

spina bifida

Overview of spina bifida and the nervous system

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 group 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

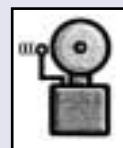
As neural tube defects affect the entire length of the brain and spinal cord, central nervous malformations are very common in people with spina bifida. One of the commonest types, the Arnold–Chiari malformation, often results in raised intracranial pressure early in childhood, requiring the surgical 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.

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.



Good continence control usually involves fairly 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^{1,2}

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?

Cerebrospinal fluid 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

Cerebrospinal fluid flow results from a pressure gradient that exists between the ventricular system and venous channels. The fluid flows from the lateral ventricles through foramina (foramina of Monro) into the third ventricle, and then passes 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.

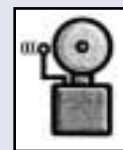
Hydrocephalus in spina bifida

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.

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 intracranial 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 within the first few months of life. In these individuals, the concern is that the shunt may become blocked, resulting in increased intracranial pressure.

Raised intracranial pressure secondary to Arnold–Chiari malformations can occasionally occur in later life resulting in spasticity, and abnormalities in gait 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.

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 to 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 management³

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 and cognitive problems that may 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. A spina bifida specialist centre can help to address these issues, and contact with the centre will provide assistance in overcoming problems.

To help in GP consultations, a list of common problems and some concrete strategies follows.

Common cognitive problems encountered in people with spina bifida

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

Short attention span and distractability interferes with the ability to pay attention to important details of a new task. Brief attention spans mean the person may not learn all of the necessary information, or may forget or hear only

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
- decreased motor and sensory function
- fits and seizures
- lower extremity hypertonia with generalised hyperreflexia.
- 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

part of any instructions given, as well as taking longer to complete tasks. Distractability can be internal, from the person's own thoughts, as well as from the environment.

Ringling patients with a gentle reminder about appointment times may help them to remember to attend, 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 supplement called *Passport to success*.

Language skills may be deceptive. 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 words said. 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 with inhibiting the flow of thoughts going through the mind or difficulty focusing on and comprehending relevant aspects of a situation

Difficulty in answering questions, following instructions, participating in back and forth conversation, or misinterpreting information or responding inappropriately to situations may indicate difficulties in actual comprehension.

Problems with abstract reasoning may cause difficulties in analysing and synthesising information and distinguishing between relevant and irrelevant information. This can also cause problems of generalisation 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 judgments 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, doing things at the last minute, or not keeping to 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 difficult situations are encountered, 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 diagrams and pictures, rather than repeating the same words over and over again. This will also help with any visual-spatial problems.

Ask the person to explain to you previous instructions to establish the level of comprehension. 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 success not only as an outcome, but also as an attempt. Empathise with any frustration felt during the attempt, even when the desired outcome has not 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 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 and remove the need to take notes and may help them to remain attentive during listening.

Be conscious of the 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 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 or www.racgp.org.au

References

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