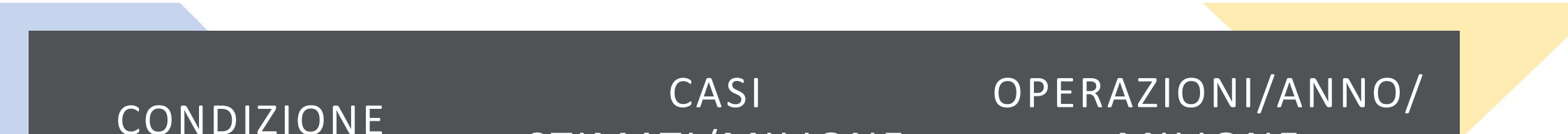




Chiari e Idrocefalo

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CONDIZIONE	CASI STIMATI/MILIONE	OPERAZIONI/ANNO/ MILIONE
Chiari asintomatico	~8000	0
Chiari sintomatico/siringomielia	~1.000	~15
Chiari sintomatico + idrocefalo	~70	~1

Wilkinson 2017 – Sadler 2020 – Greuter 2025



Persistenza di
idrocefalo dopo
l'intervento



Fattore Prognostico negativo

Associata a circa 40 volte meno probabilità di
avere un outcome favorevole!



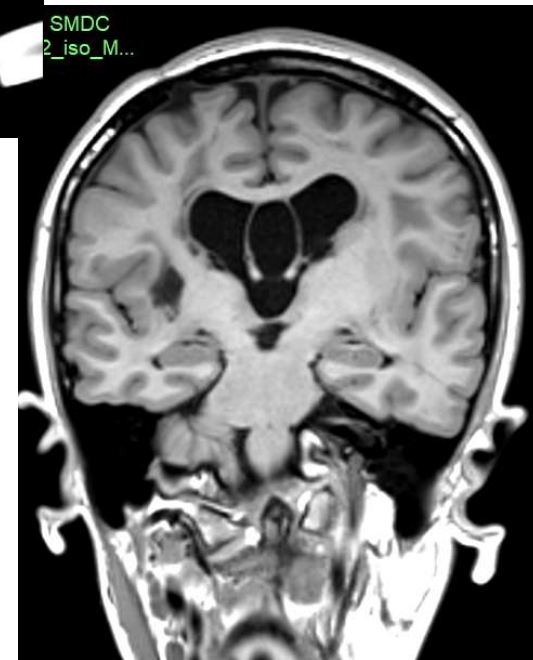
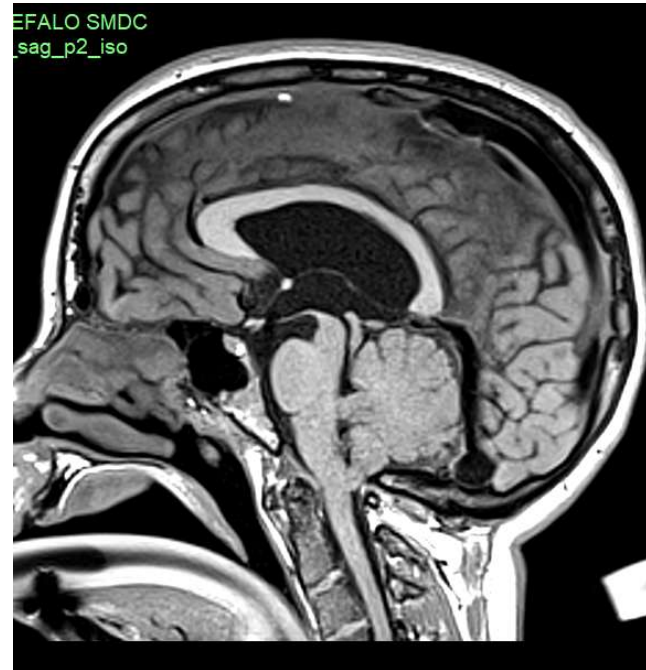
Long-term outcomes following posterior fossa decompression in pediatric patients with Chiari malformation type 1, a population-based cohort study

Victor Gabriel El-Hajj¹ · Erik Öhlén¹ · Ulrika Sandvik¹ · Jenny Pettersson-Segerlind¹ · Elias Atallah² · Pascal Jabbour² · Mohamad Bydon³ · David J. Daniels³ · Adrian Elmi-Terander^{1,4} · Erik Edström¹

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Domande cui rispondere

- Cosa trattare? 
 - L'idrocefalo?
 - Il Chiari?
- Come trattare l'idrocefalo?
- E se coesiste una siringomielia?





The management of pediatric Chiari I malformation with concomitant hydrocephalus: a multicenter retrospective analysis

Ladina Greuter^{1,2} · Tim Jonas Hallenberger¹ · Maria Licci^{1,2} · Ulrich-Wilhelm Thomale³ · Valentina Pennacchiotti³ · Andrea Bartoli⁴ · Mahmoud Messerer⁵ · Agne Andriuskeviciute⁵ · Marta Garvayo⁵ · Malte Ottenhausen⁶ · Verena Fassel⁶ · Conor Mallucci⁷ · Arthur R. Kurzbuch⁷ · Giuseppe Cinalli⁸ · Giuseppe Mirone⁸ · Christopher Chandler⁹ · Sanjeev Bassi⁹ · Martina Messing-Juenger¹⁰ · Katharina Lutz^{10,11} · Dieter Class¹² · Jonathan Roth¹³ · Shlomi Constantini¹³ · Carla Richetta¹³ · Raphael Guzman^{1,2,14} · Jehuda Soleman^{1,2,14}

patients with hydrocephalus, where the dilemma of “what came first - the chicken or the egg” remains: Is the hydrocephalus caused by a narrowing of the foramen magnum due to the CIM or is the CIM caused by the hydrocephalus pushing the tonsils downwards?



Chiari type I and hydrocephalus

Luca Massimi^{1,2,3} · Giovanni Pennisi² · Paolo Frassanito² · Gianpiero Tamburrini^{1,2} · Concezio Di Rocco³ · Massimo Caldarelli^{1,2}

Childs Nerv Syst (2019) 35:1701–1709

1703

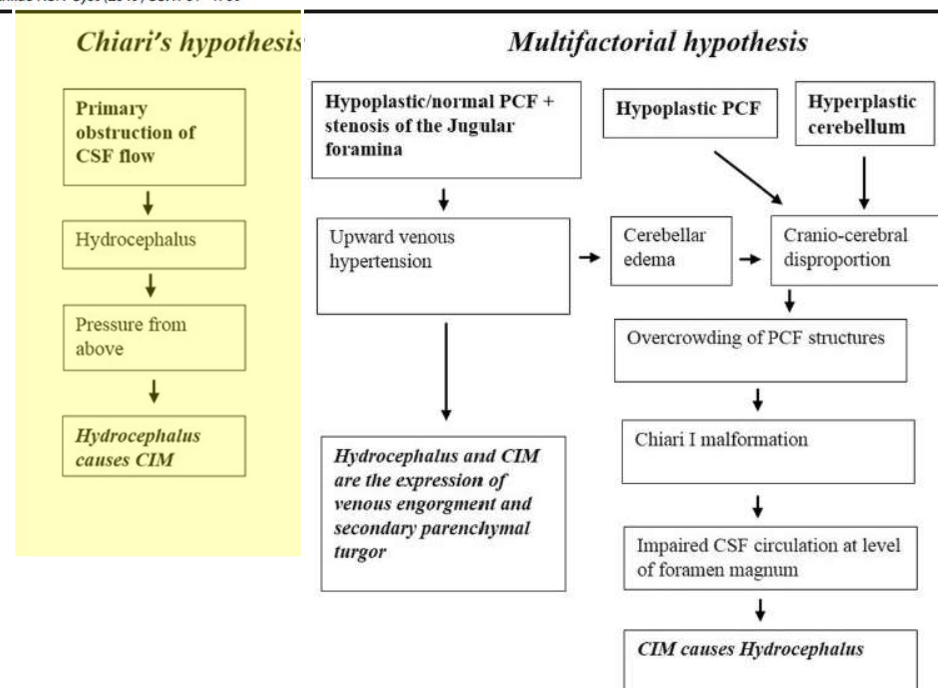


Fig. 1 Pathophysiological hypotheses (modified from 18)

Velo aracnoideo: membrana sottile che ricopre l'outlet del IV ventricolo

Presente in subset di pazienti con Chiari I

Causa di idrocefalo ostruttivo



«Velo»





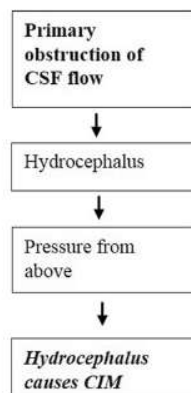
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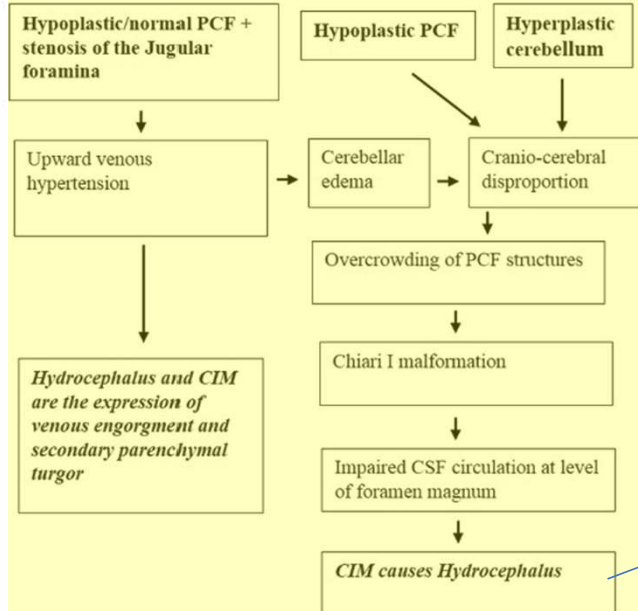
Childs Nerv Syst (2019) 35:1701–1709

17

Chiari's hypothesis



Multifactorial hypothesis



movements [31, 80]. The obstruction of the foramen magnum would account for the compression of the veins, which lose their ability to distend to compensate an increased intracranial pressure. Subsequently, according to the unifying hypothesis for hydrocephalus, Chiari malformation, and syringomyelia proposed by Williams on 2008, the hydrocephalus could be a condition of edema of brain depending on the venous insufficiency [79]. Such a failure of the venous drainage could act as a transient “pseudotumor cerebri” (raised ICP without ventricular enlargement) that, if severe and repeated, would contribute to the hydrocephalus as well as to the rare case of abrupt clinical deterioration in CIM patients [4, 8, 22, 38, 82]. Unfortunately, this venous insufficiency is hard to demonstrate radiologically or experimentally.

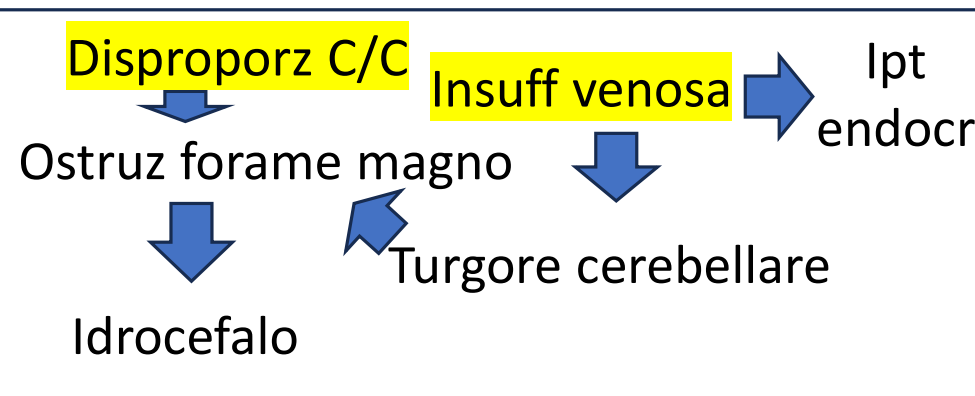


Fig. 1 Pathophysiological hypotheses (modified from 18)

Opzioni Terapeutiche: Trattamento dell'Idrocefalo

Strategia Gerarchica

- **PRIMA** trattare l'idrocefalo se sintomatico
- **POI** valutare se la sintomatologia da Chiari persiste

Endoscopic Third Ventriculostomy (ETV)

Prima linea per idrocefalo ostruttivo (stenosi acquedotto, outlet 4° ventricolo).

Tassi di successo: circa 80%

Vantaggi: minore tasso di complicanze, preserva anatomia naturale.

Derivazione Ventricolo-Peritoneale (VP Shunt)

Indicata se idrocefalo comunicante o ETV fallisce.

Considerazioni: valvole programmabili + dispositivo anti-sifone.

Complicazioni: overdrainage, infezione, necessità di revisioni (47-48%).

ETV: COSA ASPETTARSI?

Table 2

Measurement	Pre-ETV	Post-ETV	P value
Presence of Chiari I (absolute frequency)	12/12	11/12	
Degree of tonsillar herniation in cm (median and IQ range)	1.14 (0.91–1.49)	0.94 (0.73–1.53)	0.1
Evan's index (median and IQ range)	0.4 (0.34–0.43)	0.36 (0.31–0.39)	0.005
Diameter III ventricle in cm (median and IQ range)	1.35 (1.12–1.52)	0.76 (0.63–0.95)	0.003
Presence of syrinx (absolute frequency)	7/12	6/12	
Length of the syrinx in body levels (median and IQ range)	5 (1–18)	1 (0–4)	0.052
Transverse diameter of the syrinx in cm (median and IQ range)	0.75 (0.34–1.12)	0.32 (0–0.46)	0.03



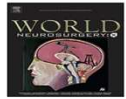
Rapporti fra Chiari, Siringa ed ETV



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Endoscopic third ventriculostomy for the management of children with cerebrospinal fluid disorders, ventriculomegaly, and associated Chiari I malformation



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MC, Siringomielia, Pressione endocranica, Idrocefalo

sono in un
equilibrio
mediato da



**Outflow
venoso**

**Circolazione liquorale fra
ventricoli e spazi subaracnoidei
cerebrali e spinali**

Identificare il pattern fisiopatologico:

Idrocefalo primario → trattare prima idrocefalo (ETV/Shunt)

Chiari primario → considerare FMD ± previo trattamento idrocefalo

Pattern simultaneo → approccio individualizzato

Selezione procedura per idrocefalo:

Ostruttivo → ETV preferibile

Comunicante → Shunt VP

Fallimento ETV → Shunt VP

Algoritmo Terapeutico Decisionale

1. Valutare tipo di idrocefalo (ostruttivo vs comunicante) Se possibile!!!

↓ Se **ostruttivo** → ETV prima linea

↓ Se **comunicante/non trattabile** → VP shunt

2. ETV riuscita? / VP shunt posizionato?

↓ Sì → Follow-up: RMN cine, controllo sintomi

↓ No → Riconsiderare VP shunt o revisione ETV

3. Sintomi da Chiari persistono?

↓ Sì → Decompressione forame magnum ± duraplasty

↓ No → Follow-up regolare

4. Post-op: Follow-up a 3-6 mesi con RMN

Valutare ectopia, flusso CSF, siringomielia

Grazie

