

Syringomyelia and Chiari malformation : advances in surgery

Meeting in Turin in CTO / CRESSC, 9th of June 2016

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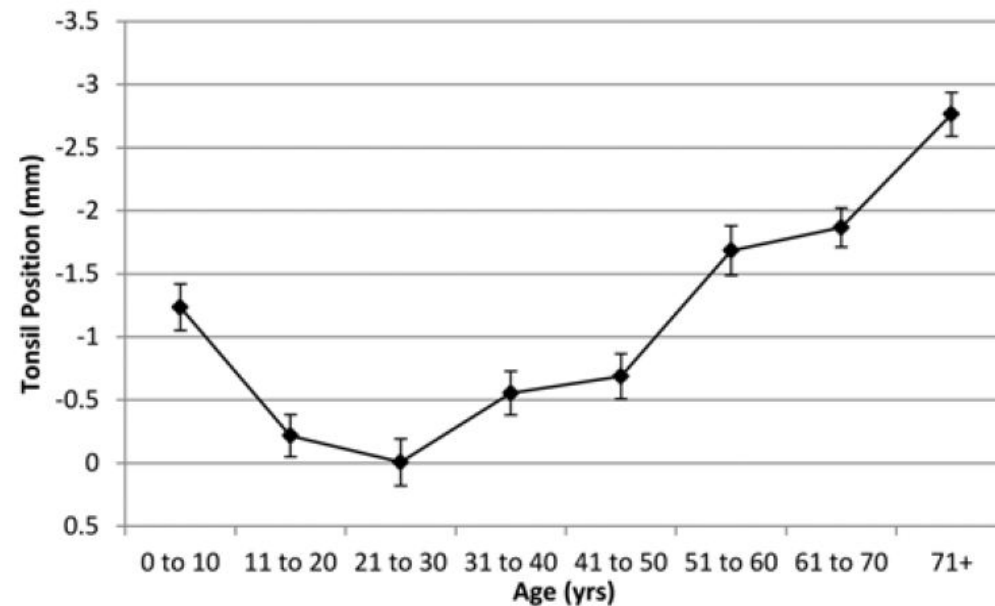
I - Better understanding of cranicervical junction malformation 's epidemiology : Tonsils 'morphology in the general population



Pegged tonsils (amygdales pointues)

Population générale : 1%

Probabilité CM > 5mm : 85%

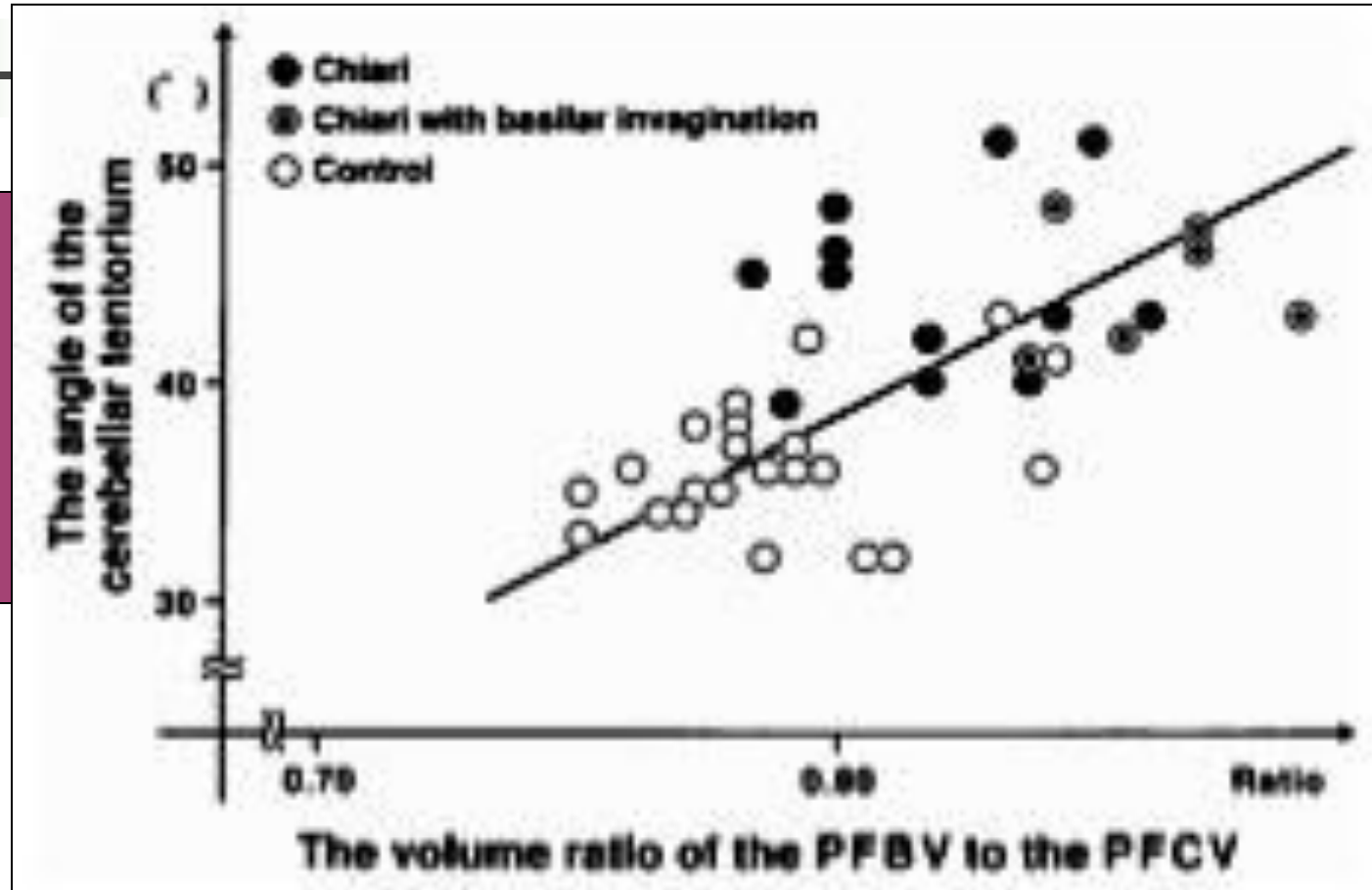


Position moyenne la plus basse selon la décade
N = 2378 personnes

30% of CM 5 to 10 mm are asymptatique

The craniocerebral disproportion of the FP

Mesodermic origine: occipital hypoplasia without Neurectodermic abnormalities



Opasie Occipitale

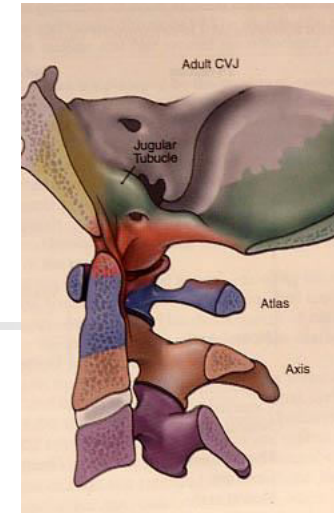
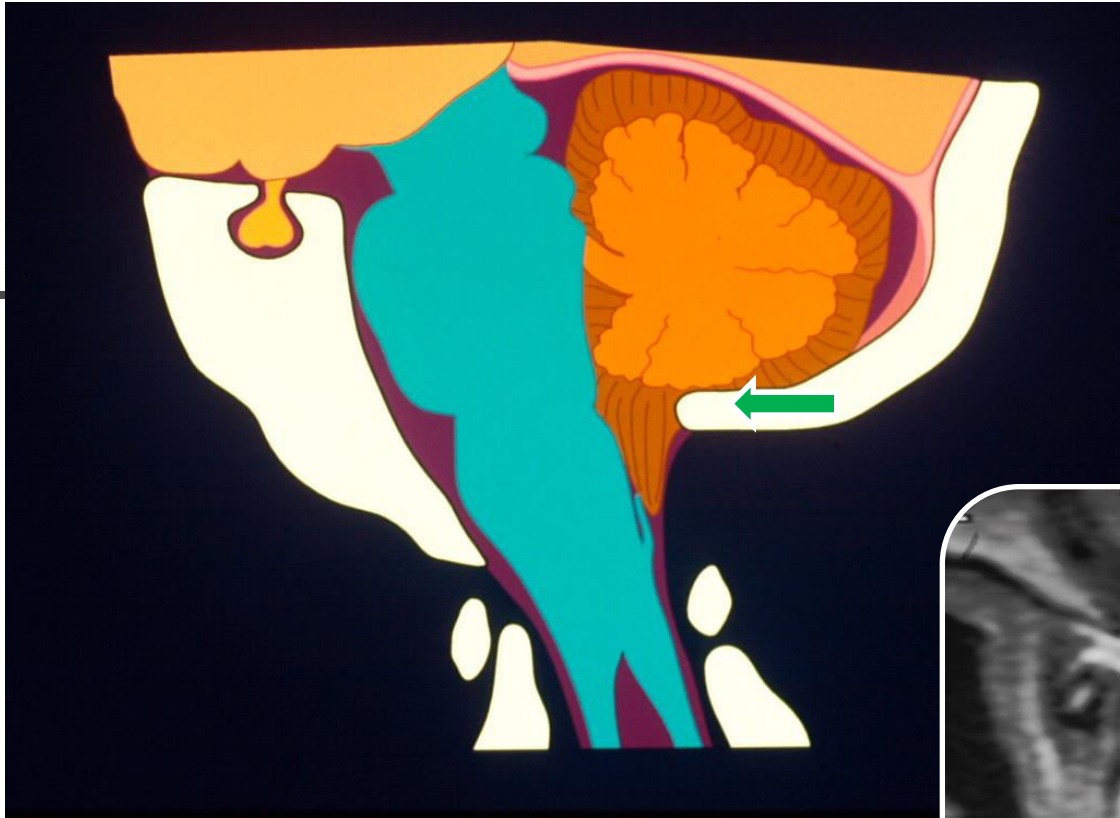
ou Clivus Court

teur FP



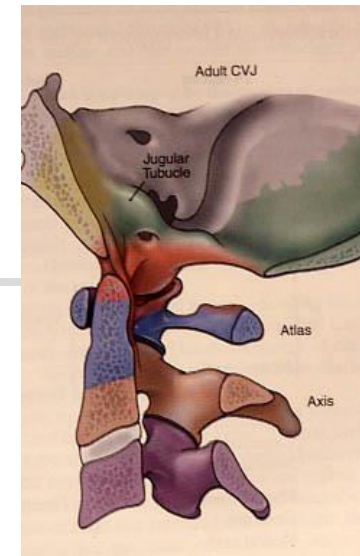
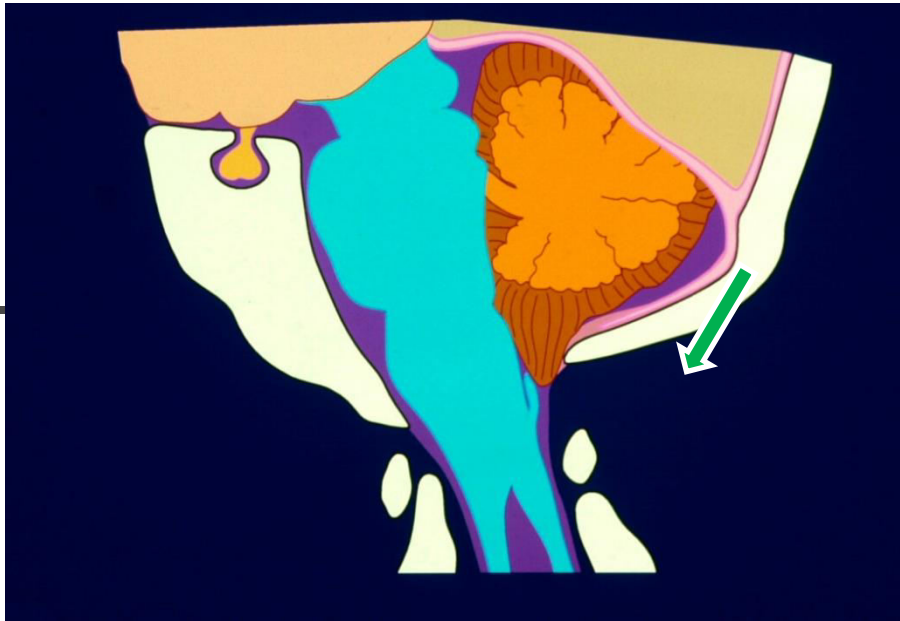
Signe du ménisque au pôle inf. cerebelle =
Preuve suffisante du surremplissage de la FP

- Diminution Volume moins constante
- Niveau Hernie Tonsillaire mal corrélé à Taille FP



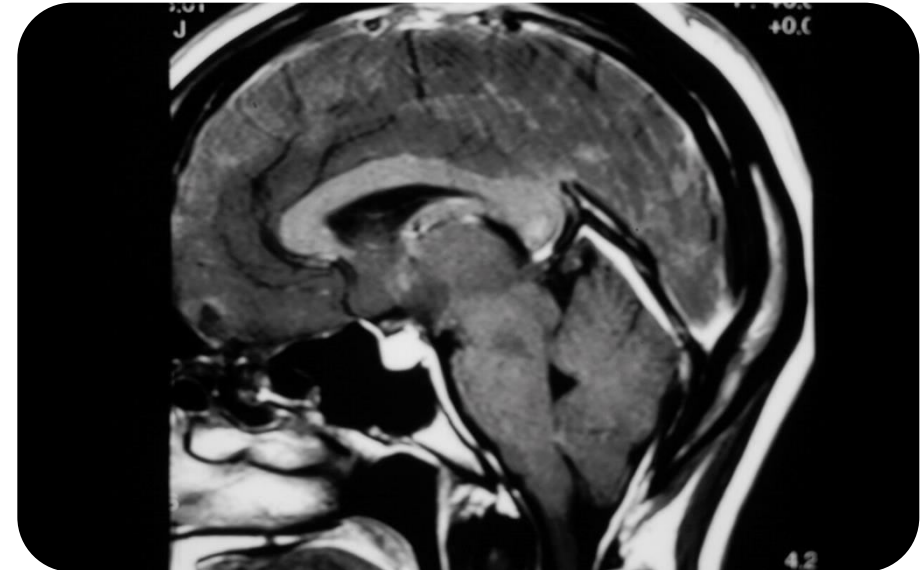
Architecture : « Floor Type »

Os occipital horizontalisé
épaissi et compressif

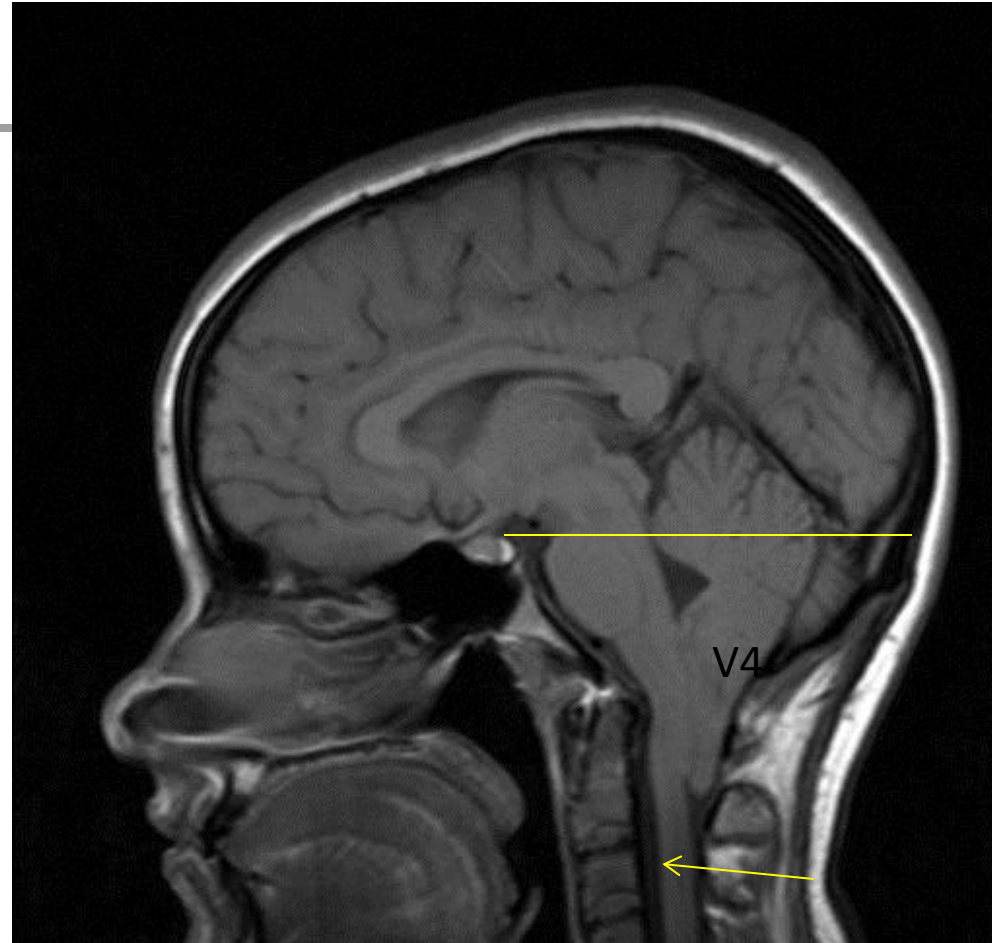
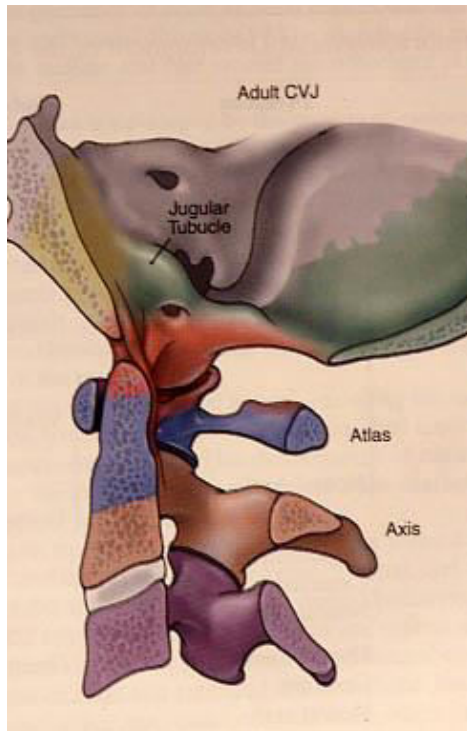


Architecture « roof type »

Insertion basse du torcular
Verticalisation de la tente

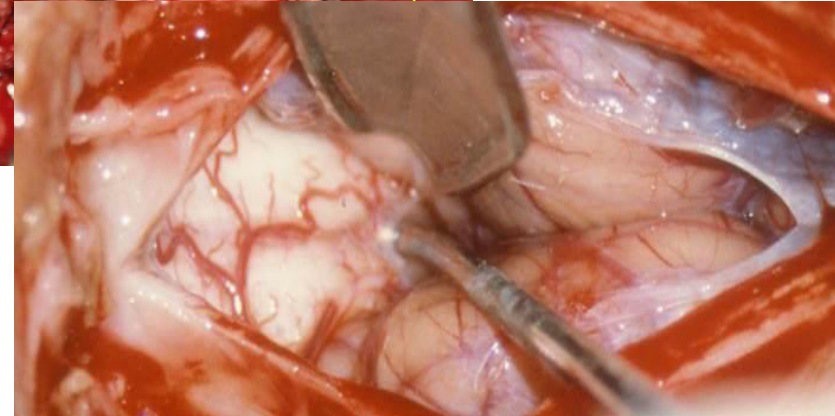
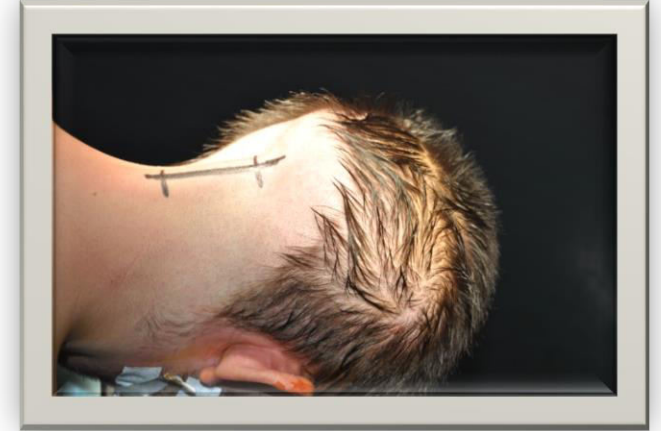
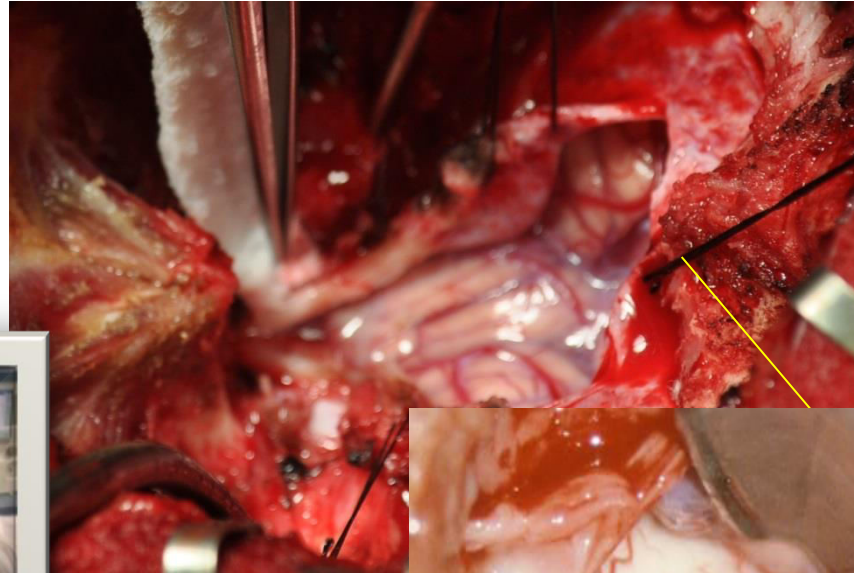
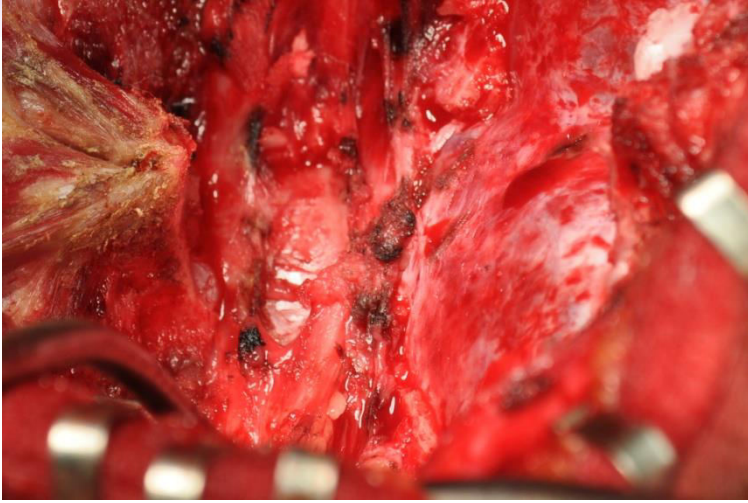


Chiari Type 2

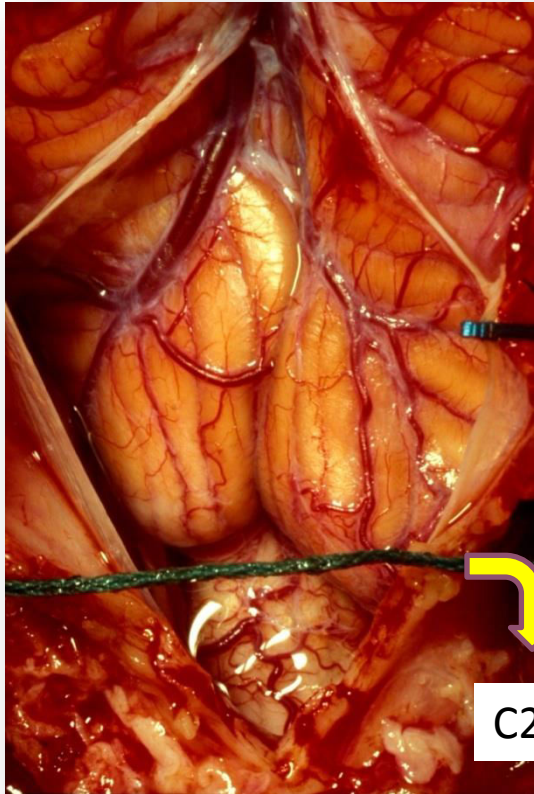


Résection / coagulation des amygdales

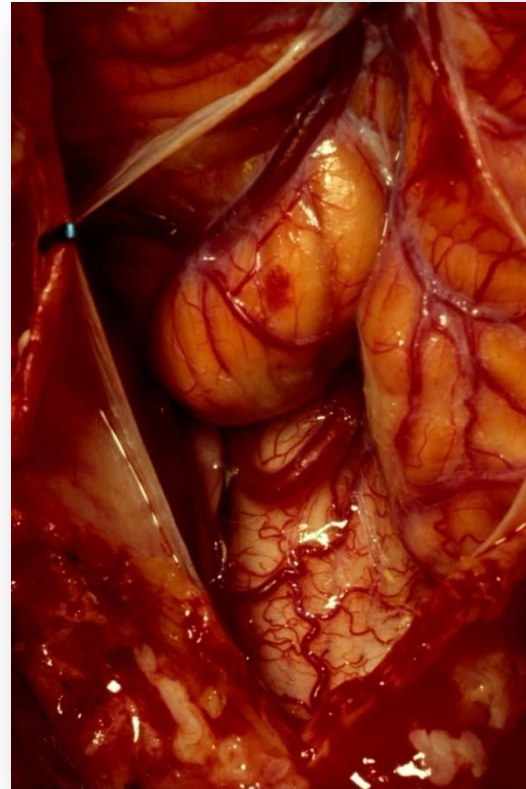
II – A safer surgical procedure in routine



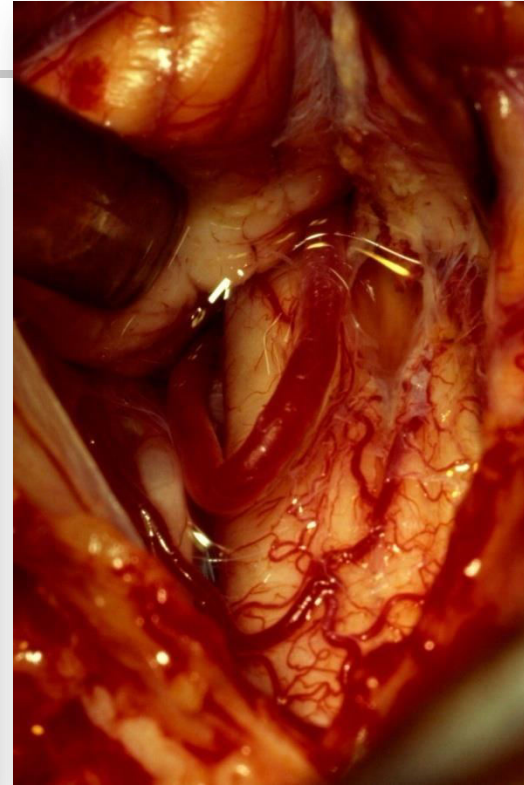
Ouvrir la Dure Mère



C2

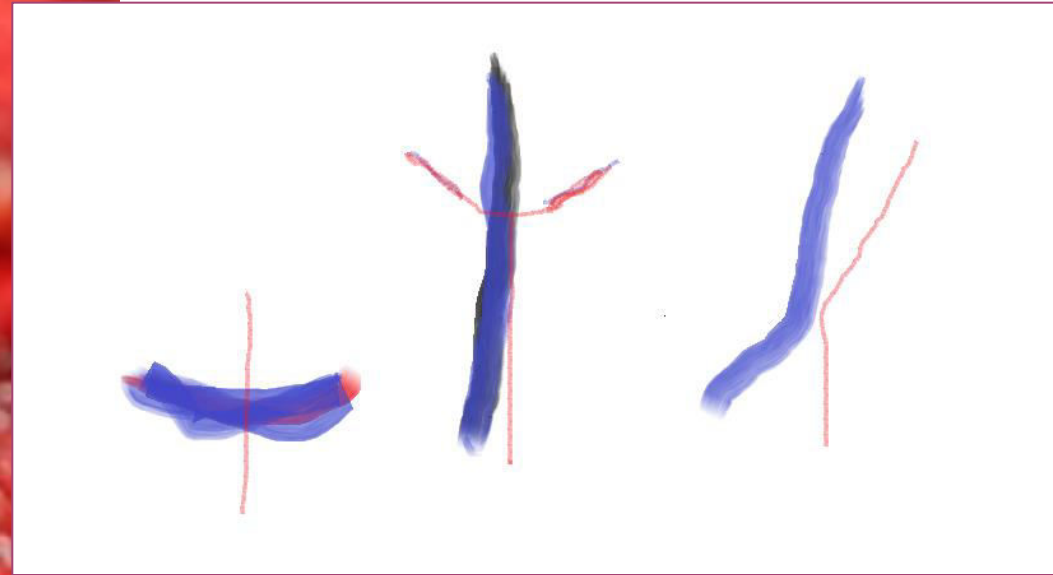
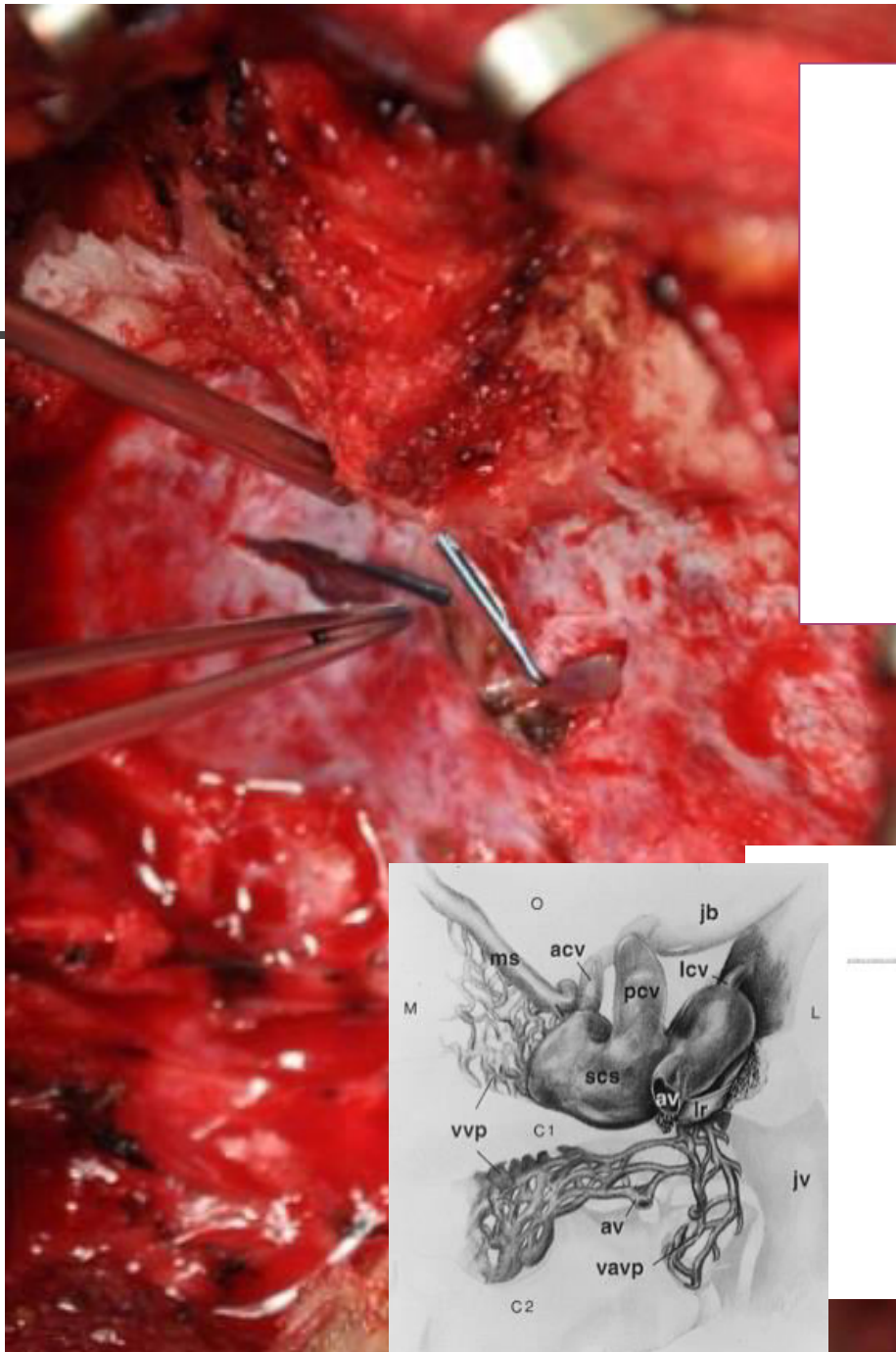


Tonsil's
coagulation

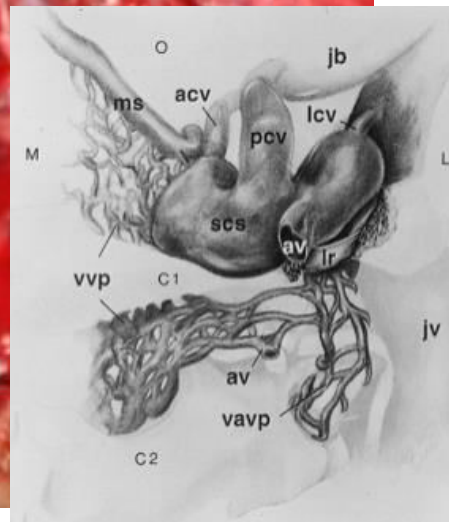


Check the Magendie





Veinous sinus : 15% cases



III- A better understanding of the future :
LONG-TERM FOLLOW-UP OF CHIARI-RELATED SYRINGOMYELIA IN
ADULTS: ANALYSIS OF 157 SURGICALLY TREATED CASES

Mean Follow-up
7 years et 3 month (82 - 204 mois)

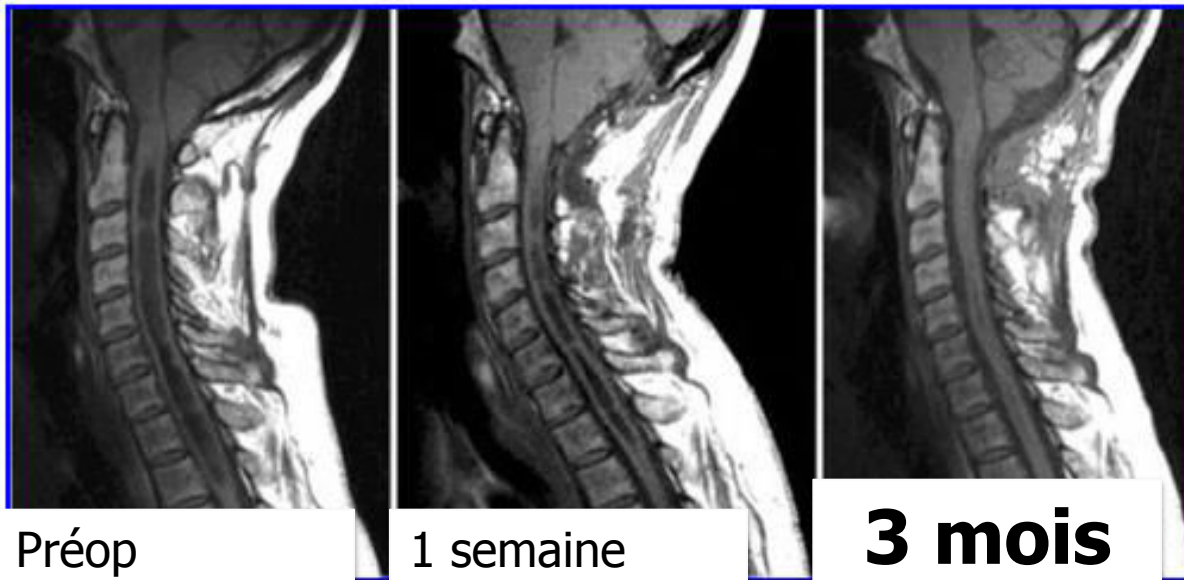
- 99 patients improved : 63%
- 48 patients stabilized : 30%
- 9 patients worsened : 6%
- 1 patient died : 0.6%

N. Aghakhani, F. Parker, P. David, S.Morar, M.Tadié
Neurosurgery, Vol 64(2) , February 2009, p 308–315

Post op evolution of the cavity after dural opening in CM1

- Wetjen, Heiss, Oldfield JNS, feb 2008 (1), 2

Surgical Outcome	Follow-Up Visit		
	3-6 Mos	1 Yr	≥ 2 Yrs
clinical result			
improved	27/28 (96)*	21/22 (95)*	17/18 (94)*
no change	1 (4)	1 (5)	1 (6)
worsened	0	0	0
neuroimaging result			
syrinx resolved†	25/29 (86)	21/23 (91)	19/19 (100)



Post-op evolution of the syrinx after dural grafting

Grade Post-op

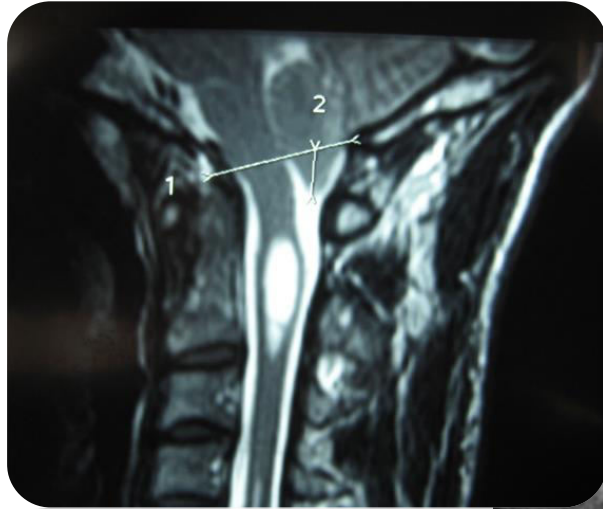
Grade
Pré-op

	A	B	C	D	Total
A	10	44	17	20	91
B	3	13	28	6	50
C	0	2	10	3	15
D	0	0	0	0	0
Total	13	59	55	29	156

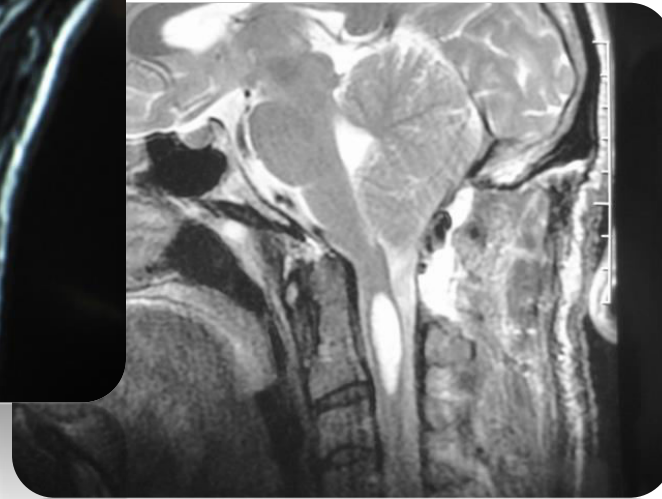
Neurosurgery ,
Vol 64(2) , Feb 2009, p 308–315

reduced	:	75%	Complete resorbtion : 19%
Unchanged	:	21%	
increased	:	3%	

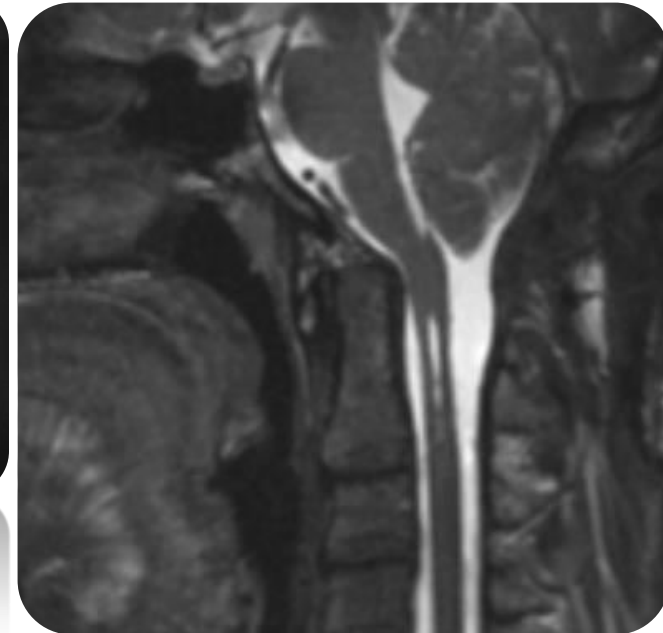
IV- A better knowledge of new surgical approach and their limits :
Extradural decompression +/- dural splitting



Pré-op



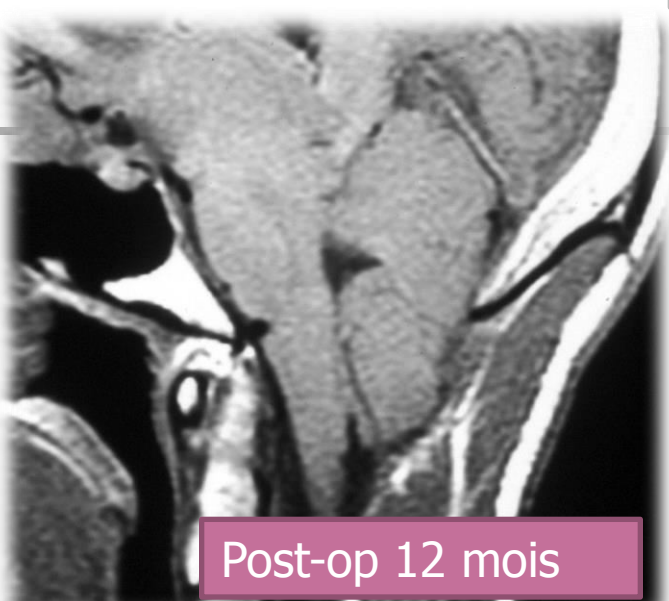
6 months



18 months

Extradural decompression : higher risk of incomplete decompression

groupe adultes



Réintervention pour	2008	2009	2010	2014
Décompression osseuse extradurale inadéquate	2	4	6	14
Décompression ostéodurale inadéquate	1	2	3	12
Méningocèle	1	1	2	2
Arachnoïdite	2	2	2	3



Post-op2 after dural plasty

Intra-opérative ultrasound recording

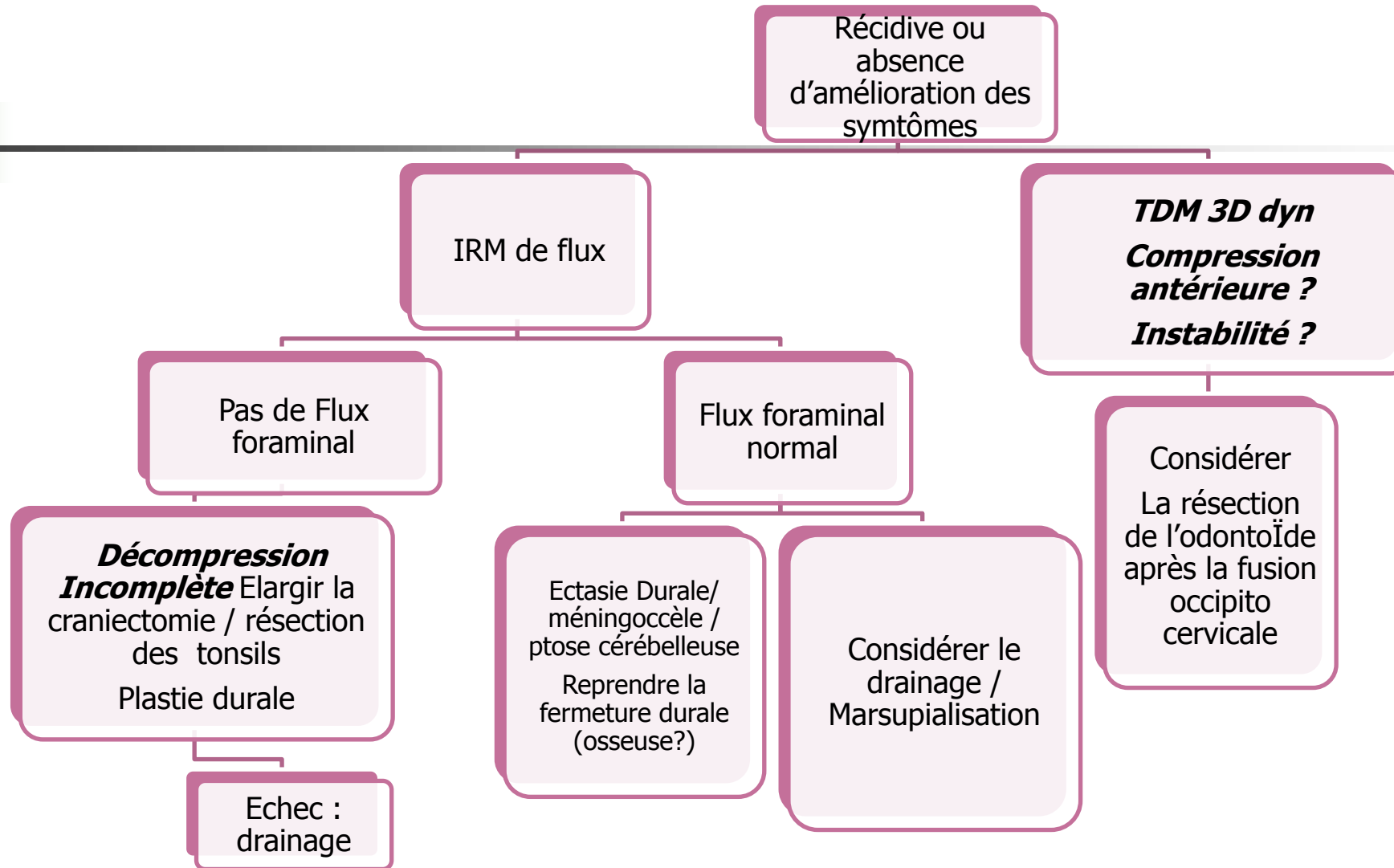
Etude de la dynamique des mouvements des structures anatomiques et des flux de LCS



Procédure extradurale / Intradurale

"E. Oldfield, J Neurosurg 80:3-15, 1994"

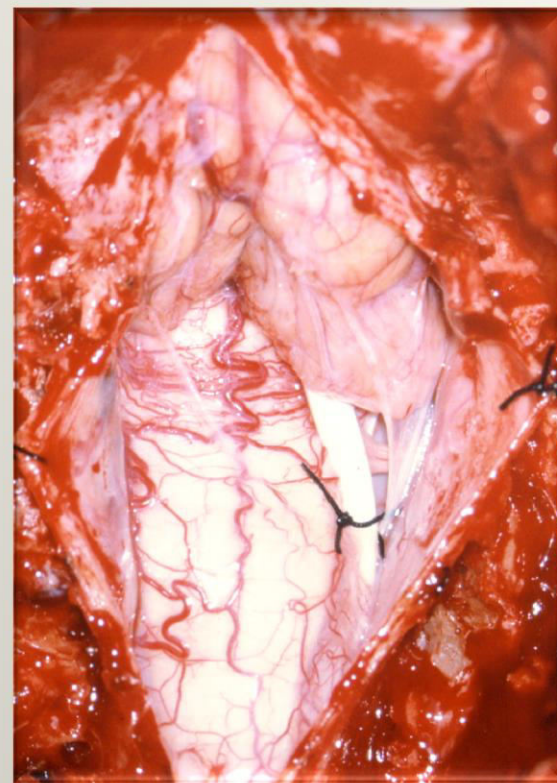
V – A BETTER MANAGEMENT IN CASE OF FAILURE



Persistence of symptoms and syrinx : « focal arachnoïditis of the CCJ »



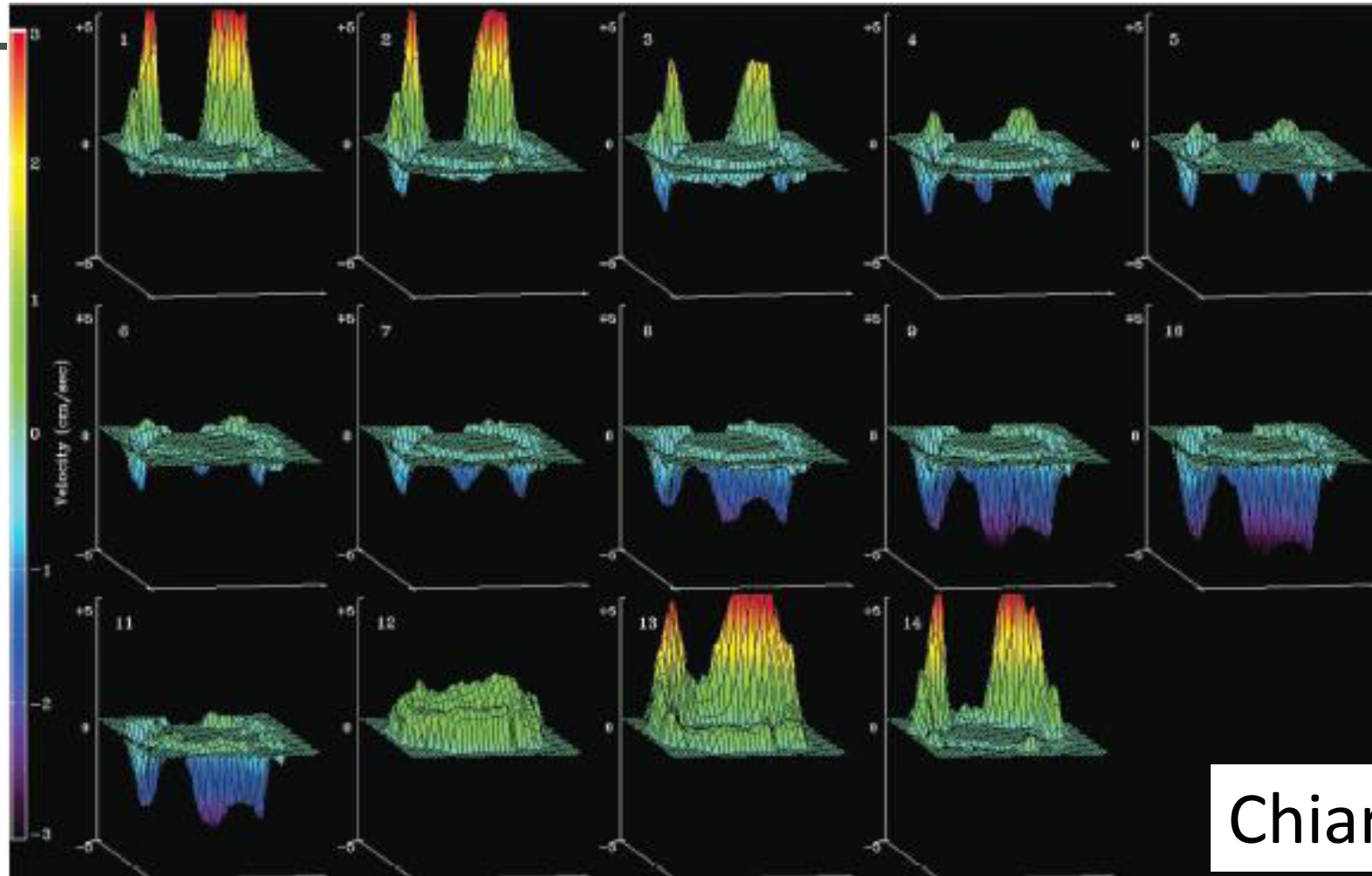
Post-op 1



Post-op 2

Arachnoidolyse / Drain V4 – ESA cervicaux

CSF flow at the foramen magnum level



Chiari I

From Quigley, Radiology 232:229–236, 2004

Arachnoïdite Foraminale

Principale cause d'échec

Série de Bicêtre
(n=153)

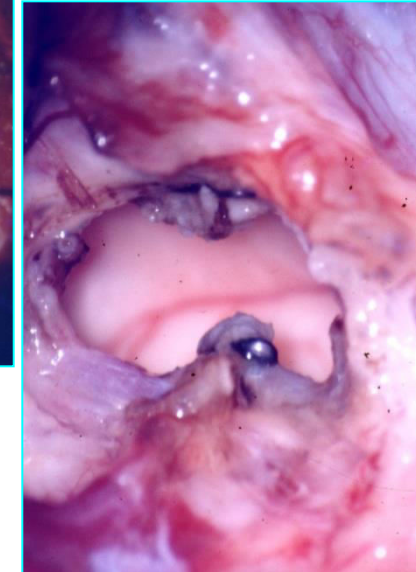
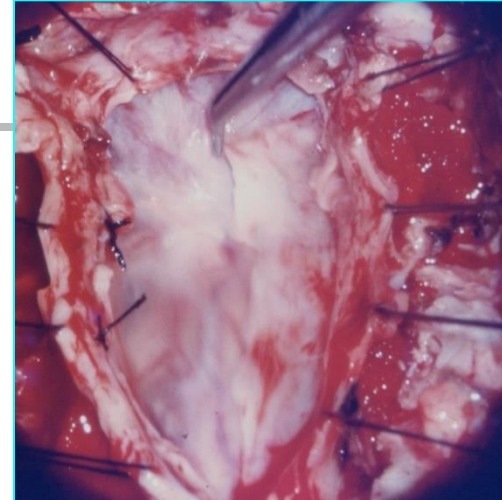
Archnoïdite (13%)

- Dense et diffuse : 5%
- Focale : 8%

*Obliteration du 4ème
Ventricule*

- Complète (1/4 avec hydrocéphalie) ou
- partielle

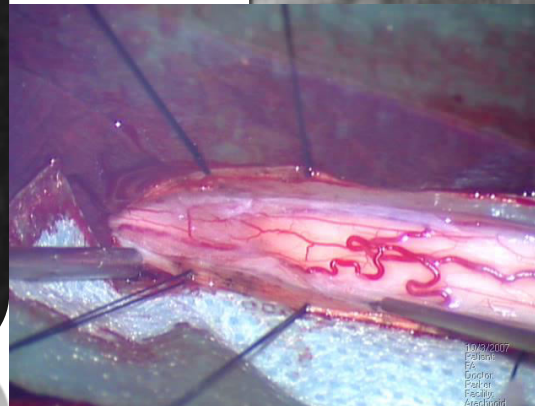
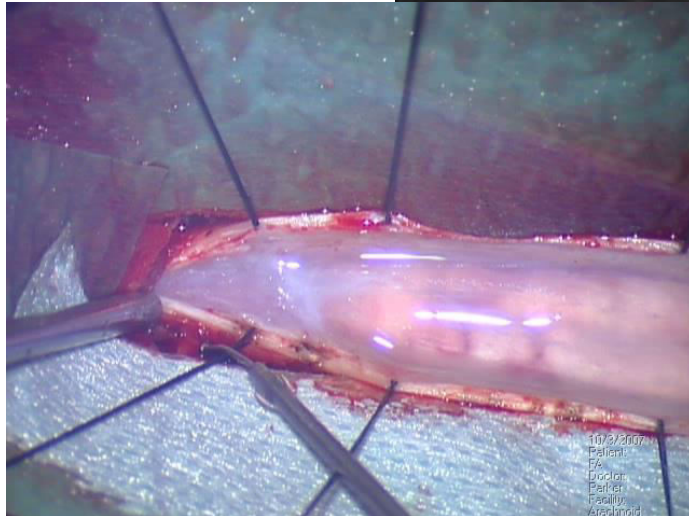
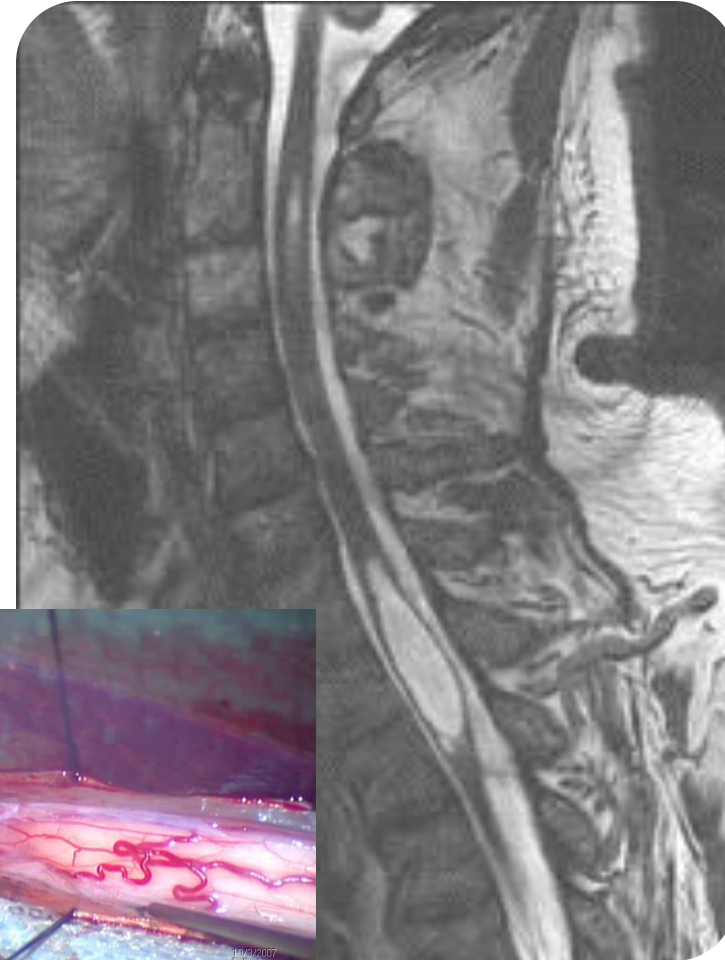
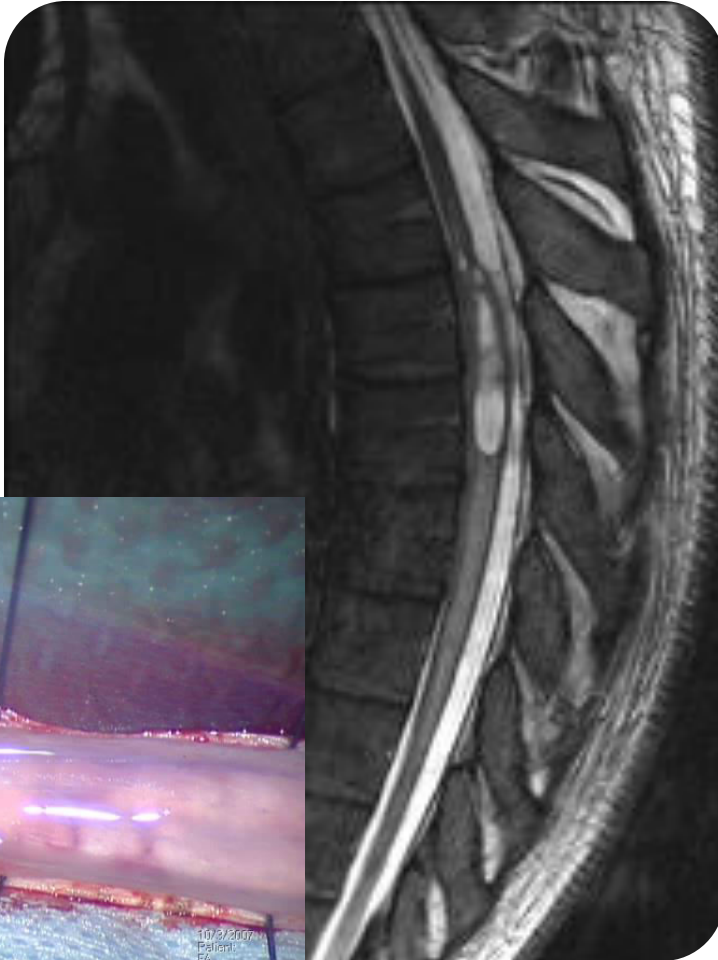
CM1 + Syrxinx



**Résection
amygdales
Ouverture V4**



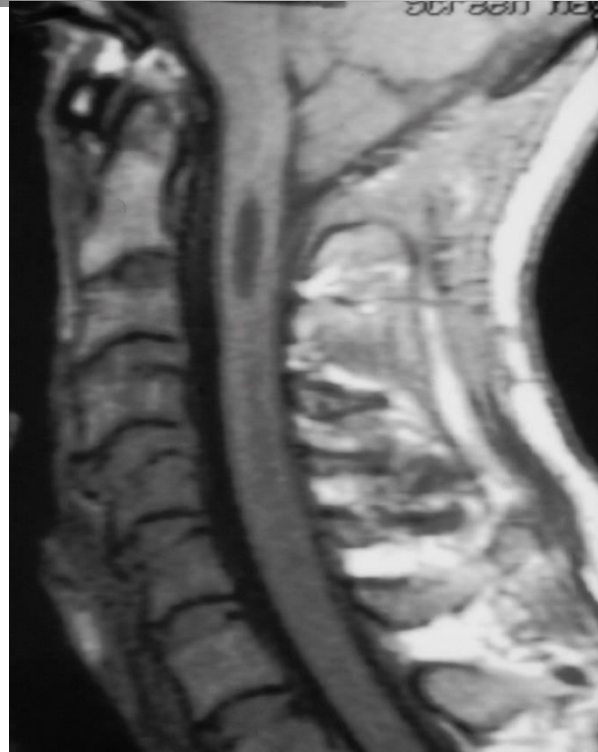
VI- Focal arachnoiditis : a rare cause of failure



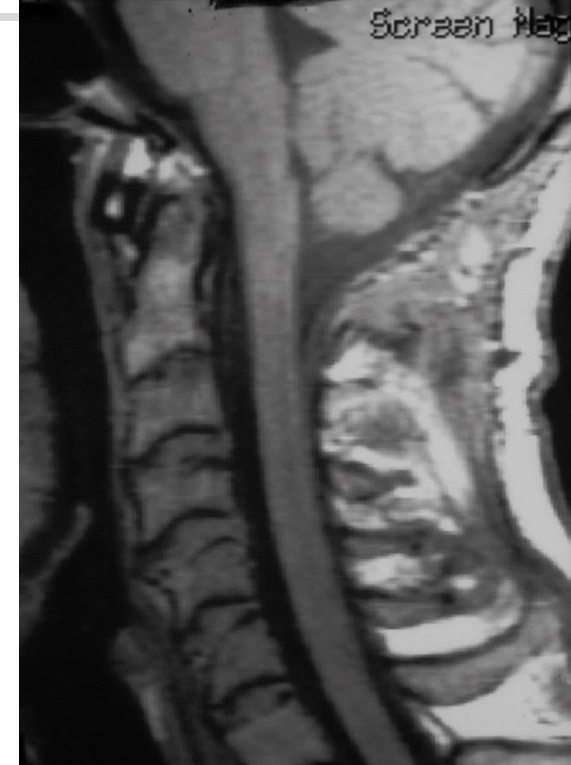
Persistence of syrinx + symptoms : Incomplete decompression



Pre op IRM



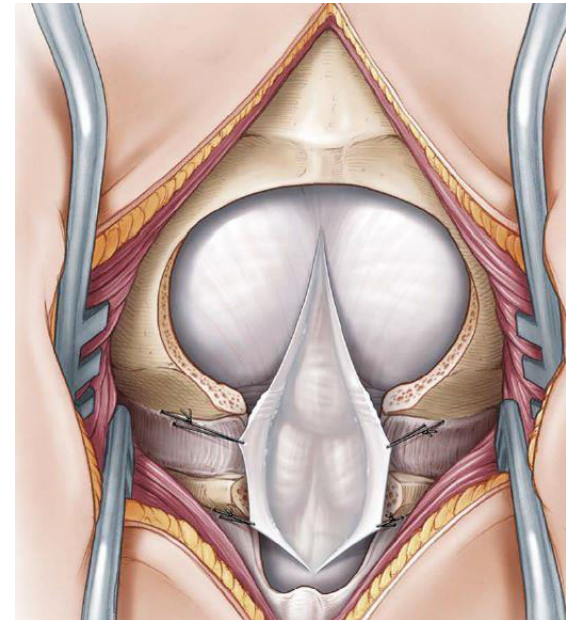
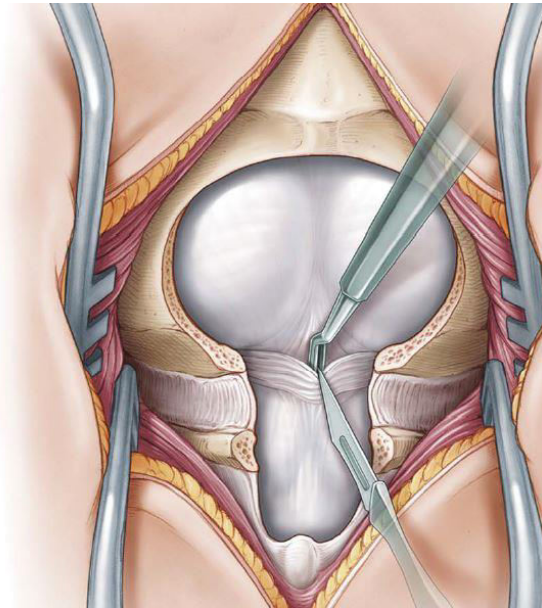
After first decompression



After tonsils resection

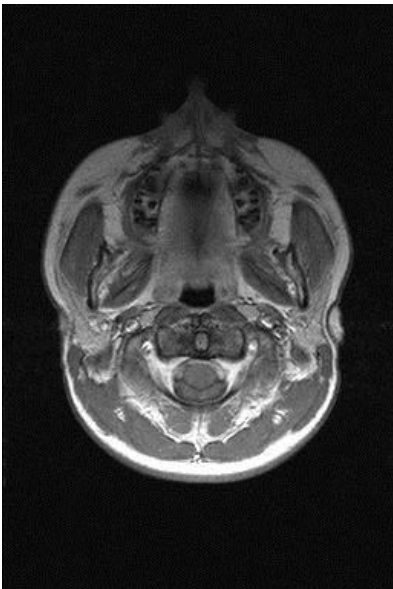
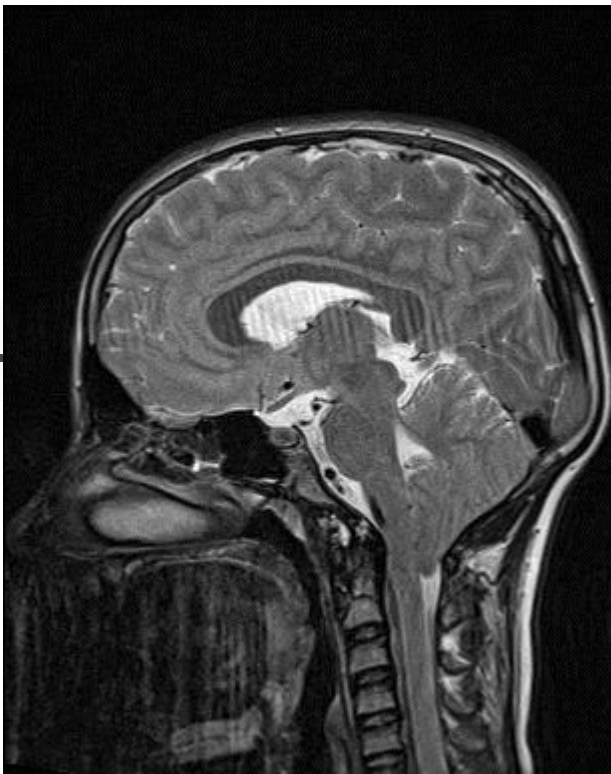
Dura splitting decompression for Chiari 1 malformation without syringomyelia in Pediatric Population :

Zachary N. Litvack, Neurosurgery 72:922-929, 2013



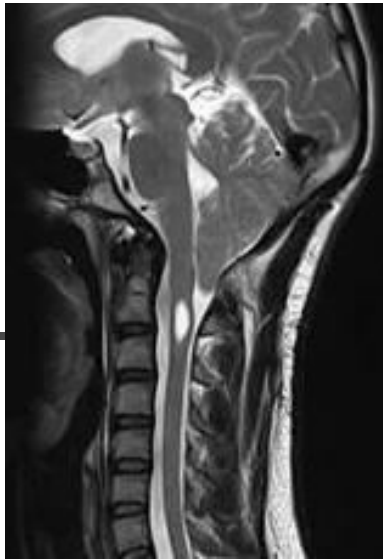
Cost savings of \$ 3026 per patient : less operating room time and shorter hospitalisation stay

Cas N°1 : Jean, 17 ans



- Préop
 - Céphalées déclenchées par effort rire toux, vertiges, vomissements,
 - ex normal rot vifs
- CHIR 2014 Chiari
 - Résection occipito-C1,
 - « dédoublement » dural
 - Pulsatilité « satisfaisante » sur echo per-op

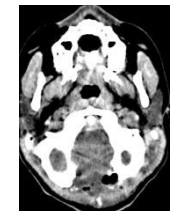
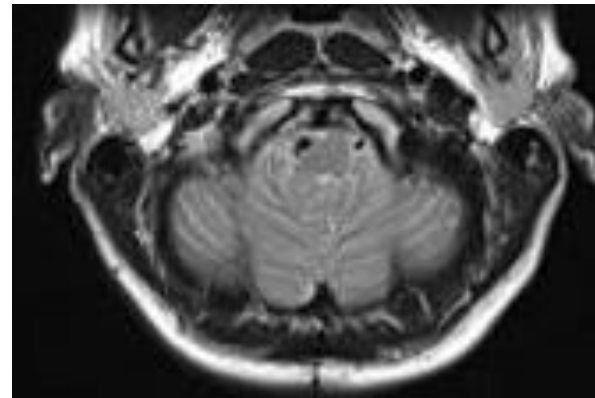
Cas N°1 : post-op 12 mois



Apparition d'une cavité

■ Post op

- Amélioration : disparition des céphalées, vertiges, vomissements et paresthésies, a repris le sport
- Mais apparition d'une cavité centro médullaire qui augmente progressivement



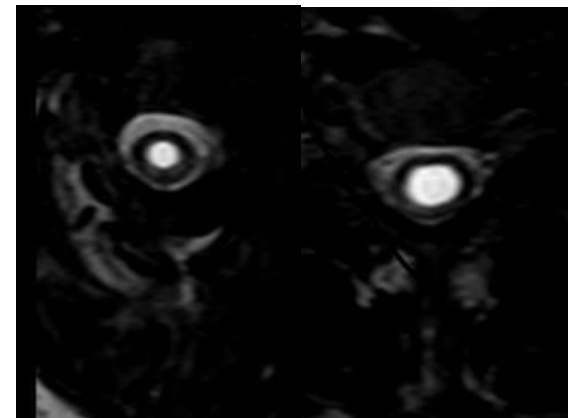


Pré-op

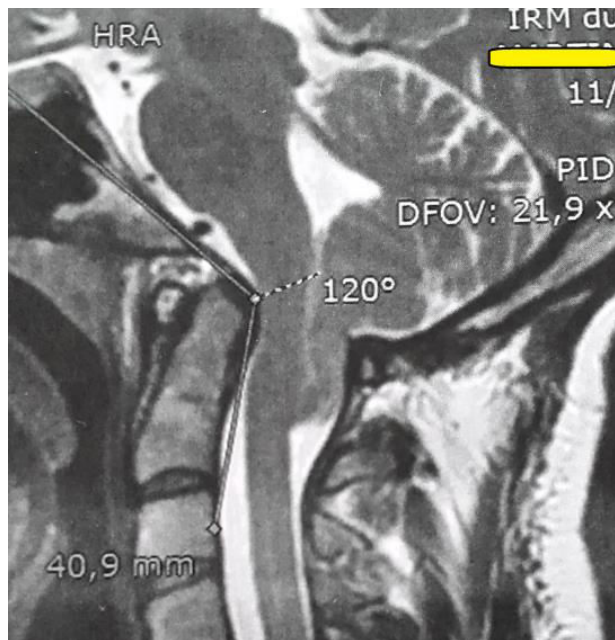


Post-op à 12 mois

- Pre op: céphalées, à l'effort, vertiges, troubles de déglutition, paresthésies des mains, ataxie,
- Chirurgie 2014 : ouverture large, laminectomie C1 et C2 **dédoublement dural**
- Post op: amélioration, puis ré aggravation
- 2 mois post op : nuchalgies, paresthésies des mains, fonte des avant-bras, asymétrie voile.

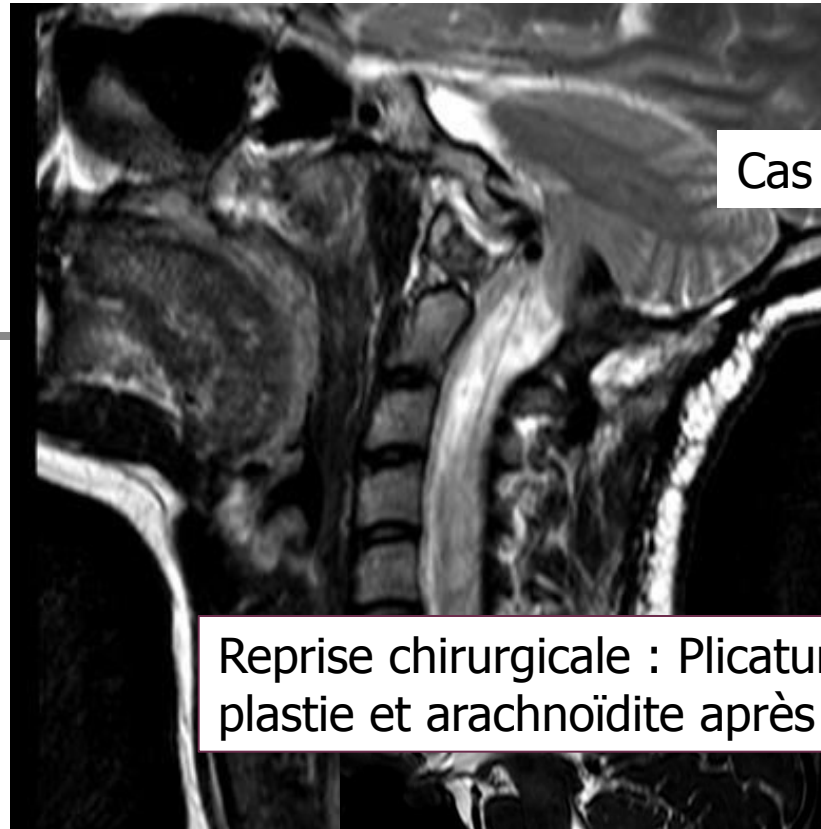


Cas N°3 : MARTINE : pré op : Chiari sans syrx



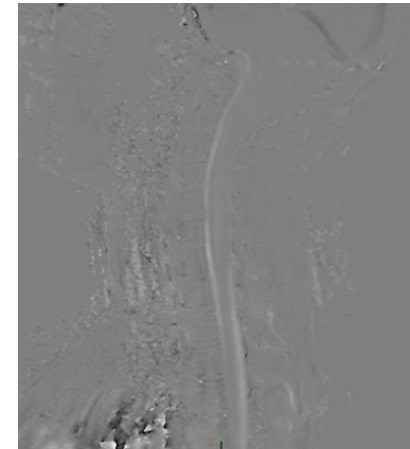
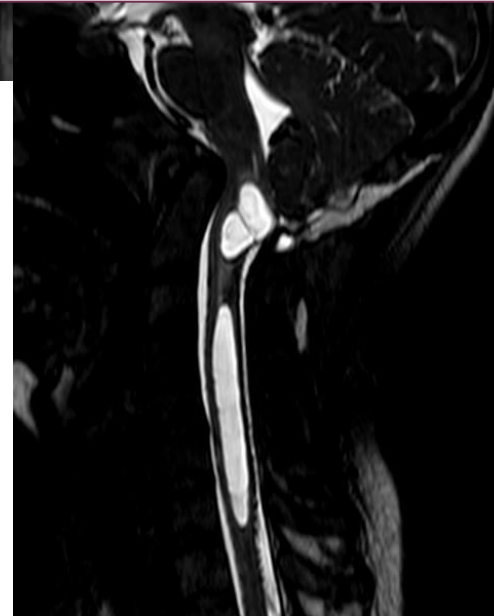
PRE OP :

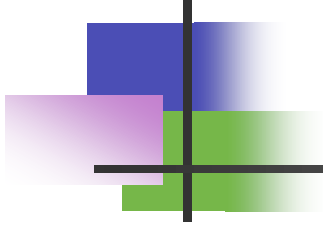
- 2013 : Céphalées morphino-requérantes
- Chiari 1 sans syrx
- Chir 1 : décompression ouverture du foramen, résection C1 et une amygdale, plastie durale, pas d'arachnoïdite, encollage
- Suites post op : persistance des céphalées, hydrocéphalie, et ***apparition syrx***

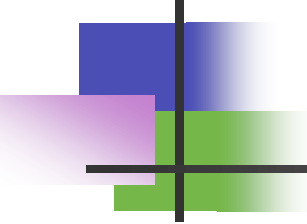


Cas N°3 : IRM à 12 mois

Reprise chirurgicale : Plicature compressive de la
plastie et arachnoïdite après encollage





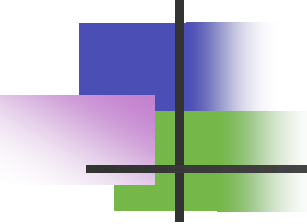


Discussion

Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation Type I in pediatric patients : a meta-analysis J Neurosurg Pediatr 2008 :

« PFD + duraplasty is associated with a lower risk of reoperation but a greater risk for cerebrospinal fluid-related complications. »

Durham SR et col



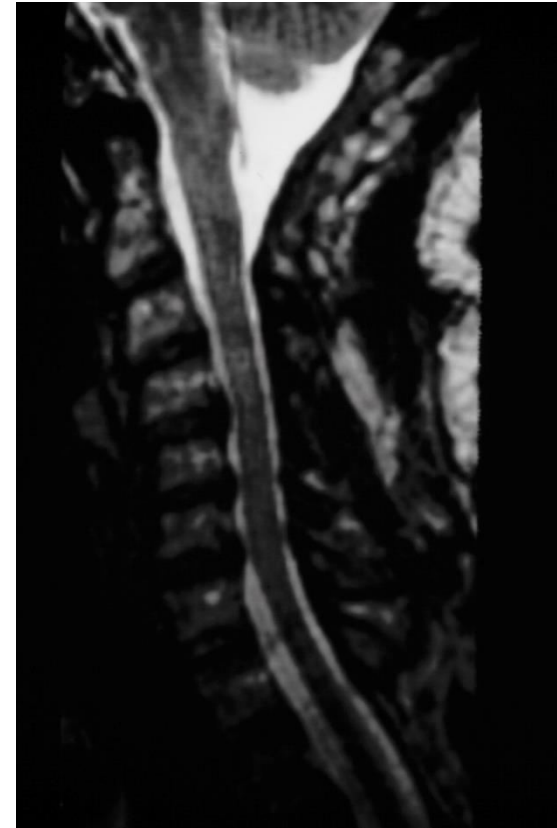
Discussion

Decompression + dural plasty :

KEKLAMP *Neurosurgery* 2012 : n=371

- ❖ Amélioration à 3 mois dans 82% des cas
- ❖ Récidive : 7% à 5 ans et 8% à 10 ans.

Céphalées persistantes post-opératoires positionnelles "Méningocele / Pseudoméningocele"



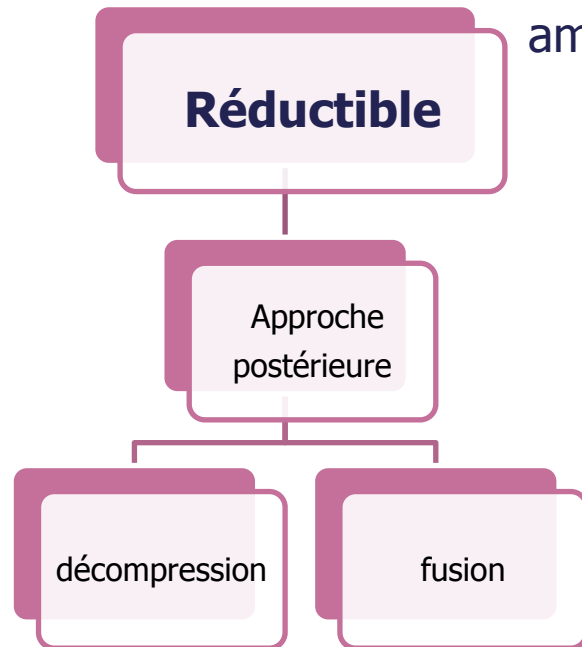
Basilar Invagination : reversible instability?

dynamic study / traction

Traction cervicale axiale progressive sous contrôle clinique et radio

3 à 4 j avant l'intervention / Halo MR compatible

IB sans CM : 82% réductible et
amélioration clinique sous traction (Goel, JNS, 1998)



Irréductible

1 - Décompression
Antérieure
transoropharyngée

2 - fusion
postérieure
Dans le même
temps

80% des enfants < âge 14 ans (Menezes)

BI + CM : 5% réductible et
amélioration clinique sous traction (Goel)

Anterior bony malformation associated in 30% of Chiari

If Chiari + anterior bony compression = 46% of lower nerve dysfunction



Risk of trachostomy if odontoidectomy

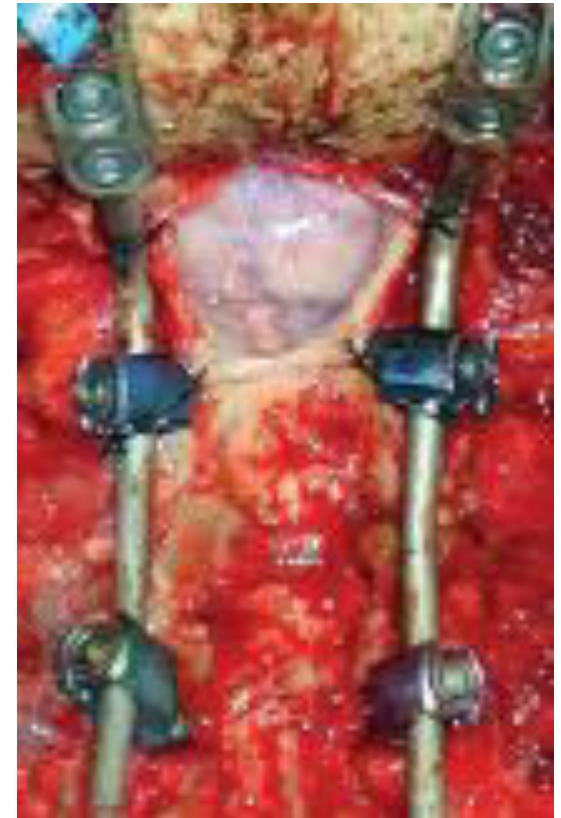
- If Chiari + ventral compression non reversible :

- transoral decompression +
Posterior decompression :

Perrini et al Neurosurgery May 2009

- posterior decompression + fixation :

Personnal experience : 18 case with anterior decompression
in only 2 cases



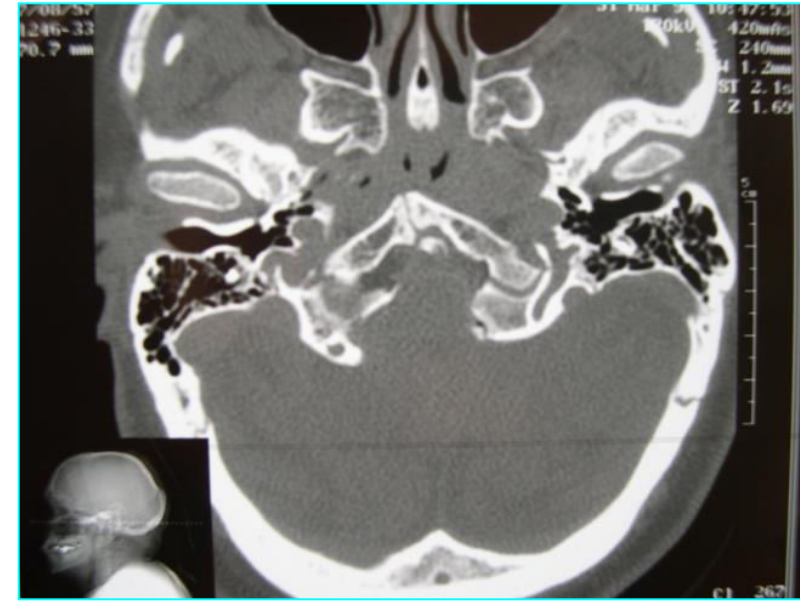
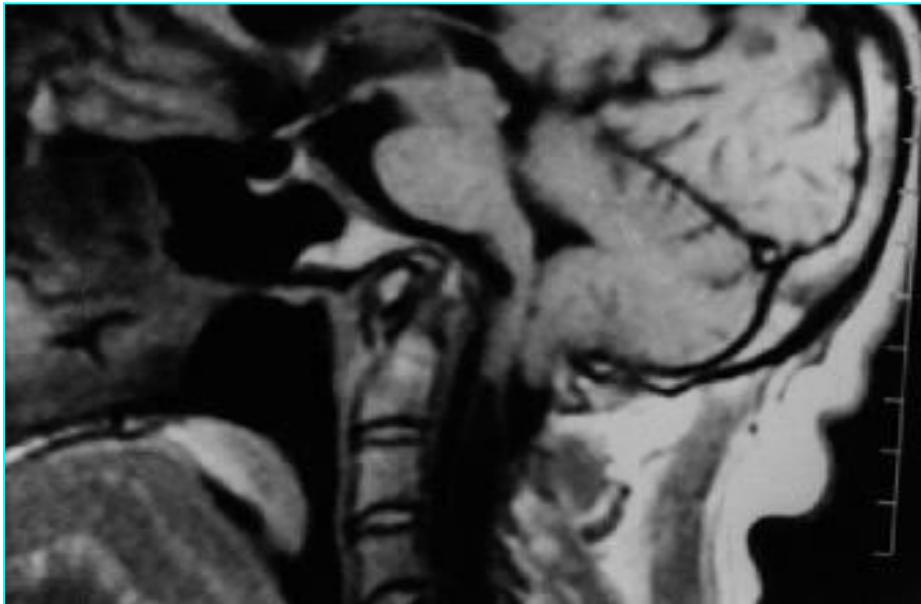
Chiari + basilar Invagination in adulte

Etude dynamique TDM Rx IRM

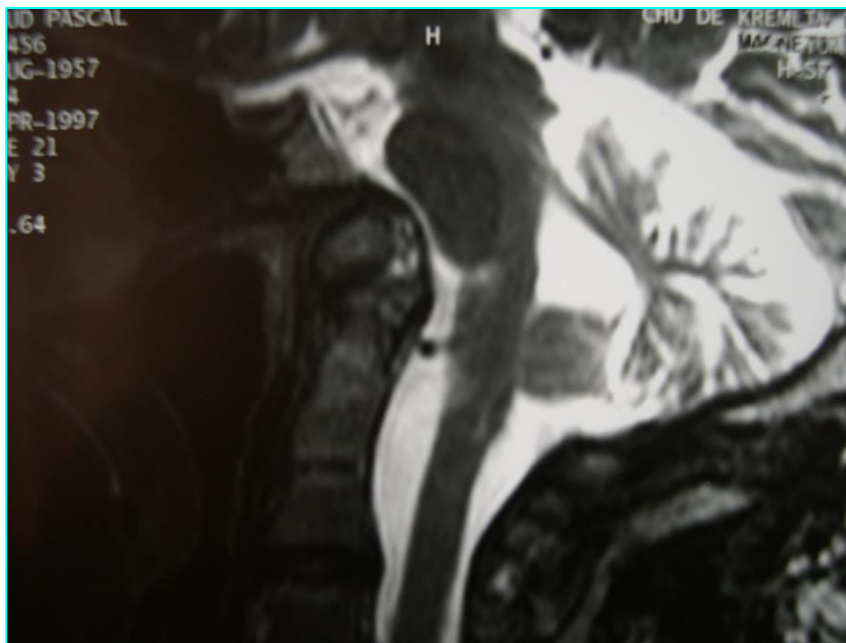
Analyser l'étiologie de l'IB

- Si instabilité Atlantoaxoïdienne réductible
: fixation postérieure + décompression
postérieure
- SI IB non réductible :
 - Gros Chiari + petite IB :
decompression + fixation post
 - Forte IB + petit Chiari :
odontoïdectomie + fixation post
même temps

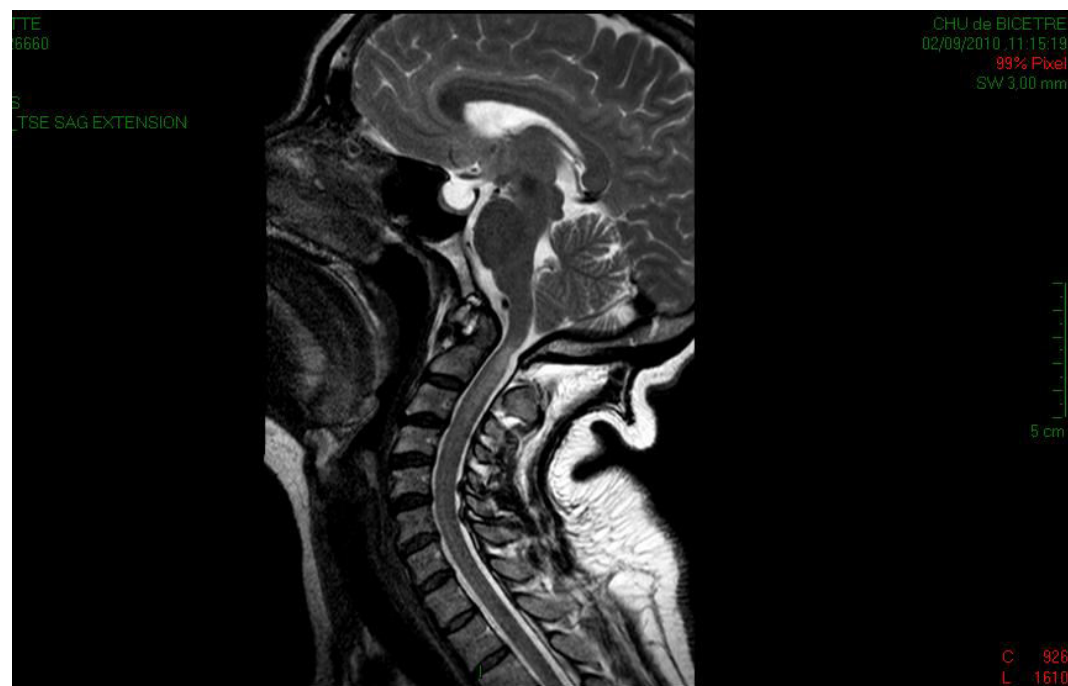
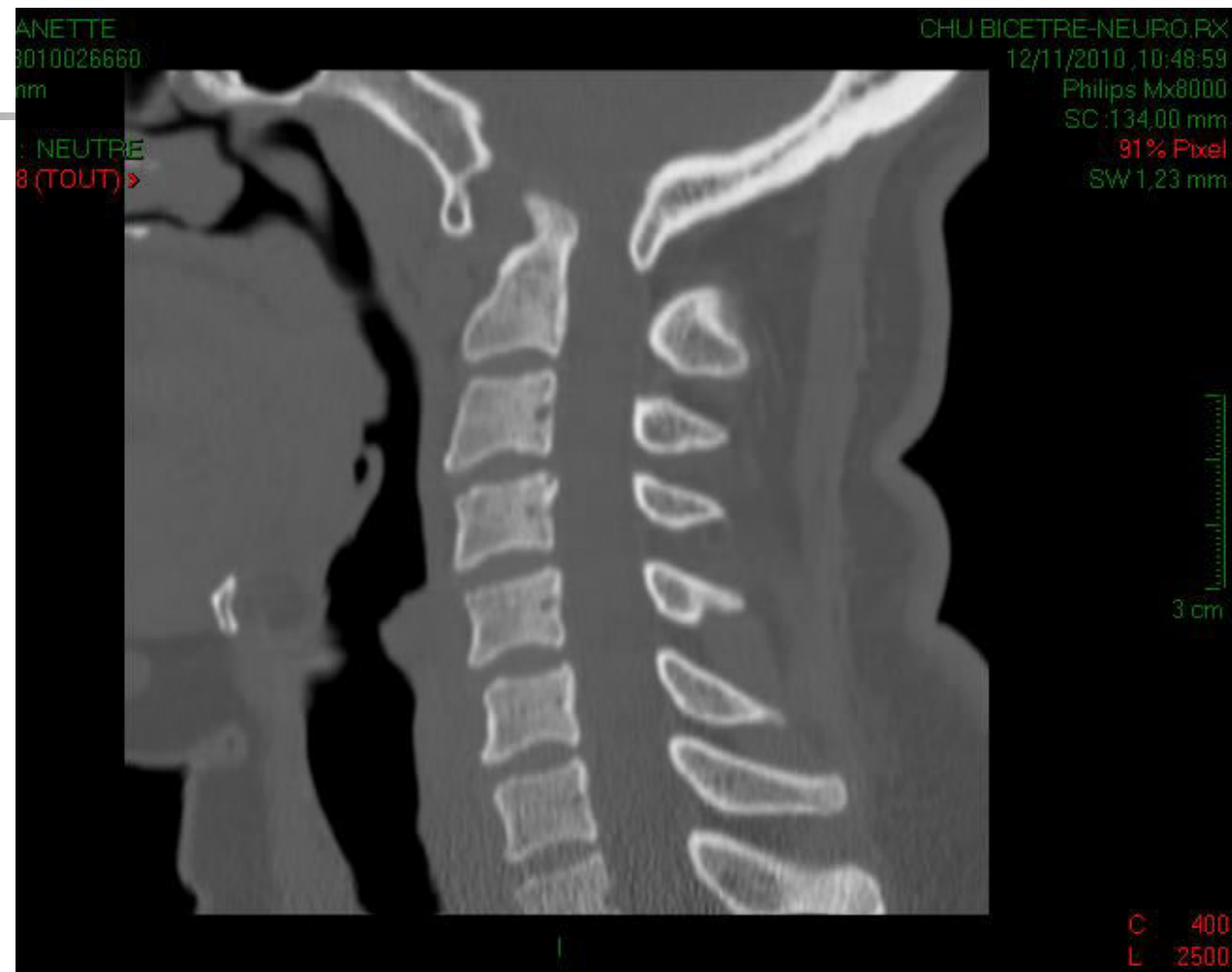




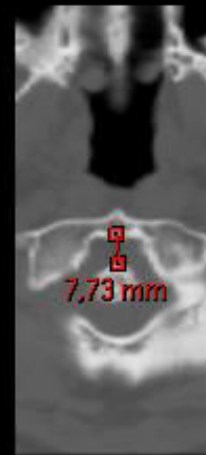
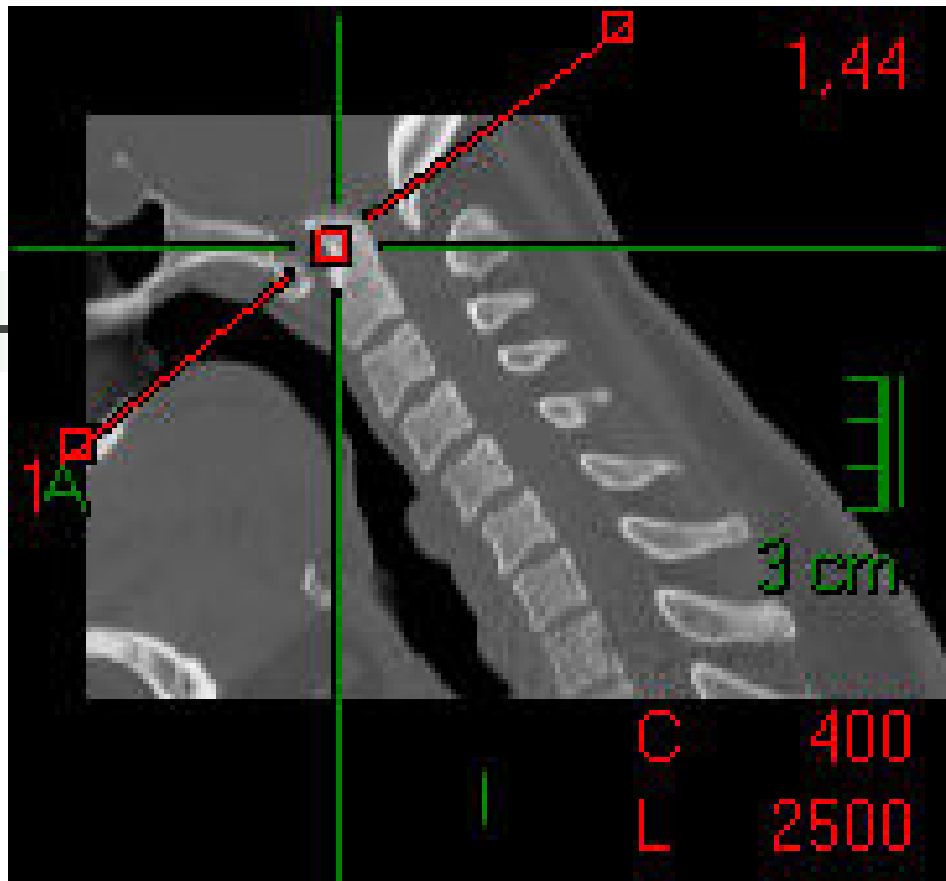
Pré-opératoire



Post-opératoire après décompression postérieure + plastie durale + tonsillectomie
+ greffe laçage vis résorbable



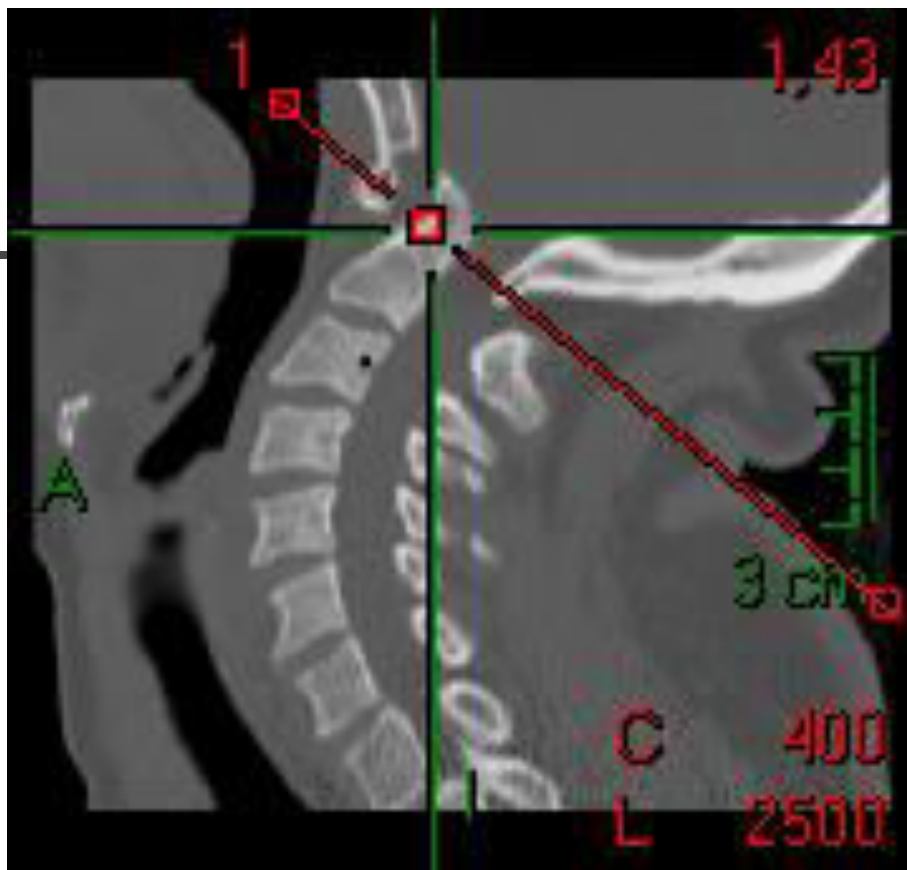
IRM extension

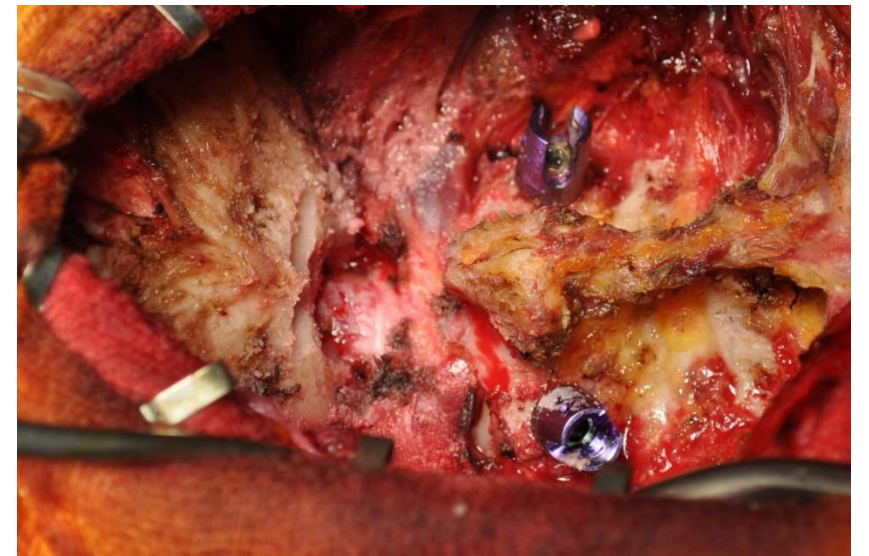
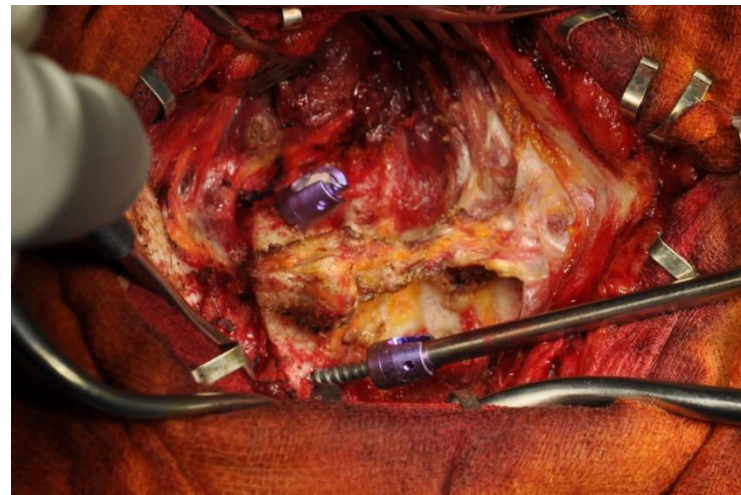
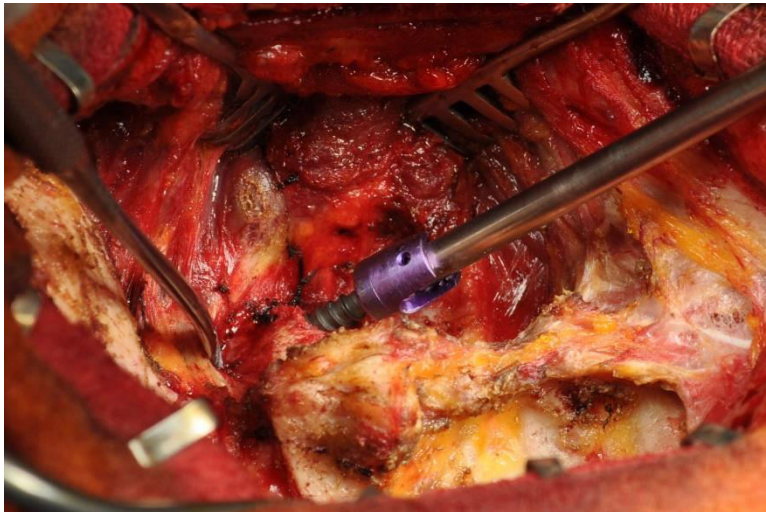
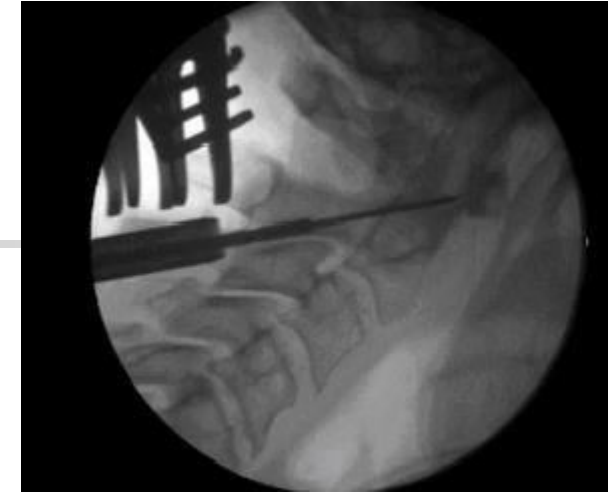
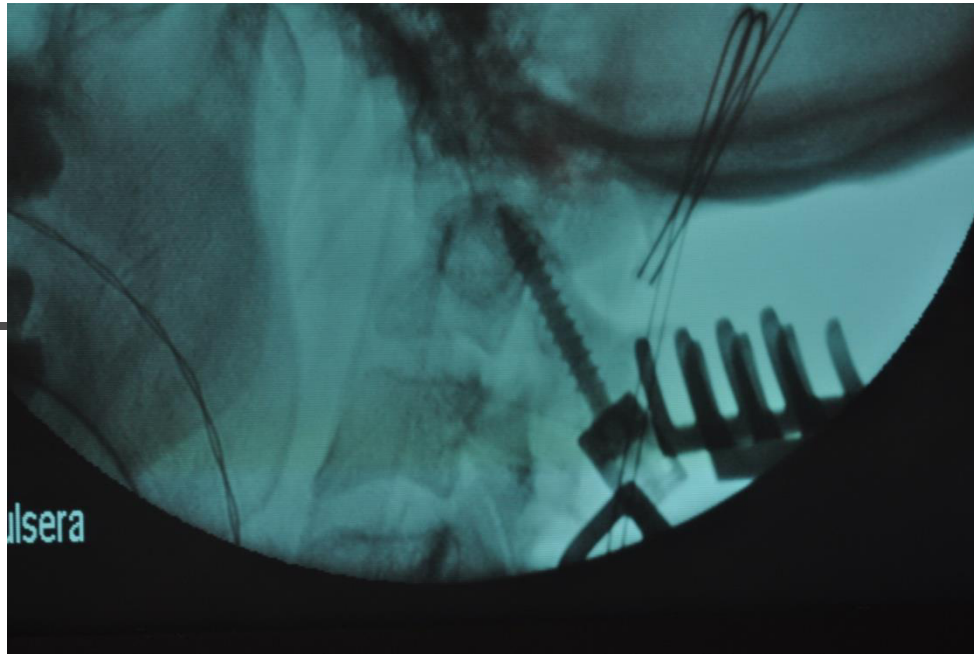
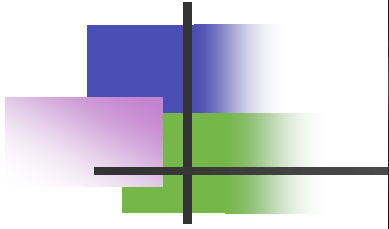


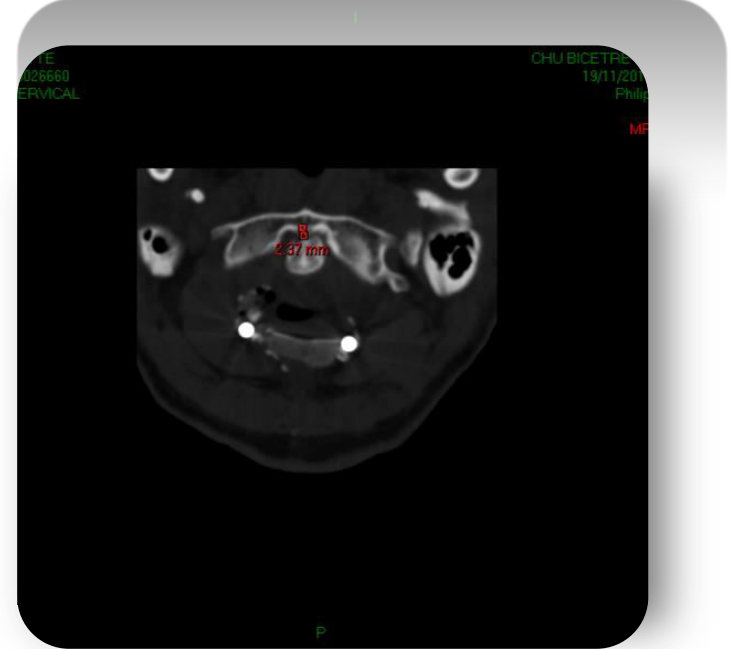
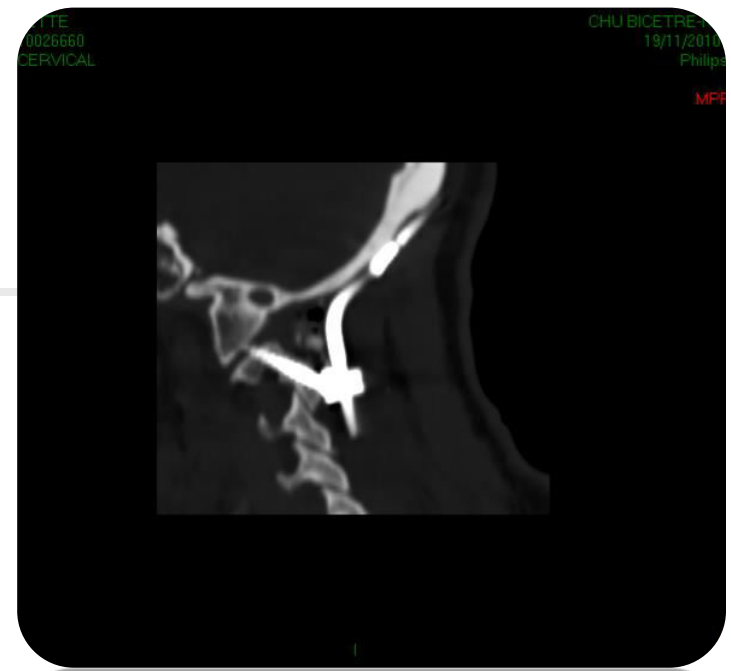
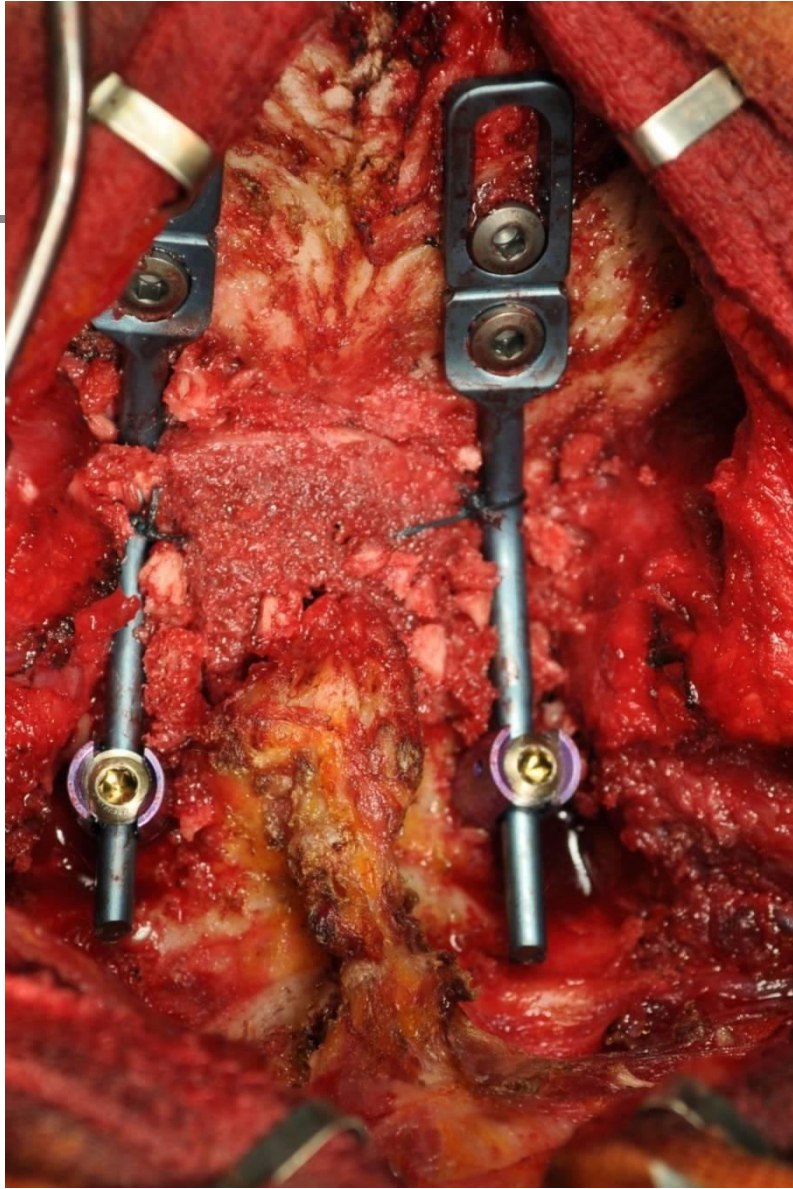
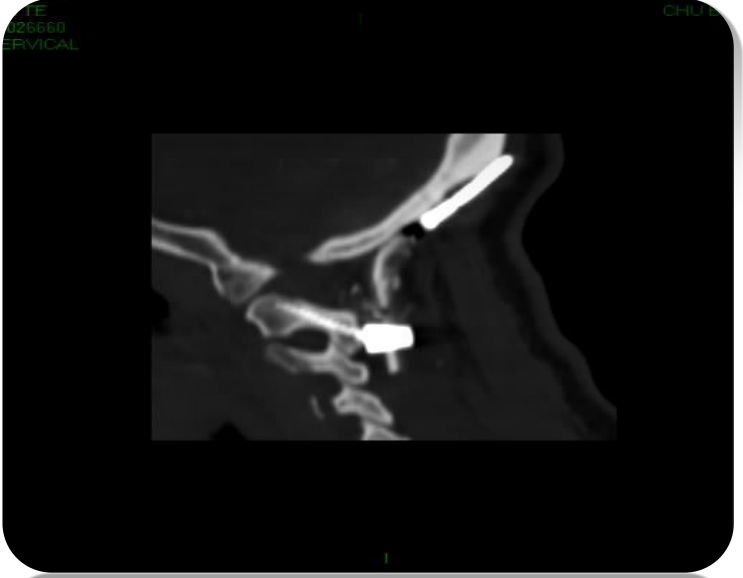
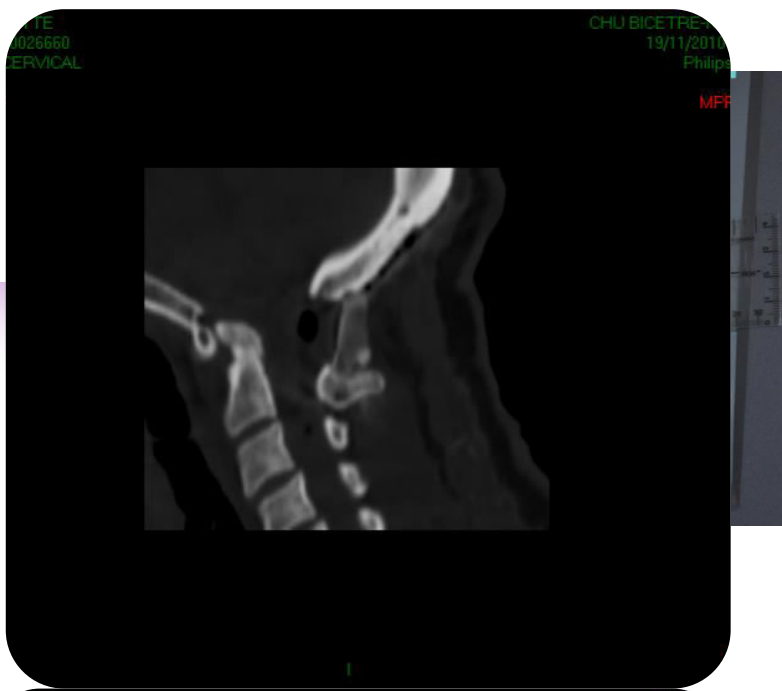
La dislocation est elle réductible?
TDM dynamique

flexion

PS









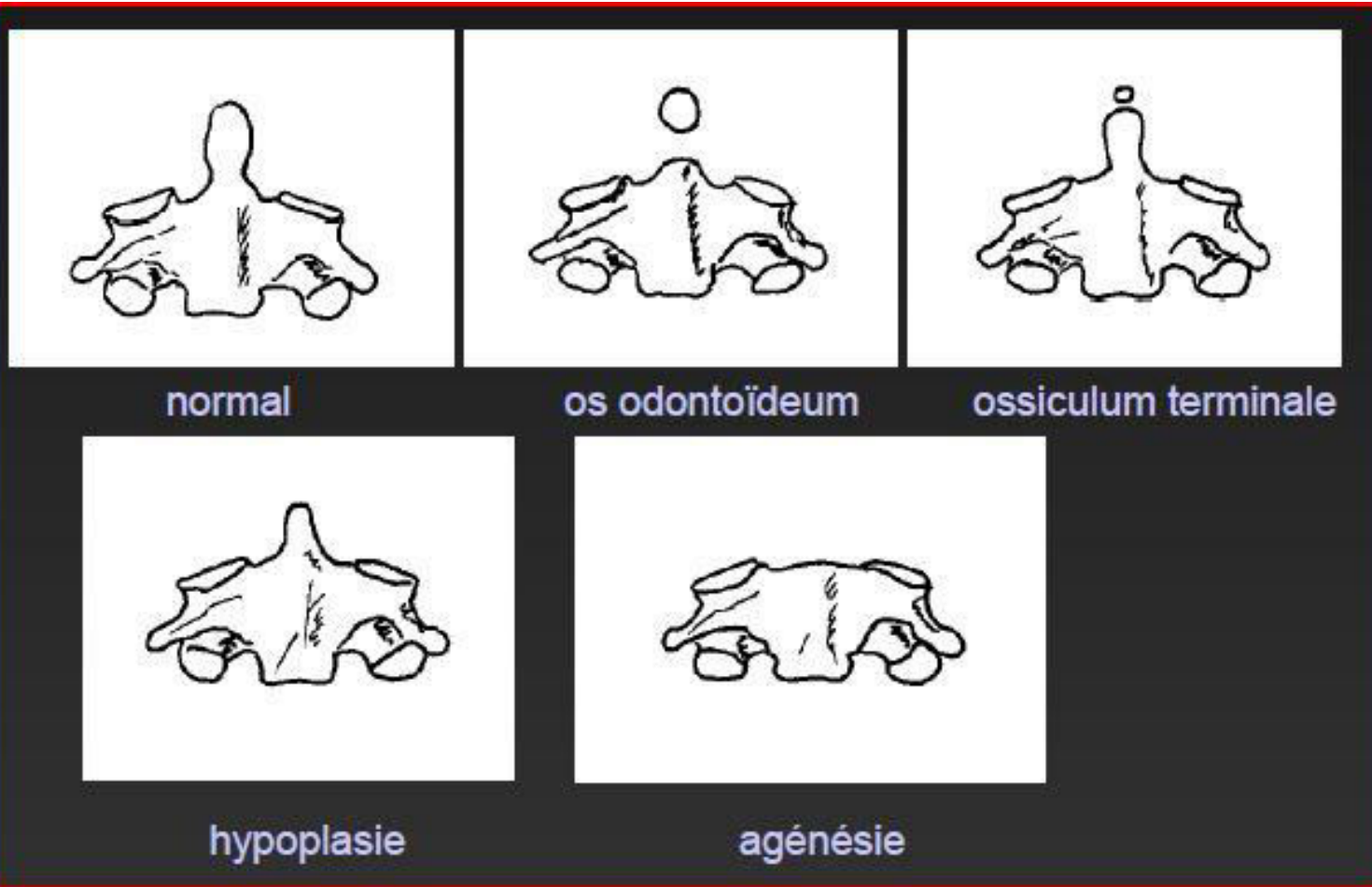
2000



Pseudarthrose congénitale de
l'odontoïde avec déplacement
en avant : en hyperextension
réduction du déplacement .



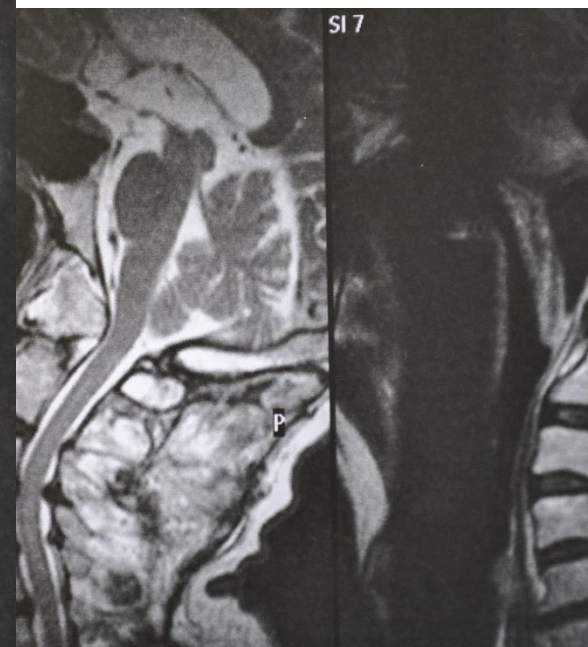
Malformations de l'Odontoïde



Hyperplasie / vestiges du disque atlantoaxoïdien (non pathogène)



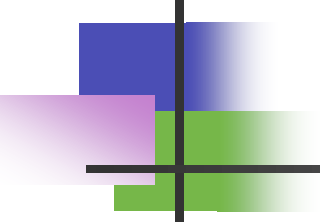
2010 : 8 ans après laçage greffe
C1-C2





Regression des symptomes





VIII- A better organization in research and European collaboration

- Filum terminal section protocol
- Eurogroup /E-Crf / Palma Ciaramitaro
- Association of patients

Thank you for your attention