

Syringomyelia Demystified

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Conflict of Interest

The study of the Physics of CSF Dynamics and Intracranial pressure has been the subject I have been researching for over 30 years. Other than that I perceive no conflict of interest either economically or intellectually with this presentation

Syringomyelia

Pathophysiology and Management



Syringomyelia Is Hydrocephalus Of the Spinal Cord



Multiple Theories of Pathogenesis

- As of 1966 Syringomyelia was described in neurosurgical textbooks as a degenerative disease with no known cause or treatment
- Much hot debate from very diverse theories from multiple researchers
- Each new theory seeks to drive out and discount previous theories
- Why can't we just get along?
- Our job is to have a single unifying theory that explains all the observations!!!!!!

SYRINX: From Classical Greek mythology:

A mountain nymph of Arcadia who was transformed into a reed to protect her virtue from Pan. Subsequently Pan used the reed to create the Panpipe (flute)



PROPOSED THEORIES OF PATHOGENESIS

- Gardner: Hydrodynamic theory based on arterial pulsations
- Williams: Dissociation of pressure waves in the spinal subarachnoid spaces by standing up, coughing and straining
- Ball and Dayan: CSF from SSAS into cord blocked from going intracranial
- Oldfield: Cerebellar tonsils act as a piston with each systolic pulse

OPERATIONS BASED ON GARDNER'S THEORY

- Posterior Fossa Craniotomy with plugging of the obex (*sic*. Outlet of the central canal into the fourth ventricle)
- “Terminal Ventriculostomy” (Resection of the Filum Terminale)
- Both work from time to time, the PFD much more reliably than “terminal ventriculostomy”
- Source of CSF: The ventricles

OPERATION BASED ON THE WILLIAMS HYPOTHESIS

- Posterior Fossa Craniotomy in which the dura is opened widely and left open
- Source of CSF: The cerebral ventricles with a one way valve at the level of obstruction by tonsils or arachnoid pouches

OPERATION BASED ON BALL AND DAYAN HYPOTHESIS AS WELL AS OLDFIELD

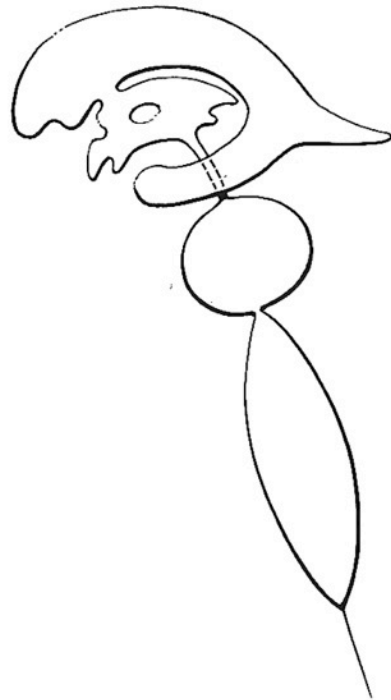
- Posterior fossa craniotomy with shunting of the outlets of the fourth ventricle to the spinal subarachnoid spaces
- Lumboperitoneal shunts
- Source of CSF: CSF derived from the ventricular system which enters the syrinx cavity via distended Virchow-Robin Spaces through the pia of the spinal cord

OLDFIELD RECOMMENDATIONS

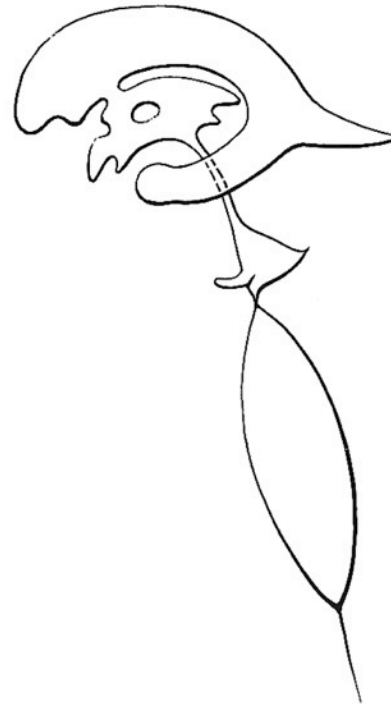
- Posterior fossa decompression
- Dural opening without violation of the arachnoid
- Source of CSF is from the spinal subarachnoid spaces through the pia via dilated Virchow-Robin spaces

HYDROMYELIC HYDROCEPHALUS

Oi Based on Rekate Model



Syringomyelia
Associated with
Isolated Fourth
Ventricle



Isolated Central
Canal Dilatation

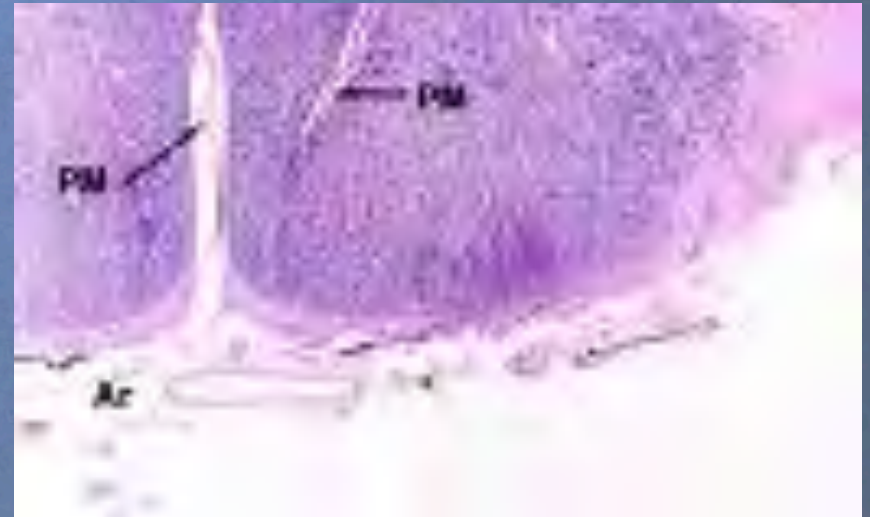


SYRINGOMYELIA AS HYDROCEPHALUS OF CORD

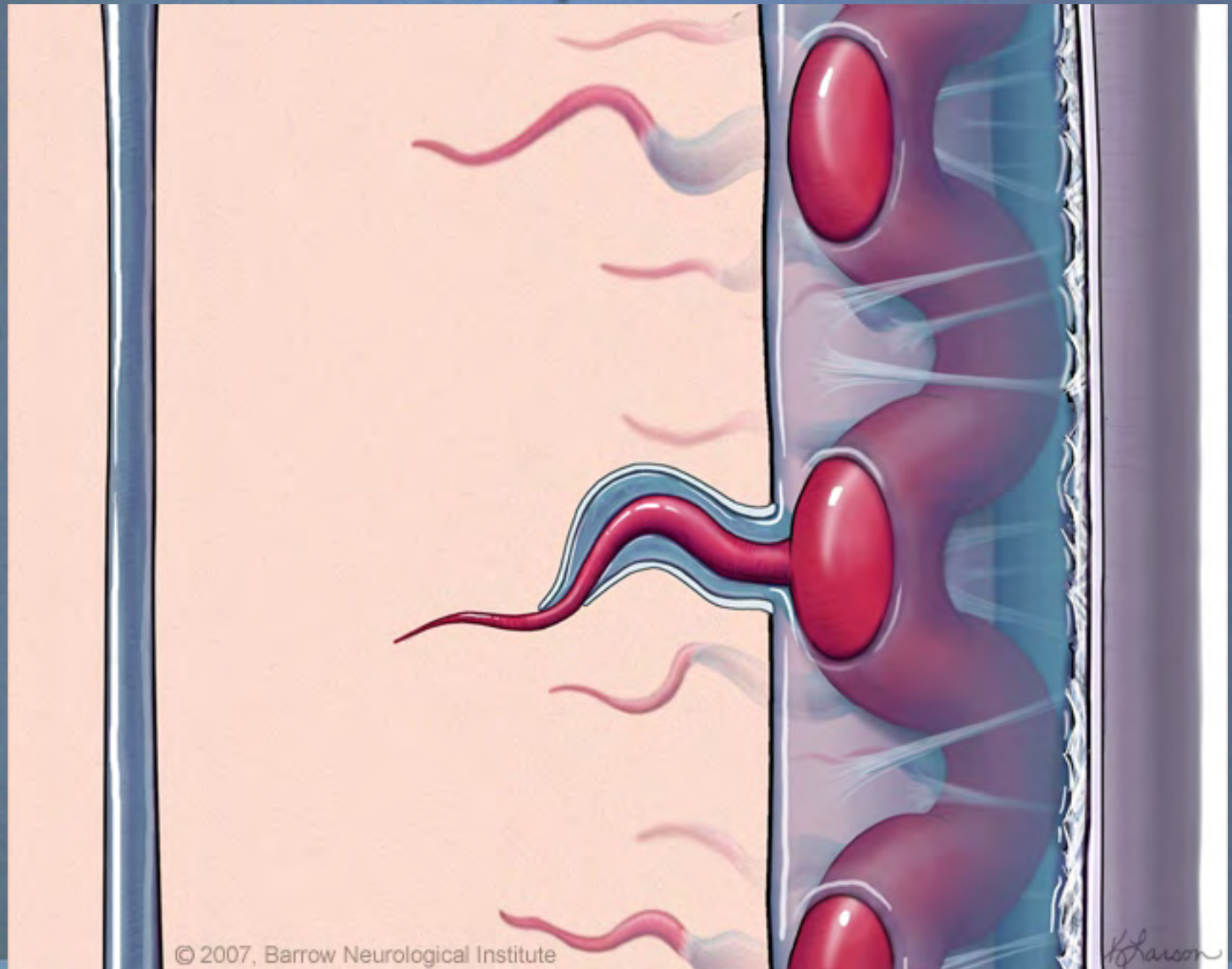
- Central canal is histologically a ventricle in infants
- The central canal of the Conus Medullaris is embryologically the “Ventriculus terminalis”
- While it does not contain choroid plexus, there is compelling evidence of the spinal cord as a source of CSF

The Pial Surface

- The Pia itself is impervious to flow of CSF
- The vascular supply of spinal cord enters through the pia creating the “Virchow-Robin Space
- VR space distends with increases in ICP
- CSF can enter spinal cord through VR spaces by pulsatile flow



Virchow Robin Spaces



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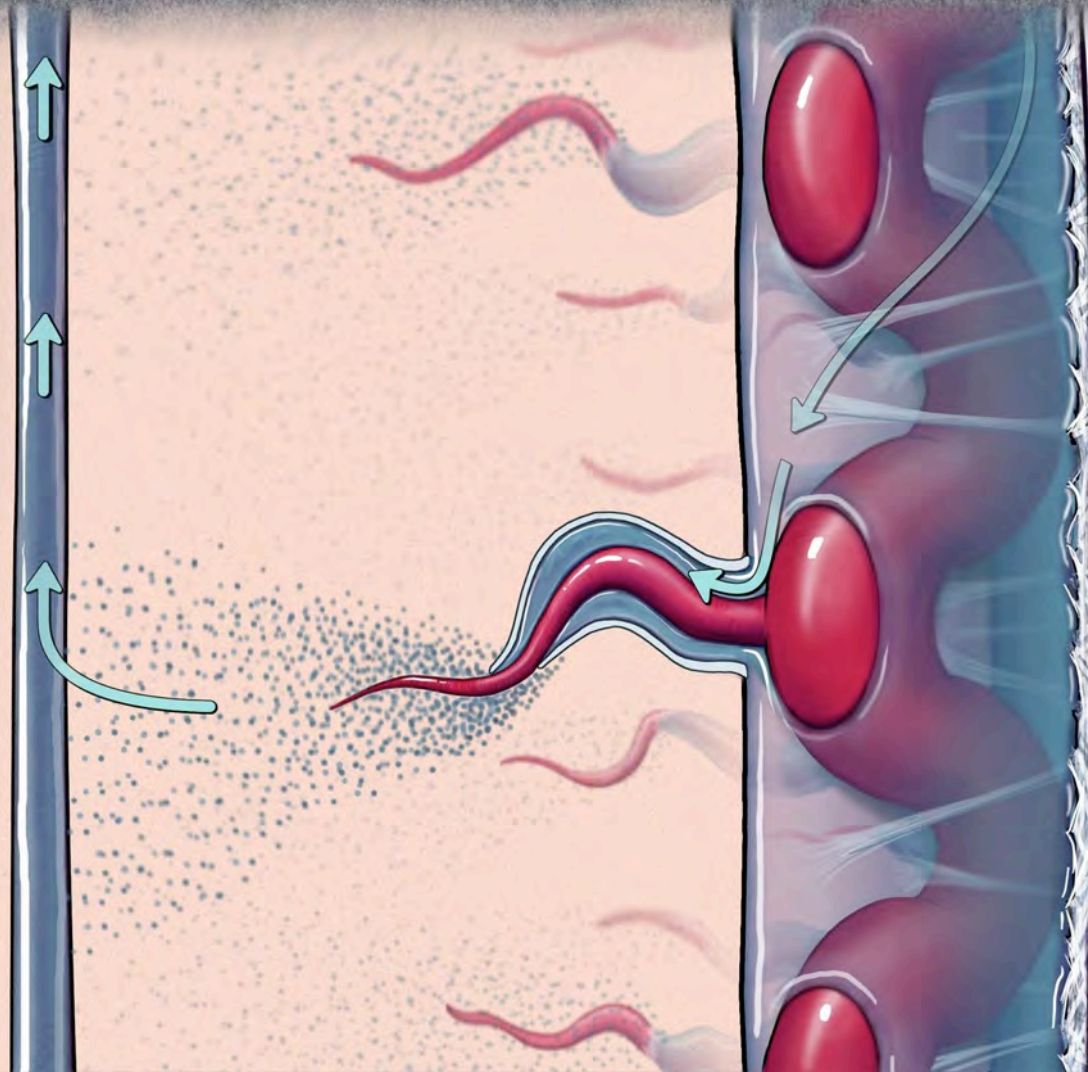
ShoreLIJ Institute

North Shore-LIJ Long Island Jewish Health System

SCHOOL of MEDICINE

Oldfield, Ball and Dayan

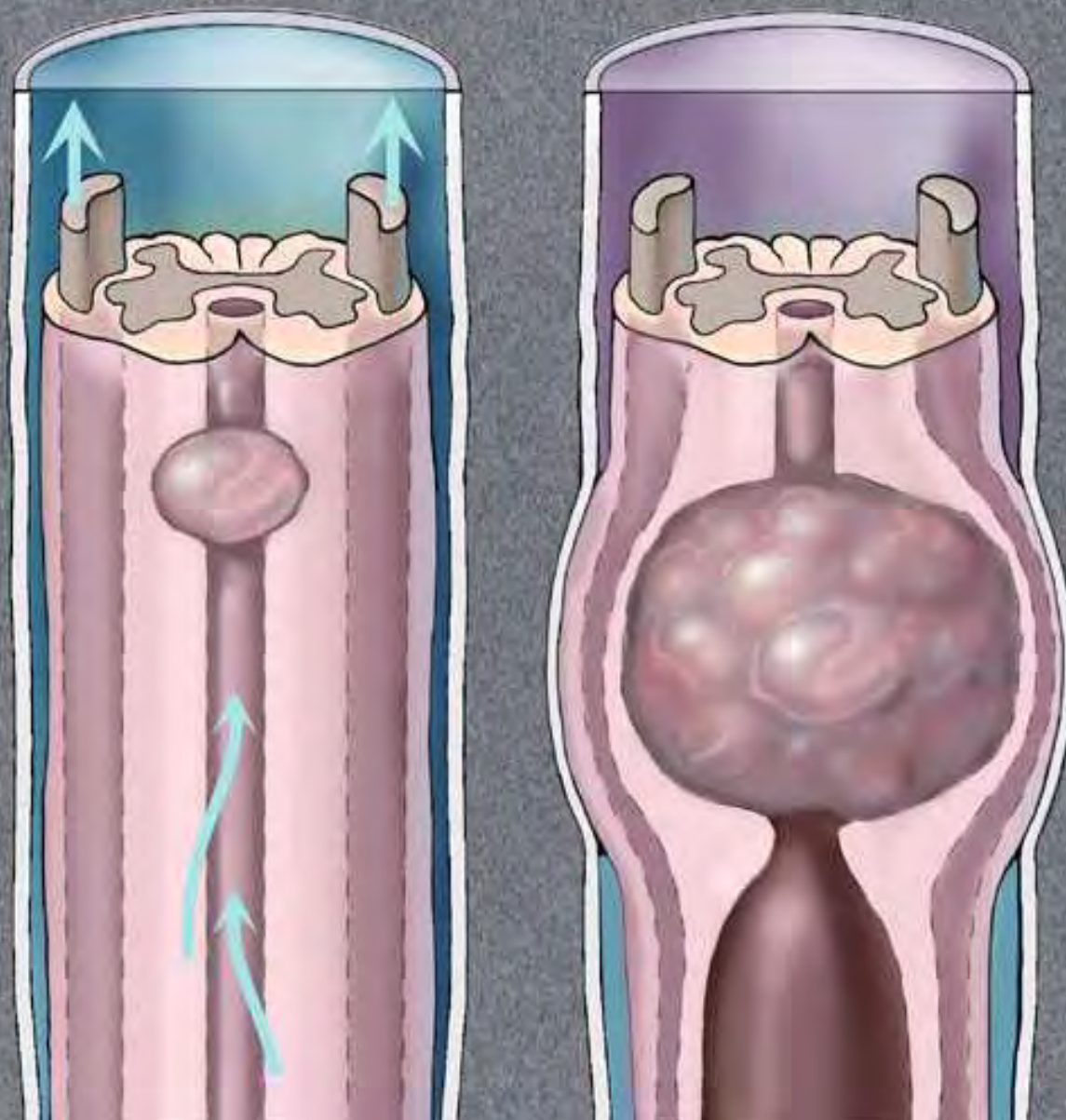
Virchow-Robbins Spaces



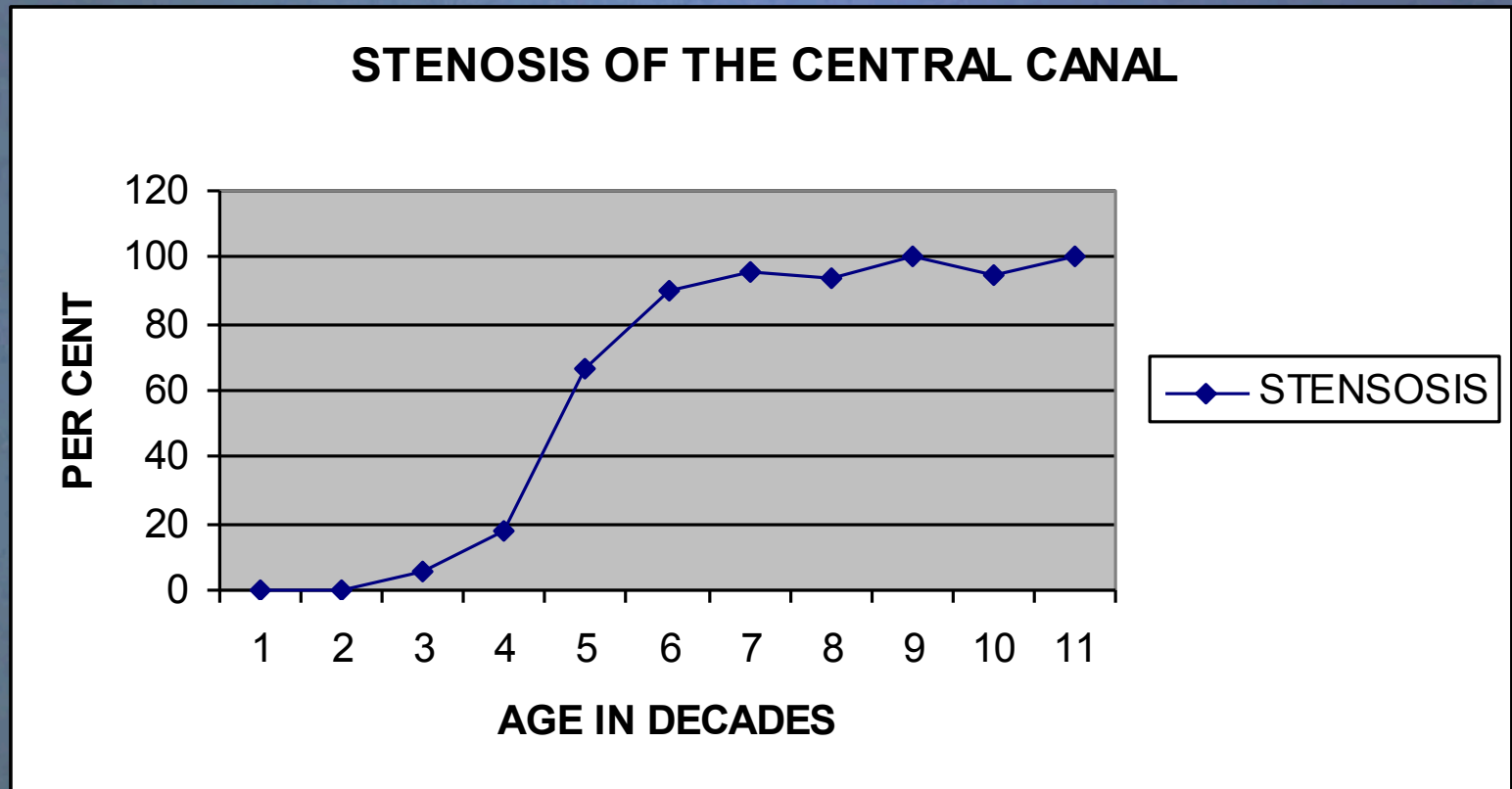
CSF PRODUCTION BY SPINAL CORD

- Estimates of CSF production as a byproduct of CNS metabolism range from 5-8% (Davson) to 30-50% (Milhorat)
- In brain CSF empties into ventricle by bulk flow. Same mechanism likely in cord
- A great deal of experimental evidence shows that CSF can pass through the ependyma but not out through the pia.
- It may be able to pass from CSAS to brain parenchyma
- Volume of daily CSF production
 - 2.0 to 3.2 cc/day (Davson)- 0.5-0.8% total
 - 16-20 cc/day (Milhorat) 4.0-5.0% total

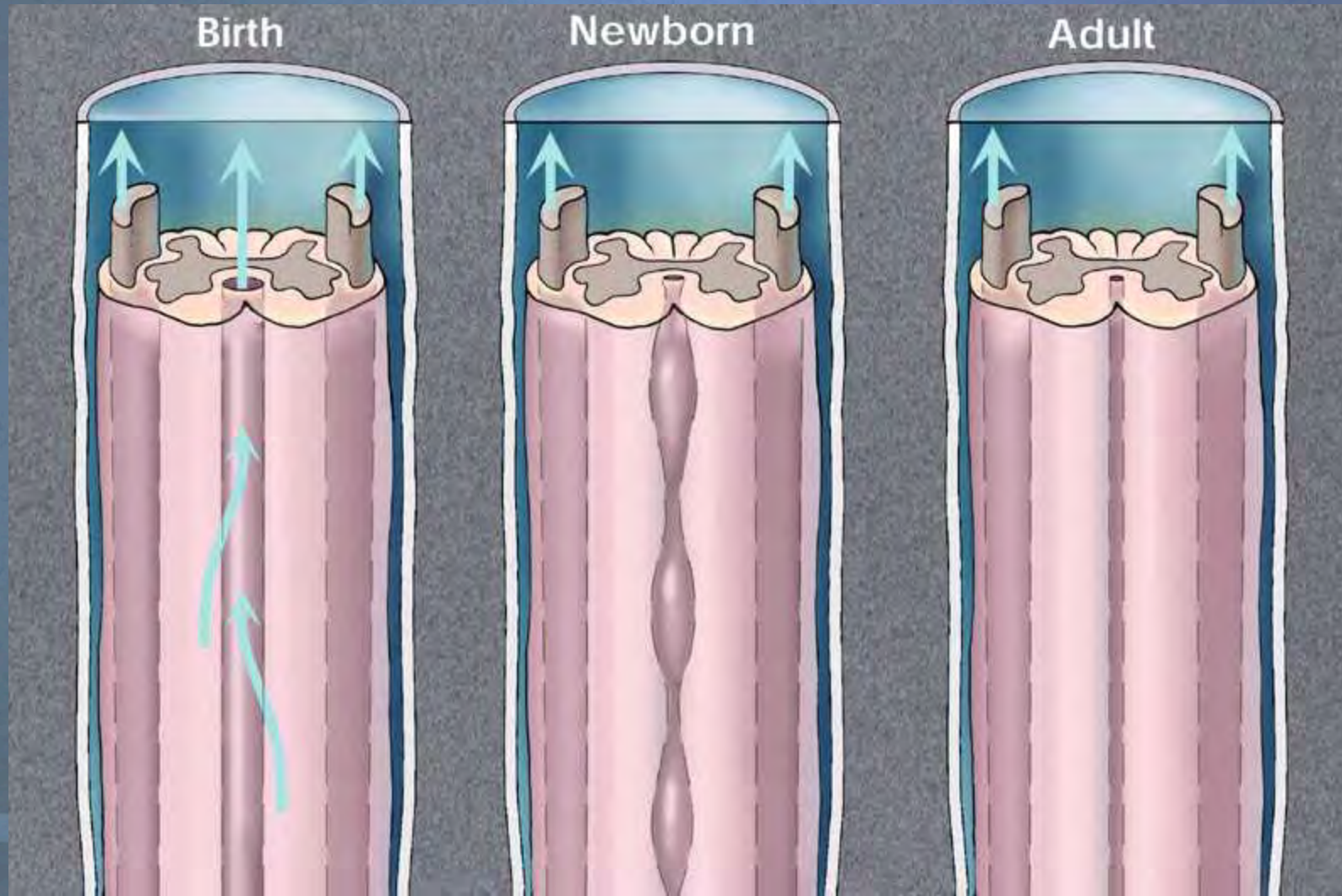
Newborn



PATENCY OF CENTRAL CANAL RELATIVE TO AGE

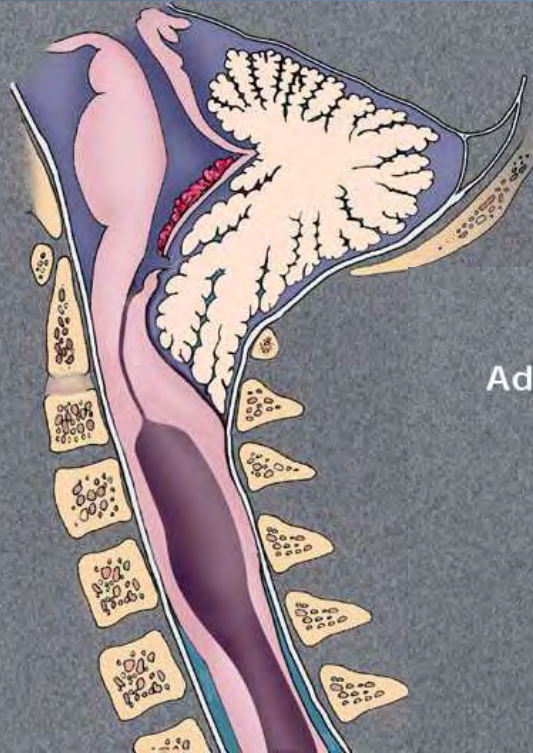


THE CENTRAL CANAL AS A VENTRICLE

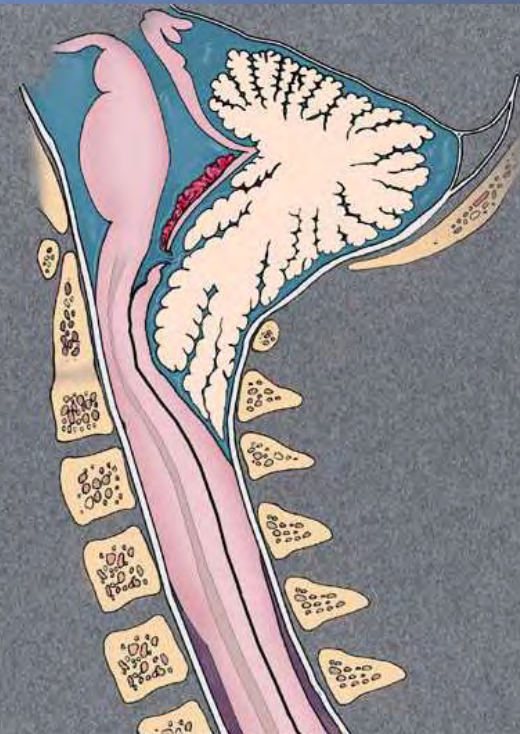


ADULT VS CHILDHOOD TONSILLAR HERNIATION

Newborn Chiari



Adult Chiari



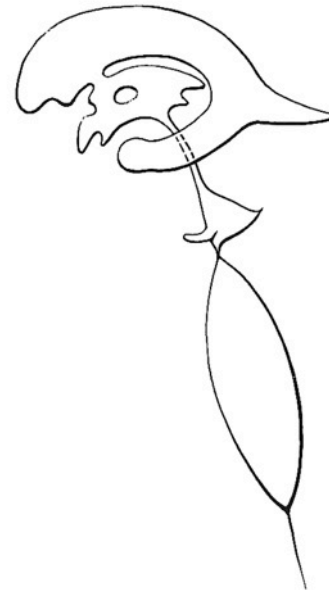
SOURCE OF CSF MAY BE VENTRICLE OR SPINAL CORD OR CORTICAL OR SPINAL SUBARACHNOID SPACE

Communicating syringomyelia



Syringomyelia
Associated with
Isolated Fourth
Ventricle

Non-communicating syringomyelia



Isolated Central
Canal Dilatation



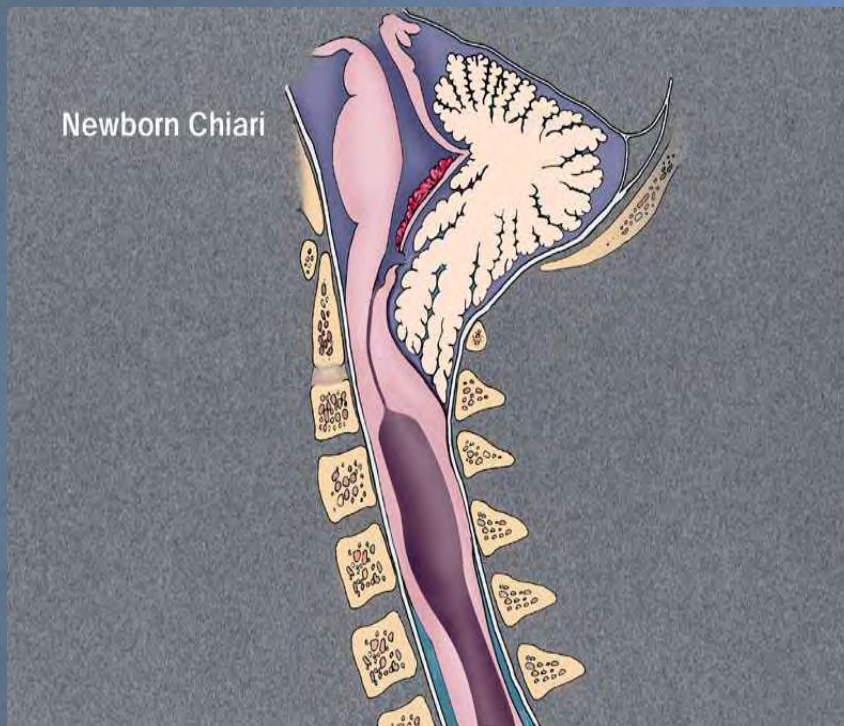
SYRINGOMYELIA

- Central canal is a fifth ventricle embryologically
- 20-50% of CSF is produced in white matter not requiring choroid plexus
- White matter CSF cleared by bulk flow through ependymal surface
- Blockage involves proximal flow in the central canal and white matter.

OPERATIONS BASED ON THE OI / REKATE HYPOTHESIS

- All of the operations described for the treatment of syringomyelia should work except those with effective plugging of the obex with a muscle plug
- Sources of CSF:
 - From ventricles with obstruction of outlets of fourth ventricle
 - From spinal cord with blockage of both central canal and white matter flow
 - From cortical subarachnoid space with Severe Chiari malformation and increased pressure in subarachnoid space

TREATMENTS ANALYZED

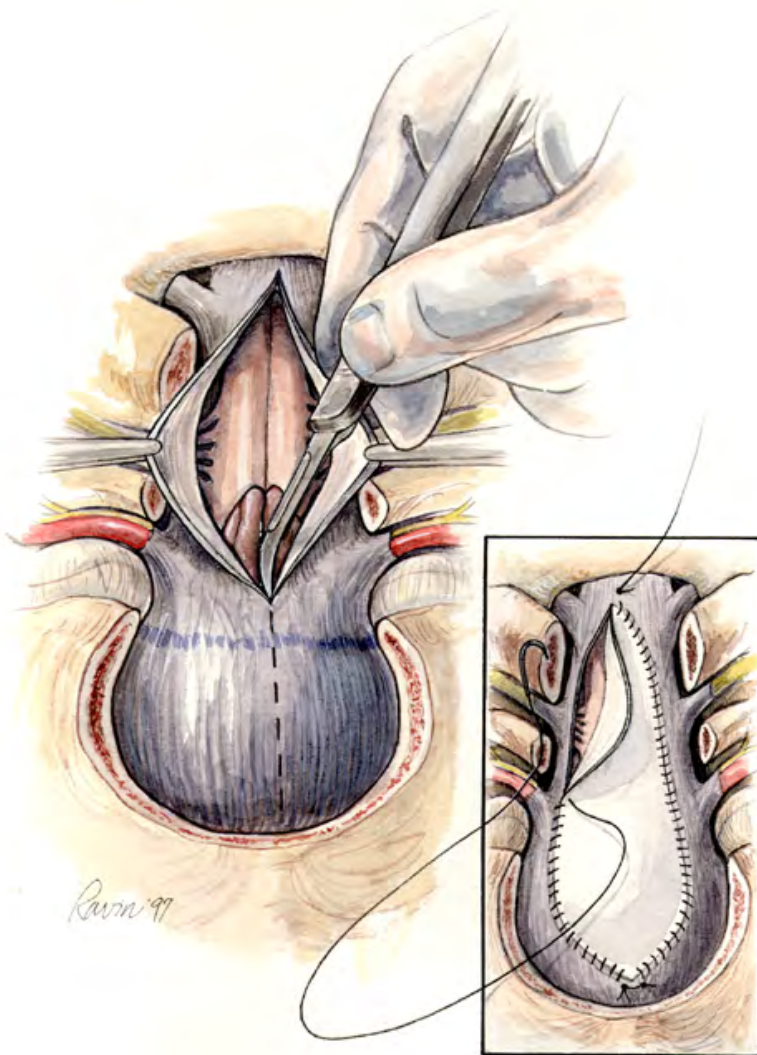


- Terminal ventriculostomy
- Posterior Fossa Decompression
- Syringosubarachnoid shunt (SS)
- Syringoperitoneal or pleural shunt (SP)

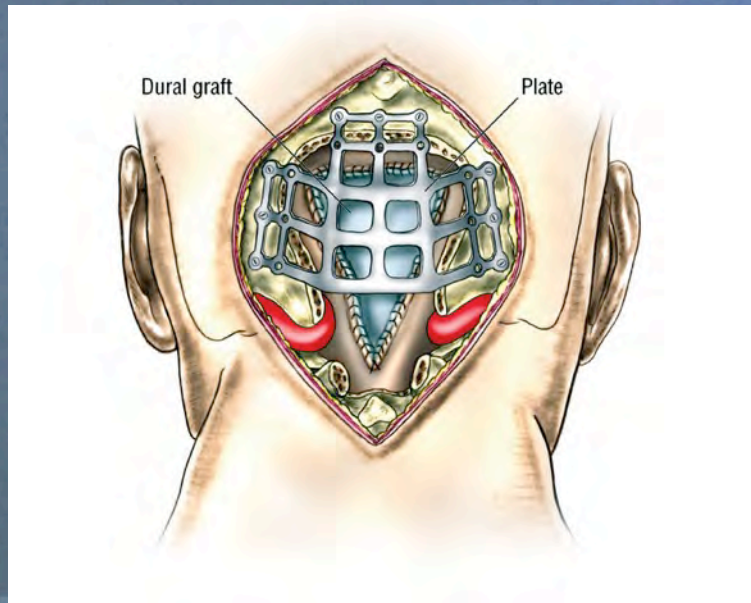
TERMINAL VENTRICULOSTOMY

- Two papers analyzed outcome of the treatment
- Gardner (1977): 10 of 12 patients improved (5 as primary procedure)
- Williams (1983): 3/31 objective improvement, 18/31 subjective improvement/ Only 5 sustained improvement
- MOSTLY ABANDONED

POSTERIOR FOSSA DECOMPRESSION



Posterior Fossa Reconstruction



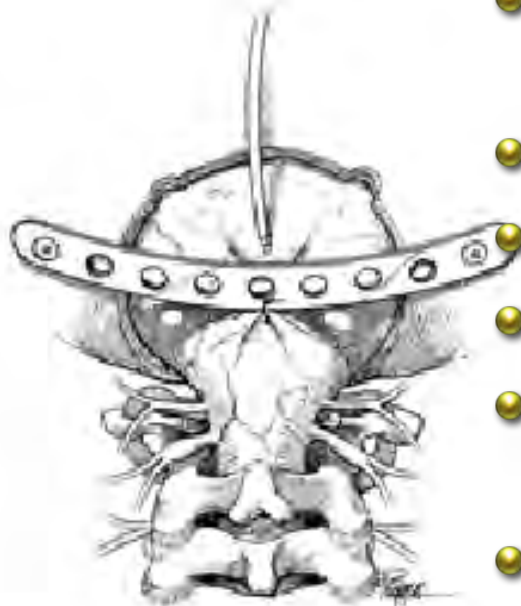
Use of the Chiari Plate

- The ultimate goal of Chiari decompression with or without syringomyelia is to create a new cisterna magna
- The Chiari plate protects the integrity of this new finding from mechanical distortion by lying on the back of the neck
- It holds the patch back against the plate to maintain the Cisterna magna

POSTERIOR FOSSA DECOMPRESSION

- Found to be **guideline** when associated with Chiari I malformation
- Found to be **option** when Chiari II or when there is no cerebellar tonsillar herniation
 - “Chiari 0 malformation
- Decompression of the syrinx may take weeks or months as in third ventriculostomy in hydrocephalus
- Response nearly always maintained.

ADJUNCTS TO PFD: ALL ARE OPTIONS



- Leaving dura open
- Dural patching with several substances
- Plugging of “Obex”
- “Shrinking” of the tonsils
- Stent of the fourth ventricle
- Shunting the syrinx at the area of maximum distension
- Posterior fossa reconstruction

id #177

Rekate HL, Nadkarni T, Wallace D. Severe intracranial hypertension in slit ventricle syndrome managed using a cisterna magna-ventricle-peritoneum shunt. *J Neurosurg (Pediatrics)*. 2006 Apr;104(4 Suppl):240-4.

PLUGGING THE OBEX

- Dural opening and dissection of the arachnoid into the fourth ventricle necessary to plug
- Assumes all CSF is derived from the ventricles
- If effective would block the CSF produced in Spinal Cord
- High Complication rate

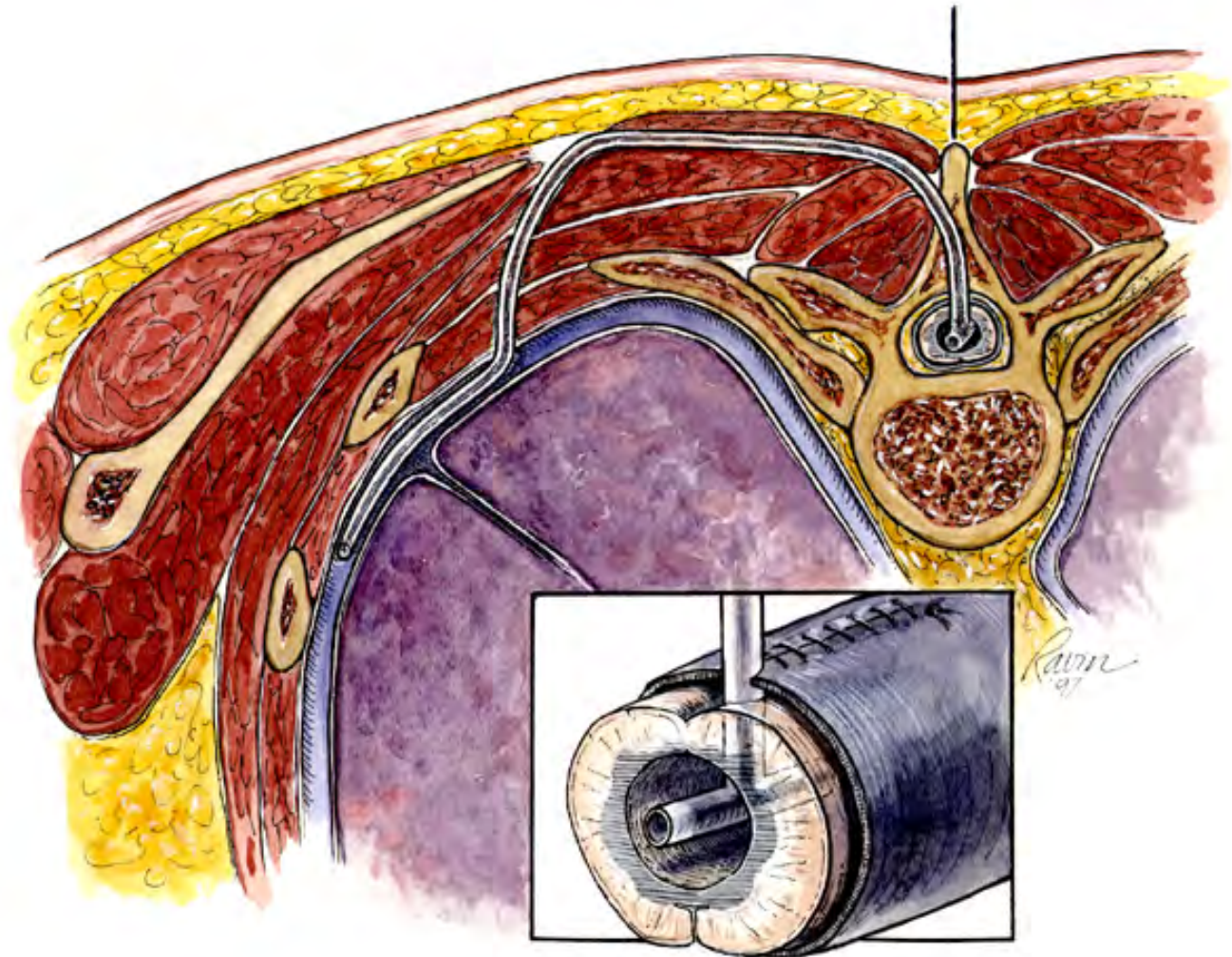
STENTING OF THE FORAMEN OF MAGENDIE

- Leads to increase speed of syringeal decompression
- High failure rate
- Probably not necessary except in the case of complicating arachnoiditis
- May lead to tethering of the brainstem

WHAT TO DO WITH THE DURA

- No clearcut evidence that any mechanism is superior to any other
- Some recurrences due to pseudomeningocele Helped by reconstruction
- Individual case reports of resolution of syrinx with purely extradural operations

SYRINX SHUNTING



WHAT TYPE OF SHUNT?

- SS shunt assumes adequate drainage from subarachnoid space
- SP shunt is to a lower pressure system and works faster and more reliably
- Three articles about postoperative tethering
- Long term success in <50%

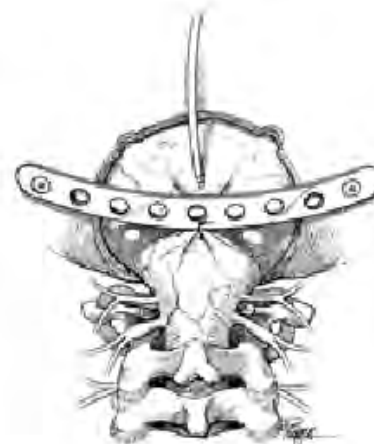
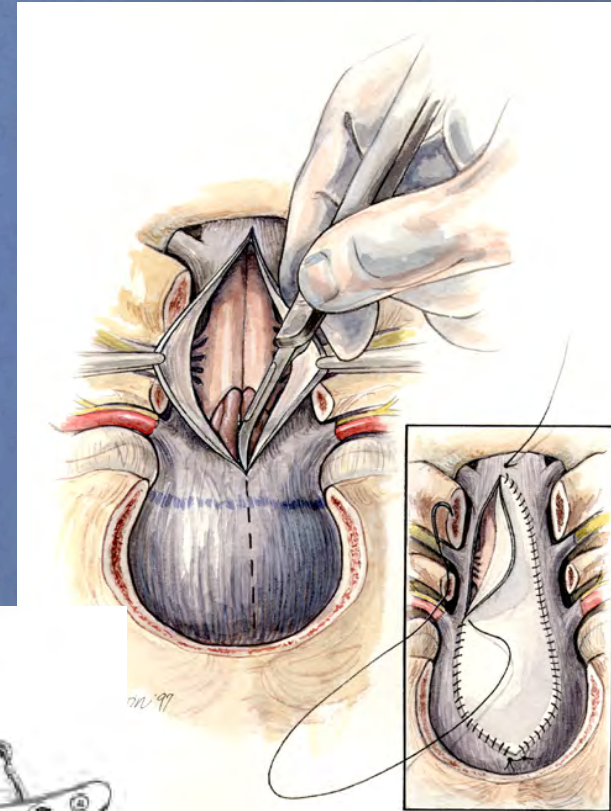
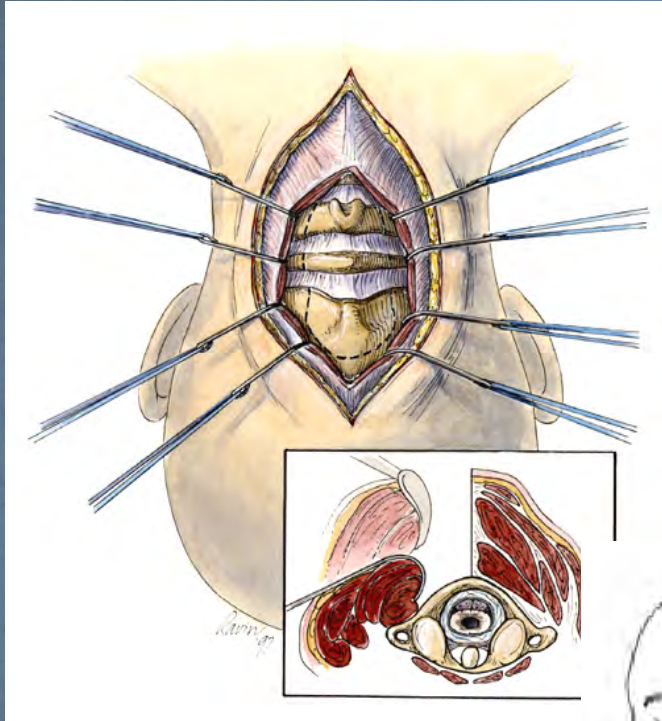
COMPLICATIONS OF SYRINX SHUNTS

- Infection: Rarely reported but reported at essentially the same rate as for ventricular shunts
- Low pressure states
- Increased pain and dysesthesia
- Symptomatic tethering in about 10%
- 50% long term patency

Chiari Zero

- Patients with syrinx but without tonsillar herniation
- Based on information of syrinx in association with traumatic delivery and arachnoiditis
- PFD with opening of the foramen of Magendie works even without Chiari

BE REASONABLE: DO IT MY WAY



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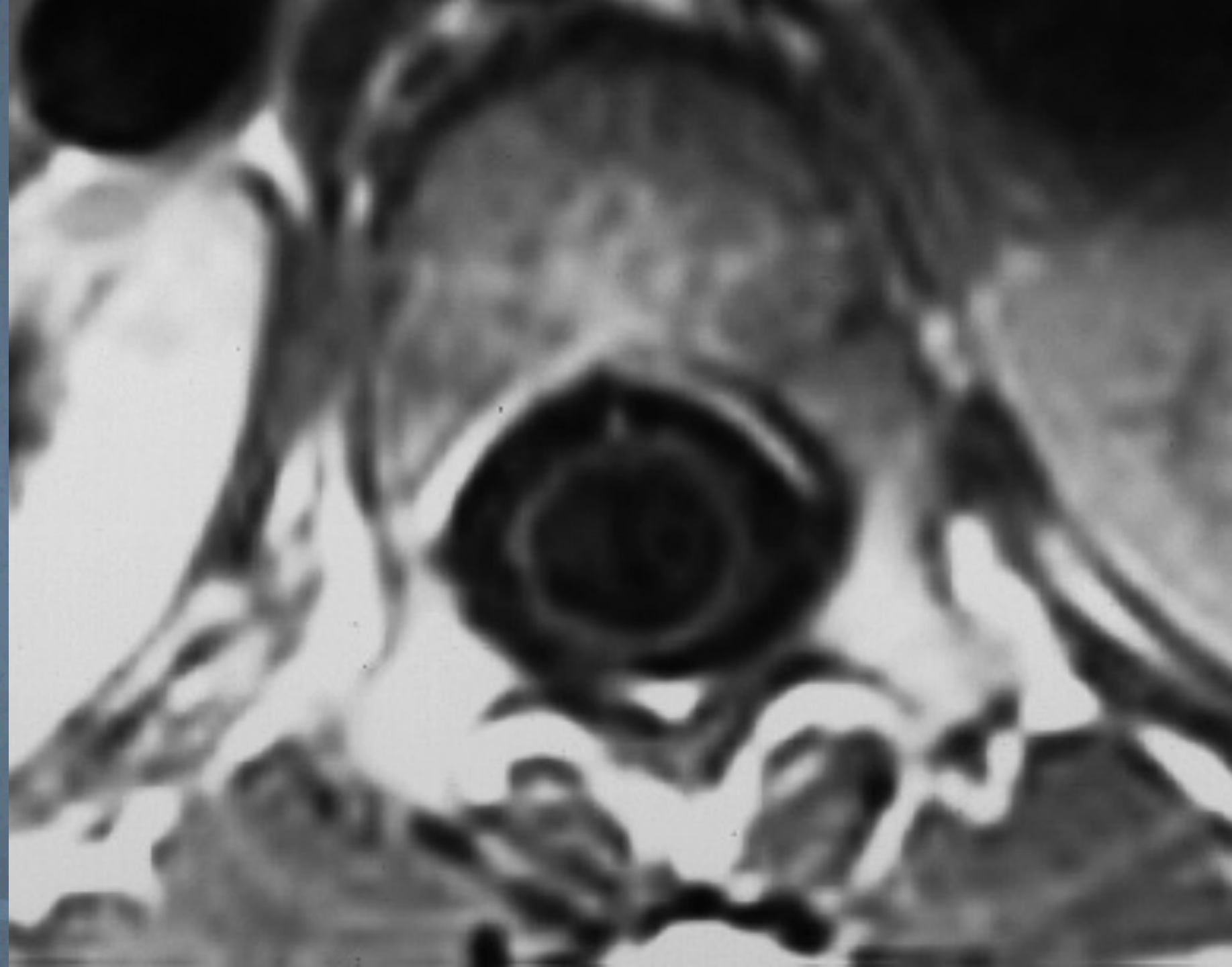
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HOSPITAL OF LONG ISLAND
SCHOOL of MEDICINE

MY SUGGESTIONS

- Just as in hydrocephalus, decisions as to risks and benefits need to be individualized
- In syrinx due to simple Chiari I (not associated with other complications), PFD alone. Open foramen of Magendie
- Patch is patulous
- Prefer pericranium
- Reserve shunting for cases in which foramen magnum cannot be approached



Conclusions

- Syringomyelia like hydrocephalus is always an “obstructive process”
- Syringomyelia requires that the central canal is open and distensible
- The pia is a one way system not allowing CSF to flow to the spinal subarachnoid space except under high pressures and then through the Virchow-Robin Space
- No flow of CSF from spinal cord to CSAS
- The ependyma likewise is a one way system which allows flow into the central canal but not back again.
- Treatment options are similar to those of hydrocephalus: Always look for a direct way to manage the obstruction.

Conclusions: Treatment

- Operations that permit flow from the spinal cord to the fourth ventricle are preferred
- Shunts to the subarachnoid space are rarely effective
- Shunts to the subarachnoid space often complicated by spinal cord tethering
- Shunts to the pleura or peritoneum are effective but may lead to low intracranial pressure.