

# NSW Guidelines for Management of Adults with Spina Bifida

*'If I lived in a society where being in a wheelchair was no more remarkable than wearing glasses, and if the community was accepting and accessible, my disability would be an inconvenience and not much more than that. It is society which handicaps me, far more seriously and completely than the fact that I have Spina Bifida'. [1]*

This document is a general guide to appropriate practice to be followed only subject to the clinicians' expert judgement in each individual case. The guidelines are designed to provide information to assist decision making and are based on the best information available at the date of publication

## **Acknowledgements**

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Joanne Brady, Clinical Nurse Consultant, Spina Bifida Adult Resource Team, Northcott

Jade Baynes, Occupational Therapist, Spina Bifida Adult Resource Team, Northcott

Dr Carolyn West, Adult Rehabilitation Specialist, Royal Prince Alfred Hospital

Julie Dicker, Spina Bifida Clinical Nurse Consultant, Children's Hospital Westmead

Lynne Brodie, ACI Transition Care Network Manager

## **Reviewers**

- NSW Statewide Spina Bifida Collaborative
- Policy, Technology and Support Unit (PTSU)
- State-wide Services Development Branch NSW Health
- Northcott Disability Services

# **Executive Summary**

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## Aims, Context and Scope of this model

This document aims to guide the holistic health management of adults with Spina Bifida throughout NSW. While specifically focusing on health, the context and scope of the guidelines includes related aspects that impact on health such as supported living arrangements, opportunities for study, work, socialisation and recreation.

The guidelines provide an outline for a range of evidence based interventions for management of health issues for people with Spina Bifida that have shown to be effective within the Australian context. The intention is that each Local Health Network and Primary Care Organisation across NSW will be able to review, select and implement locally appropriate service changes to ensure that health issues are addressed for all adults with Spina Bifida living in metropolitan, rural, regional or remote areas of the State.

The population to be targeted is young people and adults aged 18 years or older. The guidelines are applicable in all settings where these people are managed that best meet the local context such as outpatient departments, community health centres, general practice, private rooms or relevant other government or non-government organisations.

# Background to Development of the Guidelines

## **Incidence, morbidity and complications associated with Spina Bifida**

Spina bifida is a neural tube defect and is the most common, complex, congenital condition affecting the development of the brain and the spinal cord in developed countries. A neural tube defect is an opening in the spinal cord or brain that occurs very early in human development. Normally, the neural tube closes during the fourth week of pregnancy, and develops into the brain, spinal cord, and back bones. When the neural tube does not close completely, a neural tube defect develops. In Australia approximately 1:500 pregnancies are affected with a neural tube defect.

There are two types of neural tube defects: open, which are more common, and closed. Open neural tube defects occur when the brain and/or spinal cord are exposed at birth through a defect in the skull or vertebrae (back bones). Examples of open neural tube defects are anencephaly, encephalocele and spina bifida. Closed neural tube defects occur when the spinal defect is covered by skin. Common examples of closed neural tube defects are lipomyelomeningocele, lipomeningocele, and tethered cord. Despite differences in primary care, both open and closed neural defects do share similarities in secondary care issues such as tethered cords, bowel and bladder dysfunction and mobility issues. However it is the adults with the most common type of Spina Bifida that will present to hospitals across NSW.

Spina Bifida, which is derived from the Latin term meaning “split spine”, is a form of neural tube defect that occurs when there is incomplete development of the spinal cord, the bones forming the spinal column (vertebrae), and often the overlying skin. In spina bifida the bones of the baby’s spine do not close over the spinal cord properly and parts of the spinal cord may be exposed. This can happen anywhere along the length of the spine and the location will determine how the child is affected.

There are different types of spina bifida. Myelomeningocele is the most common and serious type of spina Bifida and is generally what people are referring to when they talk about spina bifida.

These patients have multifaceted health care needs and disability, resulting in paresis or paralysis limiting mobility, sensory deficits in the lower limbs causing pressure areas, burns and fractures, hydrocephalus requiring a shunt and compromising cognition and complex spinal complications. Neurogenic bladder and bowel present in up to 95% of patients with spina bifida [3] and incontinence of bladder and bowel is a major management issue.

Australian morbidity data from 2007 described the first cohort of adult patients surviving childhood with complex spina bifida and hydrocephalus requiring shunting, and who were above 18yrs of age. Of 29 known deaths over a period of 25 years, 33% were from chronic renal failure, 14% died from pressure area associated sepsis, 10.4% from chronic respiratory failure and 7% from acute shunt dysfunction (Personal communication, Dr Carolyn West, Head, Spina Bifida Services, The Children's Hospital at Westmead, Sydney, Australia).

## **NSW DATA PRIOR TO ESTABLISHMENT OF THE SPINA BIFIDA ADULT RESOURCE TEAM (SBART) IN 2009.**

### **The GMCT Transition Workforce Study:**

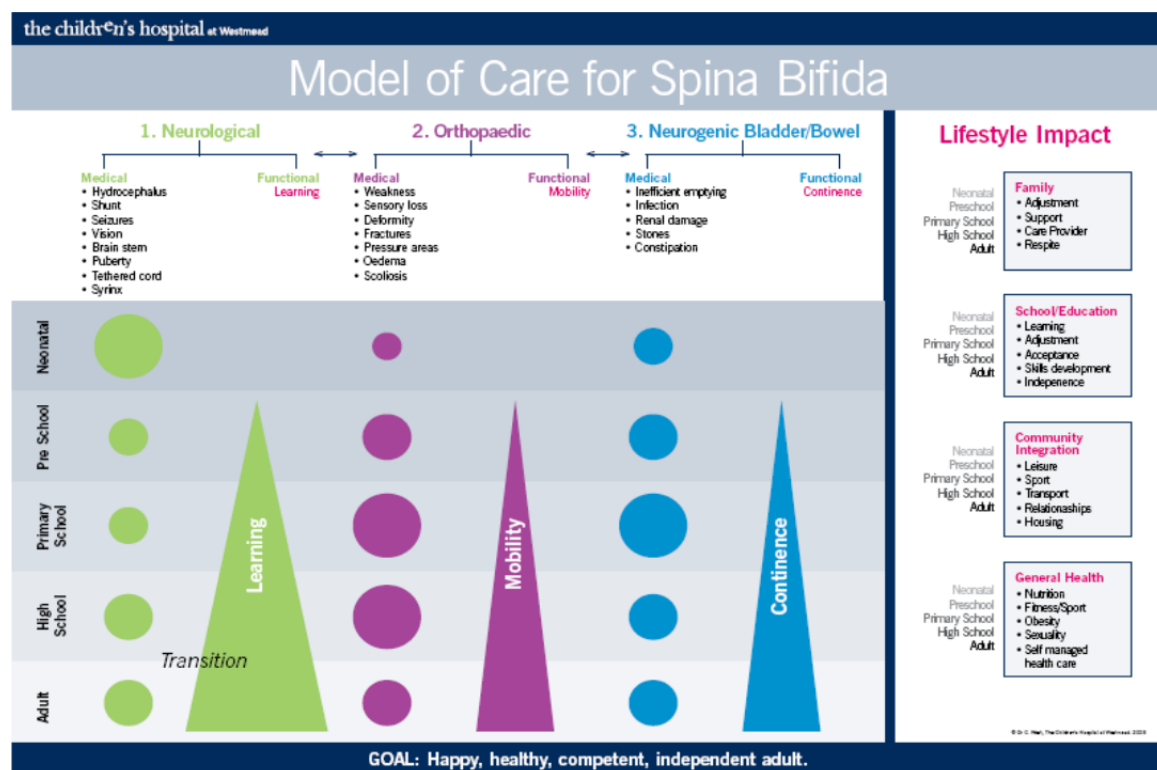
In 2007, 288 patients aged 12-18 years were registered with the three tertiary paediatric spina bifida clinics in NSW. The largest clinic had 195 registered, but of these 50 were considered lost to follow up because they had not been in touch with the service in any manner and were unable to be contacted. The remaining 238 patients aged 12-18 years attended a paediatric service at least once during 2007. An additional 31 patients over 18 years also attended paediatric clinics in the same year. Seventeen young people were transitioned to adult care in the 12 months. There were 549 patients aged 16-24 years registered with adult clinics, although only 40% attended appointments in the year of the data collection and none of these were less than 16 years.

In 2007 there were 740 inpatient admissions in NSW for young people aged 16-24 with spina bifida, with an average length of stay of five days. The commonest causes

for admission were chronic renal failure, pressure areas/sepsis, chronic respiratory failure and acute shunt dysfunction. Anecdotal evidence suggests that many of these admissions were unplanned through the emergency departments. The AIHW reported in 2006 (from figures obtained 2004-2005) that the average cost to the health system per patient separation was \$AUD3410.00 (with an estimated per annum increase of nearly 5%) [16]. Annual spina bifida admission costs conservatively were thus estimated at over \$AUD 2.5 million.

## Model of Care for Spina Bifida

Adapted from the Children's Hospital Westmead, this model of care highlights the medical and life style impacts of spina bifida.



## The Need for Change – Patient Journeys

The following two case studies highlight the difficulties faced by young people in negotiating the health system.

### “Mitchell” Case Study

## What is the problem?

Young man with severe spina bifida fell while transferring from his wheelchair and was taken by ambulance to a Northern Sydney Hospital in June 2010:

### Why did he present?

Mitchell had been diagnosed mid June with a UTI and was feeling very ill. He lives independently in Dept of Housing accommodation. Transferring from his bed to her wheelchair, he slipped, couldn't move himself and was in severe pain so he called an ambulance and was taken to her local ED.

June to July 2010

### What was the diagnosis?

He spent the night in ED, had X rays of his bruised face and neck which showed no fractures. The diagnosis was bruising following fall + pre-existing UTI.

Spent very uncomfortable 24 hrs in ED on "hard bed".

Transferred to ward after 24 hrs and pressure mattress organised

Mitchell spent a month in hospital before being transferred to a rehab bed "to build up his strength". Discharged on 19/7/2010



## History continued

### What happened after discharge?

Mitchell's GP requested that he return to Hospital for additional rehab to help his wheelchair transfers and also for dietary review.

A routine colonoscopy was performed at the request of the GP and he was discharged 2 days later.

August to October 2010

In August Mitchell was returning to his home in a taxi when he felt severe neck pain. He asked the taxi driver to take him to ED. According to her mother "he was given painkillers (propenol) was discharged and told to take 2 now and the rest later."

It is unclear what happened next. It seems Mitchell took more tablets and was found in a confused state by the home care attendants on Monday at 5pm. Mitchell was incoherent and rolling his eyes. They called her parents who told them to call 000

Has now spent nearly 7 weeks in hospital

Mitchell taken by ambulance to ED





## History (cont)

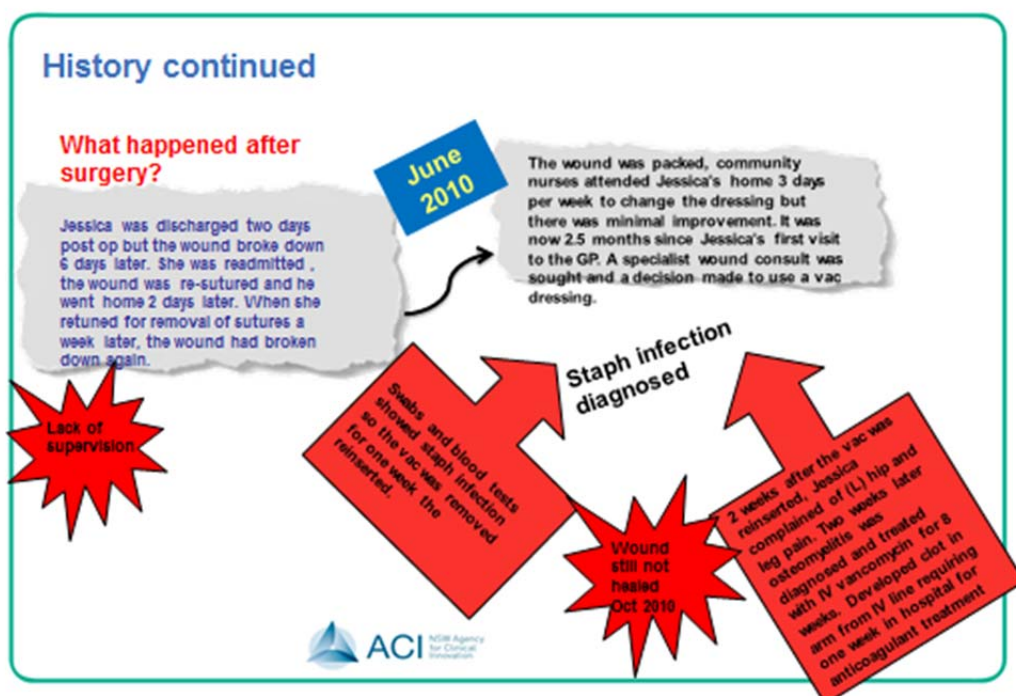
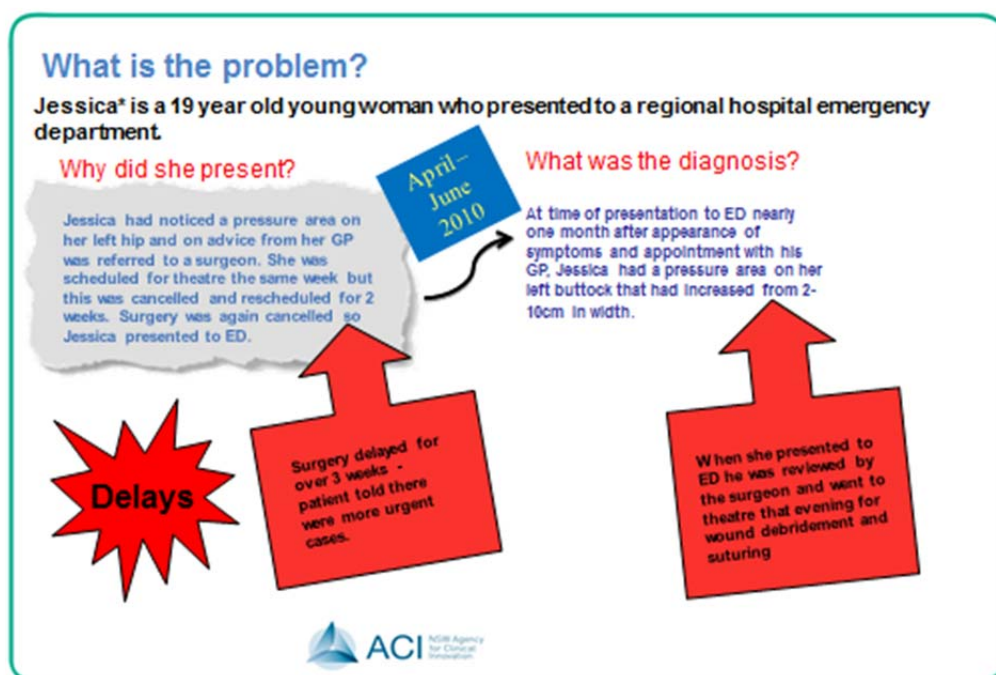
CT scan showed hairline fracture of C1. Confirmed on MRI and neck brace "fitted" which restricted eating and speech. **Treatment for UTI commenced.** Moved to ICU....parents told it was for more intensive treatment of the UTI.

Team meeting, decision made to move to another Sydney hospital and he was transferred September 2010 for assessment of his neck. Spent 6.5 hours in ED on **hard mattress** and developed sacral pressure area.

Transferred back to original hospital and eventually moved to rehabilitation ward.



## “Jessica” Case Study



## A: Jessica - Complete Story

Jessica is a 19 year old young woman with spina bifida who lives in regional NSW. She has a VP shunt due to hydrocephalus which impacts on her executive functioning skills but she is otherwise a bright young woman who prior to this incident, was able to walk.

In mid April 2010 she presented to her GP with a pressure sore on her left buttock. Her GP referred her to a local surgeon. She was seen by the surgeon within a week and scheduled for surgery the same week. However surgery was cancelled and re-scheduled for 2 weeks and then cancelled again for a further 2 weeks. The reason given to Jessica was that there were "more urgent cases."

Jessica then presented to ED as she was concerned about her wound. After assessment by the triage nurse her surgeon was informed and reviewed Jessica. According to Jessica "the surgeon was shocked when he saw the wound had gone from 2cm to 10cm wide."

Surgery was performed that evening to debride and close the wound and Jessica was discharged after 2 days. Six days later she re-presented to ED with wound breakdown and was readmitted. The wound was re-sutured and she was discharged two days later. One week later she returned to the surgeon's rooms for removal of the stitches but was informed that the wound had again broken down and she needed to return to hospital. This time the wound was packed and Jessica was discharged to the care of Capac (community nursing service) who visited every 3 days to attend to the dressings.

At approximately 2.5 months after initially presenting to her GP, the wound was still far from being healed and a decision was made to use a vac dressing. One month later it was obvious that the wound was infected and bloods and a wound swab were done. Staph was diagnosed and the vac was removed for a week according to Jessica "to rest the wound". The vac was then reinserted.



## A: Jessica - Complete Story (cont)

Two weeks later Jessica experienced left hip and leg pain. She was told by the nurses that it was due to the wound and related to his disability. Jessica refuted this but according to her she stated "I was continually ignored".

She then contacted the team leader of the State Wide Adult Spina Bifida team in Sydney (SBART) who spoke to the rehab/wound CNC in the regional area. She liaised with Jessica's team who organised blood tests and a scan of the affected area.

Ostomyitis was diagnosed and he was re-admitted. A pic line was inserted and Vancomycin commenced. Jessica was discharged and community nurses visited to check the wound and IV line. After several days Jessica complained of soreness in her arm and on review by the specialist, the arm was found to be red and painful and arrangements were made to insert a new line. This required transfer to another hospital as the first attempt to replace the line was unsuccessful. On removal of the initial line an extensive clot was discovered that extended the length of the line.

Jessica was commenced on warfarin and several other medications to treat the clot and infection. She spent a week in hospital and experienced incontinence as a result of the medications which caused her embarrassment. On one occasion she suffered the indignity of being left in a soiled bed overnight despite requests to staff to help her.

Her future is uncertain. More surgery may be required and she remains off work, socially isolated and is now likely to require long term use of a wheelchair when she was previously ambulant.



# Common Medical Conditions in Adults with Spina Bifida

## **ORTHOPAEDIC ISSUES**

### **Sensory function**

Sensory feedback is essential for a number of functional activities. The level of sensory impairment is usually similar to the motor level. All modalities of sensation are affected in spina bifida. Sensory feedback from the lower limbs is important for mobility and protecting the lower limbs from harm. Adults with spina bifida do not have these warning symptoms in their lower limbs. Fractures, pressure areas, burns, chilblains and other trauma are more common.

### **Fractures**

Lower limbs in adults with spina bifida are osteopaenic due to reduced weight bearing resulting in an increased risk of fractures. Fractures can be missed because there is no report of pain. X-ray is indicated if swelling and/or deformity is present. Other causes of a swollen leg are cellulitis, DVT or lymphedema.

Upright mobility, adequate calcium in diet and sunlight exposure for vitamin D is useful in maintaining bone health. In some adults calcium/vitamin D supplementation may be necessary. Bisphosphonates may be indicated if recurrent fractures occur.

### **Neuropathic (Charcot's) joint**

Repeated trauma to a joint without sensation can cause deformity and further limit mobility.

### **Circulation**

Inadequate circulation in the lower limbs can result in swelling, poor healing, skin breakdown and chilblains. Management includes elevation of the lower limbs, warm clothing and the use of support stockings.

## NEUROLOGICAL ASPECTS OF SPINA BIFIDA

### **Arnold Chiari Malformation and Hydrocephalus**

This malformation includes a small posterior fossa, descent of the cerebellar tonsils and brain stem into the spinal canal and partial obstruction of the foramina of Lushca and Magendie. This frequently leads to slow circulation of the CSF because of the obstruction of the foramina resulting in ventriculomegaly either during the pregnancy or following surgical closure of the myelomeningocele after delivery.

Surgical treatment is often required for ventriculomegaly as it usually results in raised intercranial pressure, an enlarged head and compromised brain development.

Treatment involves surgical insertion of a shunt to divert to CSF to another body cavity, usually the peritoneal cavity. The shunt is usually located in the right side and is called a ventriculoperitoneal (V/P) shunt

Hydrocephalus may also be treated by an endoscopic third ventriculostomy.

Shunt function is monitored by clinical symptoms such as lethargy, headache, nausea, vomiting, blurred vision or neck pain. Level of consciousness, cranial nerve signs, papilloedema, discontinuity of shunt tubing are signs. Imaging such as ultrasound, CT scan or MRI will confirm enlarging ventricles.

Ventriculomegaly on imaging can assist with diagnosis, however some adults will have dysfunction despite minimal changes in ventricular size. Further investigations maybe required such as pressure monitoring or isotope shunt studies. A blocked shunt should always be considered with a symptomatic patient. A blocked shunt can lead to compression of the brainstem, coning and death.

### **Tethered cord**

Adults with myelomeningocele have a low lying spinal cord. The end of the spinal cord is usually located at the lesion and may be fixed in this position by fatty or scar tissue. During growth the spinal cord can be put on stretch. Symptoms can include lower back pain, worse during sitting and relieved by changing position with limited

relief from analgesics, deterioration of motor or sensory function in lower limbs, or otherwise unexplained change in continence or sexual functioning. Only a few adults will develop these symptoms. It is important to recognise these adults as surgical treatment can assist in preserving function. Diagnosis is clinical – change in neurological symptoms and signs from usual baseline status. Treatment is surgical release of the tethered cord by a neurosurgeon. Diagnosis and surgical treatment can preserve function and prevent further deterioration. There may be some return of recently lost function if the patient presents soon after the onset of symptoms. Tethered cord may present at any age but is more common during rapidly growing years.

### **Syringomyelia**

Syringomyelia occurs when CSF collects in small pockets within the spinal cord due to abnormal circulation of CSF fluid. Due to the abnormal circulation of this fluid, it can collect in small pockets called a syringomyelia. As it enlarges the syringomyelia can press on the cells of the spinal cord and compromise function.

Symptoms can include deterioration in motor or sensory function in lower limbs, continence issues, scoliosis and upper limb dysfunction if located in the cervical area. . Treatment includes posterior fossa decompression, draining or shunting the syrinx. Despite surgical intervention, Syringomyelia can result in slow neurological decline over years presumably due to atrophy of the spinal cord.

## **NEUROGENIC BLADDER AND BOWEL**

### **Assessing the neurogenic bladder**

Clinically in adults there will be incontinence, urine infections and excoriated skin. During adulthood each adult should have a renal ultrasound annually. If the adult's management is stable and the ultrasound is normal then this may be sufficient investigation. However if the adult has a high pressure bladder further monitoring is required to adjust management to maximise continence and prevent renal damage.



## **Management - clean intermittent catheterisation (CIC)**

This procedure is carried out with a small disposable plastic catheter which is inserted into the bladder 3-4 times per day to empty the bladder completely. Adults are usually taught in their childhood on how to do this in a clean and hygienic fashion however it is not a sterile procedure. Adults will often need to be retrained in this procedure as over time they become lazy, their technique is poor which leads to ongoing urine infections.

CIC reduces pressures in bladder, reduces infection by more efficient emptying and improves continence. This is the management of choice in over half the patients with a neurogenic bladder.

Anticholinergics are also used to relax the detrusor muscle, reduce bladder pressures, increase bladder capacity and improve continence.

## **SURGICAL INTERVENTIONS**

### **Suprapubic catheter**

For an acute obstruction, urethral stricture or CIC failure a supra pubic catheter can be used. It can be used permanently when other methods are not practical. Supra pubic catheters need to be changed every 6 weeks. Complications may include infection, calculi and blocking of the catheter.

### **Mitrofanoff procedure**

This involves surgically creating an alternate catheterisable channel using the appendix (or small bowel). One end of the appendix is attached to the anterior wall of the bladder and the other end to the abdominal wall near the umbilicus as a catheterisable stoma.

This operation is suitable in the very disabled patient who has problems accessing the urethra for self- catheterisation because of difficulty with balance, transfers to toilet or poor co-ordination. It may enable the patient to self- catheterise in the

wheelchair without the help of a carer and hence improve independence. Patients with urethral stricture may also be considered for this procedure.

### **Sphincterotomy**

Can facilitate bladder emptying however the patient will be wet from incontinence at all times. They can be useful in the older male patient who can use a penile appliance to drain the urine into a leg bag.

### **Bladder augmentation**

A surgical procedure to enlarge the bladder using a segment of the bowel, It is used in high pressure bladders where despite appropriate management renal damage is occurring. Bladder augmentation enlarges bladder capacity, reduces detrusor contractions, increases compliance of the bladder and improves continence.

Complications include mucous formation (clogging the catheter, increasing infection), bladder rupture if patient forgets to catheterize and possible metabolic acidosis and/or longer term vitamin B12 and folate deficiency from reduced small bowel absorption. Metaplasia of the augmented bladder is a risk requiring annual cystoscopy from 10 years after surgery.

### **Ileal conduit**

The ureters are divided from the bladder and diverted into an isolated loop of bowel which acts as a conduit to an abdominal stoma where the urine can drain into a bag. This protects the kidneys from high bladder pressures and provides a method of managing the incontinence.

There can be long term problems with conduits including strictures, calculi, conduit enlargement and stomal stenosis or prolapse. Ileal conduits are still occasionally used to manage incontinence or prevent further renal damage.

### **Provision of incontinence equipment**

Ongoing supplies of continence equipment are expensive. There are various government assistance schemes available such as Continence Aids Payment



Scheme or ENABLE. Contact your local continence nurse or the Spina Bifida Adult Resource Team for further continence management.

### **Bowel care**

Adults with spina bifida have poor lower bowel mobility, abnormal anal tone and sensation. These factors all contribute to incontinence.

Management includes:

1. Firm to soft stool consistency. Consider dietary modification, stool softeners or combined agents such as Movicol. Regular medication is best for most patients.
2. Regular routine of bowel emptying. Patients may require a suppository or micro enema to initiate adequate emptying.

A few patients are not well managed by these simple methods and may need large volume washouts per rectum or antegrade washouts via a caecostomy button.

## Prevention of Common Complications

- **Shunt malfunction** – difficult to prevent but clients need to be taught from an early age the signs and symptoms of shunt malfunction and to seek medical help early.
- **Urinary tract infection** – Infections are more common in the patient with a neurogenic bladder. Causes are inadequate bladder emptying, introduction via catheter, poor hygiene with catheters, calculi, deteriorating bladder function e.g. high pressure. For those on intermittent catheterisation treat symptomatic infections only. As catheterisation is clean rather than sterile most patients on intermittent catheterisation will have organisms in the urine. Treating these organisms if an adult is asymptomatic can often lead to resistant organisms. If recurrent UTIs check catheterisation technique. All patients should have annual renal ultrasound, serum creatinine and urea and check blood pressure.
- **Bowel problems** – avoid constipation by healthy diet and regular bowel emptying daily or second daily [see above]
- **Skin problems and decubitus ulcers** – client needs to check skin daily for skin ulceration after showering, maintain continence to stay clean and dry. Visit a podiatrist regularly for foot care. Wear low compression below knee stockings and massage legs daily to prevent lymphoedema to lower limbs. Spend some time during the day out of the wheelchair [with legs elevated if possible] to relieve pressure and to help blood circulation. In winter avoid chilblains by wearing warm socks and massage legs prior to showering.

## Management of Common Complications

- **Shunt Malfunction** – regular medical visits to GP and seek medical attention if signs of shunt not working properly e.g. recurrent headaches and vomiting.
- **Cord tethering** – only relevant for those with good mobility. See neurosurgeon if walking changes or sensation in lower limbs changes.[see above]
- **Urinary tract infection** – following correct CIC procedure and only treat symptomatic bacteraemia.
- **Bowel problems** – correct constipation through diet and/or laxatives like “Movicol” and daily elimination using suppository or enema.
- **Skin disorders and decubitus ulcers** – treat skin ulceration immediately, seek medical advice and remove pressure to aid healing. Visit podiatrist regularly for foot care. Utilise an occupational therapist for pressure problems related to a client’s bed, seating or wheelchair.

# Ideal Patient Journey



## Hospital Management Plan for young people and adults with Spina Bifida

### DEFINITION:

Spina Bifida, which is derived from the Latin term meaning 'split spine' is a form of neural tube defect that occurs when there is incomplete development of the spinal cord, the bones forming the spinal column (vertebrae) and often the overlying skin.

### FUNCTIONAL DEFICITS:

Spina Bifida frequently results in paralysis or paresis of the lower limbs, neurogenic bladder and bowel and some cognitive compromise. The disability issues include impaired walking, incontinence of bladder and bowel and learning issues particularly related to executive functioning. There is a range of disability from very mild to severe depending on the level of the lesion and the neurological complications.

SYMPTOMS					
PERSISTENT HEADACHES Presents with:	PERSISTENT HEADACHES Presents with:	ABDOMINAL PAIN	SKIN BREAKDOWN	LOWER LIMB SWELLING	OTHER CONSIDERATIONS
<ul style="list-style-type: none"> <li>Nausea and vomiting</li> <li>Decreased level consciousness</li> <li>Burred Vision</li> </ul>	Urinary Tract Infection (UTI) <b>NOTE:</b> Organisms in the urine are common in patients doing clean intermittent self catheterisations (CISC). No treatment is indicated unless there are systemic symptoms.	Faecal Loading UTI, renal calculi Shunt Dysfunction Other including: oesophageal reflux, gall stones, appendicitis and bladder rupture (if bladder augmentation)	Pressure areas Lymphoedema Cellulitis Burns <b>NOTE:</b> Most pressure areas are of a result of poor pressure care. Ensure that the adult is referred to an Occupational Therapist for follow up and review of their pressure care equipment.	Fracture Lymphoedema Cellulitis Deep Ven Thrombosis Other including: chilblains, eczema and poor circulation	When treating the acute medical problem other aspects of care need to be considered.
Shunt Dysfunction—needs a neurosurgical review					<b>DOES THE PATIENT NEED ASSISTANCE WITH MOBILITY?</b> <ul style="list-style-type: none"> <li>walking frame / crutches / wheelchair</li> <li>assistance with transfers</li> <li>consider referring to rehabilitation team for further assessment</li> </ul> <b>DOES THE PATIENT NEED PRESSURE AREA CARE?</b> <ul style="list-style-type: none"> <li>air mattress (ideal for when inpatients as help with protection against pressure areas on sacrum, legs and heels)</li> <li>as an inpatient consider all chairs that client is sitting on. Wheelchair cushions can be used as pressure relief on other chairs</li> </ul> <b>DOES PATIENT NEED DVT PROPHYLAXIS?</b>
					<b>DOES PATIENT NEED ASSISTANCE WITH CONTINENCE CARE?</b> <ul style="list-style-type: none"> <li>clean intermittent catheterisation</li> <li>enema/bowel without</li> </ul> <b>NOTE:</b> If an inpatient and is confined to hospital bed or has intravenous drip and/or fluids running may also need help with personal care.
					<b>WHAT MEDICATION DOES THE PATIENT TAKE?</b>
CONSIDERATIONS FOR TREATMENT					
					When considering discharge planning ensure that: <ul style="list-style-type: none"> <li>Adults are assessed to determine how they will cope once discharged from hospital. <b>Attention should be given to their mobility and personal/domestic care needs.</b></li> <li>All adults should have their discharge summary sent to their GP and any relevant specialist</li> <li>The Spina Bifida Adult Resource team is contacted to provide support to the adult in the community and ensure they are followed up to prevent further hospital admissions</li> </ul>

For any further information please contact the Spina Bifida Adult Resource Team on 94725214 or 94725215

# Cognitive Functioning in Spina Bifida

## **Assessment**

Adults with spina bifida often have an IQ in the average to low range. Cognitive deficits can be seen in the area of executive functioning. Executive functioning includes goal directed behaviour, problem solving, flexibility, organising and planning. These skills are important for independence in adulthood. Executive functioning skills are required to manage the personal care and health issues related to spina bifida, such as ordering equipment and catheter supplies. Structure and routine will also assist in managing executive functioning deficits in both the home and workplace.

Neuropsychological assessment is useful in identifying cognitive difficulties and setting realistic goals. Assessment should occur in adolescence when cognitive abilities are significantly developed.

Cognitive difficulties and their implications for independent living should also be considered when attempting discharge planning.

## **Anxiety and depression**

Managing the challenges of adult life is stressful for people with spina bifida. Issues with cognition lead to difficulties with independence, employment and relationships.

Depression can result in a disinterest in personal care leading to medical complications such as urinary tract infections and pressure areas. Psychological assessment may be required. Social support can assist in improving life skills and building social networks.

## Transition to Adult Services

Adult health services, in contrast with the developmental, family centred approach of paediatric services, are frequently problem orientated, individual patient focused and may not be aware of the specific needs of adults with spina bifida. There are a few adult physicians who specialise in the care of adults with spina bifida and adult spina bifida clinics are in Sydney metropolitan areas. Access to specialist services for rural and regional patients becomes difficult and expensive.

As with many chronic childhood disabilities, it is harder for spina bifida youth to achieve autonomy from parents or carers [5,6,7]. Parents and carers tend to continue responsibility for the daily activities of living rather than act as facilitators to independence. These learned dependencies reinforced from an early age, require high levels of assessment and interventions in self-management training and support to enhance self-management and achieve behaviour change [8]. There is evidence that young people with spina bifida are significantly disadvantaged in all areas of life studied, and suffer much higher levels of discrimination, social exclusion and isolation than their able bodied peers. [ 9,10]. Adverse health outcomes contribute significantly to their inability to find work, form relationships and live independently [11]. Health care professionals involved with young people with spina bifida in NSW, and like many around the world, have long recognised the need for health support services for this group to maximise health care, reduce hospitalisation and enable access to appropriate equipment.

## Formation of Spina Bifida Adult Resource Team (SBART)

After negotiations and discussions around the specific needs of young adults with spina bifida, the Spina Bifida Adult Resource Team (SBART) in NSW was funded by NSW Health in 2008. The major identified needs were 1) the cognitive difficulties inherent to the condition which interfere with self-management, 2) the increased personal health care costs when young people leave the heavily subsidised paediatric system and 3) pathways to avoid avoidable acute hospitalisations. The principles of the SBART proposal were 1) affordable easy access to a specialist multidisciplinary team, 2) continuity of care with close collaboration between paediatric and adult health services during the transition period and 3) a case manager/ co-ordinator to liaise with young people and their families, to provide support and education to help them to remain engaged with appropriate health services. In practical terms the SBART team has a complex brief which includes numerous responsibilities as summarised in table 2.

Table 2

<b>Responsibilities of the SBART team</b>	
i)	facilitating access to general practitioners, spina bifida clinics, specialists (including urologists and neurosurgeons)
ii)	improving the young person's ability to access equipment provision and equipment maintenance (wheelchairs, pressure cushions, toileting aids, continence equipment, modification to the home and work place, post-surgery equipment needs)
iii)	assisting in the adaptation to changes in life situation (pregnancy, parenthood, deterioration in mobility, and change in accommodation status and support services where elderly parents become unable to provide the level of necessary care).
iv)	playing an educative role and providing information specific to the needs of patients with spina bifida to health professionals in more generic services who may be able to provide the care closer to home.
v)	developing a directory of appropriate services able to provide care e.g. equipment supplies and maintenance, government benefits, continence assistance schemes and programs of aids for disabled persons.
vi)	Improving access to respite services, identifying supported accommodation and educating new carers.

### **Key objective of the service**

The key objective of the service is to provide clinical consultation, education, support, and preventive health strategies to young adults with spina bifida in an effort to facilitate their effective transition from paediatric to adult health services across NSW. The key elements of this model are summarised in table 3.



Table 3

<b>Key elements of the SBART Model</b>	
i)	facilitate and coordinate the effective transition of young people with spina bifida from paediatric to adult health services across NSW
ii)	provide clinical consultation, education and support, and preventive health strategies for adults with spina bifida living in the community across NSW.
iii)	facilitate participation in healthy lifestyle activities.

### **Clinical outcome measures**

The effectiveness of this program is monitored through key clinical indicators as listed in table 4.

Table 4

<b>Clinical outcome measures</b>	
i)	improved clinic attendance rates
ii)	decreased hospital admissions and decrease complications
iii)	improvements in self-care, self-esteem and fitness
iv)	increased independence and decreased reliance on carer support
v)	Increased employment and decrease on social service benefits

Appendices:

**Referral to Spina Bifida Adult Resource Team**

**Date:**

<b>Hospital or Service Provider Name</b>			
<b>Surname</b>	<b>First name</b>	<b>Date of birth</b>	<b>Gender</b> Male      Female
<b>Address</b>			
<b>Home phone number</b>		<b>Mobile number and email address</b>	
<b>Other reliable contact</b>		<b>Phone number of other contact</b> <b>Mobile number</b>	
<b>Interpreter required</b> Yes      No		<b>Language Spoken at Home:</b>	
<b>Primary diagnosis</b>		<b>Co-morbidities</b>	
<b>Reason for Referral to Spina Bifida Adult Resource Team and any priorities for management.</b>			
<b>Has a Spina Bifida Transition Plan been completed?</b>			
<b>Education / Employment Status</b> <input type="checkbox"/> School (which year) <input type="checkbox"/> Preparing for Uni /TAFE <input type="checkbox"/> TAFE <input type="checkbox"/> UNI <input type="checkbox"/> Work (? Full or part –time) What type and Where ..... <input type="checkbox"/> Other .....		<b>Living Arrangements</b> <input type="checkbox"/> Lives with family <input type="checkbox"/> Lives Alone <input type="checkbox"/> Lives with other (Not Family Member)  <b>Receives Personal Care</b> Yes <input type="checkbox"/> No <input type="checkbox"/> If Yes where from <input type="checkbox"/> Home Care/ Attendant Care <input type="checkbox"/> Family	
<b>Paediatric/Adult service details (if applicable list all clinicians involved and their speciality)</b>		<b>Proposed adult service details (if applicable list all relevant clinicians and their speciality)</b>	
<b>Recommended first appointment at adult service</b> ____/____/____			
<b>Suggested Place for Initial Contact and Assessment.</b> <input type="checkbox"/> Next Clinic Appointment <input type="checkbox"/> Home Visit <input type="checkbox"/> Other .....			

**Client Consent:** I, the client agree for this referral to be passed onto the Spina Bifida Adult Resource Team and to be contacted by the staff of the Spina Bifida Adult Resource Team.

**Name:**

**Signature:**

Please fax or email the completed form to Joanne Brady, Team Leader SB Adult Resource Team on 9482 9177.



## Spina Bifida Adult Resource Team

The Spina Bifida Adult Resource Team is a new initiative funded by NSW Health and commenced in August 2009. The team, which consists of a Clinical Nurse Consultant and a Senior Occupational Therapist, aims to provide clinical consultation, education, support and preventative health strategies to adults with spina bifida.

### Eligibility

Adults aged 18 and over who have Spina Bifida and who are living in NSW.

### Geographical area

The Spina Bifida Adult Resource Team is a state-wide resource team and covers all areas of NSW.

### Services

- Assist any young person making the move from paediatric to adult services
- Orientate any young person to their new adult health service
- Assist with any medical concerns as well as provide education and support to any adult with spina bifida
- Linking adults to appropriate health services as required
- Promote healthy lifestyles for all clients with Spina Bifida
- Attend all adult Spina Bifida Clinics across NSW
- Education to community services about spina bifida

### Referrals

Referrals are taken directly by the Spina Bifida Adult Resource Team. People can refer themselves or any health or community service working with the client can make the referral with the client's permission.

### Cost

There is no charge to use the Spina Bifida Adult Resource Team

### Funding

The service is funded by NSW Health and supported by Northcott Disability Services.

### Contact

**Joanne Brady**  
Team Leader/ Clinical Nurse Consultant  
Ph: (02) 9472 5214  
Mobile: 0418 429 960  
Email: joanne.brady@northcott.com.au

**Jade Baynes**  
Senior Occupational Therapist

Ph: (02) 9472 5215  
Mobile: 0418 408 738  
Email: jade.baynes@northcott.com.au



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