Malformazione di Chiari 1:

Indicazioni & Trattamento nell'Adulto

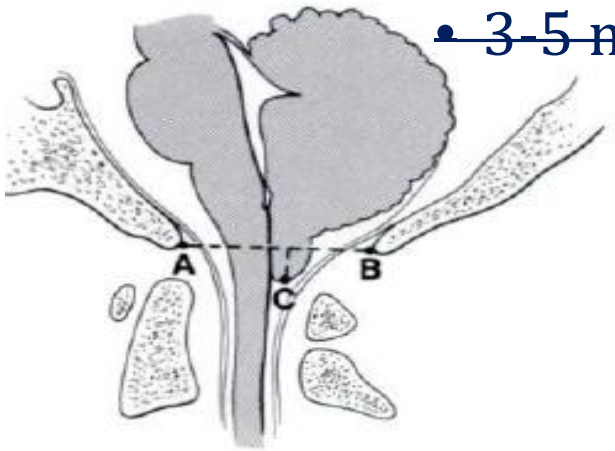
L.G.VALENTINI

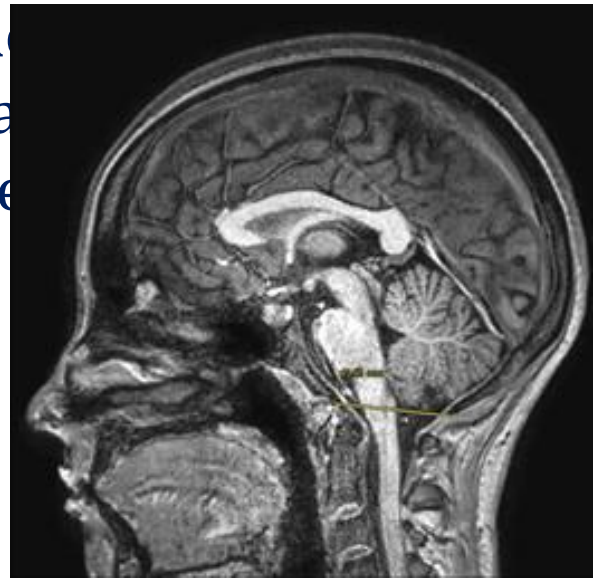
FONDAZIONE I.R.C.C.S. ISTITUTO NEUROLOGICO CARLO BESTA

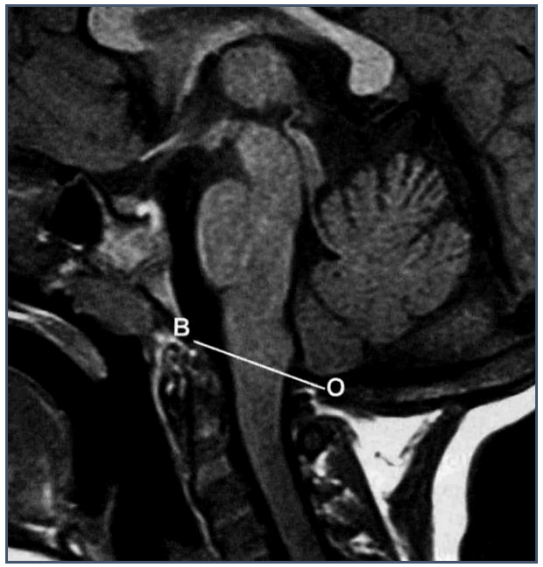
Verso una Diagnosi Funzionale :

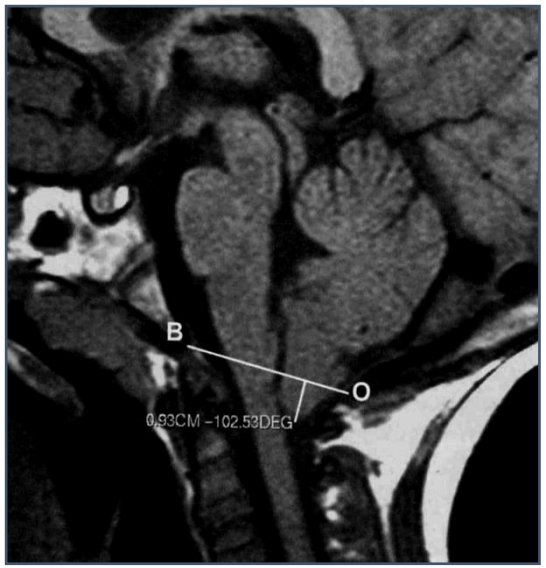
La cosiddetta 'Chiari' (malformazione, anomalia, variante..) è una sindrome compartimentale causata da una «segregazione» del comparto encefalico rispetto a quello midollare per un ostacolo alla circolazione del liquor a livello del Forame Magno, causato da:

- **Affollamento del Foramen Magnum per discesa delle Tonsille CBLari**
- **Entità dell'Erniazione:** quanti mm?
  - **< 5 mm n**
  - > 5 mm pa
  - ~~3-5 mm be~~









Basion-opisthion sulla **Linea Mediana**

In pochi casi non c'è discesa tonsillare, ma il blocco del Forame Magno è causato da

- **Aracnoidite Forame Magno:** Rarissimo, **CM0**

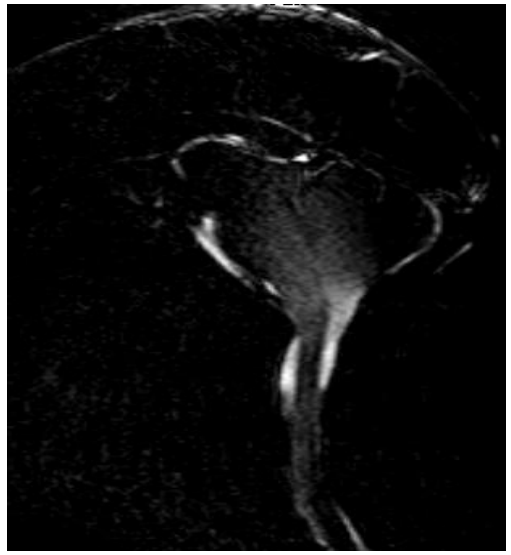
FONDAZIONE I.R.C.C.S. ISTITUTO NEUROLOGICO CARLO BESTA

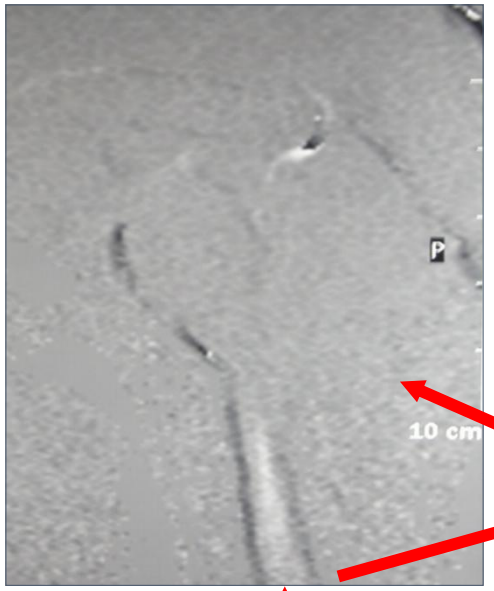
Verso una Diagnosi Funzionale :

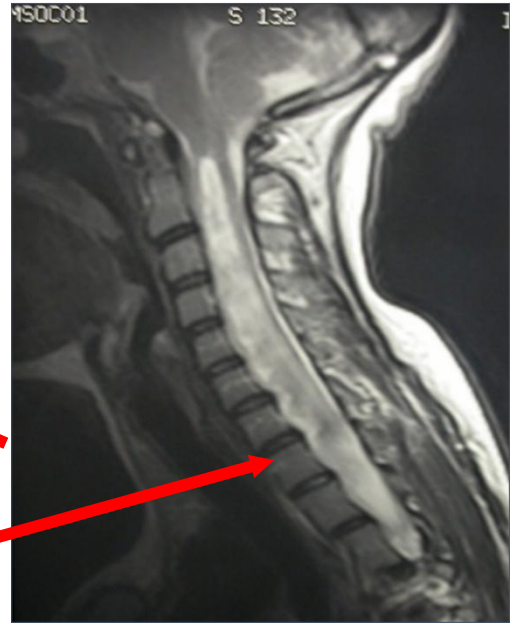
Questo blocco comporta:
*un Delta pressorio fra comparto intracranico ↑ e spinale ↓ con un gradiente a livello Bulbare che causa i **Sintomi** ed impedisce i riarrangiamenti pressori con i cambi posturali,*
*ed una alterazione del flusso liquorale posteriore in salita, con diversione verso il canale centro-midollare, e conseguente genesi della **Siringomielia***

RM con Studio del flusso liquorale

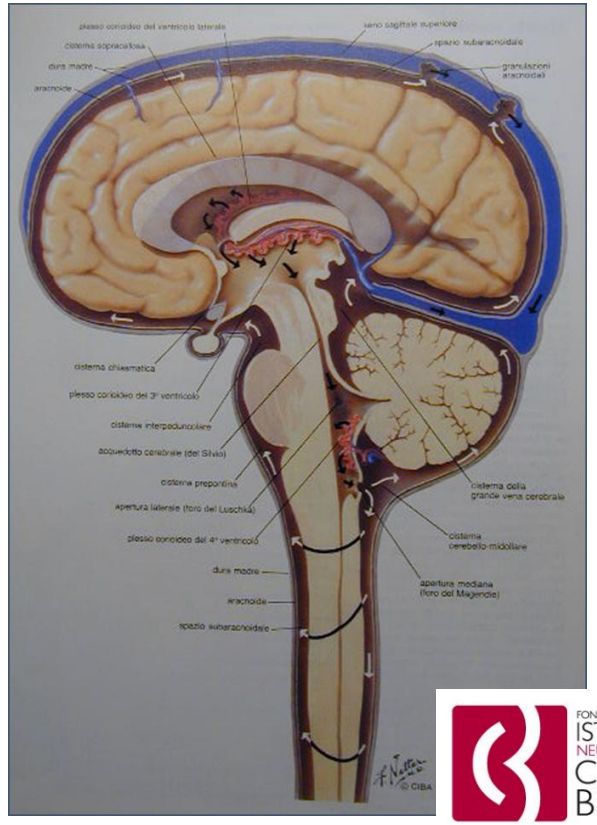
Ostacolo Parziale







Blocco Completo



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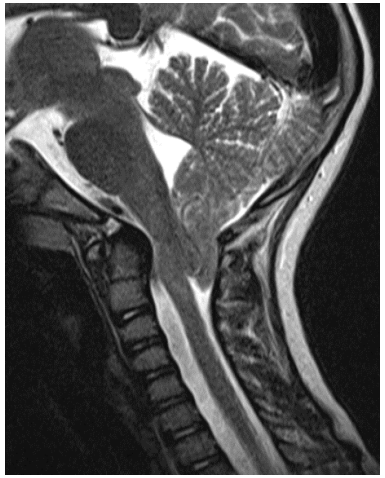
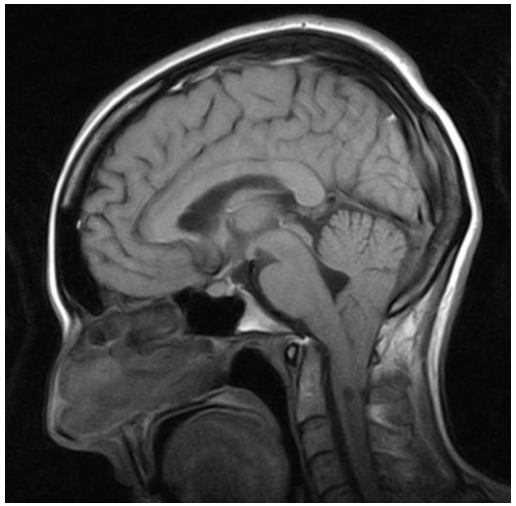
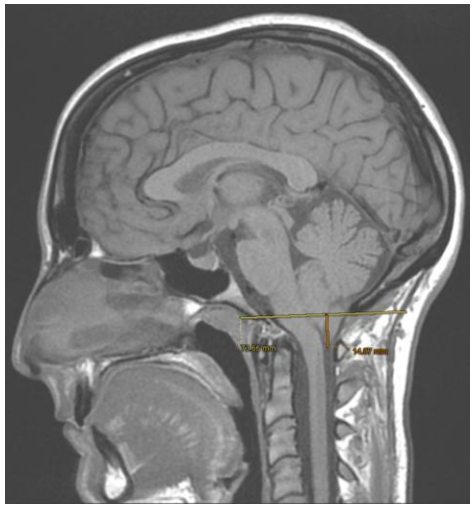
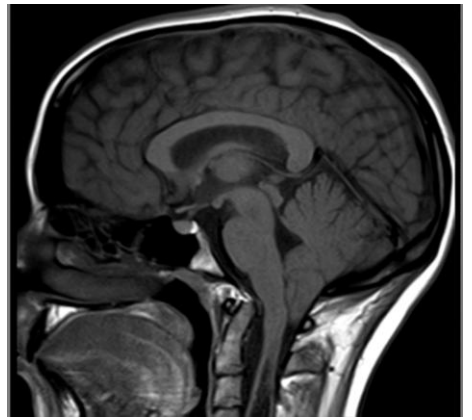
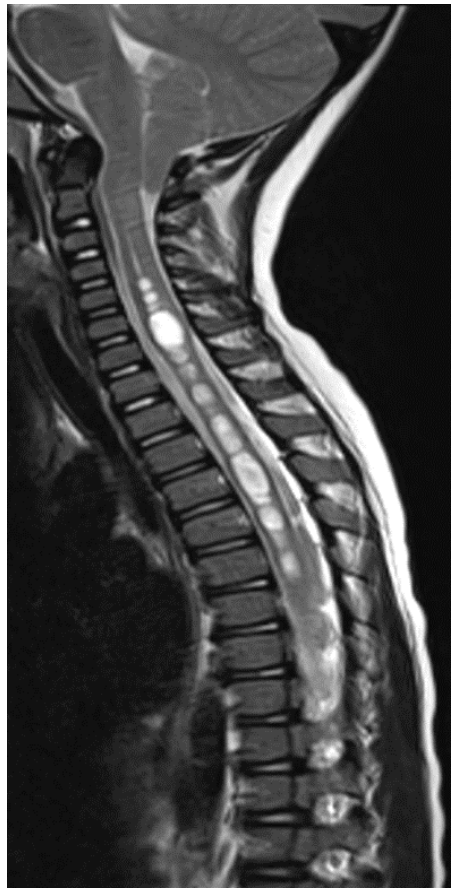
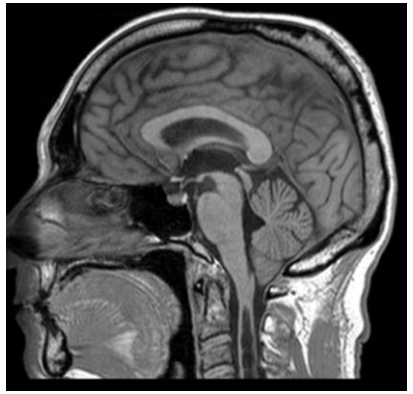
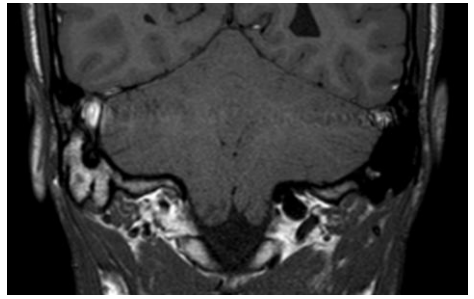
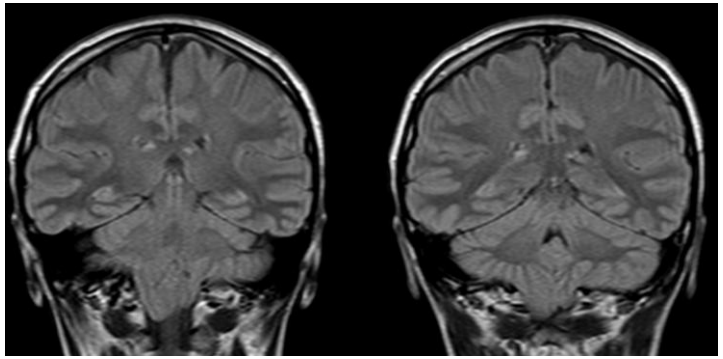
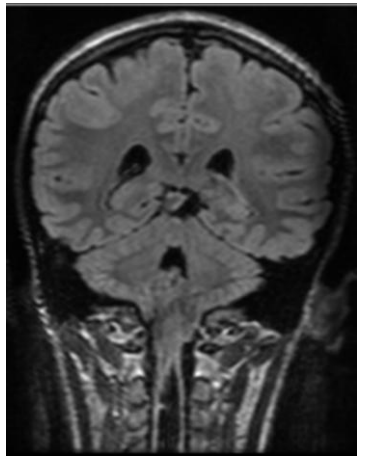
Verso una Diagnosi Funzionale :

*i **Sintomi** sono dovuti all'Ipertensione nel comparto intracranico*
ed al Gradiente Pressorio che si crea nel parenchima al passaggio fra i due comparti,
*la **Siringomielia** all'ostacolo alla risalita del CSF verso il vertice*

Ostacolo Parziale, Blocco Completo

Ma il Blocco DEVE esserci!

MALFORMAZIONE DI CHIARI 1 (CM1): CONTROVERSIE SULL'INTERVENTO

PERCHE'?

- Come evolve la Storia Naturale negli anni?
- Quali Sintomi sono dovuti al CM1?

QUANDO?

- Profilattico/Sintomatici +- Siringomielia
- C'è un Rischio di Deterioramento Acuto?

CHE TIPO DI INTERVENTO?

- DCV/DCV+Duroplastica/DCV+Duroplastica+Resezione/CoagulazioneTonsillare?
- Fissazione CranioVertebrale
- Espansione della Volta Cranica?

QUALI SONO I RISULTATI CHIRURGICI A LUNGO TERMINE?

- Criteri per valutare successo/fallimenti
- Percentuale di Re-Do?
- Quanto deve essere lungo il follow-up? Quando si è **Guariti**?

CONSENSUS CONFERENCE ON

CHIARI MALFORMATION & SYRINGOMYELIA

- the Lesson Learned in Ten Years (2009-2019) -

SCIENTIFIC COMMITTEE

LAURA GRAZIA VALENTINI *MILAN* **LUCA MASSIMI** *ROME*
PALMA CIARAMITARO *TURIN* **PAOLA PERETTA** *TURIN*
LUISA CHIAPPARINI *MILAN* **VERONICA SALETTI** *MILAN*

NOVEMBER 11-12-13 | 2019

Milan, Italy - University of the Studies, Greppi Palace

- Scientific Committee prepared the questionnaires to answer the 7 PICOs from the Association with a Dephi Method
- Pooled **50** multidisciplinary Experts
- in **2** separate groups for Children and Adults
- from **12** different countries
- who are Referral Centers for their national patients' Associations
- Authorship of > **100** papers on CM1 & Syringomyelia
- With cumulative experience of **27.660** followed patients,
- **10.200** of whom operated, **3.500** Children and **6.700** Adults

Symptoms
and/or
Syringomyelia
SURGERY

No Symptoms
No Syringomyelia
Follow Up

Gruppo Multidisciplinare
Malformazione di Chiari & Siringomielia

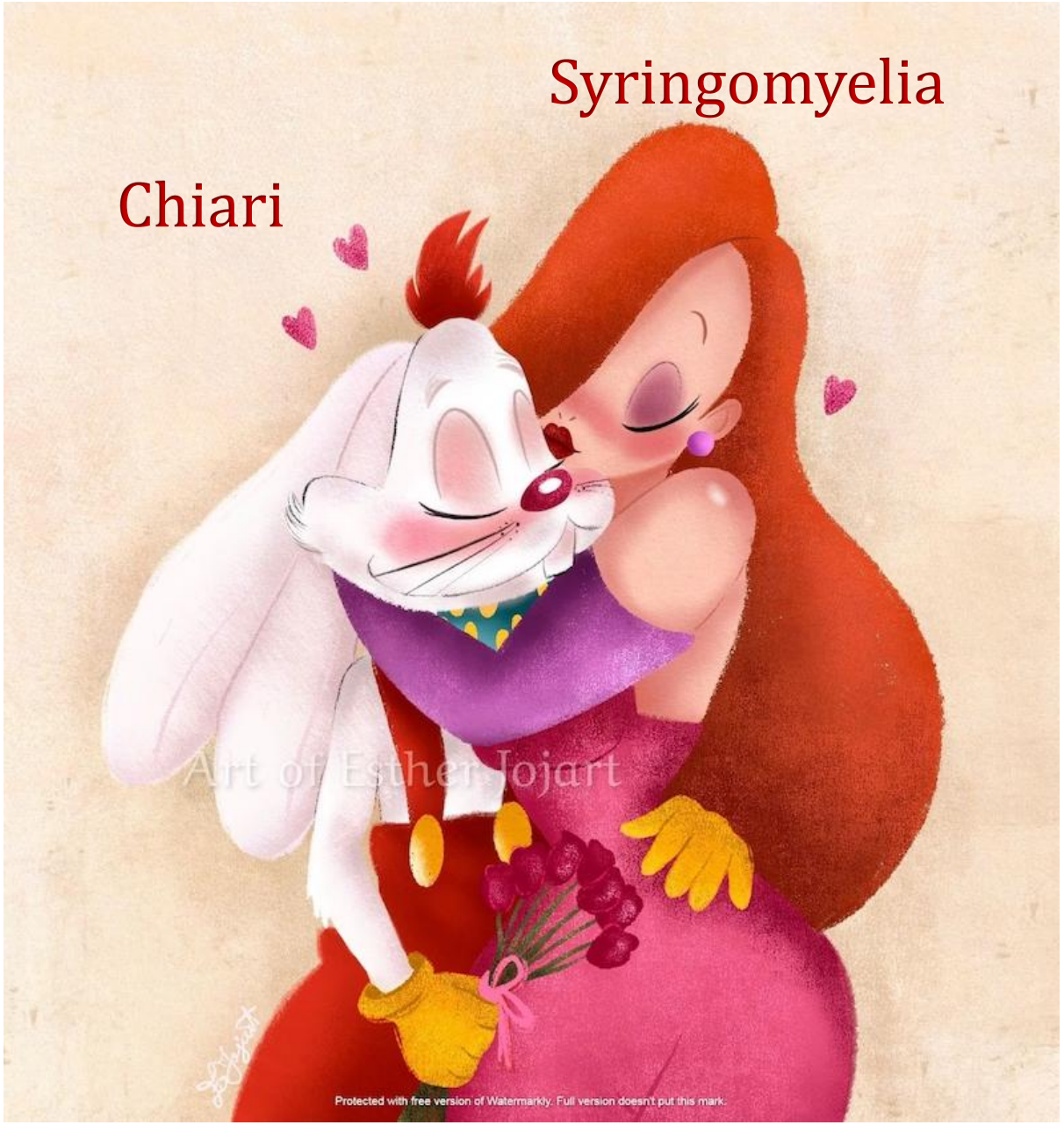
NeuroCHirurghi: Dr Valentini, Dr Vetrano, Dr Galbiati
NeuroPsichiatra Infantile: Dr Saletti
Neuroradiologi: Dr Chiapparini
Neurologi: Dr Braccia, Dr Curone (Dr Piscosquito, Dr Golfre, Dr Straccia)
Neurofisiologo: Dr Devigili, Dr Lanteri
Neurortopedico: Dr Costa

Specialisti Esterni

Ortopedico: Dr Luca
Psicologo: Dr Gaudino

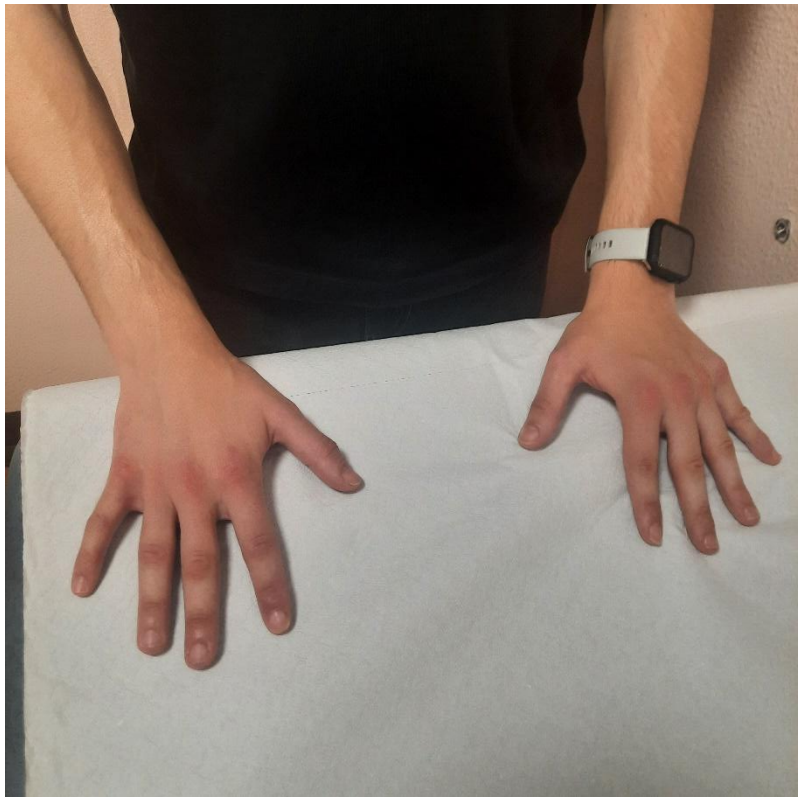
Chiari:

- Headache
- Dizzines
- Gait disturbance
- Ataxia
- Diplopia
- Nystagmus
- Bulbar Signs



Syringomyelia:

- Scoliosis
- Motor deficits
- Sensory Deficits
- Bulbar signs
- Cronic Pain

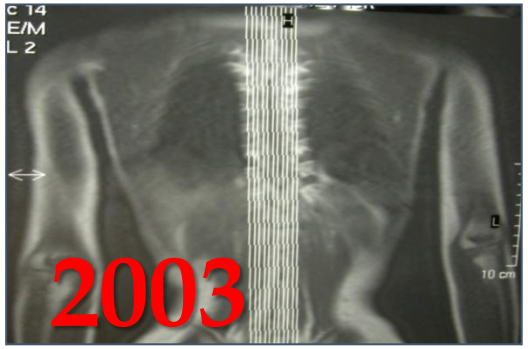


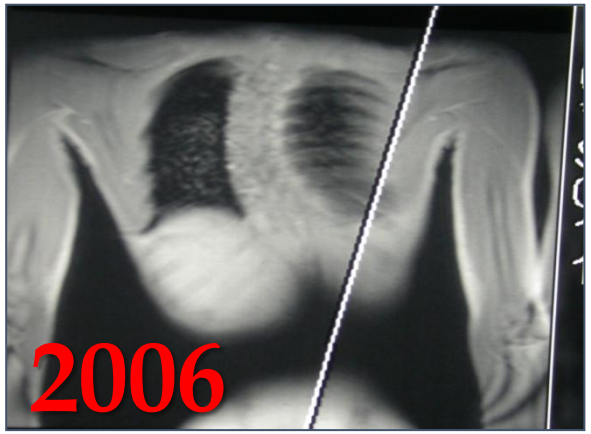
Syringomyelia:

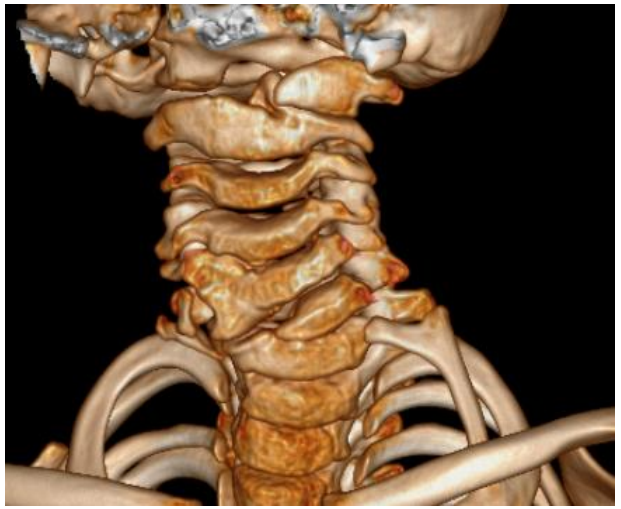
- Scoliosis
- Motor deficits
- Sensory Deficits
- Bulbar signs
- Cronic Pain

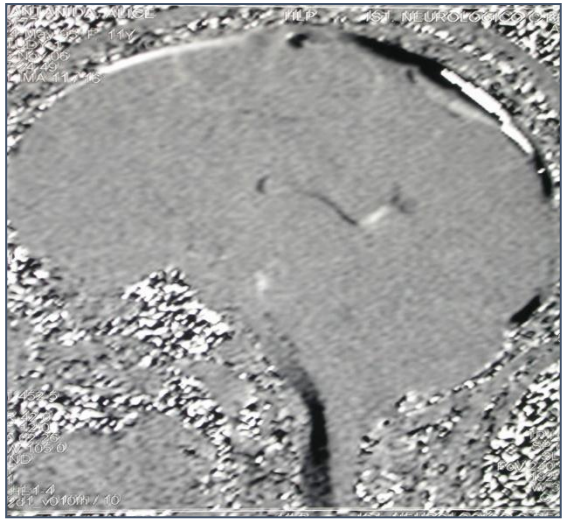
Siringomielia: Ruolo dell'**Ortopedico**

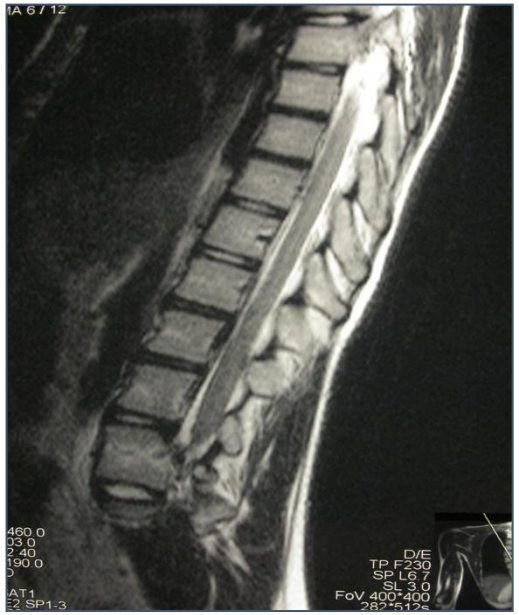
Scoliosi
Rapidamente
Evolutiva

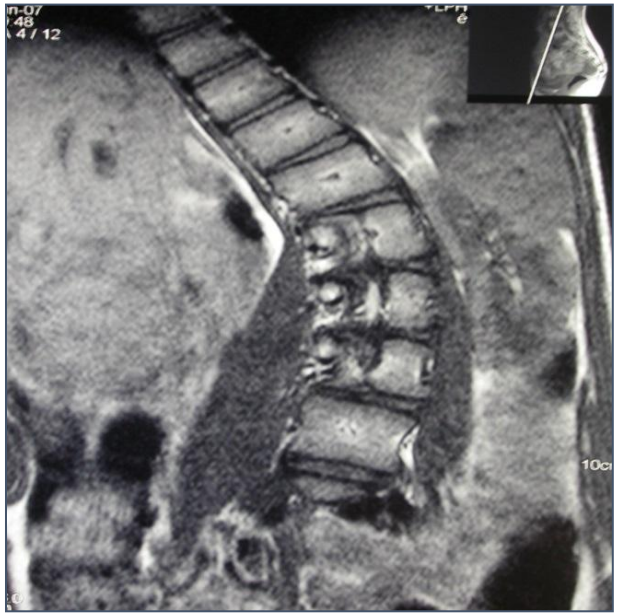












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PEDIATRIC NEUROLOGY & NEUROSURGERY, MILAN

THE ORIGINS - DISCLAIMER

Istituto Neurologico IRCCS Carlo Besta, MILAN

The Origins

- **1918** Carlo Besta, Neuroscientist, founds the
- r CNS and
- , Milan
- urrent
- ychiatry,
- 882)
- osurgery,
- ning, 1900)
- , 1934)

1937:
First Chiari Patient!
Foramen Magnum
Decompression

Institute Neurologico IRCCS Carlo Besta, MILAN

The Chiari's Experience

Besta's Series

Children: 360 pts

ReDos: 63 pts

69 41 altri, 28 FINCB

607

Adults: 620 pts

Istituto Neurologico IRCCS Carlo Besta, MILANO

Chiari & Siringomielia

MIGL 33%

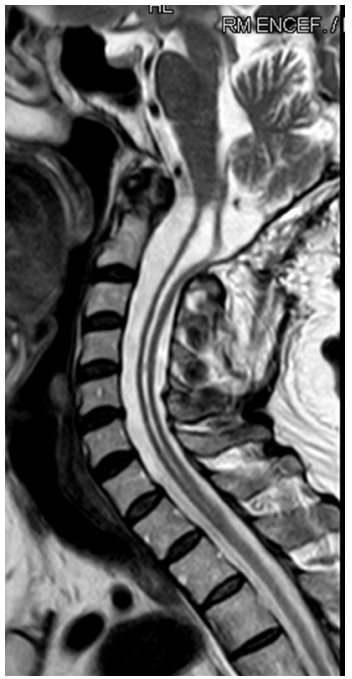
%?

Sproporzione CBL-Fossa Posteriore → Asintomatici, Sintomi Lievi → Sintomi Tipici, Iniziale Siringomielia → Sintomi Invalidanti, Siringomielia → Deficit Neurologici, Scoliosi → Deformità, Dolore Cronico, Atrofia Midollare

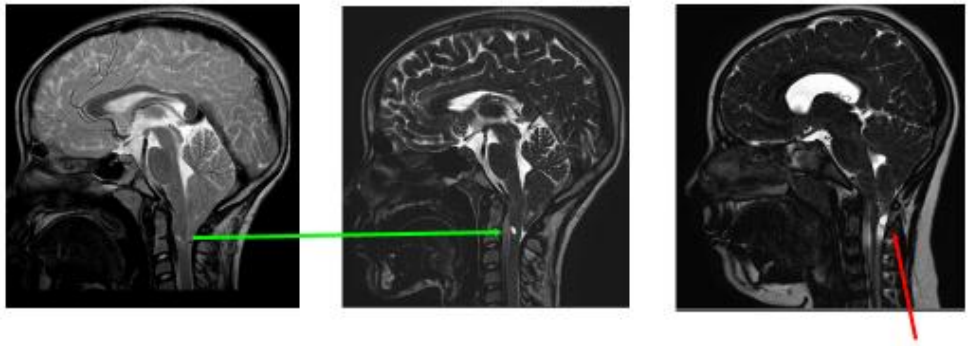
%?

PEGG 17%

Indicazione all'Intervento



Symptomatic Phase due ProgressiveTonsils Descent
Critical Period along growth in adolescence



Tonsils seems Suckled By Spinal Hypotension
..Cystic Transformation..

Rachicentesi, Traumi

AR con Instabilità, Osteopetrosi

CAUSE PEGGIORAMENTO

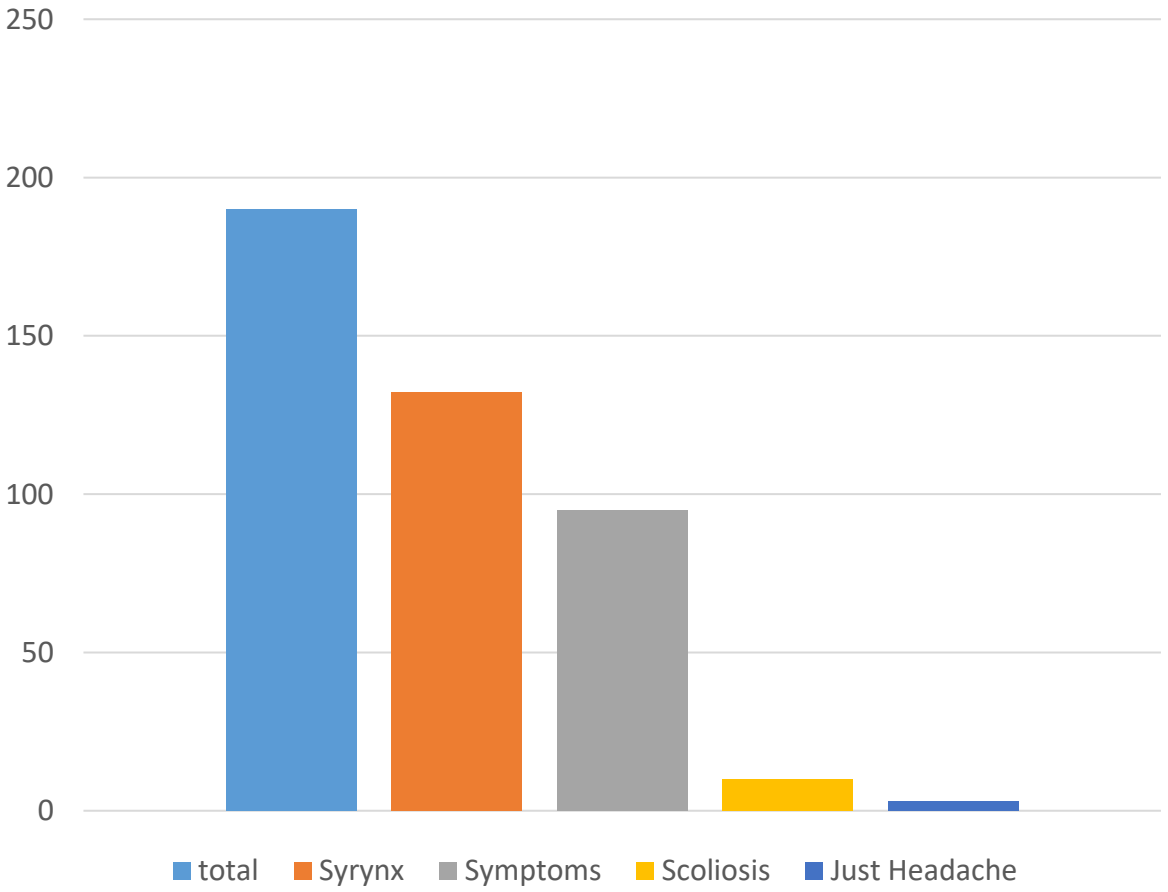
Besta's Series

Chiari Malformations & Syringomyelia & Symptoms

Children: 190 pts

Mean age: 10,8 yrs

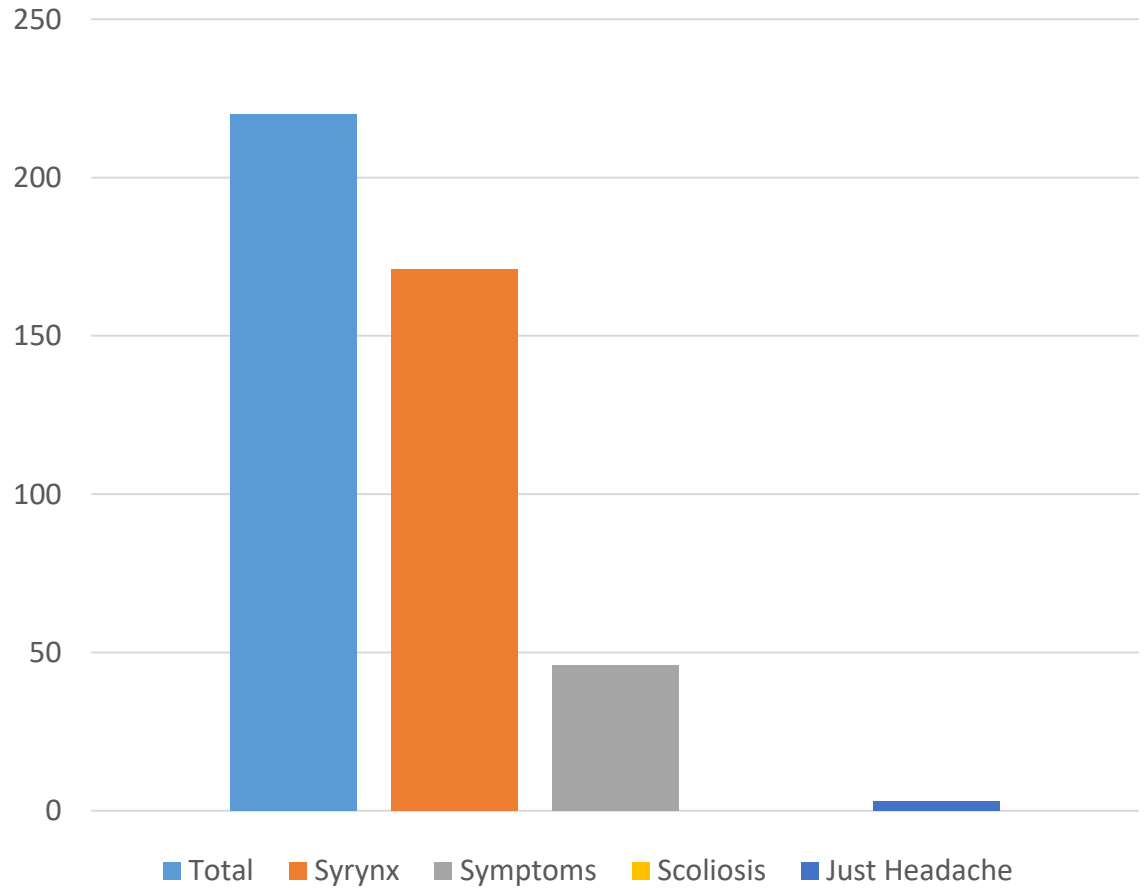
Very Young <6: 23%



Adults: 220 pts

Mean age: 44 yrs

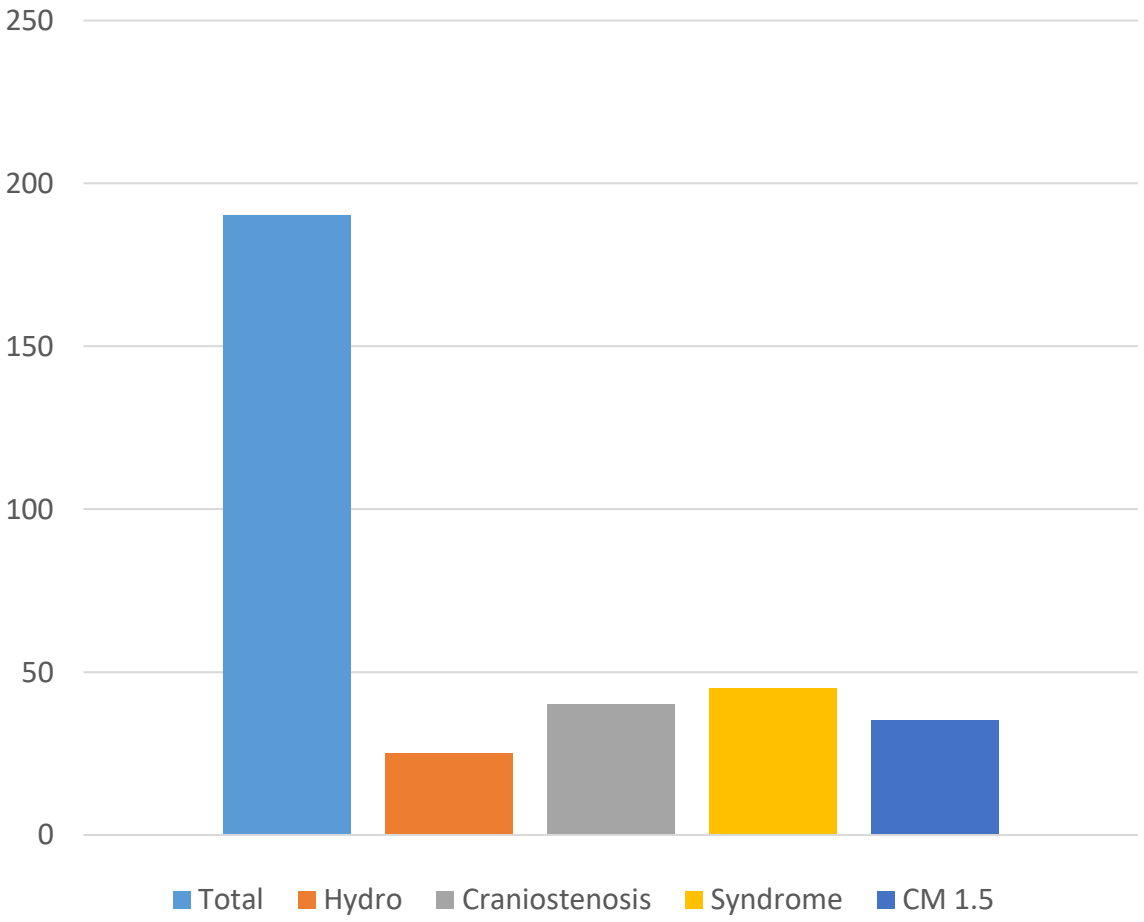
Females: 70%



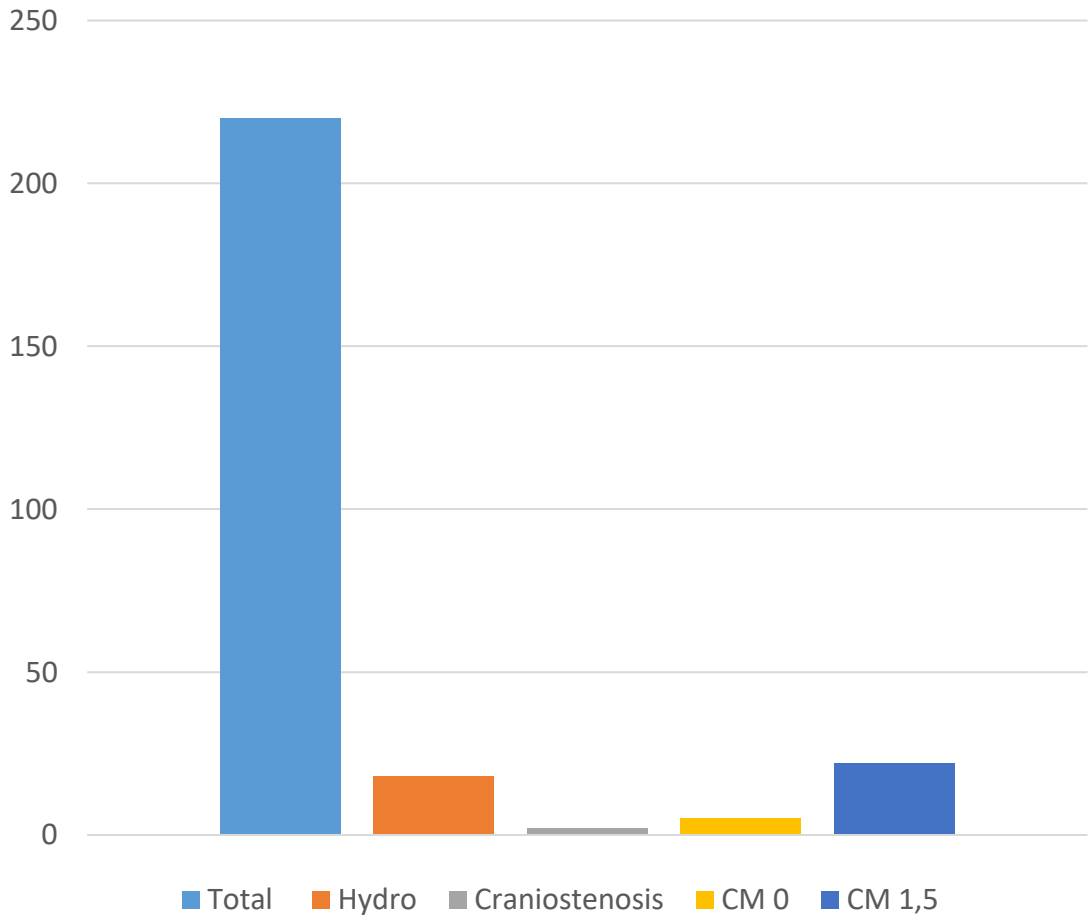
Besta's Series

CM1 & Associated Malformations

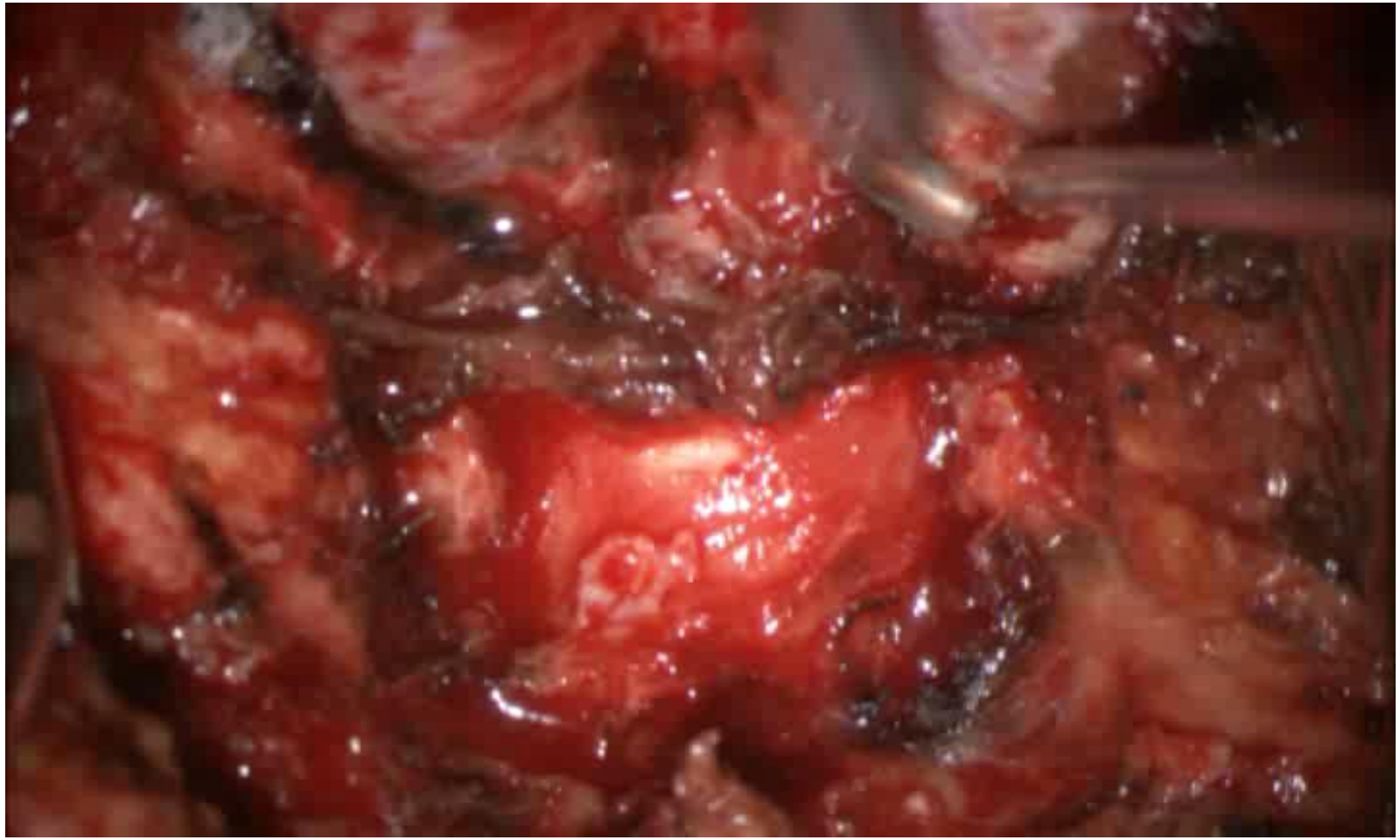
Children: 190 pts



Adults: 220 pts

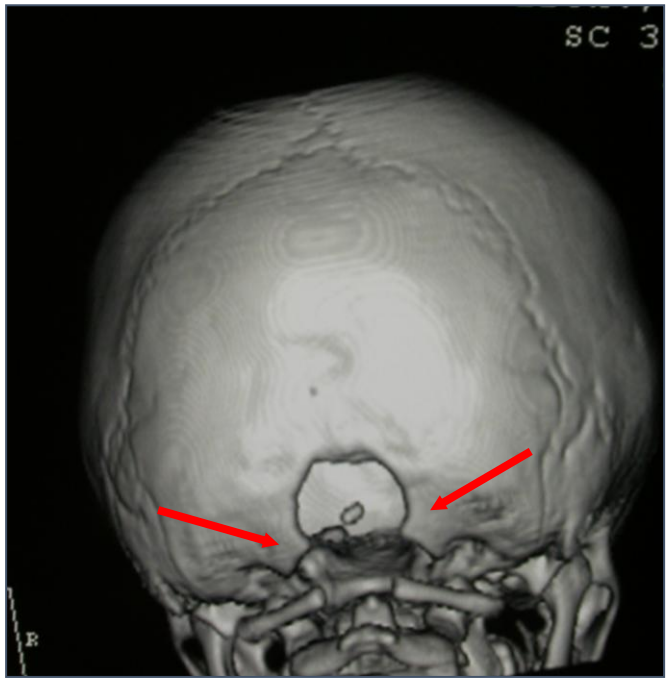


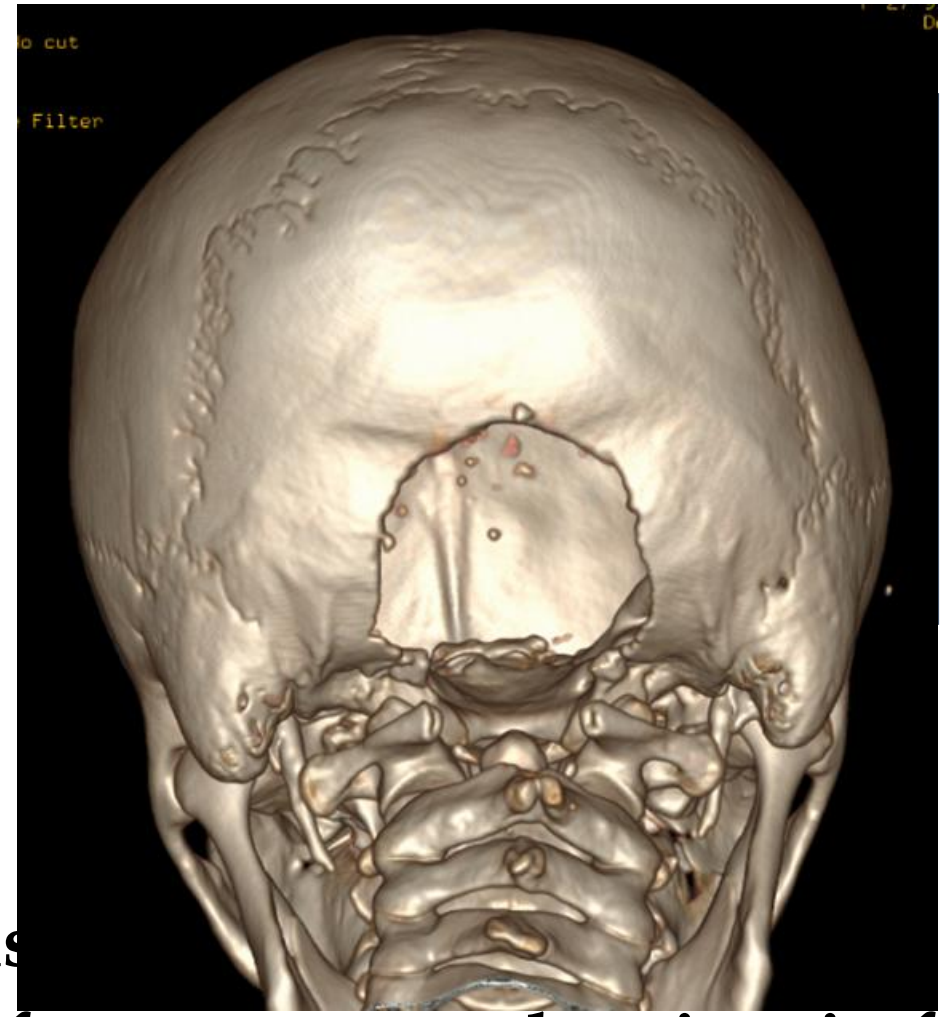
SURGICAL TECHNIQUE
Bone Decompression

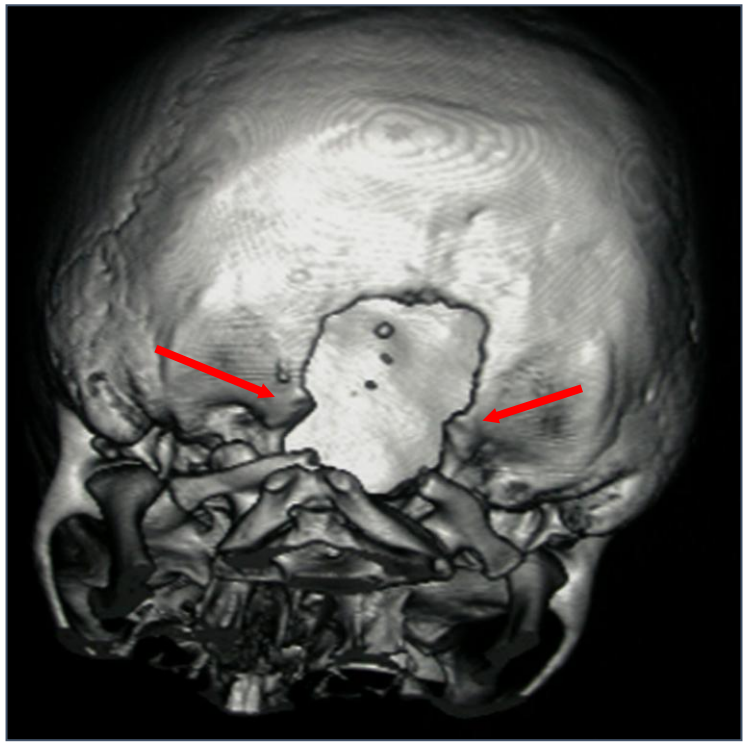


Step 1 BONE DECOMPRESSION

Enlarged enough at the foramen ??







In 75 cases CSF puls

→ rapid regression of symptoms and syrinx in few months

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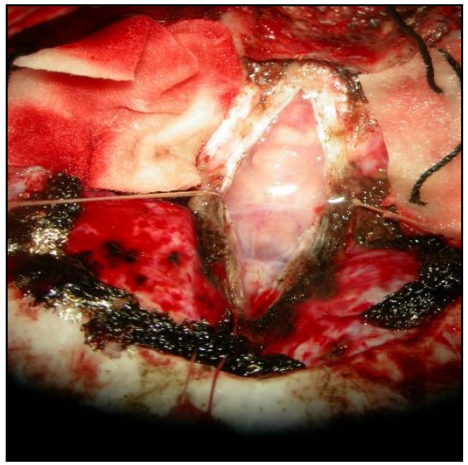
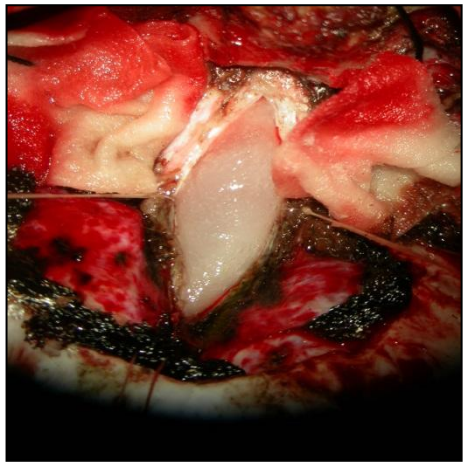
CM1

A couple of tricks to prevent CSF Leakage

1





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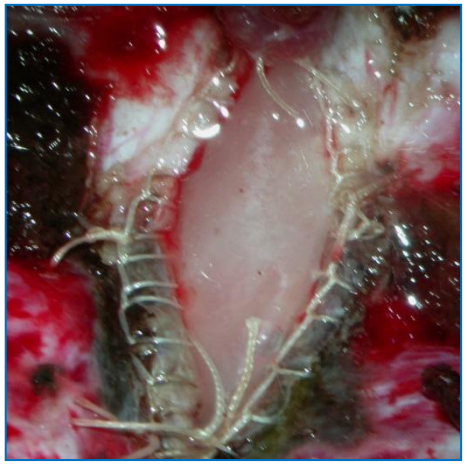
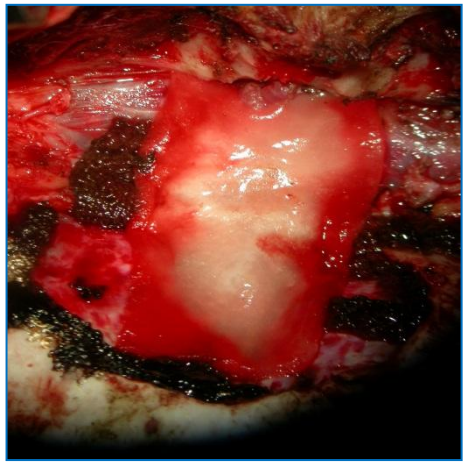
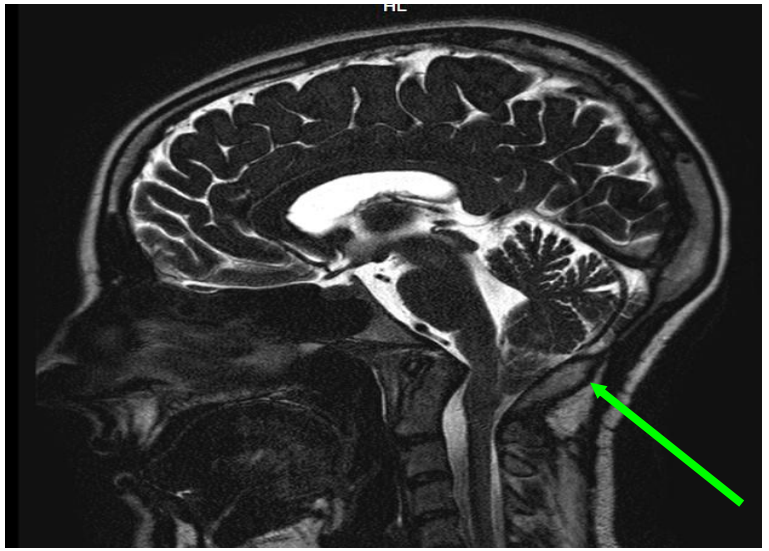


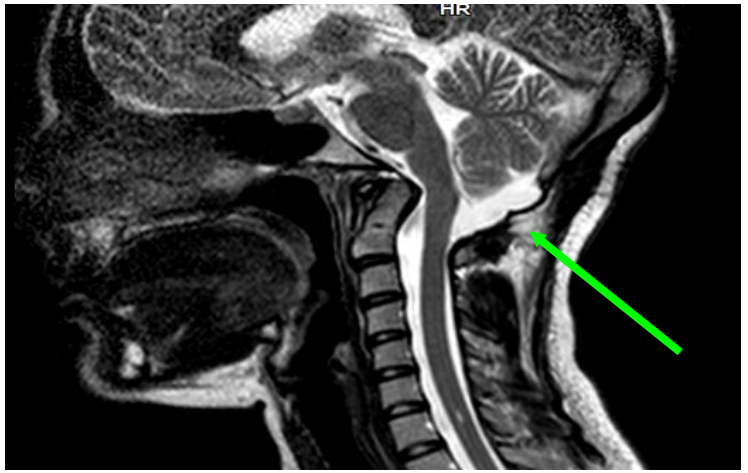


1. Insertion of Dural Substitute between Arachnoid and Duroplasty

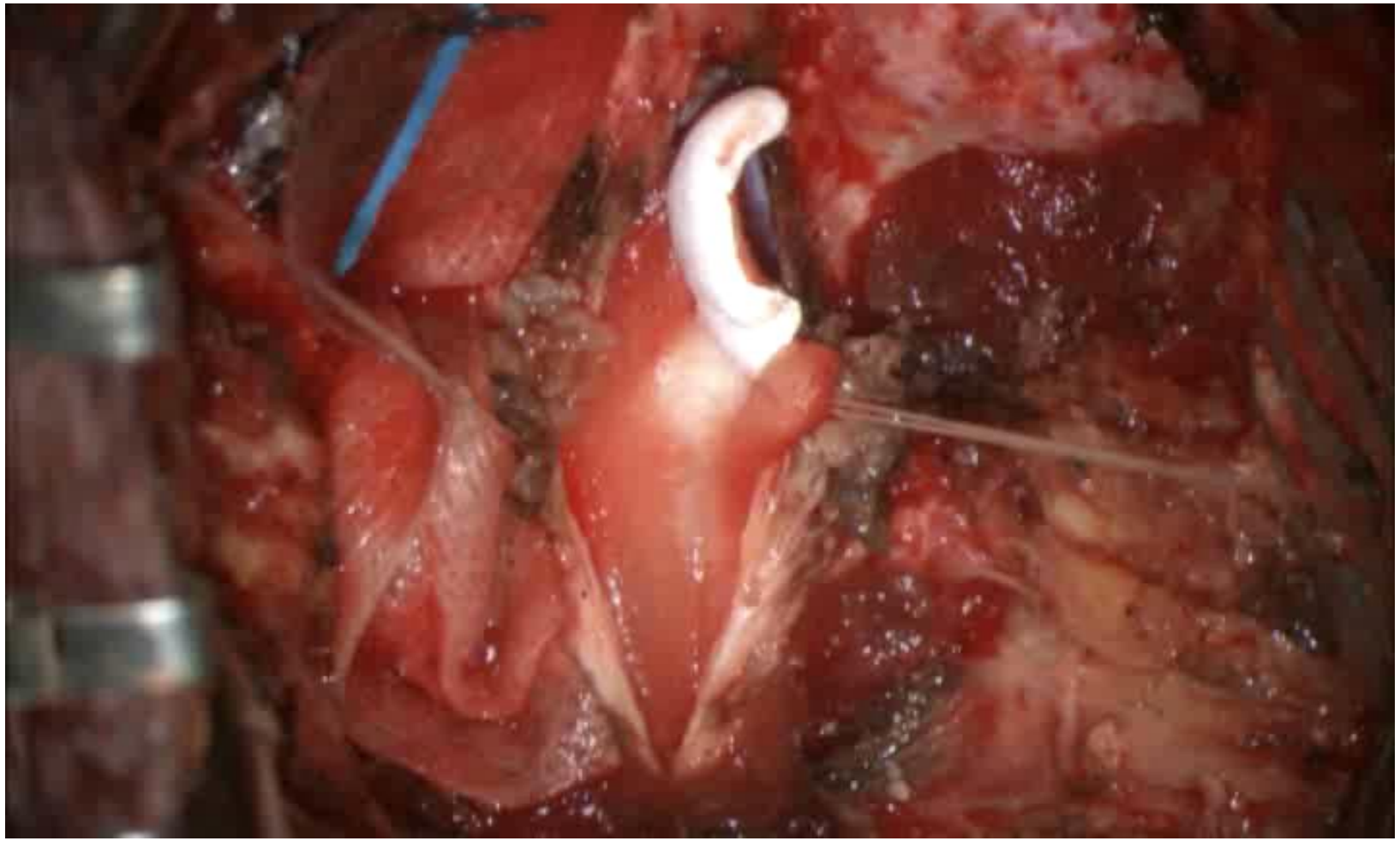
2. Suturing the Plasty by an unreasorbable stitches (Prolene or Goretex)

No CFS collections!





ARACHNOIDAL REPAIRING



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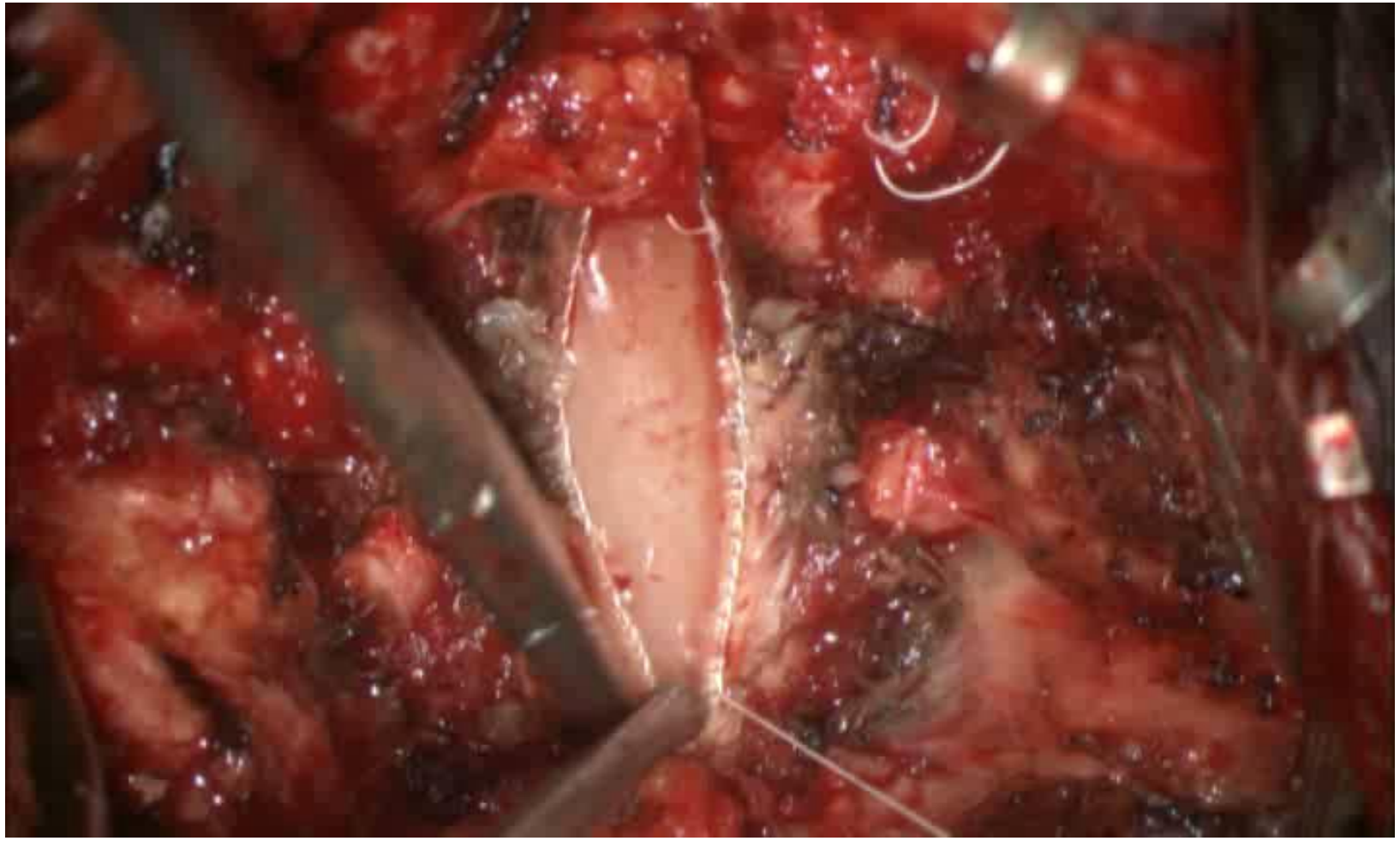
ISTITUTO
NEUROLOGICO
CARLO
BESTA

OBSESSIVE DURAL CLOSURE



+ Compressive Bandage

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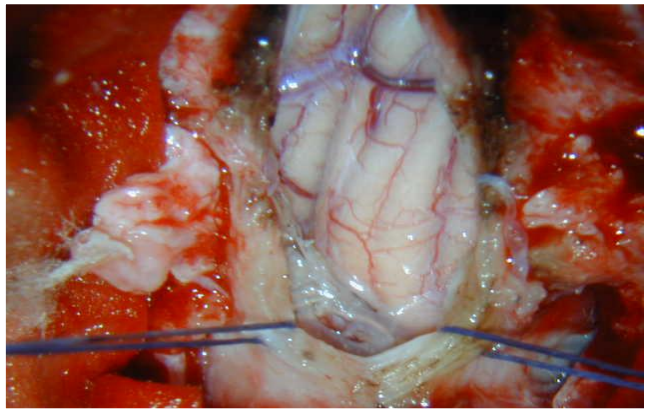
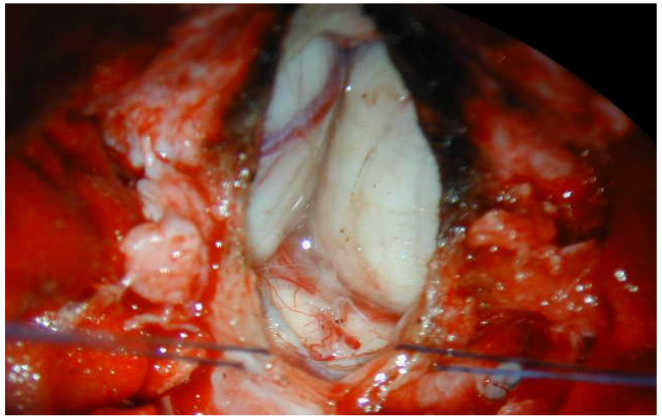
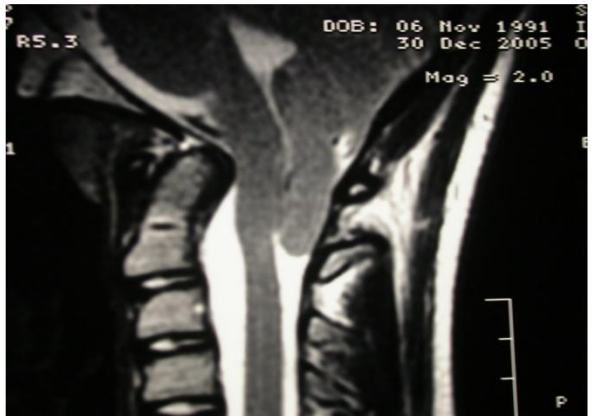
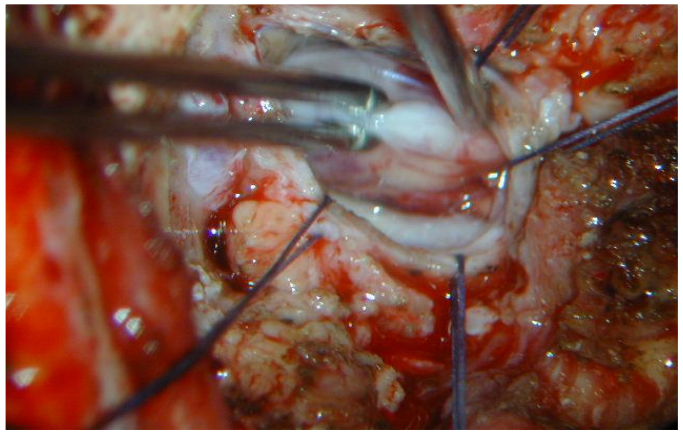
CVD + RESECTION OF CBL TONSILS

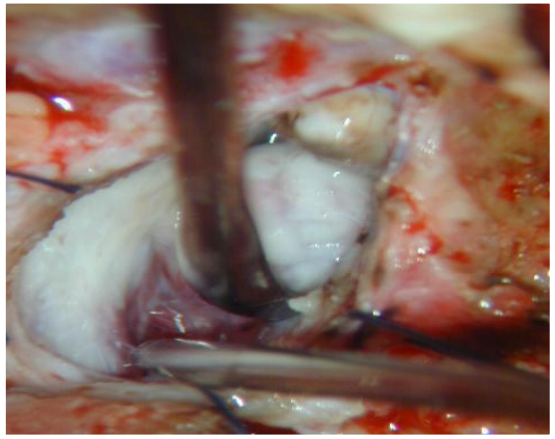


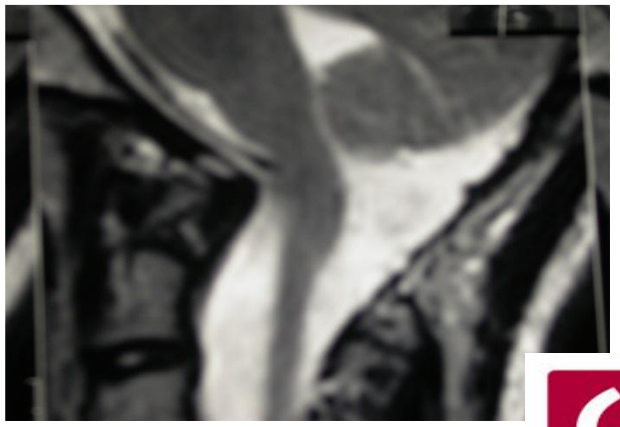




Coagulation and/or resection of CBL tonsils:
Second Choice
Or for relevant tosillar migration - severe crowding







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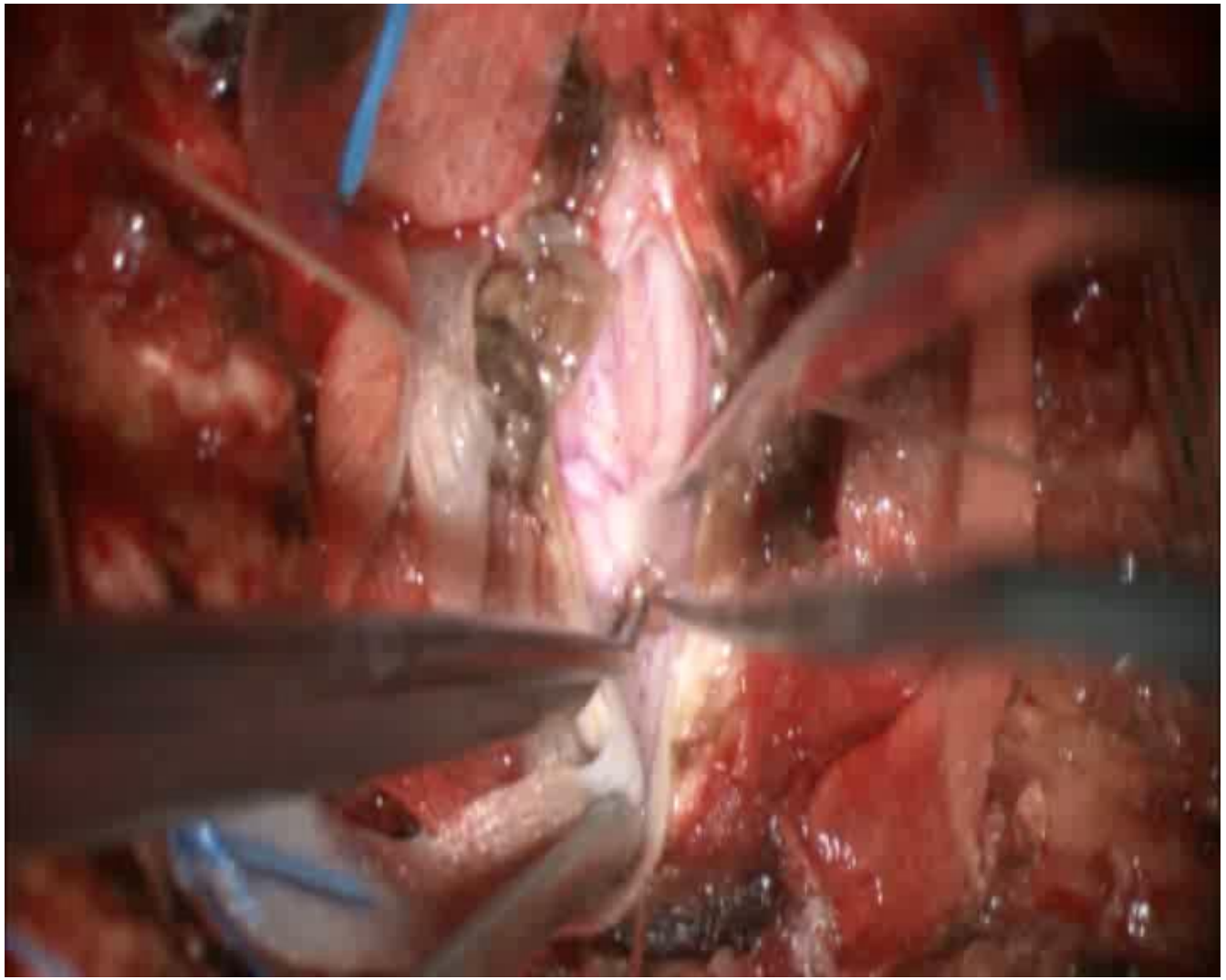
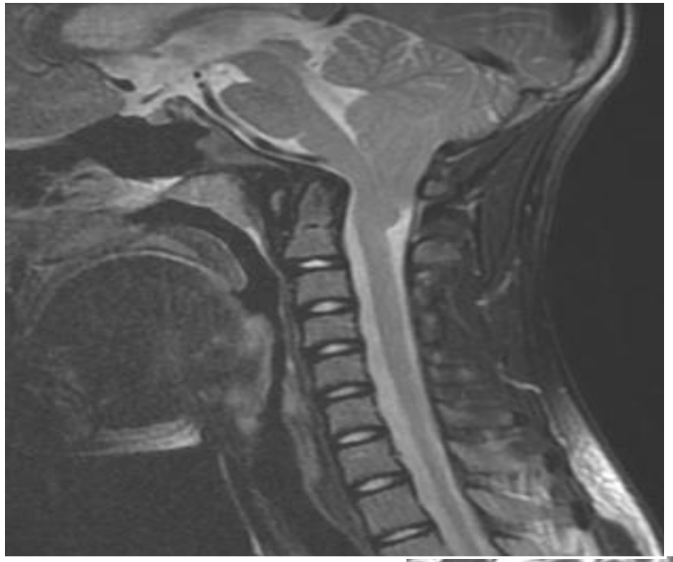
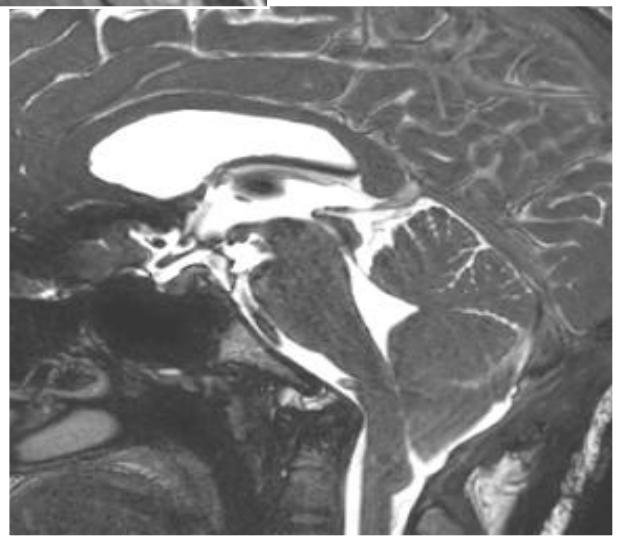
COAGULATION OF CBL TONSILS:

32% very low lying tonsils, severe overcrowding due to CM 1.5







…TRANS-ARACHNOIDAL

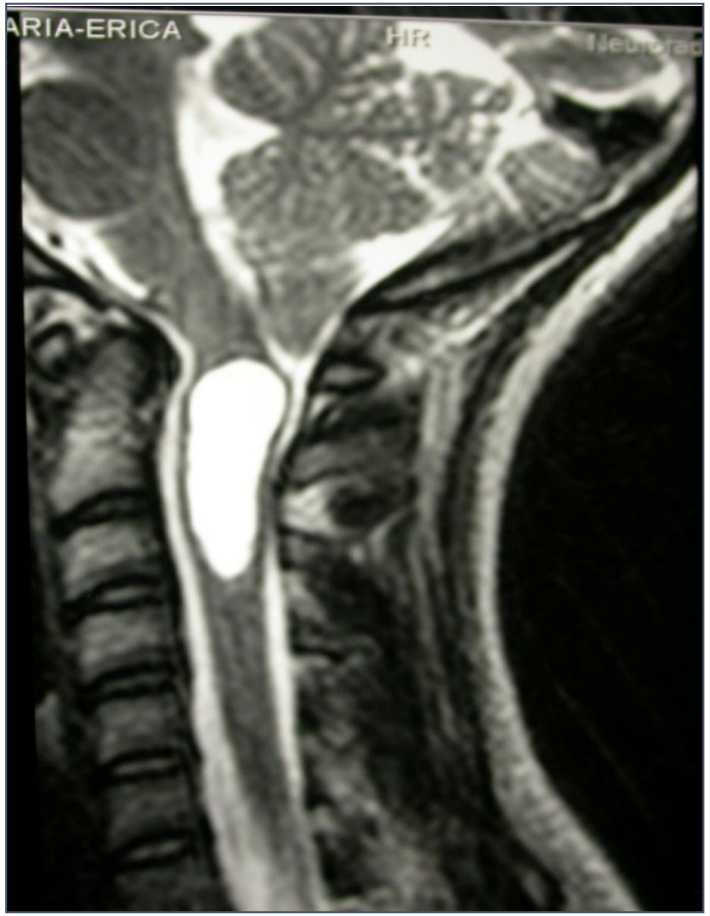
Besta's Series

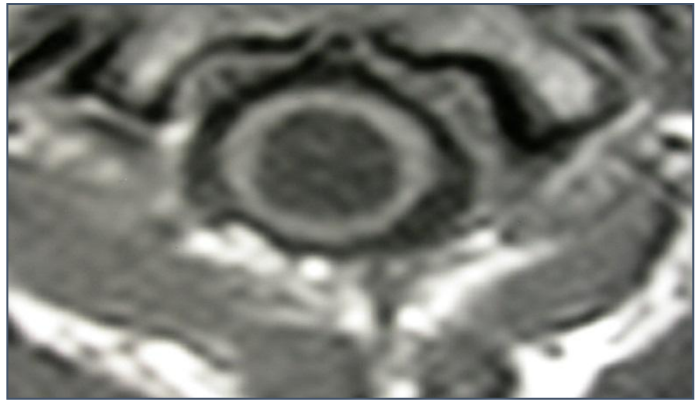
Type of Surgery & Results

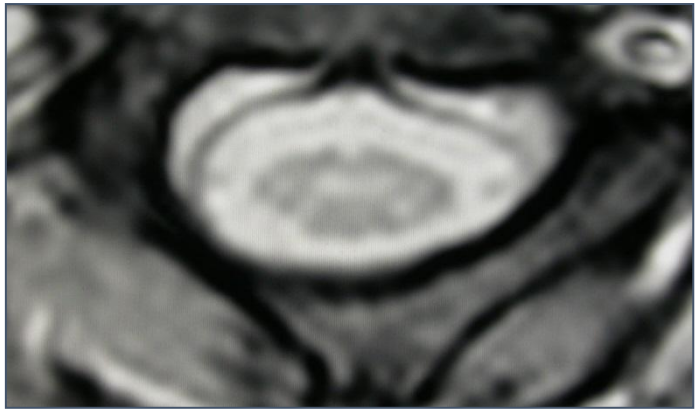
- Posterior Fossa Decompression with Durplasty (PFDD) was performed 96% of the cases IN HIGH ACCORDANCE WITH CONSENSUS DOCUMENT
- No mayor complications
- Minor Complication Rate (CSF leakage/collection) 10%, mainly solved by medication and bendage, only 2.3% deserving review surgery
- Mean Time of Hospitalization: 4,5 days
- Associated Hydrocephalus in 10,5% of cases, treated before PFDD just in 28 cases (65%) and the same occurred for associate Craniosynostosis LOW ACCORDANCE WITH CONSENSUS DOCUMENT

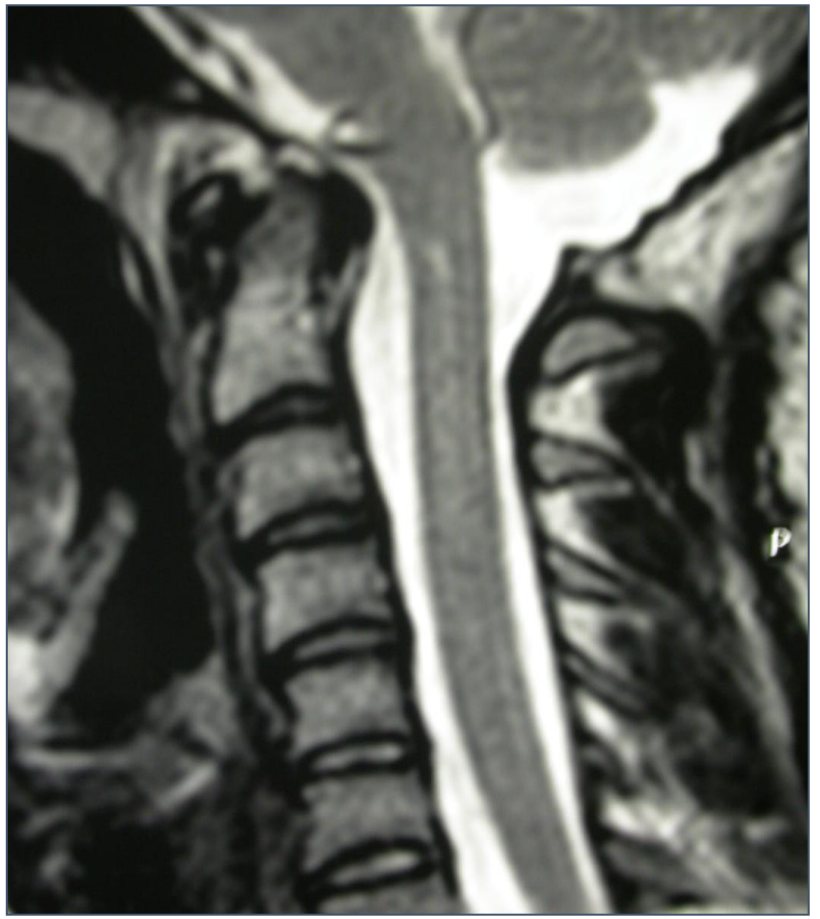
Besta's Series

Results on Syringomyelia: *Shrinkage 206 (96%)*









D.P.E., 13 years, Female:

Syrinx disappearance 8 months after surgery

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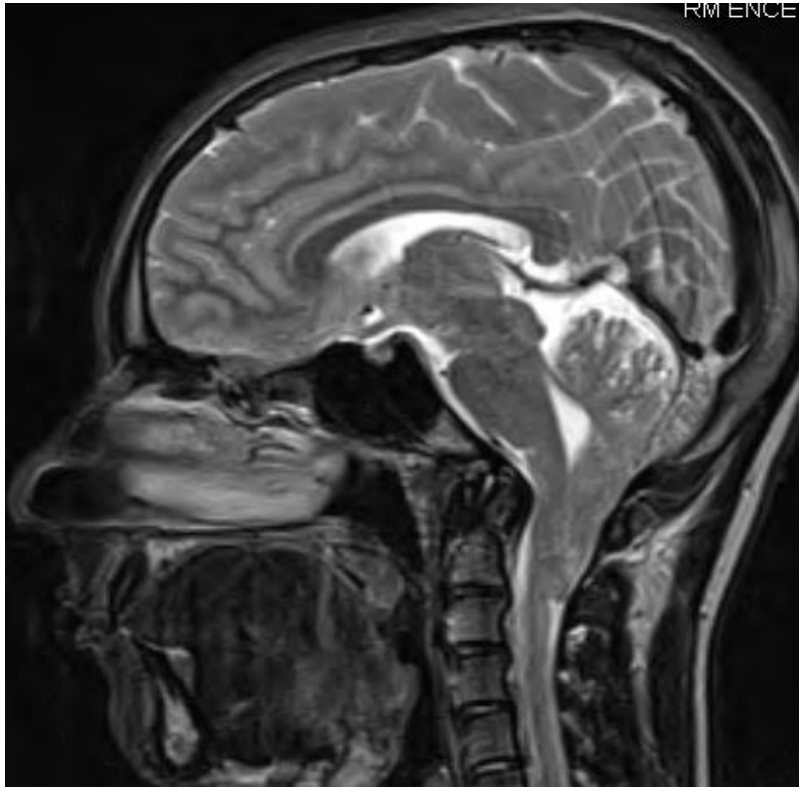
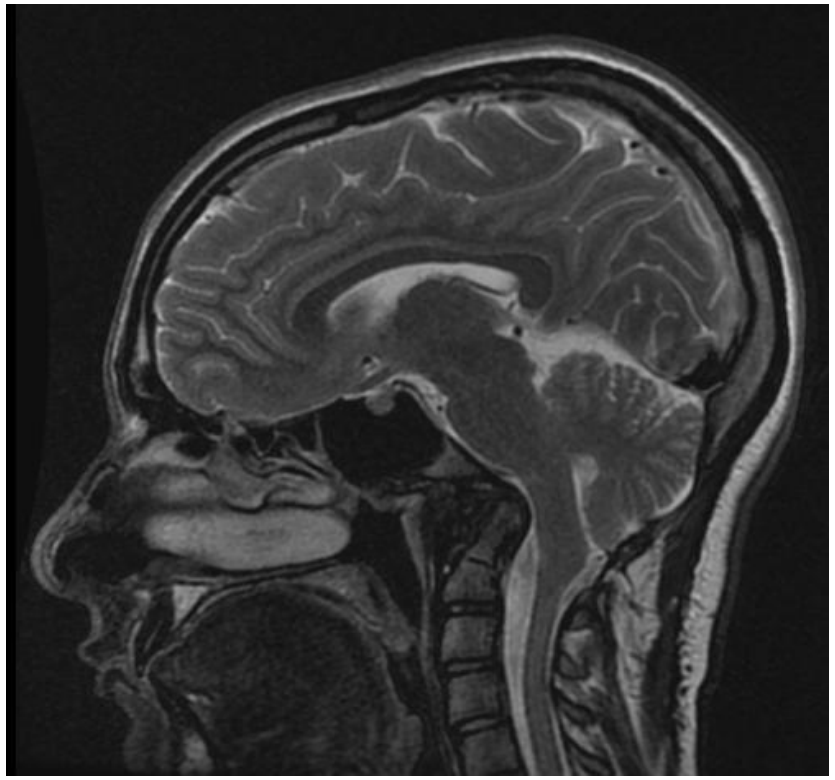
Besta's Series

Results on Symptoms: *Resolution 95%*





C. D., 11 yrs, male: cerebellar syndrome

→ Regressed after surgery

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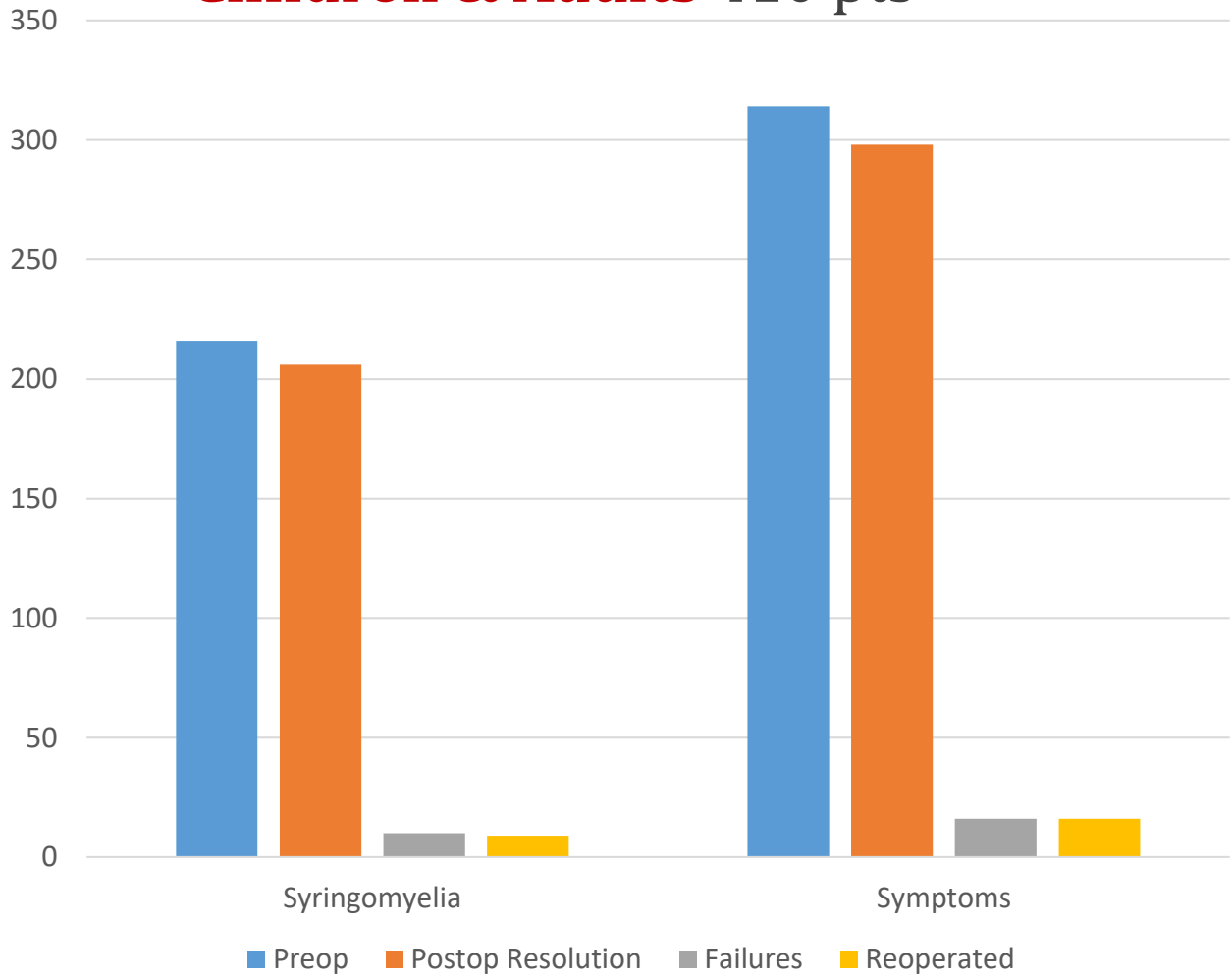
Besta's Series

Surgical CM1 - Long Term Results

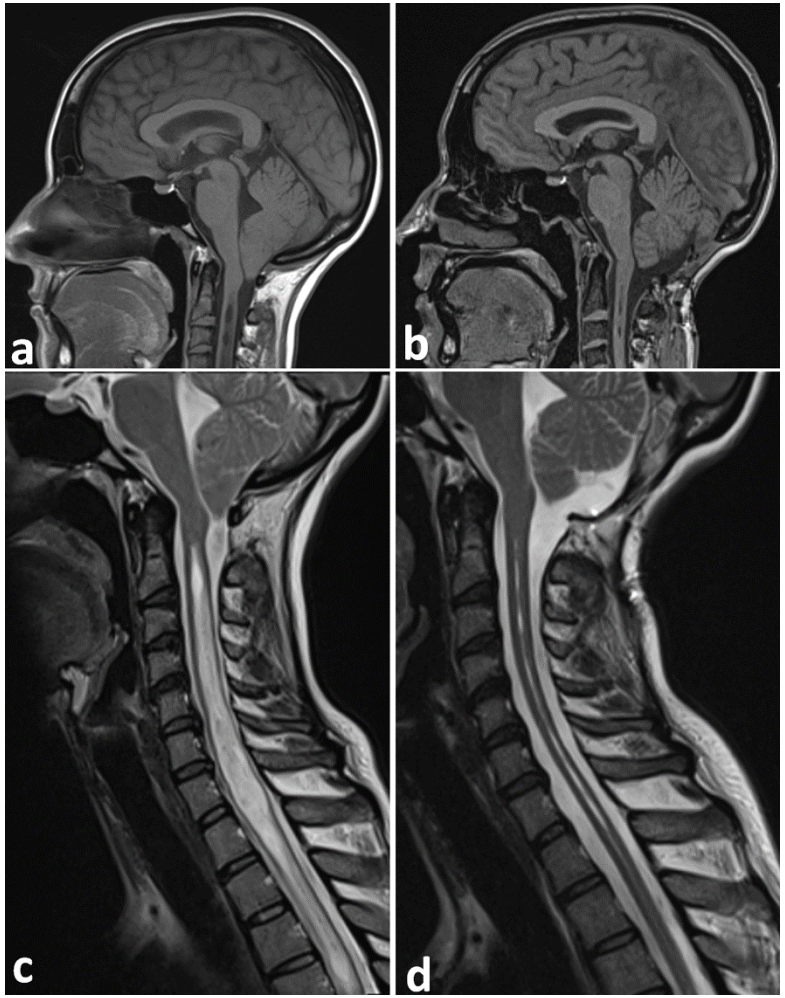
Mean F.Up 4,5 yrs

Children & Adults 410 pts



Pediatric CM1:

What happens if the operation is postponed in adulthood?



RESULTS OF SURGERY CHILDREN vs ADULTS

CM1: personal Surgical Series – Results of First Surgery

	190 Children	220 Adults	Total
Sex: Males Females	89 46,8% 101 53,2%	65 29,5% **155 70,5%**	410 pts
Age: Range Mean	9 m-18 yrs 44 Very Young <6yrs 10,8 yrs	19-77 yrs 41 yrs	
Associations: Syringomyelia Hydrocephaus Craniosynostosis	130 68,4% 25 13,1% **40 21%**	195 88,6% 18 8,1% 2 0,9%	325 79%
Complications: Not Reopetated Reoperated			43 10,9% 9 2,3%
Clinical Results: Improved Worsened			314 16
Syringomyelia: Reduced Increased	96 92,3% **8 7,7%**	**120 98,3%** 2 1,7%	216 95,5% 10 4,5%

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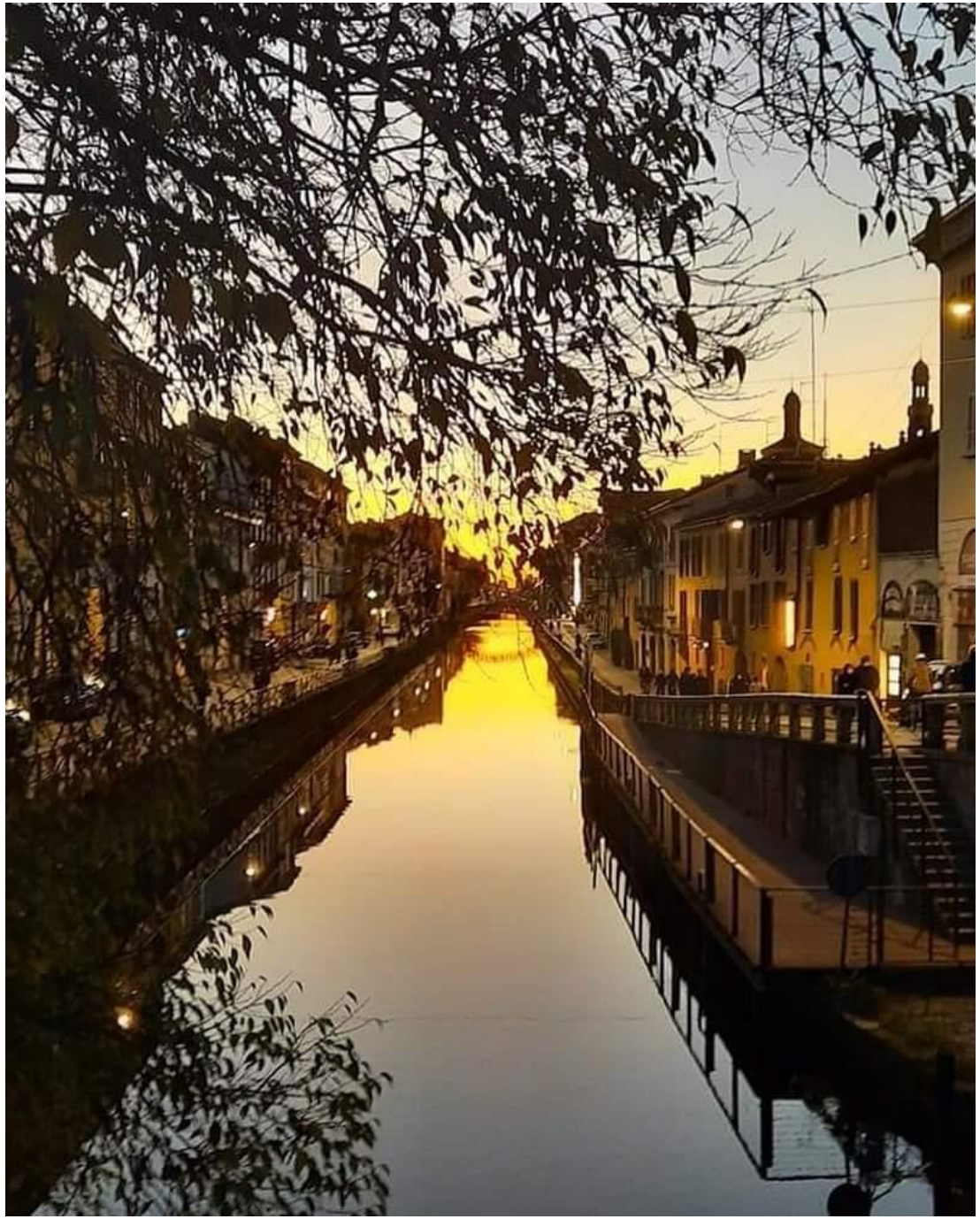
Cosa abbiamo imparato dai nostri:
Chiari Operati

- L'analisi della nostra casistica ha mostrato che i risultati a lungo termine della PFDD stati buoni, sia nel controllare i Sintomi, che per risolvere la Siringomielia
- Questo risultato è legato all'alto volume di casi trattati al Besta, che ci ha permesso di ottimizzare nel tempo la tecnica per la PFDD
- Ma sicuramente il buon risultato clinico è dovuto all'indicazione chirurgica, frutto di un'analisi precisa del gruppo multidisciplinare
- che ha seguito attentamente il Chiari Consensus Document 2019, uno strumento utile per standardizzare la cura dei pazienti affetti da Malformazione di Chiari e Siringomielia.



Grazie per l'attenzione !

..e per la collaborazione

Neurochirurghi: Dr Vetrano Dr Galbiati

NeuroPsichiatra Infantile: Dr Saletti

Neuroradiologi: Dr Chiapparini Dr Erbetta Dr Moscatelli Dr Doniselli Dr Parazzini Dr Righini

Neurologi: Dr Braccia Dr Curone

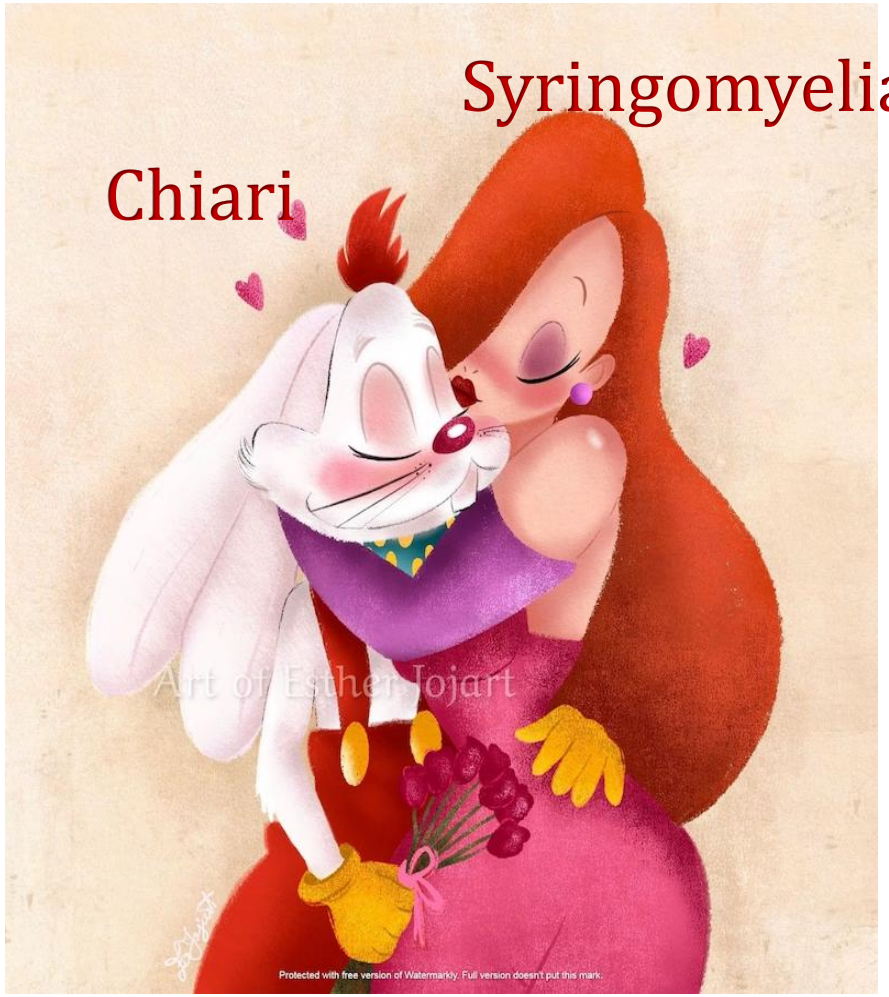
Epidemiologo: Mariangela Farinotti

Case Manager NCH Pediatrica: Sabrina Mariani

Complex Chiari = the Chiari PLUS..

Associated Malformatios:

- Craniosynostosis (Complex & Single)
- Hydrocephalus
- IICP



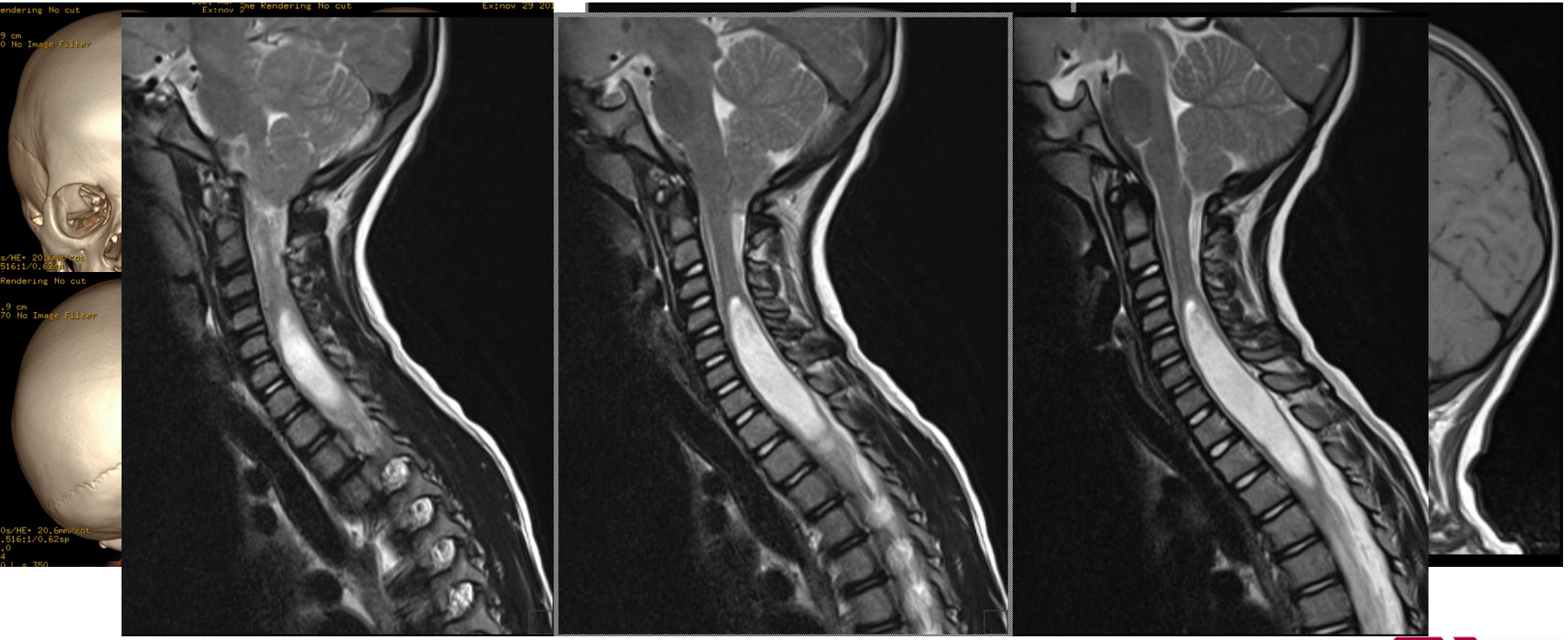
Chiari:

- Headache
- Dizzines
- Gait disturbance
- Ataxia
- Diplopia
- Nystagmus
- Bulbar Signs

Syringomyelia:

- Scoliosis
- Motor deficits
- Sensory Deficits
- Bulbar signs
- Cronic Pain

POLISYNOSTOSIS: how to evaluate the Chiari & Syringomyelia?



Extend the MRI on the whole Spine!

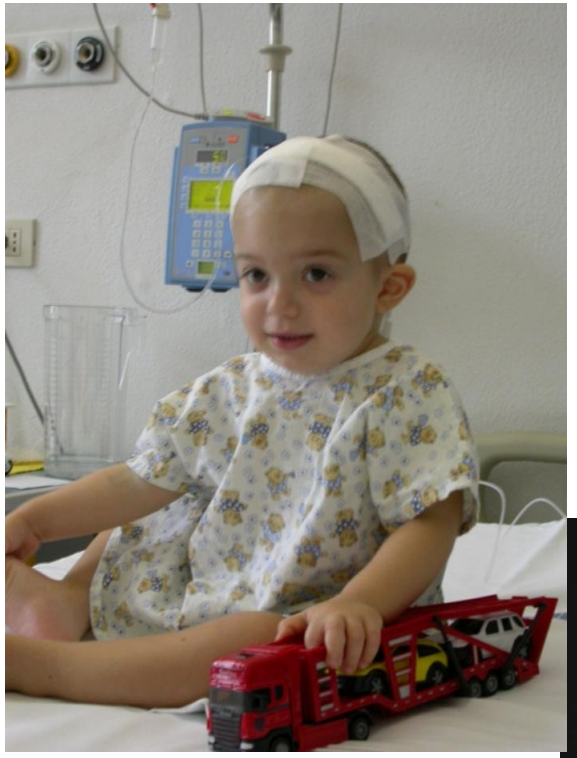
Posterior Calvarial Distraction **Surgical Series**

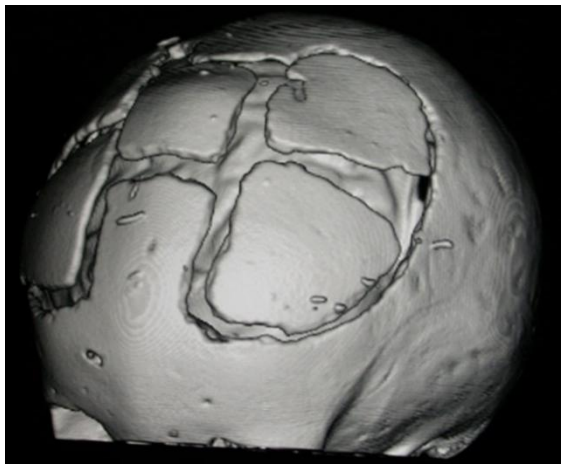
Retrospective study on **10 Children** affected by polisynostosis

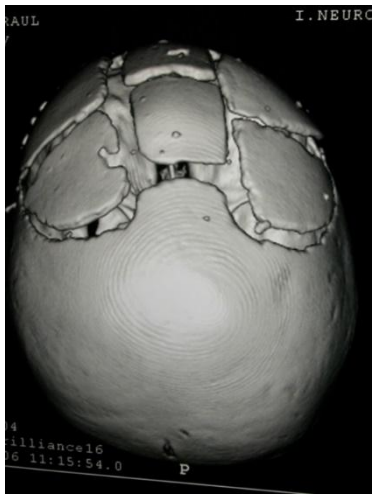
	Sex	Synostosis	Age/m	Syndrome	CM1	Syrinx	Hydro	ICP
B M	M	Bicoronal + Bilambdoidal	9	**Pfeiffer**	Y	N	Y	Y
L E	F	Plagio Ant + Bilambdoidal	48	**Muenke**	Y	N	N	Y
M D	M	Bicoronal+ Bilambdoidal	11	**Saethre Chotzen**	Y	N	N	Y
U R	M	Oxicefaly	145	**ERF**	Y	Y	N	Y
Y M	M	Plagio Ant → Bitemporal Sagittal & Bilambdoidal	13	**Hypophosphatemic Rickets, Rayne**	Y	N	Y	Y
O C	F	Bicoronal	7	**Frontonasal Dysplasia**	N	N	N	Y
C An	M	Bicoronal + Sagittal	52	**Noonan**	Y	Y	N	Y
S E	F	Plagio Ant → Sagittal & Bilambdoidal	119	**Saethre Chotzen**	Y	N	N	Y
C Au	F	Bicoronal	15	**Muenke**	N	N	N	Y
I S	M	Oxicefaly	20	**Hypophosphatemic Rickets**	Y	N	Y	Y

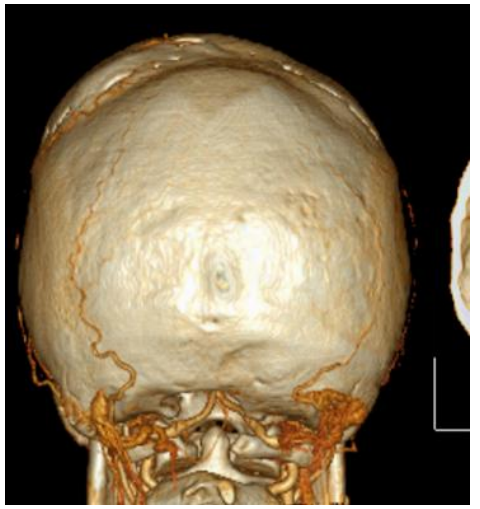
Mean Age at Surgery **44 months**

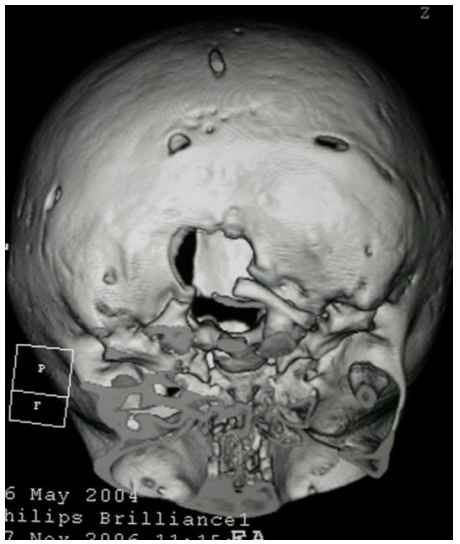
OXICEFALY: UR, Pansynostosis-ERF

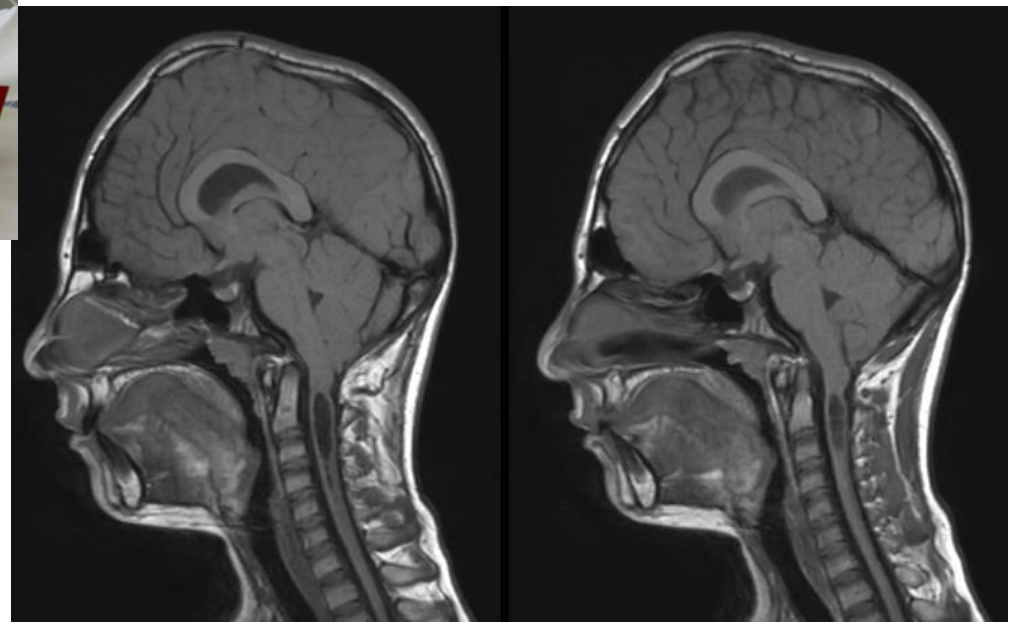


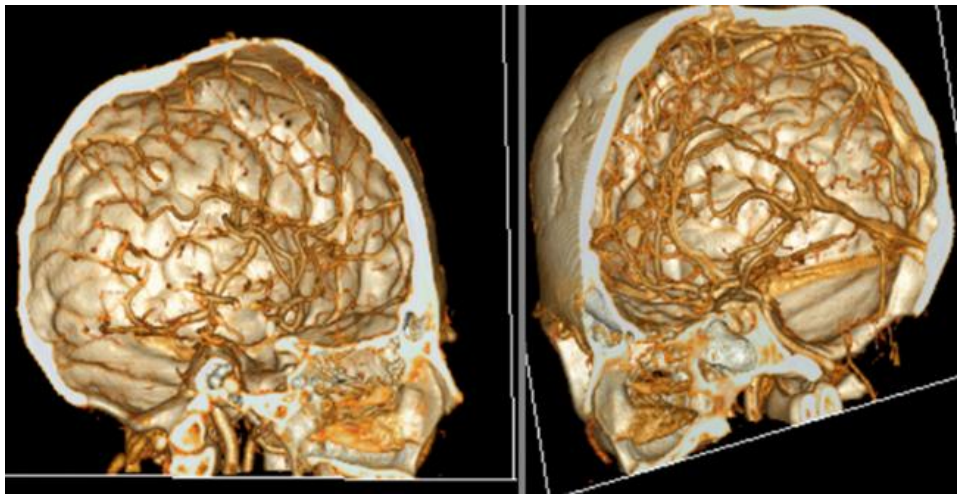












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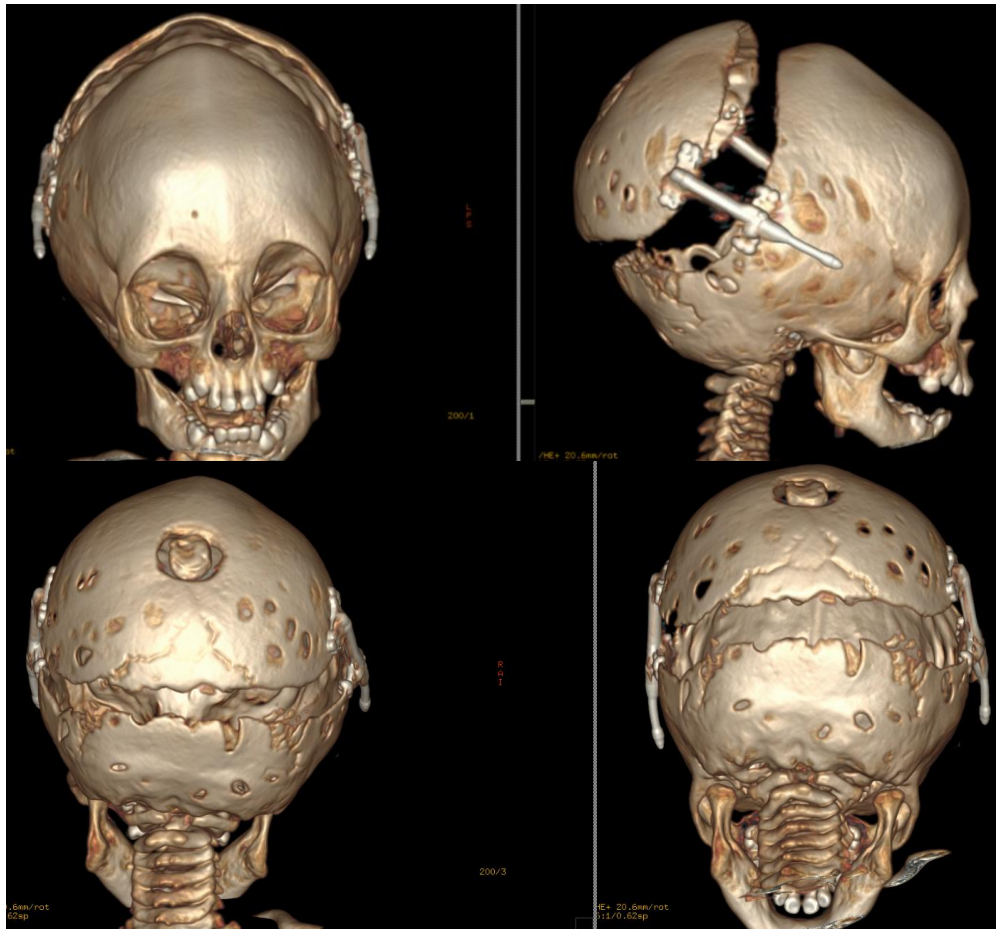
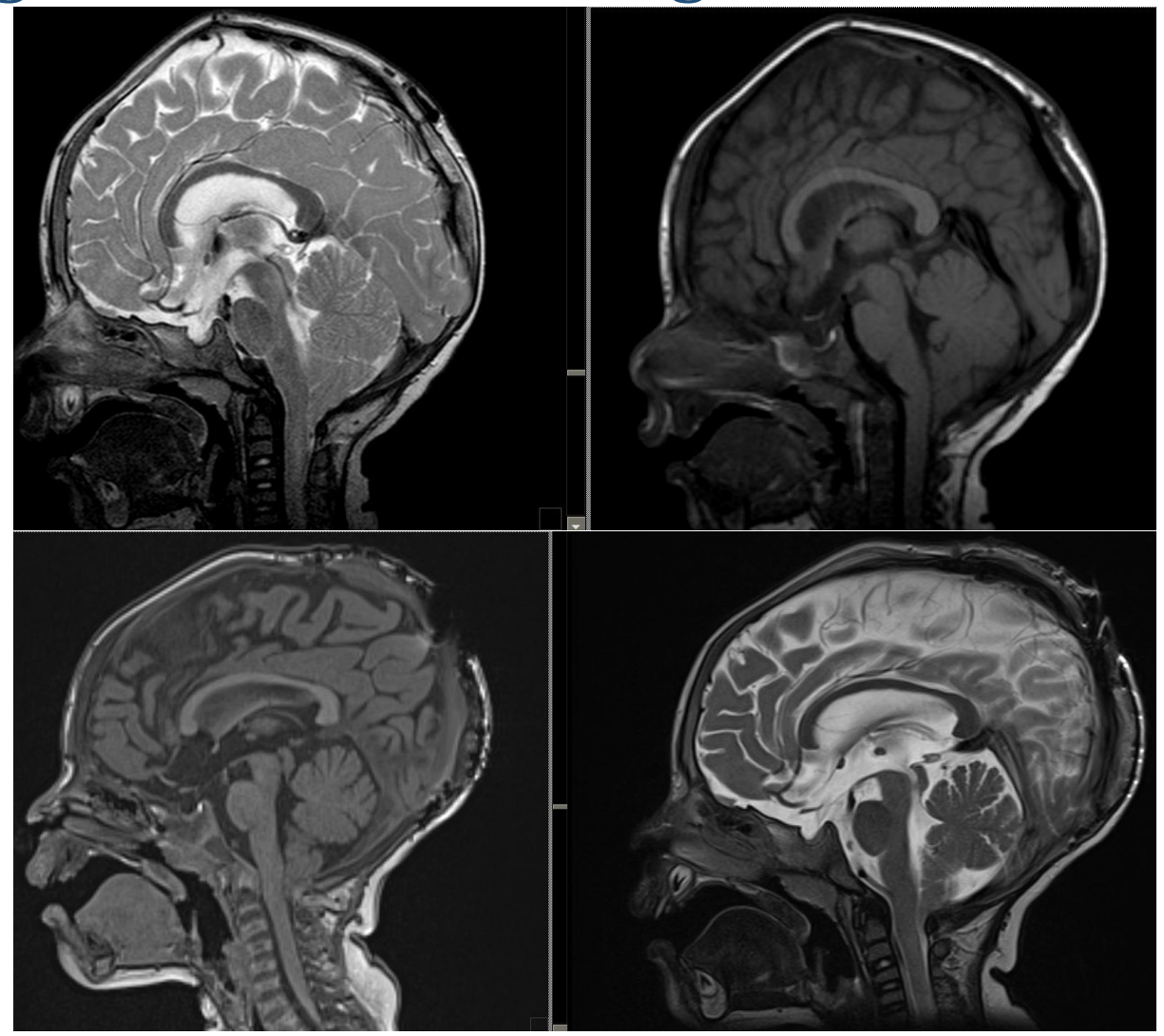
EUROPEAN CLEFT PALATE CRANIOFACIAL ASSOCIATION

Posterior Calvarial Distraction

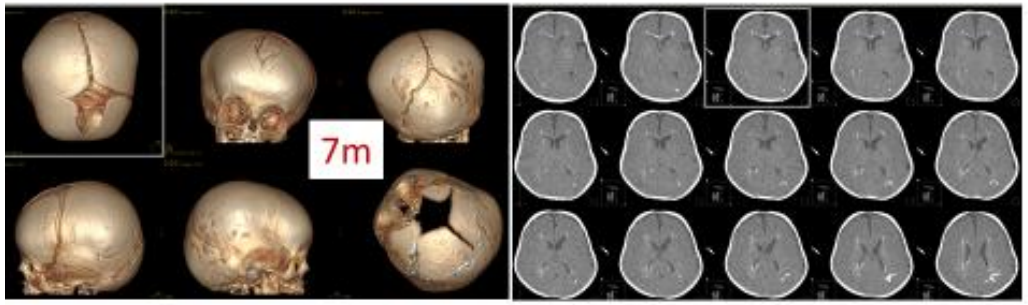
Reossification 10/10

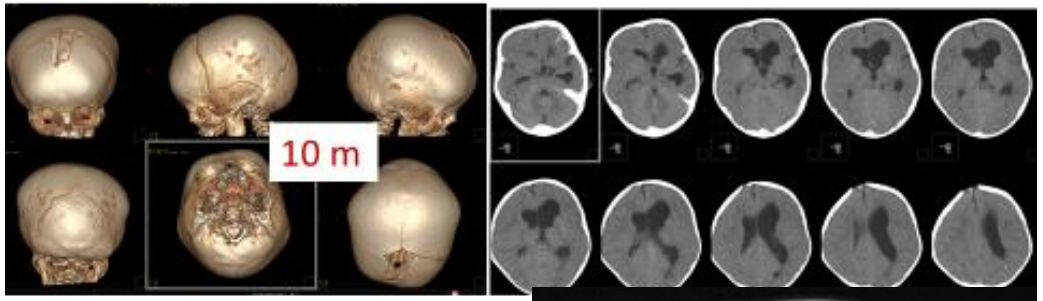
POSTERIOR CALVARIAL AUGMENTATION:
Care for Pathological Venous Drainage

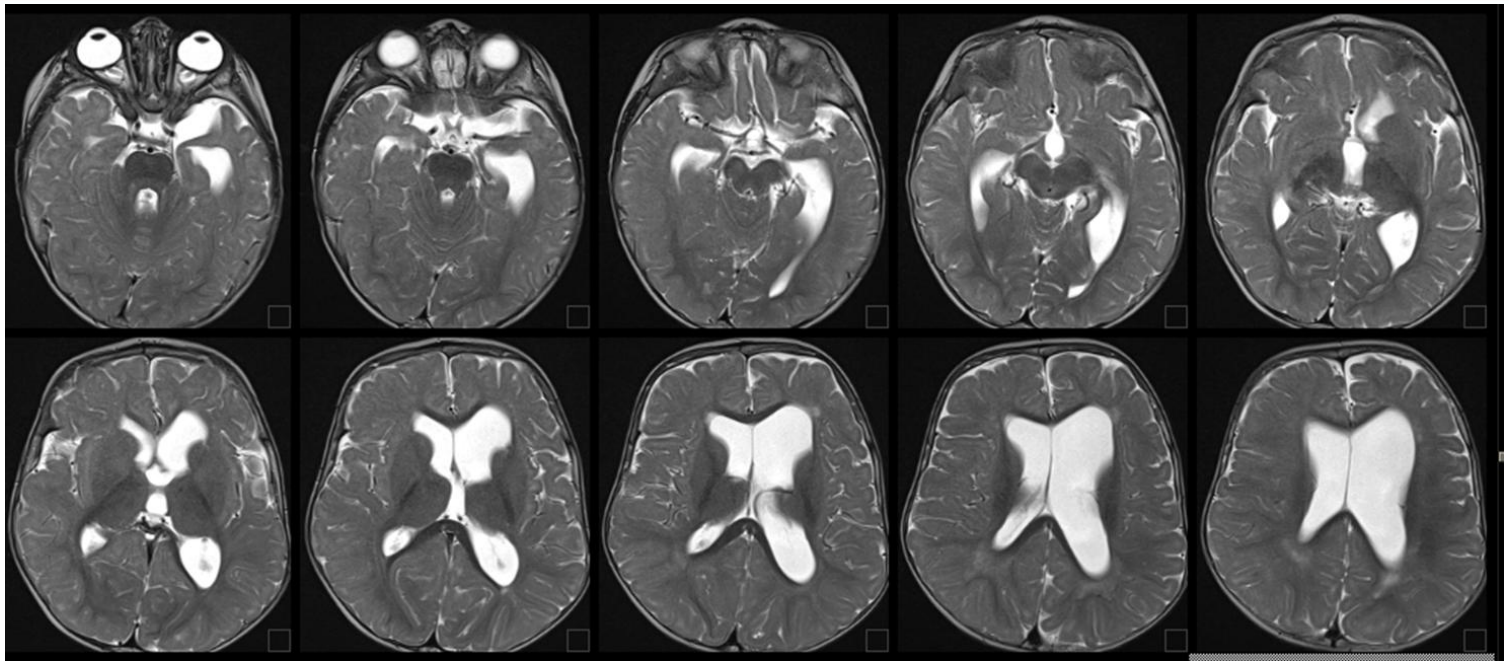




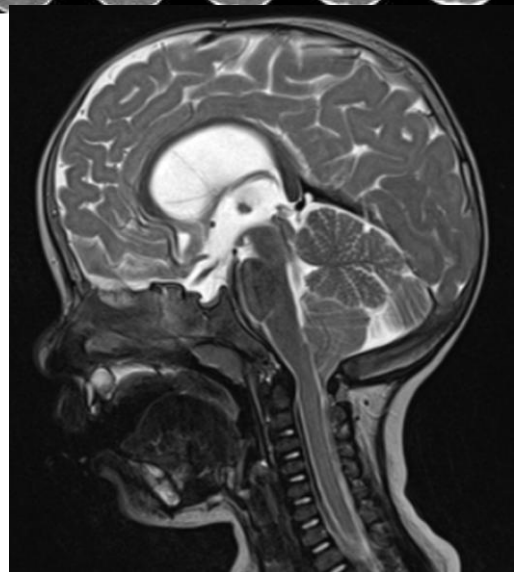
Y.M.: Hypophosphatemic Rickets, Rayne Syndrome

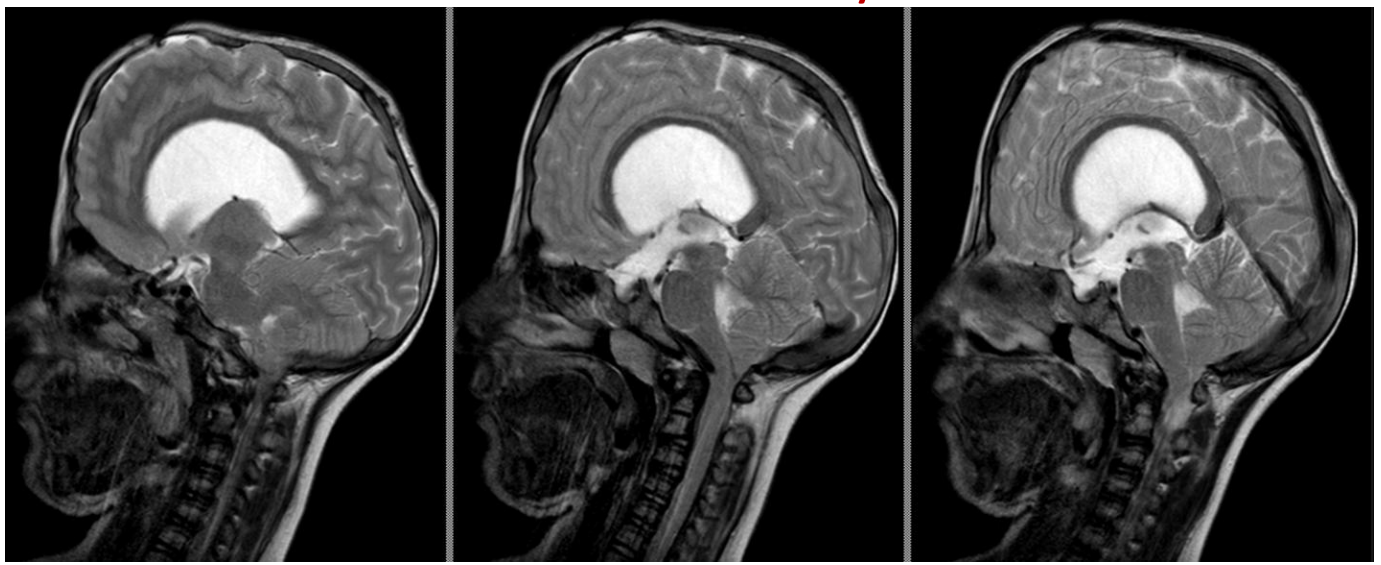






and his sister B.S. 13 yo





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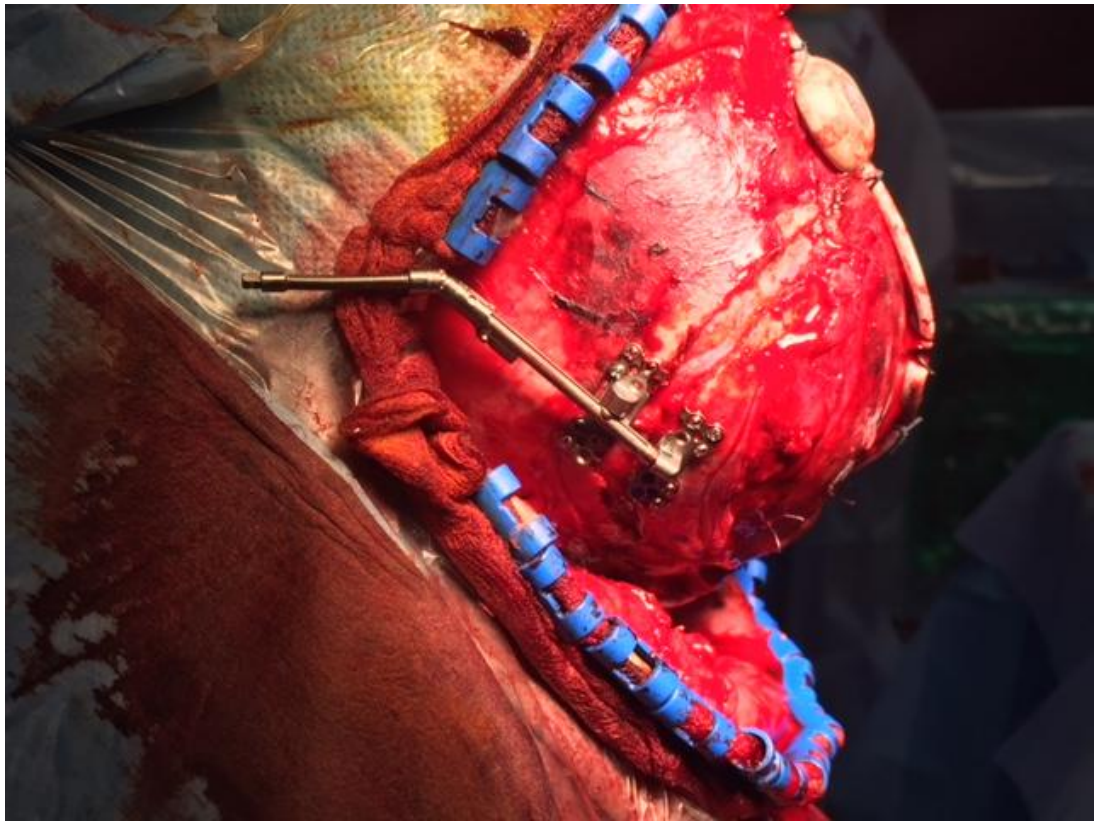
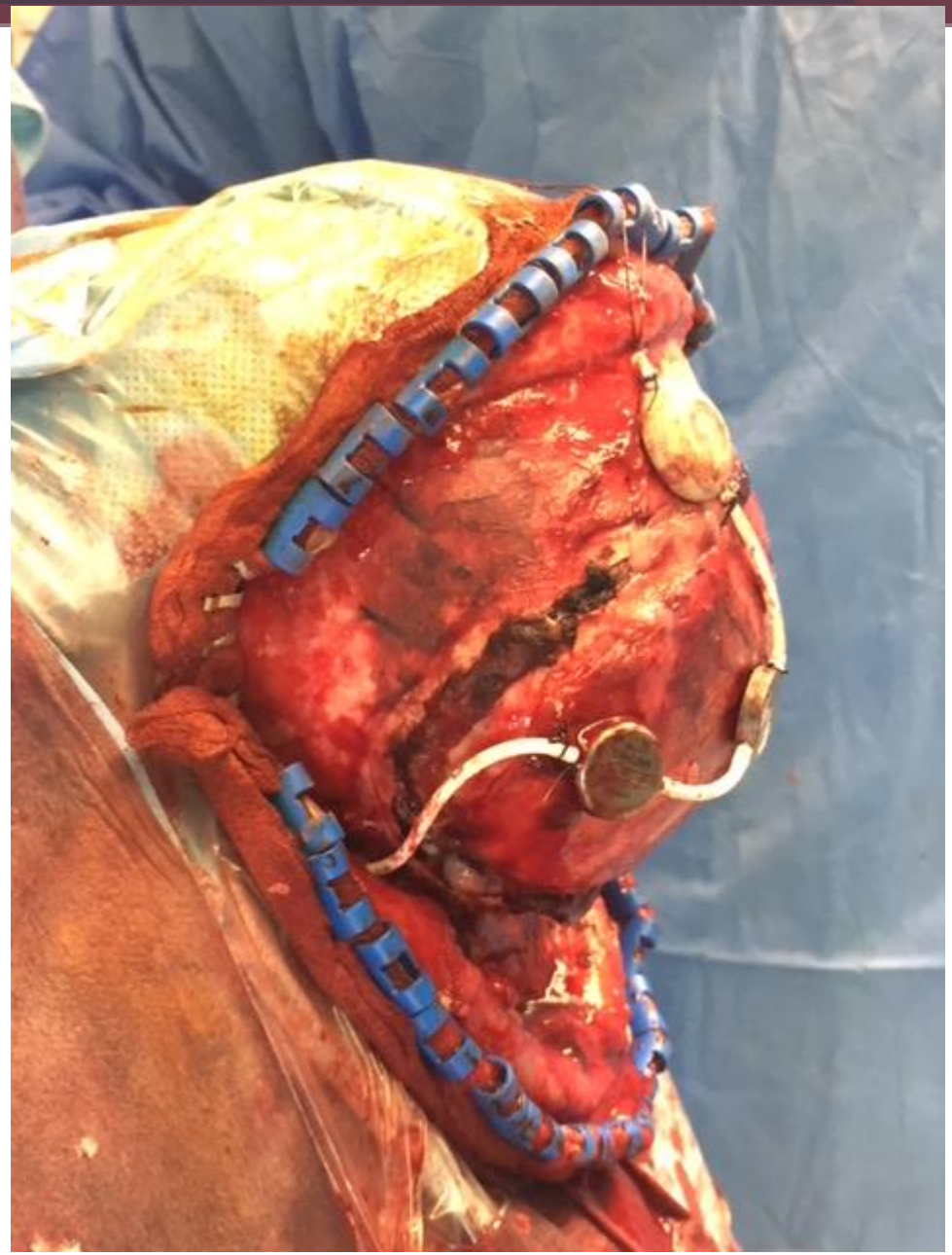
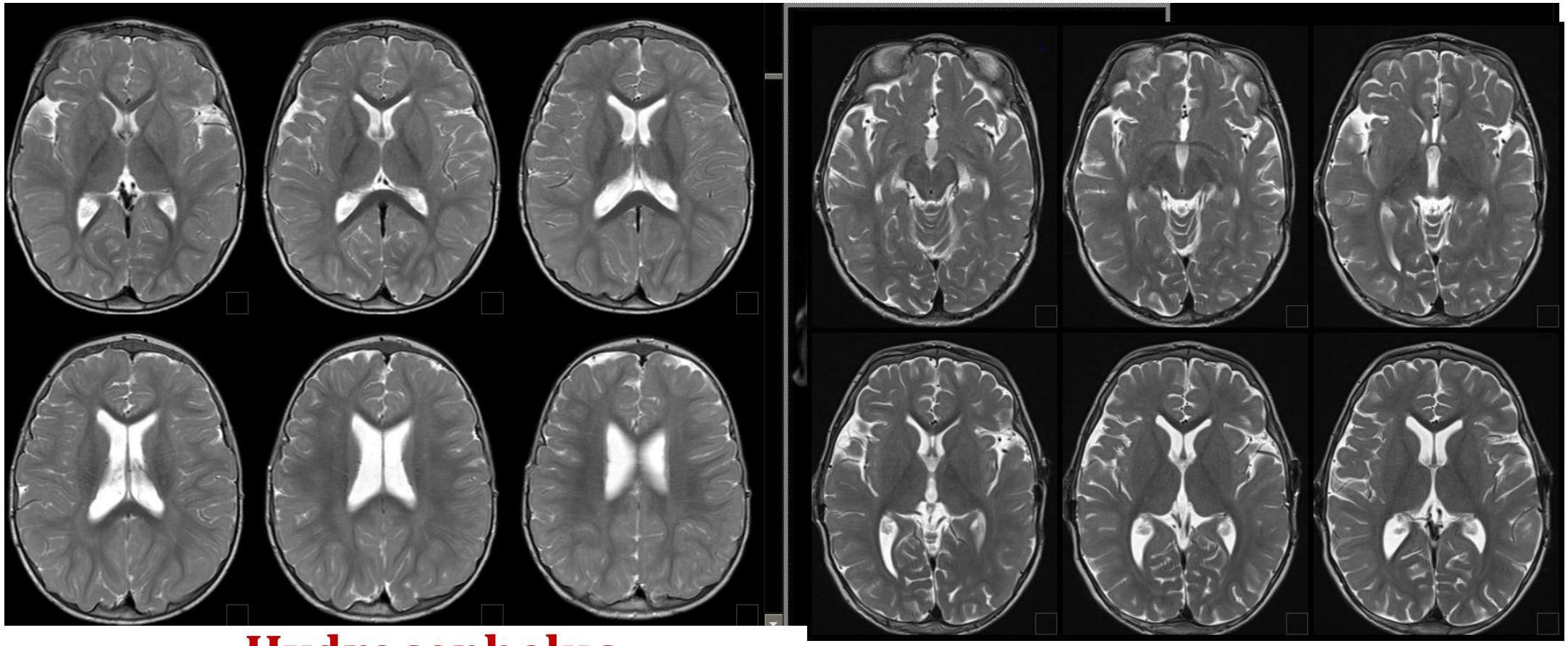
EUROPEAN CLEFT PALATE CRANIOFACIAL ASSOCIATION

POSTERIOR CALVARIAL AUGMENTATION:
How to deal with Hydro: Temporary Shunts





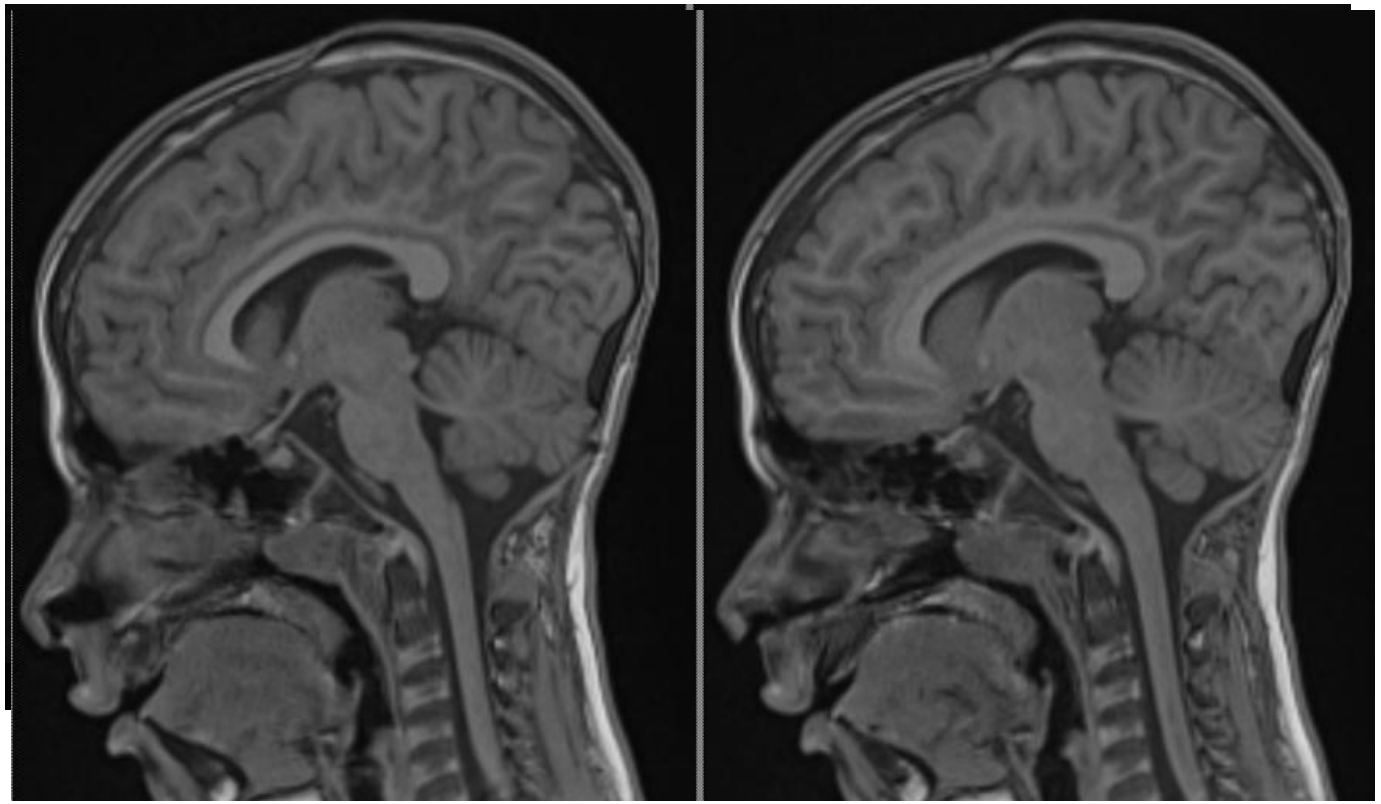
Posterior Calvarial Distraction:
Effects on Hydrocephalus 3/10 & ICP 10/10



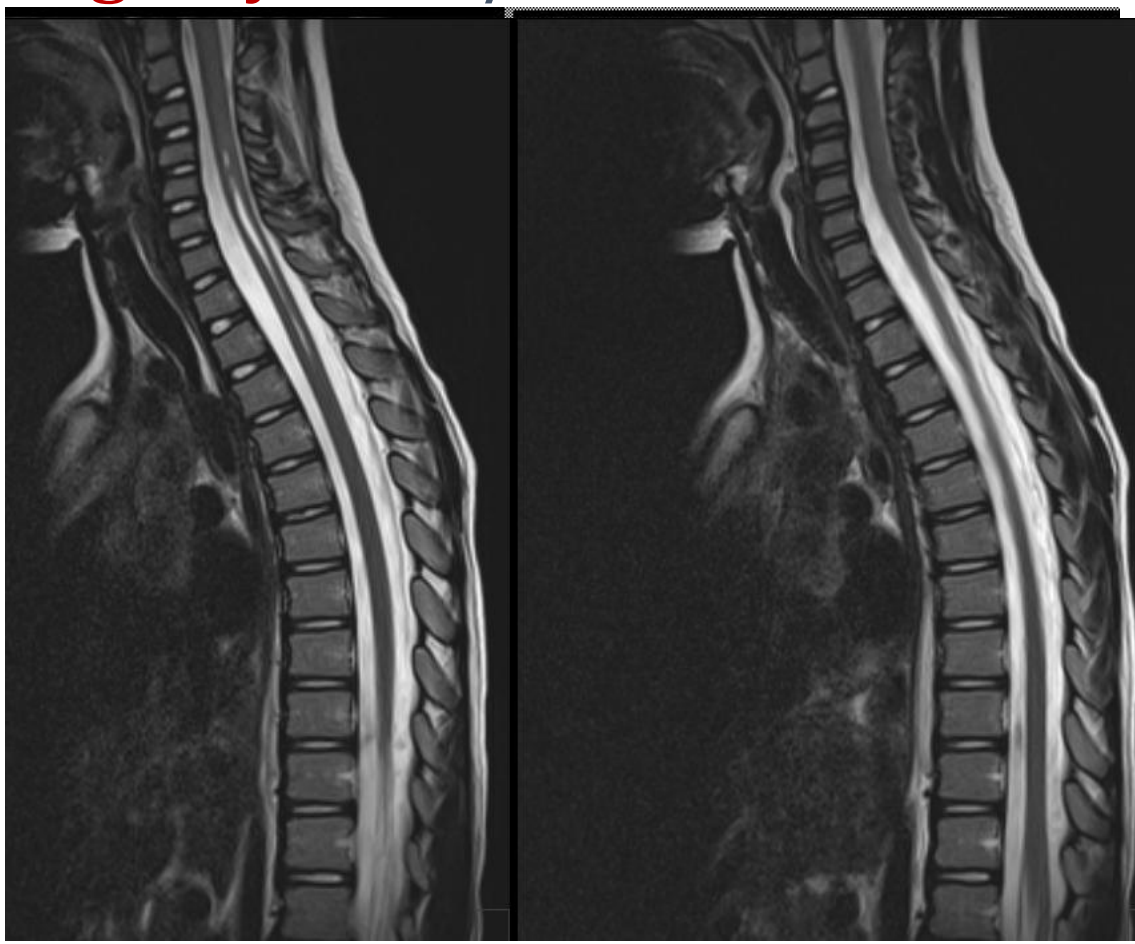
Hydrocephalus:
improvement **3/3**
Shunt closure **2/2**

ICP (Fundus):
improvement **10/10**

Posterior Calvarial Distraction:
Effects on CM1 8/10 & Syringomyelia 2/10

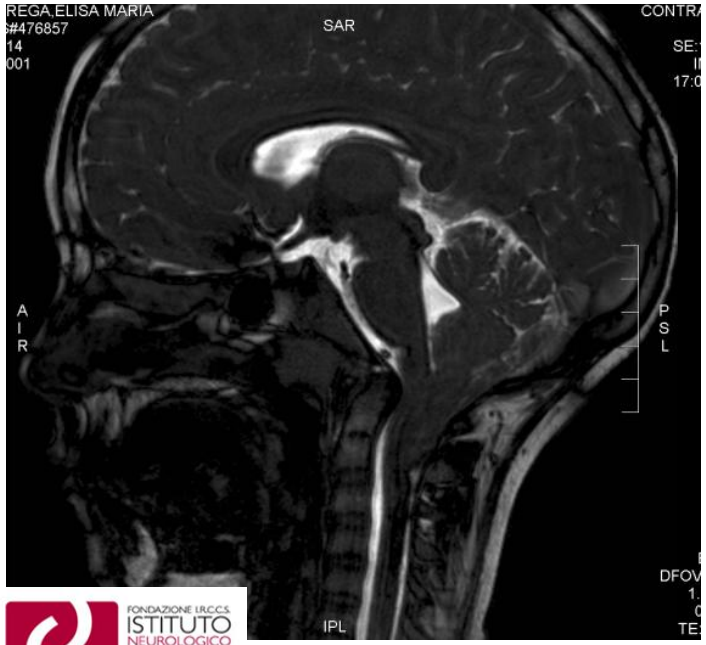


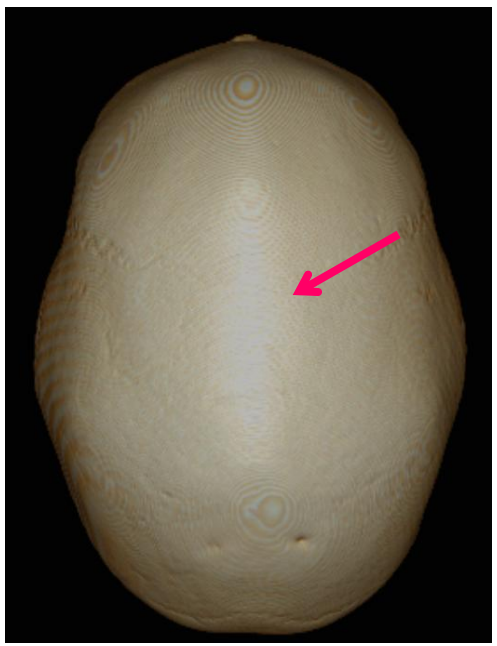
CM1:
improvement **8/8**

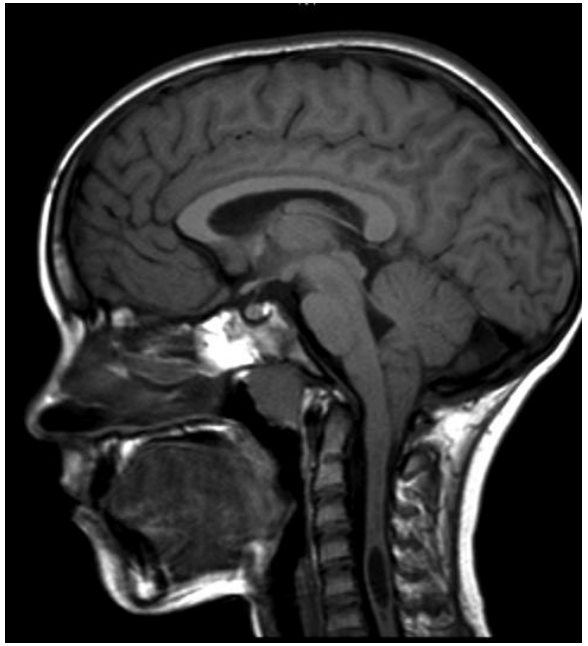


Syringomyelia:
shrinkage **2/2**

CM1 & UNTREATED SAGITTAL SYNOSTOSIS (USS)







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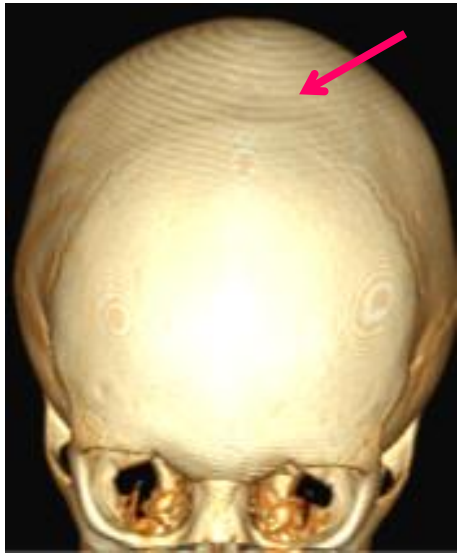
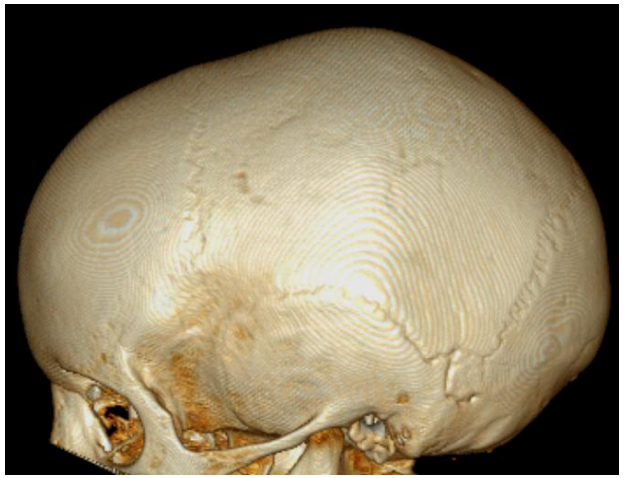
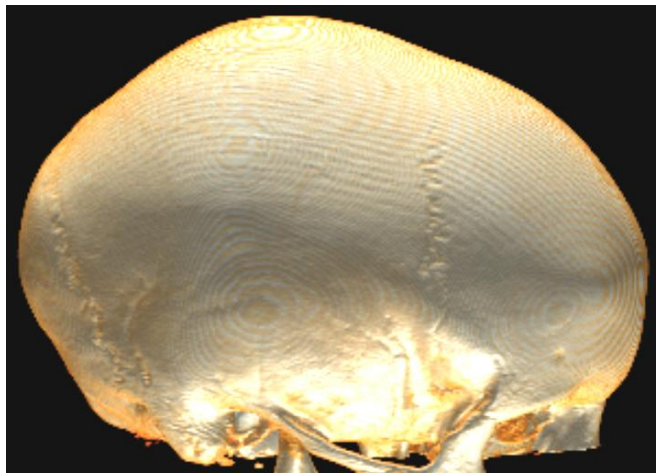
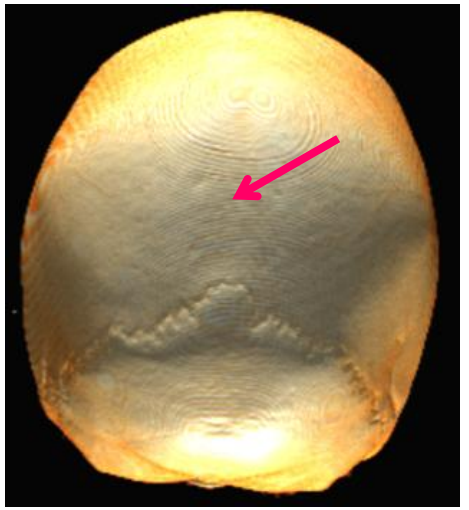
E & D, twins, 9 yo: USS → **Chiari Malformation & Syringomyelia**





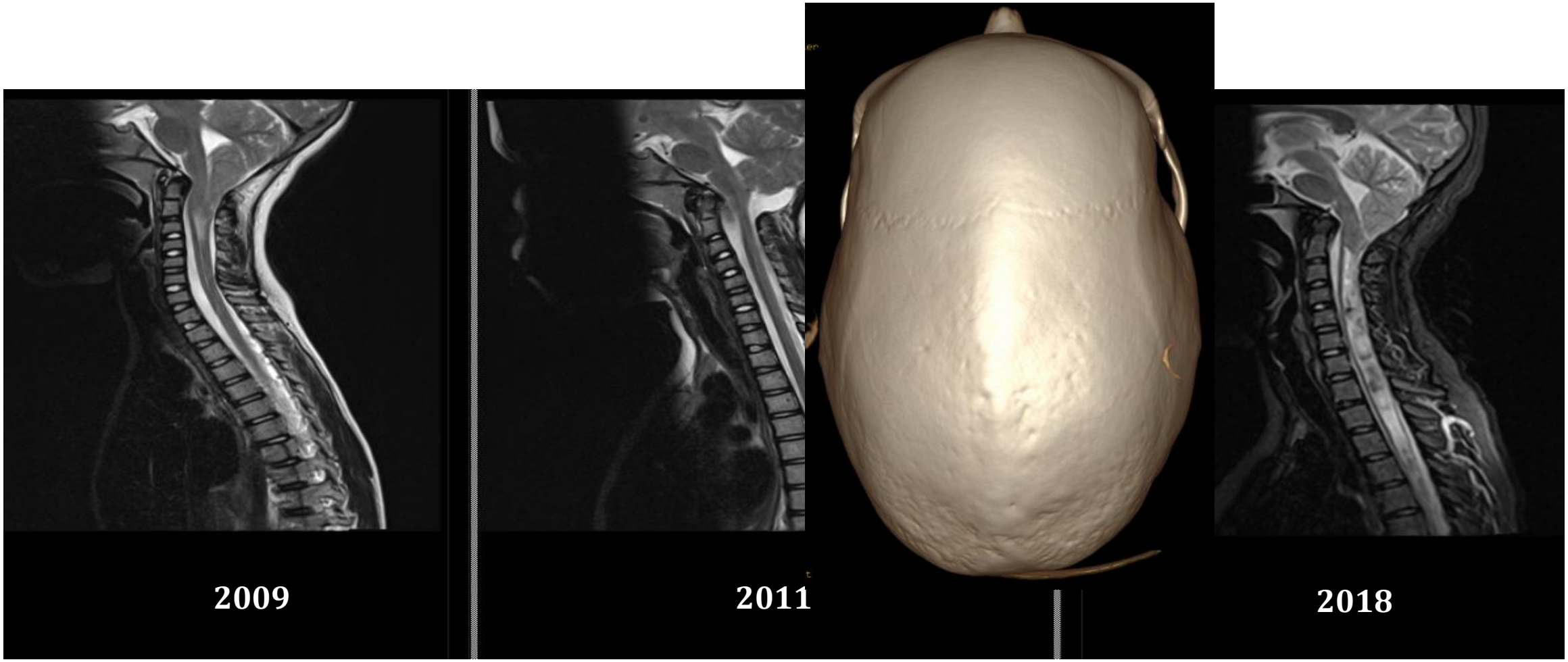


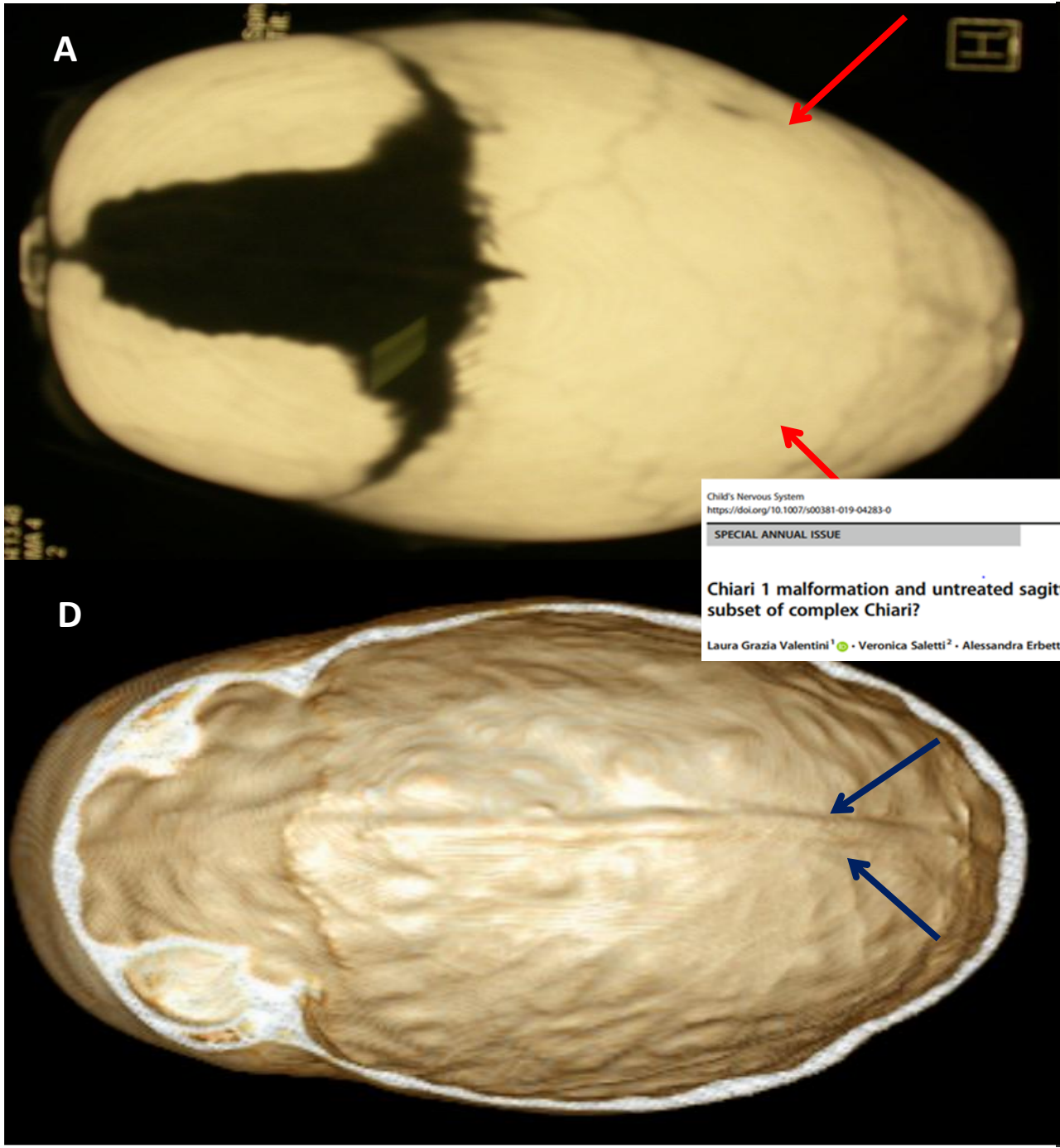


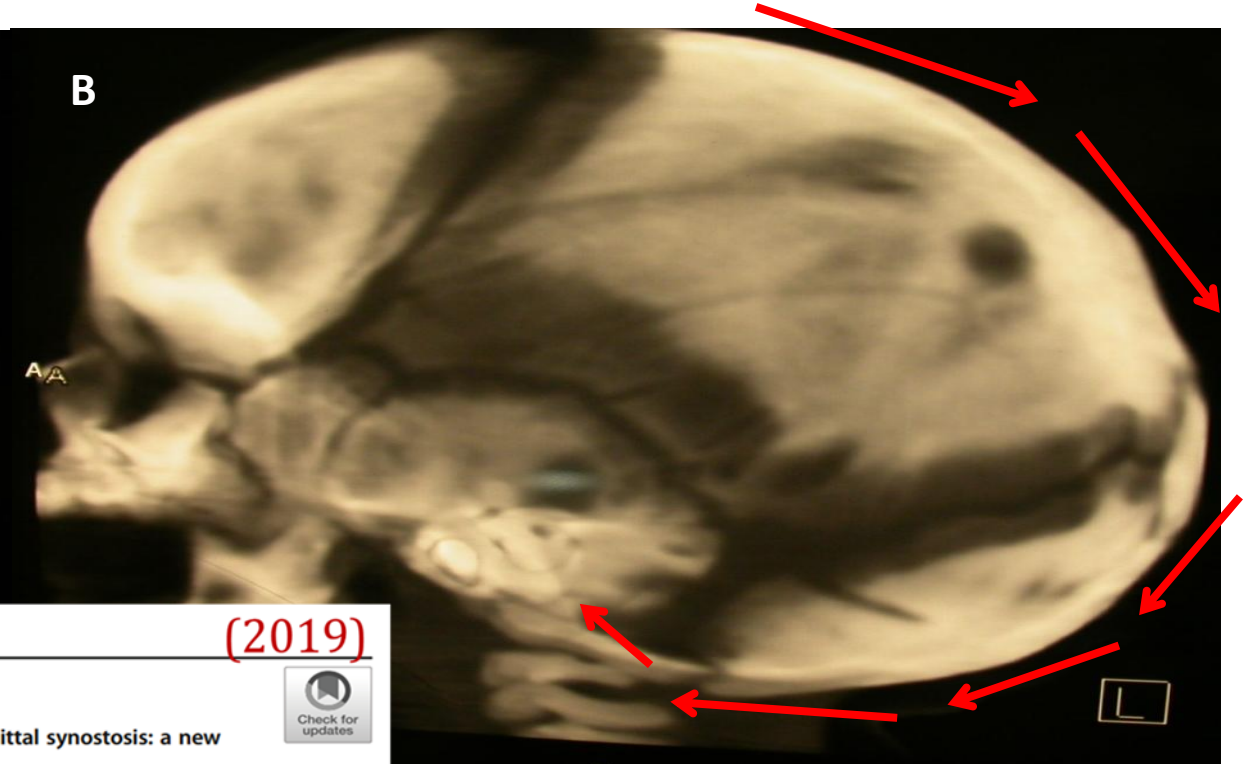
Pediatric CM1:

How long should we follow our children ?

P.C. delayed Recurrence of CM1 & Syringomyelia





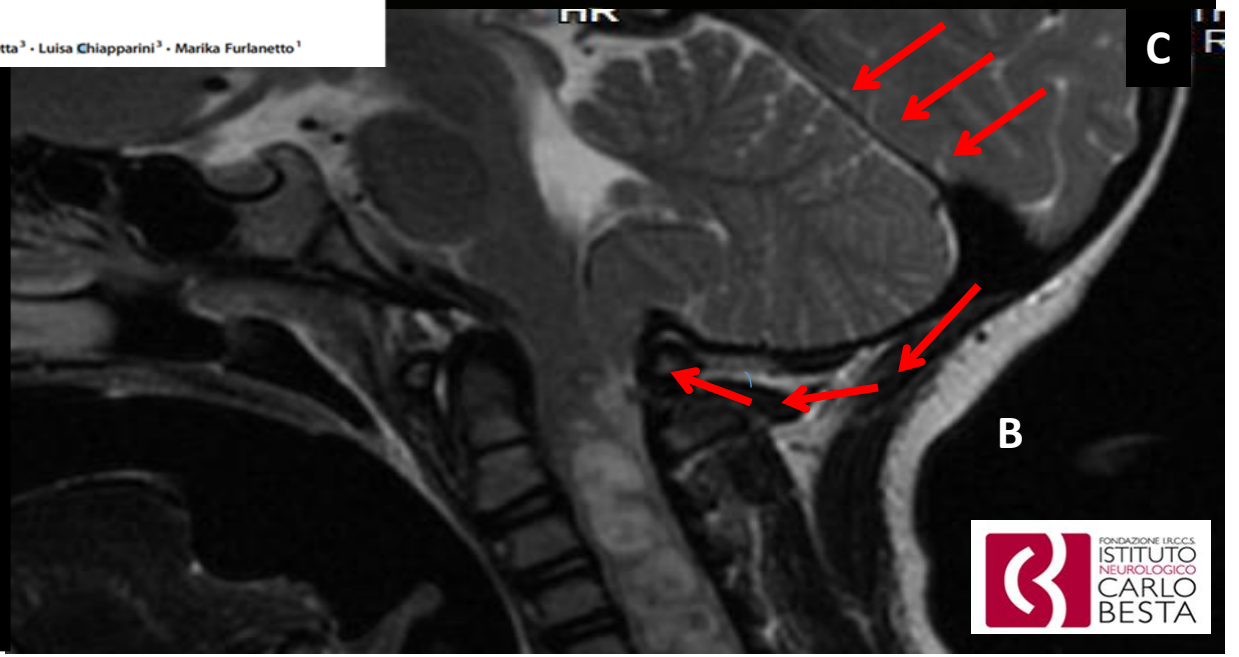


Child's Nervous System
https://doi.org/10.1007/s00381-019-04283-0
(2019)
SPECIAL ANNUAL ISSUE

Check for updates

Chiari 1 malformation and untreated sagittal synostosis: a new subset of complex Chiari?

Laura Grazia Valentini[1] · Veronica Saletti[2] · Alessandra Erbetta[3] · Luisa Chiapparini[3] · Marika Furlanetto[1]



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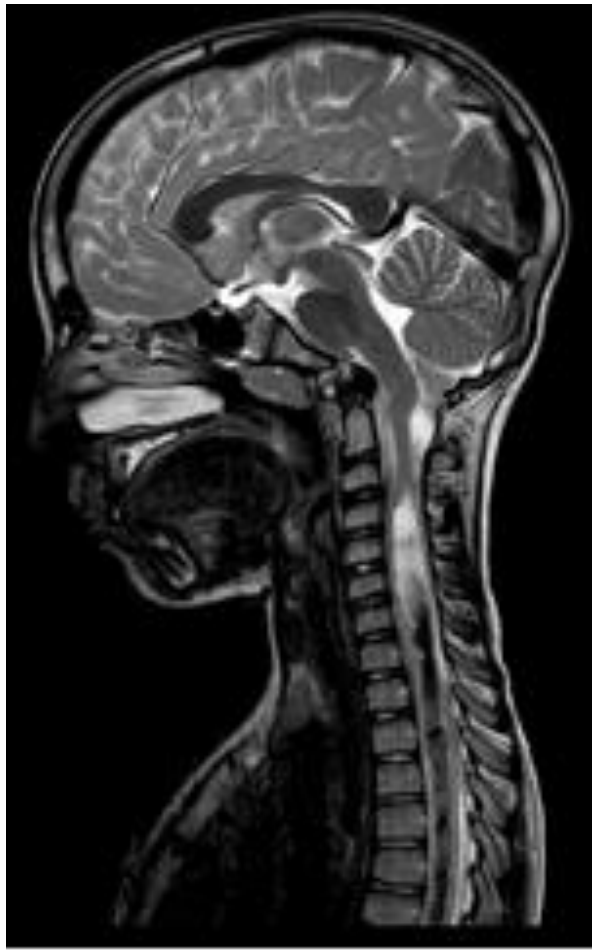
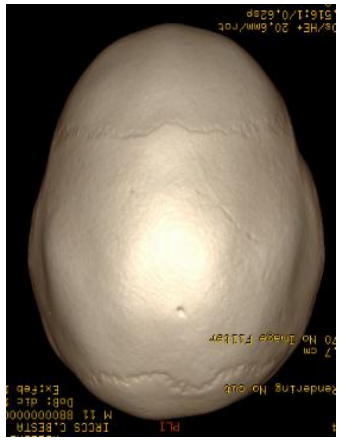
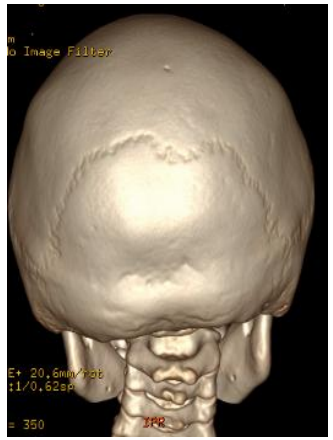
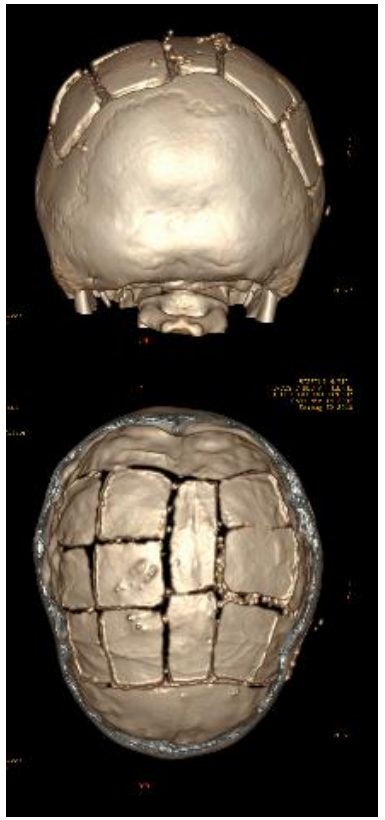
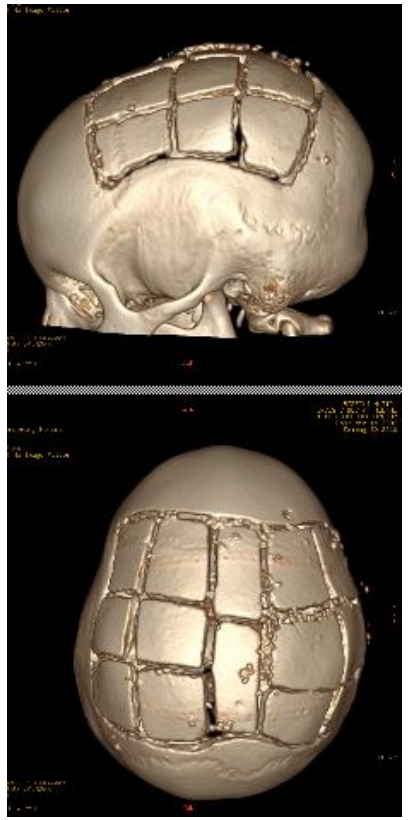
R.M. 11yrs, CM1 & Scaphocephaly

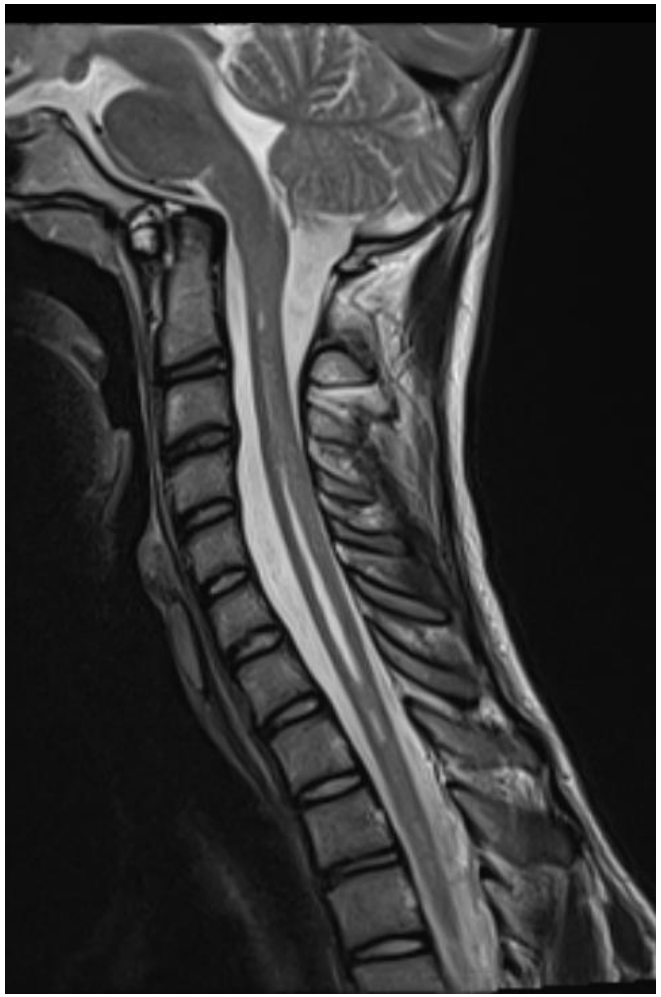












Just Free Flap Cranioplasty,
NO PFD

2 yrs after: Syringomyelia Shrinkage!

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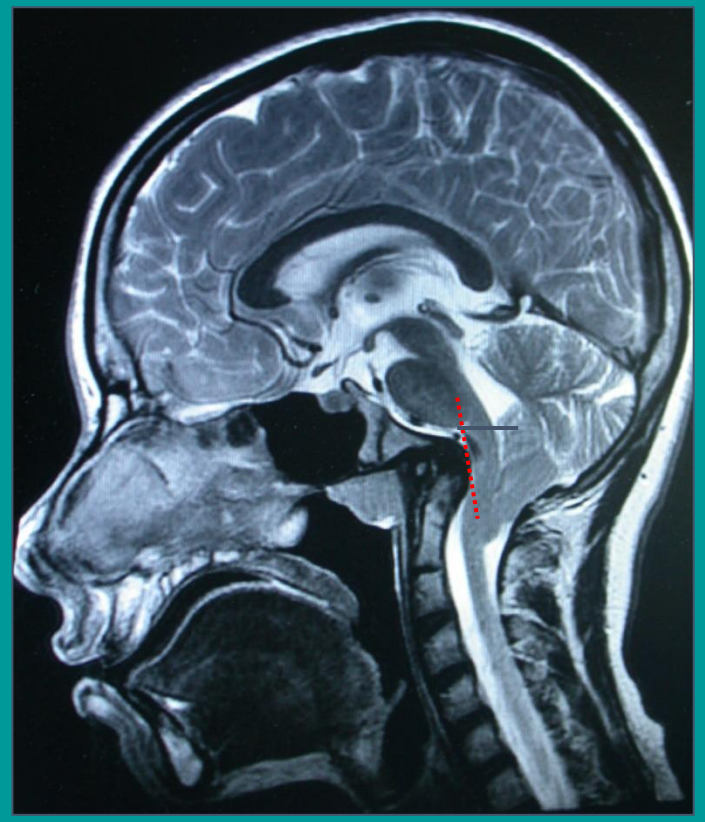
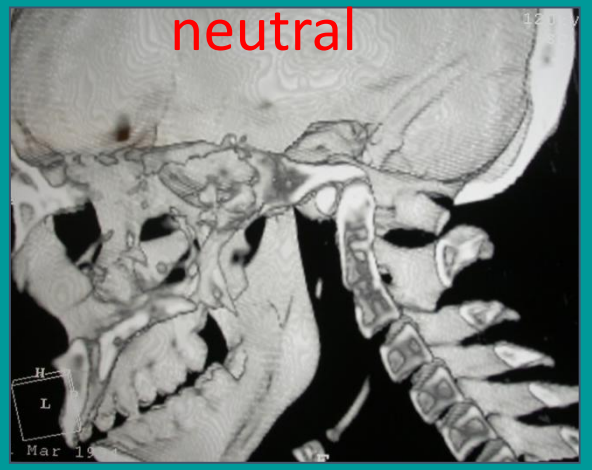
Clinical question 5	For CM1 children with Craniovertebral Junction Malformation (CVJM) is Craniovertebral Decompression more effective than other surgical approaches?	
1	**CM 1.5** may be associated with basilar invagination or impressio basilaris. Only cases with **related symptoms** need to be operated.	*Agreement: **95.2***
2	The preferred, etiological, surgical option for symptomatic basilar invagination associated with CM1 without atlanto-axial instability, could be anterior decompression, when posterior reduction has already failed.	*Agreement: **85.7***
3	The preferred surgical option for basilar invagination with atlanto-axial dislocation is posterior fixation.	*Agreement: **100***
4	Craniovertebral junction **(CVJ) instability** is a mobile dislocation between C0, C1, C2 (according to neuro-radiological exams) leading to neuro axial compression, neurological deficits, progressive deformity, or structural pain.	*Agreement: **85.7***
5	The standard diagnostic work-up for CVJ instability in CM should include (other than MRI) **dynamic** X-rays + dynamic CT scan with 3D reconstructions.	*Agreement: **81%***
6	CVJ fixation, with or without posterior decompression, is **not indicated** in CM1 patients **without** a documented CVJ instability.	*Agreement: **90.5***
7	**Posterior decompression and CVJ fixation** is the preferred surgical option for CM patient **with** CVJ instability and related symptoms.	*Agreement: **90.5***
8	In order to identify the best surgical option for CVJ instrumentation in a CM patient it is mandatory to identify: A) the vertebral artery course by preoperative neuroradiological studies (Angio-MRI, Angio-CT); B) the bone thickness of the occipital cresta, the C2 istmus diameter, the volume of C3 lateral masses.	*Agreement: **90.5***
9	To identify the best surgical option of C1–C2 instrumentation it is mandatory to define: A) the vertebral artery course by preoperative imaging (Angio-MRI, Angio-CT); B) the C2 istmus diameter.	*Agreement: **85.7***
10	Fixations by C0-C3 or C1-C2 in CM patient with CVJ instability should be decided on the basis of local anatomy.	*Agreement: **95.2***

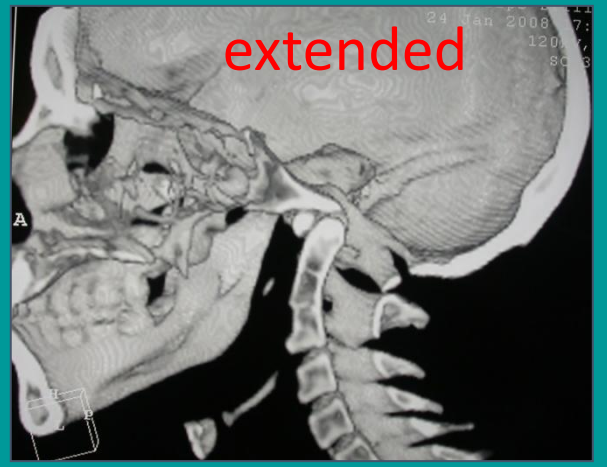
AISMAC
ASSOCIAZIONE ITALIANA SIRINGOMIELIA E ARNOLD CHIARI

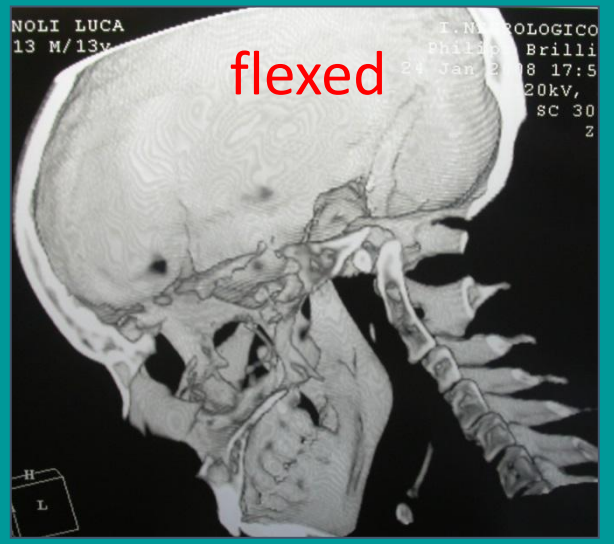
Pediatric plus CVJ Malformation
What is the **NeuroRadiologist** doing for CM1 pts ?



Grabb, '99: pB- C2 > 9mm







G. L., 13 years, Male:
basilar invagination and instability

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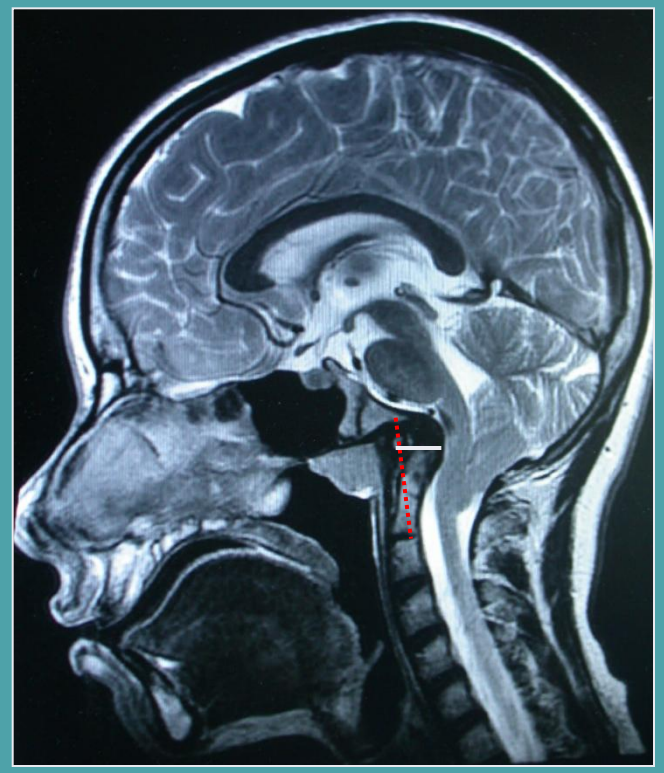
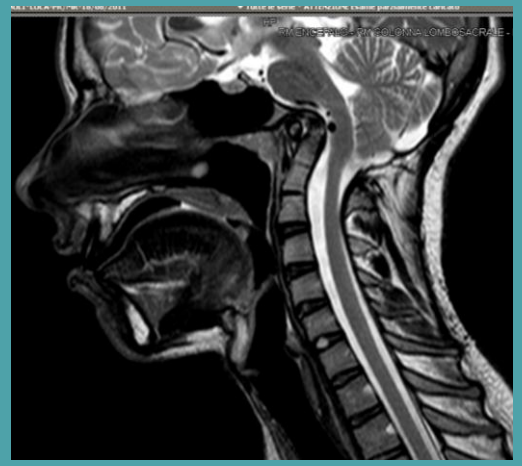
Pediatric CM1,5 :

Ventral Brain Stem Compression

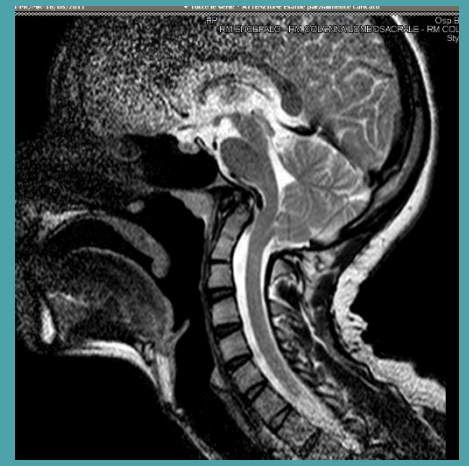
Grabb, '99: pB- C2 > 9mm



G. L., 13 years, Male: basilar invagination and instability



neutral



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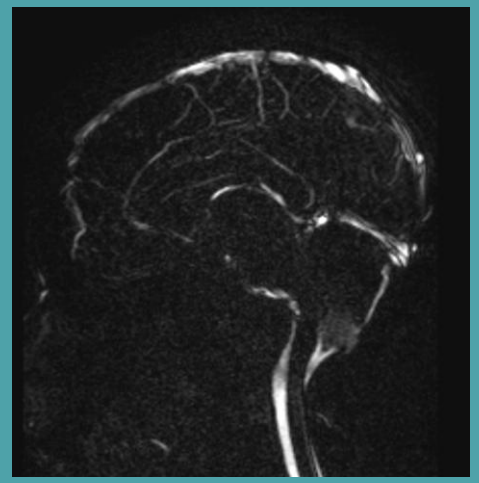

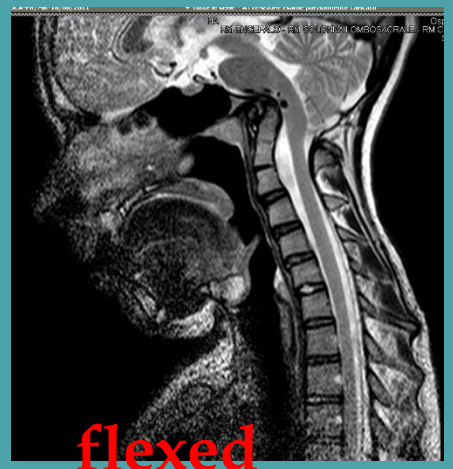

flexed

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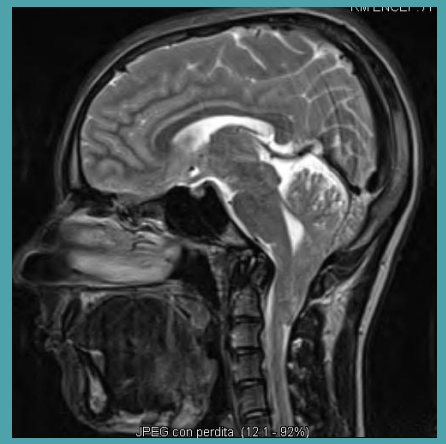
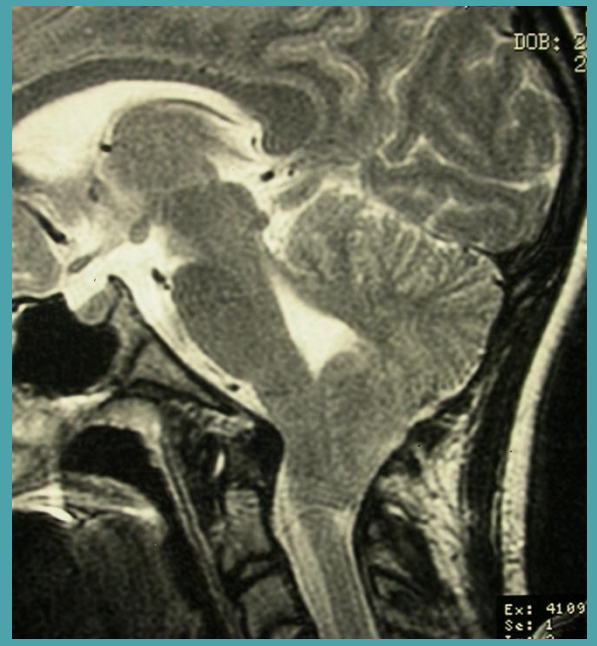
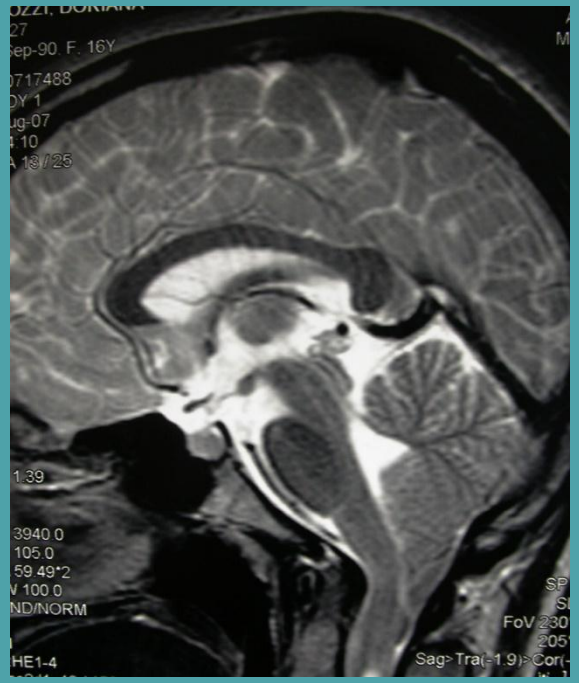
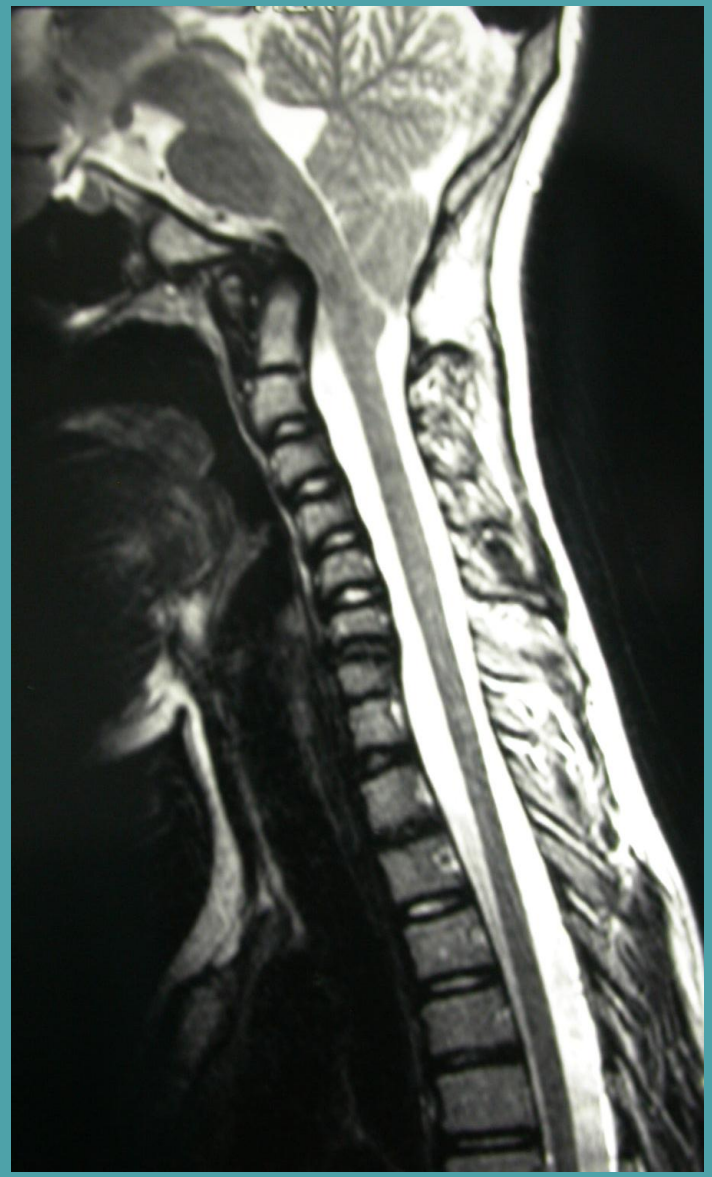
Pediatric CM 1,5:

73/174 (43%)





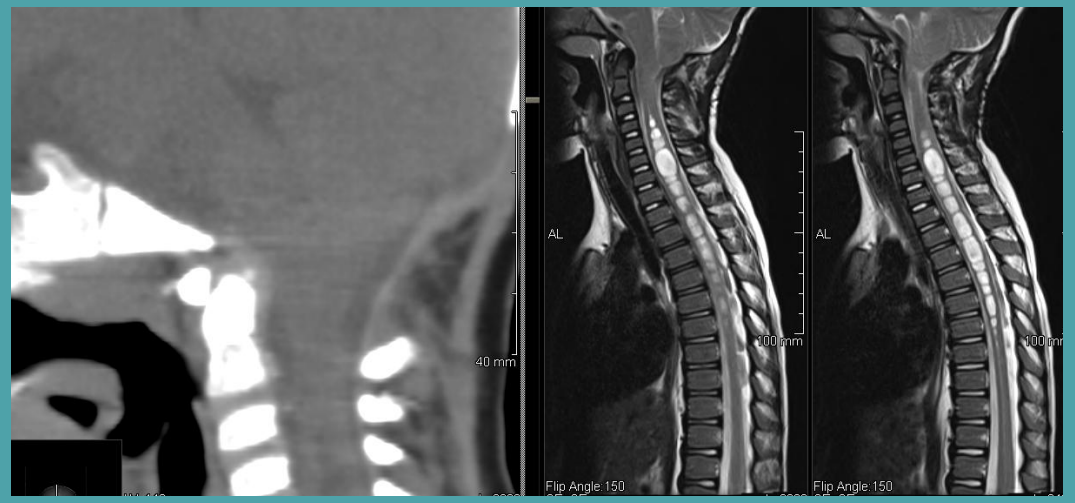




- **58%** various degrees of CVJ anomalies
- **None** had anterior decompression or fixation up to now
- **2/190** Operated by FMD needed CVJ Fixationation (1 postraumatic Instability, 1 Syndromic

Postoperative CV Instability or CVJ Hypermobility ?

flexded



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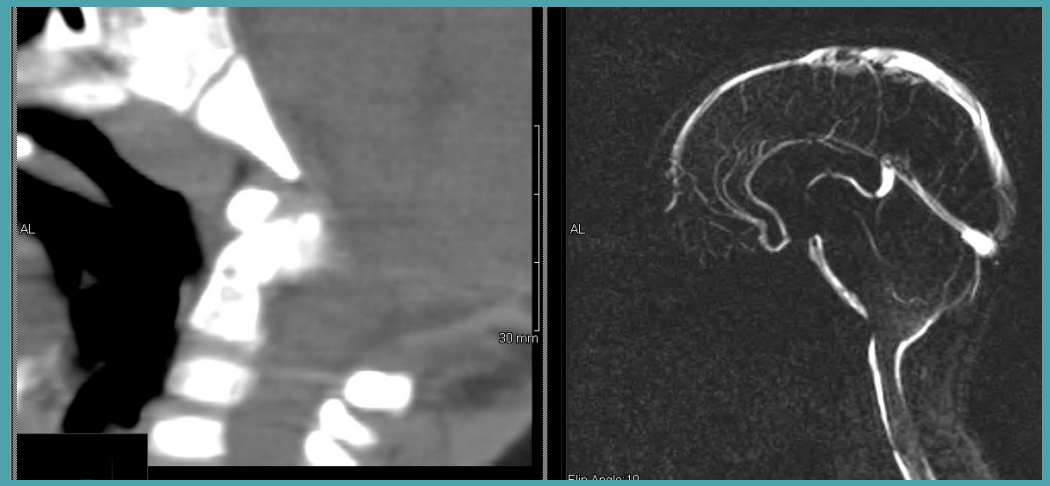

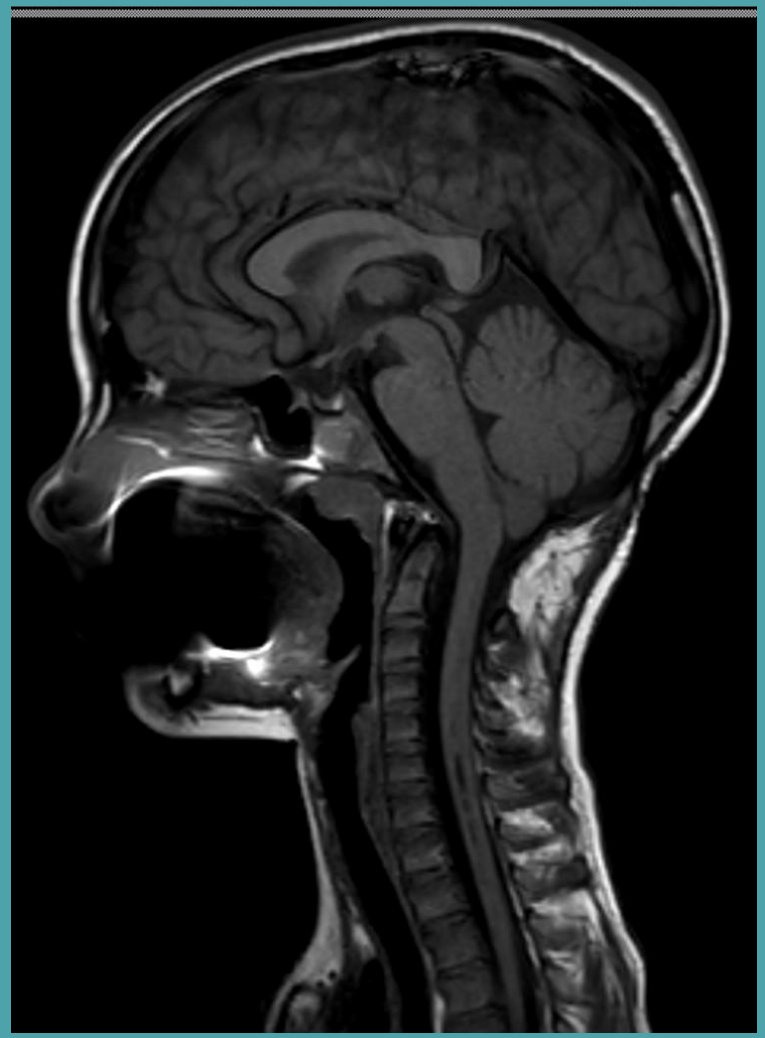

B.T, 4 y, Male: Postoperative CVJ Instability?

10 years later → No Fixation, No Symptoms


QUIET NAIL.
WAIT! HAMMER NO!

Chiari type I Malformation: Should We Operate Pictures or Children?

L.G.VALENTINI, A. ERBETTA T. GALBIATI I.G.VETRANO **V. SALETTI**

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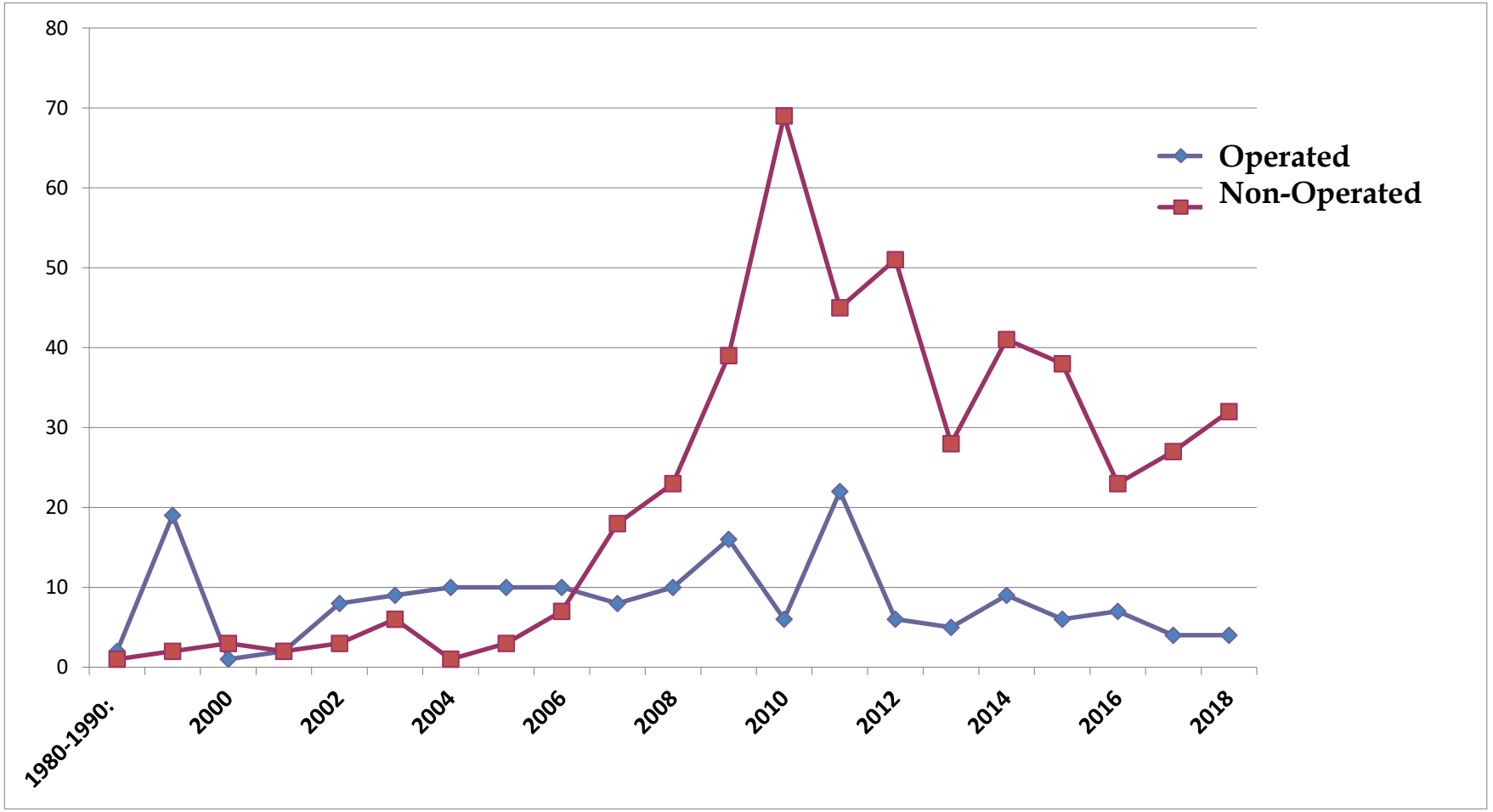
PEDIATRIC CM 1

Surgical Series 1994-2019



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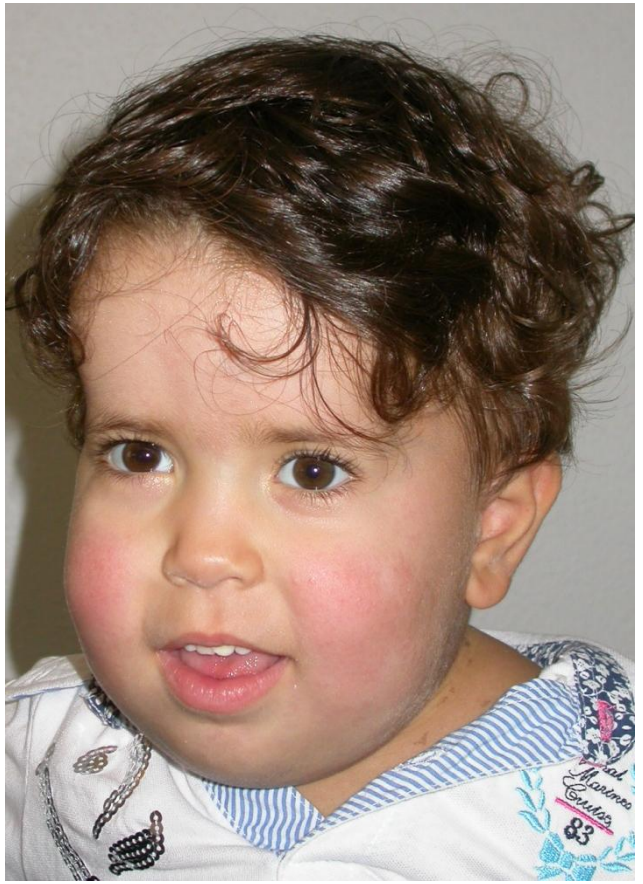
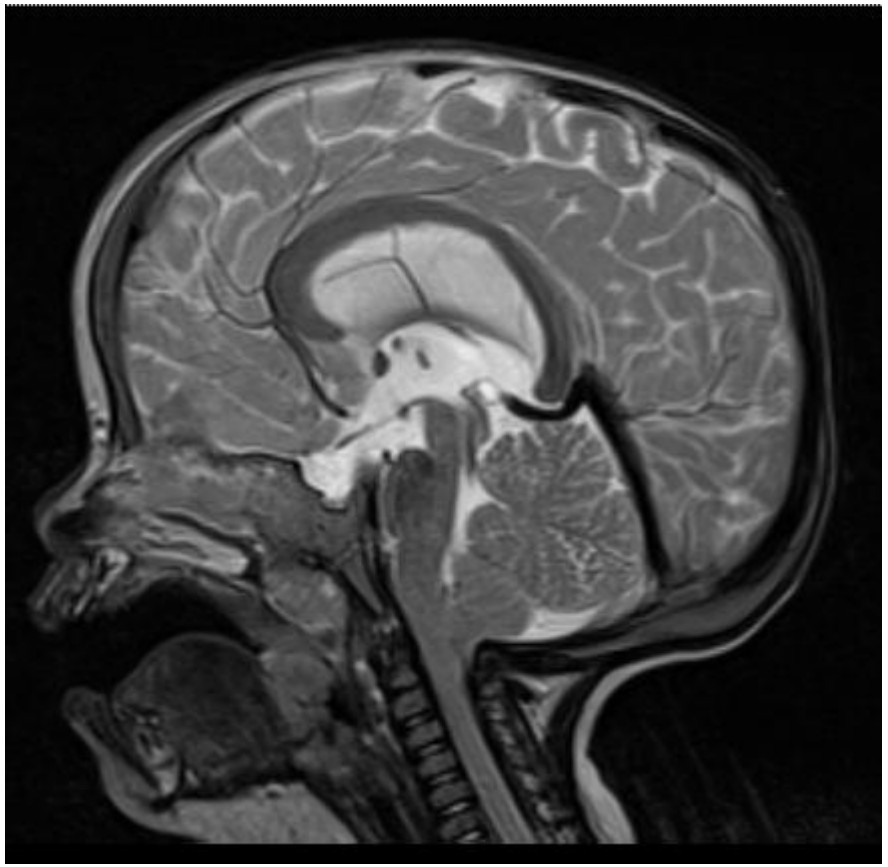
Pediatric CM1:

Cause leading to Diagnosis in 250 Children

Headache
Mental Retardation
Growing Delay GH
Precocious Puberty
Epilepsy
NF
Autism
Behaviour Disorder
Trauma

>30%
Associated Syndrome





CI, 5y,Cutis Marmorata

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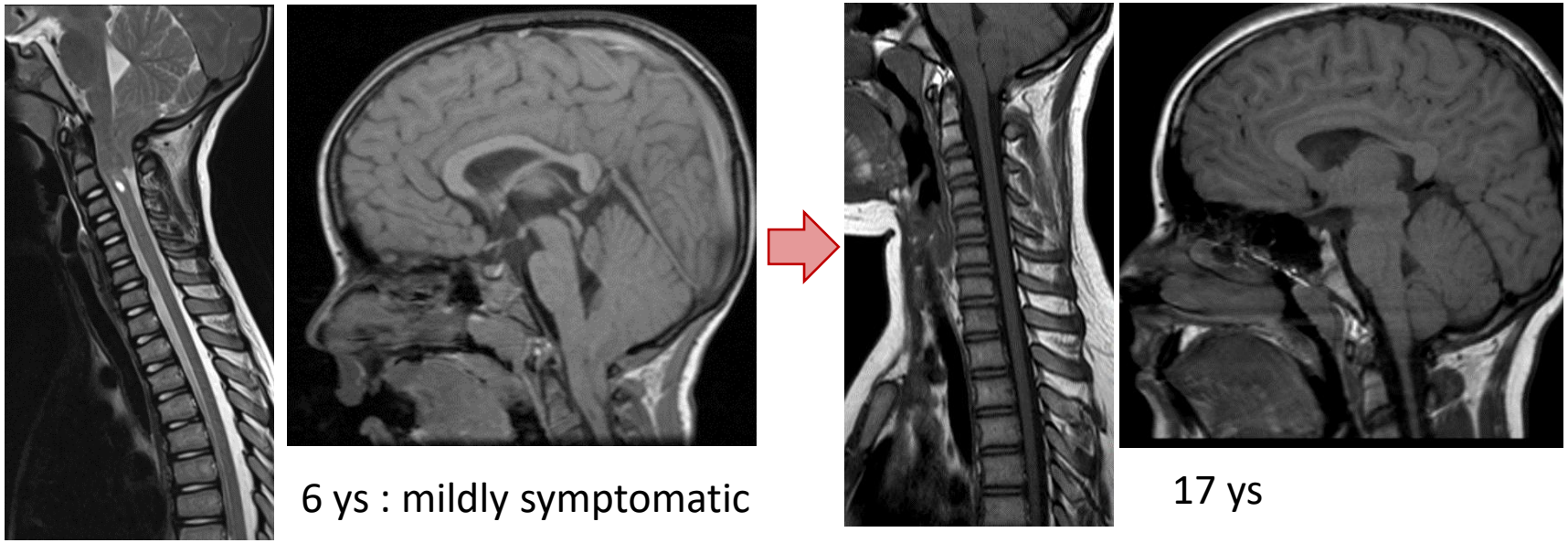
Should we operate MRI or Children ?

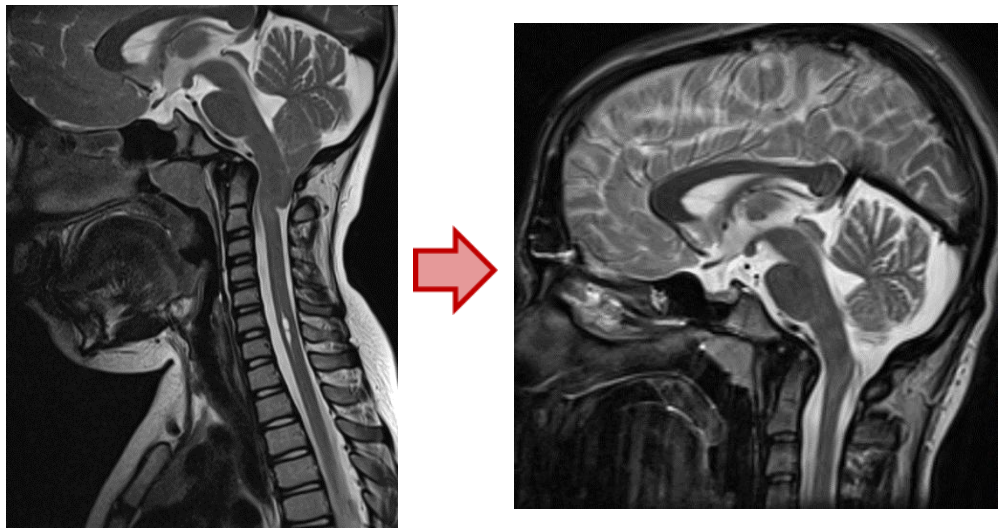
5,6%: Spontaneous resolution of CMI



6 ys : mildly symptomatic

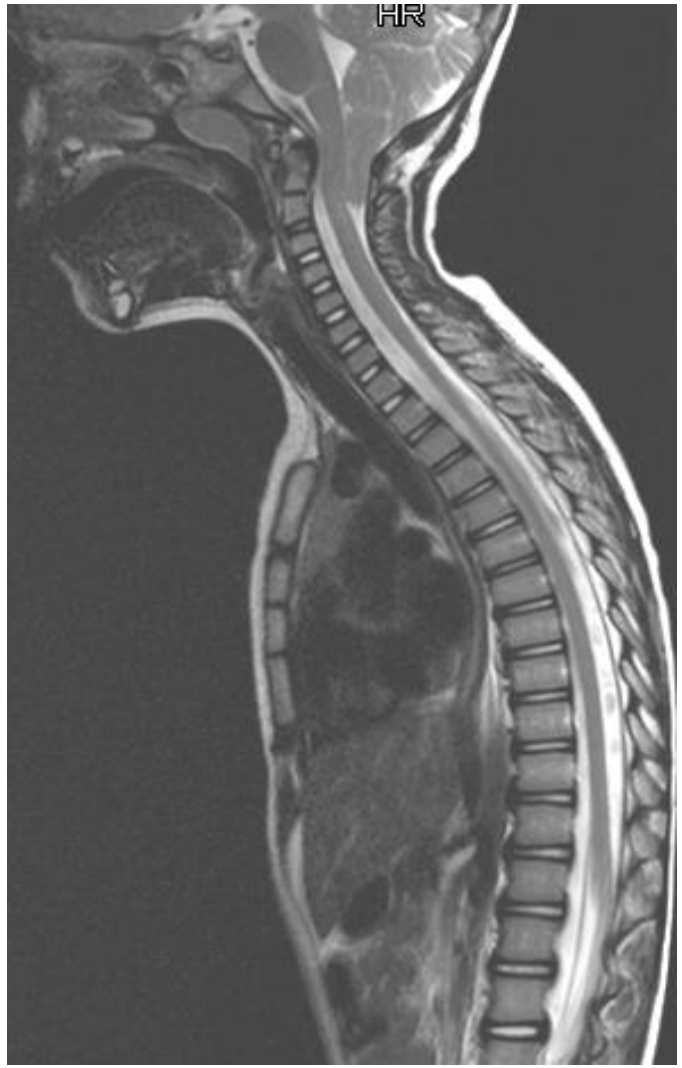
17 ys

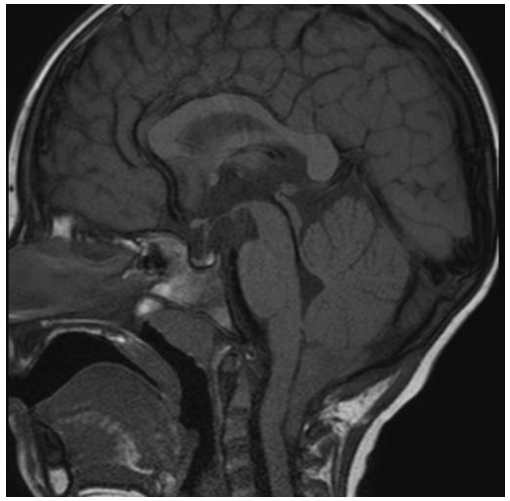
11 ys

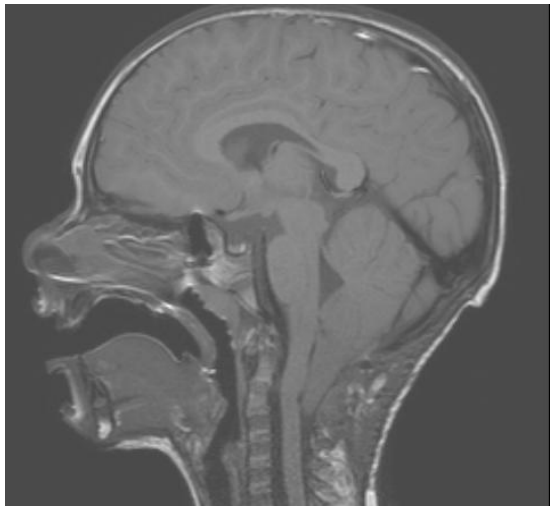


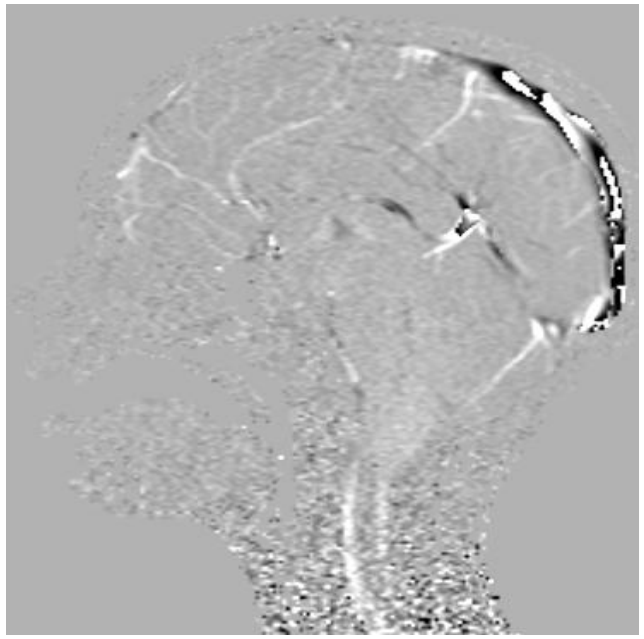
18 ys

Lack of Anatomical and Clininical Efficacy of Pure Bone Decompression









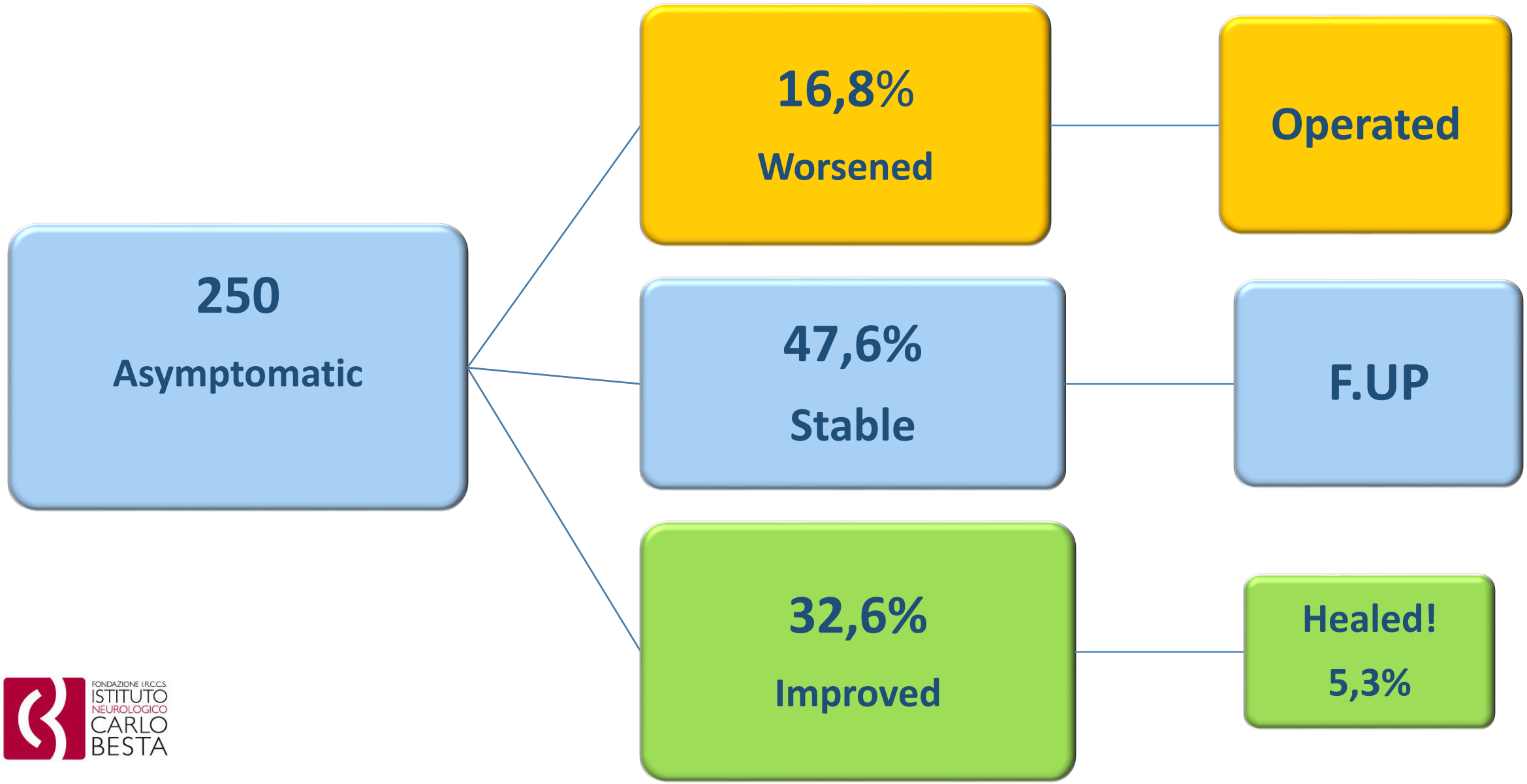
TA, 4y:

No syryx, no symptoms: **a typical case to be followed !**

After a PBD: the same tonsil descent and lack of PF flow

Just increasing complexity and risks of a CURATIVE surgery, when eventually needed

Pediatric CM1: Outcome



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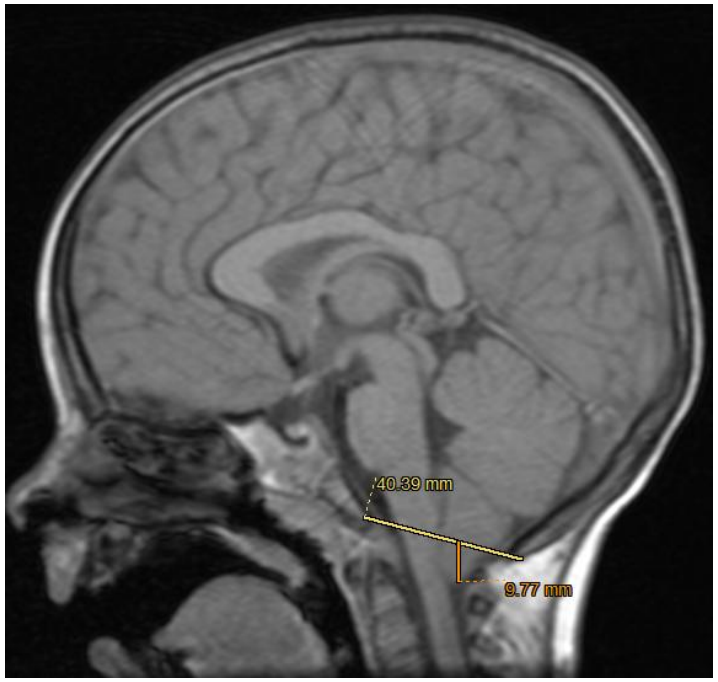
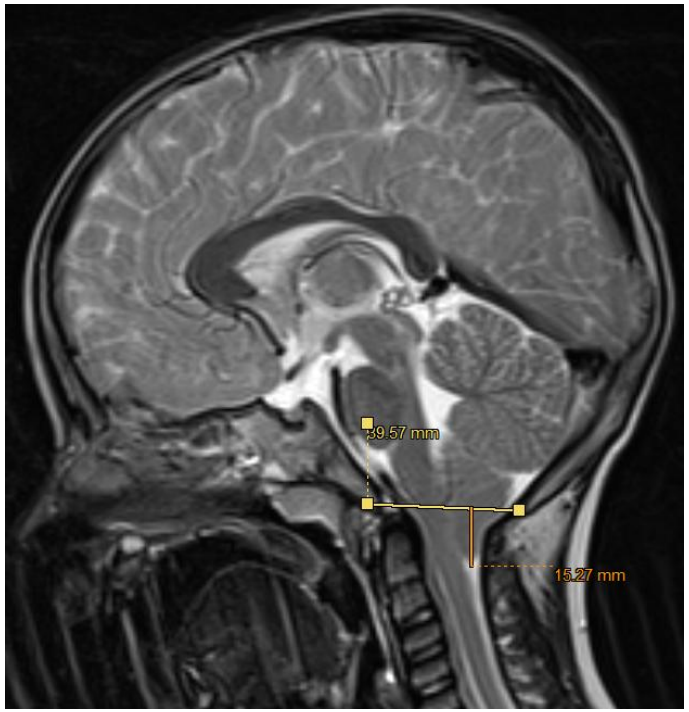
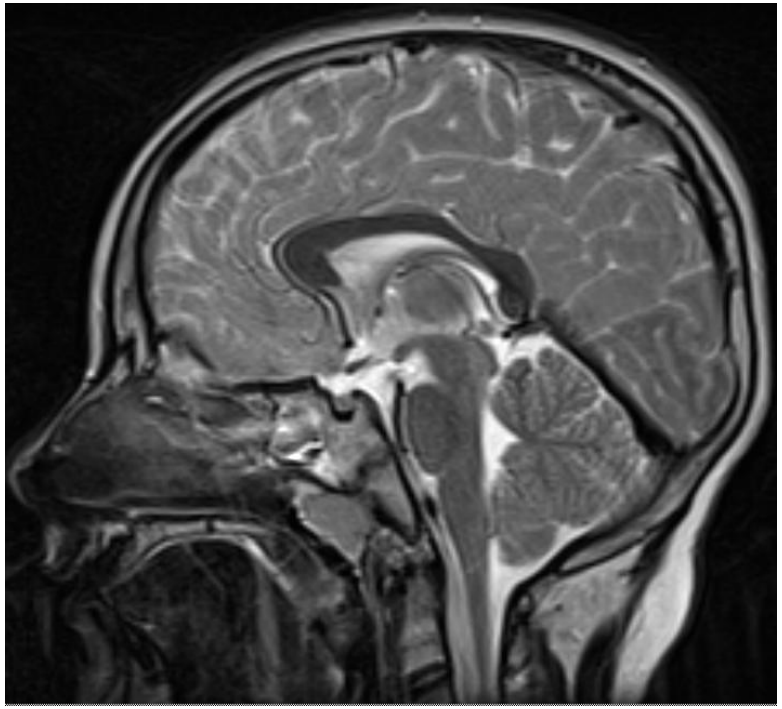
32,6 %: Spontaneous Healing of CMI



2 ys : mildly symptomatic



3 ys: asymptomatic



10 ys: asymptomatic

CM1: Influence of Risk Factors on Outcome

	Worsened	Stable	Improved
Global Series:	16,8%	47,6%	32,6%
Tonsils Herniation: Mild Moderate Severe	10,2% 20,4% 32,0%		29,2% 39,8% 12,0%
CVJ Malformation: 16,8%	26,2%	47,4%	26,4%
Hydrocephalus: 5,6%	35,7%	42,9%	21,4%
CM 1,5: 21,2%	34,0%	39,6%	26,4%
Untreated Craniosynostosis: 6%	53,3%	20,1%	26,6%

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Lesson Learned
Surgical Chiari

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-Higher Complexity due to associated Syndromes & other Malformations
-Chance of Spontaneous Resolution
→**No need for Prophylactic Surgery**
→ **don't scare families about Emergency**

-More prone to permanent psychological scar and CSF complications

-> Do less Surgeries!
-> But Effective
-> Evaluate and Follow CM1 in Pediatric Specialized Multidisciplinary Teams!

Pediatric CM1:
CONSENSUS CONFERENCE CONCLUSIONS

WHO should we operate?

- Just Children with **CM1** plus **CM1-Symptoms** and/or **Syringomyelia**

HOW to do the Clinical and Radiological Evaluation*?

- Pediatric Neurology evaluation
- Entire Nevraxis MRI +/- dynamic CVJ study (just in case of associated CVJM)
- ++Respiratory Studies, +-Neurophysiology

HOW and WHEN to do the clinico-diagnostic Follow-Up?

- All the Asymptomatics
- Yearly by the same clinico-diagnostic evaluation*
- Until the end of growth OR resolution of CM1 picture

Asymptomatic CM1: Take Home Messages

- CM1 behaved as a **dynamic condition**, more than a fixed malformation
- As we increase follow-up time, we appreciate better these changes:
    - 16,8% worsening, 32,6% improvement with 5,3% normalization
    - Just 47,6% remained stable
- The risk factors analysis on the present large series dislayed that:

  - **Entity of Tonsil Herniation** influenced evolution, with 10,2% **healing** in mild-moderate and 32% worsening in **severe** herniation
- Association with a Syndrome increased the rate of worsening and with:
  - **CVJ Malformation** is frequent 26% but true instability was quite rare (1/42)
  - **Brainstem Kinking CM1,5** doubled the chance of **worsening (34%)**
  - **Ventricular dilatation** increased the **worsening (35%)** chance as well
  - **Untreated Craniosynostosis** increased **worsening (53%)** chance three folds

→ Taylored Follow Up?

0% Acute Deterioration!

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the Chiari & Syringomyelia International Conferences
dealed by Associations

- 2006 Rugby, UK (ANN CONROY TRUST)
- 2007 Paris, FR (APAISER)
- 2009 Consensus Conference Milan, IT (AISMAC)
- 2010 Berlin, DEU
- 2013 Syndey, AUS
- 2017 New York, USA (ASAP)
- 2018 Birmingham, UK (ANN CONROY TRUST)
- 2019 Niagara Falls, USA (Columns of Hope)

Monothematic Meetings gathering Experts

coming from international Centers with high volumes of cases

Aim of the Chiari & Syringomyelia Conferences

Learning from each others

International Congresses organized by the Chiari Associations,

Where Experts presented their experiences that work:

- Sharing Tips & Trics

And that didn’t work:

- Learning from **mistakes**
- In Literature the reports of failures are rare (eg: plugging obex)

A sage learns from the mistakes of others, a fool from his own (Plauto)

→ Surgical mistakes creates so much suffering for patients and families

→ they should become a resource for the entire Medical community