



Malformazione di CHIARI e SIRINGOMIELIA

diagnosi, trattamento, vita quotidiana

10-11 APRILE 2026

Seraphicum, Collegio Serafico di San Francesco,
Via del Serafico, 1 - ROMA



Cosa chiedere alla radiologia

Paola Sciortino

Neuroradiologia CDC AFFIDEA TORINO

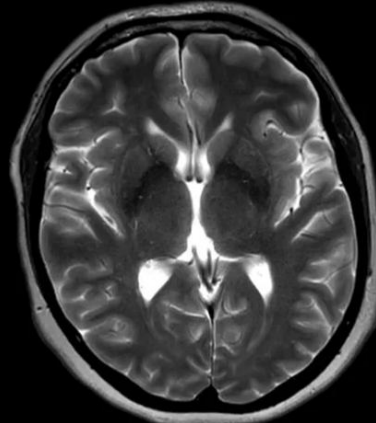
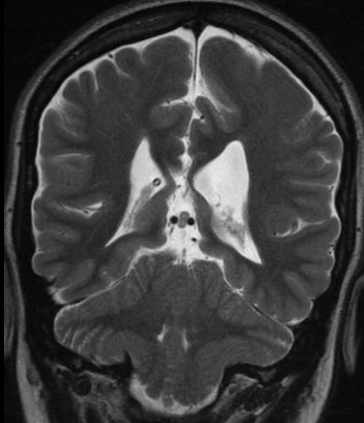
Cosa chiedere alla radiologia

- *Diagnosi e classificazione*
- *Monitoraggio nel tempo*
- **Supporto** alla Decisione Chirurgica
- *Follow-up post-chirurgico*
- *Fattori prognostici*



Protocollo MR

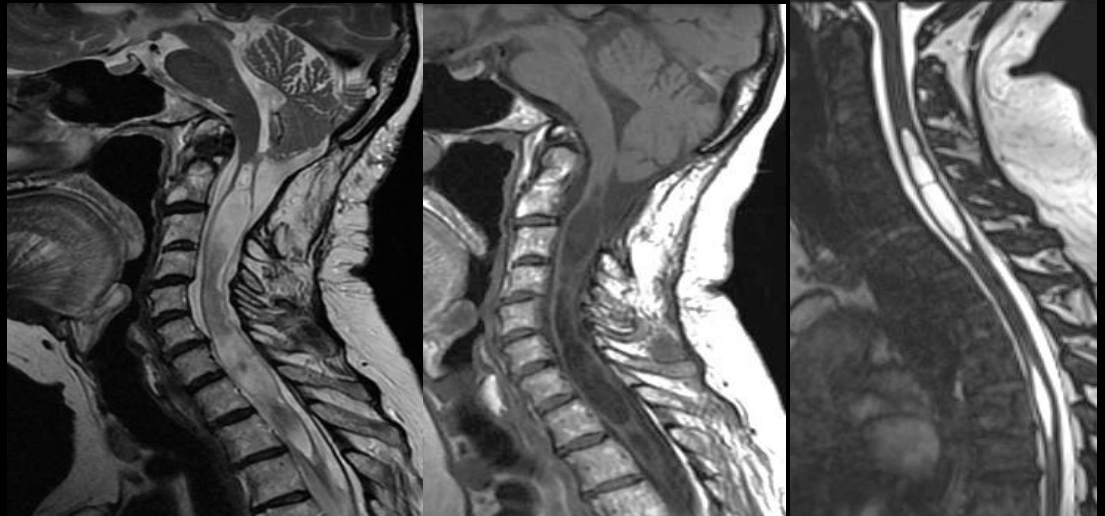
MR ENCEFALO COLONNA



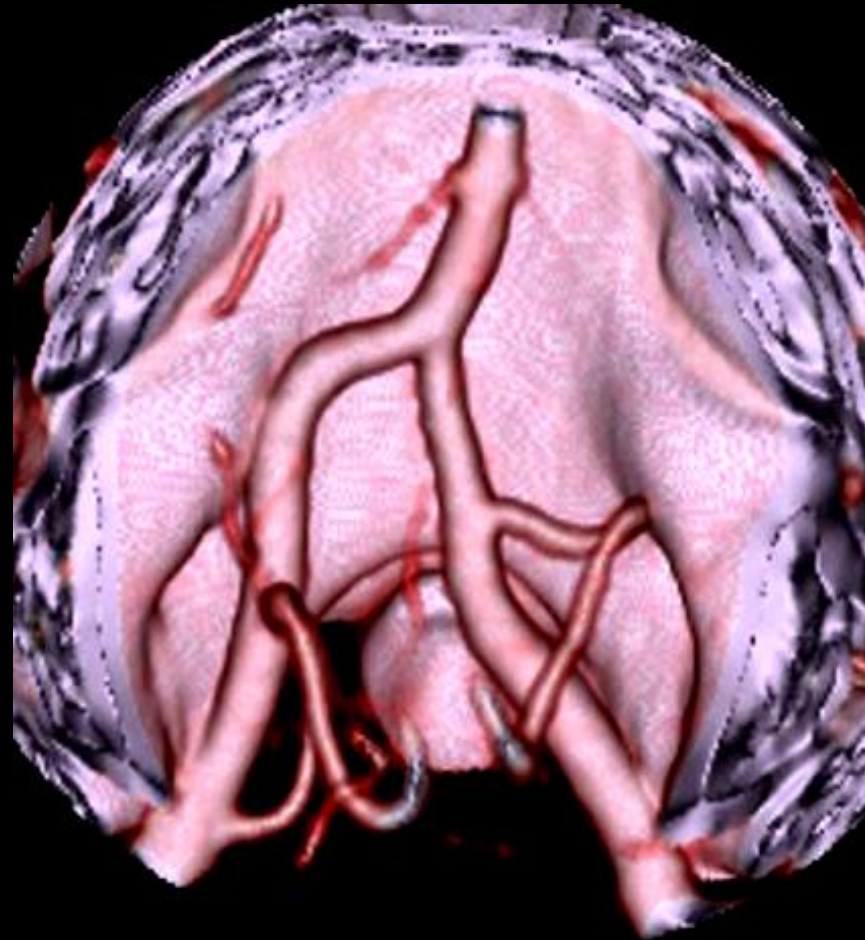
**Phase Contrast
Cine RM**

**Immagini assiali,
sagittali, coronali T1, T2
pesate**

Fiesta, Drive, Ciss 3D



TC e Angio TC



Diagnosi e classificazione

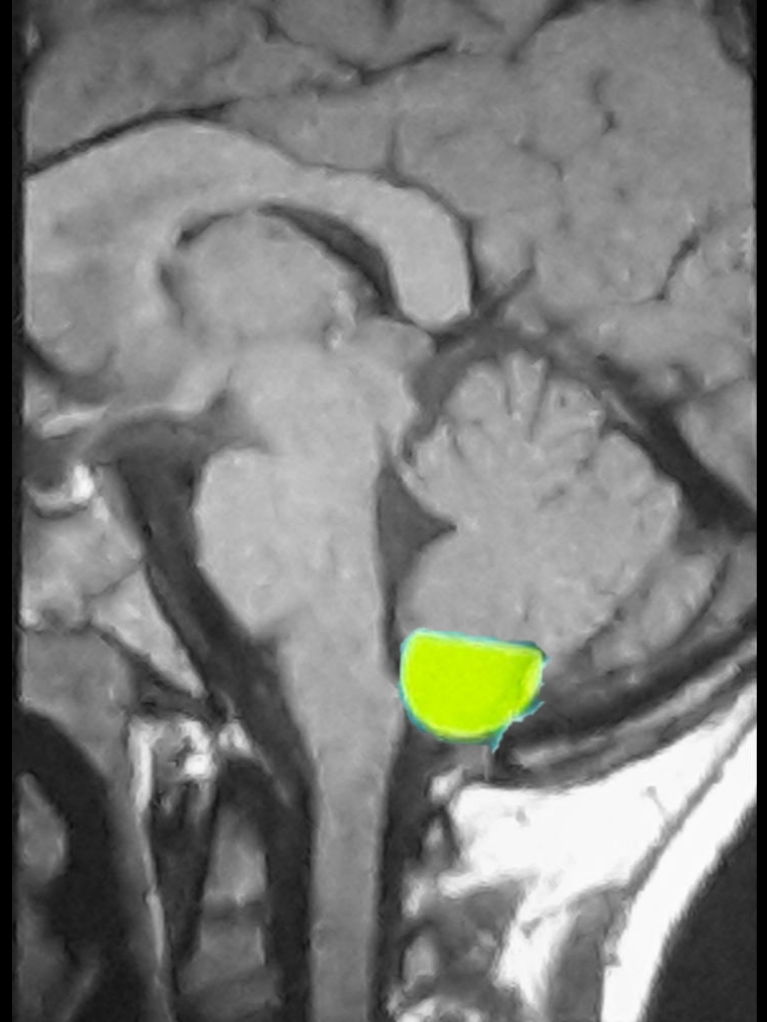
- **Identificazione dell'erniazione: rilevare la discesa delle tonsille cerebellari attraverso il forame magno**
- **Classificazione**
- **Diagnosi differenziale: distinguere la malformazione di Chiari da altre patologie per evitare errori diagnostici**
- **Studio del flusso del liquor attraverso la Cine-RM evidenziando ostruzioni a livello del forame magno**
- **Ricerca di condizioni associate: idrocefalo, midollo ancorato, scoliosi**
- **Siringomielia**

Identificazione dell'erniazione

Chiari **vs** **Normal**



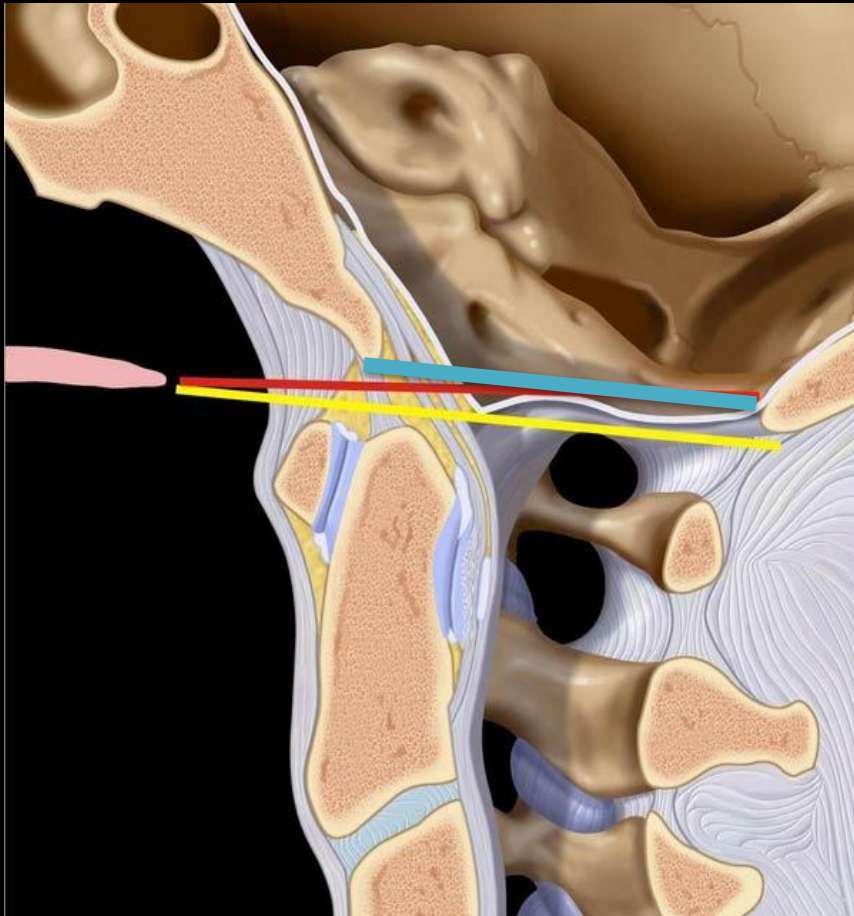
deformazione peg-like delle tonsille



morfologia arrotondata

Quantificazione dell'erniazione

Rilevare la discesa delle tonsille cerebellari attraverso il forame magno rispetto alla linea basion-opisthion = *McRae line*



McRae line



Sag. T1

Cerebellar tonsils herniation below the McRae line:

≥ 5 mm: CM1;

< 3 mm: physiological variation;

3-5 mm: borderline ectopia is definitely pathologic if at least one of the following findings is associated:

- peg-like deformation of the tonsillar profile
- presence of subarachnoid space crowding in the area of the cranio-cervical junction



Cerebellar tonsils herniation below the McRae line:

> 5 mm: CM1:

< 3mm: physiological variation:

3-5 mm: borderline ectopia is definitely pathological findings is associated:

- peg-like deformation of the tonsillar projection
- presence of subarachnoid space crowding at the junction



Cerebellar tonsils herniation below the McRae line:

≥ 5 mm: CM1;

< 3 mm: physiological variation;

3-5 mm: borderline ectopia is definitely pathologic if at least one of the following findings is associated:

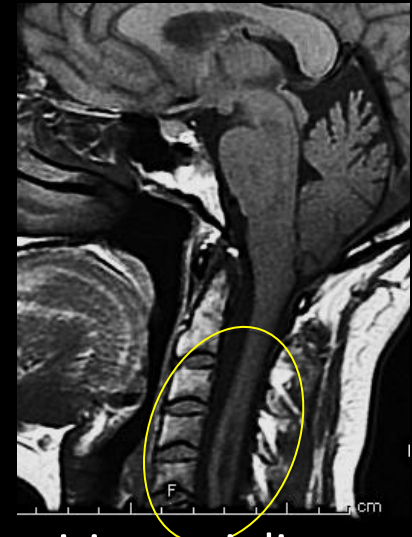
- peg-like deformation of the tonsillar profile
- presence of subarachnoid space crowding in the area of the cranio-cervical junction
- syrinx



deformazione
peg-like



affollamento degli spazi cisternali
della giunzione cranio cervicale



siringomielia

Classificazione

Neurological Sciences
<https://doi.org/10.1007/s10072-021-05347-3>

ORIGINAL ARTICLE

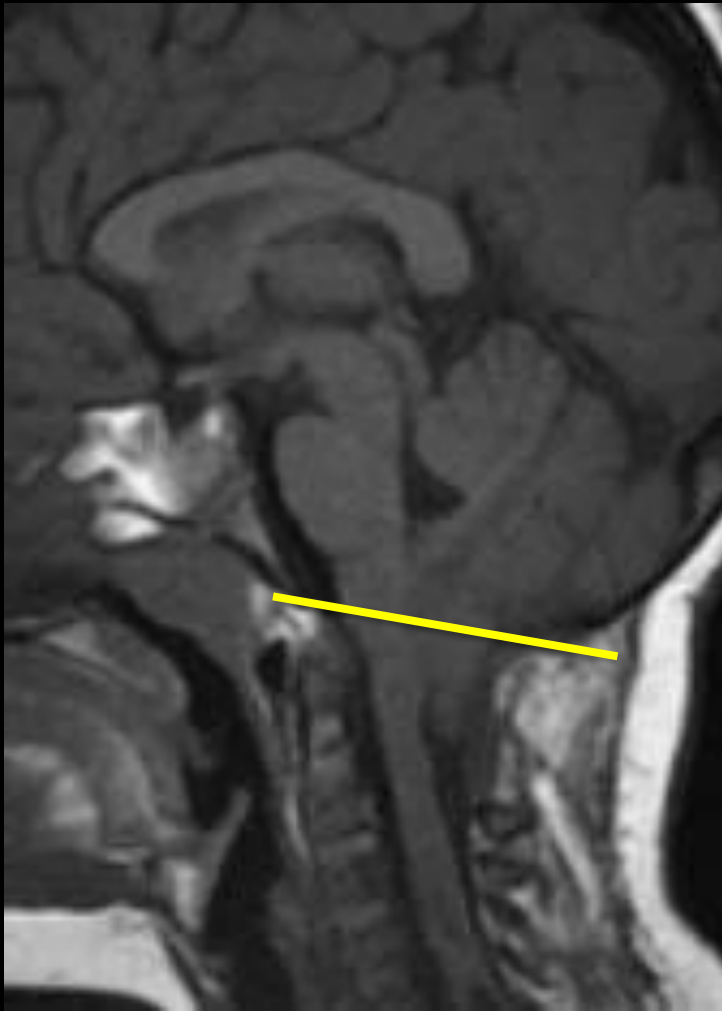


Diagnosis and treatment of Chiari malformation and syringomyelia in adults: international consensus document

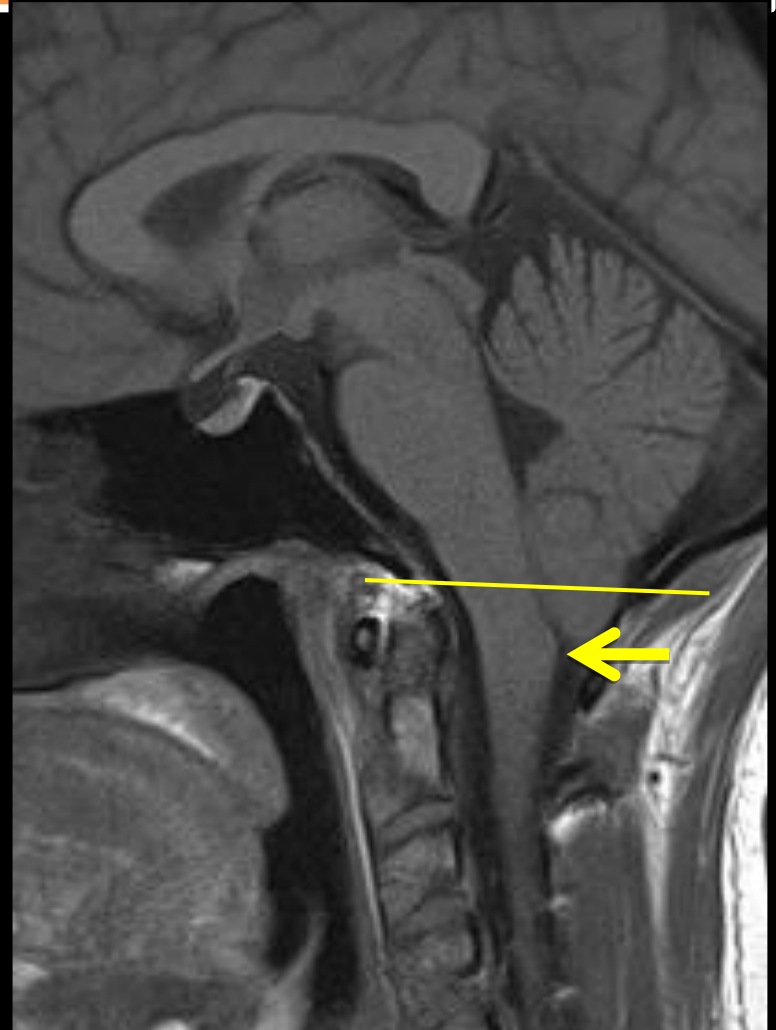
Palma Ciaramitaro^{1,2} • Luca Massimi³ • Alessandro Bertuccio⁴ • Alessandra Solari⁵ • Mariangela Farinotti⁵ • Paola Peretta⁶ • Veronica Saletti⁷ • Luisa Chiapparini⁸ • Andrea Barbanera⁴ • Diego Garbossa¹ • Paolo Bolognese⁹ • Andrew Brodbelt¹⁰ • Carlo Celada¹¹ • Dario Cocito¹² • Marcella Curone¹³ • Grazia Devigili¹⁴ • Alessandra Erbetta¹⁵ • Marilena Ferraris¹⁶ • Marika Furlanetto¹⁷ • Mado Gilanton¹⁸ • George Jallo¹⁹ • Marieta Karadjova²⁰ • Jorg Klekamp²¹ • Fulvio Massaro²² • Sylvia Morar²³ • Fabrice Parker²³ • Paolo Perrini²⁴ • Maria Antonia Poca²⁵ • Juan Sahuquillo²⁵ • Marcus Stoodley²⁶ • Giuseppe Talamonti²⁷ • Fabio Triulzi²⁸ • Maria Consuelo Valentini¹⁶ • Massimiliano Visocchi²⁹ • Laura Valentini¹⁷ • On behalf of the International Experts Jury of the Chiari Syringomyelia Consensus Conference,

- C1** **CM1:** paraxial mesoderm disorder, with abnormalities of the posterior cranial fossa (mostly small) and the consequent herniation of the cerebellar tonsils: **CM1-A:** with syringomyelia on MRI; **CM1-B:** without syringomyelia on MRI.
- C2** **CM2:** due to prenatal CSF hypotension always associated with myelomeningocele and, in some cases, with hydrocephalus and syringomyelia. Other types of intracranial defects (hypoplastic tentorium cerebelli, cranial lacunae, anomalies of the Sylvius aqueduct) may coexist (1).
- C3** **CM-1.5:** cerebellar tonsils and brainstem herniation (kinking) below the McRae line; defined “complex” if also skeletal deformities of craniovertebral junction and cervical spine (Klippel-Feil anomaly, atlanto-occipital fusion, basilar invagination, retroversion of the odontoid process) are present.
- C4** **CM-0:** obliteration of the cisterna magna (due to arachnoid adhesions) and/or volumetrically small posterior fossa, with cerebellar tonsils positioned at the foramen magnum and a syringomyelia in the cervical spinal cord.

CM 1 : erniazione delle tonsille isolata



**CM 1.5 : erniazione delle tonsille+
dislocazione del bulbo con basso
posizionamento dell'obex**





Diagnosis and treatment of Chiari malformation and syringomyelia in adults: international consensus document

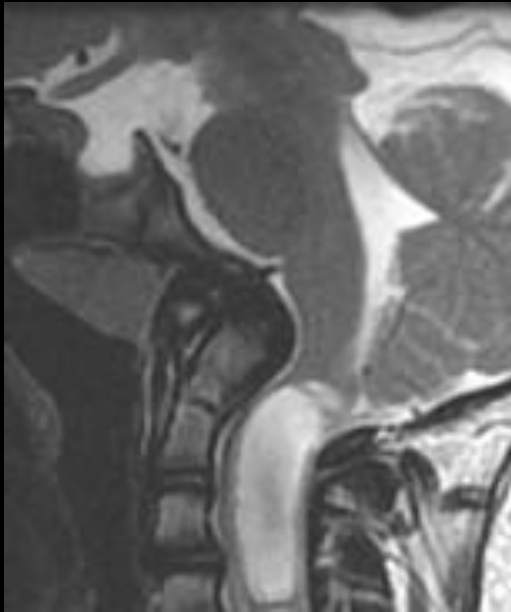
Palma Ciarra^{1,2} • Luca Massimi³ • Alessandro Bertuccio⁴ • Alessandra Solari⁵ • Mariangela Farinotti⁵ • Paola Peretta⁶ • Veronica Saletti⁷ • Luisa Chiapparini⁸ • Andrea Barbanera⁴ • Diego Garbossa¹ • Paolo Bolognese⁹ • Andrew Brodbelt¹⁰ • Carlo Celada¹¹ • Dario Cocito¹² • Marcella Curone¹³ • Grazia Devigili¹⁴ • Alessandra Erbetta¹⁵ • Marilena Ferraris¹⁶ • Marika Furlanetto¹⁷ • Mado Gilanton¹⁸ • George Jallo¹⁹ • Marieta Karadjova²⁰ • Jorg Klekamp²¹ • Fulvio Massaro²² • Sylvia Morar²³ • Fabrice Parker²³ • Paolo Perrini²⁴ • Maria Antonia Poca²⁵ • Juan Sahuquillo²⁵ • Marcus Stoodley²⁶ • Giuseppe Talamonti²⁷ • Fabio Triulzi²⁸ • Maria Consuelo Valentini¹⁶ • Massimiliano Visocchi²⁹ • Laura Valentini¹⁷ • On behalf of the International Experts Jury of the Chiari Syringomyelia Consensus Conference,

Table 1 Classifications and Definitions

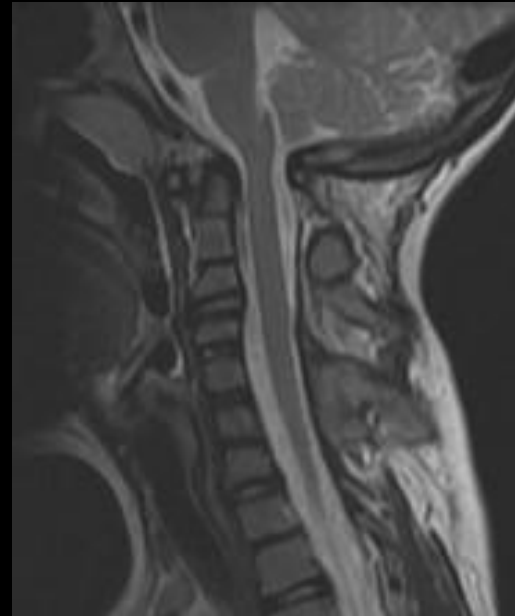
Chiari Malformation classification

- C1** **CM1:** paraxial mesoderm disorder, with abnormalities of the posterior cranial fossa (mostly small) and the consequent herniation of the cerebellar tonsils: **CM1-A:** with syringomyelia on MRI; **CM1-B:** without syringomyelia on MRI.
- C2** **CM2:** due to prenatal CSF hypotension always associated with myelomeningocele and, in some cases, with hydrocephalus and syringomyelia. Other types of intracranial defects (hypoplastic tentorium cerebelli, cranial lacunae, anomalies of the Sylvius aqueduct) may coexist (1).
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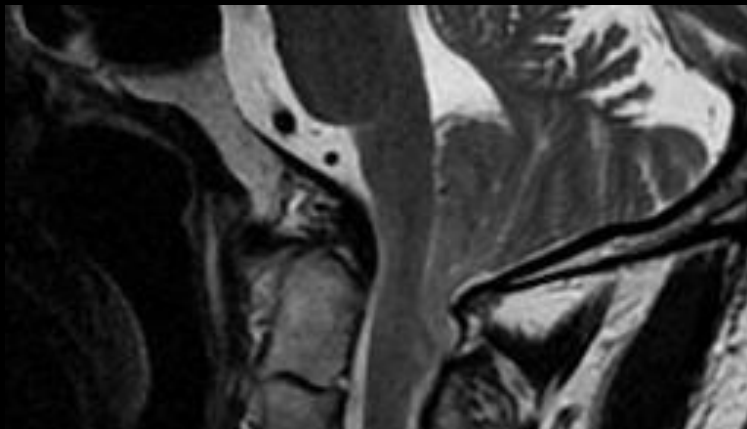
Anomalie della giunzione cranio-cervicale e del rachide cervicale



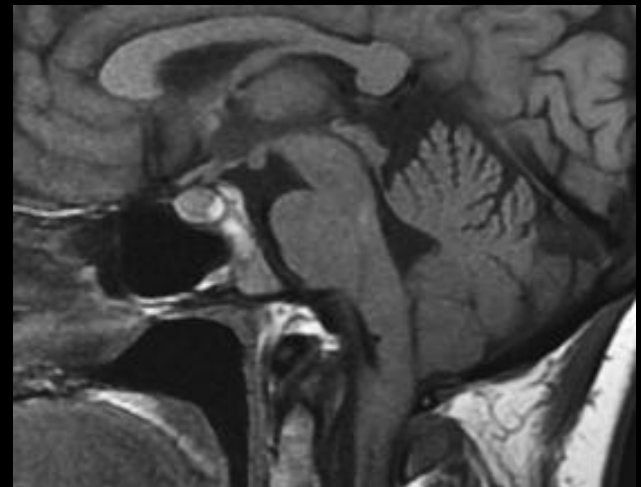
Dente retroflesso



klippel-feil

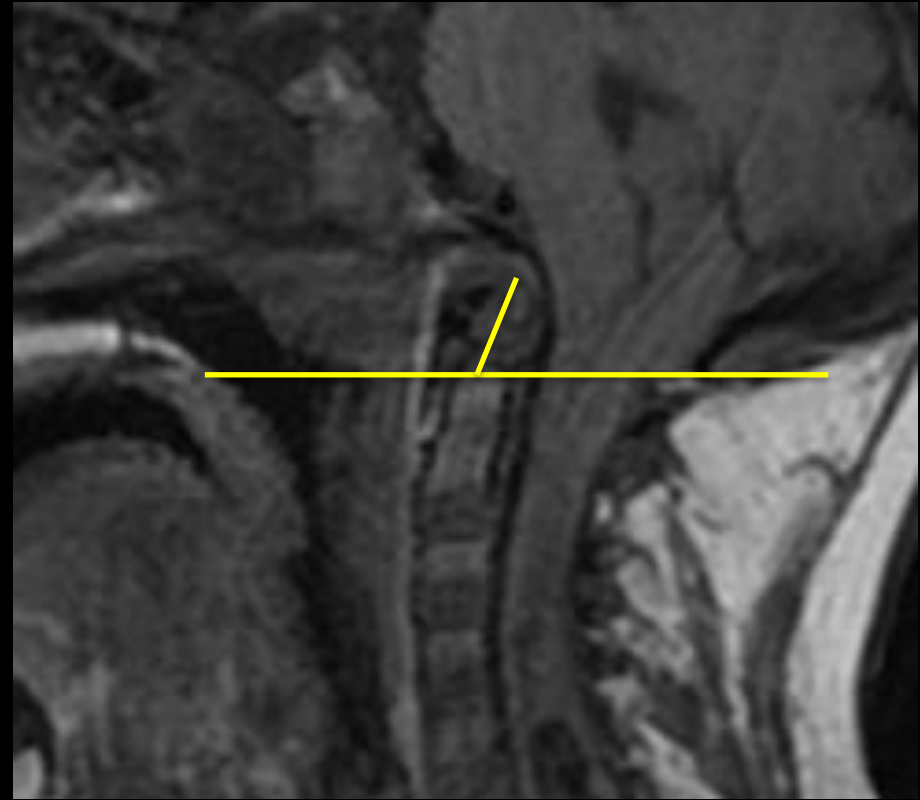
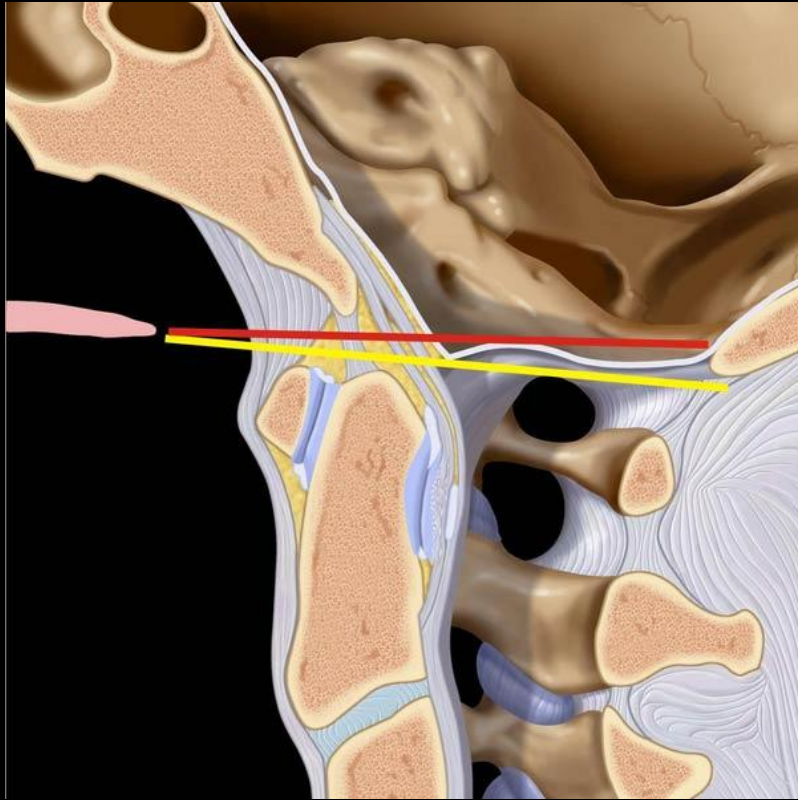


Assimilazione di C1



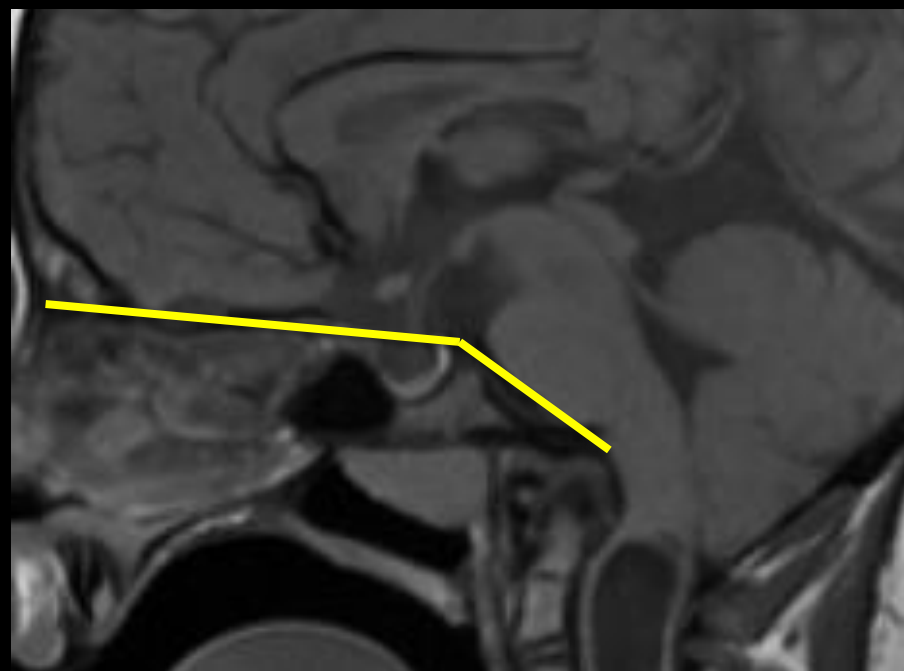
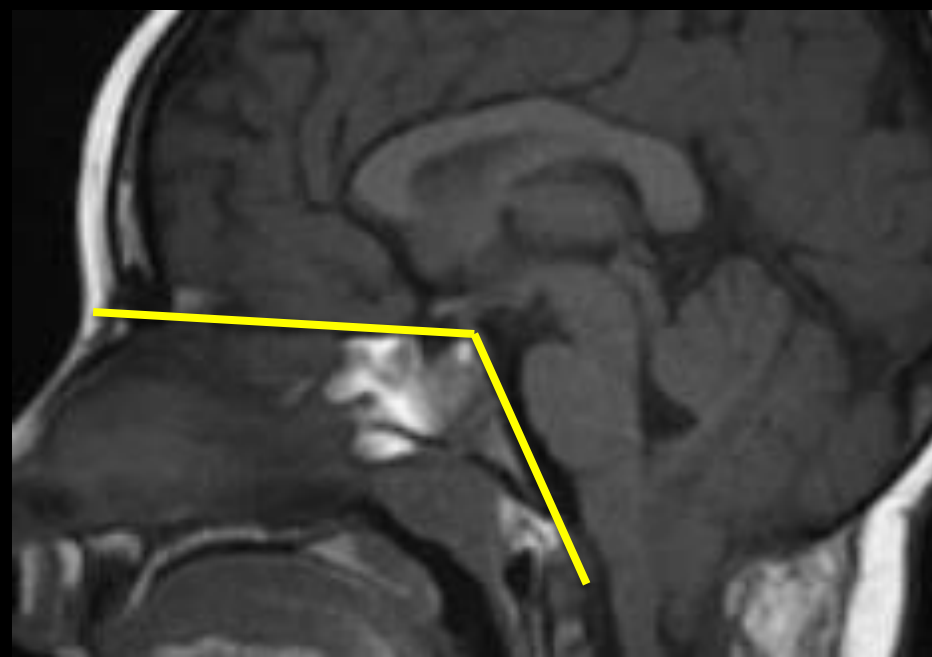
Ipoplasi del basioccipite

Invaginazione basilare

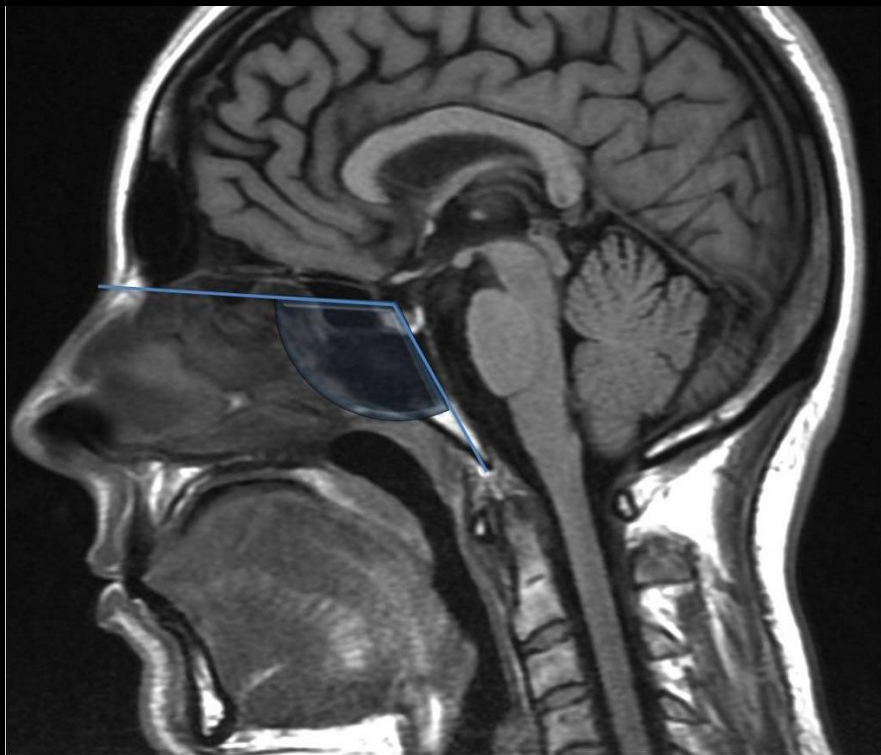


Risalita del dente di C2 > 5 mm al di sopra della linea di Chamberlain

PLATIBASIA



aumento dell'angolo basale



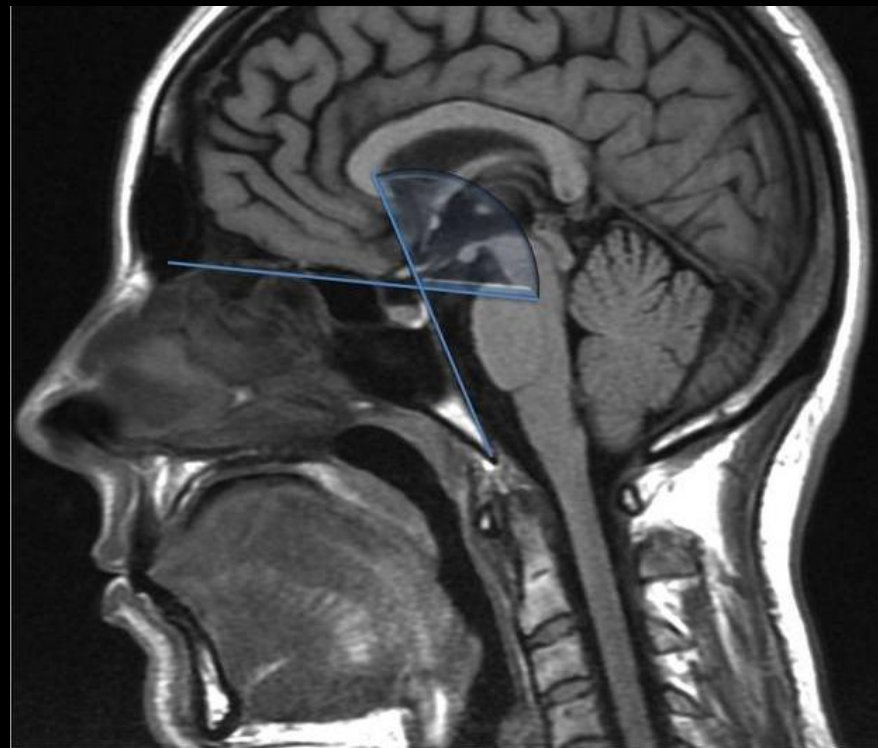
Metodo Standard

Linea nasion-centro della sella turcica
Linea lungo il margine posteriore del
clivus

Normale 125° – 143°

Platibasia $>143^{\circ}$

Cifosi $<125^{\circ}$



Metodo Modificato

Linea nasion-dorso della sella turcica
Linea lungo il margine posteriore del
clivus

Normale:

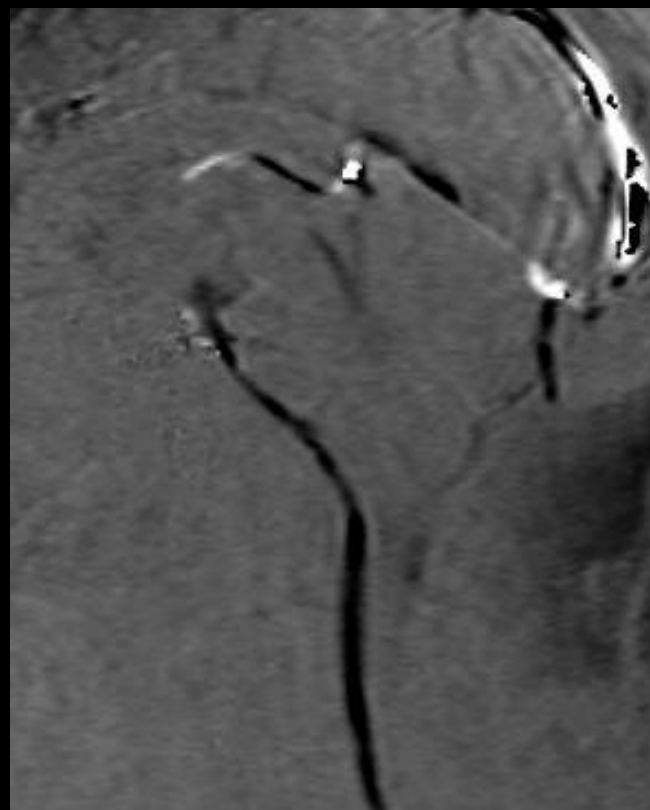
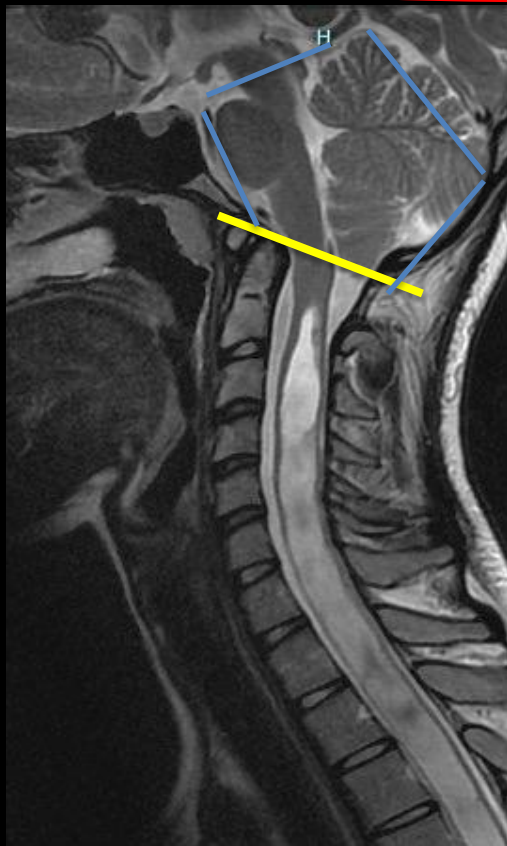
range 116° – 118° per adulti

113° – 115° per bambini

Table 1 Classifications and Definitions

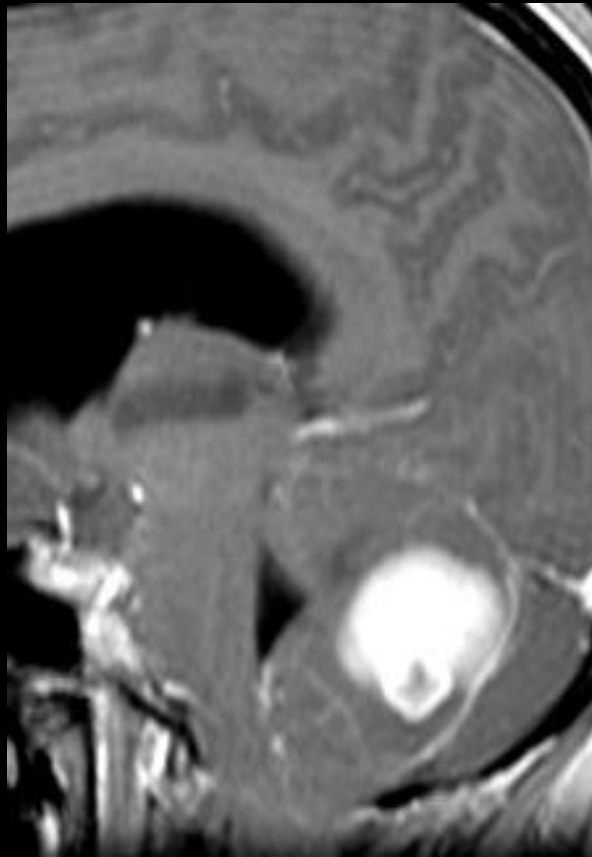
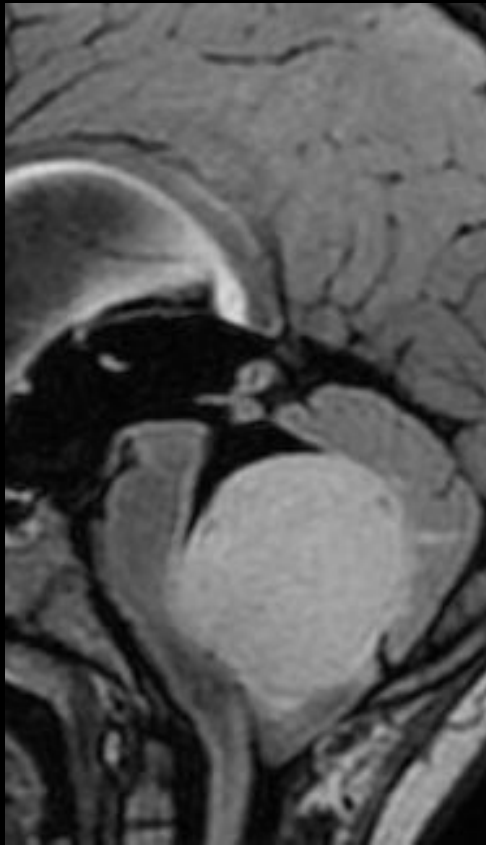
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C2	CM2: due to prenatal CSF hypotension always associated with myelomeningocele and, in some cases, with hydrocephalus and syringomyelia. Other types of intracranial defects (hypoplastic tentorium cerebelli, cranial lacunae, anomalies of the Sylvius aqueduct) may coexist (1).	<i>Agreement:</i> 85.4% Grade D
C3	CM-1.5: cerebellar tonsils and brainstem herniation (kinking) below the McRae line; defined “complex” if also skeletal deformities of craniovertebral junction and cervical spine (Klippel-Feil anomaly, atlanto-occipital fusion, basilar invagination, retroversion of the odontoid process) are present.	<i>Agreement:</i> 81.3% Grade D
C4	CM-0: obliteration of the cisterna magna (due to arachnoid adhesions) and/or volumetrically small posterior fossa, with cerebellar tonsils positioned at the foramen magnum and a syringomyelia in the cervical spinal cord.	<i>Agreement:</i> 81.3% Grade D



Erniazione acquisita

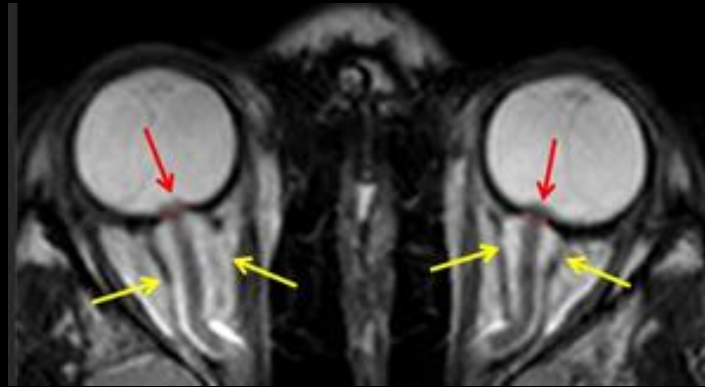
Secondaria ad un tumore della fossa posteriore / cisti aracnoidea

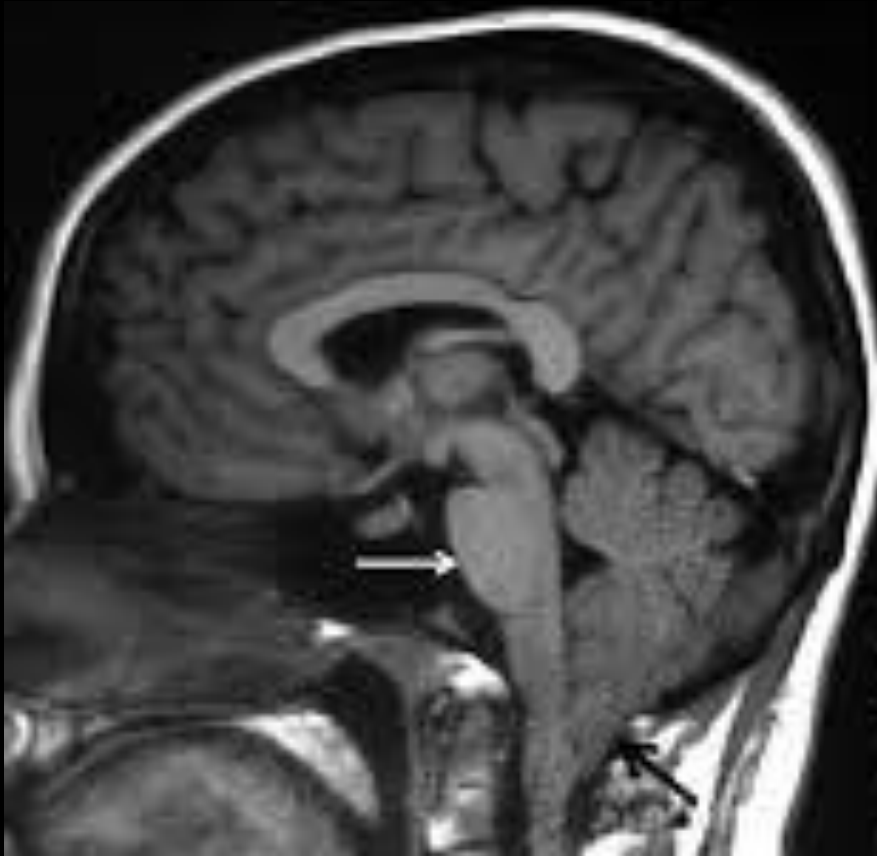


Iperensione Intracranica Idiopatica

vari segni neuroradiologici associati

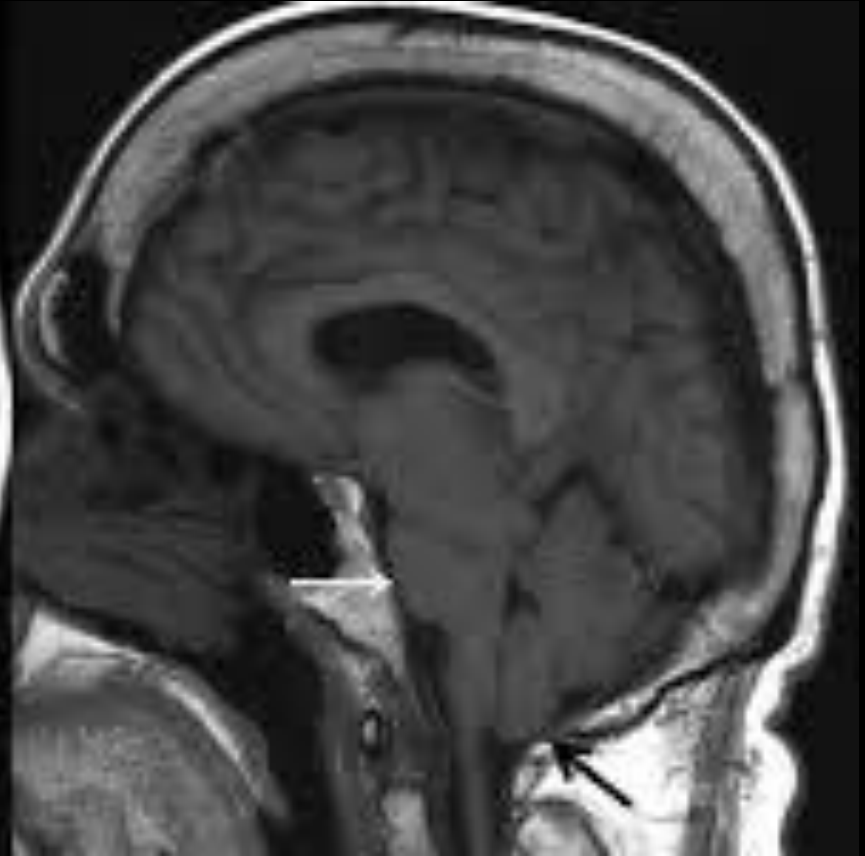
papilledema, distensione delle guaine periottiche, decorso tortuoso dei nervi ottici
stenosi dei seni venosi, sella vuota, riduzione ventricoli





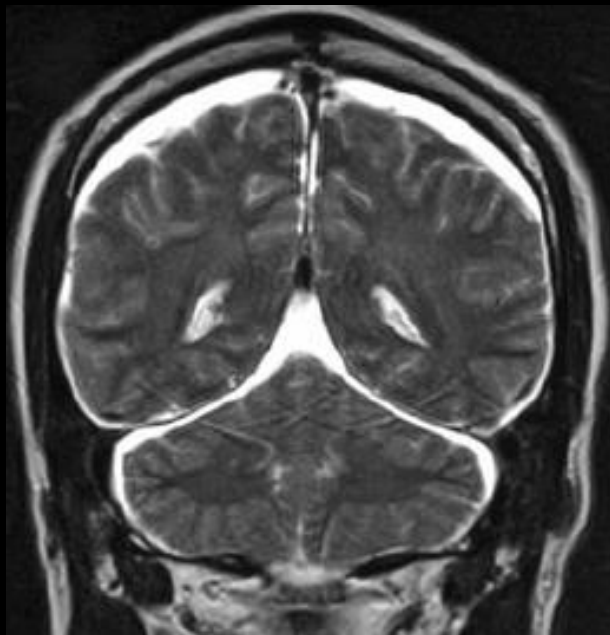
Chiari 1

- *Cisterna prepontina di regolare ampiezza*
- *Protuberanza pontina*
- *Deformazione "peg like"*

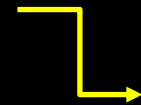


Ipotensione Intracranica

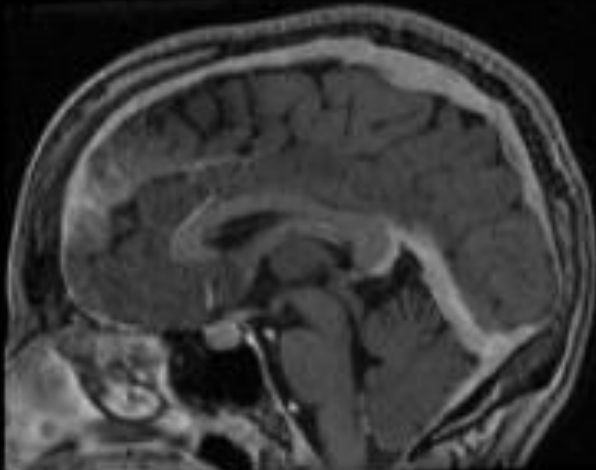
- *Cisterna prepontina oblitterata*
- *Protuberanza pontina appiattita*
- *Riduzione volume del IV ventricolo*
- *Tonsille cerebellari di regolare morfologia*



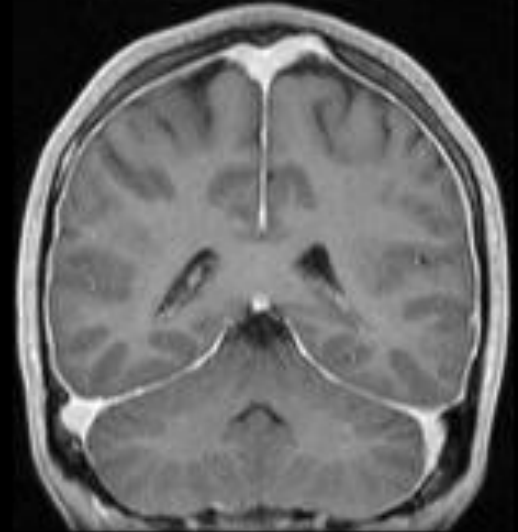
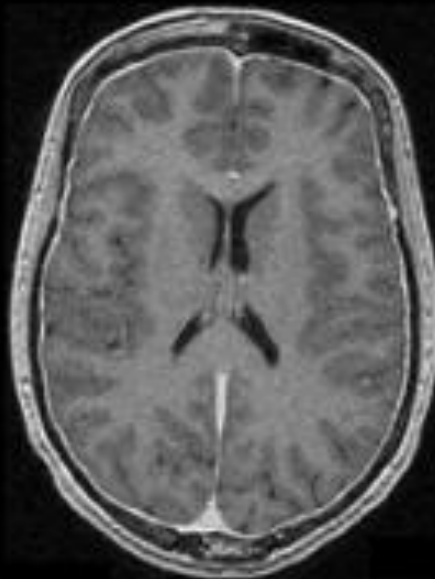
Chiari vs Ipotensione Intracranica



c.e.della dura



T1 C+

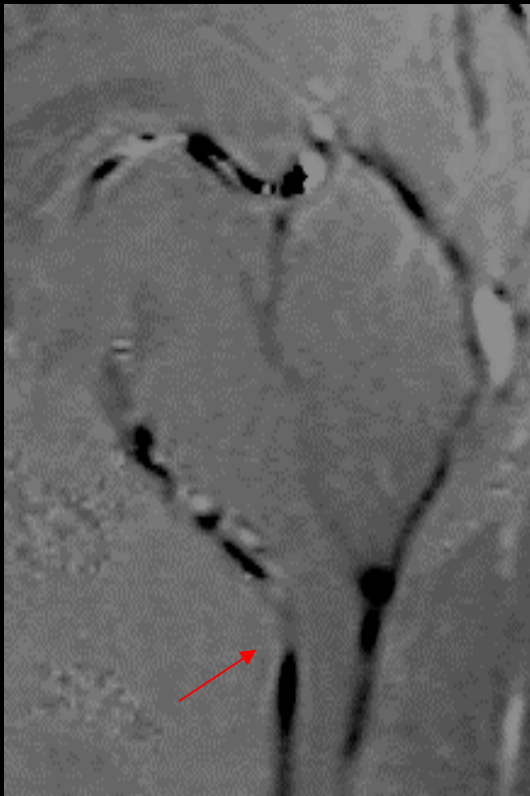


Phase Contrast cine MR

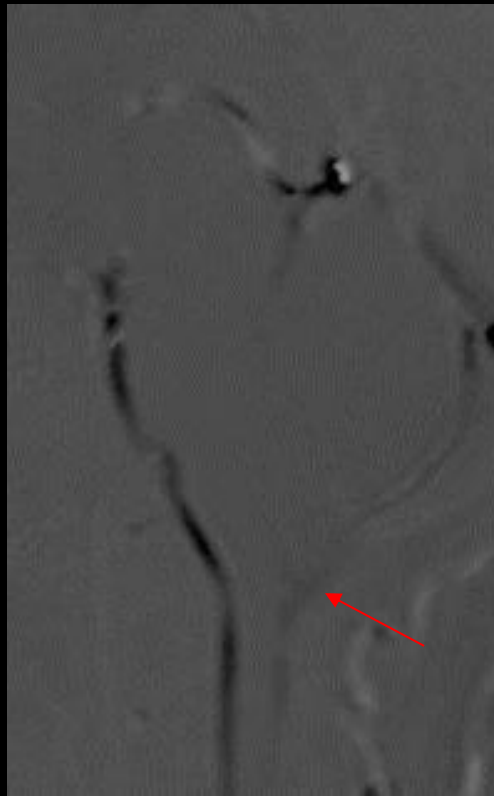
Anomalie del flusso Liquorale

Riduzione

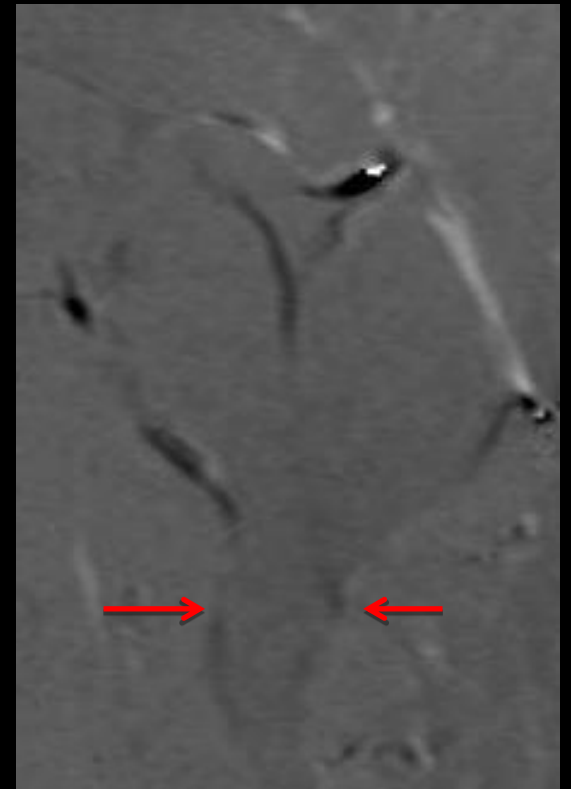
anteriore



posteriore

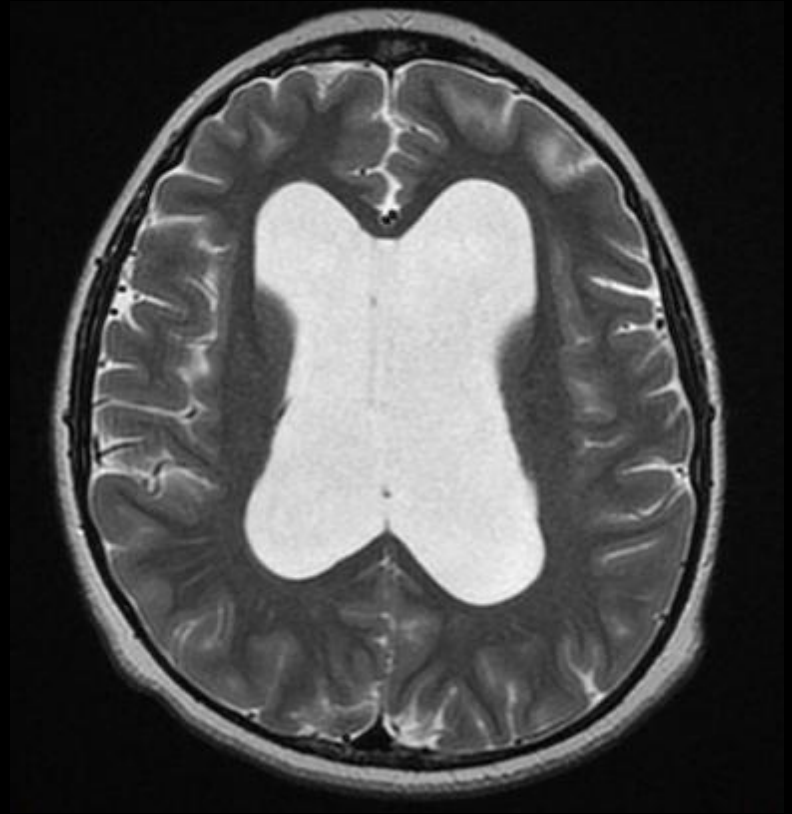


Ostruzione

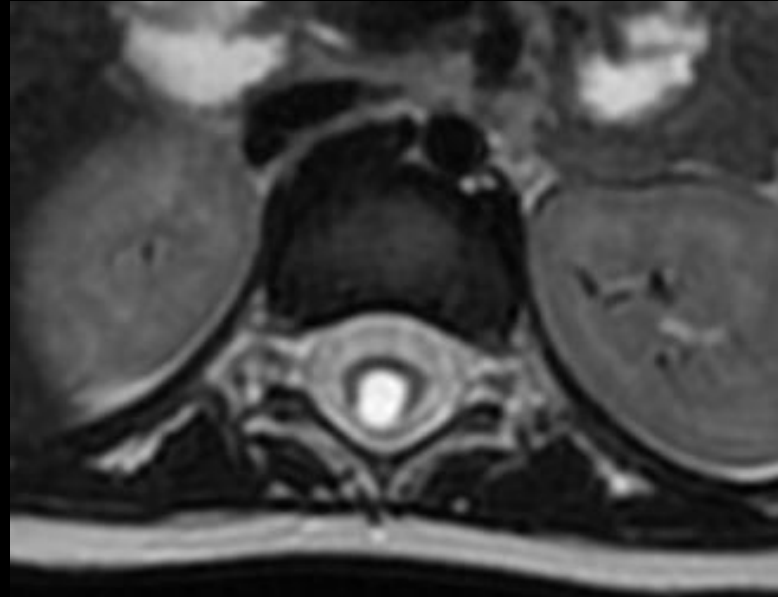


Ricerca di anomalie associate

IDROCEALO?



Tethered cord



Midollo ancorato posteriormente a S1
con un filum terminale ispessito e
lipomatoso

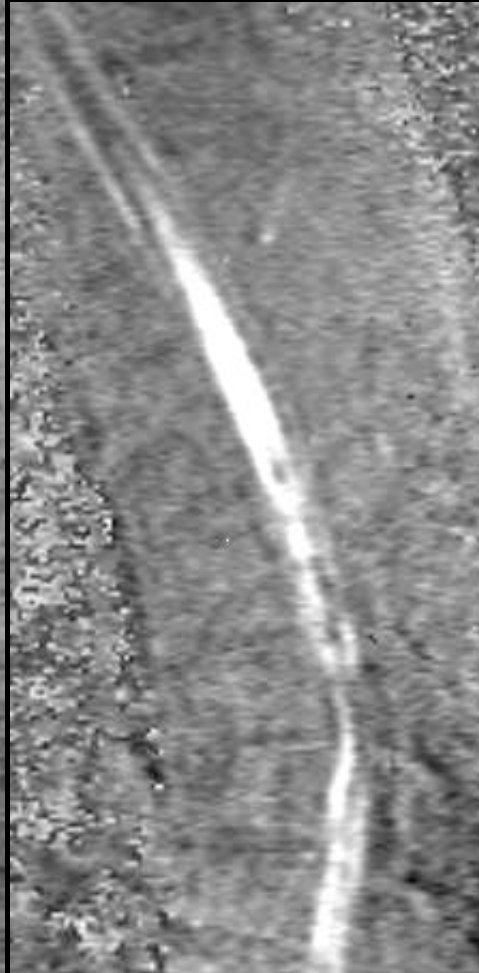
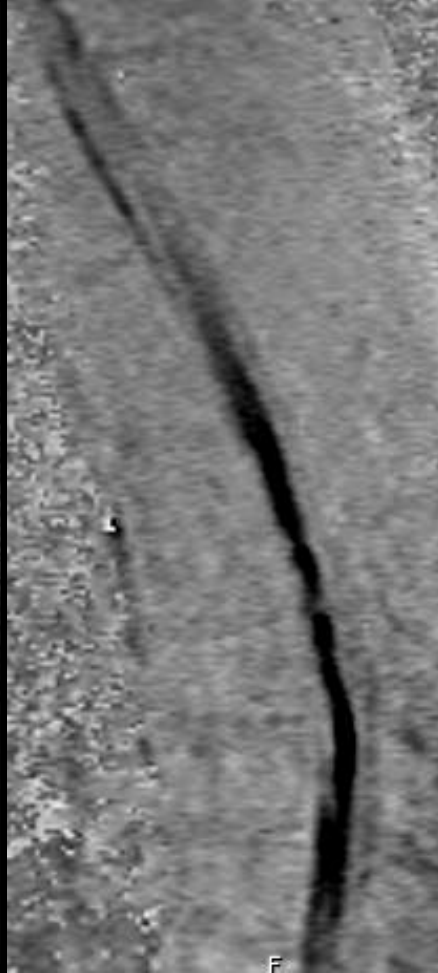
Scoliosi



35% Syr,
28% CM1 con Syr,
25% CM1 isolata

Siringomielia

**Cavità endomidollare a segnale isoliquorale
con flusso allo studio Cine- RM
priva di enhancement dopo mdc**



Siringomielia secondaria (Tipo III)





Diagnosis and treatment of Chiari malformation and syringomyelia in adults: international consensus document

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Siringomielia-Classificazione

Type I: syringomyelia with obstruction of the foramen magnum and dilation of the central spinal canal:

a) Associated with CM1;

b) Associated with other obstructive lesions of the foramen magnum;

Type II: syringomyelia without obstruction of the foramen magnum, or **idiopathic**;

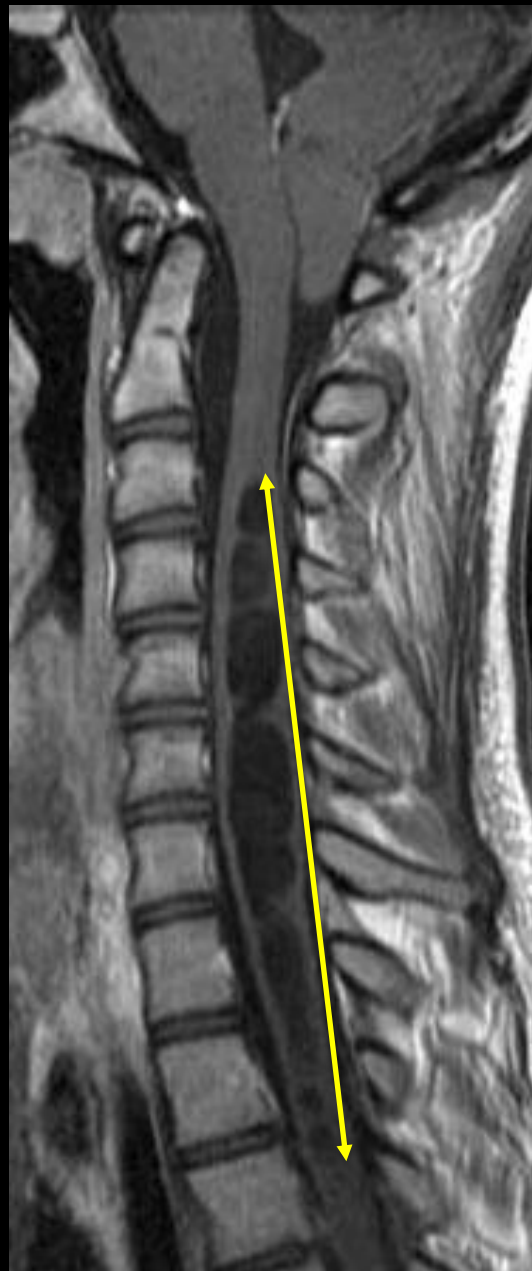
Type III: syringomyelia with other diseases of the spinal cord (**secondary**):

a) Spinal cord tumours (usually intraspinal);

b) Traumatic myelopathy;

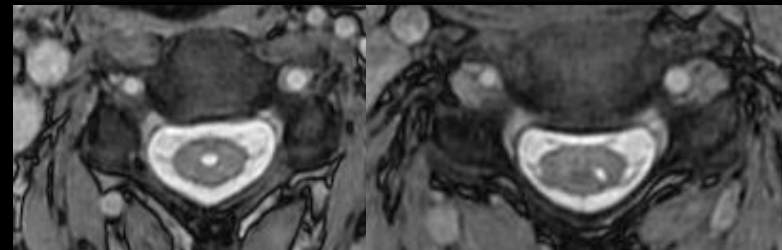
c) Spinal arachnoiditis and pachymeningitis;

d) Myelomalacia due to compression of the spinal cord (tumor, spondylosis);



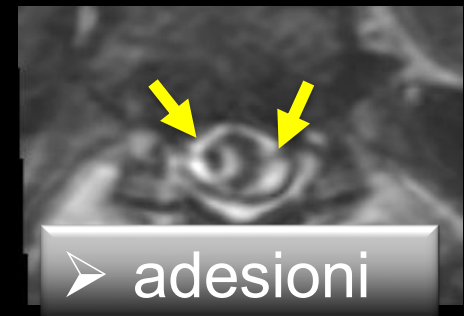
- Estensione
- Diametro AP
- Setti

- Sede: posizione nelle immagini ax



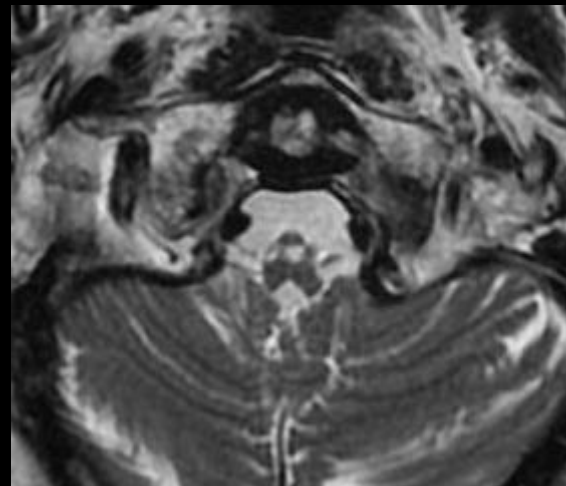
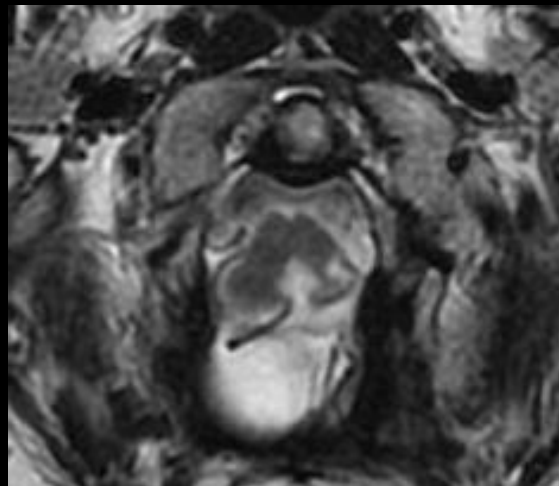
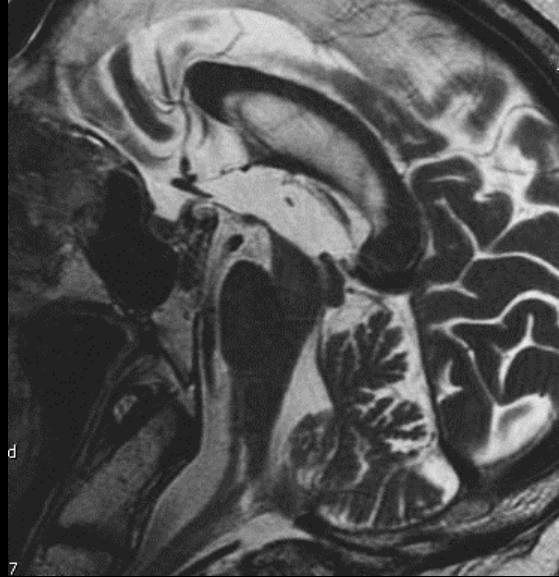
centrale

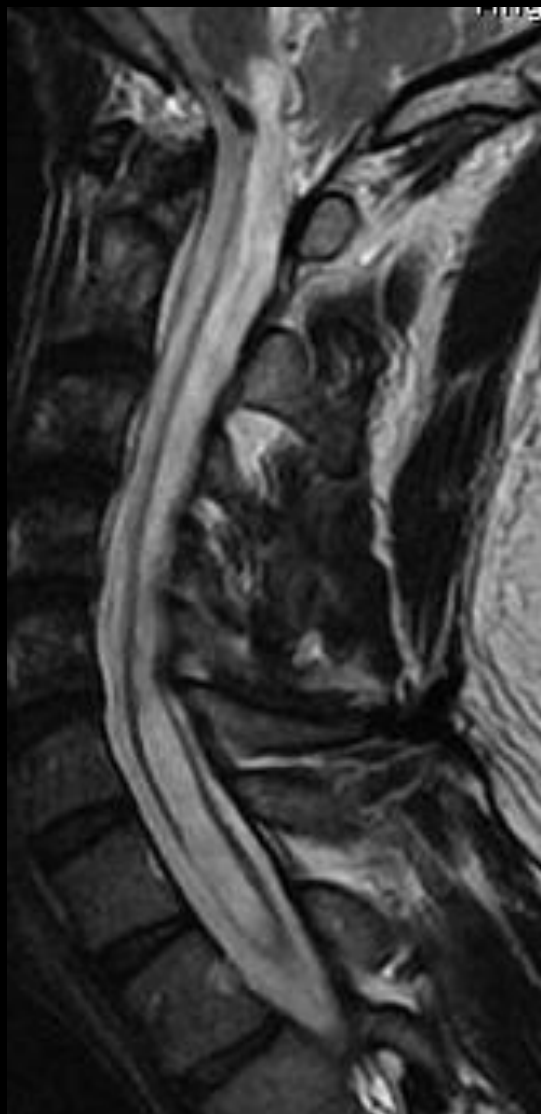
eccentrica



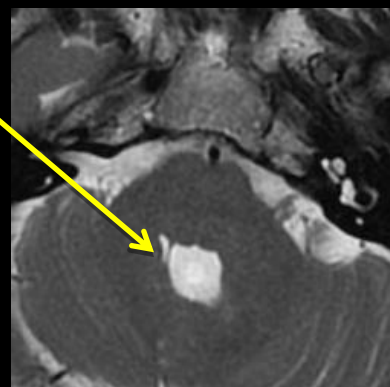
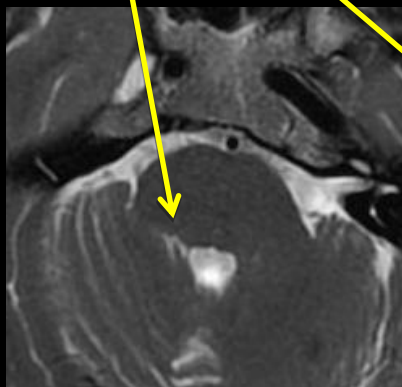
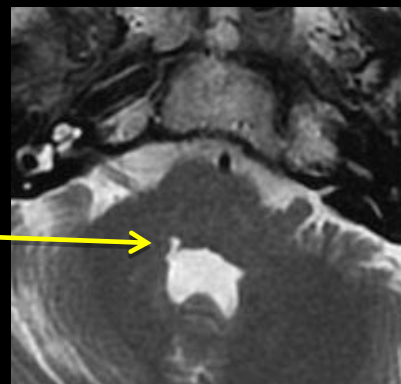
- adesioni

estensione al bulbo → Siringobulbia





**Estensione al ponte
(rara)**



Dilatazione del canale ependimale centrale

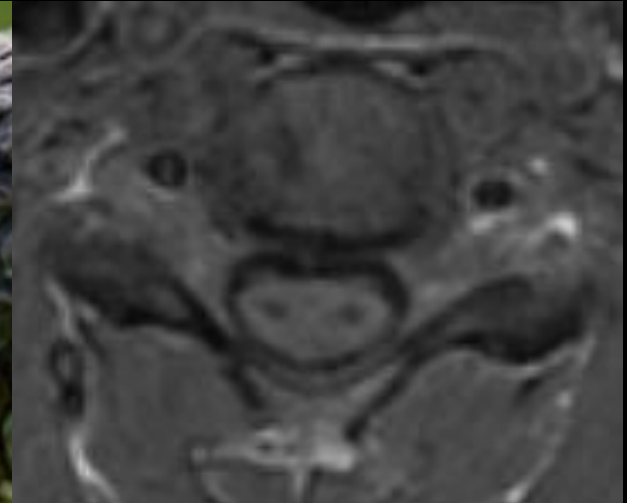
- Presente alla nascita, progressiva chiusura progressiva ma può persistere
- Non associata a sintomi clinici
- Posizione leggermente anteriore nel midollo cervicale, centrale nel midollo dorsale
- Fusiforme sul piano sagittale, rotonda sul piano assiale
- Di pochi millimetri di diametro (1-2 mm)
- Spesso localizzata in un breve segmento del midollo spinale
- Non si associa rigonfiamento del midollo spinale



Siringomielia ?

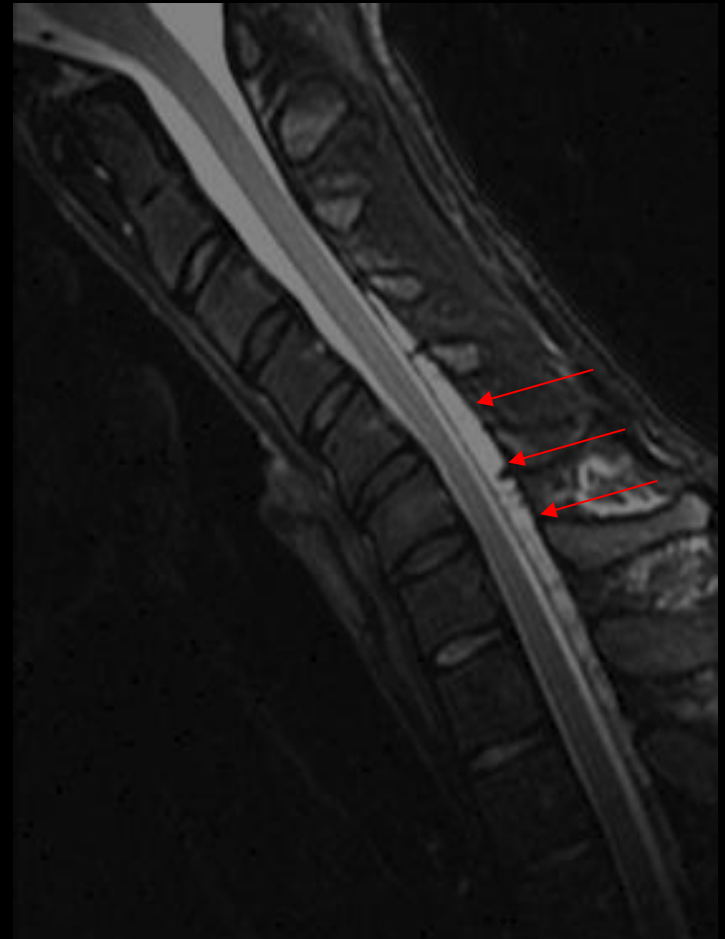
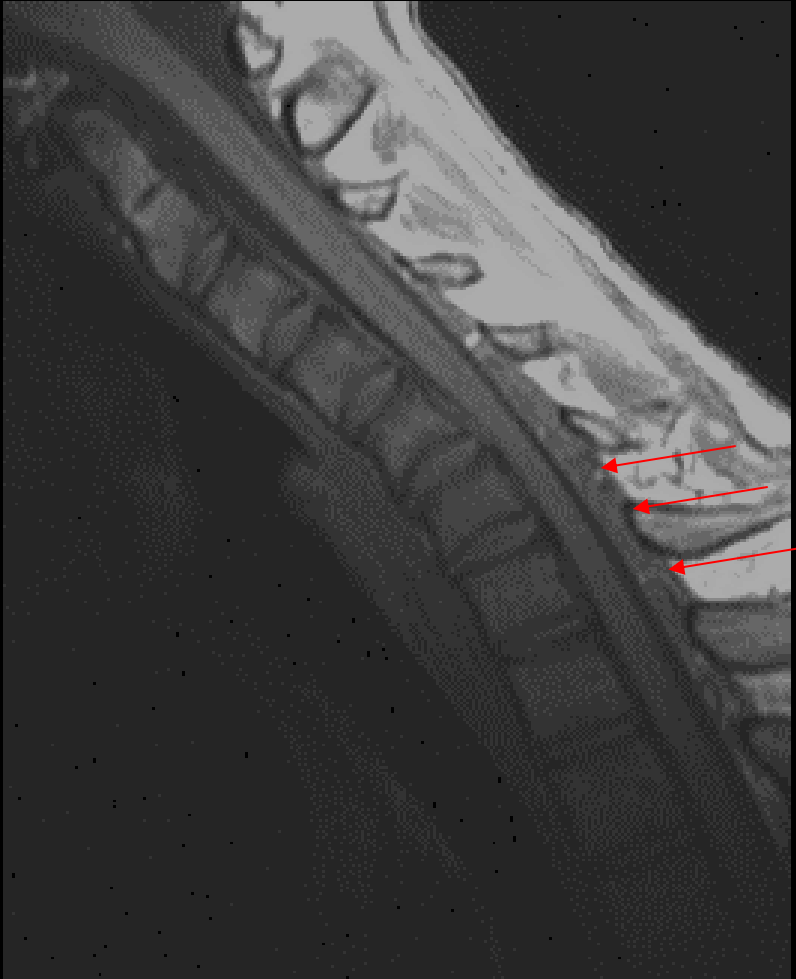


Snake eyes sign



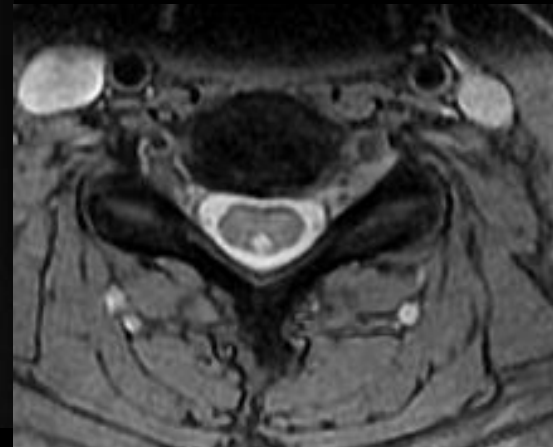
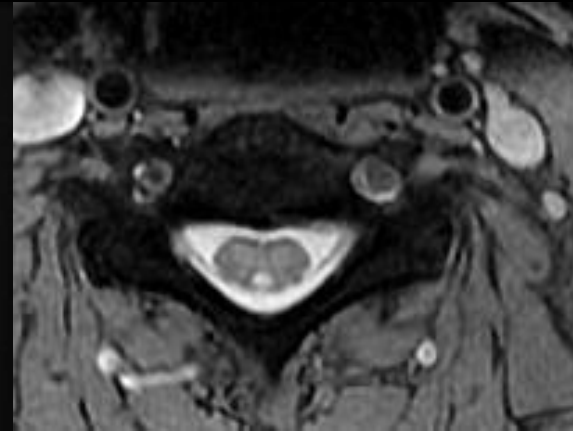
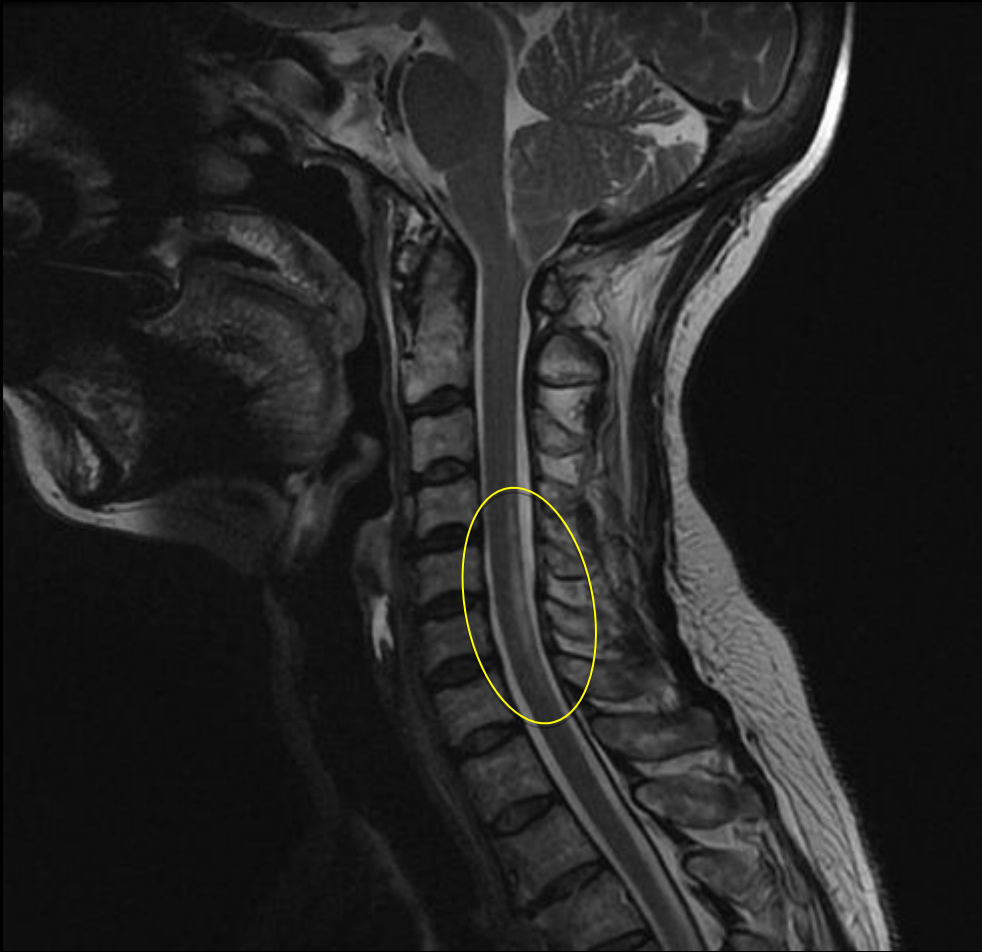
Malattia di Hirayama (Amiotrofia monomelica)

Malattia di Hirayama



Nella posizione in iperflessione si ha un incremento delle dimensioni del plesso venoso nello spazio epidurale posteriore con dislocazione anteriore, compressione del midollo spinale, ischemica cronica con sofferenza delle corna anteriori e necrosi

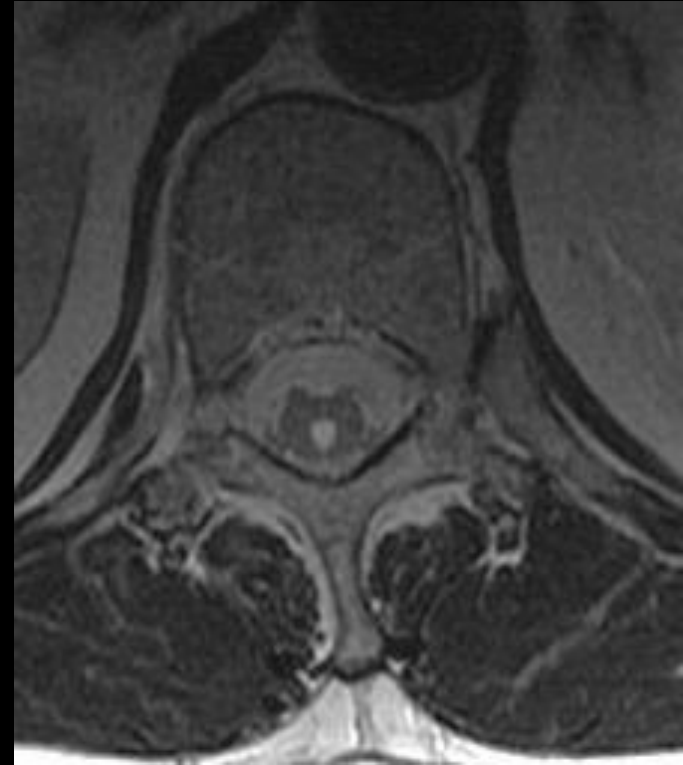
Siringomielia ?



Mielopatia da deficit di vitamin B12

Degenerazione dei cordoni laterali e posteriori dovuta a demielinizzazione

Siringomielia terminale vs Ventricolo Terminale Persistente



Dilatazione del canale centrale ependimale a livello del cono
In genere regredisce durante la prima settimana di vita

Monitoraggio nel tempo



Edema presiringomielico



Comparsa cavità siringomielica



MR Giugno 2020



MR Febbraio 2021



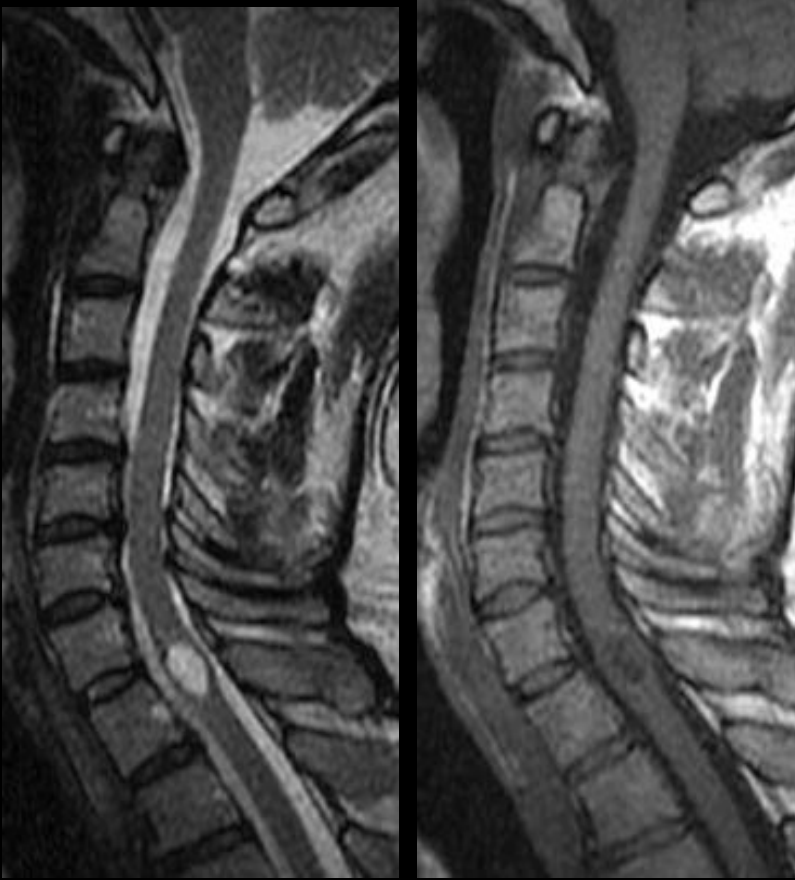
Peggioramento della Siringomielia



L'estensione cranio- caudale di una syr nota è preceduta dall'edema presyr



Siringomielia idiopatica?



1 anno dopo....



3 anni dopo....

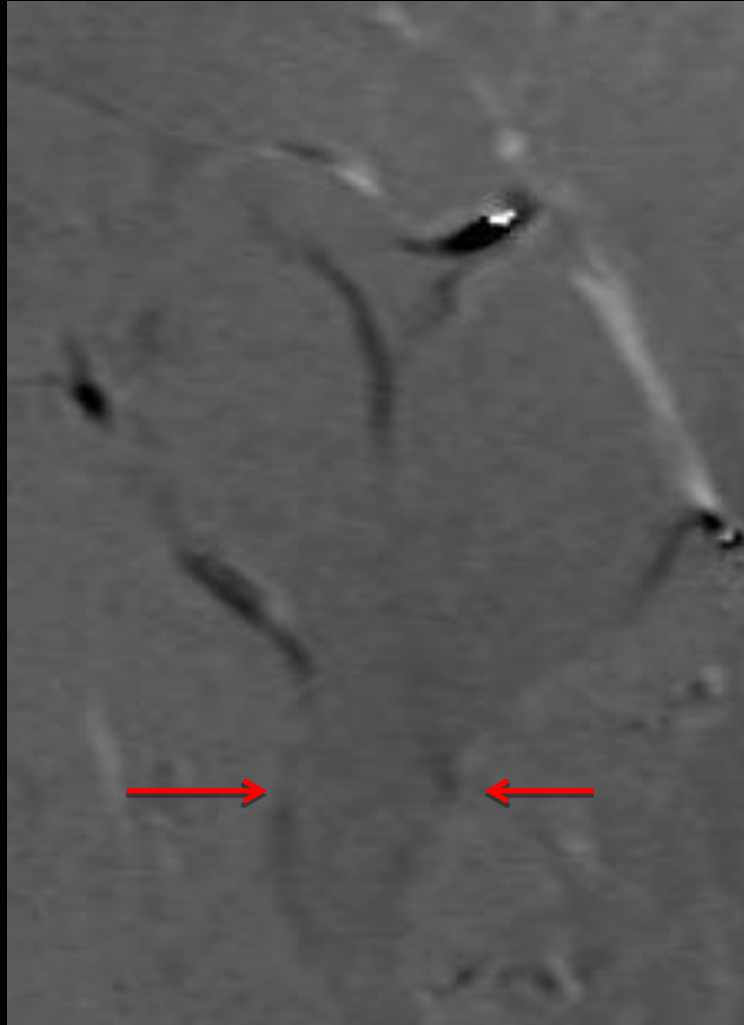
Tumore



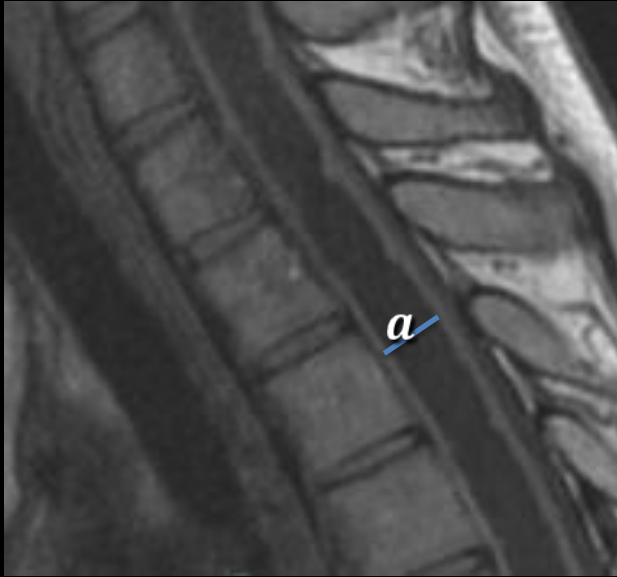
Supporto alla Decisione Chirurgica



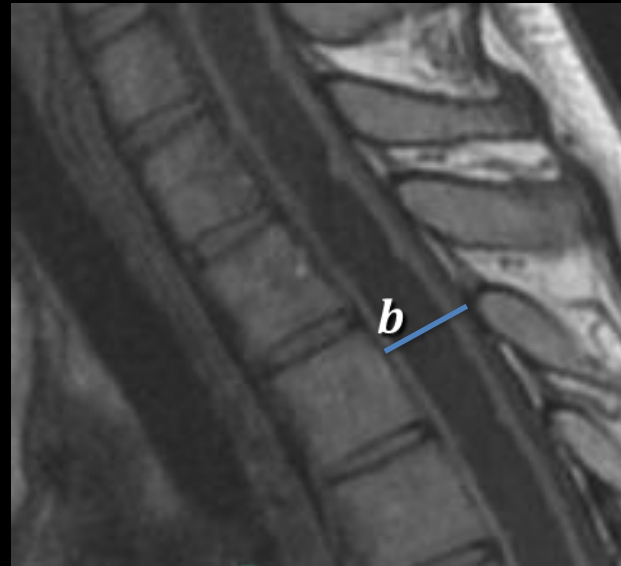
Ostruzione del flusso liquorale



VAQUERO INDEX



Rapporto tra il diametro ap della cavita' syr nel suo punto di maggior ampiezza e il diametro del canale vertebrale



Sag T1
linea mediana

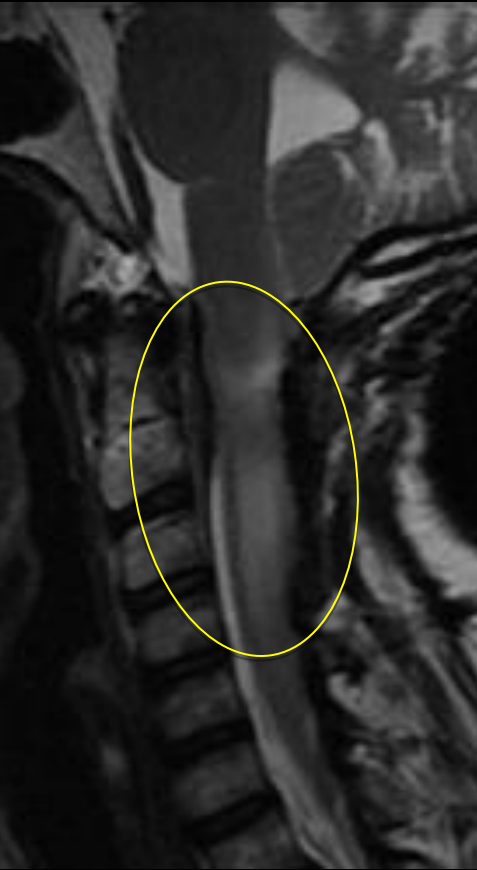
$$\frac{a}{b} \geq 0,5$$



**INTERVENTO
CHIRURGICO**

Progressione radiologica

- Comparsa di syr in paziente con CM inizialmente isolata
- Espansione della siringomielia nel follow-up



Gennaio '22

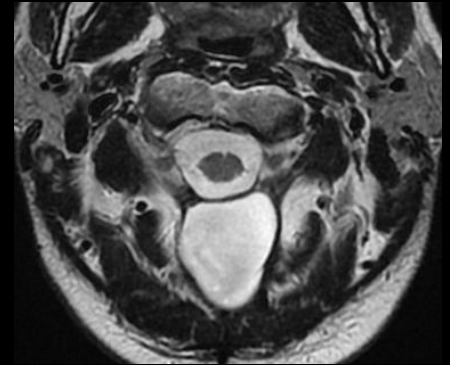
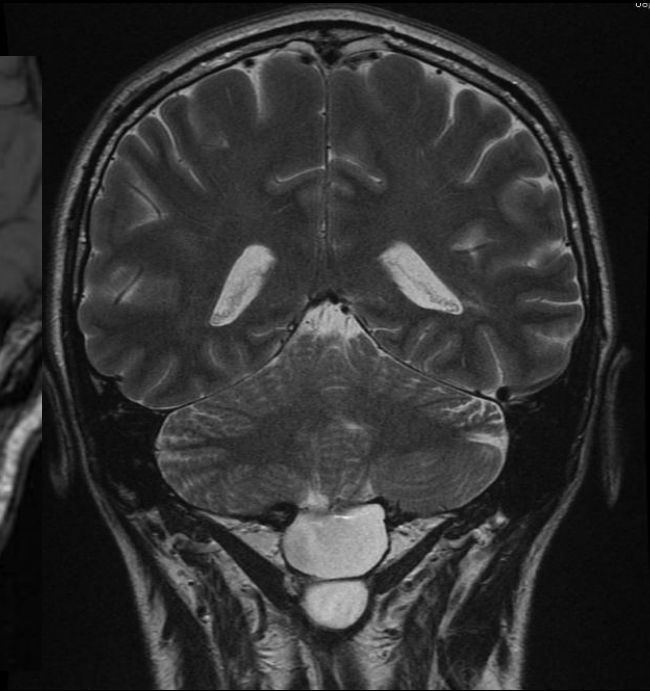
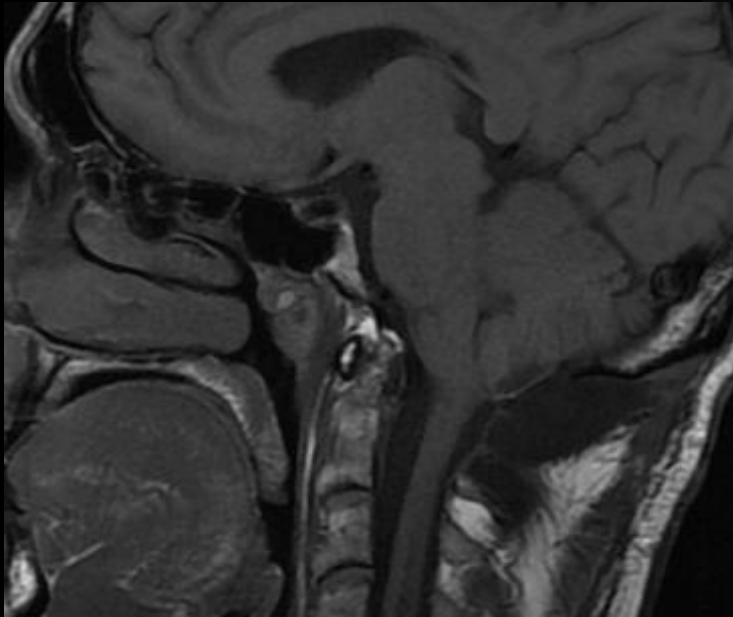


Febbraio '22



Follow-up post-chirurgico

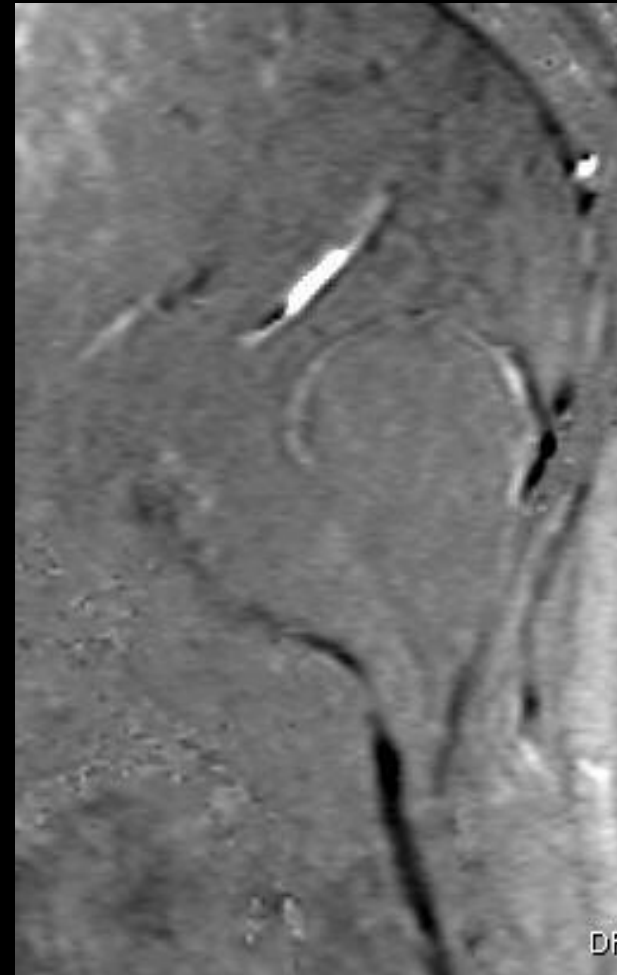
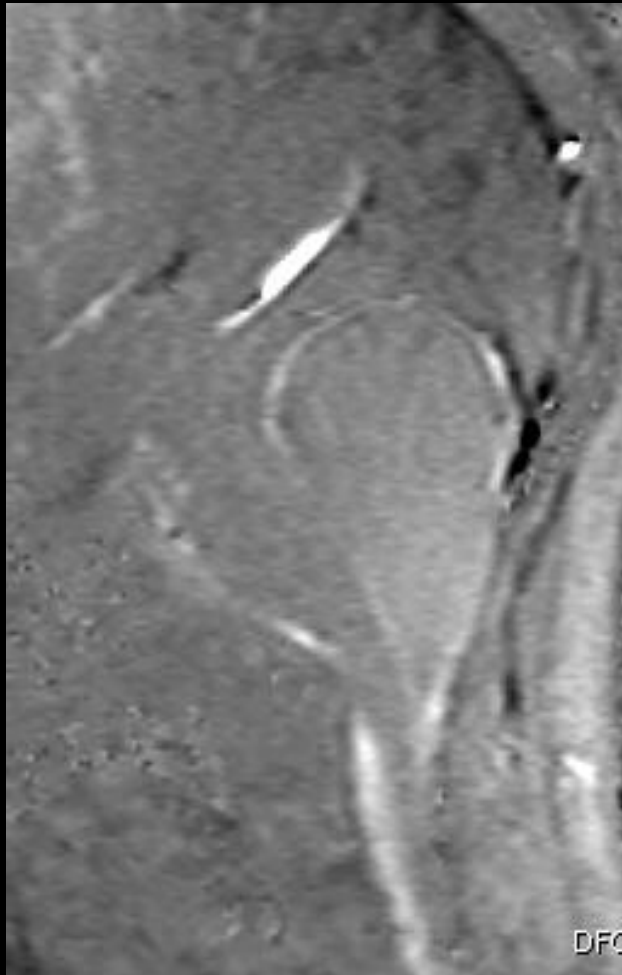
Complicazioni post-chirurgiche



Efficacia della decompressione della fossa cranica posteriore

Phase Contrast Cine MR

Ripristino flusso liquorale



**Riduzione del diametro e
dell'estensione della siringomielia**



Riduzione dell'Indice di Vaquero

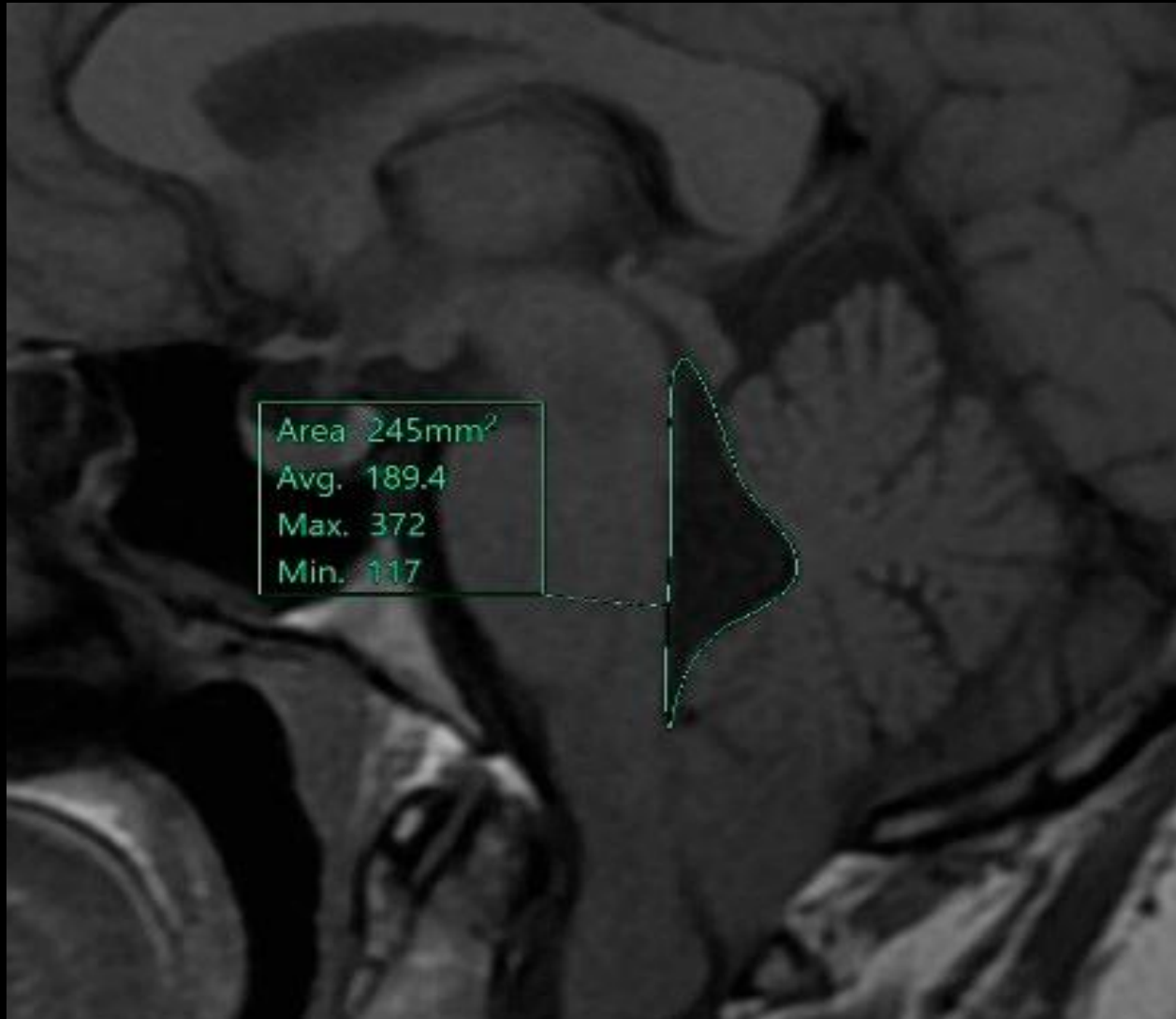
Prima int.ch.(VI = 0.75)



Dopo int.ch.(VI = 0.1)



Fattori prognostici



Valutazione sulla Sag T1 pesata

Syringomyelia Associated with Chiari 1 Malformation in Adults: Positive Outcome Predictors after Posterior Fossa Decompression with Duraplasty

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Malformation in Adults: Positive Outcome Predictors after Posterior Fossa Decompression with Duraplasty. *J. Clin. Med.* **2023**, *12*, 3019. <https://doi.org/10.3390/jcm12083019>

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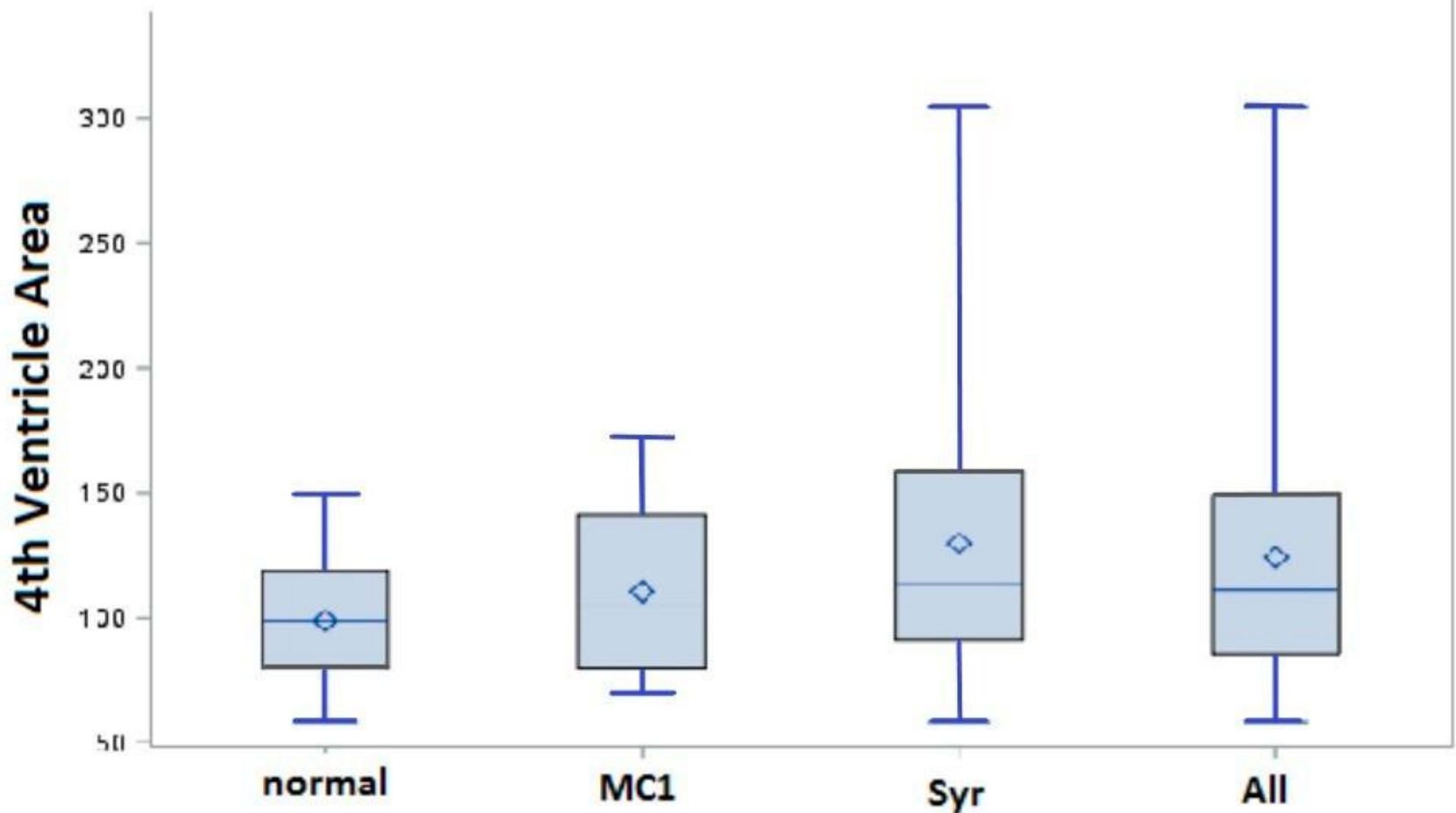
Published: 21 April 2023



Abstract: Background: Syringomyelia (Syr) in patients with Chiari 1 malformation (CM1) may be attributable to abnormal dynamics of cerebrospinal fluid (CSF) in the upper cervical segment; fourth ventricle enlargement has been reported in association with a worse clinical and radiological presentation, independently of the posterior fossa volume. In this study, we analyzed presurgery hydrodynamic markers to evaluate if their changes could be associated with clinical and radiological improvement after posterior fossa decompression and duraplasty (PFDD). As a primary endpoint, we aimed to correlate improvement in the fourth ventricle area with positive clinical outcomes. **Methods:** In total, in this study, we enrolled 36 consecutive adults with Syr and CM1 who were followed by a multidisciplinary team. All the patients were prospectively evaluated with clinical scales and neuroimaging, including CSF flow, the fourth ventricle area, and the Vaquero Index by using a phase-contrast MRI before (T0) and after surgical treatment (T1-Tlast, with a range of 12–108 months). The CSF flow at the craniocervical junction (CCJ), the fourth ventricle area, and the Vaquero Index changes were statistically analyzed and compared to the clinical and quality of life improvement after surgery. The good outcome prediction ability of presurgical radiological variables was tested. **Results:** Surgery was associated with positive clinical and radiological outcomes in more than 90% of cases. The fourth ventricle area significantly reduced after surgery (T0-Tlast, $p = 0.0093$), but no significant associations with clinical improvement were found. The presurgical presence of CSF flow at the CCJ was able to predict a good outcome (AUC = 0.68, 95% CI 0.50–0.87 and LH+ = 2.1, IC 95% 1.16–3.07) and was also significantly associated with post-surgical pain relief ($\rho = 0.61$ and $p = 0.0144$). **Conclusions:** Presurgery CSF flow at the CCJ is proposed as a radiological marker with the ability to predict a positive outcome after PFDD in adults with syringomyelia and CM1. Measurements of the fourth ventricle area could be useful additional information for evaluating surgical long-term follow-up; further experience on larger cohorts is required to better define the prognostic yield of this radiological parameter.

Keywords: syringomyelia; Chiari 1 malformation; PFDD; surgery; PC-MRI; outcome; adults

Area del IV ventricolo è maggiore nei pazienti con MC1 e soprattutto nei pazienti con Syr rispetto ai controlli normali



3.2. Follow-Up: Clinical and Radiological Outcomes

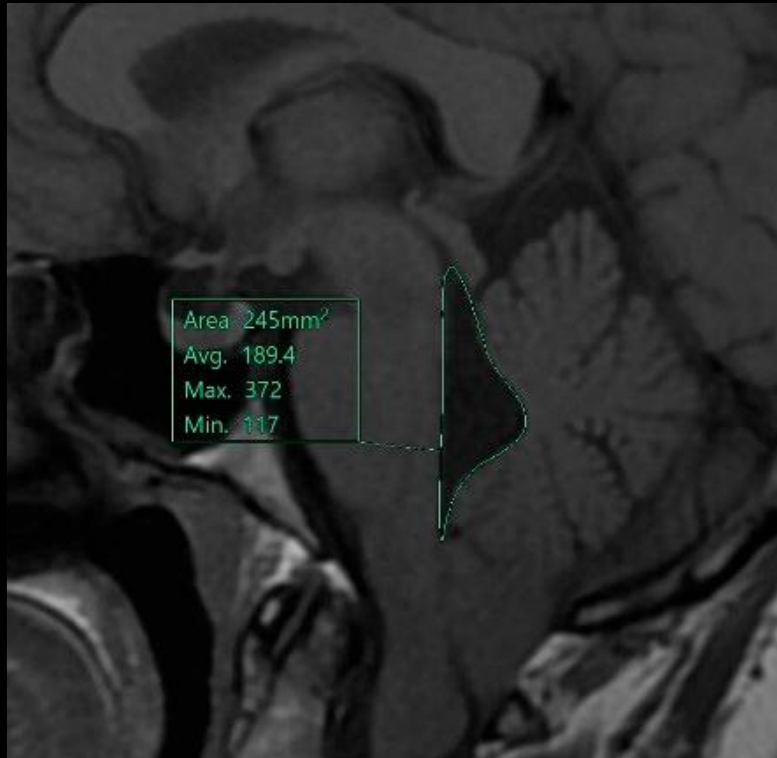
The statistical pre/post-surgery comparisons among clinical and radiological variables are reported in Table 5. The VAS and CSS scores were significantly reduced, both at short-term and long-term follow-up (FU). The fourth ventricle area (Figure 2(1)) presented a significant reduction at short-term FU. The Vaquero Index (Figure 2(2)) was significantly reduced at short- and long-term FU; CSF flow at the CCJ was significantly improved at short- and long-term FU.

Table 5. Radiological and clinical changes at T0-T1, T0-Tlast, T1-Tlast (statistical comparison).

Parameters	T0	T1	Tlast	<i>p</i> -Value		
	Mean (SD)	Mean (SD)	Mean (SD)	T0-T1	T1-Tlast	T0-Tlast
Fourth Ventricle Area (mm ²)	124.23 (52.63)	116.64 (50.57)	110.63 (40.27)	0.0571	0.0373	0.0093
CSF flow score	2.3 (0.59)	1.92 (0.65)	1.69 (0.74)	0.0009	0.0325	<0.0001
Vaquero Index	0.49 (0.17)	0.30 (0.18)	0.28 (0.21)	<0.0001	0.4196	0.0002
CSS	1.19 (0.92)	0.94(0.83)	0.94 (0.88)	0.0016	0.572	0.0091
VAS	3.29 (2.87)	2.31 (2.58)	1.97 (2.92)	0.0188	0.449	0.0266
mRS	1.33 (0.53)	1.31 (0.75)	1.28 (0.92)	0.7677	0.4882	0.5

Radiological and clinical variations at pre-surgery and post-surgery follow-up. Mean and SD of all the parameters are reported. All the radiological parameters significantly improved after surgery as well as clinical and pain scores. Bold face characters indicate statistically significant differences. CSF, cerebro-spinal fluid; CSS, clinical sign score; VAS, visual analog scale; mRS: modified Rankin scale.

Prima int.ch



Area del IV ventricolo 245 mm²

Dopo int.ch



Area del IV ventricolo 139 mm²

with a marked improvement in headache after decompressive surgery.

However, due to the pilot nature of this study, the obtained results should be interpreted with the awareness of some limitations. The first limitation is the retrospective measurement of the fourth ventricle area. To overcome this limitation, as mentioned in the text, all the measurement were performed by a single radiologist; moreover, because this is the first study specifically aimed to describe this new technique, a normal control group was identified and matched to patients by size, sex, and age, as described in details in the Methods section, statistical analysis ("control group selection"). The second limitation of this study is the relatively small sample size of patients. Although our patients were admitted at a single Institution and investigated homogeneously according to a multidisciplinary and standardized protocol, the small cohort could not provide reliable results on the statistical

analysis, in primis, on the prognostic role of the presurgical fourth ventricle area. Finally, fourth ventricle area variability was wider in our patients than we expected based on the literature. We are planning to quantify and compare the fourth ventricle volume with the fourth ventricle area in a larger series of patients in a prospective, collaborative, multicentric study in order to better define the predictive role of theses radiological parameters.

In summary, presurgery CSF flow at the CCJ is proposed as a radiological marker that can predict a positive outcome after PFDD in adults with syringomyelia and CM1 at short- and long-term follow-up. Phase-contrast brain MRI should be proposed in the preoperative diagnostic path. Nevertheless, surgical indication and treatment of syringomyelia with CM1 must include careful preoperative analysis of a single clinical case, involving all the

Acknowledgment



Presidio ospedaliero CTO Torino
(Centro Traumatologico Ortopedico)



**GRAZIE
PER L'ATTENZIONE!**

Corallo cervello
Mar Rosso