



Indicazioni e trattamento chirurgico in età pediatrica

Paola Peretta

Neurochirurgia Pediatrica

Ospedale Infantile Regina Margherita

Torino

Age as a predictor of reoperations and complications in surgically managed pediatric Chiari malformation type I

Thomas Johnstone, BS,¹ Maria Isabel Barros Guinle, BA,¹ Laura M. Prolo, MD, PhD,² and Gerald A. Grant, MD²

¹Department of Neurosurgery, Stanford University School of Medicine, Stanford, California; and ²Department of Neurosurgery, Duke University, Durham, North Carolina

Child's Nervous System (2024) 40:1443–1448
<https://doi.org/10.1007/s00381-023-06273-9>

RESEARCH



Natural history of Chiari I malformation with syrinx and dilatation of the central canal in the pediatric population: the CHEO experience

Maria Fernanda Dien Esquivel¹ · Neetika Gupta¹ · Christian Alfred O'Brien² · Vid Bijelić³ · Nick Barrowman³ · Nagwa Wilson¹ · Albert Tu⁴

Foramen Magnum Decompression for Chiari Malformation Type 1: Is There a Superior Surgical Technique?

Venetia Giannakaki¹, Jack Wildman^{1,4}, K Thejasvin², Georgios Pexas³, Justin Nissen¹, Nicholas Ross¹, Patrick Mitchell¹

Acta Neurochirurgica (2024) 166:460
<https://doi.org/10.1007/s00701-024-06332-3>

RESEARCH



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

Received: 24 September 2024 / Accepted: 25 October 2024 / Published online: 18 November 2024
© The Author(s) 2024

ORIGINAL ARTICLE



Pediatric Chiari I malformation: novel and traditional measurements associated with syrinx and surgery

Maria F. Dien Esquivel^{1,2} · Neetika Gupta^{1,2} · Nagwa Wilson^{1,2,4} · Christian Alfred O'Brien^{1,5} · Maria Gladkikh¹ · Nick Barrowman⁴ · Vid Bijelić⁴ · Albert Tu^{1,3,4}

Received: 30 December 2021 / Accepted: 3 August 2022 / Published online: 12 August 2022
© The Author(s), under exclusive licence to Springer-Verlag GmbH Germany, part of Springer Nature 2022

Surgical Outcomes in Chiari 1 and Chiari 1.5 Malformation Treated by Posterior Fossa Reconstruction: A Comprehensive Analysis of 110 Pediatric Cases and Literature Review

Maria A. Poca^{1,2,3,*} · Diego Lopez-Bermeo^{1,2} · Dulce Moncho^{2,4} · Alex Ferre^{2,4,5} · Angel Sanchez-Montañez⁶ · Olga Mestres^{2,3} · Sandra Galve⁷ and Juan Sahuquillo^{1,2,3}

Pediatric Chiari Malformation Management WFNS Spine Committee Recommendations

Ricardo Gepp, MD,^a June Ho Lee, MD,^b Jutty Parthiban, MD,^c Francesco Costa, MD,^d Fernando Dantas, MD,^{e,f} Francisco Sampaio, MD,^g Zan Chen, MD,^h Joachim Oertel, MD,ⁱ Salman Sharif, MD,^j Jörg Klekamp, MD,^k Ricardo Botelho, MD,^l Mehmet Zileli, MD,^m and Óscar L. Alves, MD^{n,o}

Child's Nervous System (2025) 41:378
<https://doi.org/10.1007/s00381-025-07052-4>

RESEARCH



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 Pennacchietti³ · 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}

Pediatric Chiari Malformation Management

WFNS Spine Committee Recommendations

*Ricardo Gepp, MD,^a June Ho Lee, MD,^b Jutty Parthiban, MD,^c Francesco Costa, MD,^d
Fernando Dantas, MD,^{e,f} Francisco Sampaio, MD,^g Zan Chen, MD,^h Joachim Oertel, MD,ⁱ
Salman Sharif, MD,^j Jörg Klekamp, MD,^k Ricardo Botelho, MD,^l Mehmet Zileli, MD,^m and
Óscar L. Alves, MD^{n,o}*

The following questions were discussed by the expert's panel:

- 1.If concomitant scoliosis is present, can surgery avoid the progression of scoliosis in children with Chiari I with or without syringomyelia?
- 2.Can children with Chiari I associated with syringomyelia be treated only with osteoligamentar decompression without duraplasty?
- 3.In type 2 Chiari, can decompression alone be recommended?
- 4.Is cranial vault expansion recommended in pediatric Chiari malformation and craniosynostosis?
- 5.Can children with Chiari malformation play sports?

Recommendations

- ***Surgery before puberty slows the progression of scoliosis*** in children with CM1, and decompression of the foramen magnum is advised. If no other cause of scoliosis is found, children with Chiari type 1 without syringomyelia may benefit from surgery. (70% totally agree; 30% more than agree)
- ***Decompression of foramen magnum with duraplasty is recommended in children with CM type 1 associated with syringomyelia.*** (70% totally agree; 30% more than agree).
- In children with CM type 2, the recommendation is to perform urgently tailored decompression of foramen magnum and cervical canal after after hydrocephalus and valve dysfunction were ruled out. (40% totally agree; 50% more than agree; 10% agree).
- ***Cranial vault expansion is recommended*** in pediatric CM associated with craniosynostosis. (100% strongly agree).
- ***Children with CM type 1 can play sports*** as there is a low risk of clinical worsening. (100% strongly agree).

Conclusion

Pediatric age is a modifier for the management of Chiari malformation (CM). Prompt diagnosis and **appropriate decompressive surgery with duraplasty before puberty** are essential to mitigate the impact of the condition on the child's wellbeing. Increased awareness among health care professionals, timely access to specialized expertise in neurosurgical interventions are crucial, especially for type 2 CM patients that require urgent decompression of CVJ and cervical spine.

Quali sintomi/segni andare a ricercare?



Neurol Sci (2022) 43:1311–1326

1315

Table 1 Planning for CM1 in children: indications to surgery

1	In <i>asymptomatic</i> children with incidentally discovered, isolated CM1 and <i>no syringomyelia</i> , surgery is not indicated	Agreement: 94.1%
2	In asymptomatic children with incidentally discovered CM1 and syringomyelia, surgery is indicated in cases of <i>syrinx larger than 5–8 mm</i> , and smaller syrinx increasing in size	Agreement: 82.4%
3	In children with <i>epilepsy</i> and CM1, surgical treatment of CM1 does not improve the seizure disorder	Agreement: 94.1%
4	In children with CM1 and <i>cognitive</i> and/or <i>behavioral</i> disorders (such as autism) not able to correctly report their symptoms because of communication impairment, careful clinical and instrumental assessments are mandatory to detect symptoms and signs of cerebellar tonsils ectopia	Agreement: 97.1%
5	In children with CM1 and cognitive and/or behavioral disorders (such as autism) and no clear CM1 symptoms, surgery is not indicated to improve the clinical picture	Agreement: 91.2%
6	Surgical treatment is not decided on <i>MRI pattern</i> in asymptomatic children with tonsillar ectopia ≥ 20 mm without syringomyelia	Agreement: 88.2%

... individuare i sintomi dei nostri pazienti al fine di selezionare i bambini candidati alla chirurgia ...

Research Submission

Headache in Children With Chiari I Malformation

Irene Toldo, MD PhD; Marta Tangari, MD; Rodica Mardari, MD; Egle Perissinotto, MD;
Stefano Sartori, MD PhD; Michela Gatta, MD; Milena Calderone, MD; Pier Antonio Battistella, MD

However, 67-100% of patients with suboccipital headache were reported to benefit from surgery,^{22,24} whereas patients with frontal headache and patients with a long history of headache have experienced recurrent headache after surgery.^{9,22}

*... la **cefalea** è il sintomo più comune e va molto bene tipizzata, è molto preziosa la collaborazione con il neuropediatra*

Table 1 Planning for CM1 in children: indications to surgery

1	In <i>asymptomatic</i> children with incidentally discovered, isolated CM1 and <i>no syringomyelia</i> , surgery is not indicated	Agreement: 94.1%
2	In asymptomatic children with incidentally discovered CM1 and syringomyelia, surgery is indicated in cases of <i>syrinx larger than 5–8 mm</i> , and smaller syrinx increasing in size	Agreement: 82.4%
3	In children with <i>epilepsy</i> and CM1, surgical treatment of CM1 does not improve the seizure disorder	Agreement: 94.1%
4	In children with CM1 and <i>cognitive</i> and/or <i>behavioral</i> disorders (such as autism) not able to correctly report their symptoms because of communication impairment, careful clinical and instrumental assessments are mandatory to detect symptoms and signs of cerebellar tonsils ectopia	Agreement: 97.1%
5	In children with CM1 and cognitive and/or behavioral disorders (such as autism) and no clear CM1 symptoms, surgery is not indicated to improve the clinical picture	Agreement: 91.2%
6	Surgical treatment is not decided on <i>MRI pattern</i> in asymptomatic children with tonsillar ectopia ≥ 20 mm without syringomyelia	Agreement: 88.2%

... anche la **siringomielia** è un sintomo e la scelta del trattamento è strettamente connesso alle sue caratteristiche e alla clinica concomitante

Table 1 Planning for CM1 in children: indications to surgery

1	In <i>asymptomatic</i> children with incidentally discovered, isolated CM1 and <i>no syringomyelia</i> , surgery is not indicated	Agreement: 94.1%
2	In asymptomatic children with incidentally discovered CM1 and syringomyelia, surgery is indicated in cases of <i>syrinx larger than 5–8 mm</i> , and smaller syrx increasing in size	Agreement: 82.4%
3	In children with <i>epilepsy</i> and CM1, surgical treatment of CM1 does not improve the seizure disorder	Agreement: 94.1%
4	In children with CM1 and <i>cognitive</i> and/or <i>behavioral</i> disorders (such as autism) not able to correctly report their symptoms because of communication impairment, careful clinical and instrumental assessments are mandatory to detect symptoms and signs of cerebellar tonsils ectopia	Agreement: 97.1%
5	In children with CM1 and cognitive and/or behavioral disorders (such as autism) and no clear CM1 symptoms, surgery is not indicated to improve the clinical picture	Agreement: 91.2%
6	Surgical treatment is not decided on <i>MRI pattern</i> in asymptomatic children with tonsillar ectopia ≥ 20 mm without syringomyelia	Agreement: 88.2%

... epilessia, disturbi del comportamento, autismo vanno contestualizzati e valutati con il neuropediatra/NPI

Table 1 Planning for CM1 in children: indications to surgery

1	In <i>asymptomatic</i> children with incidentally discovered, isolated CM1 and <i>no syringomyelia</i> , surgery is not indicated	Agreement: 94.1%
2	In asymptomatic children with incidentally discovered CM1 and syringomyelia, surgery is indicated in cases of <i>syrinx larger than 5–8 mm</i> , and smaller syrinx increasing in size	Agreement: 82.4%
3	In children with <i>epilepsy</i> and CM1, surgical treatment of CM1 does not improve the seizure disorder	Agreement: 94.1%
4	In children with CM1 and <i>cognitive</i> and/or <i>behavioral</i> disorders (such as autism) not able to correctly report their symptoms because of communication impairment, careful clinical and instrumental assessments are mandatory to detect symptoms and signs of cerebellar tonsils ectopia	Agreement: 97.1%
5	In children with CM1 and cognitive and/or behavioral disorders (such as autism) and no clear CM1 symptoms, surgery is not indicated to improve the clinical picture	Agreement: 91.2%
6	Surgical treatment is not decided on <i>MRI pattern</i> in asymptomatic children with tonsillar ectopia ≥ 20 mm without syringomyelia	Agreement: 88.2%

... il quadro RMN va sempre contestualizzato con la clinica del paziente

Prima di approcciare chirurgicamente una M. di Chiari

... escludere la presenza di:

- IPERTENSIONE ENDOCRANICA

- E una causa di fallimento
- Approfondimento diagnostico con Angio-RMN e TC3D per escludere altre cause come: pseudotumor, idrocefalia o craniostenosi

- IPOTENSIONE LIQUORALE

- Valutare la clinica
- Effettuare RMN di encefalo e midollo con mdc

- INSTABILITA' CRANIO-CERVICALE

- Studio della dinamica della cerniera cervicale specialmente nei pazienti con Chiari 1,5

Prima di approcciare chirurgicamente una M. di Chiari

- Idrocefalia

- Craniostenosi

- Tethered cord

Table 3 Planning for CM1 in children: associated malformations

1	In symptomatic children with CM1 and hydrocephalus, it is recommended to treat hydrocephalus first to relieve raised intracranial pressure and to avoid post-operative complications. CM1 can be treated afterwards if symptoms do not disappear	Agreement: 90.9%
2	In CM1 children with non-syndromic craniosynostosis (sagittal or lambdoid synostosis, oxycephaly), the craniosynostosis is better treated before the CM1	Agreement: 100%
3	In infants with syndromic craniosynostosis and CM1, the surgery should first increase the cranial volume, with the proper vault remodeling for each syndrome	Agreement: 90.9%
4	In case of persistent CM1, in children with syndromic craniosynostosis already submitted to cranioplasty, the surgical approach depends on the syndrome, cranial volume obtained by previous operations, posterior fossa volume, and CM1 symptoms/associated syringomyelia	Agreement: 100%
5	In CM1 toddlers with polysynostosis and prevalent brachycephaly (and in selected cases in the other age classes), a posterior vault osteogenic distraction could be considered before performing a posterior fossa decompression	Agreement: 95.2%
6	Angio-RM or angio-CT is mandatory to rule out anomalous subcutaneous drainage before craniocervical decompression in CM1 children with syndromic craniosynostosis	Agreement: 90.9%
7	CM1 is rarely associated with dysraphisms*and tethered cord syndrome (*tethering of the medulla at any level due to split cord malformation, limited dorsal myeloschisis, retained medullary cord, terminal myelocystocele, conus lipomas, thickened, and fatty filum with a low-lying conus (below L3))	Agreement: 100%
8	In the rare occurrence of CM1 in children with tethered cord syndrome, the de-tethering procedure should be performed to prevent neuro-urological deterioration and has no influence on CM1.	Agreement: 95.2%
9	The association of CM1 with "occult tethered spinal cord" (a tethering syndrome with a specific urodynamic pattern with a conus normally positioned above L2) has never been demonstrated.	Agreement: 81.8%
10	The intradural section of the filum terminale in CM1 children is recommended just to treat tethered cord syndrome and it plays no role in the management of a possible CM1 syndrome.	Agreement: 100%
11	The extradural section of the filum terminale in CM1 children is not recommended either to treat tethered cord syndrome or for the management of a possible CM1 syndrome.	Agreement: 100%

Quale Tecnica Chirurgica per i pazienti pediatrici?

1318

Neurol Sci (2022) 43:1311–1326

Table 4 Surgery for CM1 in children: techniques



1	In symptomatic CM1 children without syringomyelia, the bony decompression of the posterior fossa alone could be performed for the low complication rate, if the family accepts the perspective of possible second surgery	Agreement: 80.9%
2	In CM1 children with syringomyelia, bony decompression + duraplasty is preferable	Agreement: 81.8%
3	The extent of the bony decompression of the posterior fossa should be wide on the foramen, always including C1 laminectomy, and never extended to C2 for the risk of CVJ instability.	Agreement: 86.4%
4	In CM1 without arachnoiditis, it is indicated to preserve the arachnoid membrane to avoid CSF leakage and delayed scarring	Agreement: 86.4%
5	Cerebellar tonsils coagulation/resection is indicated in cases of very low-lying tonsils and recurrent or residual syringomyelia	Agreement: 86.4%
6	Autologous and allograft dural patches are preferable to artificial graft; both are suitable, according to the surgeon's preference, while there are experiences suggesting to avoid the artificial ones	Agreement: 81.8%
7	A watertight suture helps preventing CSF leakage, by non-resorbable stitches, together with a strict muscle and soft tissue closure.	Agreement: 95.5%

Decompressione ossea: razionale

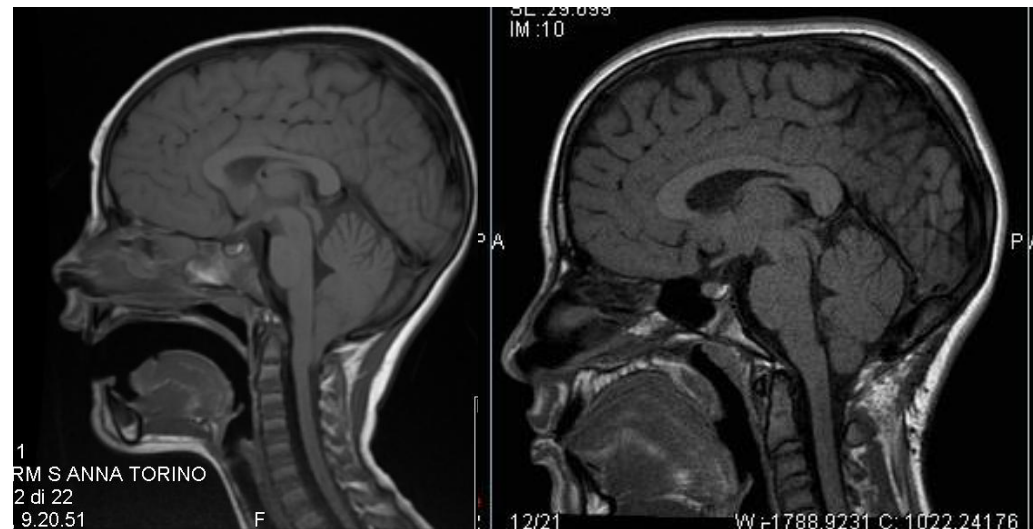
*Il razionale della decompressione ossea è quello di **ricreare indirettamente la cisterna magna** ...*



Decompressione ossea: razionale

La ragione del successo della decompressione ossea in età pediatrica potrebbe essere imputata al progressivo allargamento della fossa posteriore dovuta alla fisiologica crescita del cranio.

A differenza della popolazione adulta la decompressione ossea può favorire l'allargamento osseo e dare spazio alle strutture interne.



Decompressione ossea: indicazioni

La decompressione ossea è suggerita:

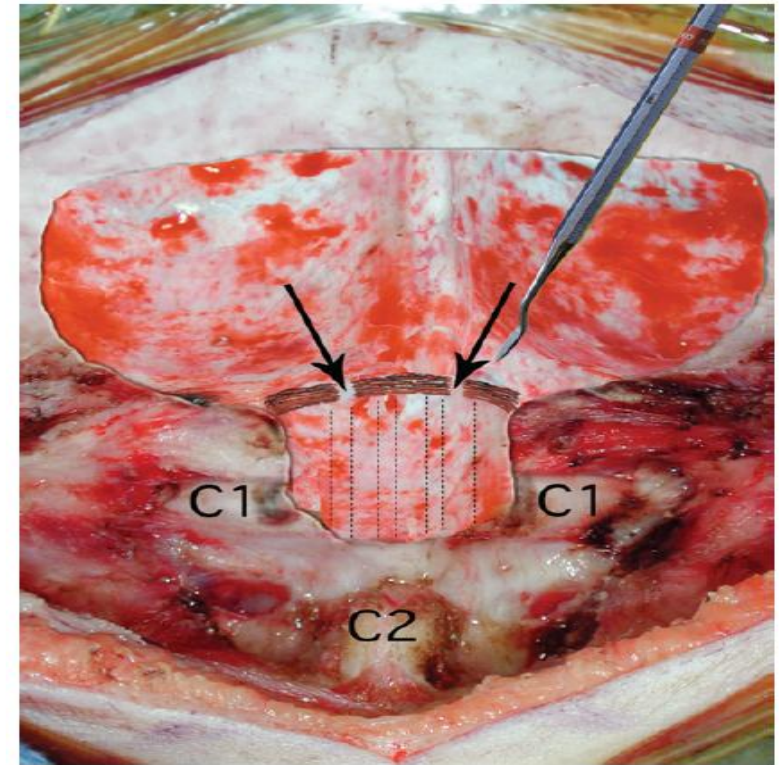
- in pazienti senza siringomielia
- in assenza di sintomi severi
- prendendo in considerazione il rischio di una possibile ricorrenza dei sintomi e la necessità di reintervenire.

... ma la strategia siringomielia = duroplastica non costituisce un assioma

Siringomieliie piccole o asintomatiche possono essere trattate con successo mediante la decompressione ossea.

Decompressione ossea: tecnica

- Apertura del forame magno 2,5/3 per 2,5/3 cm
- Rimozione dell'arco posteriore di C1
- Slaminamento della membrana atlo-occipitale
- La laminectomia di C2 va evitata per ridurre il rischio di instabilità
- Possibile la valutazione intraoperatoria ecografica della pulsatilità tonsillare per decidere la strategia chirurgica

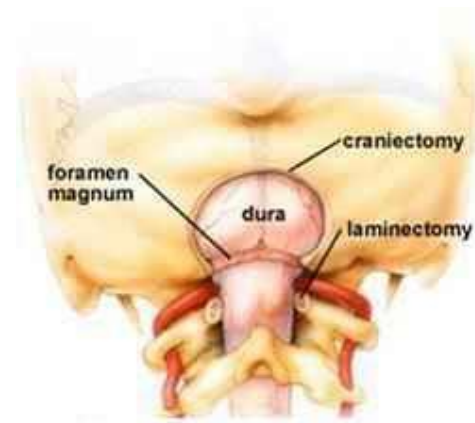


Decompressione ossea: tecnica

... la principale causa di fallimento è in relazione ad una decompressione non sufficiente soprattutto a livello del forame occipitale o anche a una ricrescita ossea

... attenzione a non decomprimere troppo per evitare la ptosi tonsillare

In alcuni pazienti potrebbe essere utile una TC 3D per valutare al meglio l'entità della decompressione e pianificare la chirurgia ... **APPROCCIO SU MISURA**



Apertura durale

1318

Neurol Sci (2022) 43:1311–1326

Table 4 Surgery for CM1 in children: techniques

1	In <i>symptomatic CM1 children without syringomyelia</i> , the bony decompression of the posterior fossa alone could be performed for the low complication rate, if the family accents the perspective of possible second surgery	Agreement: 80.9%
2	In CM1 children with syringomyelia, bony decompression + duraplasty is preferable	Agreement: 81.8%
3	The extent of the bony decompression of the posterior fossa should be wide on the foramen, always including C1 laminectomy, and never extended to C2 for the risk of CVJ instability.	Agreement: 86.4%
4	In CM1 without arachnoiditis, it is indicated to preserve the arachnoid membrane to avoid CSF leakage and delayed scarring	Agreement: 86.4%
5	Cerebellar tonsils coagulation/resection is indicated in cases of very low-lying tonsils and recurrent or residual syringomyelia	Agreement: 86.4%
6	Autologous and allograft dural patches are preferable to artificial graft; both are suitable, according to the surgeon's preference, while there are experiences suggesting to avoid the artificial ones	Agreement: 81.8%
7	A watertight suture helps preventing CSF leakage, by non-resorbable stitches, together with a strict muscle and soft tissue closure.	Agreement: 95.5%

Apertura durale: razionale e indicazioni

Il razionale della plastica durale è quello di ricreare direttamente la cisterna magna.

La **creazione diretta dello spazio cisternale** rende più rapido il ripristino della circolazione liquorale e di conseguenza il miglioramento clinico e l'effetto sulla siringomielia.

Rischio di reintervento, l'accordo su questo punto è alto (80%):

- La duroplastica con o senza coagulazione delle tonsille è preferita per evitare il rischio di reinterventi ed essere più efficaci nel risolvere la sintomatologia in tempi più brevi come la riduzione della siringomielia ...

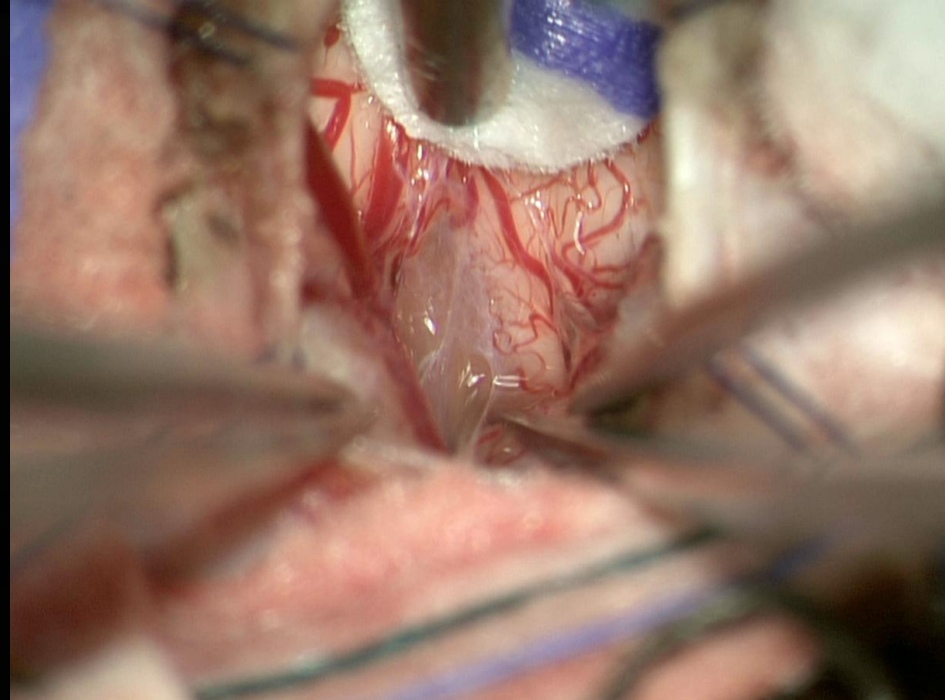
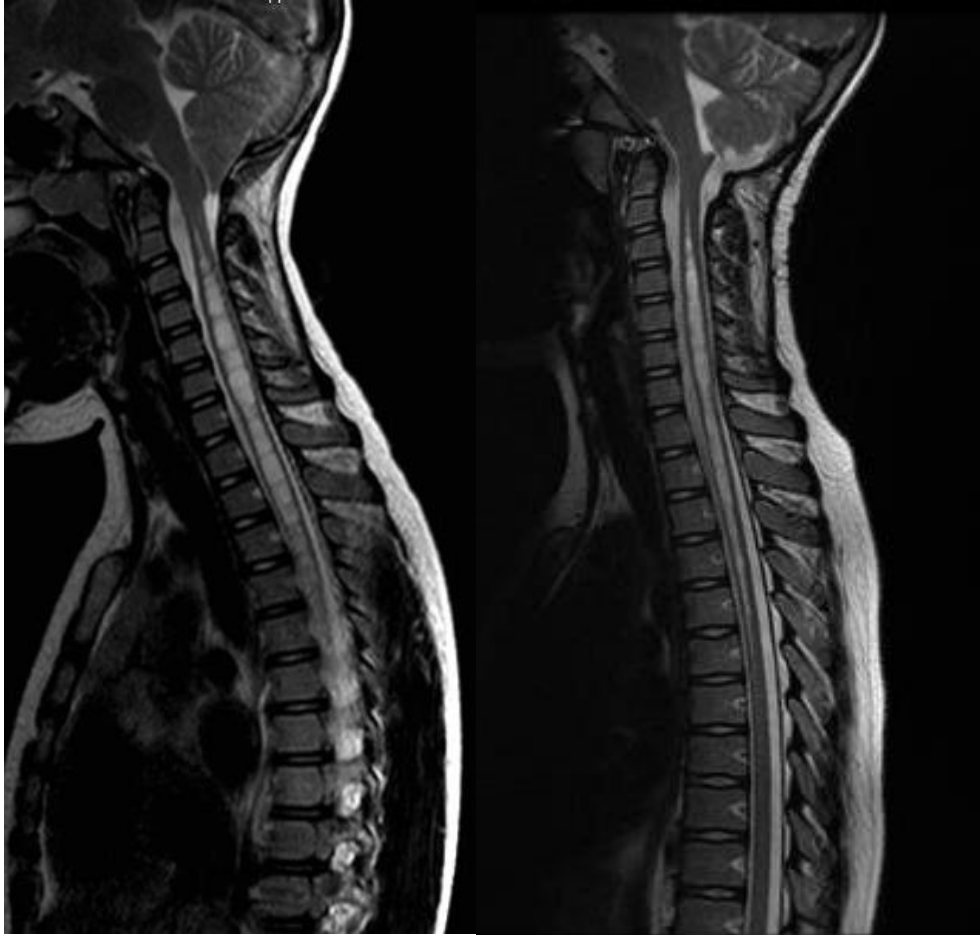
... ma, la duroplastica, si associa ad un più elevato rischio di fistola durale

Coagulazione delle tonsille

Table 4 Surgery for CM1 in children: techniques

1	In <i>symptomatic CM1 children without syringomyelia</i> , the bony decompression of the posterior fossa alone could be performed for the low complication rate, if the family accepts the perspective of possible second surgery	Agreement: 80.9%
2	In CM1 children with syringomyelia, bony decompression + duraplasty is preferable	Agreement: 81.8%
3	The extent of the bony decompression of the posterior fossa should be wide on the foramen, always including C1 laminectomy, and never extended to C2 for the risk of CVJ instability.	Agreement: 86.4%
4	In CM1 without arachnoiditis, it is indicated to preserve the arachnoid membrane to avoid CSF leakage and delayed scarring	Agreement: 86.4%
5	Cerebellar tonsils coagulation/resection is indicated in cases of very low-lying tonsils and recurrent or residual syringomyelia	Agreement: 86.4%
6	Autologous and allograft dural patches are preferable to artificial graft; both are suitable, according to the surgeon's preference, while there are experiences suggesting to avoid the artificial ones	Agreement: 81.8%
7	A watertight suture helps preventing CSF leakage, by non-resorbable stitches, together with a strict muscle and soft tissue closure.	Agreement: 95.5%

Duraplasica e Coagulazione delle tonsille



Ostruzione del IV° ventricolo da un
velo aracnoideo

Quali sono le cause di insuccesso?

- DECOMPRESSIONE OSSEA INSUFFICIENTE

- E' una delle cause di insuccesso. La TC3D permette di fare una corretta diagnosi

- FISTOLA LIQUORALE

- Predisporre all'infezione e al fallimento chirurgico per la formazione di aderenze

- ARACNOIDITE ... ma a volte anche prima della chirurgia

- Lisi delle aderenze, coagulazione delle tonsille ed eventuale posizionamento di uno stent tra IV vn e spazi perimidollari

Reintervento-Fallimento-Outcome

Table 5 Surgery for CM1 in children: outcomes, failure, re-intervention

Surgical outcomes		
1	The early surgical outcome is assessed at 6 and 12 months by clinical evaluation and whole neuraxis MRI; a cine-MRI may be helpful.	Agreement: 98.9%
2	Surgical failure is clinically defined by the persistence of Chiari symptoms 12 months post-surgery.	Agreement: 98.9%
3	Surgical failure is radiologically defined by persistence of low tonsils and unchanged syringomyelia at 24 months post-surgical MRI.	Agreement: 88.7%
Diagnosis and treatment of the main causes of surgical failure		
4	Insufficient bone decompression is one of the causes of failure; it is diagnosed by 3D-CT scan and the treatment is widening its extension.	Agreement: 82.4%
5	Posterior fossa arachnoiditis is one of the causes of failure: its treatment is arachnoidysis and/or tonsil resection; possible adjunctive procedures may be starting of the IV ventricle with perimodullary space.	Agreement: 87.9%
6	Postoperative CSF leakage is a predisposing factor for infections and surgical failure due to arachnoiditis.	Agreement: 97.1%
7	An intracranial hypertension (IHH) may be a cause of failure. When intracranial hypertension is present, angio-MRI for venous study and 3DCT scan should be performed to exclude other causes of raised ICP such as pseudotumor, hydrocephalus, or craniosynostosis.	Agreement: 97.1%
8	A CVJ instability is one of the causes of failure. To diagnose it, a dynamic CVJ study is indicated, especially in CM 1.5 patients.	Agreement: 88.2%
Surgical re-intervention		
9	In case of symptomatic CSF leakage, a new operation is necessary.	Agreement: 91.2%
10	In case of asymptomatic CSF collection, conservative management is indicated as the first option.	Agreement: 91.2%
11	In case of unresolved/increasing CSF collection despite conservative treatment, a diagnostic re-evaluation is necessary to decide on the surgical option.	Agreement: 94.1%
12	Asymptomatic children without syrinx with persistent tonsil descent should be followed up for at least 24 months to monitor the appearance of clinical symptoms.	Agreement: 82.4%
13	Asymptomatic children with persistent syrinx at MRI at 12 months follow-up should be strictly followed up, and re-operated if scoliosis and/or symptoms occur.	Agreement: 85.3%
14	Children with persistent symptoms and unchanged MRI (no tonsils ascent and absent flow) at 6 or 12 months follow-up should be re-operated on.	Agreement: 85.3%
15	Children with persistent symptoms and syringomyelia at MRI at 6 or 12 months follow-up should be re-operated on.	Agreement: 85.3%
16	In case of success of surgery, the long-term postoperative follow-up is performed by a clinical examination and whole neuraxis MRI for at least 10 years, or until the end of growth, with a timetable depending on clinical and MRI patterns.	Agreement: 88.2%

Reinterventi

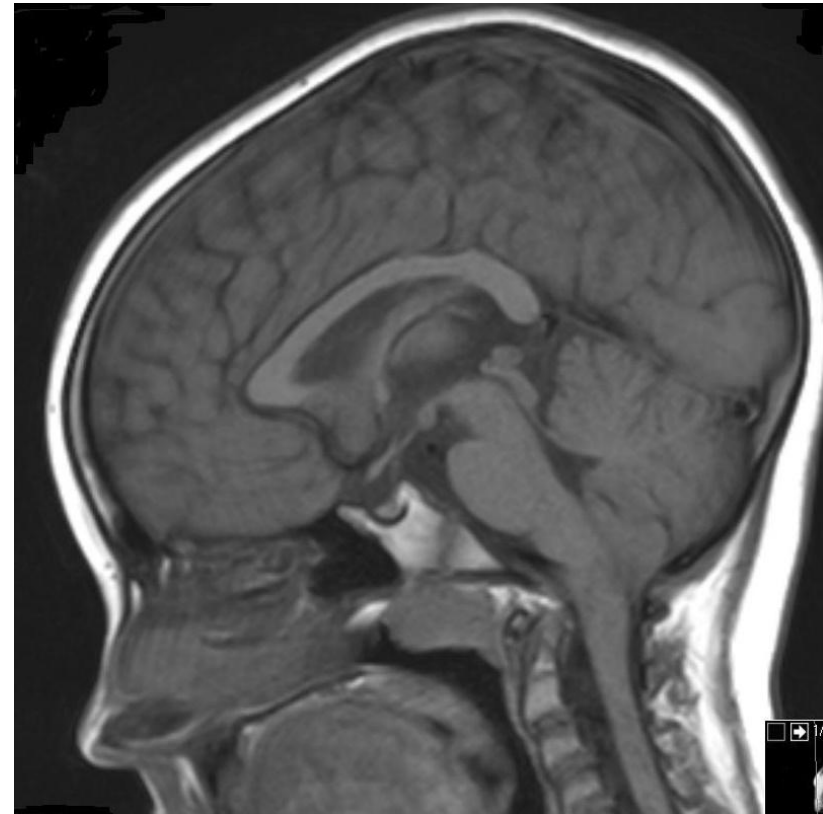
Il tasso di reinterventi e' del 7–15% ma può raggiungere il 50% nelle serie dove sono inclusi i pazienti con siringomielia

E' chiaro che lo scarso miglioramento dei pazienti affetti da siringomielia in alcuni studi potrebbe dipendere dal troppo breve follow-up. Infatti, nelle serie dove il follow-up è superiore ai 5 anni sono riportati buoni risultati anche in relazione alla riduzione/scomparsa della siringomielia.

Caso Clinico 1

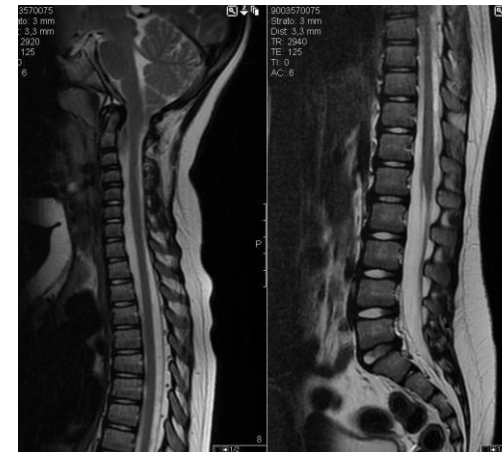
A.D.K, F 5 aa

- Giunge in DEA per frequenti episodi cefalalgici (tempie e nuca) prevalentemente mattutini, associati a dubbio episodio di apnea (riferito dalla madre) e deviazione del capo-collo verso dx
- Alla RMN encefalo riscontro di CM1 con erniazione delle tonsille cerebellari di circa 1,5 cm ed associata anomalia della giunzione occipito-cervicale.



Caso Clinico 1

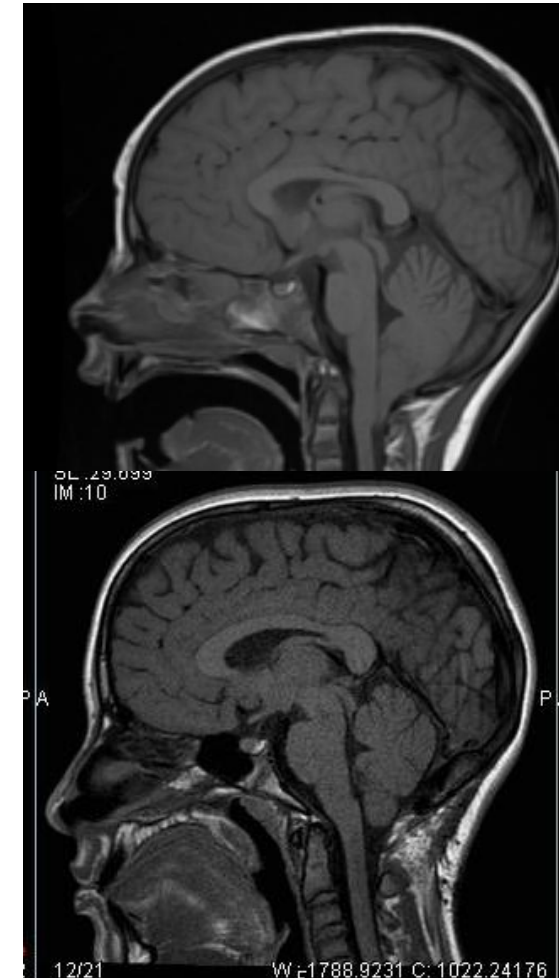
- TC giunzione: dismorfismo delle masse laterali con tendenza alla platibasia
- RX dinamico: non instabilità
- RMN del rachide in toto: di norma
- PESS e BAEP di norma
- Monitoraggio saturazione notturna: nella norma
- Valutazione NPI: paziente cefalgica. Terapia sintomatica. Miglioramento



Caso Clinico 2

M.M. 4 aa, LLA

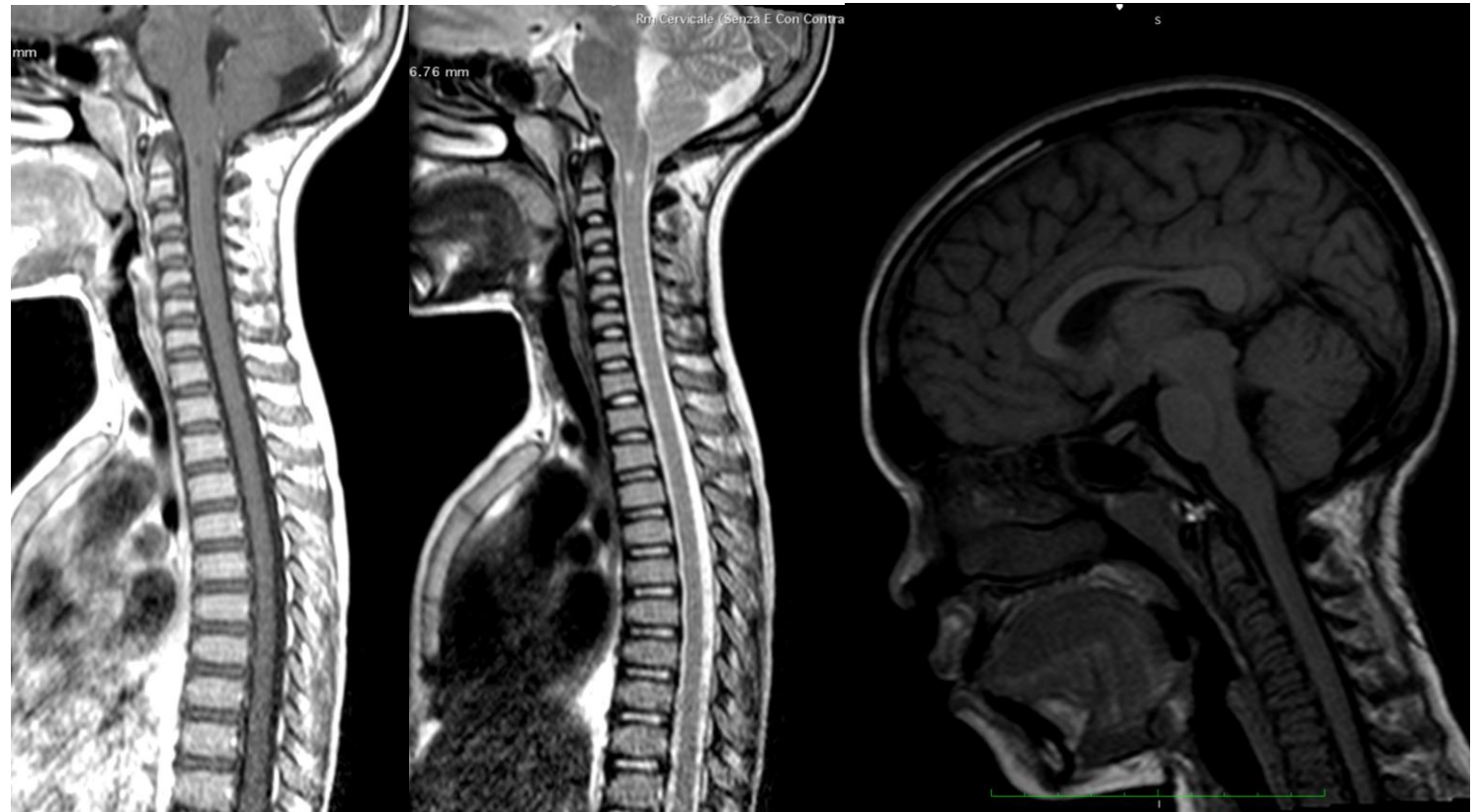
- frequenti episodi cefalgici nicali progressivamente ingravescenti non alleviate dalla terapia medica mirata.
- Alla RMN encefalo riscontro di CM1 con erniazione delle tonsille cerebellari di circa 1 cm, non siringomelia associata.
- Intervento chirurgico: decompressione ossea.
- Successivo miglioramento della sintomatologia e scomparsa della cefalea.



Caso Clinico 3

M. 6 aa,

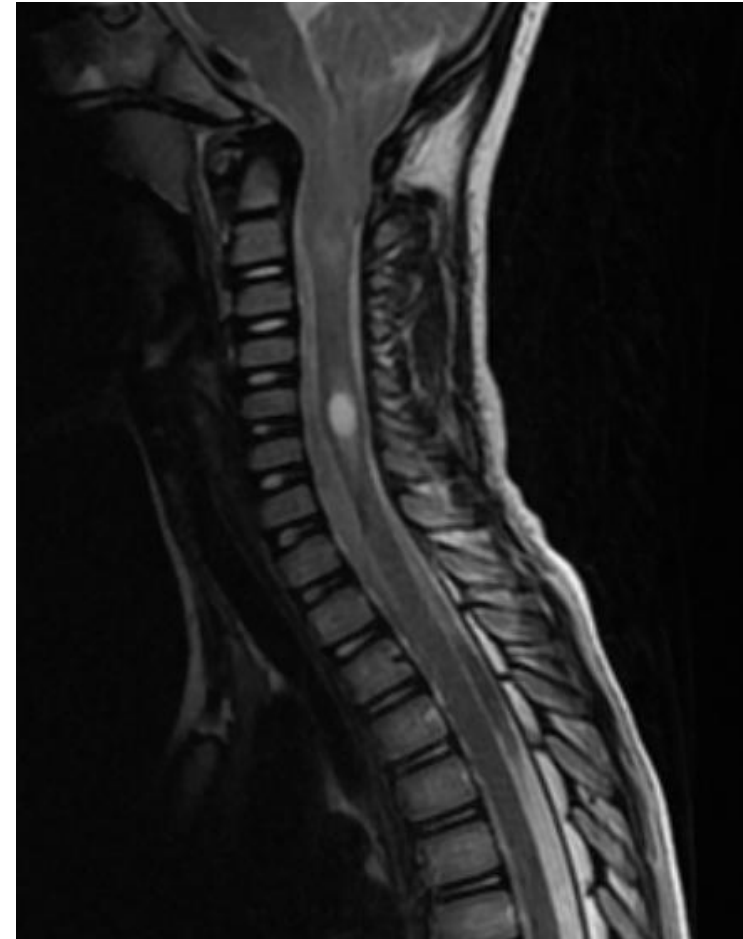
- Episodi cefalgici nucali progressivamente ingravescenti
- Singhiozzo
- Intervento chirurgico: decompressione ossea.
- Miglioramento della sintomatologia



Caso clinico 4

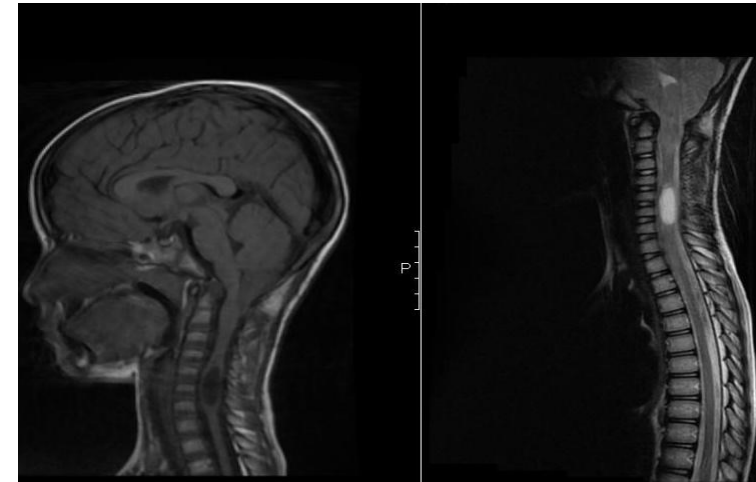
C.S. 5 aa e ½

- Episodi di cefalea frontale correlati a manovra di Valsalva
- RMN encefalo e rachide in toto: Chiari I (erniazione fino a C2), siringomielia C5-C6
- 1° intervento: solo decompressione ossea

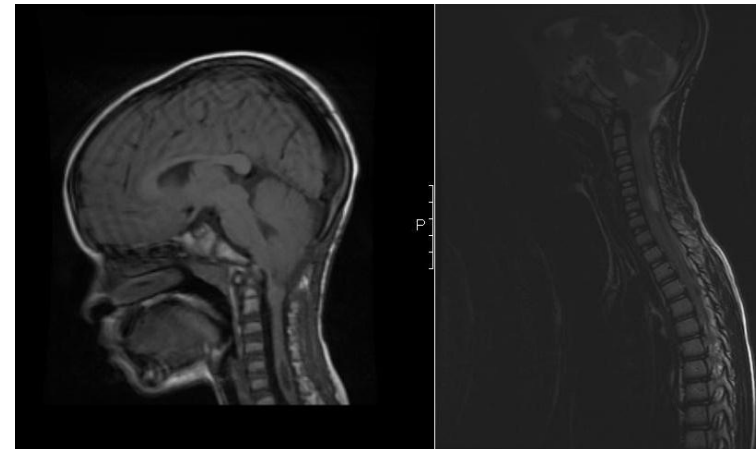


Caso Clinico 4

- RMN post 1° intervento:
incremento della siringomielia



- 2° intervento: coagulazione
delle tonsille e plastica durale
- Riduzione della siringomielia



... alcune riflessioni

- La Malformazione di Chiari è sempre più frequentemente diagnosticata in modo occasionale in bambini asintomatici.
- Abbiamo la responsabilità di non mettere etichette di malattia che possono condizionare la vita di questi piccoli pazienti e allo stesso tempo porre molta attenzione ad individuare i sintomi al loro manifestarsi ...

... una diagnosi accurata, clinica e neuroradiologica, porta ad una scelta chirurgica appropriata migliorando l'esito dell'intervento ...

... di concerto con un approccio multidisciplinare

Indicazione chirurgica

... la scelta chirurgica avviene in continuum decisionale in cui tutti gli elementi diagnostici e clinici vengono presi in considerazione ...

La multidisciplinarietà è una preziosa e indispensabile risorsa.



The Chiari's Road

Grazie per l'attenzione

Clinica

Diagnosi

Trattamento

