

#### **Chiari Type 1 Malformation with Papilledema in Children**

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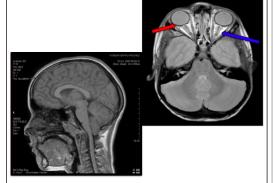
#### **Introduction**

Patients with Chiari type 1 malformation (CM1) may present primarily with visual symptoms.

The presence of papilledema without hydrocephalus, with associated visual troubles, indicating raised intracranial pressure (ICP), has been rarely reported in the literature.

The purpose of this study was to follow the clinical evolution after decompressive surgery and to examine the relationship between CM1 and papilledema.

# Encephalic MRI: CM1 and ICP



Red arrow: flattening of the posterior
 aspect of the globes
 Blue
 arrow: dilatation of the optic nerve sheaths

#### **Methods**

Series of 5 children, aged 5 to 14 years.

Visuals symptoms: headache, diminished visual acuity, diplopia or retro-orbital pain.

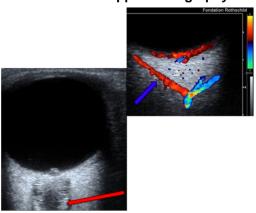
Magnetic resonance imaging (MRI): CM1, 7 to 12 mm of tonsillar herniation, dilatation of the optic nerve sheaths and flattening of the posterior aspect of the globes,

<u>Ophtalmological evaluation</u>: bilateral papilledema,

<u>Intraorbital Doppler sonography:</u> reversed flow in the superior ophthalmic vein in two children.

Surgery consisted in posterior fossa decompression with C1 laminectomy and duraplasty.

#### Intraorbital Doppler sonography



Red arrow: dilatation of the optic nerve
sheaths
Blue arrow:
reverse flow in the superior ophtalmic vein

#### **Results**

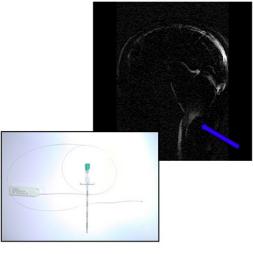
Following surgery (follow-up: 1 year to 5 years), all children improved clinically.

Papilledema completely resolve in only one case and improved in another. In the remaining three, papilledema was unchanged and intraorbital Doppler sonography demonstrated a persisting reversed flow of the superior ophthalmic vein, despite normalized intracranial pressure during postoperative monitoring with Codman® monitor in two children.

## **Conclusions**

Patients with CM1 and papilledema from increased ICP may benefit from suboccipital decompression.

# Postoperative encephalic flow MRI and Codman® monitor



Blue arrow: flow in the cisterna

Following surgery, the significance of a persisting papilledema despite clinical and radiological improvement, as we encountered in three of our patients, remains enigmatic.

### **Learning Objectives**

By the conclusion of the session, participants should be able to appreciate in children with CM1 the existance of

- 1) predominantly visual symptoms,
- 2) the spectrum of opthalmological diagnostic tools in this context,
- 3) the presence of papilledema without hydrocephalus,

and they should discuss persisting papilledema without raised ICP in the postoperative course.

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