

Module III.7

Characteristics of spina bifida and spinal cord injury at early ages.



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1.0. Introduction.

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1.0. 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.



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1.1. A. Spina Bifida definition

During embryonic development, the vertebrae close posteriorly, protecting the contents of the neural canal (meninges and spinal cord), however this does not occur in cases of Spina Bifida (SB) and the contents are exposed.

Therefore, SB can 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, whereby the contents of the neural canal are exposed to the outside (Gallar Pérez-Albaladejo, M.,2016).



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Classification of Spina Bifida

Depending on whether the content of the spinal canal comes out or not, SB are 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 on a spinal X-ray. There are no associated symptoms.

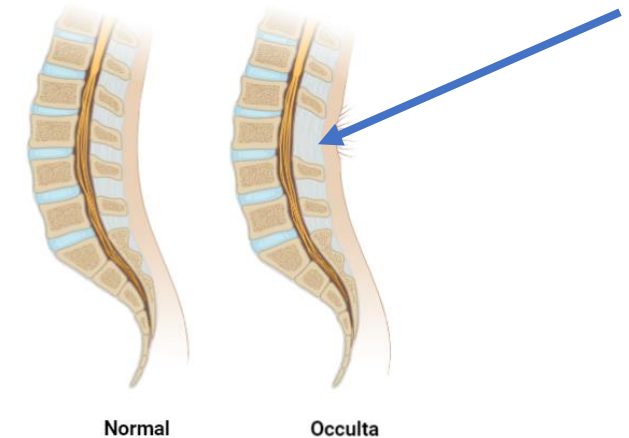


Image 1



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Classification of Spina Bifida

- **Open spina bifida.** In these cases, the lesion appears 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**. This is the most serious of all and has numerous consequences.

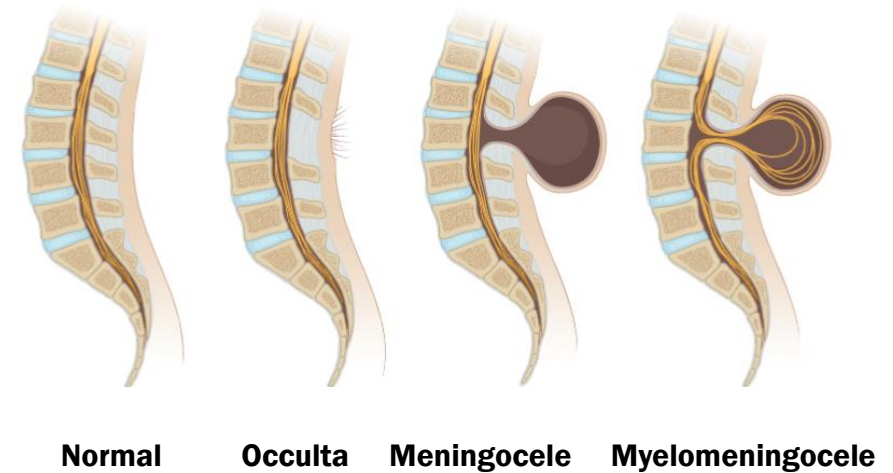


Image 2



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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.
- Extension: the greater the extension, the greater the sequelae.
- Presence of other associated malformations, such as Hydrocephalus or Arnold Chiari disease. Presence of these is associated with greater functional consequences.



Image 3



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Prevalence of Spina Bifida

In Spain between 8 and 10 out of every 10,000 live newborns have some neural tube malformation, of which more than half are affected by SB (according to the Spanish Collaborative Study of Congenital Malformations) (AMSB, 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 SB.



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1.2. Causes and prevention factors of SB

Etiology

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

Prevention factors

Prevention would therefore involve taking folic acid if pregnancy is being considered, and the evaluation of alternative medication by specialists.

Early diagnosis of SB is 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.



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1.3. Treatment of SB

In the case of open lesions, the baby must undergo surgery immediately after birth to close the cyst.

This is a complex operation, and the functional prognosis of the child will also depend on its outcome.

Since SB has multiple, very complex consequences, treatment must be approached by a multi-professional team:

- Medical treatment: various specialists are involved, such as neurosurgeons, urologists, traumatologists, rehabilitation specialists, and pediatricians, among others.
- Rehabilitation: physiotherapy, occupational therapy, orthopedic and psychological treatment.

It is important for rehabilitation to start an early care program as soon as possible to give them the best possible chance to work in all areas.



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1.4. Consequences and Functional Implications of SB

SB is considered a polydeforming disease, with multiple organ involvement as a consequence of the neurological involvement resulting from the fact that the meninges and roots have been exposed. Generally speaking, the consequences are usually as follows (Gallar Pérez-Albaladejo, M., 2016):

- Muscle weakness or even complete muscle paralysis below the injury. The higher the injury, the more difficulties it will present, so that if the injury is high, patients will not be able to walk and may even present weakness in the arms.



Image 4



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1.4. Consequences and Functional Implications of SB

- Loss of sensitivity below the lesion. May entail risks of skin lesions and burns, among other things.
- Weakness of the bladder and intestinal tract muscles. May present urinary and fecal incontinence, which means significant care in this regard.
- Hydrocephalus. This complication appears in 70% of children with SB. It is treated a little later.
- Other sequelae: precocious puberty, tendency to obesity, Arnold Chiari malformation, among others.



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1.5. Hydrocephalus

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

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

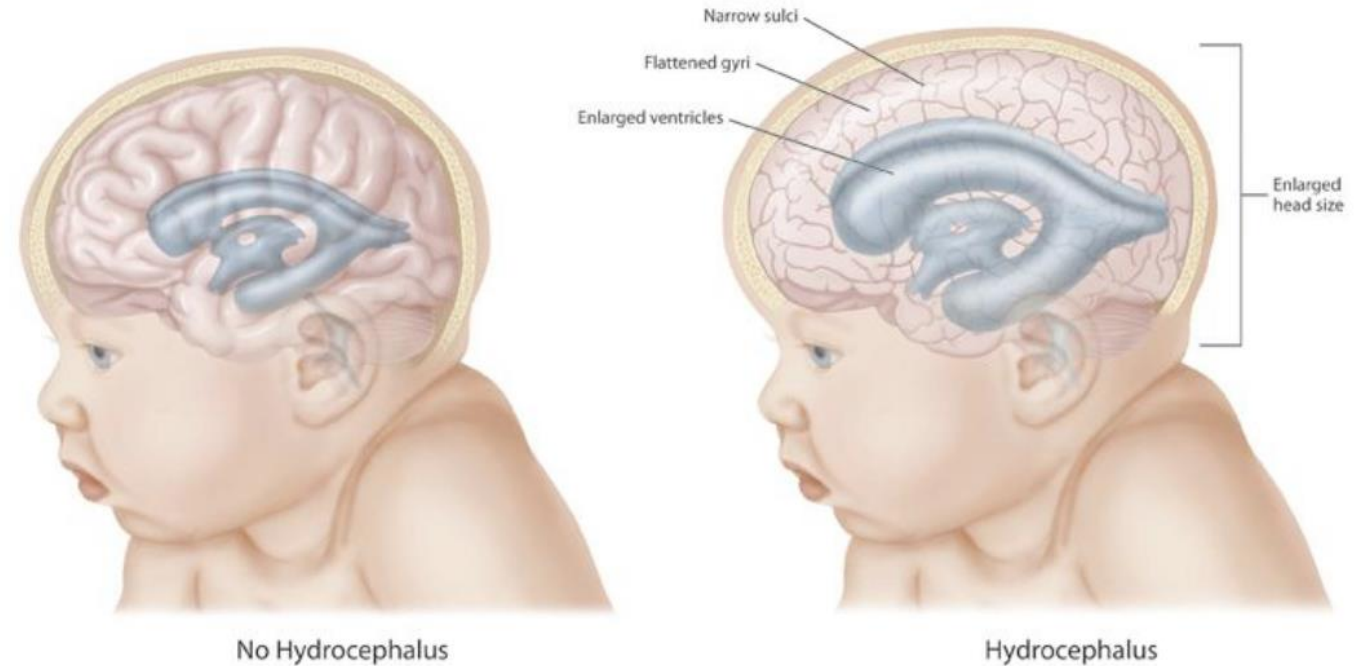


Image 5



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1.5 Hydrocephalus

This increase in CSF means an increase in the size of the cerebral ventricles (where this fluid is produced) and this leads to increased pressure on the brain, deforming the skull. This situation must be addressed urgently in order to avoid brain lesions. To do this, a valve is placed in the cerebral ventricles which evacuates the excess CSF to the peritoneal cavity or to the vena cava, which is done by means of a new operation from the neurosurgery department.

Sometimes hydrocephalus is also associated with delays in motor and cognitive development, which, if associated with SB, would add to the consequences of the latter.



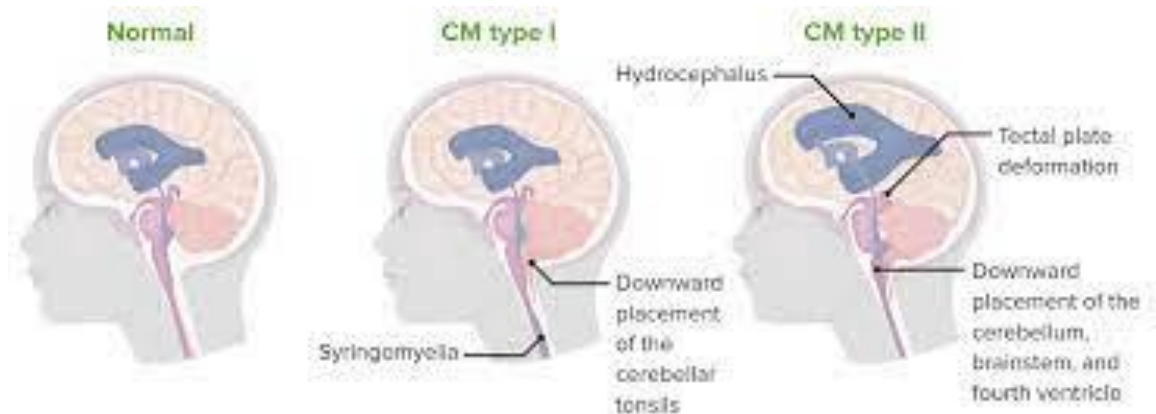
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1.6. Arnold Chiari

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

It does not have to be associated with any other symptomatology, but sometimes there are difficulties in swallowing or breathing and weakness in the arms.

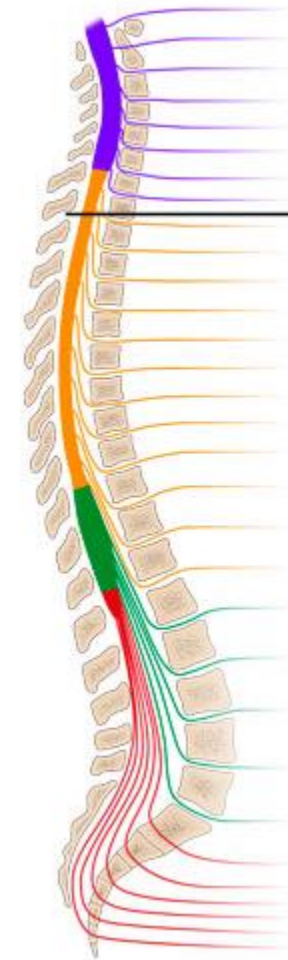
Image 6



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1.7. Definition of pediatric spinal cord injury

Spinal cord injury is a term that refers to the presence of damage to the spinal cord as a consequence of a traumatic or non-traumatic process. SB could be included among spinal cord injuries of non-traumatic origin. However, injuries of traumatic origin (car accident, falls, etc.) are more common in young adults, and the proportion in early childhood is low, it is important to be aware that there are also children with spinal cord injuries of traumatic origin.



Quadriplegia:
loss of sensation and
movement in all four
extremities



Paraplegia:
loss of movement and
sensation in the lower half of
the body



Image 7



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1.7. Definition of paediatric spinal cord injury

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

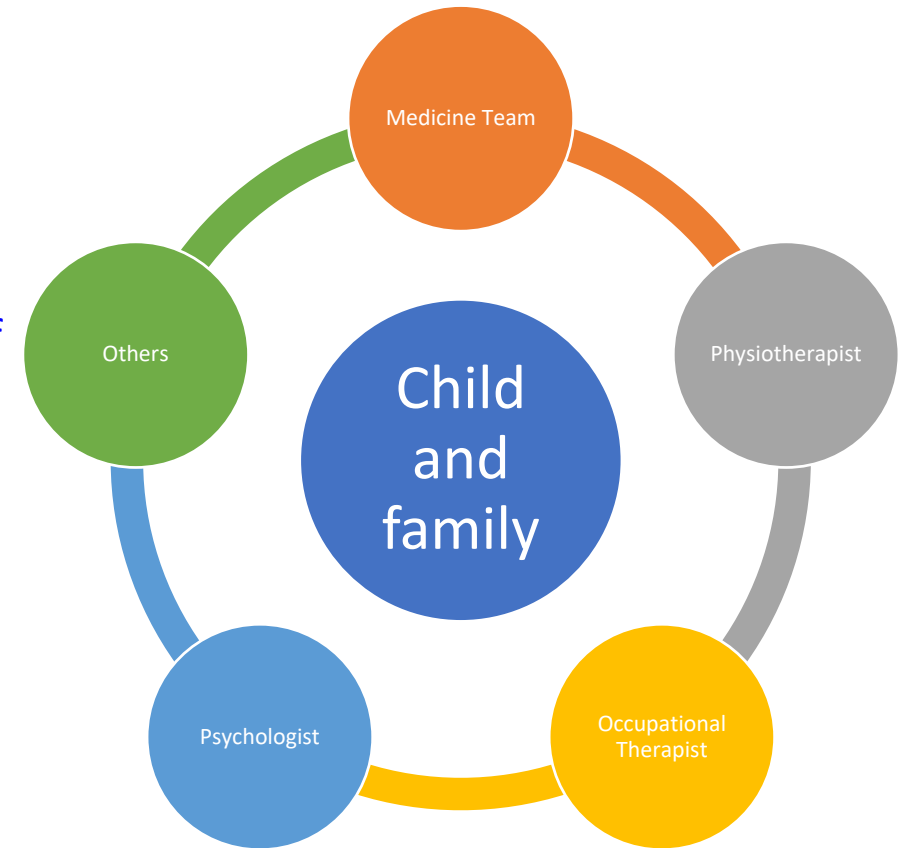
Like SB, they also require a multi-professional approach to provide children with intervention programs that facilitate development and the acquisition of independence.



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1.8. Intervention proposals in SB and spinal cord injury.

The approach to both SB and spinal cord injury should be multidisciplinary. In SB, the first year of the child's life, and with spinal cord injuries, 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.).



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1.8. Intervention proposals in SB and spinal cord injury.

Once the injury is medically stabilized, it is advisable for children to begin stimulation programs as soon as possible to encourage development of their full potential.

From a **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 orthopedic aids.
- Avoid orthopedic deformities. Ideally, this can be done from birth, even while the child is in hospital.



Image 8



Characteristics of spina bifida and spinal cord injury at early ages.

1.8. Intervention proposals in SB and spinal cord injury.

From an **occupational therapy** point of view, an intervention program should have at least the following objectives:

- Achieve independence in Activities of Daily Living (ADLs), always respecting the pace of development (see module 6).
- Advise and train in the use of assistive devices needed 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.
- Adapt the environment and its materials to facilitate this independence.



Image 9. Assistive device to handle eraser. Made for a 6-year-old girl with spinal cord injury.



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1.8. Intervention proposals in SB and spinal cord injury.

It is also important for multidisciplinary programs to include **the family** as part of the intervention.

The presence of a child with a disability produces 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, even at such early ages, need **psychological help** to cope with the changes brought about by their situation.



Image 9.



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1.8. Intervention proposals in SB and spinal cord injury.

eEarlyCare web application

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 SB and spinal cord injury. A more detailed study of the tool is presented in Module VII. 3.



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1.8. Intervention proposals in SB and spinal cord injury..

Other

- The intervention of more professionals may be necessary, depending on the extent of the injury, and the presence of other complications:
- If cognitive delay is detected, the program should also include this part.
- Nursing staff should also take care of lesions that may appear on the skin, for example pressure sores.
- Dieticians should help children with SB avoid becoming obese.



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Images

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Image 6: <https://www.lecturio.com/es/concepts/malformaciones-de-chiari/>

Image 7: <https://www.stanfordchildrens.org/es/topic/default?id=spinalcordinjury-85-P04278>

Image 8: <https://www.fisioterapiaduque.com/clinica-fisioterapia-burgos/>

Image 9: Own production

Image 10: <https://www.freepik.es/foto-gratis/padres-felices-riendo-junto-hija_13309072.htm#query=familia&position=4&from_view=search>Image de rawpixel.com in Freepik

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