Multiloculated Hydrocephalus management strategy

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The term "multiloculated hydrocephalus" refers to the presence of an isolated CSF compartment or compartments within the ventricular system that may progressively enlarge.

Multiloculated hydrocephalus (MLH) is the most difficult form of hydrocephalus to treat.

The incidence

Neonatal meningitis 0.13% to 2.24%.

Post neonatal meningitis hydrocephalus is over 31%.

MLH is variable 7% and 20% according to some series.

Classification

- **Multiloculated hydrocephalus:**
  - multiple cysts isolated by multiple intraventricular septations:
    - post infectious
    - post inflammatory,
    - post hemorrhage

- **Uniloculated hydrocephalus:**
  - single cyst inside the ventricular system

The clinical features of MLH are not specific and are often difficult to recognize:

- full fontanel, head enlargement
- psychomotor retardation,
- seizures
- papilledema, headaches, vomiting, and decreased consciousness.

The MLH usually arise in infants or children neurologically compromised by:

- Neonatal meningitis,
- Intraventricular haemorrhage
- Hydrocephalus.

The presenting symptoms are usually insidious, although some cases of sudden death are reported.

HOW TO DO THE DIAGNOSIS???

Diagnostic studies

- Ultrasoundography
- Computed tomography scan
- Magnetic resonance imaging
- Contrast CT ventriculography
**Ultrasonography:** is used for neonates and patients with open fontanel

Some authors have demonstrated its superiority over the CT scan

BUT, it is operator-dependant and it cannot be considered as a definitive preoperative diagnosis modality.

**Computed tomography scan:** shows disproportionate hydrocephalus, and can be used for screening of patients. But it is unable to identify communication or non-communication between cavities.


Contrast CT ventriculo-graphy

Contrast CT ventriculo-graphy, by the injection into the cavities of Metrozamide, allows direct visualization of sequestrated ventricular compartments.

This is an invasive technique that requires puncture of the different cysts.

Magnetic resonance imaging

MRI with gadolinium is the method of choice for patients with complex hydrocephalus.

The more widespread high-resolution MRI technique, in particular (CISS) MRI, or fiesta MRI are used for the diagnosis and preoperative evaluation of loculated hydrocephalus.

Treatment

The goal of treatment

- The control of hydrocephalus and signs and symptoms of increased intracranial pressure
- Simplify complex shunt
  - Replacing multiple shunts with a single shunt
  - Reducing shunt revision rate.
  - Avoiding implanting shunts if possible.
  - Decreasing operative morbidity & mortality

The significant neurological deficits are related to the primary CNS insult and cannot be reversed by treatment of the hydrocephalus.
Several operative approaches have been described:

- Multiple shunt placement,
- Multi-perforated ventricular catheter,
- Stereotactic aspiration,
- Craniotomy trans-callosal fenestration of intraventricular septations
- Endoscopic fenestration.

Whatever technique is preferred??
In loculated hydrocephalus CSF cell count and biochemistry can vary in different cavities.

If one of the CSF samples gives evidence of CSF infection, the infected shunt system should be removed and replaced with an external ventricular drainage.

An appropriate antibiotic treatment administered based on CSF cultures and antibiograms.

After resolution of the CSF infection, the most appropriate surgical technique should be selected.
Multiple Shunt Placement

Is the classical method for treating MLH with shunt placement in the isolated compartments in full expansion.

BUT with high rate of mechanical shunt failure, infection and high rate of mortality and morbidity

The medians shunt revision were 2.75 and 3.04 by patient and per year (1)

The mortality rate was over 54% with the remaining patients severely impaired (2)


Our illustrative case: 6 months old boy born with ruptured myelomeningocele and had developed meningitis. The IMR showed multiloculated hydrocephalus.

After 1 month he presented seizures and head enlargement. The CT scan showed the expansion of the left cyst.

He was treated by double shunt with Y connection.

VP shunt was placed in the right side.

With stabilization of his hydrocephalus.
Multi-perforated ventricular catheter,

The use of multi perforated ventricular catheter might obviate the need for multiple shunting.

But we can't proven that is reliable method.

We had operated one case by this technique and we had a good result

Stereotactic Aspiration

Stereotactic aspiration alone fails to create a large fenestration and to devascularize the septal walls, the risk of recurrence is high.

Mobile cysts with thick walls resist to the Stereotactic puncture


Stereotactic aspiration is considered to be a blind technique.
Craniotomy

**Craniotomy and trans-callosal fenestration** was first performed by Rhoton and Gomez in 1972

This approach was reviewed by Nida and Haines in 1993

**Following open surgery the shunt revision rate was reduced from 2.74 to 0.25 per patient per year.**


However, trans-callosal surgery is quite an invasive option and carries potential risks:

- venous infarction from sacrificing bridging veins,
- damage to the pericallosal artery, fornices, and subcortical nuclei.
- Subdural collections are frequent (because the cortical mantle is thinned by the hydrocephalus).
- the loss of CSF during cyst decompression leads to collapse of the ventricular walls


Endoscopic fenestration combines both advantages:

The minimal invasiveness of stereotactic fenestration,
The effectiveness of microsurgery

It is a useful approach to avoid the morbidity, the mortality, and the long recovery period associated with open surgery.

AND

- to insert a new shunt under direct vision,
- to perform a third ventriculostomy when needed.
- to simplify the treatment of loculated hydrocephalus, permitting the use of a single shunt to drain multiple compartments and sometimes avoiding or eliminating the need for shunt

1 year old girl with head enlargement and seizures the MRI and CT scan showed uniloculated hydrocephalus. She was treated at first by endoscopy with a large septotomy.
The success rate of neuroendoscopy

The conversion of multilocular to unilocular hydrocephalus, allowing simplification of the shunt system to a single ventricular catheter

Varied between 61.8% to 100% in the published series.

The keys to the success of endoscopic treatment are early diagnosis and early treatment.

Foda et al.: Endoscopic management of loculated hydrocephalus. Thesis submitted to the Faculty of Medicine University of Alexandria 2009.
## Complications

<table>
<thead>
<tr>
<th>Complications</th>
<th>CSF leak</th>
<th>CNS infection</th>
<th>Haemorrhage</th>
<th>Wound infection</th>
<th>Death</th>
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<tbody>
<tr>
<td>Lewis AJ et al 1995 34 cases</td>
<td>3%</td>
<td>3%</td>
<td></td>
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<td>0%</td>
</tr>
<tr>
<td>Valenzuela S and Trellez A 1999</td>
<td>5%</td>
<td>12%</td>
<td>4%</td>
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<td>Nowoslawka et al 2003</td>
<td>6%</td>
<td>9%</td>
<td></td>
<td></td>
<td>0%</td>
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<tr>
<td>Spennato et al 2004 (18 cases)</td>
<td>1 case</td>
<td>3 cases</td>
<td>1 thalamic hematoma</td>
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<td></td>
</tr>
<tr>
<td>M A. Eshra 2014 (14 cases)</td>
<td>2 cases</td>
<td></td>
<td></td>
<td>1 case</td>
<td>0%</td>
</tr>
<tr>
<td>El-Ghandour 2008 (24 cases)</td>
<td>8%</td>
<td>8%</td>
<td></td>
<td></td>
<td>0%</td>
</tr>
</tbody>
</table>

Prognosis and Outcome

Despite the progress in surgical technique the prognosis of these children remains poor.

**Nowadays mortality has decreased**

But many children have cognitive deficits which range from:
- profound psychomotor retardation
- to mild learning disability
- most are seriously affected.
Conclusion

✧ Complex hydrocephalus is a challenging problem in pediatric neurosurgery.
✧ Early diagnosis and treatment are the keys for a better prognosis,
✧ Therefore, we must be vigilant in dealing with infants with meningitis, intraventricular hemorrhage or premature..
✧ Multi-planar MR imaging is the preferred diagnostic modality.
Conclusion

- The definitive treatment is surgical, yet the approach remains controversial.
- Cyst fenestration is the main strategy of treatment, and it can be done by multiple strategies, with the aim to improve the management of complex hydrocephalus, and to reduce number of shunts and shunt revision rate.
- Nowadays the endoscopic treatment is the best option.
THANK YOU

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