Guideline

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Visual atlas and recommendations for the follow-up of the newborn with fetoscopic myelomeningocele surgery

Atlas visual y recomendaciones para el seguimiento del recién nacido con cirugía fetoscópica de mielomeningocele

Atlas visual e recomendações para acompanhamento do recém-nascido com cirurgia fetoscópica de mielomeningocele

Catalina Vaz Ferreira[1], Paula Couchet[1], Mario Moraes[2], Gonzalo Costa[3], Alfredo Cerisola[4], Grupo Fetología Clínica Álvarez Caldeyro Barcia[5]

Abstract

Myelomeningocele is the most common malformation of the central nervous system, associated with functional limitations and neuromusculoskeletal, nephourological, and intestinal complications with the consequent social and psychological repercussions in the individuals who present it. Worldwide, and in our country, fetal surgery with repair of the defect is currently the first-line treatment option. In Uruguay, more than 20 fetal surgeries have been performed with the SAFER technique. The objective of this atlas is to guide clinicians in the appropriate neonatal evaluation and treatment of patients who underwent surgery in the fetal period. A postnatal multidisciplinary management guide for newborns is presented, including illustrative images and recommendations on neonatal follow-up and long-term neurosurgical, neuropediatric, and urological care, among others. We highlight the importance of a comprehensive, individualized, and multidisciplinary assessment required by these patients to optimize surgical and functional outcomes.

Keywords: Meningocele
Postnatal Care

Resumen

El mielomeningocele es la malformación del sistema nervioso central más frecuente que se acompaña de limitaciones funcionales y complicaciones neuromusculoesqueléticas, nefrourológicas e intestinales con la consecuente repercusión social y psicológica en los individuos que la presentan. La cirugía fetal con reparación del defecto es a nivel mundial y en nuestro país la primera opción de tratamiento en la actualidad. En Uruguay se han realizado más de 20 cirugías fetales con técnica SAFER. El objetivo de este atlas es guiar a los clínicos en la evaluación y el tratamiento neonatal acorde de los pacientes que fueron intervenidos en el período fetal. Se

1. Asist. Dra. Unidad Académica Neonatología. CHPR. Facultad de Medicina. UDELAR.
2. Prof. Dr. Unidad Académica Neonatología. CHPR. Facultad de Medicina. UDELAR.
3. Prof. Agdo. Dr. Cátedra de Neurocirugía. CHPR. Facultad de Medicina. UDELAR.
4. Prof. Agdo. Dr. Cátedra de Neuropediatría. CHPR. Facultad de Medicina. UDELAR.
5. Grupo Fetología Clínica Álvarez Caldeyro Barcia.
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presenta una guía de abordaje multidisciplinario posnatal del recién nacido con imágenes ilustrativas y recomendaciones sobre el seguimiento neonatológico y a largo plazo neuroquirúrgico, neuropediátrico y urológico, entre otros. Destacamos la importancia de la valoración integral, individualizada y multidisciplinaria que requieren estos pacientes para optimizar los resultados quirúrgicos y funcionales.

Palabras claves: Meningocele
Atención Postnatal

Resumo

A mielomeningocele é a malformação mais comum do sistema nervoso central, que é acompanhada de limitações funcionais e complicações neuro-músculo-esqueléticas, nefro-urológicas e intestinais com as consequentes repercussões sociais e psicológicas nos indivíduos que a apresentam. A cirurgia fetal com correção do defeito é, internacionalmente e em Uruguai, a primeira opção de tratamento atualmente. No Uruguai, quase 20 cirurgias fetais foram realizadas com a técnica SAFER. O objetivo deste atlas é orientar os médicos na avaliação e tratamento neonatal adequado de pacientes submetidos a cirurgia no período fetal. É apresentado um guia de abordagem multidisciplinar pós-natal para o recém-nascido com imagens ilustrativas e recomendações sobre acompanhamento neonatológico e neurocirúrgico de longo prazo, neuropediátrico e urológico, entre outros. Destacamos a importância da avaliação abrangente, individualizada e multidisciplinar que estes pacientes necessitam para otimizar os resultados cirúrgicos e funcionais.

Palavras chave: Meningocele
Cuidados Pós-natais

Introduction

Within neural tube defects, spinal dysraphisms encompass congenital malformations that affect the spine, spinal cord, and/or meninges. It is a non-progressive pathology, but its secondary alterations can develop over time.

There are different types depending on the affected structures:

- Spina bifida occulta without spinal cord involvement.
- Meningocele (involves the meninges).
- Myelomeningocele (MMC) (involves the spinal cord and meninges).
- Lipomyelomeningocele (involvement of the spinal cord, includes an accumulation of adipose tissue -lipoma- associated with the neural placode and the meninges).

The MMC is the most common congenital malformation of the central nervous system, associated with spinal cord injury at the level of the lesion, as well as hindbrain herniation. Ventriculomegaly is associated with up to 90% of MMC cases, with the defect at the lumbosacral level being the most frequent.

Its importance lies in its frequency, functional limitations, and neuro-musculoskeletal, nephro-urological, and intestinal complications that may arise, as well as its social and psychological impact, considering its prolonged survival given the technological advances. In Uruguay, the prevalence of MMC is estimated at 8/10,000 births. Worldwide, 300,000 children are born annually with this condition, contributing to the burden of infant mortality (41,000deaths/year).

The etiology is multifactorial; however, folate deficiency is identified as the main risk factor. Periconceptional maternal supplementation is the primary prevention strategy, reducing the risk by 50% to 70%. Other factors involved are environmental, genetic, and pharmacological ones.

Early diagnosis in the first trimester is key for family counseling, coordination with the multidisciplinary team, and determining the timing and surgical approach[1-3].

The prognosis depends on multiple factors, including the level of the defect, which will determine the extent of sensory, motor, and autonomic dysfunction.

The affected neurological level is key for functional prognosis. Different classifications have been proposed to categorize them. We highlight the National Spina Bifida Patient Registry (NSBPR) Scale, which divides them into:

a. Thoracic (no lower limb mobility)
b. High lumbar (hip flexion present)
c. Mid-lumbar (knee extension)
d. Low lumbar (ankle dorsiflexion)
e. Sacral (ankle plantar flexion)

It is important to highlight that the functional ambulation prognosis is better the lower the lesion level.

Regarding the neurological lesion level:

1) Patients with lumbosacral MMC typically present with sensory-motor deficits in the lower limbs, urinary and intestinal dysfunction, and possible lower limb deformities.

2) Independent and unaided ambulation is expected when lesions are below S1.

3) Partial or total dependence on wheelchair mobility is expected when the lesion level is above L2. In these cases, scoliosis is also likely to develop at some point.

4) When the lesion is at the L4-L5 level, ambulation (with or without assistive devices) is achieved approximately 50% of the time.

Conditions associated with the brain may include:

1) Chiari II malformation (80%): a complex developmental brain malformation characterized by a small posterior fossa and downward displacement of the cerebellum and brainstem through an enlarged foramen magnum.

2) Hydrocephalus: it may result from compression of posterior fossa structures in the context of a Chiari II malformation (including obstruction of the fourth ventricle or its outlets, or impaired cerebrospinal fluid [CSF] circulation around the brainstem at the foramen magnum level), or aqueductal stenosis (present in 40% - 75% of cases).

3) Corpus callosum dysgenesis.

4) Neuronal migration disorders (polymicrogyria, heterotopic gray matter).

These conditions may present clinically with:

1) Lower cranial nerves involvement, including swallowing disorders, respiratory stridor, and apnea.

2) Cervical compression, which may lead to spinal paresis, opisthotonus, and hydro/syringomyelia.

3) Macrocephaly and other signs of intracranial hypertension.

4) Global developmental delay and/or cognitive impairment, associated with underlying malformations and complications of hydrocephalus shunt systems (especially ventriculitis).

5) Seizures (20% of cases).

6) Visual complications.

Conditions associated with the spine and spinal cord may include:

1) Scoliosis

2) Hydro/syringomyelia

3) Diastematomyelia

4) Tethered cord syndrome

5) Urinary dysfunction

6) Intestinal dysfunction

7) Sexual dysfunction

The main cause of morbidity and mortality in these patients is secondary to hydrocephalus and its complications, mainly infectious ones. Other associated morbidities include cognitive deficits in up to 40% of cases and motor disability[4-7].

Prenatal repair has been shown to halt spinal cord damage and reverse hindbrain herniation, improving survival and morbidity burden in these patients compared to those who were intervened postnatally. Early defect closure in the second trimester halts the loss of motor neurons in areas adjacent to the lesion.

According to the MOMS trial, fetuses who underwent prenatal surgery had better outcomes, including a lower need for ventriculoperitoneal shunting by one year of age, a decreased incidence of hindbrain herniation (Chiari), and a higher rate of ambulation without orthotic devices. Likewise, the secondary analysis of the MOMS trial showed long-term benefits of *in utero* neuroprotection in terms of motor level and functional skills at school age[8-11]. Since 2021, more than 20 minimally invasive fetoscopic MMC repair surgeries (SAFER technique or skin-over-biocellulose) have been performed in our country. This technique has demonstrated neurological outcomes comparable to those of open prenatal repair (according to MOMS) with greater obstetric advantages such as the potential for vaginal delivery and reduced risk of uterine suture dehiscence.

The objective of this atlas is to pr ovide clinicians with illustrative and educational material, serving as a guideline for a standardized approach to patients who undergo prenatal intervention and will receive postnatal care from a multidisciplinary team[12-15].

Scenario 1. Repair with artificial skin (Nevelia® matrix) (Figure 1).

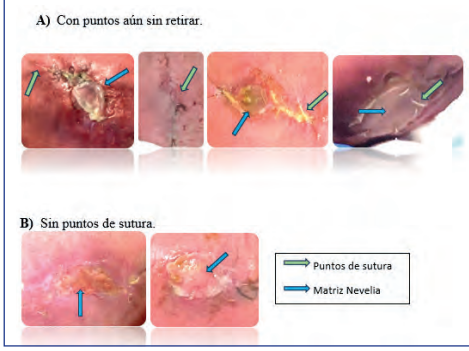
It consists of a porous bovine collagen layer to promote and guide dermal regeneration and a reinforced silicone layer that acts as a pseudoepidermis. This matrix serves as a support for cellular infiltration, becoming a dermis-like tissue and being reabsorbed 2-3 weeks after implantation.

It is used in large defects that cannot be closed initially[16-18].

Care and management at birth

- The patient should be handled with sterile nitrile gloves.
- The surgical wound should be cleaned with saline solution.

Figure 1. Presentation at birth.



- The stitches should be cut at birth. The suture is continuous and performed with a non-absorbable transparent thread. The stitches should be removed by the neurosurgeon of the clinical fetology team when the defect is completely closed. To remove the stitches, simply cut the suture thread once it becomes visible over the silicone.
- The silicone will spontaneously detach from the matrix after the stitches are removed. Epithelialization occurs by secondary intention[19-21].
- If epithelialization occurs earlier, the stitches should be removed.
- Apply a foam dressing for sensitive skin, such as Mepilex® Border or Allevyn Gentle Border (Figure 2).
- The dressing should be available at the time of birth. It should not be replaced with porous dressings to avoid the risk of direct contact between the wound and feces or urine.

The dressing should be changed every two or three days or sooner if it becomes contaminated with feces or urine.

In cases where the defect is very low, it is suggested to place a small "roll" of sterile gauze in the intergluteal cleft closest to the wound and then cover it with the dressing (Figure 3).

If a foam dressing like Mepilex® Border or Allevyn Gentle Border is not available, the best dressing option will be discussed with the clinical fetology team, considering the newborn's skin immaturity and whether the wound has areas of dehiscence.

An option is to use a hydrocolloid dressing like Duoderm or a non-adhesive Allevyn Classic dressing, which can be covered with Duoderm or another adhesive.

Figure 2. Application of foam dressing for sensitive skin.

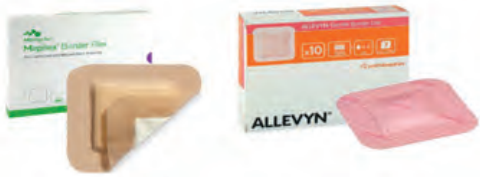
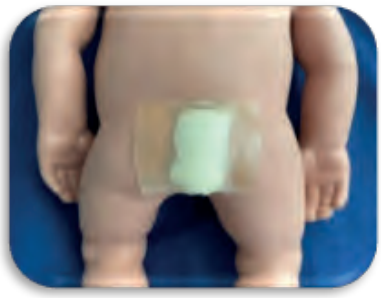


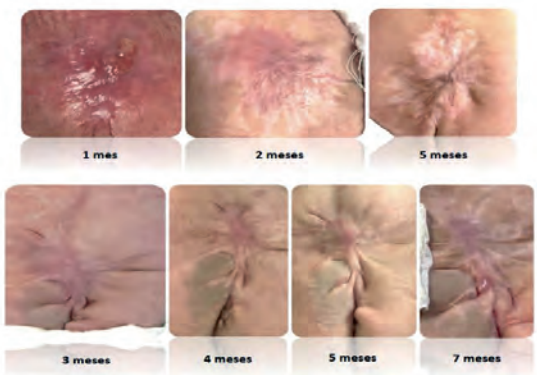
Figure 3. Dressing.



It is not recommended to cover the defect with a sterile polyethylene bag or antibiotic dressings.

Natural progression of epithelialization (Figure 4)**.**

Figure 4. Natural progression of epithelialization.



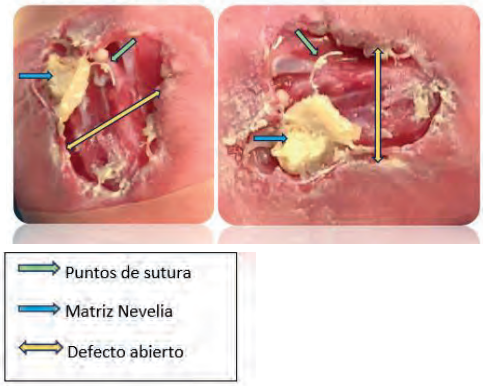
Complications

In case of suspected systemic infection/sepsis, it is recommended to collect samples for blood culture (at least 1 ml of blood by aseptic peripheral puncture), wound culture (and Gram stain), and urine culture by bladder catheterization (Figure 5).

It is recommended to use an empirical antibiotic coverage plan with activity against both Gram+ and Gram– microorganisms, with good penetration in CSF, such as ampicillin + cefotaxime if the patient is admitted from home, or vancomycin + meropenem if the patient is already hospitalized, expanding coverage to coagulase-negative Staphylococcus, MRSA, and Gram– with ESBL+ resistance. It is recommended

Figure 5. Complications.

A) Failure of intrauterine suturing. CSF leakage.



B) Infectious complications.

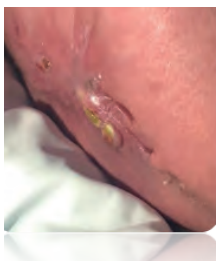
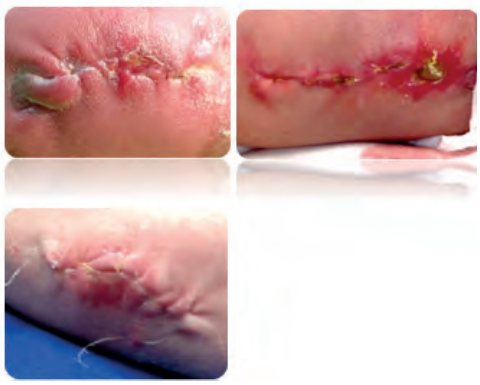


Figure 6. Care of the newborn with fetal surgery scar.



to adjust antibiotic treatment according to the sensitivity profile of the isolated microorganism as soon as possible.

If systemic infectious complication is ruled out with negative cultures, it is recommended to discontinue intravenous antibiotic therapy and switch to topical treatment, according to the skin culture from the lesion area.

Lumbar puncture is not recommended in case of infectious complications; the best puncture site for CSF collection should be evaluated with the treating neurosurgeon.

In case of any complication, the clinical fetology team should be contacted for a joint evaluation[22-23].

Scenario 2. Care in a newborn with fetal surgery scar (without matrix) (Figure 6).

Care and management at birth

• At birth, clean the skin with saline solution and cover with Mepilex® Border or Allevyn Gentle Border foam dressing.

• It is not recommended to remove the skin suture; excess thread at the edges of the lesion may be cut.

Long-term evolution

If any area remains open or has not completely closed:

We do not recommend attempting new sutures to bring the edges together. We do not recommend using steri-strips to try to approximate the edges. With the application of Mepilex® Border or Allevyn Gentle Border dressings, healing by secondary intention will occur (Figure 7).

Latex allergy

Latex allergy is observed in 10%-73% of patients with MMC, so these patients should be considered allergic, ensuring care throughout the hospital stay and subsequent discharge, without products that may contain latex in their composition. It is important to ensure that latex-free gloves and resuscitation materials are available in the delivery room and to educate the family on avoiding contact with products that may contain latex protein, as well as recognizing allergy/anaphylaxis symptoms.

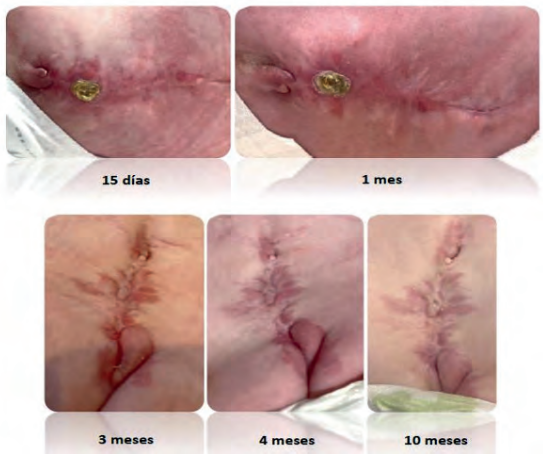
Follow-up recommendations

It is suggested that the clinical fetology team evaluates and follows up on the motor and urinary development, the potential requirement of a shunt (VP shunt), and other interventions, regardless of the initial healthcare provider for the newborn.

Given the heterogeneity of possible clinical scenarios related to MMC and the associated conditions and complications, some general criteria following the immediate perinatal period are:

1) The follow-up of patients with MMC requires an interdisciplinary team with a case-by-case personalized follow-up plan, according to the level of injury and

Figura 7. Long-term evolution.



function, and the associated conditions and complications. The general objectives include optimizing functions, preventing complications, and promoting the overall well-being of the patients.

2) Progressive hydrocephalus with intracranial hypertension requires the placement of a CSF shunt system. Stabilized hydrocephalus may not require the placement of a CSF shunt system.

3) Bladder functionality should be evaluated conservatively, initially through a simple urinary tract ultrasound, and post-void residual measurement can be performed. Routine bladder catheterization and early urodynamic study are not recommended.

4) Chiari II malformation, which presents other symptoms besides hydrocephalus, may require posterior fossa decompression surgery.

5) The tethered spinal cord syndrome may manifest over time through spinal deformity, acquired hip dislocation, worsening urinary or intestinal dysfunction, and worsening sensory-motor deficits (Table 1).

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Table 1. Follow-up recommendations.

EVALUATION AND EXAMS	DISCHARGE	6M	9M	12M	15M	18M	24M
TF cranial ultrasound	X	X		X		X	
Brain MRI				X			
Standing spine X-ray				X			
Renal and bladder ultrasound with post-void residual measurement	X	X	X	X	X	X	X
CONSULTATIONS	**DISCHARGE**	**6M**	**9M**	**12M**	**15M**	**18M**	**24M**
Urology		X		X			X
Neurosurgery	X	X		X		X	
Ped. Neurology	X	X		X			X
PM&R		X	X	X	X	X	X
Orthopedics		X		X			

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Correspondence: Dra. Catalina Vaz Ferreira.
E-mail: catalinavazferreira@gmail.com

Data availability
The dataset supporting the results of this study is NOT available in open-access repositories.

Author contribution
All authors of this manuscript contributed to its conception and critical revision, and approved the final version for publication.

Catalina Vaz Ferreira, ORCID 0000-0002-9724-7047.
Paula Couchet, ORCID 0000-0001-6366-3731.
Mario Moraes, ORCID 0000-0002-5174-2405.
Gonzalo Costa, ORCID 0000-0002-6143-8490.
Alfredo Cerisola, ORCID 0000-0003-1277-2828.