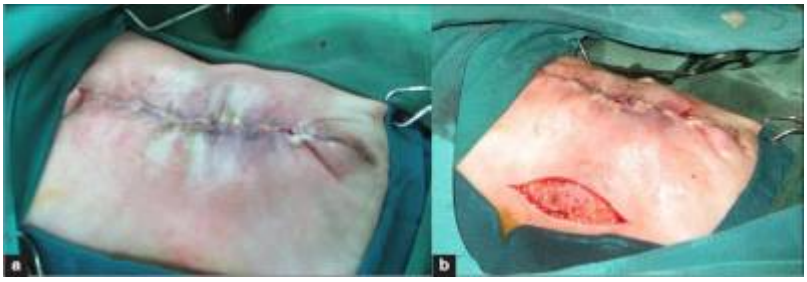
Myelomeningocele repair

- Functional Outcomes of Minimally Invasive Percutaneous Surgical Correction of Late-Presenting Severely Deformed Foot in a Patient with Myelomeningocele
- The repair of large meningomyelocele defects using perforator artery-based non-island fasciocutaneous flaps
- Endoscopic assessment of ventricular anomalies diagnosed by MRI in hydrocephalus associated with myelomeningocele
- PRENATAL REPAIR OF SPINA BIFIDA USING UTERINE WOUND RETRACTOR: A SINGLE-CENTER COHORT STUDY
- Differences in brain development and need for CSF diversion based on MMC level: Comparison between prenatal and postnatal repair
- Benefits and complications of fetal and postnatal surgery for open spina bifida: systematic review and proportional meta-analysis
- Isolation and characterization of human cKIT positive amniotic fluid stem cells obtained from pregnancies with spina bifida
- Presentation and management of neural tube defects in the middle belt of Ghana



Surgical repair and closure of myelomeningocele (MMC) defects are important and vital, as the mortality rate is as high as 65%-70% in untreated patients. Closure of large MMC defects is challenging for pediatric neurosurgeons and plastic surgeons.

The closure of the skin defect in myelomeningocele (MMC) repair is an essential step that determines the quality of the surgical result. The success of surgical results is related to the decision to use the most suitable techniques, namely flaps or primary closure.

The Management of Myelomeningocele Study (MOMS) demonstrated that fetal myelomeningocele closure results in improved hydrocephalus and hindbrain herniation when compared to postnatal closure.

Chiari type 2 malformation patients are now operated on with the first detectable symptom or evidence of a syrinx, and yet medullary dysfunction from the Chiari II malformation remains the leading cause of death in myelomeningocele treatment today. Our knowledge of the natural history of the untreated conditions and the increased safety of the operation has made surgical intervention a much more viable option for this group of patients [1)].

Classification

Myelomeningocele repair can be classified based on several criteria, including timing, approach, and technique. Here's a structured classification:

1. Based on Timing of Repair Prenatal (Fetal) Repair: Performed in utero before birth, usually between 19-26 weeks of gestation. Postnatal Repair: Performed within the first 48 hours of life to prevent infection and further neural damage.

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2. Based on Surgical Approach Open Repair (Standard Approach) Traditional surgical technique where the lesion is closed directly after birth. Minimally Invasive Fetal Repair Includes fetoscopic repair with a small incision in the uterus to reduce maternal and fetal risks.

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3. Based on Techniques for Closure Primary Closure: Direct closure of the defect without tissue expansion. Skin Flap Reconstruction: Utilized in large defects where primary closure is not feasible. Duraplasty and Fascia Closure: Reinforcement of the dural layer to protect the neural elements. Muscle Flap Reconstruction: Used when additional tissue coverage is needed. Skin Grafting: In extensive defects where local tissue is insufficient.

Fetoscopic Myelomeningocele Repair

Fetoscopic Myelomeningocele Repair.

Open fetal surgery

Open fetal surgery

Key concepts

Critical goals:

1) free placode from dura (to avoid tethering)

2) Water-tight dural closure

3) skin closure (can be accomplished in essentially all cases). Closure does not restore any neurologic function

● timing goal:surgical closure with latex-free setup ideally ≤36 hours after birth.

● helpful tips: start at normal dura, open as wide as the defect, trim placode if necessary to close

dura, undermine skin to achieve closure (avoid trapping skin →dermoid tumor).

● post-op Cerebrospinal fluid fistula usually means a shunt is required.

Timing

Myelomeningocele repair timing.

General principles

Prevent desiccation – keep the exposed neural tissue moist. Use latex-free environment (reduces development of latex allergy, as well as attack by maternal antibodies that may have crossed through the placenta). Do not allow scrub solutions or chemical antimicrobials to contact neural placode. Do not use monopolar cautery. At every point during the closure, avoid plac- ing tension on the neural placode.

Multiple layer closure is advocated, 5 layers should be attempted, although occasionally only 2 or so layers may be closed. There is no evidence that multiple layer closure either improves neurologic function or prevents later tethering, but there is a suggestion that when tethering does occur, it may be easier to release when a previous multilayered closure was performed. Silastic does not prevent adherence in series with long follow-up (>6 yrs), and may even render untethering procedures more difficult.

Begin by dividing the abnormal epithelial covering from the normal skin. The pia-arachnoid may be separated from the neural tissue. The placode is folded into a tube and the pia-arachnoid is then approximated around it with 7–0 suture (absorbable suture, e.g. PDS, may make future re-operation easier). It often helps to start with normal dura above, and then work down. The dura can then be isolated around the periphery and followed deep to the spinal canal superiorly. The dura is then also formed into a tube and approximated in a water-tight closure. If the dura cannot be closed, the placode may be judiciously trimmed. The filum terminale should be divided if it can be located. The skin is then mobilized and closed. Dermoid tumors may result from retained skin during the closure, but alternatively dermoids may also be present congenitally.

If there is a kyphotic deformity, it is repaired at the same sitting as the MM defect closure. The kyphotic bone is rongeured, and 2–0 Vicryl is used to suture the adjacent bones. Some surgeons use a brace post-op, some do not.

Post-op management

1. keep patient off all incisions

2. bladder catheterization regimen

3. daily OFC measurements

4. avoid narcotics (midbrain malformation renders these patient more sensitive to respiratory

depression from narcotics)

5. if not shunted

a) regular head U/S (twice weekly to weekly)

b) keep patient flat to ↓ CSF pressure on incision

6. if a kyphectomy was done, use of a brace is optional (surgeon preference)

Issues

Late problems/issues include:

1. hydrocephalus: ALWAYS RULE OUT SHUNT MALFUNCTION when a MM patient deteriorates

2. syringomyelia (and/or syringobulbia)

3. Tethered cord syndrome as many as 70% of MM patients have a tethered cord radiographically (some quote 10–20%), but only a minority are symptomatic. Unfortunately there is no good test to check for symptomatic retethering (SSEPs may deteriorate,15 myelography may help)

a) scoliosis: early untethering of cord may improve scoliosis

b) symptomatic tethering may manifest as delayed neurological deterioration

4. dermoid tumor at the MM site: incidence ≈ 16%

5. medullary compression at foramen magnum, see symptomatic Chiari II malformation

6. use of growth hormone to increase stature is controversial

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Some prefer to place the shunt and close the defect in the same procedure, it reduces the risks inherent to exposure to anesthesia, reduces hospital stay, and related costs. If there is a suspicious of infection, they do not place the shunt on the same procedure [2].

Prenatal therapeutic strategies that interrupt progressive pathological processes offer an appealing approach for treatment of MMC. However, a thorough understanding of pathological progression of MMC is mandatory for appropriate treatment to be rendered [3].

Closure of the defect.

Complications

Myelomeningocele repair complications.

MOMS Trial

see MOMS Trial.

References

1)
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