

# **COGNITIVE FUNCTIONS IN SPINA BIFIDA**

**The cognitive is just as important as the physical**



## **Authors**

**Barbro Lindquist, neuropsychologist**

**Marie Peny-Dahlstrand, occupational therapist**

**In collaboration with**



**MMCUP**

Kvalitetsregistret för MMC  
och annan neuralrörsdefekt

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## **Introduction**

Arising early in fetal development, spina bifida is a malformation of the spinal column, spinal cord and brain. It manifests as a bulge in the spinal column and can be open (no skin covering) or closed. This sac usually contains spinal tissue and nerves. It prevents the working of nerve impulses to and from the brain. Symptoms vary depending on the spinal column position of the sac. Mobility is affected by reduced muscle function and sensation. Most people with spina bifida have impaired bladder and bowel function. Consequently, certain procedures for emptying both bladder and bowel have to be learnt. Regular emptying of the bladder via a catheter (clean intermittent catheterisation – CIC) is one example.

Most people with open spina bifida have increased fluid in the brain (hydrocephalus). To drain the fluid to the abdominal cavity, surgical insertion of a shunt and subcutaneous tube is usually required.

Spina bifida also has an impact on the brain (primarily the central and posterior parts). This can result in difficulties with several cognitive functions.

On giving birth to a child with spina bifida, it is usual for the first thoughts of relatives to be about the physical difficulties and the bladder and bowel problems. However, spina bifida also entails characteristic cognitive difficulties. Cognitive difficulties are not the same as low intellectual ability. It is possible to be highly gifted, but still to have cognitive difficulties in certain areas. Around one third of all people with spina bifida have an intellectual disability that means they find it harder than other people to understand and learn things (both theoretical and practical).

## **Cognitive functions**

Cognitive functions are mental processes in the brain that, for example, enable us to: perceive, interpret and process stimuli; think about and solve problems; understand speech; and communicate.

### **Cognitive functions in spina bifida**

For the person with spina bifida, cognitive difficulties can present everyday obstacles. Those around the person can experience this as frustrating, e.g., if a child does not get things done or has problems concentrating. Unfortunately, when they cannot do things that their friends do, a number of children can feel that they are inferior.

Thus, it is important that each child himself/herself understands what he/she is good at and what he/she finds difficult. Both at home and at school, each child should also get help with strategies. The more that parents know about their child's cognitive abilities, the better they can support the child's development and ensure that what they ask of the child is reasonable.

### **Common difficulties**

#### **Language**

As they often speak early with correct grammar and like talking and communicating, children with spina bifida seem to have good language ability. However, it is common for them not to understand the real meaning of words. Consequently, they may not grasp part or all of what a conversation is really about. For this reason, it is important to ensure that a child with spina bifida understands what is said or written and that what the child is seeking to communicate is truly understood.



## Memory

Both visual and auditory memory difficulties (i.e., as regards what is seen and what is heard) are common. They are often noticed early on. This applies to both long-term and short-term memory. Working memory is also often affected. This can make it difficult to remember several instructions given at a time or to remember an instruction for as long as it is needed to carry out a task.

## Activity regulation and concentration

Conscious regulation of activity is often difficult for children with spina bifida. Their attention soon wanders, and they are easily distracted. When they do get into a task, they can demonstrate good perseverance and concentration. However, it can then be difficult for them to switch focus and start something else.

## Visual perception and visuospatial ability



Perception is being aware of, interpreting and creating meaning from what we see, hear and feel. Problems with visual perception are particularly common with spina bifida. Interpretation of what is stationary (e.g., recognising faces, shapes and size) is usually fine, but visuospatial ability is affected. This limits spatial perception and spatial thinking. In turn, this entails difficulties in assessing distances and

direction; navigating; using maps; and, determining the speed at which something is moving. At home and in school, children with spina bifida can take time to learn to put clothes on the right way round and find things in cupboards and drawers.

## Executive and everyday functioning

Executive functions are the processes enabling us to: have an overview of our lives; move from idea to action; plan and use strategies; and evaluate and develop our strategies. Here, so that we can do what we want to do and what we need to do, most of our cognitive functions interact with each other.

The smooth working of these executive functions requires a good working memory and the abilities to grasp and think about several things simultaneously as well as to select and regulate own thoughts and impulses.

Our days are full of tasks and activities such as brushing teeth, completing homework, shopping, playing, etc. Doing absolutely anything is a process comprising various phases. We first need an idea regarding what we want to do. We then have to plan how this is to be

done, which steps are to be involved and the order in which they are to be carried out. Next, we must initiate i.e., go from thinking to action. With idea being turned into action, implementation/performing follows. Problems sometimes arise here. Things may not go as we expect. We must then rethink and plan all over again. We must also be able to terminate an activity, know and decide when enough has been done. Last, but not least, we must, throughout the process, manage timings – when to start, when to finish, when to hurry up and, possibly, when to omit a less important step so that we finish on time.

Anyone with **executive difficulties** such as those that are common in spina bifida has difficulties with several phases in each such process. Coming up with an idea can be difficult. Especially for new, unfamiliar activities, it is often hard to plan what is to be done and divide it into steps. Starting, i.e., moving from idea to action, is one of the most difficult things for many people with spina bifida. They know and also want to do what has to be done. Nonetheless, it is not seen through. Once something is started, performance is not usually difficult. Often, sometimes with a little training, it is possible to learn to do most things. However, even in very familiar activities, an unexpected difficulty that necessitates rethinking and making a new plan can halt things. Many people with spina bifida also relate that stopping is slightly difficult when something is underway. Finally, time management is commonly difficult for people with spina bifida. Individuals with spina bifida often learn to tell the time but may have difficulty relating to, and not least planning, timings.

***Difficulties with executive functioning are what cause most problems in the everyday lives of people with spina bifida. This applies regardless of the level of intellectual ability.***

The everyday difficulties faced by a person with spina bifida can be summarised as: *He/she can do it, but it somehow just never gets done.* For example, even though the ability exists, a snack may not be prepared or, despite the procedure and its great importance being well known, toilet needs may not be regularly attended to. Without nagging,

homework does not get done. Individuals with spina bifida often do not independently think of calling a friend. Movie invitations might be rejected because the person does not know how to get there.

This affects many facets of everyday life. Unfortunately, people with spina bifida are often incorrectly labelled as lazy, apathetic or unmotivated. Nagging and even over-protective are words that parents apply to themselves.



### **How is learning affected?**

As a rule, children with spina bifida learn early to decode and read fluently. However, they often demonstrate poor reading comprehension and have difficulty making connections. This is due to problems with, inter alia, working memory. Free composition is a challenge, both relating events (“What I did in the summer holidays” – memory) and telling a story (executive functioning).

Memory difficulties mean that learning facts, even if they are understood, is extremely time-consuming. In tests, recalling what has been learnt is also difficult. Sums and counting usually go well provided the task is concrete and clear. Problem-solving tasks are more difficult. Where mathematics requires spatial thinking, individuals with spina bifida find it difficult to: understand relationships between numbers, geometric shapes and diagrams; make rough estimates; and,



assess plausibility. The working-memory problem makes it difficult to cram tables, learn weight and dimension systems and understand money. In sports, correct direction and motion can be hard to maintain. In crafts, following joinery or sewing instructions can be problematic.

### **How to help?**

Problems in the various areas of cognition are interrelated. It is important that children with spina bifida are given the possibility of neuropsychological assessment to determine why some things work well and others are difficult. People often measure themselves against other people. In this connection, if children are not given the correct information, it often leads to unfortunate misconceptions that can affect self-image and self-confidence. In Sweden's Myelomeningocele Follow-Up Program (MMCUP), assessment is recommended at 4, 6 and 11-12 years of age. Ahead of starting school, the habilitation service and parents need to provide information about the future student's potential, difficulties and needs. Then, throughout schooling, regular follow-up meetings are recommended.

### **Preschool/school**

- One instruction at a time. Check that the child has understood. Visual aids?
- Give frequent feedback so that the child knows it is on the right track and does not need to redo anything because it has forgotten the instructions.
- Check reading comprehension:
  - Supplement with listening?
  - Visual aids?
  - Split texts up?
  - Rather than asking questions about what has just been read, ask questions first and then search for answers.
- Oral tests (with hints if necessary).
- Tests with multiple choice questions. It is easier to recognise knowledge than it is to dig it out.

- Quick reference guides for mathematics. Trying to learn things by heart is an inefficient use of time.
- Get the child to showcase his/her strengths.
- Clear teaching materials with little text.
- Everything in its place.
- If necessary, color code drawers and shelves.
- Set seats in classrooms and cafeteria.
- Give strategies for finding color coding (if any).
- Set routines!
- Time assistive devices.
- Calendars and schedules for clear step-by-step planning.

***Even if they have good understanding and intellectual ability, children with spina bifida often need individual adult support in school.***

### **Everyday life**

- Help children with spina bifida by giving them strategies to find their way around (both inside and outside home).
- Organise clothes and toys on an “everything in its place” basis. This also applies to “shared spaces” such as wardrobes, cupboards, drawers and refrigerators.
- Set routines.
- Calendars and timetables for clear step-by-step planning.
- Time assistive devices.
- Alarm clocks are common and can be of use. However, sometimes they do not help. Many people with spina bifida feel it is too easy to turn them off. If they ring when the person is in the middle of something else, finishing can be difficult and forgetting then plays its part.

Support with starting things is often invaluable, as also is support with reasoning to: find solutions when problems are encountered; or, remember what has been read for homework (but cannot be recalled).

Often, **extremely little support** is needed in these cases, but it can make all the difference.

### **Difficulties throughout life?**



The motor and cognitive difficulties caused by spina bifida do not change over time. Even though they manifest themselves differently and have different consequences depending on the stage of the person's life, they remain into adulthood. For everyday life to work smoothly (e.g., maintaining order in procedures, keeping to times and planning activities, get started), adults also usually require cognitive support. At all times, it is important for the person to: know and understand themselves; and get help with learning and with developing strategies. They need to find their own ways. Many need help to develop their own solutions,

rather than being told what to do.

### **Where is support available?**

Assessments are usually carried out in collaboration with the **habilitation team or the school healthcare service**. A psychologist carries out intellectual ability and perception tests and assesses executive functioning. There is also collaboration with: an occupational therapist (assessment of the child's skills and measures to improve these in everyday activities); a special needs teacher (to devise good

pedagogic strategies for play and school); a speech therapist (to map out language); and a physical therapist to clarify the child's motor potential. The occupational therapist and speech therapist can recommend and prescribe aids to help with time perception and communication.

Methods based on the individuals setting their own goals and getting help to find their own strategies have proven successful. The CO-OP method seeks to get the individuals to use own strategies and, with the help of guiding questions, analyze why something is not working (and then come up with new plans). It has been shown to improve the person's feeling of self-efficacy and can be offered by many habilitation services.

Parents, habilitation service and preschool/school then collaborate for the child to have every opportunity to develop to his or her full potential. This is a continuous process that needs to be adapted throughout adolescence and, not least, in preparation for the transition to adulthood when it is time to choose education courses and get own accommodation.

It is important that individuals with spina bifida can fulfil aspirations and be hopeful about and confident in their abilities. It has to be possible to develop what is fun rather than just seek to overcome difficulties.

### **More information about spina bifida can be found at:**

- [www.mmcup.se](http://www.mmcup.se).
- Spina bifida – Sweden's National Board of Health and Welfare.
- Spina bifida – 1177 (Sweden's web-based healthcare facility).

### **MMCUP**

MMCUP is a follow-up program and national quality register for spina bifida and other neural tube defects. The register seeks to increase the quality of healthcare so that, for people of all ages throughout Sweden,

complications can be avoided or, if necessary, treated in accordance with best practice.

### **Your local psychologist**

If you would like more information about cognitive function and what an assessment entails, contact your local habilitation service or local school healthcare service.

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### **If you have any questions about the contents of this booklet, please contact:**

Ann Alriksson-Schmidt.

Email: [ann.alriksson-schmidt@med.lu.se](mailto:ann.alriksson-schmidt@med.lu.se).

### **For more information booklets, please contact:**

Linda Sandström.

E-mail: [linda.sandstrom@med.lu.se](mailto:linda.sandstrom@med.lu.se).