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e-EarlyCare-T

Specialized and updated training on supporting advance technologies for early childhood education and care professionals and graduates

MODULE III.7

Spina bifid and spinal cord injury in children

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CONTENTS

I. Introduction

This topic deals with the definition, classification and aetiology of Spina Bifida (SB), as well as its treatment and functional consequences. It also defines and briefly explains some pathologies associated with SB such as **Hydrocephalus** and Arnold Chiari malformation. Finally, the bases of a multidisciplinary intervention program in early stimulation for children with BS or infantile spinal cord injury are proposed.

II. Objectives

The objectives of this unit are:

- To understand what is Spina Bifida and its main functional consequences.
- To approach the keys of a multidisciplinary program of early stimulation for children from 0-6 years old.

III. Specific contents of the theme

1. III. Definition and classification of Spina Bifida

During embryonic development, the vertebrae close at the back, thus protecting the contents of the **neural canal** (**meninges** and spinal cord), however, in cases of Spina Bifida (SB), this does not occur and the contents are exposed. Babies are born with a cyst on the back (Figure 1) that must be surgically operated within the first hours/days of birth.

Figure 1. baby with SB, before the operation.

Therefore, SB could be defined as a congenital malformation characterized by the lack of fusion of one or more vertebral arches, with or without protrusion of the **meninges** or

spinal cord, and whereby the contents of the **neural canal** are exposed to the outside (Gallar Pérez-Albaladejo, M.,2016).

1.1 Classification of Spina Bifida

Depending on whether or not the contents of the medullary canal come out, the SB is classified as follows (Gallar Pérez-Albaladejo, M.,2016):

- **Spina bifida occulta.** Some of the vertebral arches have not fused, and the lesion is covered by skin along its entire length. It may go unnoticed throughout life, or it may be detected accidentally in a spinal X-ray. No symptoms are associated with it, except that sometimes a little hair or a patch of skin may appear in that area (Figure 2).
- **Open spina bifida.** In these cases, the lesion is covered by membranes in the form of a cyst. If this cyst contains only the **meninges**, it is called a **meningocele**, but if, in addition to the **meninges**, it also contains part of the spinal cord, it is called a **myelomeningocele** (Figure 2). This second case is the most serious of all and has numerous consequences**.**

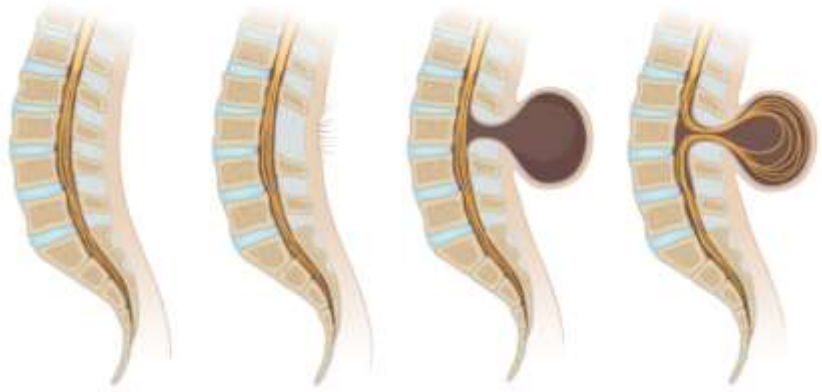

Figure 2. Spina Bifida classification: normal, occult, **meningocele**, **meningocele**, **myelomeningocele**.

The severity of **meningocele** or **myelomeningocele** depends on several factors (Gallar Pérez-Albaladejo, M.,2016):

- Location: the higher up (cervical area), the greater the sequelae, due to it affects more nerve roots.
- Extension: the greater the extension, the greater the sequelae. It depends on the number of nerve roots in the cyst.
- Presence or not of other associated malformations, such as **hydrocephalus** or Arnold Chiari malformation. The presence of these is associated with greater functional consequences. In addition to these two malformations that sometimes appear associated, there are also other complications such as tethered medulla that also cause greater functional complications.

2. III. Causes and prevention factors of Spina Bifida

The prevalence of neural tube malformations in Spain is estimated to be between 8 and 10 out of every 10,000 live new-borns, of which more than half of them are affected by BS (according to the Spanish Collaborative Study of Congenital Malformations) (AMEB, 2022). Other neural tube defects, such as **Anencephaly** or **Encephalocele**, are considered low prevalence diseases, and their sequelae are much more severe than those of BS.

2.1 Aetiology and prevention factors

The cause of SB is unknown, although its appearance has been related to different factors such as folic acid deficiency in the mother, the intake of valproic acid (antiepileptic) or etetrinate (psoriasis or acne drug) during pregnancy.

Prevention would therefore involve taking folic acid if pregnancy is being considered (it should be taken for at least one year prior to pregnancy) and the specialists should evaluate other alternative medication.

On the other hand, early diagnosis of SB during pregnancy is made through biochemical methods by determining the amount of alpha-fetoprotein in the mother. Ultrasound scans can also detect it, but it is difficult to see in the first weeks of pregnancy.

3. III. Spina Bifida Treatment

In the case of open lesions, as already mentioned, the baby must undergo surgery as soon as he/she is born, to close the cyst. This is a complex operation and the functional prognosis of the child will also depend on its outcome.

Since the consequences of SB are multiple and very complex, the treatment must be approached by a multi-professional team:

- Medical treatment: numerous specialists are involved in the SB process from birth, such as the neurosurgeon, in the first instance, but later and due to the manifestations of SB, other areas such as the urologist, traumatologist, rehabilitation physician and paediatrician, among others, will also intervene.

- Rehabilitative treatment: children with BS will have to receive rehabilitative treatment throughout early childhood, and possibly also later on, which should include the areas of physiotherapy, occupational therapy, orthopaedics and psychology.

In terms of rehabilitation, it is important to start an early care program as soon as possible in order to enhance their abilities and promote their development in all areas.

4. III. Consequences and functional implications

SB is considered a polydeforming disease, which has multiple organ involvement as a consequence of the neurological involvement resulting from the fact that the **meninges**

and nerve roots have been exposed. In general, the consequences that usually occur are as follows (Gallar Pérez-Albaladejo, M., 2016):

- Muscle weakness or even complete muscle paralysis below the injury. The higher the injury is located, the more difficulties they will have, so that if the injury is high, they will not be able to walk and may even have problems with arm weakness. On many occasions they will have to use mobility aids, whether crutches, walkers or wheelchairs. Also, as a result of this muscle weakness or paralysis, children with SB may have a variety of orthopaedic deformities such as **scoliosis**, varus or equinus feet.
- Loss of sensation below the lesion. May carry risks of skin lesions and burns among others.
- Weakness of the muscles of the bladder and intestinal tract. May present urinary and faecal incontinence, which implies an important series of care in this regard, and may require, in addition to wearing diapers, having to be catheterized periodically (once or twice a day). Among the problems of the intestinal tract, they may suffer constipation that may even lead to **rectal prolapse**.
- **Hydrocephalus**. This complication appears in 70% of children with SB (explained below).
- Other sequelae: **precocious puberty**, tendency to obesity, Arnold Chiari malformation, among others.

5. Hydrocephalus

It is one of the most frequent complications of SB, but it can also appear not associated with SB as a primary pathology, also causing disability by itself.

It is an accumulation of cerebrospinal fluid (CSF) in the brain, due to poor circulation or its non-reabsorption.

This increase in CSF implies an increase of the cerebral ventricles (where this fluid is produced) and this in turn leads to an increase in pressure in the brain, deforming the skull. It is urgent to resolve this situation in order to avoid brain lesions. To do this, a valve must be placed in the cerebral ventricles, which evacuates the excess CSF into the peritoneal cavity (abdomen) or into the vena cava, which is done by a new operation from the neurosurgery department.

Occasionally, **hydrocephalus** is also associated with delays in motor and cognitive development, which, if it is associated with BS, it would add this type of consequences (see Figure 3).

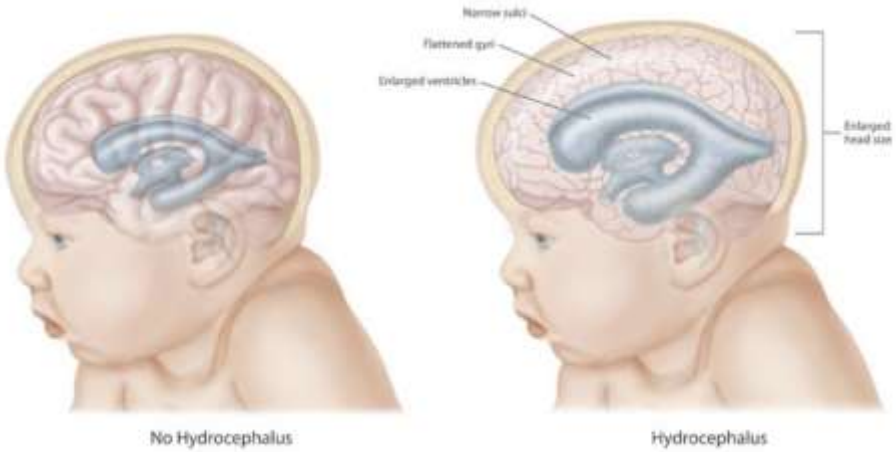

Figure 3. Brain ventricles imaging: without **hydrocephalus** with **hydrocephalus**.

6. Arnold Chiari malformation

Arnold Chiari malformation is a rare disease, which can also be found in isolation or linked to the presence of SB. When linked to SB, it is type 2, and consists of a descent of the cerebellum and the lower part of the IV cerebral ventricle into the **spinal canal**, also leading to elongation of the brainstem.

It is not necessarily associated with any other symptomatology, but sometimes there are difficulties in swallowing or breathing and weakness in the arms (see Figure 4).

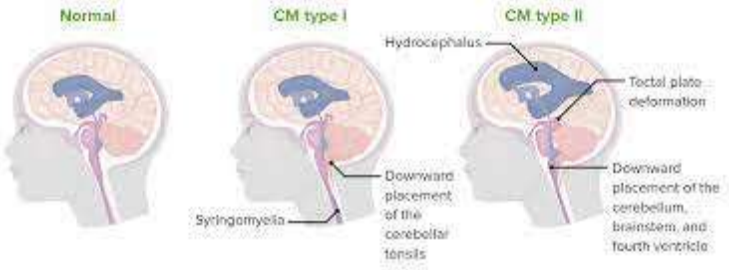

Figure 4. Image of Arnold Chiari malformation: normal, type I and type II.

7. Spinal Cord Injury in children

Spinal cord injury is a term that refers to the presence of damage to the spinal cord as a consequence of any traumatic or non-traumatic process. Therefore, SB could be included among spinal cord injuries of non-traumatic origin. However, injuries of traumatic origin (car accident, falls, etc.) are the most frequent in young adults, and although they represent a low percentage in early childhood, it is important to know that there are also children with spinal cord injuries of traumatic origin.

The consequences of these traumatic spinal cord injuries are very similar to those of SB, except that they do not involve, for example, the risk of **hydrocephalus** or other malformations. They share muscle weakness or paralysis below the lesion, loss of sensation, and weakness of the muscles of the bladder and intestinal tract.

Like SB, it will also require a multi-professional approach to provide the child with intervention programs to facilitate his/her development and acquisition of independence.

8. Intervention proposal for Spina Bifida and spinal cord injury in children

The approach to both SB and spinal cord injury should be, as already mentioned, a multidisciplinary one. In the case of SB, the first year of the child's life, and in the case of spinal cord injury, the first year after the lesion appears, will be mainly marked by medical intervention and stabilization of the lesion (closure of the cyst, treatment of **hydrocephalus** if it appears, etc).

Once the injury is medically stabilized, it is advisable for the child to start the stimulation program as soon as possible in order to favour the development of his/her full potential.

8.1 Objectives of the physiotherapy intervention program

From the physiotherapy point of view, the objectives of the program should focus mainly on:

- Enhancing all preserved musculature, starting with trunk control to promote sitting and continuing with the lower extremities and upper extremities if affected.
- Achieving independent mobility, with or without orthopaedic aids. These orthopaedic aids can be, for example, foot splints, hip-knee-ankle-foot orthosis (see Figure 5), crutches, walkers or wheelchairs, among others.

Figure 5. hip-knee-ankle-foot orthosis.

- To prevent orthopaedic deformities. Ideally, this can be done from birth, even when the child remains in the hospital. From the incubator, appropriate postures of the lower limbs can be encouraged to avoid the appearance of the dreaded deformities. Some of these deformities that may appear are:
    - Trunk deformities: **scoliosis**, lumbar hyper lordosis, dorsal kyphosis.
    - More frequent deformities in the lower extremities: hip flexion, hip dislocations, knee varus/valgus, equinus/varus/valgus foot, among others.

8.2 Objectives of the occupational therapy intervention program

From Occupational Therapy point of view, an intervention program should include at least the following objectives:

- To achieve independence in Activities of Daily Living (ADLs), always respecting the pace of development (see module 6).
- To advise and train the use of support products that may be necessary to achieve this independence. In addition to mobility products (walkers, crutches, wheelchairs), children with SB and spinal cord injury may need other products to help them in their daily life, such as adaptations of cutlery or school utensils and materials.
- To adapt the environment and its materials to facilitate this independence.
- It is also important that the multidisciplinary program includes the family, also as part of the intervention.

8.3 eEarlyCare application web

The eEarlyCare web application (Saiz-Manzanares, Marticorena-Sánchez, & Árnaiz-González, 2020;2022; Saiz-Manzanares et al., 2020), offers one of the modules on transfers that can be applied in children with BS and spinal cord injury. A more detailed study of the tool is presented in Module VII.3.

8.4 Others

The presence of a child with a disability generates a multitude of feelings, which are sometimes difficult to manage and must be addressed.

Also, especially in cases of acquired spinal cord injury, the children themselves need, even at such early ages, psychological help to cope with the changes brought about by their situation.

The intervention of more professionals may be necessary, depending on the extent of the injury and the presence of other complications:

- In case it is detected cognitive delay, the program should also include this part.
- The nursing staff should also take care of the lesions that may appear on the skin, for example the presence of pressure ulcers.
- The dietician also can work as part of the team to avoid the tendency of children with SB to become obese.

IV. Summary

The chapter defined SB and shown its classification, as well as its main functional consequences, briefly describing **hydrocephalus** and Arnold Chiari malformation, commonly associated with SB. It also established the most important objectives of an early stimulation program dedicated to children with SB or infantile spinal cord injury.

V. Glossary

Anencephaly: Malformation of the neural tube, which implies the non-formation of parts of the brain (either parts of the brain, brain stem or cerebellum) during embryonic development. It is a very serious malformation.

Spinal canal or neural canal: central hollow part of the spinal column in which the spinal cord is located.

Encephalocele: cyst-shaped protrusion in the skull, through which the **meninge**s and part of the brain protrude. It occurs during embryonic development, and is a very serious malformation.

Scoliosis: Deformity of the spine, which implies a curvature of the spine in the anterior-posterior plane in the shape of a "C" or even in the shape of an "S". It also includes a rotation of one or more vertebrae.

Sphincter: Muscle that controls the emptying of the bladder or bowel, depending on whether it is the urinary or anal **sphincter**.

Hydrocephalus: accumulation of cerebrospinal fluid in the brain.

Tethered spinal cord: A neurological condition in which the spinal cord is attached (tethered) to the surrounding tissues of the spine. This prevents the spinal cord from being able to move and grow as the child grows.

Meninge: The membrane that surrounds the spinal cord.

Meningocele: A type of open spina bifida.

Myelodysplasia: Synonym for Spina Bifida.

Myelomeningocele: type of open spina bifida.

Varus foot: deformity of the foot that adopts a "C" position, with the front part of the foot turned inward.

Equinus Foot: Deformity of the foot, where the foot appears stretched with the tip of the foot downward (as if on tiptoe).

Rectal prolapse: When passing stool, part of the rectum comes out. It usually occurs in infants and when they control constipation, it disappears.

Precocious puberty: Consists in the advancement of puberty, it occurs in girls with Spina Bifida, with the consequent advancement of menstruation at 8 or 9 years of age.

Raquisisis: Synonym of Spina Bifida.

VI. Bibliography

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6.2. Recommended bibliography

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VII. Images

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Image 5: https://www.ortopediamostkoff.com.mx/producto/ortesis-miembros-inferiores/pierna/afo-por-sus-siglas-en-ingles-ankle-foot-orthosis-ortesis-de-tobillo-pie/

VIII. Resources/Web

Living with Spina Bifida: Data that Make a Difference: https://www.cdc.gov/ncbddd/spinabifida/documents/cdcsworklivingwithspinabifida.pdf

Free resources about SB, available in English and Spanish: https://www.cdc.gov/ncbddd/spinabifida/index.html