosis, attending for investigations and following through with treatment and management plans.
- Clinicians can adopt strategies to help improve patient communication by understanding the types of cognitive problems that are common in people with spina bifida.



#### Hydrocephalus: an almost inevitable consequence of spina bifida

Hydrocephalus is not a specific disease, but rather a consequence of a diverse groups of conditions resulting from impaired flow of cerebrospinal fluid (CSF).

Around 90% of infants born with spina bifida have hydrocephalus.

#### Raised intracranial pressure from hydrocephalus: a life threatening complication

As neural tube defects affect the entire length of the brain and spinal cord, central nervous malformations are very common in people affected with spina bifida. One of the commonest types, the Arnold-Chiari malformation often results in raised intracranial pressure early in childhood, requiring the surgical insertion of a ventriculoperitoneal shunt to divert CSF flow. Shunts can block at any time, causing a life threatening condition, as well as long term central nervous system damage which has the potential to severely interfere with independent living.

#### Impact of hydrocephalus on physical and cognitive deficits

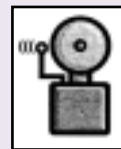
##### Impact of cognitive deficits on incontinence management.

As most people with spina bifida have some central nervous system abnormalities, cognitive deficits secondary to brain dysfunction and other physical complications — especially hydrocephalus — are common.

Good continence control usually involves fairly

Cognitive deficits secondary to complications have a major impact on compliance with diagnosis, attending for treatment investigations and following through with treatment and management plans.

Clinicians need to be aware of the extent of these often subtle effects to ensure maximal adherence to management plans.



complex procedures and, to be effective, depends upon good planning. The clinician needs to understand any potential cognitive barriers to effective continence management.

**Detecting raised intracranial pressure: a life-threatening condition<sup>1,2</sup>**

The diagnosis of raised intracranial pressure can be difficult, but a high degree of suspicion is necessary to avoid the possible severe adverse effects of complications secondary to central nervous system damage.

Raised intracranial pressure can be of gradual onset, increasing over a few months, and can be an important cause of change in overall continence status.

A clear understanding of the pathophysiology of the condition helps the clinician in diagnosis.

**Hydrocephalus and mechanisms of raised intracranial pressure**

**Where is CSF formed?**

CSF is primarily formed in the ventricular system of the brain by the choroid plexus, which is situated in the lateral third and fourth ventricles, although 25% of the CSF originates from extrachoroidal sources.

The total volume of the CSF is about 50 ml in infants and about 150 ml in adults. Most CSF is extraventricular.

**CSF flow mechanisms**

CSF flow results from a pressure gradient that exists between the ventricular system and venous channels. CSF flows from the lateral ventricles through foramina (foramina of Monro) into the third ventricle, and then pass through a narrow aqueduct (aqueduct of Sylvius) which is only 3 mm in length and 2 mm in diameter in children. The CSF then exits the fourth ventricle through three foramina (two foramina of Luschka and the midline foramen of Magendie) into cisterns at the base of the brain.

After exiting the ventricular system of the brain, the CSF then circulates over the cerebral hemispheres and spinal cord, and is absorbed by the arachnoid villi and to a lesser extent, by the lymphatic channels of the paranasal sinuses.

**Types of hydrocephalus: obstructive versus nonobstructive**

Hydrocephalus resulting from obstruction in the ventricular system is called obstructive or noncommunicating hydrocephalus. Hydrocephalus resulting from obliteration of the subarachnoid cisterns or abnormalities in functioning of the arachnoid villi is called nonobstructive or communicating hydrocephalus.

**Hydrocephalus in spina bifida**

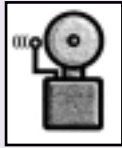
Hydrocephalus in spina bifida is usually due to the existence of the hindbrain malformation called the Arnold Chiari malformation (Type II). (see *Chapter 1*). Around a quarter of those with Arnold-Chiari malformations develop brainstem dysfunction, with symptoms often appearing in the first months of life (see clinical presentation below).

**Diagnosing raised intracranial pressure: clinician beware**

The symptoms of raised intracranial pressure can mimic many other conditions making diagnosis by even the most experienced specialist clinicians difficult. In young people and adults, raised intracranial pressure can be indolent, insidious and slow in onset, but can dramatically escalate over hours to a life threatening condition.

To avoid possible misdiagnosis, general practitioners need to have a low threshold for communication with specialist centres for assessment for advice.

If there is any suspicion of raised intra-cranial pressure at all, patients with spina bifida need to be referred to specialist centres for a full assessment. Any suspicion, no matter how small, is an indication for urgent referral.



**Causes of raised intracranial pressure in spina bifida**

Many young people and adults with spina bifida will have ventriculoperitoneal shunts inserted with the first few months of life. In these individuals, the concern is that the shunt has become blocked resulting in increased intracranial pressure.

Raised intracranial pressure secondary to Arnold-Chiari malformations can very occasionally occur in later life resulting in abnormalities of gait, spasticity and coordination during childhood.

**Presentation of increased intracranial pressure**

Clinicians need to familiarise themselves with the presentation of this life threatening situation.

Raised intracranial pressure can be a cause of change in continence patterns. Any change should be examined closely for the possibility of raised intracranial pressure or other neurological causes such as spinal tethering or other causes.

Insidious onset: change in continence patterns and other problems

**Table 2. Signs of raised intracranial pressure and VP shunt problems**

This is a life threatening situation. Symptoms can occur over weeks but can escalate over hours. Always refer urgently to a specialist centre if any suspicious symptoms occur.

**Adults, young people and children**

- Headache
- Nausea and vomiting
- Lack of appetite, refusal to eat
- Increased irritability, lethargy, drowsiness
- Personality changes
- Disorientation
- Pseudodementia
- Visual problems: nystagmus, double or blurred vision; setting sun sign in neonates
- Decreased motor and sensory function
- Fits and seizures
- Lower extremity hypertonia with generalised hypereflexia.
- Incontinence, especially a change in continence patterns

**Infants**

- Bulging fontanelle
- Increased head circumference
- Irritability
- Poor feeding
- Impaired cognitive development
- Respiratory stridor and/or high pitched cry in an infant

**Treatment: ventriculoperitoneal shunt**

Hydrocephalus is treated with the insertion of a ventriculoperitoneal (VP) shunt, usually within the first few years of life, to enable circulation of CSF and reduce the intracranial pressure.

**Signs of raised intracranial pressure and VP shunt problems**

Although this resource is aimed at health maintenance for young people and adults with spina bifida, the symptoms of raised intracranial pressure in children and infants are included, as it is such an important presentation (*Table 2*).

**Cognitive impact of hydrocephalus and other central nervous system conditions on patient management<sup>3</sup>**

Hydrocephalus can result in a series of physical complications that can adversely affect cognition. These

include memory abnormalities, attention problems, visual problems, behavioural problems including aggressive and delinquent behaviour, which all affect comprehension and adherence to any medical management plan.

While around 80% of people with spina bifida will have normal intellectual functioning, many will have subtle executive cognitive problems that can affect the outcome of any medical management.

**Other causes of cognitive problems.**

In addition, there may be other structural central nervous system abnormalities contributing to cognitive deficits. Commonly prescribed agents such as antiepileptic and anticholinergic drugs can also interfere with cognition.

Specialist help from a spina bifida specialist centre can help to address these issues, and contact with the centre can provide valuable help in overcoming any problems.

To help consultations, a list of common problems and some concrete strategies to overcome these problems follows.

**Common cognitive problems encountered in people with spina bifida**

*Organisational difficulties* interfere with the ability to think or perform activities in a fairly logical and planned way. This can cause difficulties in written language, learning sequences for procedures, keeping items and equipment in order, locating belongings or remembering to complete tasks.

Ringling patients with a gentle reminder about appointment times may help them to remember to keep appointments, although this has the potential to increase patient dependence. Encourage patients to remember their appointments by using a diary or the Health planner diary in the companion volume to this manual called *Passport to success*.

*Short attention span and distractibility* interferes with the ability to pay attention to important details of a new task. Brief attention spans mean that the person may not learn all of the necessary information, or may forget or hear only part of any instructions given, as well as taking longer to complete tasks. Distractibility can be internal, from the person's own thoughts, as well as from the environment.

*Deceptive language skills* There may be a stronger ability to say words than to comprehend their meaning. Despite what appears to be normal verbal skills, there may be a lack of comprehension of the actual meanings of words. This may reflect use of rote memory of sounds rather than their meaning, as some people with spina bifida have very good auditory memories, but poor comprehension.

**Perseveration**, or repeating information over and over, can occur and the clinician may mistakenly perceive that they are being understood.

The cocktail party syndrome describes a speech pattern characterised by the habit of repeating back phrases used, saying memorised common phrases (such as 'How are you?. Hi!) and talking about topics not always meaningful or appropriate to the situation. Cocktail party speech may be due to difficulty inhibiting the flow of thoughts going through the mind or difficulty focusing on and comprehending relevant aspects of a situation

**Difficulties in understanding language**, includes difficulty in answering questions, following instructions, participating in back and forth conversation, misinterpreting information or responding inappropriately to situations may indicate difficulties in actual comprehension.

**Difficulties with abstract reasoning** may cause difficulties in analysing and synthesising information and distinguishing between relevant and irrelevant information. This can also cause problems in generalising information and understanding complex information that involves words and concepts that cannot be seen or touched.

**Visual-spatial difficulties** can make simple tasks difficult. Tasks requiring making judgements about visual and/or spatial information such as tying shoelaces, doing up buttons and zippers without looking can become difficult. Technical procedures, such as teaching self catheterisation, need to take these difficulties into account.

**Lack of persistence** causes difficulty in focusing on tasks requiring internal motivation or have a reward that is delayed. Tasks with immediate rewards are more likely to encourage persistence.

**Lack of time management skills** may result in difficulties organising daily tasks, and may seem to do things at the last minute, or will not keep deadlines and appointment times. This behaviour can be interpreted as a lack of responsibility or caring about the needs of others, but may be a result of a deficit related to understanding the concept of time.

**Passivity, avoidance and withdrawal** may result when encountering difficulties with a situation, especially in a new context. Losing motivation to try new things may be a strategy to avoid failure. People who interact with the person may then focus on the behaviour rather than the learning difficulties that can result in unrealistic expectations or inappropriate programming.

## Strategies for maximising adherence to medical management

Learning to adapt treatment instructions to each individual's circumstances is an everyday skill for all clinicians. For people with spina bifida, there is no magic formula, but the following is a list of strategies for dealing with the commonest problems.

**Work in conjunction with multidisciplinary teams**, if possible, and be creative in finding methods that work for that individual person. Previous health personnel may have already identified the patient's learning strengths and weaknesses. Learn what you can about that person's pattern of learning. Also, if teaching a specific procedure, there may be specially qualified health personnel to assist, such as continence nurses.

**Encourage tasks that are possible**. Some people may believe that the tasks are too hard and may not have the belief that they are achievable. Helping to motivate a person's belief in themselves may be the first step to them achieving independence. Acknowledge all successes honestly and sensitively. Even if an outcome was not good, acknowledge the effort and attention involved.

**Help improve comprehension** by engaging eye contact whenever possible and have the person repeat back what was said.

**Keep verbal explanations simple**. There may be a tendency to over explain tasks in an attempt to achieve understanding, but keeping explanations simple with specific, concrete language is likely to be more effective. Adapt the methods used in the consultation to the person's strengths. For example, a person with problems comprehending language may have a better understanding by the use of explanations with diagrams and pictures, rather than repeating the same words over and over again. This will also help any visual-spatial problems.

**Get the person to explain to you previous instructions** to establish the level of comprehension of any previous instructions. This may not necessarily occur at the time of the consultation, but at follow up visits, when the person has had time to process information.

**Reward and promote persistence** to help lessen frustration and avoidance when learning new procedures.

**Reward successes** not only in successful tasks, but also when attempts have been made without successful outcomes. Empathise with any frustration due to successfully completed tasks, even though the desired outcome may not have been achieved.

**Avoid interpreting unsuccessful tasks as behavioural problems** as they may be due to cognitive deficits. This helps to focus the clinician's attention on overcoming



cognitive problems and reduces any frustration on the part of the patient.

**Break tasks down into smaller steps** and provide time and structured instruction for each step, and rewards for success as each stage is completed. Learn to do one step at a time.

**Reduce demands to realistic levels.** Rather than trying to achieve a whole series of tasks at once, try to spread tasks over time, so that such that goals are realistically achievable. This helps to lessen frustration.

**Teach organisational skills** such as ‘Everything has a place and a place for everything’. A highly structured environment may make task completion easier.

**Establish routines and structures** as much as possible.

**Use checklists, calendars and diaries.**

**Tape recorders** are useful for some people to prevent the need to take notes and to help remain attentive during listening.

**Be conscious of attention span.** With experience, the clinician may be able to judge how much information can be taken on board at each session, and then tailor the consultation to the person’s needs.

*Passport to success*, the companion patient guide to this manual includes a planning diary and has been produced in a user friendly way to help overcome the above barriers. The guide can be downloaded and provided to your patients.

[www.sbav.org.au](http://www.sbav.org.au) or [www.racgp.org.au](http://www.racgp.org.au)

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## Chapter 3: Spinal cord tethering

*Tethering of the spinal cord is an insidious cause of worsening incontinence that requires urgent medical assessment and possibly surgical intervention to prevent any further deterioration. Clinicians monitoring young people and adults with spina bifida need to maintain a high degree of suspicion to ensure the early detection of tethered cord syndrome to help promote and maintain independent living.*

### Key issues for clinicians

- Clinicians need to consider spinal cord tethering as a possible cause of change in continence patterns.
- Any suspicion of spinal cord tethering should be referred to a specialist centre for urgent assessment.
- Earlier surgical intervention in clinically demonstrated spinal cord tethering is more likely to result in an improved long term outcome.

### Tethered cord syndrome: an insidious condition

Tethering or stretching of the spinal cord in young people and adults results from fixation of the spinal cord to inelastic structures<sup>1</sup>. Spinal cord tethering is a major source of morbidity in spina bifida and clinicians need to be familiar with its presentation and maintain a high degree of suspicion when monitoring patients with spina bifida.

In the past, spinal cord tethering was thought of as mainly a condition affecting only children, especially during growth spurts, but the condition can occur in people with spina bifida at any age.

Spinal cord tethering is a particularly insidious complication of spina bifida that can have a major adverse impact on independence through its effect on mobility and continence.

### Many causes but consistent presentation

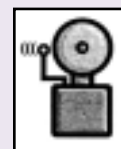
The fixation of the spinal cord in spinal tethering is due to fibrous or lipomatous tissue. In spina bifida this can be due to a myelomeningocele, lipomyelomeningocele, scar tissue, a fibroadenomatous filum terminale or many other conditions<sup>2</sup>.

### Clinical presentation of tethered cord syndrome

#### Symptoms and signs

Tethered cord syndrome typically causes a progressive loss of function at or below the level of the spinal cord defect, with or without lower lumbar pain.

Regardless of the particular mechanical cause of spinal tethering, the spectrum of clinical presentations of tethered cord syndrome are consistent and should alert the clinician to the need for IMMEDIATE neurological and neurosurgical referral.



**Table 3. Characteristics of pain in spinal cord tethering**

- Pain in lower back and legs exacerbated by physical activity, especially, any which involves flexion and extension of the lumbosacral area
- Leg pain is often in medial, lateral, anterior, or posterior aspects of thighs or legs
- Groin pain or pain in genitorectal area is common
- Pain is not less when lying supine (as opposed to disc disease).
- Straight leg raising causes no difference to pain
- In adults, pain governed by three 'B' signs (usefulness will depend upon any pre-existing neurological and orthopaedic disabilities):
  - Inability to sit with legs crossed like Buddha
  - Difficulty in bending slightly at the waist with activities such as washing dishes
  - Holding a baby or light material (< 2.5 kg) at the waist level while standing

**Table 4. Common findings in tethered cord syndrome**

- Pain (see previous table)
- Gait problems, often quite subtle.
- Progressive urinary incontinence: symptoms including urgency, frequency and enuresis
- Progressive faecal incontinence including urgency, frequency, and encopresis
- Any motor or sensory deficits in lower extremities
- Altered sensation in genital regions and during sex
- Impotence
- Muscle weakness
- Muscle atrophy
- Hyporeflexia, especially any change in pre-existing signs
- Faecal incontinence
- Scoliosis/lordosis
- Foot deformities
- Skin abnormalities — herald marks

Tethering may occur in young adults at times of growth spurts, when lengthening of the spinal column can increase spinal cord tension, but can occur at any adult age.

**Key diagnostic issue: progressive loss and change**

The key diagnostic issue in diagnosing spinal cord tethering in young people and adults with spina bifida is a progressive deterioration in neurological function at or below the level of the defect. This includes urinary and faecal incontinence.

Spinal tethering needs to be excluded in the presence of progressive deterioration of urinary or faecal incontinence.

Neurological deficits in spina bifida are usually not progressive. Any change in signs requires immediate specialist assessment.

Spinal cord tethering can also occur in adults with no known past history of spina bifida.

**Patchy distribution of clinical findings of tethered cord syndrome**

The pattern of clinical findings of spinal cord tethering often fails to follow strict dermatomal patterns like those due to compression of one or two nerve roots or a

particular level of spinal cord injury. Weakness, pain and other signs of spinal cord tethering may have a patchy distribution below the level of the lesion, rather than a strict neurotomeal pattern.

**Pain in spinal cord tethering**

Regardless of the cause, the characteristics of the associated pain are often suggestive of spinal tethering (*Table 3*).

While other causes of back pain, such as disc herniation need to be excluded, clinicians still need to organise urgent neurosurgical referral and assessment to ensure that spinal cord tethering is not missed.

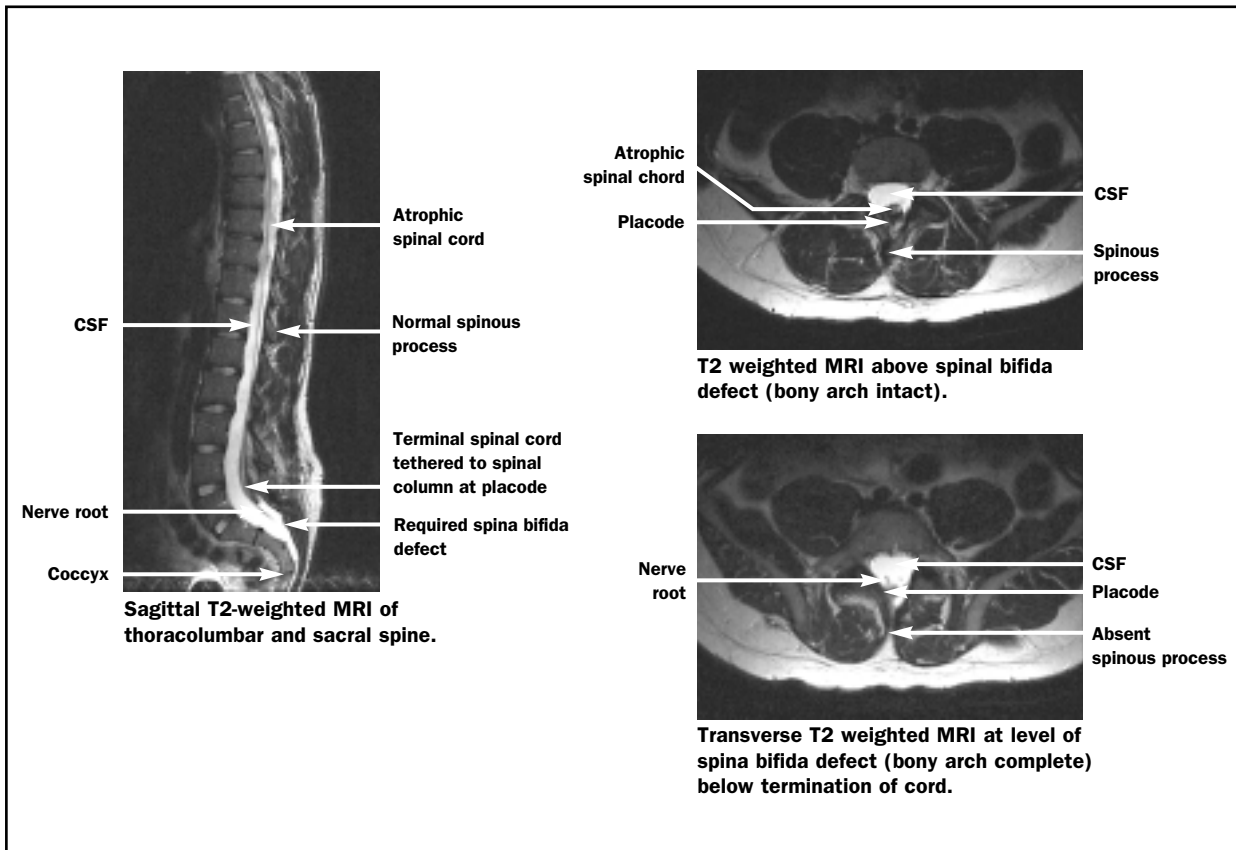
**Other common findings in tethered cord syndrome**

As highlighted above, the findings need to be considered in the context of any pre-existing neurological abnormalities, but clinicians should have a low threshold for specialist referral when there are any changes in pre-existing clinical findings (*Table 4*).

**Management: diagnostic imaging and surgical intervention**

**Magnetic resonance imaging**

Magnetic resonance imaging (MRI) is the best currently available technique for viewing the spinal



**Figure 5. Surgically repaired spina bifida defect with absent spinous process and posterior spinal arch at low sacral level. Atrophic cord tethered to placode and spinal column.** (Courtesy of Department of Diagnostic Imaging, Monash Medical Centre, Melbourne.)

cord. Most people with spina bifida will have some MRI findings suggestive of spinal tethering, but the decision to treat is based upon a combination of MRI and clinical findings. Ideally, a baseline MRI should be taken on all young people and adults with spina bifida to compare any changes with subsequent imaging, should symptoms of tethering arise.

### Typical MRI findings in spinal cord tethering

Typical MRI findings in spinal cord tethering include

- Thick filum terminale (>2 mm in diameter)
- Presence of structures such as fibrolipomatous filum terminale
- Obliteration of subarachnoid space suggesting caudal spinal cord or nerve root adhesion
- Changes in the structure of the spina bifida lesion such as dermoid, epidermoid cyst, myelomeningocele, lipomyelomeningocele or other problem
- Elongation of spinal cord
- Posterior displacement of conus medullaris with the filum pressing against the thecal lining at or near L5, or when compared with previous films.

### Surgical intervention: indications for detethering the cord

Once the diagnosis of spinal cord tethering is made, the decision for surgical intervention is based on clinical evidence. Treatment is especially indicated in the presence of new or worsening symptoms.

In studies of adults, when performed by experienced neurosurgeons, spinal cord tethering has been demonstrated to be a well tolerated, effective intervention.

Ultimately, the neurosurgeon can only confirm the presence of spinal tethering at operation. The surgery performed depends upon the intraoperative findings.

### Effect of surgical intervention on spinal cord tethering<sup>3-5</sup>

Timely surgical intervention of tethered cord syndrome can arrest, and in some cases improve neurological signs. The more long standing the neurological signs, the less the chance of resolution of symptoms and signs.

Pain improves in many people and is usually relieved within three months of the untethering, but improvements

in neurological signs can take many more months.

Patients need constant monitoring after the operation to assess recovery and help to adjust to any lifestyle changes.

**Case history: a 27 year old woman with worsening incontinence.**

M. is a 27 year woman with spina bifida. She is independently mobile, has never used calipers, has no hydrocephalus, is fully employed, and is in a steady relationship

M. presents with 12 months of progressively worsening faecal incontinence. She is now freely incontinent of faeces and manually evacuates herself before going out to avoid humiliation.

On examination, the anal tone is grossly reduced and at the time of assessment, anal sphincter reconstruction was being considered

She has also had mild urinary frequency and urgency for 6 months, as well as 8 months of reduced vaginal sensation during intercourse.

Her MRI demonstrated the presence of a tethered cord with a large neural placode. M was referred to a neurosurgeon and detethering is now planned.

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## 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 in spina bifida: the neurogenic bladder<sup>1-3</sup>

Effective control of urinary incontinence in spina bifida needs to acknowledge the special management issues related to the presence of abnormal neurological bladder function in spina bifida, 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 working, watching television or busy with other tasks. Abnormal anatomical distribution of the nerves may cause sensations to appear 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, important for continence control.

### Neurogenic bladder and sphincter abnormal function patterns in spina bifida: 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

Altered sensation affects bladder control.

One of the most serious medical conditions compatible with full life.

- 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:

- **Hyperreflexic** 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 hyperreflexic 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.

### Complications of the neurogenic bladder

Most children born with spina bifida have normal renal function. However, the presence of the neurogenic bladder can lead to urinary retention with overflow, uretric 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 may be 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

UTIs 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 quite common, any recurrent urinary tract infection, 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 secondary to 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 or stinking, 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?**

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.

The key issue is one of referral and follow up.

A 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 an 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 3 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, ongoing, active surveillance at all ages reduces morbidity and mortality.

### **Lifelong surveillance: the role of the general practitioner**

General practitioners may see patients with spina bifida who have not had regular urological monitoring or assessment. When reviewing patients with spina bifida, general practitioners need to ensure that urological



monitoring has taken place appropriate for the age group. 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, general practitioners 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 5 years of age**

Children below the age of 5 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 *Cognitive impact of hydrocephalus and other CNS conditions on patient management Chapter 2*). 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 in spina bifida**

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 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, 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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1. Donnellan S. Urological management of the patient with spina bifida. Monash Medical Centre. 2001
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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.
- Continence is best managed in conjunction with a specialist continence clinic.
- Most young people and adults with spina bifida will have already established continence 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 continence management. Clinicians need to familiarise themselves with this procedure.
- Continence procedures need to be reviewed after an urinary tract infection.
- Persistent changes in continence patterns should be referred to a specialist clinic.
- There are surgical interventions available to assist continence 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 and difficulty in coping.

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 general practitioners 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 in spina bifida<sup>1-3</sup>

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 general practitioners can greatly improve quality of life by referring patients to these specialist centres. The general practitioner 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 incontinence clinic.

### **Conservative management**

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

### **Controlling urinary continence — establishing a routine**

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

### **Continence control in spina bifida — different from routine clinical practice**

Clinicians need to be aware of differences in continence control for those affected by spina bifida. For example, the experience of many general practitioners in continence control will be in children with normal bladders. Continence 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.

### **Urinary incontinence — timing is the key**

The key to successful control of urinary continence 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 many.

Successful bladder timing — that is, the regular emptying of 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 continence control. When routines become upset, then this pattern can be used as a target to get habits back into a routine.

### **Establishing and reinforcing urinary continence control: intermittent catheterisation**

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

#### **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 non-existent.

Detrusor 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 episode of incontinence.

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

Continence 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 continence, 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 already have been using intermittent catheterisation for many years.

Many young people and adults with spina bifida will have already been using intermittent catheterisation for many years. In this case, the role of general practitioners 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.

Review catheterisation techniques after a urinary tract infection.

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 — definition and techniques

Catheterisation aims to empty the bladder to protect renal function and to achieve social independence through prevention of incontinence. Catheterisation is a simple, clean — but not sterile — method of inserting a plastic catheter several times a day to drain urine.

### 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 incontinence 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 technique is often done with the assistance of a specialist incontinence team, the general practitioner can assist by going through the patient's technique to ensure that each step is performed correctly. The following checklist has been prepared to help clinicians check that basic technique is adequate, but more detailed information and assistance is available from the specialist incontinence team.

General practitioners with any questions can contact the incontinence nurses or other health professionals of incontinence 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.

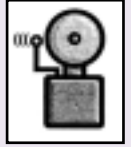
### Case history: Betty is a 26 year old woman with incontinence.

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 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.

**Table 5: Checklist for reviewing self catheterisation technique<sup>2</sup>**

- Have copies of any patient instructions for procedures included in their file as part of the routine medical history 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.
- 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 lower urine output than expected during drainage. In this case, repeat the procedure in 1/2 to 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.

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



**Table 6: Clean intermittent catheterisation — instructions for males<sup>2,4</sup>**

**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, cleaning toilet seat

**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 adverse effects that can interfere with cognition, which may 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.

**Table 7: Clean intermittent catheterisation — instructions for females<sup>2,4</sup>****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.

**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 in spina bifida**

The following is a list of common urological procedures used in spina bifida, and their commonest 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 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 ileal and colon conduits and cutaneous ureterostomy.

**Augmentation cystoplasty**

Augmentation cystoplasty involves surgically configuring a segment of bowel to augment the bladder to correct vesicoureteric reflux. When deciding upon an augmentation cystoplasty, issues to consider include which part of the bowel to use (for example, ileum, 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 mucous 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

**Surgical intervention in spina bifida**

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. 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 commonest 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.

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<sup>6</sup>.

#### 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<sup>3</sup>. 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 and may have not had 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.

#### Case history: John is a 22 year old man

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 able to be reversed, with him starting a clean intermittent catheterisation routine. Enthusiastic to find out more, he is eager to visit the adult spina bifida clinic to find out more.

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

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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 has many effects on the ability to control faecal continence. An understanding of these 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 continence management.
- Faecal continence control is best managed in conjunction with a specialist spina 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 a useful for softening and loosening stools.
- Clinicians need to familiarise themselves with continence appliances such as anal plugs.
- Surgical procedures are an important method of continence control.

### 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 in young people and adults with spina bifida

Young people and adults with spina bifida visiting their general practitioner will have already established their bowel habits, although some may not be benefiting from more recent developments in surgery and appliances. The general practitioner'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 for young people and adults: 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.

### Role of general practitioners 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 general practitioner 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 treatment of faecal incontinence. By referring to specialist clinics, general practitioners can ensure that patients can benefit from latest management techniques.

### **Faecal continence control — special issues for spina bifida<sup>1-3</sup>**

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 continence management, the presence of spina bifida adds another set of issues that need to be addressed in order to achieve continence control. The following is a list of areas that clinicians need to consider when assessing faecal continence.

#### **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, which may be for some time.

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

Diarrhoea can make the practical management of continence 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 continence 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 bowel of children with spina bifida can be slow to develop. Some children will be 8 or 9 years of age before the bowel is mature, which can mean that there are many years of faecal incontinence.

#### **Mobility, physical 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 continence 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 can also cause changes in bowel habits. Common exacerbating factors such as: 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 continence control problems. In addition, there may be difficulty in transferring to toilets if in a wheelchair.

### Overview of conservative management of faecal continence

#### 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 continence control.

#### Diet, fluids, diarrhoea and constipation<sup>3,4</sup>

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<sup>3</sup>

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*.

**Table 8. Foods which are frequently associated with causing loose stools and faecal incontinence<sup>4</sup>.**

- 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 (use foods containing high fat, high fibre, moderate sugar and higher water)<sup>4</sup>**

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	Tin 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

### Bowel emptying — overview of methods and techniques<sup>2</sup>

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 general practitioner 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 faecal continence control.

**Table 10. Dietary control of stool consistency — foods that harden stools (use foods containing high protein, low fat, moderate carbohydrate, low fibre and low water)<sup>4</sup>**

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, pump 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 (Adapted from <sup>3,4</sup>)**

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

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 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.

**Large volume fluid enemas<sup>5</sup>**

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; a colonic washout set or a specially designed set. 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 requiring cleaning.

Enemas will clean the bowel for 2 to 3 days and can often be performed by an adult without assistance. While many patients find enemas a manageable way to control

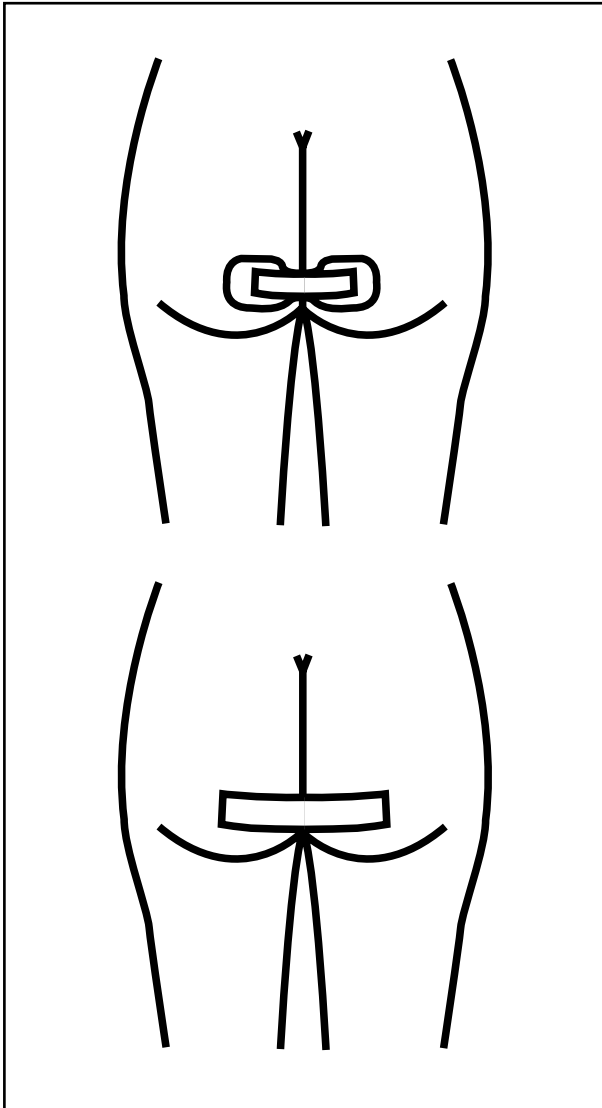


Figure 6. Buttock strapping

#### Strapping buttocks: procedure

- The tape is applied low on the buttocks so that it is 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.

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 even most of the time.

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 that 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. This allows plenty of time for removing the plug 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.

### Surgical procedures to control faecal incontinence

When bowel emptying and faecal incontinence is not successfully controlled with the above 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.<sup>6-8</sup>

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

**Case study: Peter is a 21 year old man with 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 house bound more 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 2 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.

smell as the bowel contents are sterile at the level of the appendix. The stoma can be left in place long term 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 section *Overview of common surgical procedures in spina bifida in Chapter 5 Controlling urinary incontinence*).

**Caecostomy catheters<sup>9,10</sup>**

A caecostomy catheter is a non latex, 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 incontinence 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 used to wear 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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# Other primary care issues for people with spina bifida

## Chapter 7: Health promotion strategies and other clinical issues in spina bifida

*A routine approach to reviewing spina bifida helps clinicians detect problems requiring early treatment and referral. In addition, information, such as dietary and other lifestyle information can help promote a sense of health and well being.*

### Key issues for clinicians

- Attention to general health measures may help prevent significant long term complications of spina bifida.
- Obesity is a major issue in young people and adults with spina bifida and is a significant barrier to independent mobility.
- Self care, including skin care, is an important part of general health for people with spina bifida.
- Peer support organisations can provide important access to resources and support.

### Early prevention can minimise long term problems<sup>1-3</sup>

The early detection of changes in the clinical condition and of any complications of spina bifida is a key. While monitoring of neurological and urological changes have been highlighted in earlier chapters, there are other measures that general practitioners can take to enhance the quality of life for young people and adults with spina bifida.

General practitioners often diagnose changes early, which enables them to refer these problems to specialist centres early in their course.

Some common problems that general practitioners need to be aware of are highlighted here.

### Obesity in spina bifida

Nutrition plays a vital role in spina bifida in terms of general health, bowel function regularity and the prevention of obesity.

Lack of mobility is a major contributing factor to the development of obesity in spina bifida.

Neurological and orthopaedic problems can decrease mobility, resulting in a weight gain. This weight gain can result in a decrease in mobility, contributing to further weight gain. The loss of mobility may be so severe as to result in the person being confined to a wheelchair.

The general practitioner can play an important role in monitoring this weight and intervening early to prevent weight gain.

### Promoting exercise, sports and activities

General practitioners can improve the general health of people with spina bifida by promoting the role of physical exercise.

In addition to helping to maintain mobility and prevent obesity, sport can be a major opportunity for socialisation, formation of relationships, peer support and meeting prospective partners.

### **Skin care — pressure sores and neuropathic skin**

Skin problems, often related to immobility can be a continuing source of morbidity. Early treatment is the key to successful outcomes.

Altered sensation from spina bifida can result in neuropathic, atrophic skin with ulcers and chronic infection. Poor vascular supply and abnormal autonomic function are also common in spina bifida and can contribute to skin atrophy.

Lymphoedema, secondary to immobility is common and can result in skin damage and ulceration.

Care should be taken to ensure that shoes fit properly, and that wheelchairs, general aids and aids to transfer patients do not contribute to skin damage.

Patients may benefit from referral to specialist lymphoedema clinics.

### **Latex allergies**

Allergies to latex are more common in people with spina bifida than the general population. Reactions can vary between mild reactions to severe anaphylactic shock.

Care needs to be taken to avoid the use of rubber gloves and any other latex items, such as the balloons in catheters. Some hospitals have latex free operating theatres for procedures for allergic individuals.

Clinicians need to remain alert to this possibility and to refer to specialist clinics for advice if the situation arises.

### **Self care and hygiene**

Cognitive and physical deficits can contribute to poor self care and hygiene. A deterioration in self care may point to worsening underlying medical problems. Patients may require teaching of simple grooming techniques adapted so that they are achievable within the context of any cognitive deficits.

### **Social development and educational strategies**

Peer support is one of the most important ways in which people with spina bifida learn to adopt healthy behaviours. Encouraging patients to join spina bifida associations can provide opportunities for peer support, which can increase self esteem and personal and social relationships (see list of associations in *Chapter 9*).

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### **Case study: building links through community organisations**

Toby, a handsome 26 year old man with spina bifida at the S1–2 level. He is in full time employment, had lifelong urinary incontinence, but not faeces. He presents with the difficult problem of urinary continence after he ejaculates. Toby self catheterised in the morning and at night, and used a pad during the day. As a result, he had low self esteem. He attended a spina bifida association meeting where incontinence was discussed. He was encouraged to attend a spina bifida clinic and was subsequently referred to a urologist.

Urodynamic studies were performed and full continence measures have been instituted. His resulting social and sexual function have greatly increased and he confidently anticipates the prospect of finding a girlfriend.

## Chapter 8: Sexuality and reproductive issues

*Managing issues of incontinence inevitably raises issues of sexuality. The complications of spina bifida that affect urinary and bowel function also affect sexual functioning. Routine review requires regular assessment of sexual issues while taking into account any special needs. This chapter highlights some of the common issues that general practitioners need to know when treating people with spina bifida.*

### Key issues for clinicians

- Spina bifida affects sexual function.
- Fertility is often normal in women.
- Vaginal delivery is the preferred method of delivery. A urologist should be present during a caesarean section, especially if there has been past urological surgery.
- There can be problems with erection and ejaculation in males. Ejaculation difficulties are a significant barrier to conception.
- Treatment is available for males with spina bifida.
- Assisted reproductive technologies may be of help in achieving pregnancies.

Clinicians involved in the management of spina bifida need to create an open, nonjudgmental environment that is favourable to the discussion of issues of sexuality. Dealing with issues of continence control inevitably raises questions of sexuality and the clinician needs to be prepared to deal with such important issues.

### Self esteem issues of spina bifida impact upon sexuality

The issues of body image and self esteem that arise in spina bifida profoundly influence a person's sexuality, but even people with profound disabilities are capable of active sex lives. Creating a safe environment for discussing and dealing with such issues helps to foster a sense of personal wellbeing and promote healthy sexuality.

### Creating an open environment for discussing sexuality

General practitioners can greatly improve the quality of life of their patients with spina bifida by providing a supportive environment for the discussion of sexuality.

Providing permission to discuss sexual concerns helps to normalise sex and helps to overcome fears and isolation that patients may have about sex. This is especially important for young people who are in the process of becoming independent from the parents and carers, and who might not otherwise have the opportunity to discuss sexual matters.

Taking the time to listen to sexual concerns validates the fact that the clinician is prepared to help deal with sexual issues. While the general practitioner may not have the expertise or resources to deal with complicated issues of sexual function in spina bifida, demonstrating a commitment to listen enables the general practitioner to detect any concerns early, and then refer the issues to a sexual counselling clinic if necessary.

Providing simple suggestions may be all that is necessary in some cases to assist some sexual problems. As feelings of isolation are common in sexual health



problems, minor concerns can often become major sources of anxiety. Patients may just require simple information about sexuality, such as what menstruation or a nocturnal emission is, to reassure and normalise their sexual concerns.

Many patients will require intensive therapy for specific sexual problems, and this can be arranged through specialist spina bifida clinics for adults or young people.

### Privacy concerns and young people

Some young people may not want to discuss sexual health issues accompanied by their parents. Clinicians may need to indicate to a young person that they are prepared to discuss sexual health problems in the absence of their parents, although considerable diplomacy and tact may be required with the parents when dealing with this sensitive issue.

As long as clinicians are aware that privacy may be an issue, then strategies for achieving this are likely to evolve.

### Effect of spina bifida lesion on sexual function

Vaginal or caesarean delivery?

The effect of spina bifida lesions on sexual function varies widely between patients, and often the best way to assess function is through a neurological

opinion. This can give a prognosis of the person's anatomical and physiological sexual function, which can provide the basis for developing management strategies during subsequent sexual counselling.

Males may have normal sexual function, but are commonly affected to some degree. Satisfactory erections are often possible, but without ejaculation. Other types of sexual dysfunction are also possible. In some cases, circumcision may be considered appropriate.

In males and females, issues of altered genital sensation can affect arousal patterns and sexual function.

Orthopaedic problems, for example, with lower limbs can affect the ability to use certain sexual positions.

All of these physiological and anatomical issues and others need to be taken into account when counselling on sexuality issues.

### Safe sex

Safe sex education needs to take place in the early teen years of young people, as precocious puberty is very

The high risk of neural tube defects in pregnancies of women with spina bifida makes safe sex education — in a style and content appropriate to the individual — prior to the onset of sexual activity mandatory.

common in people with spina bifida.

Safe sex education needs to be adapted to the special physical and cognitive needs of the individual with spina bifida.

For example, the use of some types of contraception, such as condoms, requires good hand eye coordination. Problems with coordination and manual dexterity need to be taken into account when recommending specific types of interventions.

Similarly, the cognitive effects of spina bifida (*Chapter 2, Impact of hydrocephalus*) need to be considered when educating about safe sex.

### Sexuality, conception and pregnancy issues

#### Issues for women

Although women with spina bifida often report altered vaginal sensation, normal sexual response is often possible.

Spina bifida generally does not affect fertility in women, and contraception and preconception counselling is therefore paramount to decrease the risk of unplanned pregnancies and neural tube defects.

The course of pregnancy in spina bifida is similar to that in women without spina bifida except for an

- increased risk of urinary tract infection
- risk of pressure sores
- increased rate of lower pelvic pain

The current recommendation is that pregnant women with spina bifida be encouraged to deliver vaginally as women who have vaginal deliveries have fewer complications, faster recovery times and shorter hospital stays.

Women delivered by caesarean section have a higher rate of complications, and surgery is often complicated if there has been past urological surgery for spina bifida. The presence of pelvic scarring often makes identification of ureters and other structures difficult.

A urologist should be present to assist the obstetrician during a caesarean section, especially in the presence of prior urological surgery in women with spina bifida.

### Is there an increased rate of birth abnormalities?

Potential parents will want to know about the possibility of birth defects, especially in view of a mother affected with spina bifida. Apart from the increased risk of neural tube defects, the question of whether there is an increased risk of birth defects is unknown.

Newer fertility techniques such as fertility drugs and

in vitro fertilisation techniques are sometimes used in people with spina bifida in conjunction with specialist spina bifida clinics.

#### Issues for males

Less is known about fertility in men with spina bifida than in affected women. Undescended testes are more common in men with spina bifida and this can contribute to poor fertility. In addition, repeated catheterisation may result in scarring and past epididymoorchitis infections from repeated urethral instrumentation can also decrease fertility.

However, men with spina bifida can still father children, although they may require the use of assisted reproductive technologies.

The major difficulties associated with conception in men with spina bifida are associated with achieving erection and ejaculation.

Difficulty in achieving an erection is a common problem for men with spina bifida, and while this may be treatable in some men, achieving ejaculation is more difficult.

The lower the level of the lesion, the more likely the male will be able to achieve an erection.

Lack of erections can be treated by using physical techniques, such as vacuum pumps, pharmaceuticals such as sildenafil or prostaglandin injections such as Caverject, and with surgical techniques such as penile prostheses. Issues of cost of treatments become important for patients, especially if they are on a low income or a pension.

Ejaculation in many men with spina bifida does not usually occur during sex. Ejaculate isolated using vibroejaculation and electroejaculation techniques often has low sperm counts, resulting in a reduced conception rate.

### Assisted reproductive technologies

Many of the issues surrounding conception in spina bifida

#### Case study: male sexual dysfunction in spina bifida.

Rodney, a 34 year old male with spina bifida at L2–3, shunted hydrocephalus and mild intellectual disability had been wheelchair bound all of his life. An ileal conduit had been constructed when he was a child.

At 34 years of age, Rodney formed a stable relationship with a nondisabled woman. He had never been able to have an erection or to ejaculate. He was prescribed Caverject injections and was counselled as how to optimise pleasure from intercourse, given his severe impairment of mobility. He and his partner enjoy a happy and fulfilling sexual life and are planning to commence a family.

are due to mechanical and anatomical difficulties in conceiving, rather than a lack of fertility. Access to, and use of, assisted reproductive technologies may be of benefit to parents with spina bifida.

### Preconception counselling

Preconception genetic counselling to decrease the risk of neural tube abnormalities in offspring (see *Chapter 2*) is generally well accepted by patients and families prior to the young person becoming sexually active.

This can provide a good opportunity for the clinician to demonstrate and emphasise that they are prepared to discuss sexual health issues at any time.

Specialist spina bifida clinics can refer at risk patients to clinicians experienced in managing pregnancy in women with spina bifida.

### Psychological and social issues

#### Young people with spina bifida and independence

Sexuality is an area where young people with spina bifida often take their first steps of independence from their parents, family and carers.

Family relationships can be disrupted when a young person begins to become sexually active. Close relationships form between parents, carers and children from many years of intimate contact that is required for the successful management of spina bifida. This relationship is often tested at this time of increasing independence as new boundaries become established.

These issues are further complicated because even when adulthood is achieved, some level of dependence may need to remain. Disharmony, disputes and tense relationships are common, and the general practitioner is in an ideal situation to provide support through this time.

#### Ending isolation: peer support and finding partners

Young people and adults often feel isolated when affected by spina bifida. Not only do they feel personally isolated due to their disability, but they may have real issues of physical isolation due to difficulty in independent transport which prevents meeting others and prospective friends and partners.

The increasing move towards incorporating disability care into mainstream organisations has increased this

Impotence is a critical fertility issue

Ejaculation in men with spina bifida

## ■ Chapter 8: Sexuality and reproductive issues

isolation for many. In the past, when people with spina bifida may have been educated together, mutual support and sharing of coping strategies was common.

### **Peer support: spina bifida associations**

Many people with spina bifida find that peer support organisations — such as spina bifida associations — as one of the best ways of overcoming this isolation. These organisations facilitate contact between members through regular newsletters and events, thereby providing opportunities for socialising.

Spina bifida associations are more than just social groups — they create a forum where affected people can offer each other mutual support, identify important common issues which then become the basis for community education and political movements. Some patients become very active in these organisations and find participation a very fulfilling part of their lives.

General practitioners can assist young people and adults with spina bifida deal with issues of social and personal isolation by encouraging them to join their local spina bifida association (see *Chapter 9 Organisations and further resources*).

### **Further resources**

Specialist spina bifida clinics can refer to appropriate sexually counselling clinics if the need arises.

Family planning clinics manage contraception issues for people with disabilities, and are often located close to families.

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## Chapter 9: Organisations and further resources

*Local organisations can put you into contact with specialist centres and other resources in your area.*

### State based Spina Bifida Associations

The spina bifida association in each state is a good first contact point when seeking information about support and services. The associations can assist those affected by spina bifida, their carers and those working with people with spina bifida. Membership is encouraged for all those with and affected by spina bifida.

#### **Spina Bifida Association of NSW**

c/o Northcott Society  
Contact: Mike Sheargold  
PO Box 4055  
Parramatta NSW 2124  
Telephone: 02 9890 0172  
Website: [www.northcott.org.au](http://www.northcott.org.au)  
Email: [mikes@northcott.org.au](mailto:mikes@northcott.org.au)

#### **Spina Bifida and Hydrocephalus Association of SA Inc.**

PO Box 272  
Torrensview Plaza SA 5031  
Telephone: 08 8443 5200  
Fax: 08 8443 5100  
Email: [info@spinabifida.asn.au](mailto:info@spinabifida.asn.au)  
Website: [www.spinabifida.asn.au/](http://www.spinabifida.asn.au/)

#### **SBH Queensland**

21 Tillot St (PO Box 8022)  
Dutton Park QLD 4102  
Telephone: 07 3844 4600  
Fax: 07 3844 4601  
Email: [bshead@spinabifida.org](mailto:bshead@spinabifida.org)  
Website: [www.spinabifida.org](http://www.spinabifida.org)

#### **Spina Bifida Association of Tasmania**

Contact: William Brown  
PO Box 450  
Sandy Bay TAS 7006  
Telephone: 03 6275 0987

#### **Spina Bifida Association of Victoria**

c/o Yooralla Society of Victoria  
705 Geelong Rd  
Brooklyn VIC 3025  
(PO Box 1101, Altona Gate VIC 3025)  
Telephone: 03 9362 6111 or Freecall 1800 686 533  
Fax: 03 9314 9825  
Email [library@yooralla.com.au](mailto:library@yooralla.com.au)  
Website: [www.sbav.org.au](http://www.sbav.org.au)

**Spina Bifida Association of WA Inc.**

37 Hampden Rd  
Nedlands WA 6009  
Telephone: 08 9389 8311  
Fax: 08 9389 8331  
Email: sbawa@swannet.com.au  
Website: www.sbawa.asn.au

**Key Australian spina bifida contact points**

**Australian Spina Bifida and Hydrocephalus Association Inc.**

The Australian Spina Bifida and Hydrocephalus Association Inc. is an incorporated body servicing the interests of state bodies. The state based associations are the best contact point for clients, their families, carers and health workers, but the website contains useful information and links to Australian websites.

Website: www.asbha.org.au

**Spina Bifida Foundation of Victoria**

The Spina Bifida Foundation of Victoria was established to improve the quality of life of the spina bifida community by raising money for and developing a range of activities and services. Activities have a strong focus on promoting independence for people with spina bifida.

Spina Bifida Foundation of Victoria  
PO Box 166  
Parkville VIC 3052  
Telephone: 03 8344 7924  
Website: www.sbav.org.au/foundation.htm

**Assistance for continence aids and funding**

**Many organisations are available to assist with obtaining continence supplies and aids.**

Ask for help when applying for continence aid schemes.

The provision of equipment, disposables and aids for successful continence management can be expensive and many programs are available to assist affected individuals

When applying for assistance for incontinence aids, the number of organisations providing assistance and their roles can be confusing. Social workers, continence nurses and doctors all have experience in applying for the various schemes, and may be asked to help when completing application forms to ensure that no errors are made that could delay payments or applications.

**Continence Aids Assistance Scheme**

The Continence Aids Assistance Scheme (CAAS) is a Commonwealth Government program offering assistance to people who have permanent incontinence due to permanent disability.

The program offers an annual subsidy to approved clients to use their subsidy to order approved continence aids listed on a schedule.

People aged 16 to 64 years of age with permanent incontinence from a permanent disability can apply for CAAS if they are eligible to receive a disability support pension through Centrelink, or a pension from the Department of Veterans' Affairs. CAAS also has other restrictions and eligibility criteria.

For further information, CAAS can be contacted through the company PQ Lifestyles at:

PQ Lifestyles  
PO Box 2082  
Milton QLD 4064  
Telephone: 1300 134 260

or contact:

The Department of Health and Aged Care  
Telephone: 1800 807 487

or write to:

CAAS  
Department of Health and Aged Care  
GPO Box 9848  
Canberra ACT 2601

**State based organisations**

Each State has aids and equipment programs, and some have additional services available to assist with incontinence management.

Health care providers will be able to identify and refer patients to these services, especially specialist continence clinics. In addition, local spina bifida associations will also be able to identify appropriate services and agencies.

**Useful websites**

The following international sites contain useful information on spina bifida and hydrocephalus.

**Spina Bifida Association of America**  
www.sbaa.org

**International Federation for Hydrocephalus and Spina Bifida**  
www.ifglobal.org

**Spina Bifida & Hydrocephalus Association of Ontario**  
<http://www.sbhao.on.ca/>

Other sites of interest include:

**Continence Foundation of Australia**

www.contfound.org.au

**Australian Council of Stoma Associations**

www.australianstoma.org.au

**Specialist spina bifida clinics**

As well as adult spina bifida clinics, paediatric hospitals are listed as they can refer adult patients to suitable facilities in their local area. If there is no specific clinic listed in your area, contact your state spina bifida association for information on local specialist health care providers.

**New South Wales**

Adult Spina Bifida Clinic  
Westmead Hospital  
Department for Rehabilitative Medicine  
Cnr Hawkesbury and Darcey Roads  
Westmead NSW 2145  
Telephone: 02 9845 7800

Spina Bifida Clinic (children)  
Sydney Children's Hospital  
High Street  
Randwick NSW 2031  
Telephone: 02 9382 1589

Spina Bifida Unit (children)  
The New Children's Hospital at Westmead  
Locked Bag 4001  
Westmead NSW 2145  
Telephone: 02 9845 2769

**Northern Territory**

Alice Springs — contact Spina Bifida Association of South Australia  
Other places — contact SBH, Queensland

**Queensland**

Princess Alexandra Hospital  
Spina Bifida Clinic for Adults  
Cornwall St  
Woolloongabba QLD 4102  
Telephone: 07 3240 2641

Mater Children's Hospital (children)  
Spina Bifida Clinic  
Raymond Terrace  
South Brisbane QLD 4101  
Telephone: 07 3840 1812

Royal Children's Hospital (children)  
Spina Bifida Clinic  
Herston Road  
Herston 4029  
Telephone: 07 3636 7818

Spina Bifida Clinic Paediatrics Outpatients (children)  
Gold Coast Hospital  
Telephone: 07 5571 8345  
Nerang Street  
Southport QLD 4215

Townsville Children's Community Therapy Services (Children)  
Spina Bifida Clinic  
Telephone: 07 4728 2681

**South Australia**

Royal Adelaide Hospital (adult)  
North Terrace  
Adelaide SA 5000  
Telephone: 08 8222 4000

Flinders Medical Centre (adult)  
Flinders Drive  
Bedford Park SA 5025  
Telephone: 08 8204 5511

Queen Elizabeth Hospital (adult)  
28 Woodville Road  
Woodville South SA 5011  
Telephone: 08 8222 6000

Continence Clinic (adult)  
Repatriation General Hospital  
Daws Road  
Daw Park SA 5041  
Telephone: 08 8275 1927

Adelaide Women's and Children's Hospital (children)  
King William Road  
North Adelaide SA 5006  
Telephone: 08 8161 7000

**Tasmania**

Contact State Spina Bifida Association

### Victoria

Adult Spina Bifida Clinic  
c/o Dr Churchyard's Secretary  
Neuropsychiatry  
Monash Medical Centre  
Clayton Road  
Clayton VIC 3168  
Telephone: 03 9594 2240

Continence Clinic  
Royal Children's Hospital  
Flemington Road  
Parkville VIC 3052  
Telephone: 03 9345 5522

### Western Australia

Spinal Rehabilitation Clinic (children)  
Princess Margaret Hospital for Children  
Thomas Street  
Subiaco WA 6008  
Telephone: 08 9340 8886

### Continence organisations

#### National Continence Helpline

The National Continence Helpline is a joint project of the Commonwealth Government and the Continence Foundation of Australia. It aims to assist people with incontinence, their families and carers, as well as health professionals. The help line can refer patients to local services.

Telephone: 1800 330 066

More help can be gained by contacting the Continence Foundation of Australia at:

Continence Foundation of Australia Ltd  
AMA House  
293 Royal Parade  
Parkville VIC 3052  
Telephone: 61 3 9347 2522  
Fax: 61 3 9347 2533  
URL: <http://www.contfound.org.au>

### Ostomy Associations

The following associations may be of use to patients for stoma care. Colostomy Associations can also help with other stoma besides just colostomies. If no ostomy association is listed close to your area, please contact your state spina bifida association for local ostomy health care providers and services.

#### Australian Capital Territory.

ACT & Districts Stoma Association Inc.  
2nd Floor, Department of Health Building  
Cnr Moore & Alinga Streets, Canberra City 2601  
PO Box 1260, Canberra City, ACT, 2601  
Telephone and fax: 02 6205 1055

#### New South Wales

Colostomy Association. of NSW Inc.  
Unit 5/7-29 Bridge Road  
Stanmore NSW 2048  
Telephone: 02 9565 4315

Ileostomy Association. of NSW Ltd  
Block E, Ozanam Village  
West St, Lewisham NSW 2049  
Telephone: 02 9568 2799

#### Queensland

Gold Coast Ostomy Association Inc.  
8 Dunkirk Close  
Arundel QLD 4214  
Telephone: 07 5594 7633  
Website: [www.ostomy.org.au](http://www.ostomy.org.au)

North Queensland Ostomy Association Inc.  
Shop 4,  
52 French Street  
Pimlico QLD 2478  
Telephone: 07 4775 2303  
Website : [www.ostomy.org.au](http://www.ostomy.org.au)  
All correspondence and orders to:  
PO Box 1017  
Hyde Park  
Castletown QLD 4812

Queensland Colostomy Association Inc.  
22 Beaudesert Road  
Moorooka QLD 4105  
Telephone: 07 3848 7178  
Email: [colostomy@primus.com.au](mailto:colostomy@primus.com.au)  
Website : [www.ostomy.org.au](http://www.ostomy.org.au)

Queensland Stoma Association Inc.  
Unit 4, 'Accent Place', 10 Valente Close, Chermside 4032  
PO Box 370, Chermside South, QLD, 4032  
Telephone: 07 3359 7570  
Emergency Telephone: 07 3359 7570  
Website : www.qldstoma.asn.au

Toowoomba & South West Ostomy Association Inc.  
c/o Epworth Nursing Home  
Stenner Street  
Toowoomba QLD 4350  
Mail address: P.O. Box 7314, Toowoomba M.C., 4352  
Telephone: 07 4636 9701  
Website : www.ostomy.org.au

Wide Bay Ostomy Association Inc.  
c/o Bundaberg Base Hospital  
Bourbong Street  
Bundaberg QLD 4670  
Telephone: 07 4150 2074  
Website : www.ostomy.org.au

#### South Australia

Colostomy Association of SA (C.A.S.A.)  
160 South Road  
Torrensville SA 5031  
Telephone: 08 8354 2618

Ileostomy Association SA Inc.  
1 Shipster Street  
Torrensville Adelaide SA 5031  
Telephone: 08 8234 2678

#### Tasmania

Ileostomy & Colostomy Association (TAS) Inc.  
Statton Bldg, Repatriation Centre  
Hampden Road  
Hobart TAS 7000  
PO Box 415, Sandy Bay, Tas, 7006  
Telephone: 03 6223 2974

#### Victoria

Ileostomy Association (VIC) Inc.  
Suite 322, 3rd Floor, The Block  
98-100 Elizabeth Street  
Melbourne, VIC 3000  
Telephone: 03 9650 9040

Ostomy Association of Melbourne Inc.  
Cabrini Medical Centre  
Isabella Street  
Malvern VIC 3144  
Telephone: 03 9508 1879

#### Western Australia

WA Ostomy Association Inc.  
15 Guildford Road  
Mount Lawley WA 6050  
Telephone: 08 9272 1833

#### Further reading for patients

Information on the following publications is available from either the Spina Bifida Association in your state, or by contacting :

Library and Information Service  
Yooralla Society of Victoria  
705 Geelong Road Brooklyn VIC 3025  
Telephone: 03 9362 6143  
Email: library@yooralla.com.au

#### **Spina bifida and hydrocephalus explained. Spina Bifida Association of Victoria.**

A booklet explaining the basics of spina bifida and hydrocephalus in plain language.

#### **Spinabilities. A young person's guide to spina bifida by Lutkenhoff M, Oppenheimer SG. Woodbine house, 1997.**

A young person's guide to all aspects of spina bifida including incontinence and sexuality. Excellent resource for teenagers with spina bifida.

#### **Hoath L, Gilbert S. Oops. A common sense approach to toilet training a child who has a 'problem'.**

This excellent plain language guide covers all aspects of the management of urinary and faecal incontinence. Although written for children and young people, it contains much information of use for the adult with spina bifida.

#### **Nolan T. Fitness training diary. Royal Children's Hospital. 1996.**

This resource contains information and a diary to help manage faecal incontinence and is available from the Royal Children's Hospital, Melbourne.



## Chapter 10: References

*While specific references are mentioned in the text, the following is a list of resources consulted during the preparation of this manual.*

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