CSF hydrodynamics in craniocervical junction anomalies

Craniocervical junction anomalies

- Congenital
  - Isolated disorders
    - Platybasia
    - Basilar invagination
    - Atlas assimilation
    - Atlas hypoplasia
    - Klippel-Feil malformation
    - Chiari malformations
    - Os odontoideum
  - General disorders
    - Achondroplasia
    - Osteogenesis imperfecta
- Acquired
  - Isolated disorder
    - Traumatic dislocation
      - Type II odontoid fx
  - General disorders
    - Rheumatoid arthritis
      - Atlantoaxial dislocation
    - Paget disease of bone
    - Acromegaly

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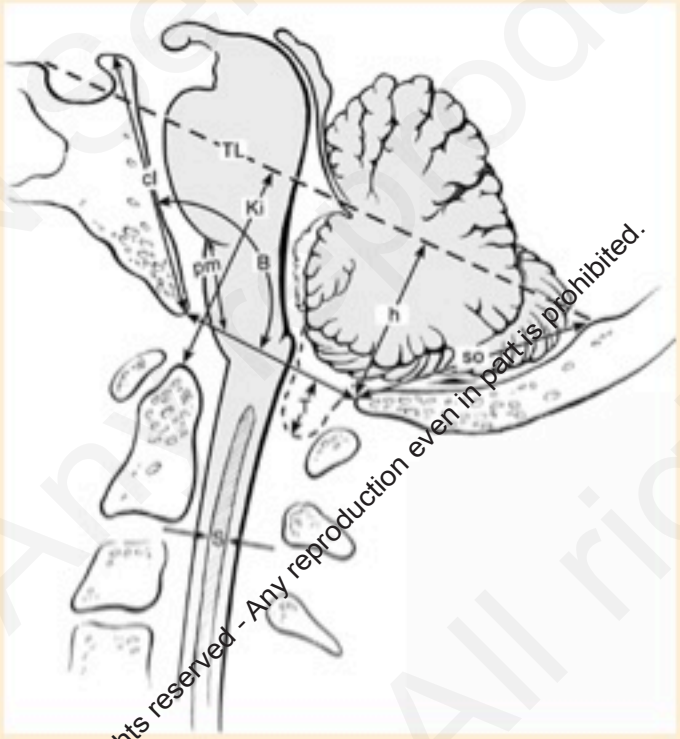
Craniocervical junction anomalies

- Symptoms
  - Non-neurological
    - Neck pain
    - Occipital headache
      - Worse with coughing and Valsalva
  - Neurological
    - Neural compression
      - Brainstem
        - » Impaired swallowing, central sleep apnea
      - Cranial nerves
      - Cerebellum
      - Medulla
    - CSF pathway compression
      - Spinal cord: Syringomyelia
    - Vascular compression



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Hydrodynamics

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CSF hydrodynamics in craniocervical junction anomalies

- Hydrodynamics
  - A branch of physics that deals with the motion of fluids and the forces acting on solid bodies immersed in fluids and in motion relative to them
  - △Pressure proportional to △ Flow (Hagen-Poiseuille)
  - △Pressure between Intracranial and cervical subarachnoid space drives CSF flow (Q) across the foramen magnum
  - Reduced radius of flow pathway drastically reduces CSF flow
- Hydraulics
  - The use of a liquid flowing under pressure to transmit power from one location to another
- Hydrostatics
  - A branch of physics that deals with the characteristics of fluids at rest and especially with the pressure in a fluid or exerted by a fluid on an immersed body:
  - △Pressure without △ Flow:
  - Fluid is pressurized: CSF pressure
  - Force and energy transmitted by pressure

$$\Delta P = \frac{8mLQ}{\pi r^4}$$

Merriam-Webster.com

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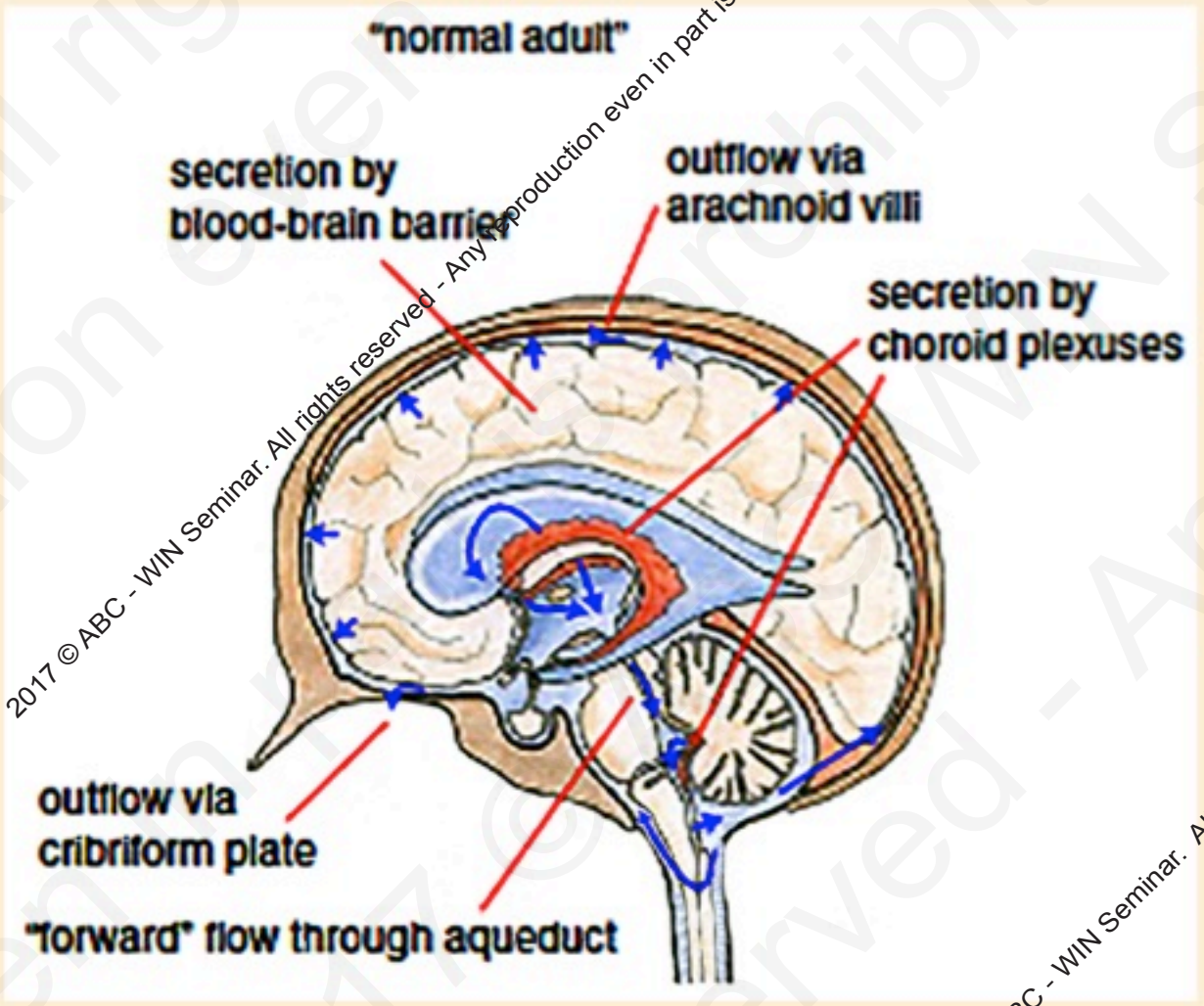
Intracranial CSF Circulation: Hydrodynamic



- Slow one-way CSF flow of 20 ml/h from choroid plexus to arachnoid villi
- CSF replaced every 8 h
- Small pressure differential drives flow between ventricles and arachnoid villi
- Blockage of flow/ absorption pathway introduces hydrostatic/ hydraulic pressure effects on ventricular and brain surface, and hydrocephalus

Figure from Strazielle N. Mol Pharm 2013; 10:1473-1491

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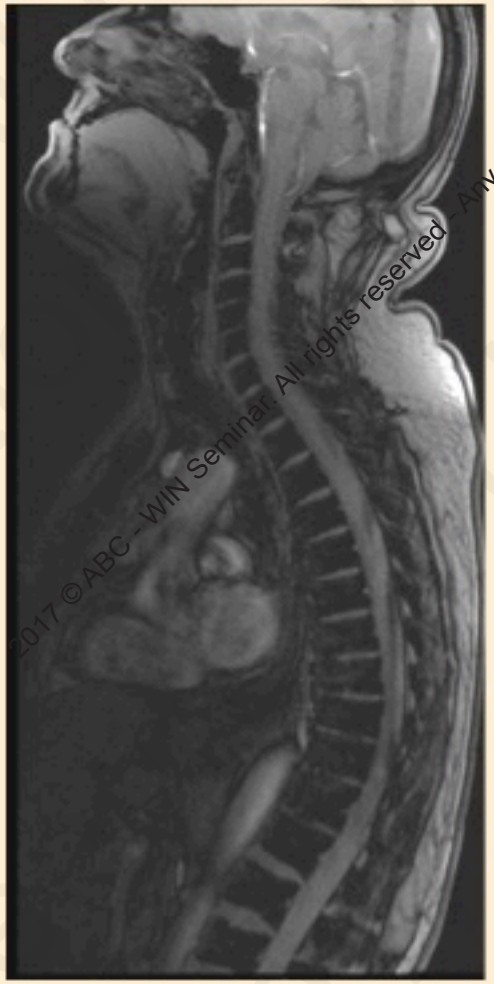
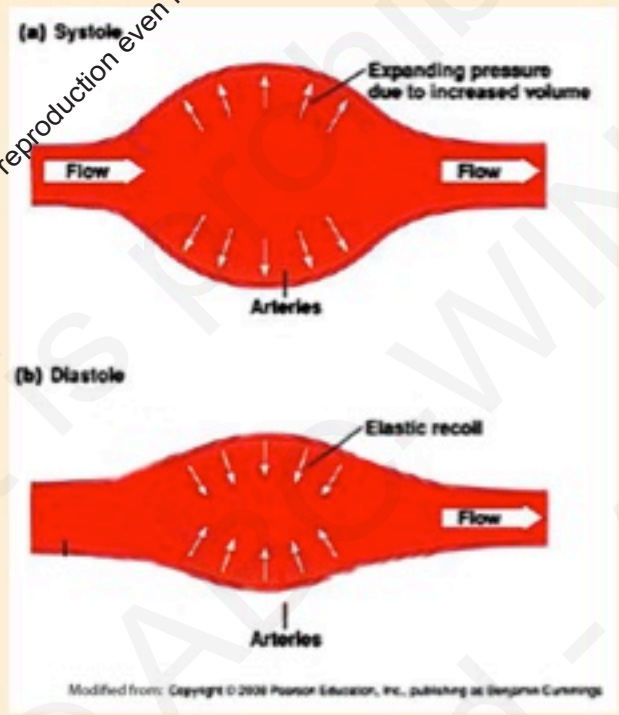
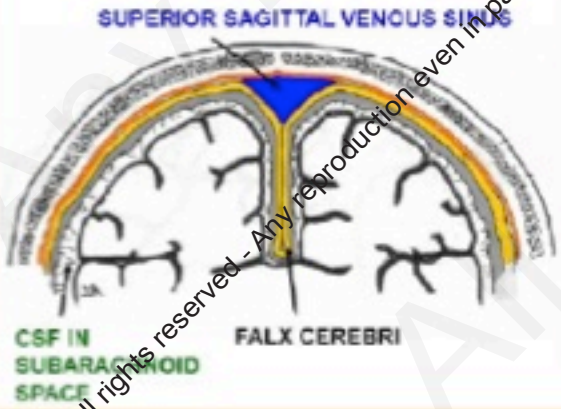
Cardiac Systole: Intracranial Pressure Pulse Dampened by CSF Outflow from Cranium to Spine





Intracranial Cavity
Skull,
Dura and Endoperiosteum Fused
except at Dural Sinuses (No Epidural Space!)
Subarachnoid Space,
Brain

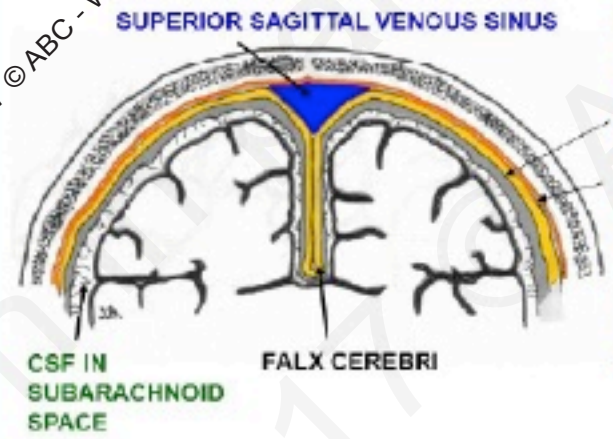


Modified from Strazielle N. Mol Pharm 2013; 10:1473-1491

Monro-Kellie doctrine

Fixed Volume Intracranial Cavity: Non-Compliant
Vs.
Expandable Volume Spinal Dura: Compliant

Intracranial Cavity
Skull,
Dura and Endoperiosteum Fused
except at Dural Sinuses (No Epidural Space!),
Subarachnoid Space,
Brain



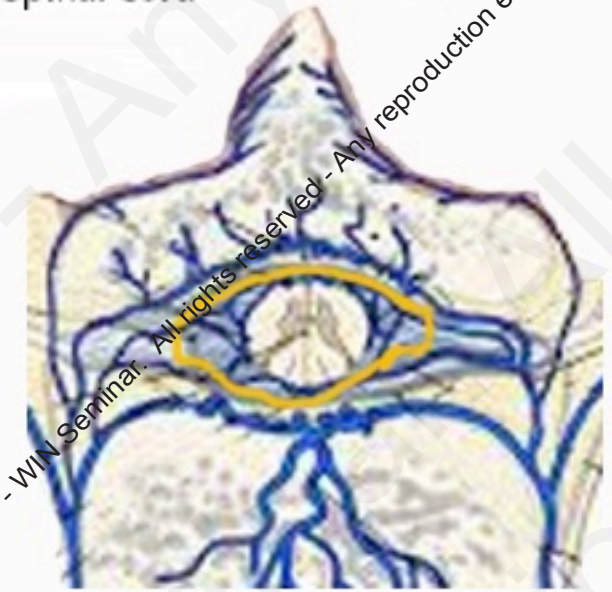
DURA MATER - tough connective tissue layer, composed of two layers -

1) INNER MEMBRANE LAYER (true dura)

2) OUTER ENDOSTEAL LAYER - periosteum on inner side of calvarium

Two layers - fused in most places - separate to form DURAL REFLECTIONS

Spinal Canal
Vertebral Components, Ligamentum Flavum,
Dura and Endoperiosteum Separate,
Epidural Space with Fat and Veins,
Subarachnoid Space,
Spinal Cord



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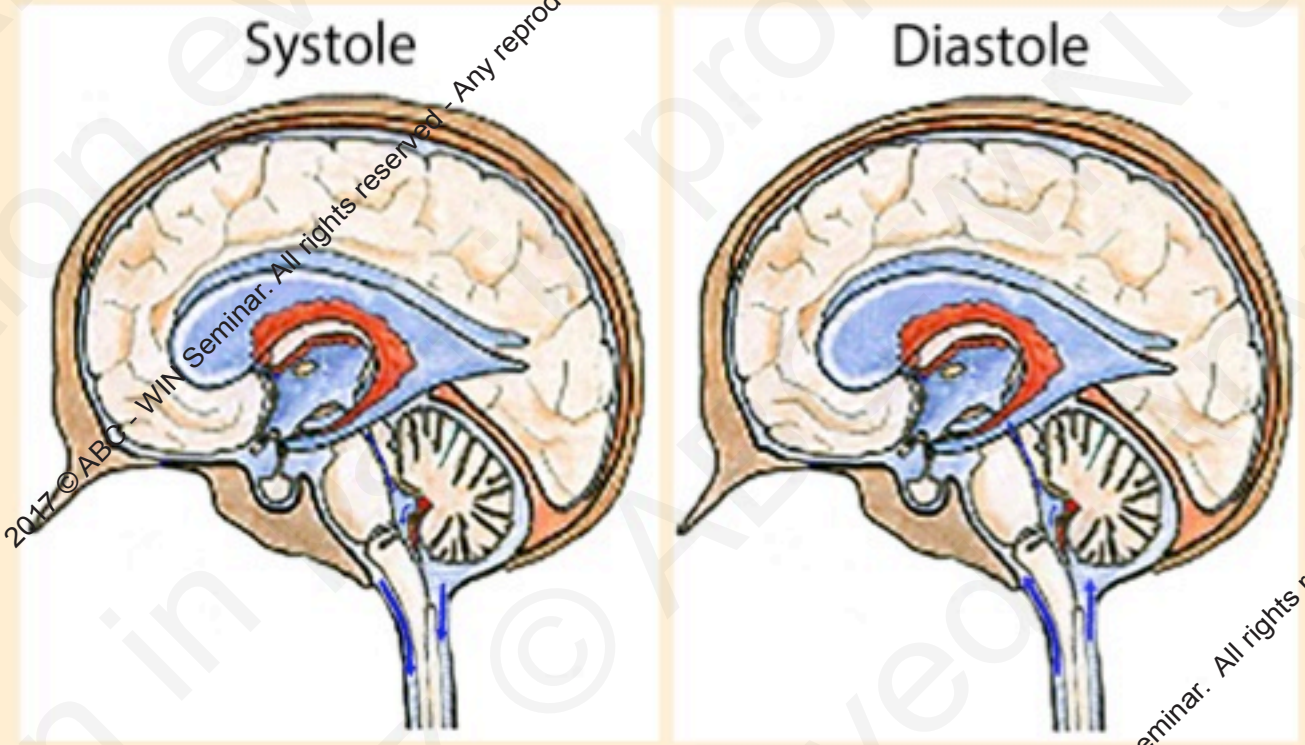
Cardiac Systole: Intracranial Pulse Pressure Dampened by CSF leaving the Intracranial Cavity



- Rapid superior-inferior CSF flow across the foramen magnum during the cardiac cycle
- Brain pulsation driven
- Blockage of flow pathway at FM leads to increased brain pulsation, hydraulic effects on the spinal cord, & syringomyelia

Craniocervical Junction Anomalies and their Relation to Syringomyelia

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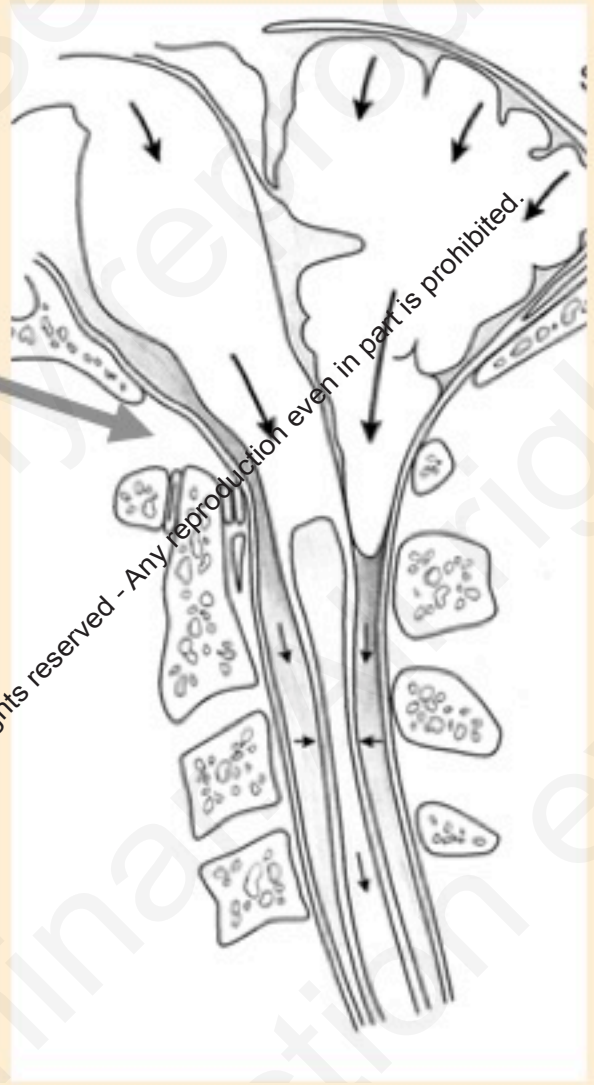
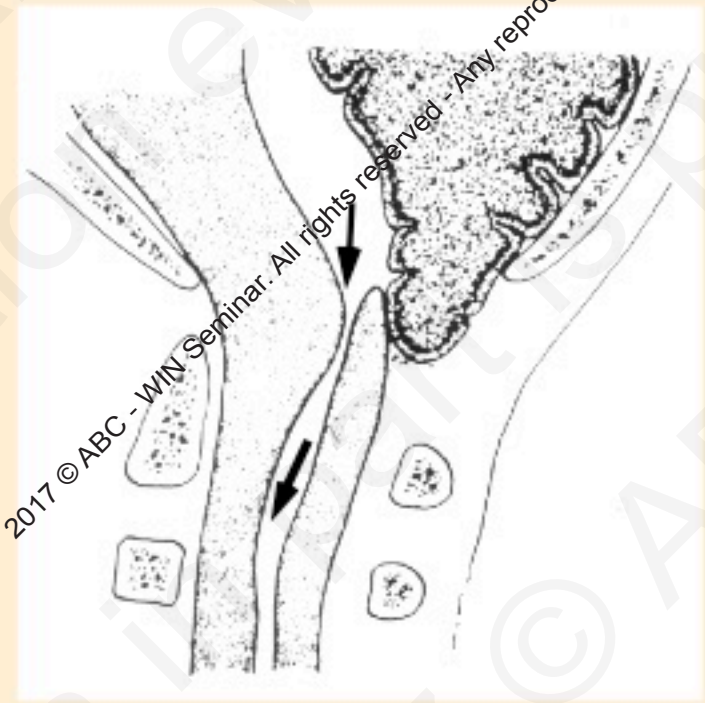
Syringomyelia Develops from an Underlying Disease

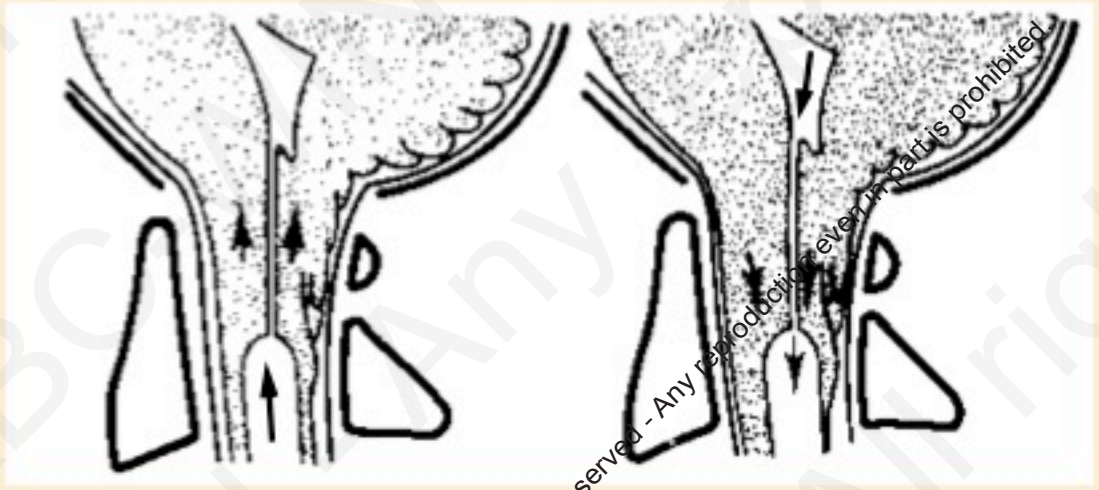
- Structure Compressing the Subarachnoid Space at the Foramen Magnum
  - Chiari I Malformation 70%
  - Basilar Invagination 10%
- Lesion Within the Subarachnoid space 12%
  - Arachnoiditis, Spinal Deformity
- Lesion of the Spinal Cord 4%
  - Tumors, Inflammatory Myelitis



Classic Theories of Syringomyelia Formation



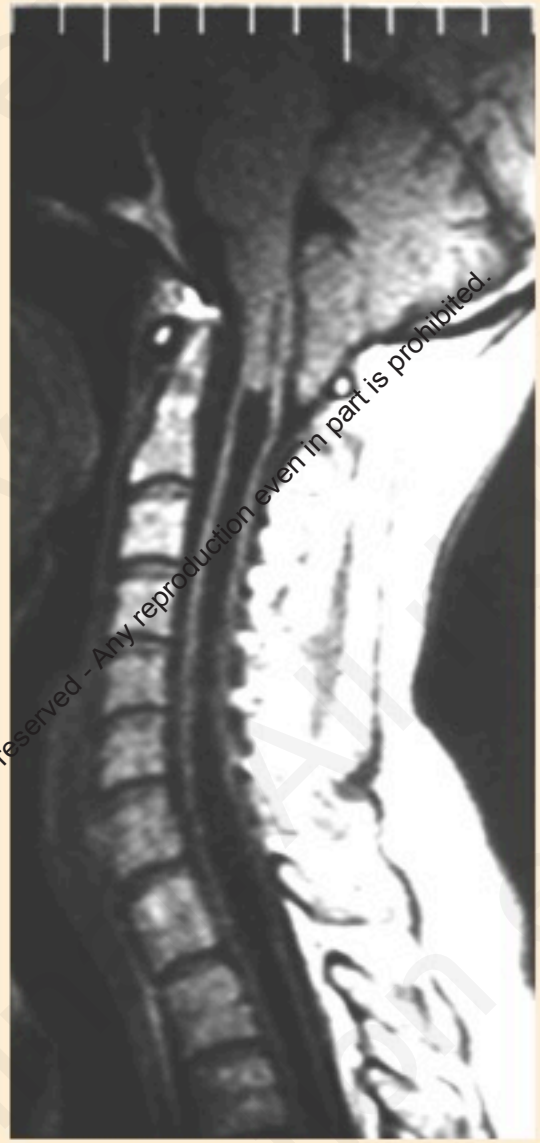
Gardner's Hydrodynamic



Williams' Craniospinal Dissociation

Chiari I-Syringomyelia Observations

- Smaller than normal posterior fossa
- Absent cisterna magna
- Ectopic tonsils narrow CSF passages at the foramen magnum
- Central canal is rarely (5% of adults) patent between 4th ventricle and syrinx
- Syrinx fluid is chemically identical to CSF
- Myelogram dye enters the syrinx (delayed)



Observation: Most syringes resolve following craniocervical decompression and duraplasty

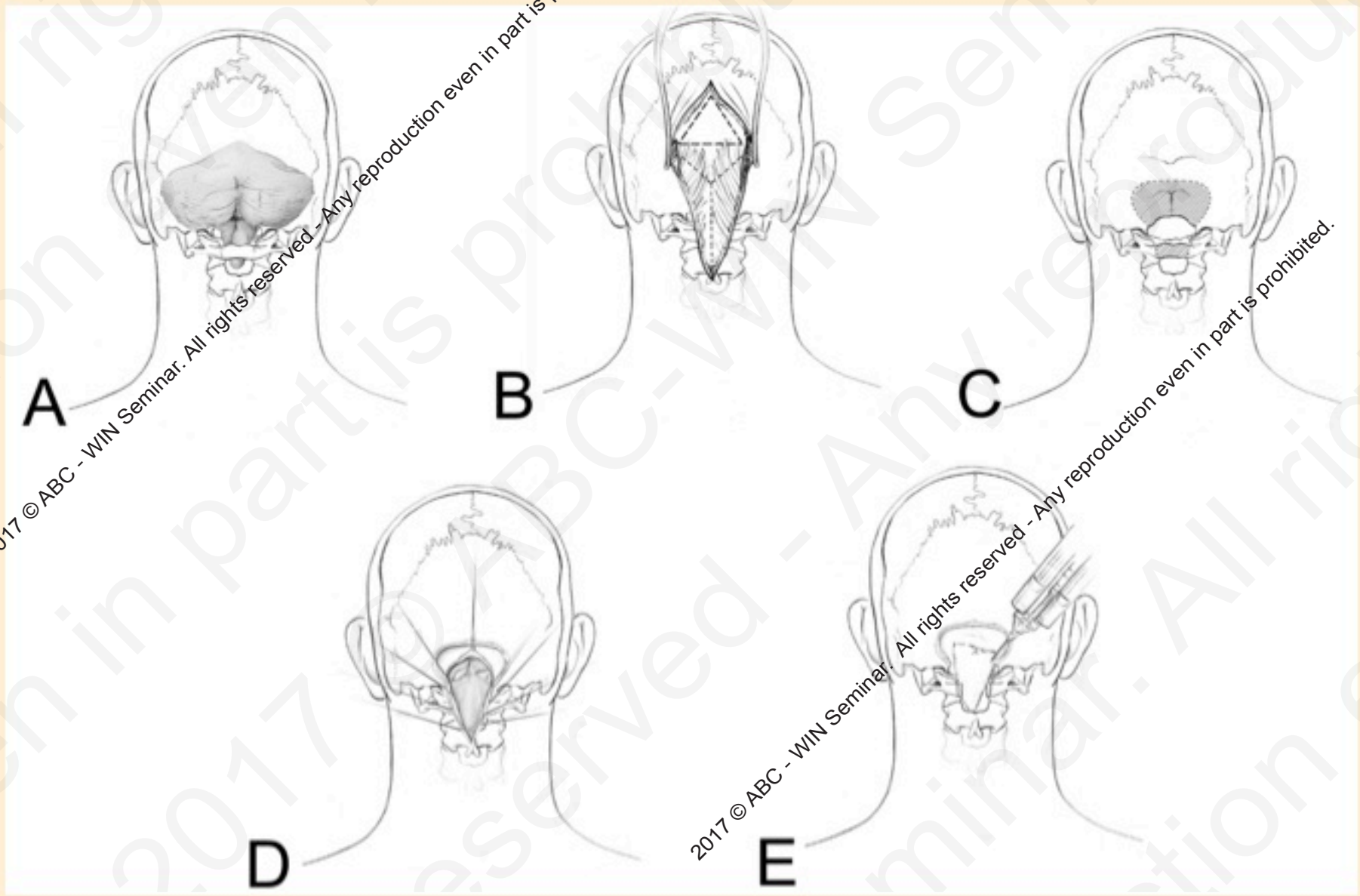
Series	Year	Decompression	Decompression Plus Shunt	Syrinx Shunt Alone	Yrs. Follow-Up
Bidzinski	1988	22/28	N/A	2/4	6.8
Hida	1995	30/33	N/A	37/37	5
Klekamp/Batzdorf	1996	72/88 (82%)**	N/A	5/22	3.2
Logue/Edwards	1981	42/51 (82%)*	4/7	N/A	3-7
Matsumoto/Symon	1989	35/60	16/28	N/A	5.9
Sgouros/Williams	1995	203/242 (83%)**	N/A	0/3	5
Van Calenbergh	1990	4/7	N/A	9/16	1.1-4
Vaquero	1990	10/15	N/A	12/15	1-5
Total		418/524 (80%)	20/35 (57%)	65/97 (67%)	1-6.8

Ratios represent number of stable or improved patients / total number of patients

* Extra-Arachnoidal

** Intra-Arachnoidal

Craniocervical Decompression



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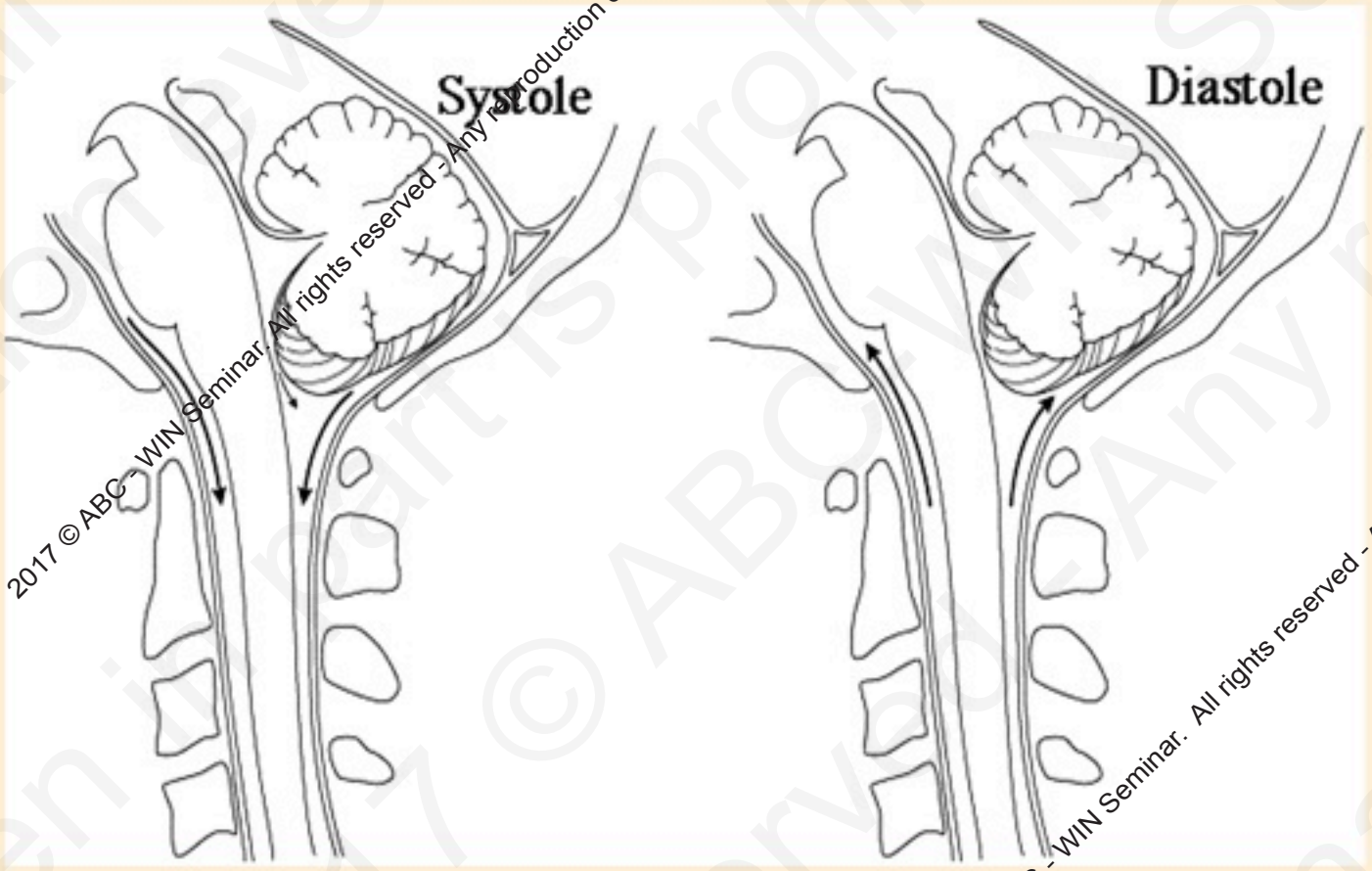
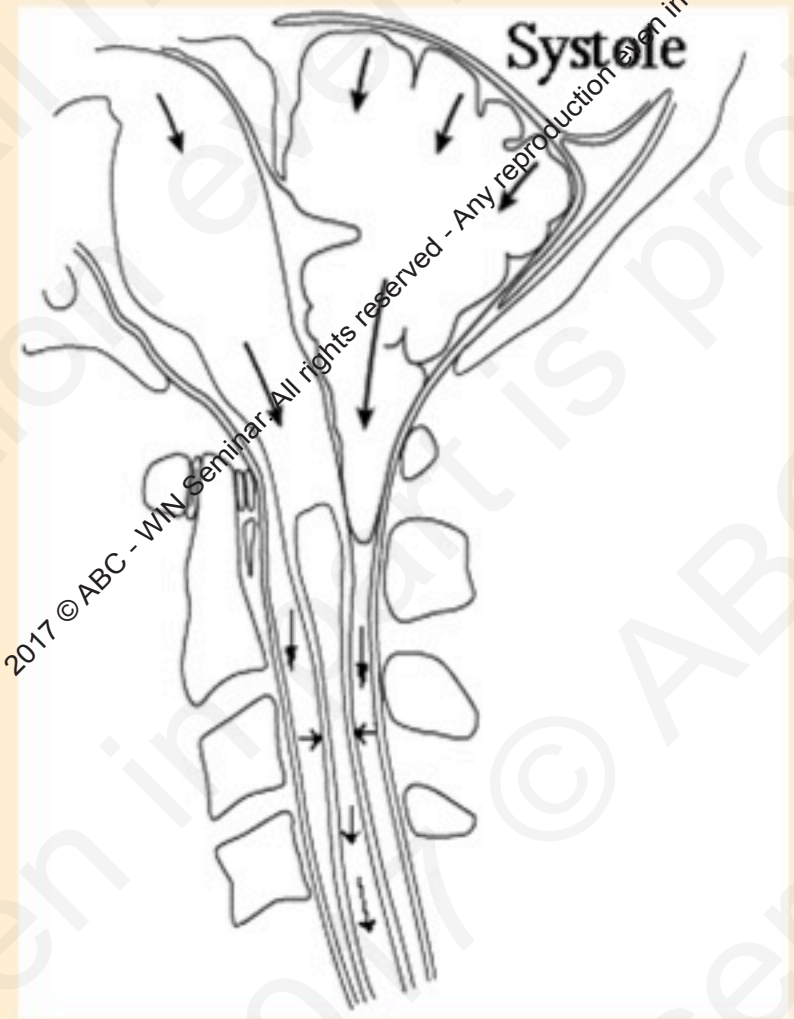
Clinical Study to Test a Theory of the Anatomic and Physiologic Effects of Chiari I Malformation on Development of Syringomyelia and of Craniocervical Decompression on Resolution of Syringomyelia

CSF Normally Moves Across Foramen Magnum During Systole to Compensate for Increase in Brain Volume



- Rapid superior-inferior CSF flow across the foramen magnum during the cardiac cycle
- Brain pulsation driven

Oldfield Theory of Chiari I & Syringomyelia



Small posterior fossa creates Chiari I → Chiari I narrows the subarachnoid space at the foramen magnum preventing normal CSF flow

↓ Compliance (pressure response to a change in volume)

Brain expansion every heart beat is not offset by CSF efflux

↑ Tonsillar pulsation

↑ Cervical subarachnoid pulse pressure

CSF moves into spinal cord

CSF coalesces into a syrinx & pulsates

Syrinx extends and progresses

Clinical Trial

- Groups Studied
  - Chiari I and Syringomyelia 20
  - Normal Subjects 18

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Testing the Theory

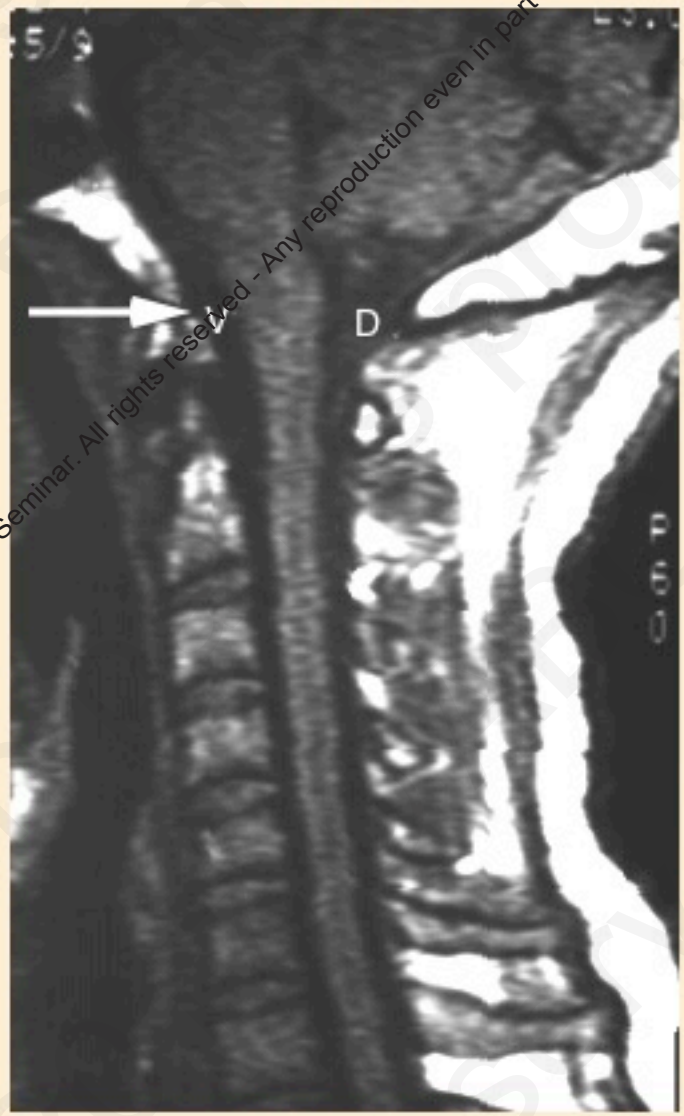
- **1. Partial Obstruction at the Foramen Magnum**
  - Anatomic MRI
- **2. Block of CSF Flow at the Foramen Magnum**
  - Cine-MRI and Queckenstedt's test
- **3. Cerebellar Tonsil Motion on a Partially Enclosed Spinal Subarachnoid Space Creates Elevated Cervical Subarachnoid Pressure Waves and Syrinx Compression**
  - Cardiac-Gated Intraoperative Ultrasound and CSF Pressure Testing
- **4. Systolic Caudal Movement of the Syrinx Fluid**
  - Cine-MRI
- **5. Extra-arachnoidal Craniocervical Decompression and Duraplasty Corrects Pathophysiologic Abnormalities and Resolves Syringomyelia**
  - 3-6 mos. after Surgery: Anatomic and Cine-MRI, CSF Pressure

Anatomy of CSF Pathways

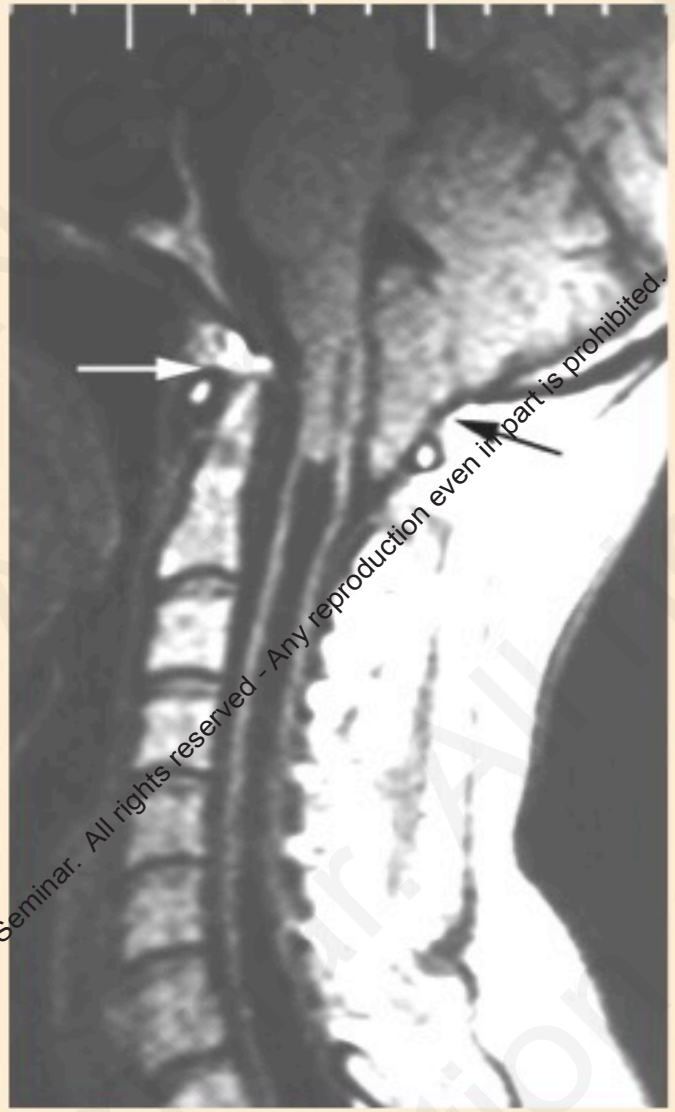
Normal

Chiari I

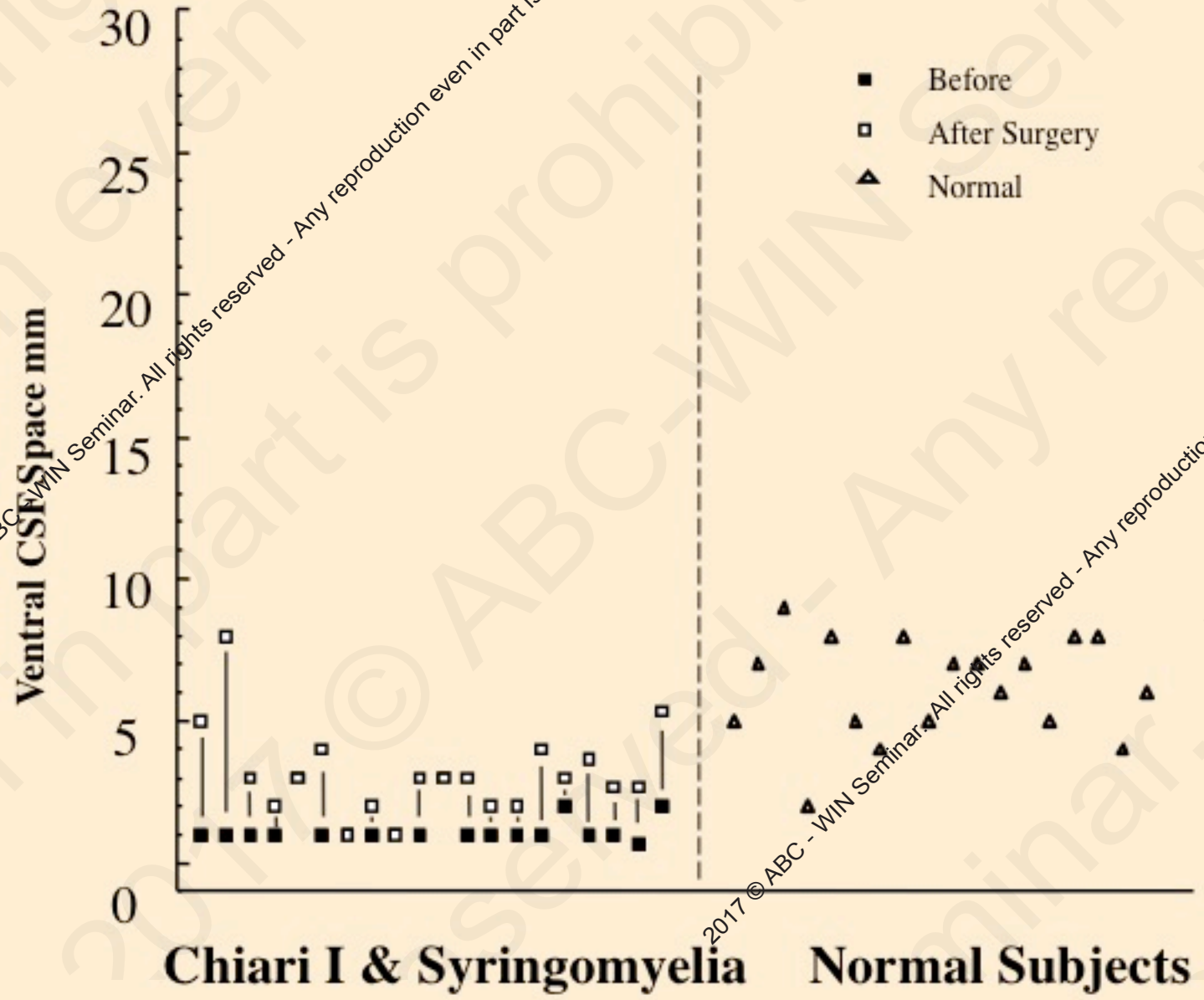
Open



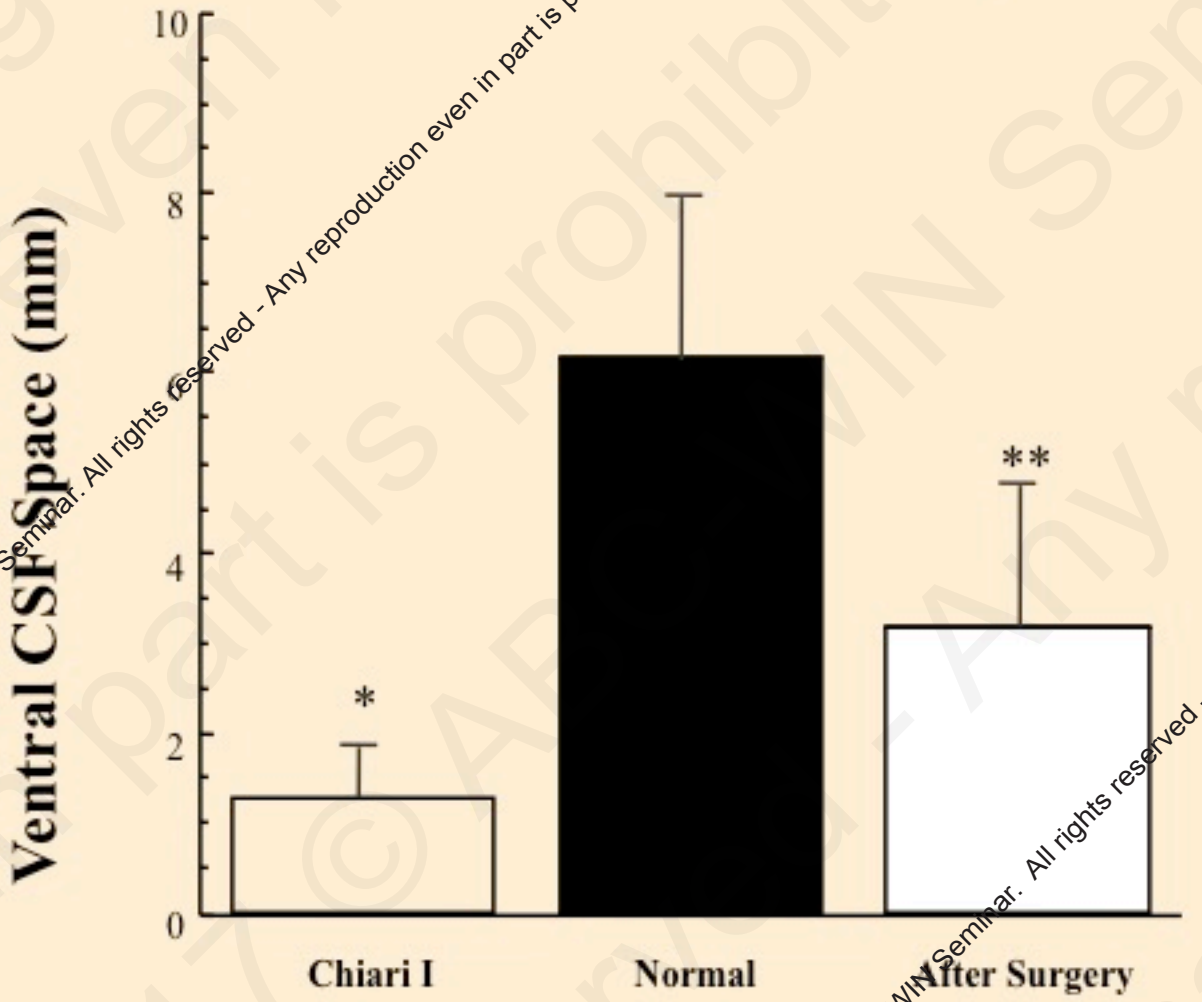
Obstructed



Ventral CSF Space



Ventral CSF Space



* p < 0.0001 Compared to Normal (unpaired t-test)

** p < 0.0001 Compared to Before Surgery (paired t-test)

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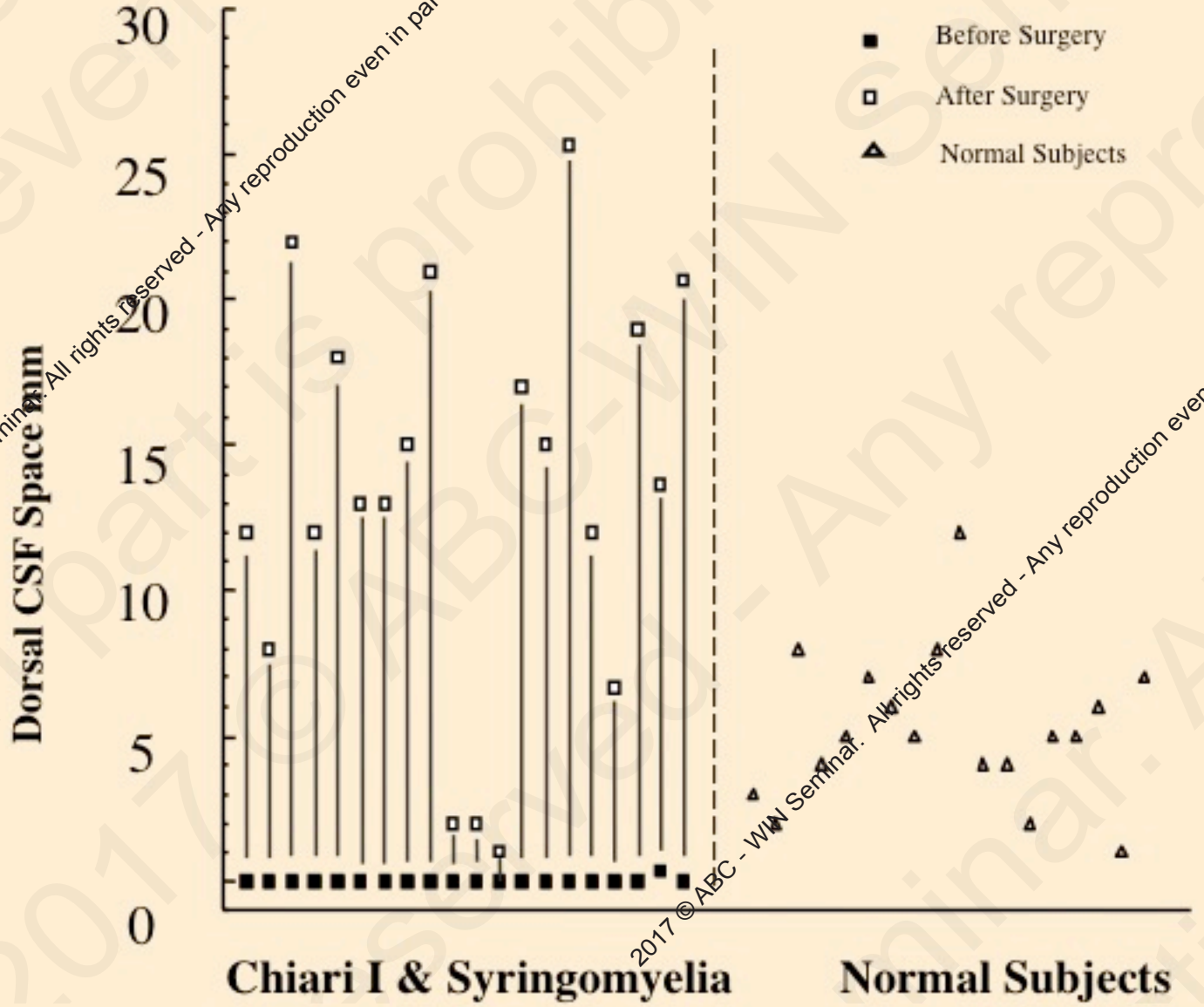
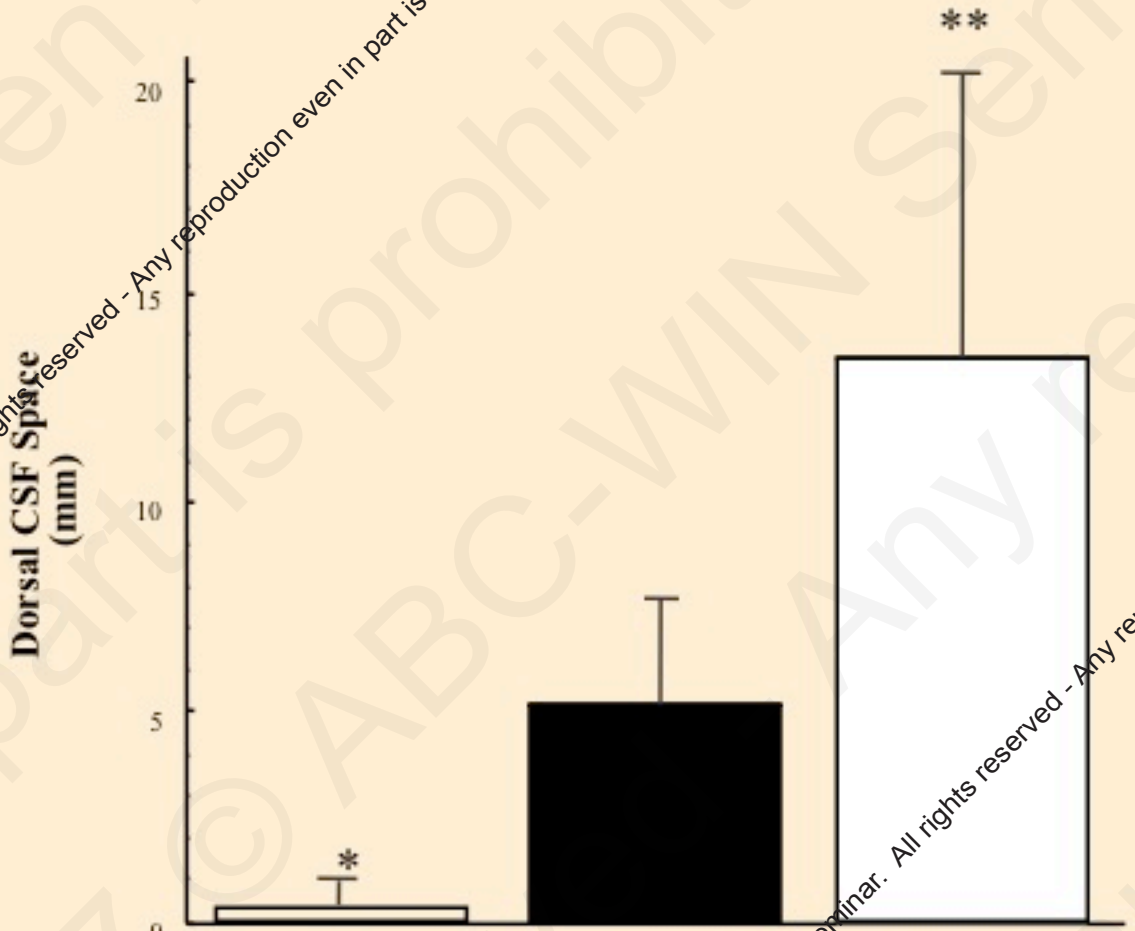
Dorsal CSF Space



Dorsal CSF Space

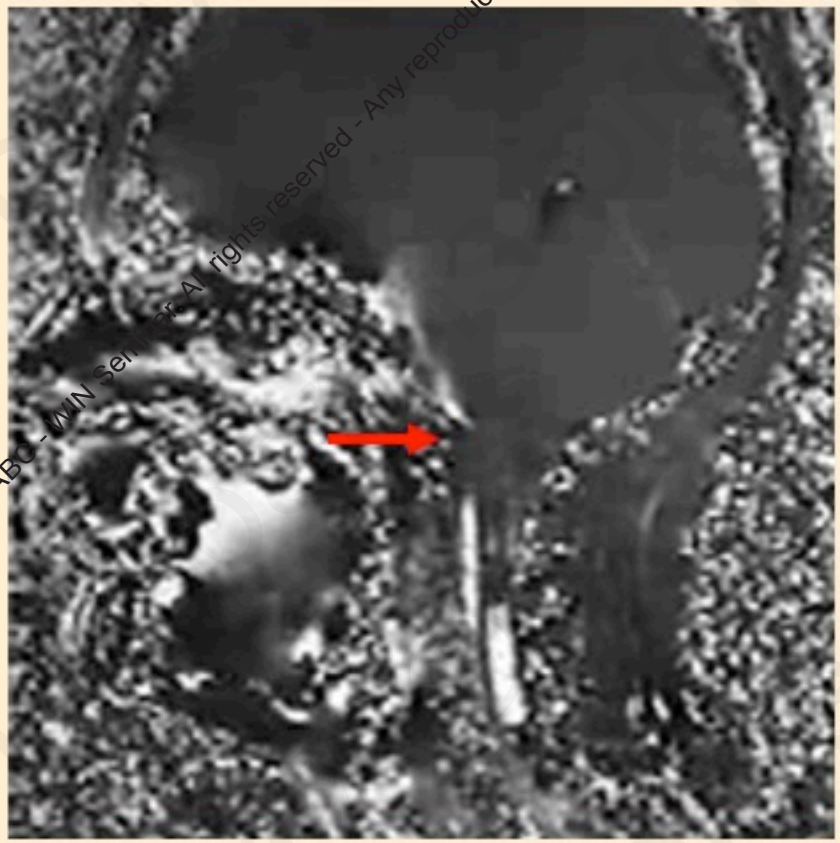


* p < 0.0001 Compared to Normal (unpaired t-test)

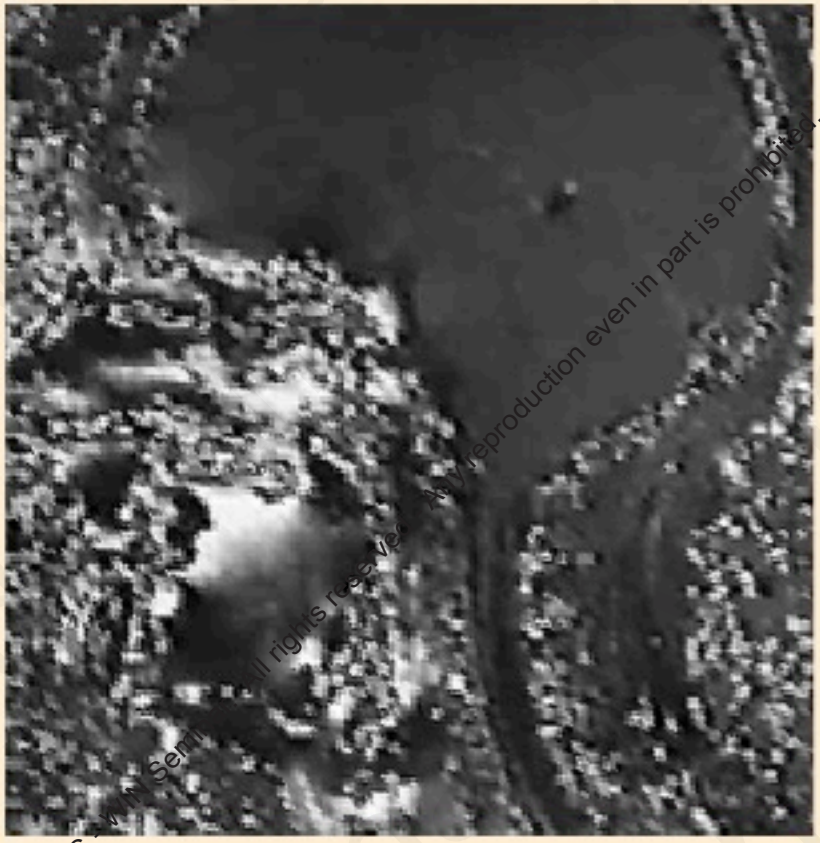
** p < 0.0001 Compared to Before Surgery (paired t-test)

Physiologic Obstruction of CSF Pathways

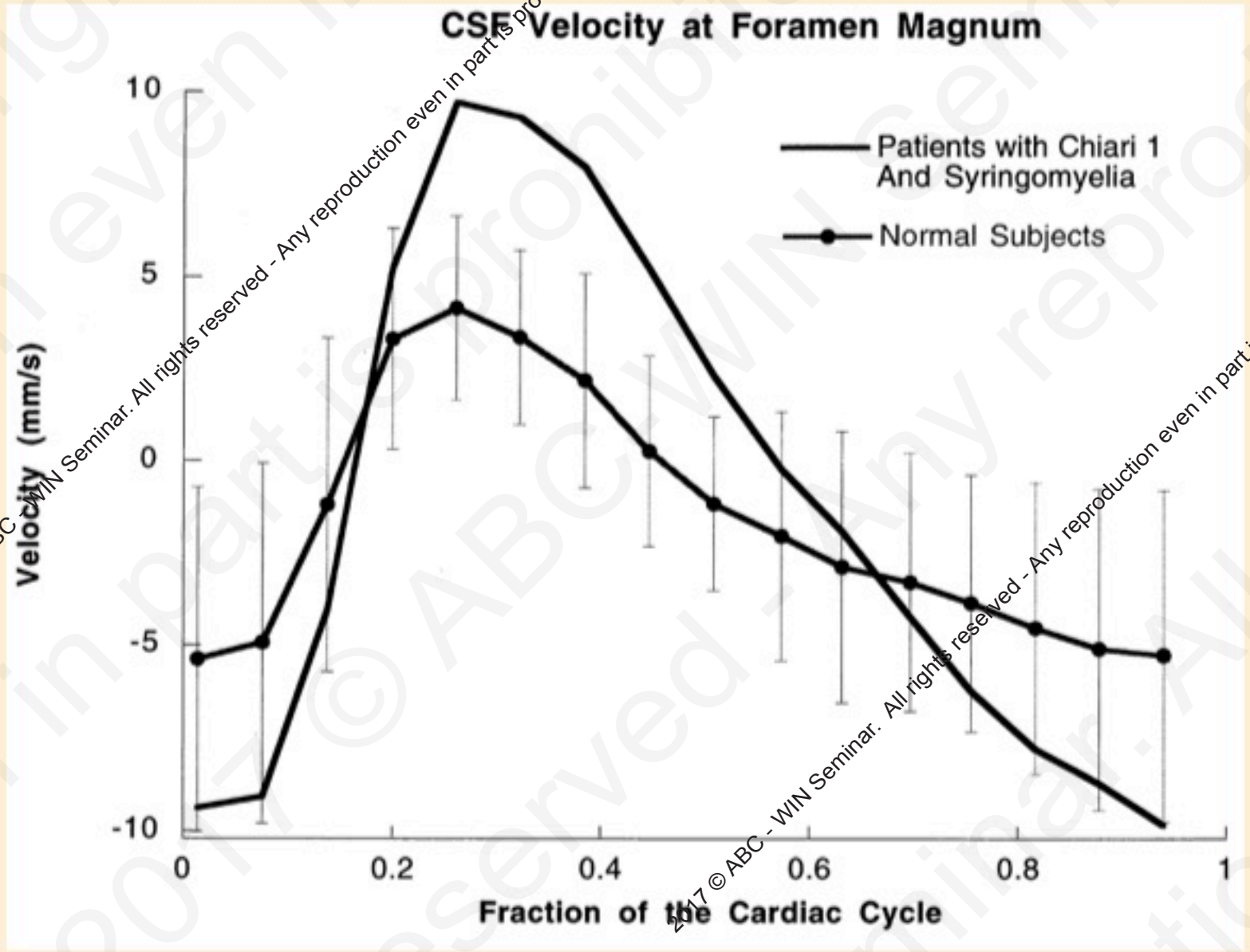
Systole



Diastole



Physiologic Obstruction of CSF Pathways



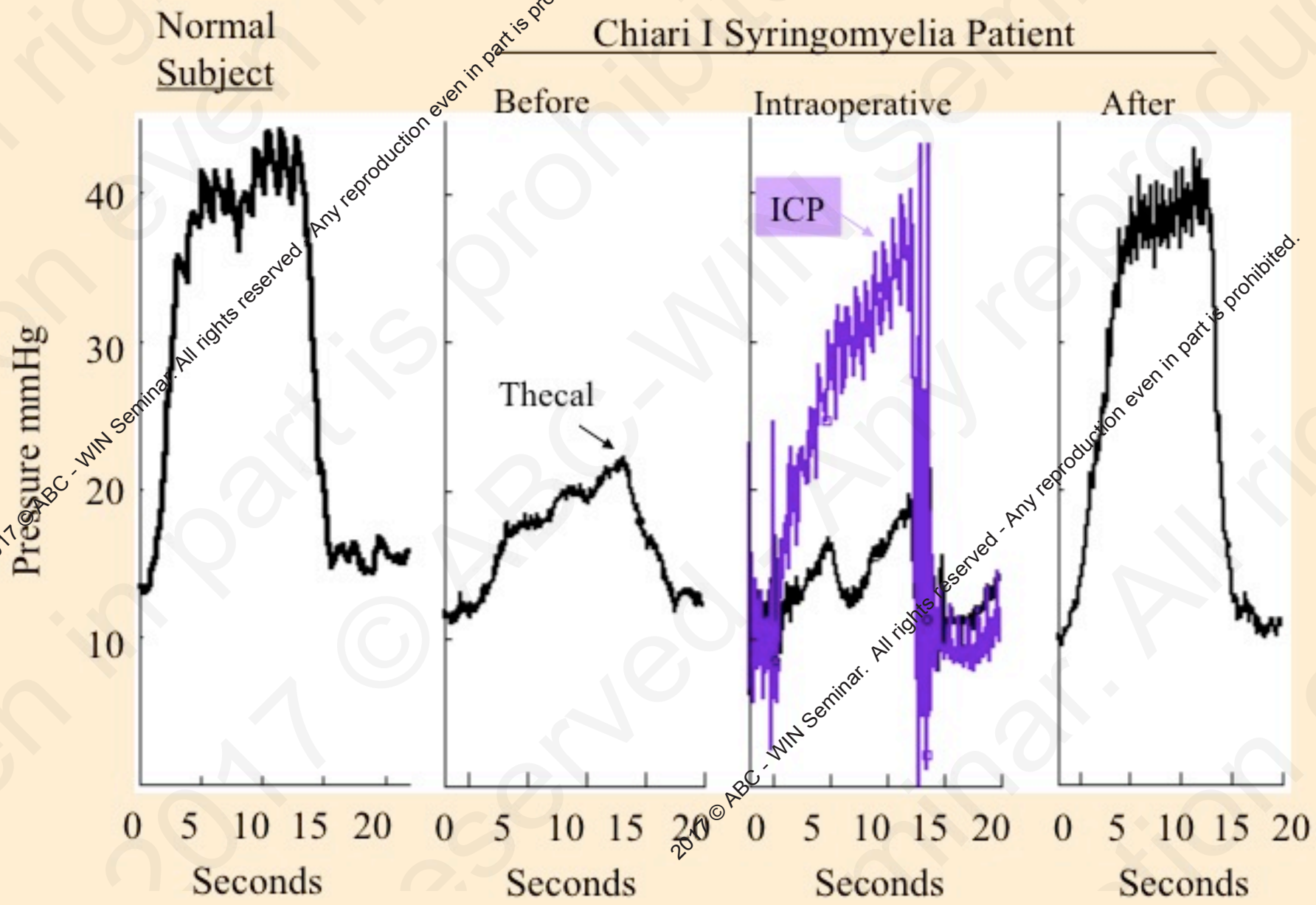
Queckenstedt's Test

Hans Heinrich Georg Queckenstedt
1875

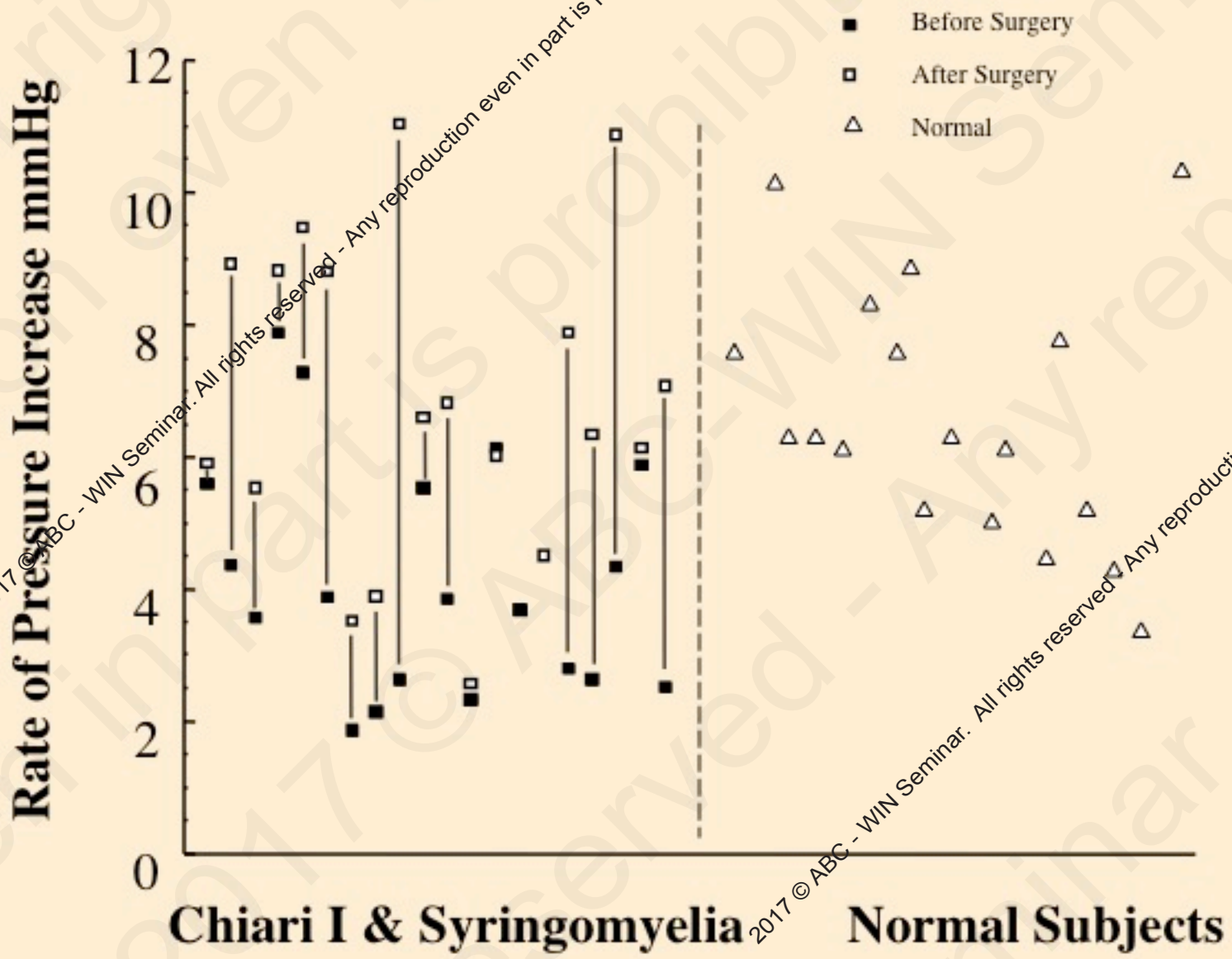
Fig. Compression cuff. Lagergren's model.

From ...sk, O., Lidvall, H. (1965): Electromanometric recordings of cerebrospinal fluid blocks in the cervical region. Acta neurol. scand., 41, suppl. 13, 107-113

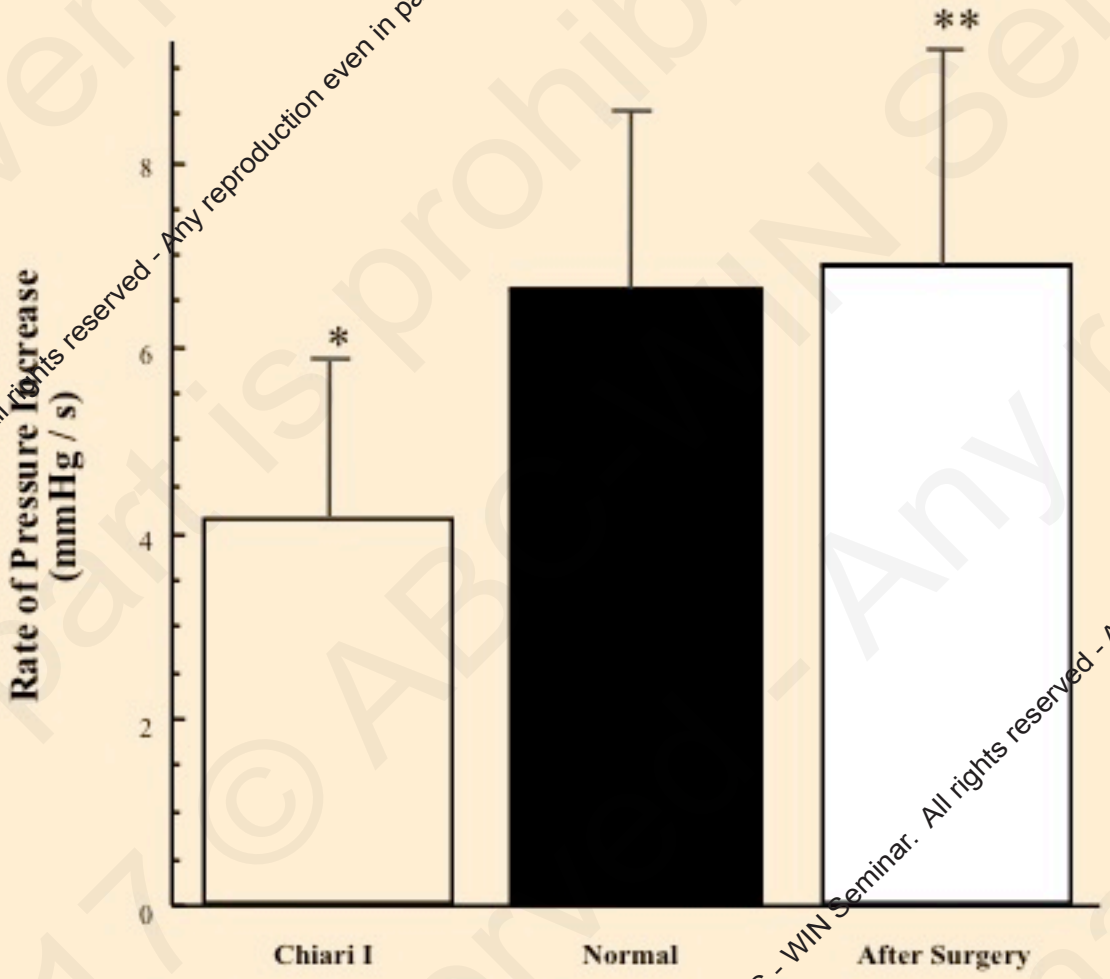
Queckenstedt's Test



Jugular Compression



Jugular Compression



* $p < 0.001$ Compared to Normal (unpaired t-test)

** $p < 0.006$ Compared to Before Surgery (paired t-test)

Cerebellar Tonsillar Motion On a Partially Enclosed Spinal Subarachnoid Space

- Reduced Compliance of Spinal Subarachnoid Space
- Elevated Cervical Subarachnoid Pressure Waves
- Syrinx Compressed During Cardiac Systole

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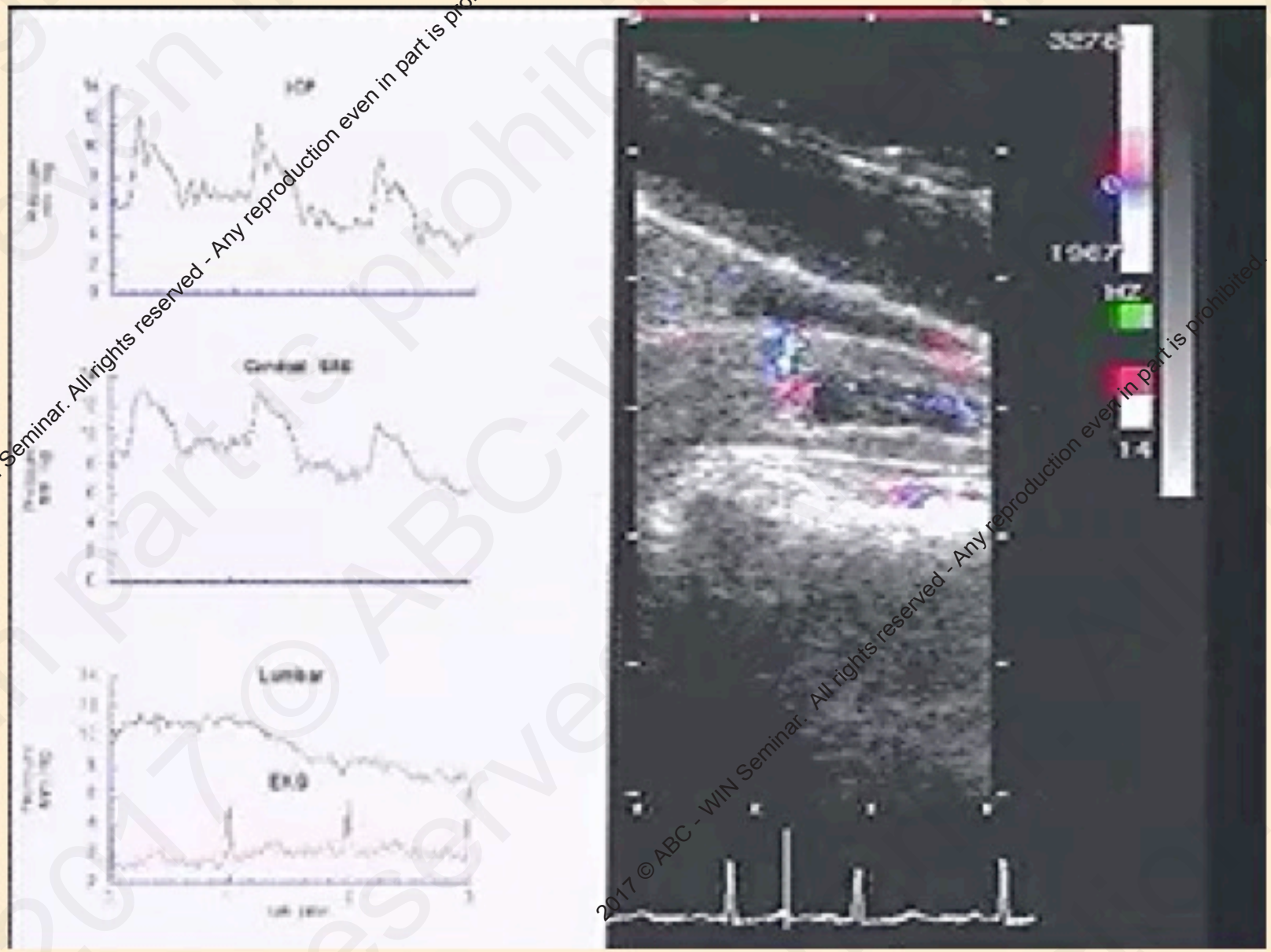
Cerebellar Tonsillar Motion



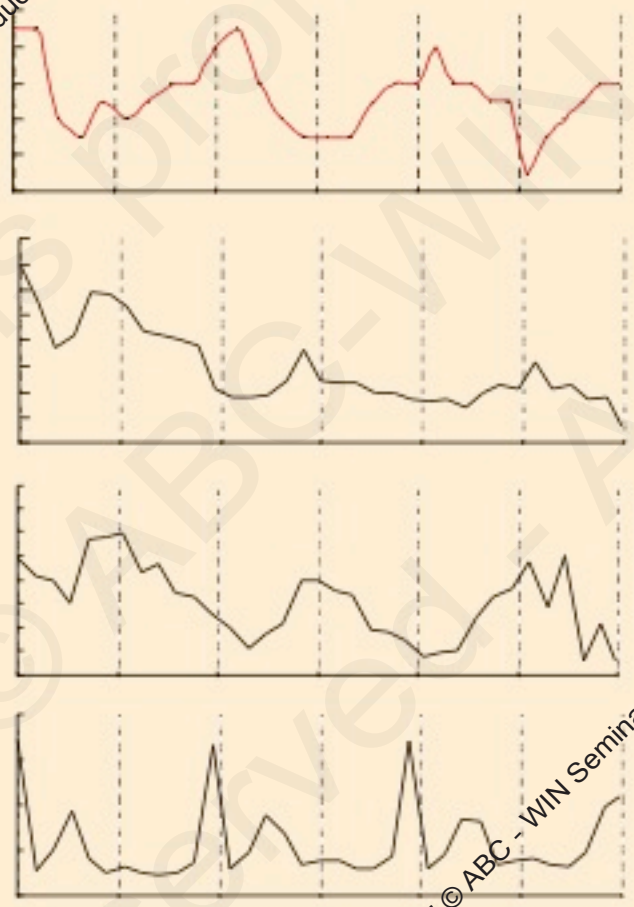
Syrinx Diameter Decreases In Cardiac Systole

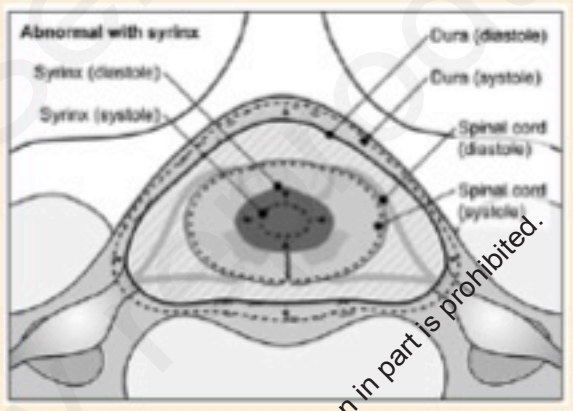
Syrinx Diameter

Cervical CSF Pressure

Syrinx Pressure

EKG





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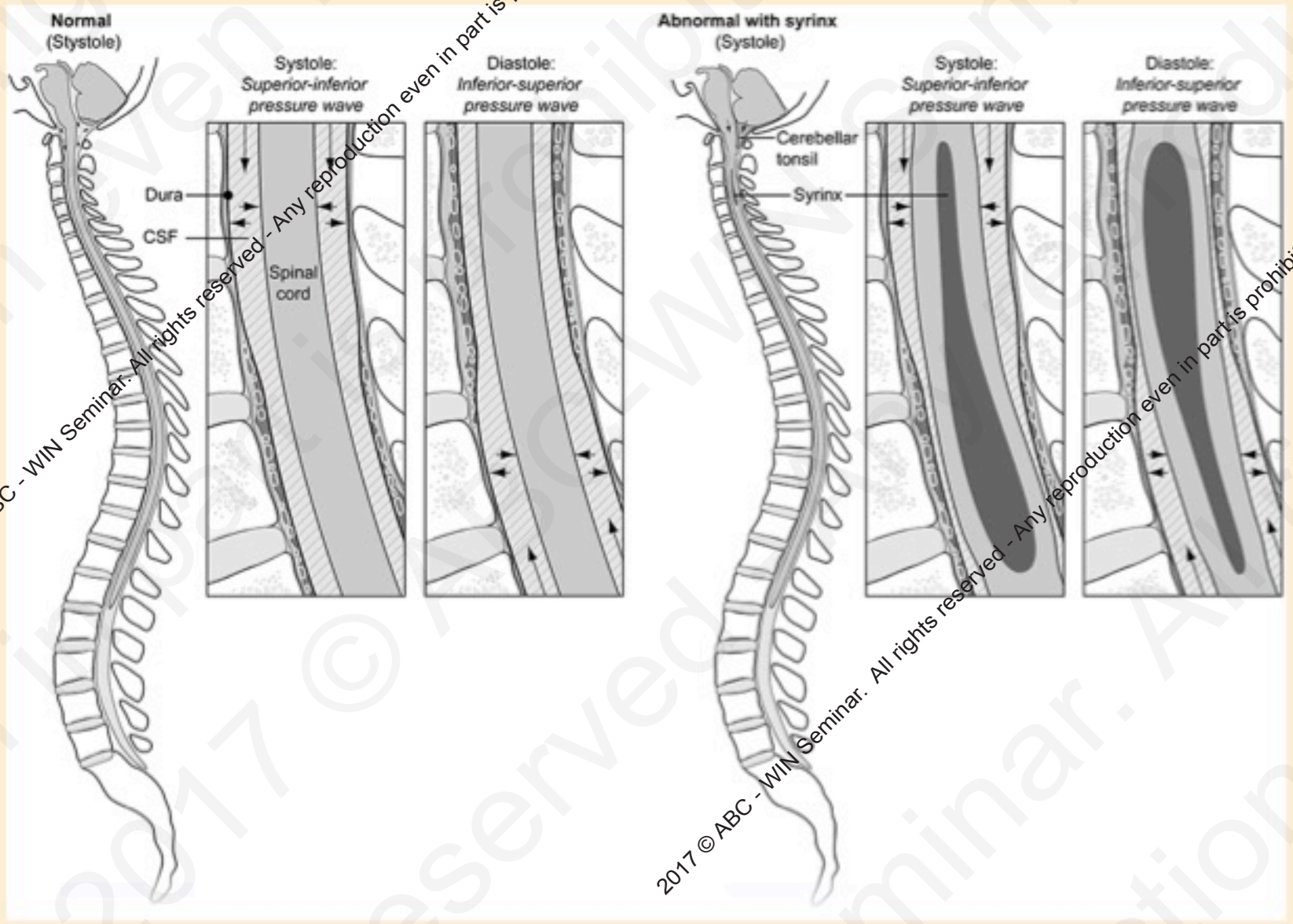
Syrinx Diameter Decreases In Cardiac Systole



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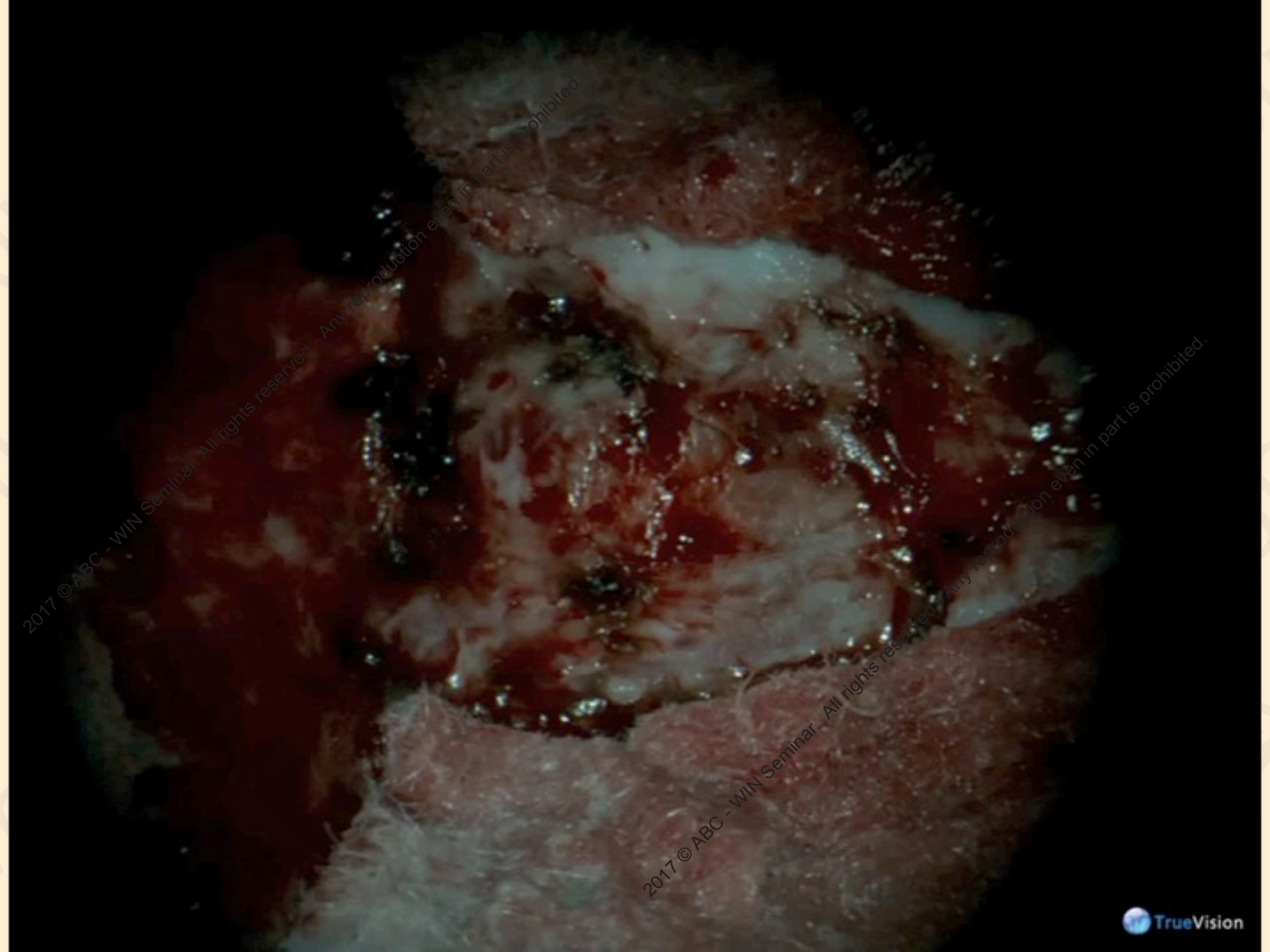
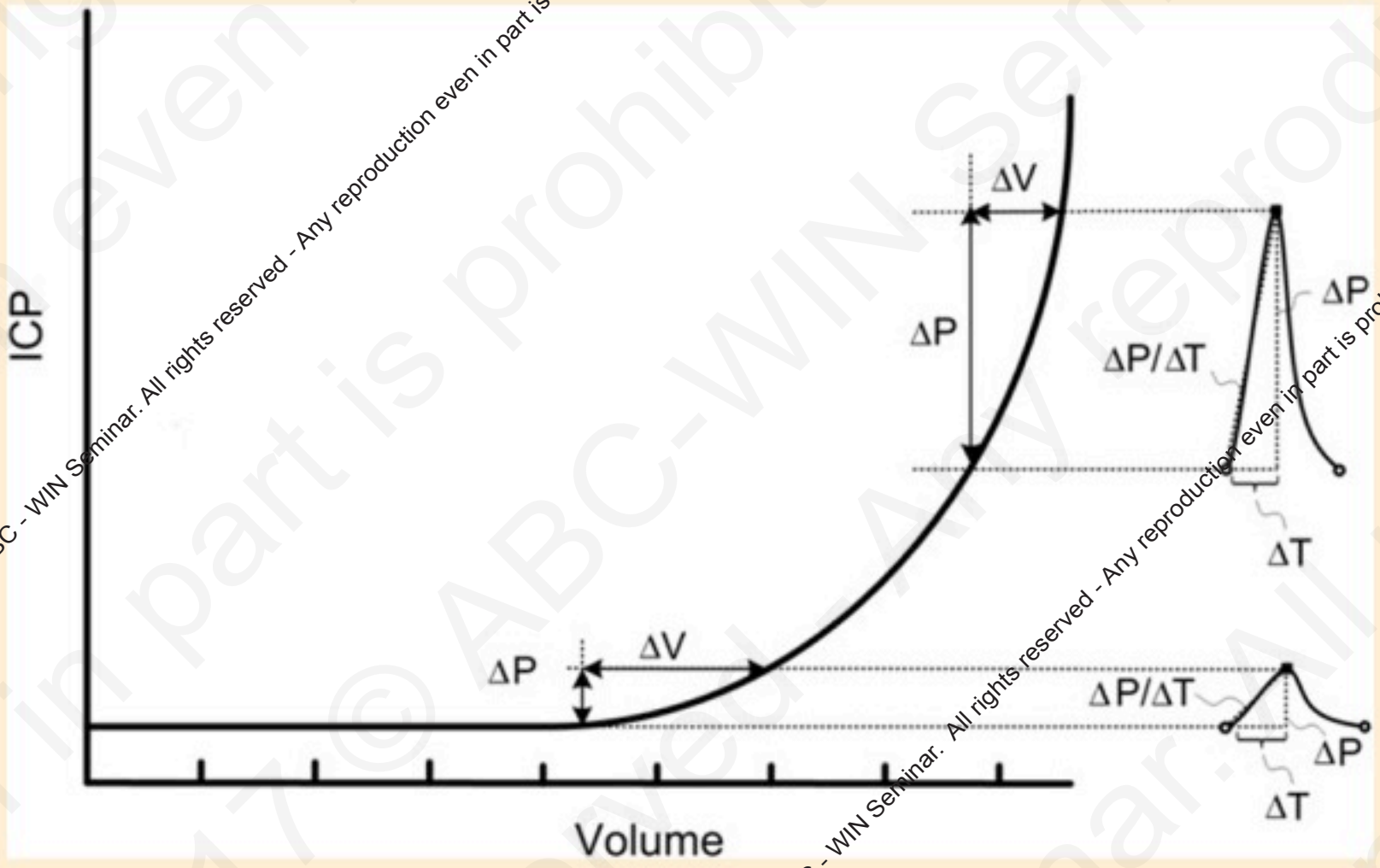


Compliance



Wagshul et al. Fluids and Barriers of the CNS 2011, 8:5
http://www.fluidsbarrierscns.com/content/8/1/5

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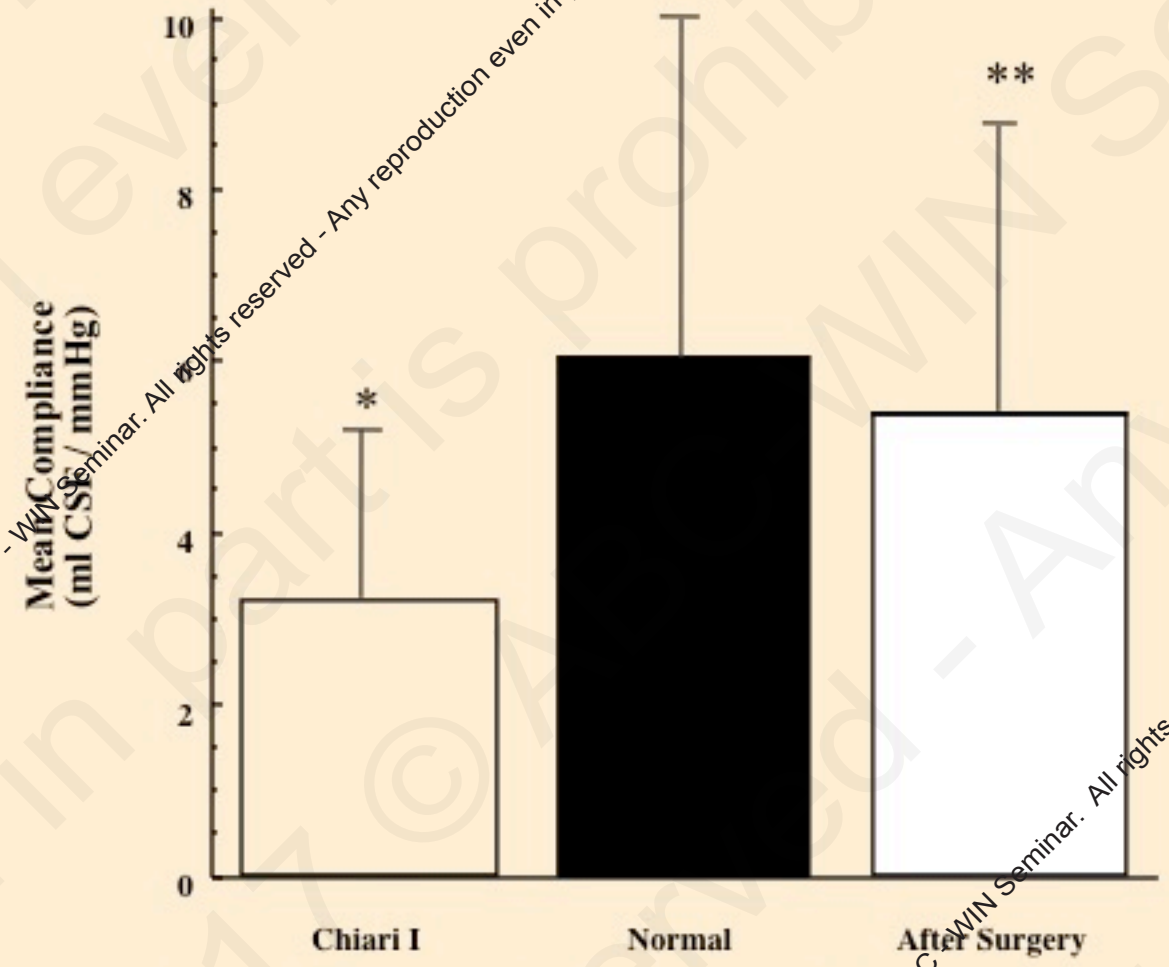
Compliance



* p < 0.02 Compared to Normal (unpaired t-test)

** p < 0.05 Compared to Before Surgery (paired t-test)

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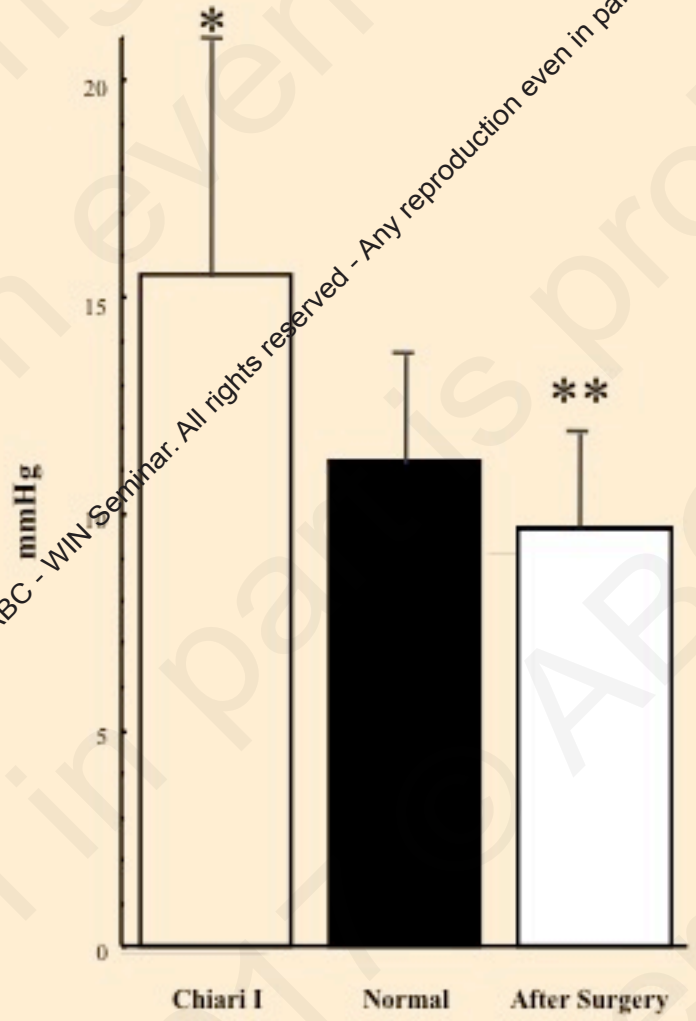
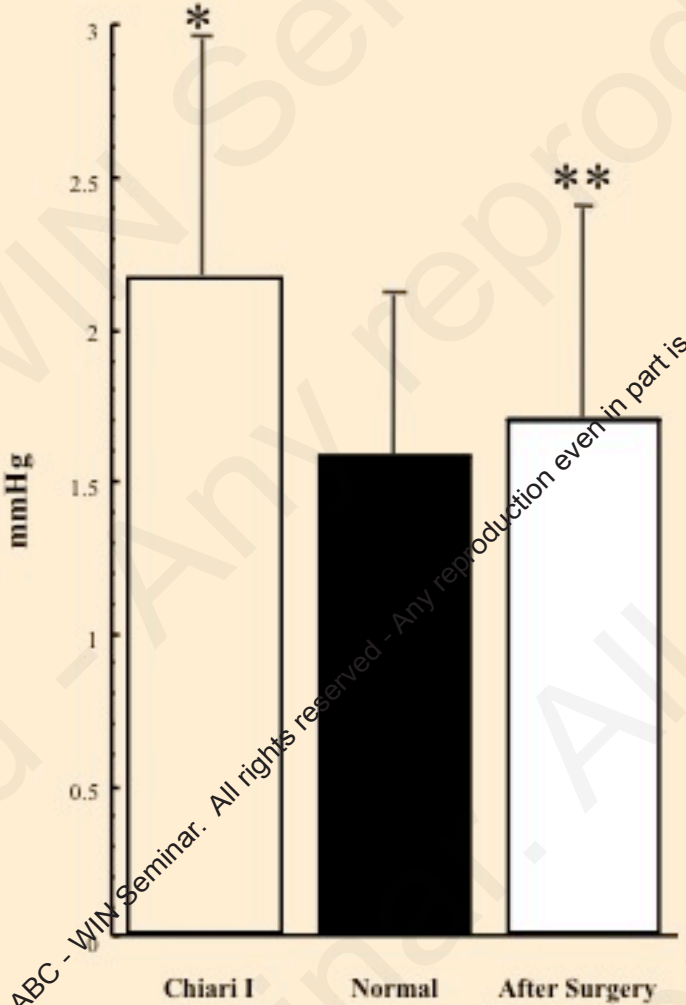
Cervical Mean Pressure



* p < 0.006 Compared to Normal (unpaired t-test)
** p < 0.02 Compared to Before Surgery (paired t-test)

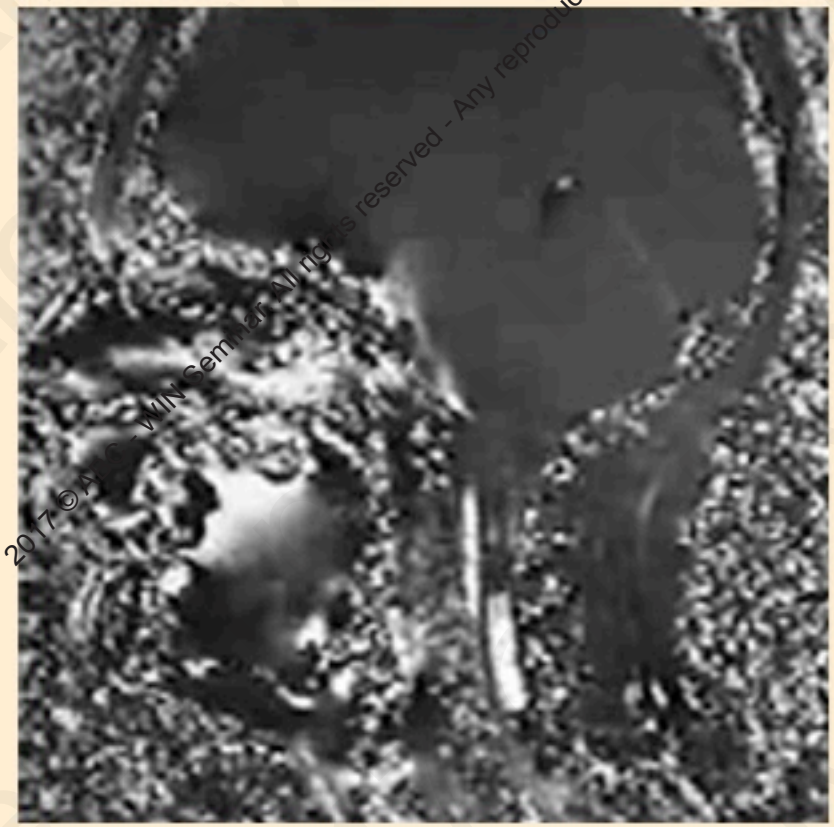
Cervical Pulse Pressure

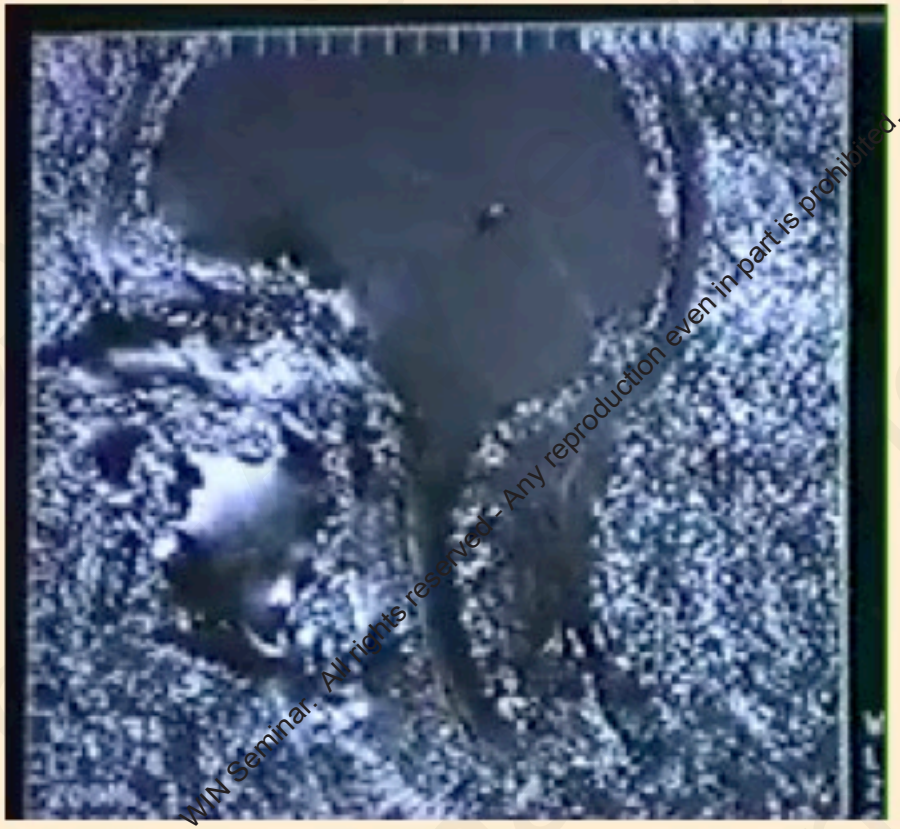


* p < 0.002 Compared to Normal (unpaired t-test)
** p < 0.04 Compared to Before Surgery (paired t-test)

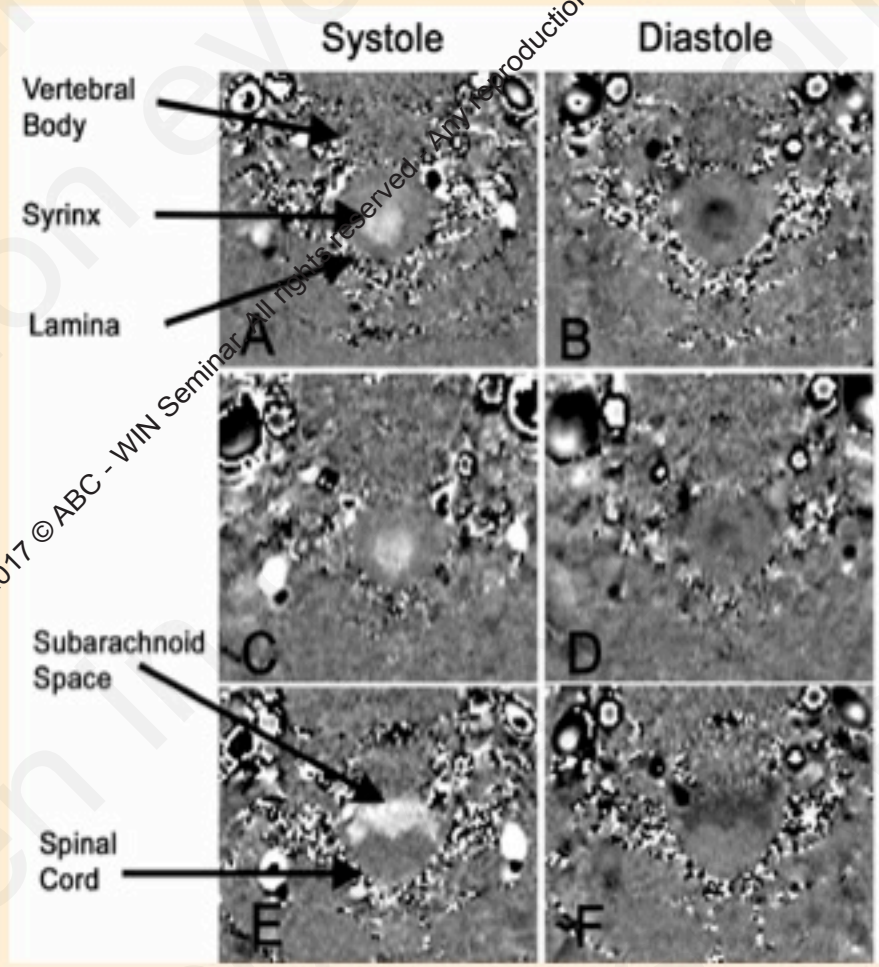
4. Caudal Flow of Syrinx Fluid During Systole

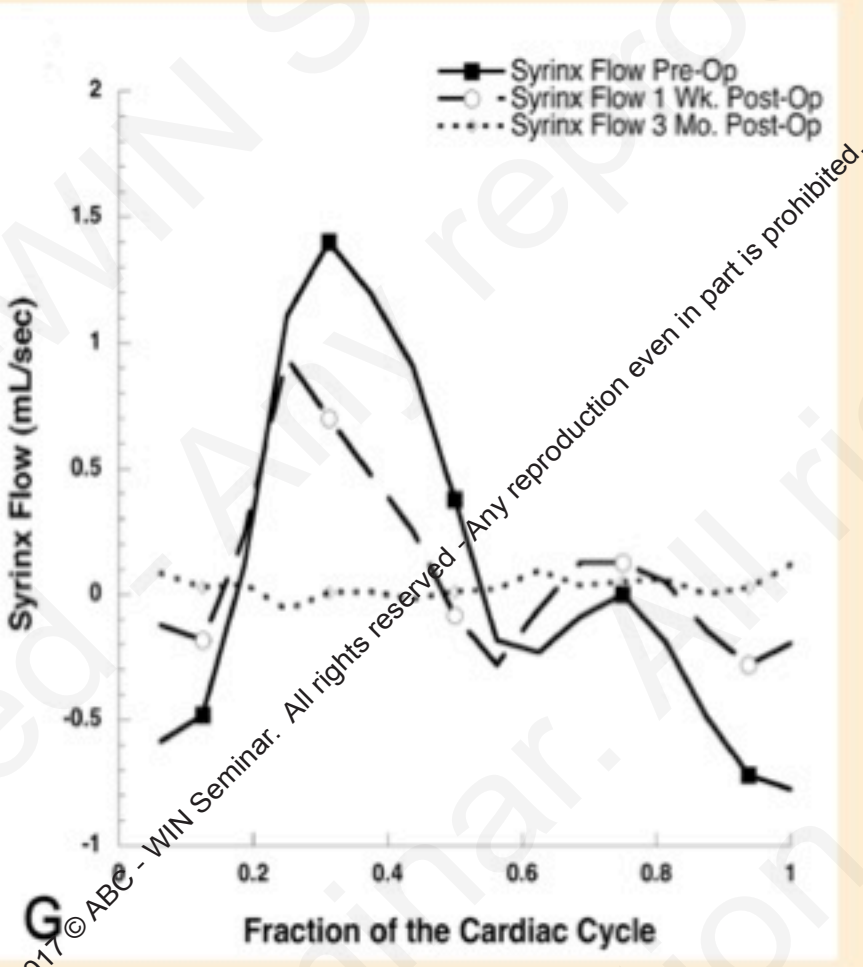
Systole





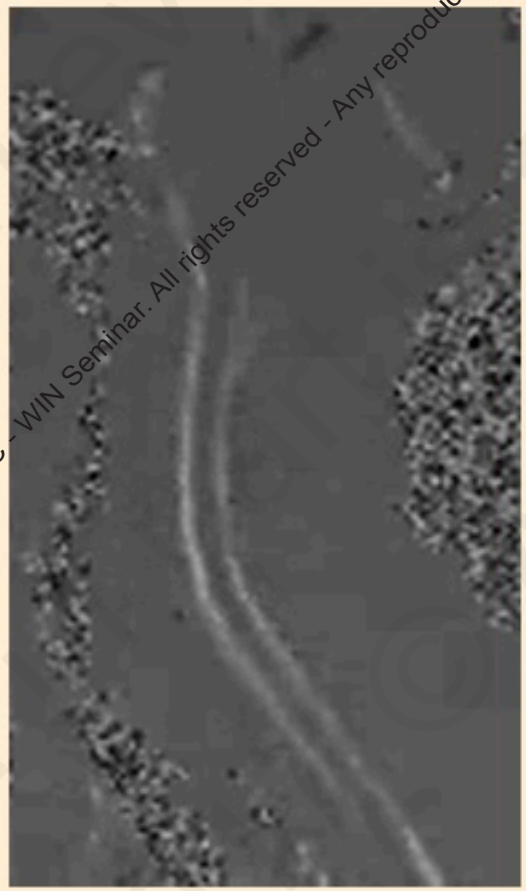
PC-MRI CSF Flow Studies





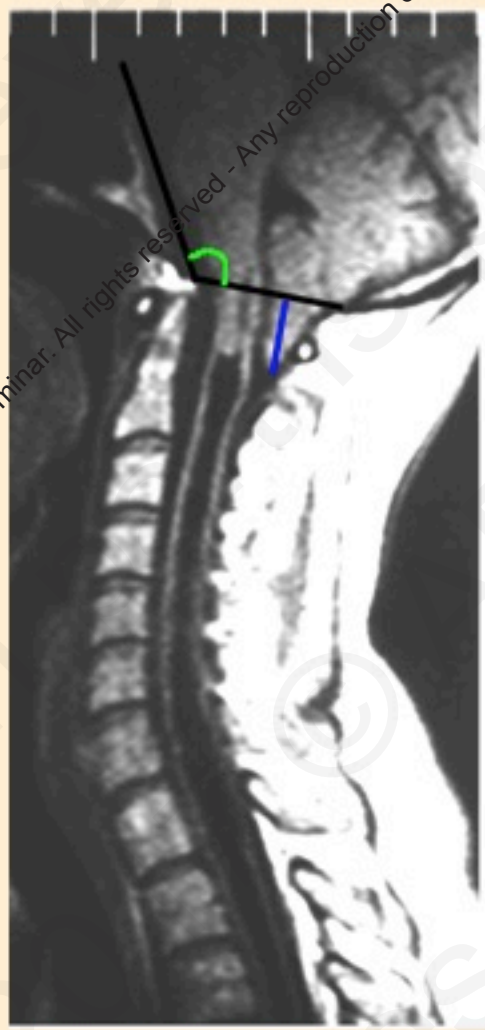
Extra-arachnoidal Craniocervical Decompression & Duraplasty Corrects Pathophysiologic Abnormalities & Resolves Syringomyelia

After Surgery



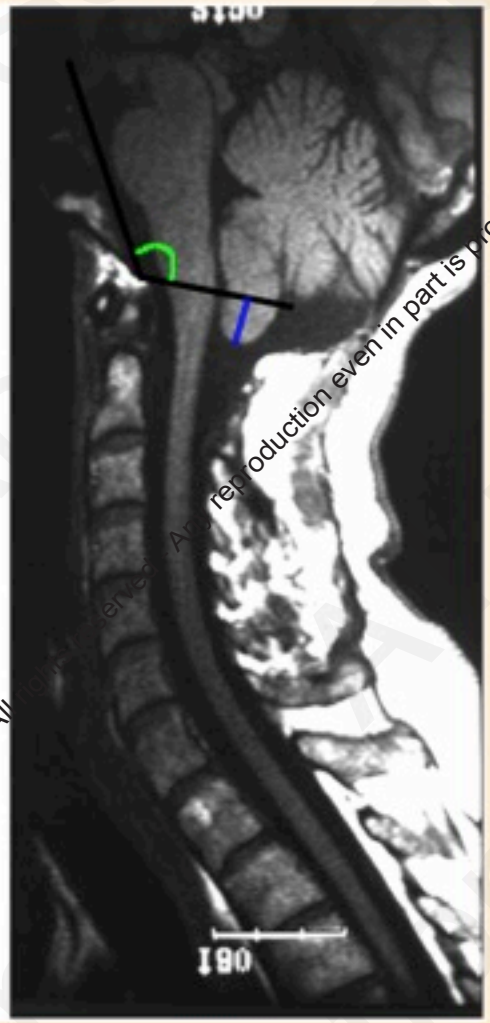
Hindbrain Deformity & Syrinx Resolve After Extra-arachnoidal Craniocervical Decompression & Duraplasty

Before Surgery



1)Tonsillar Shape
2)Tonsillar Ectopia (mm)
3) Medullary deformity

After Surgery



MRI Results

Parameter	Chiari I and Syringomyelia (N=20)		Normal (N=18)
	Before Surgery	6 Months After Surgery	
Syrinx diameter (mm)	8.6 ± 1.5	1.5 ± 1.7, p*<.0001	----------
Syrinx length (Spinal levels)	9.8 ± 4.7	2.4 ± 2.9, p*<.0001	----------
Syrinx length (cm)	17.3 ± 10.6	3.6 ± 4.7, p*<.0001	----------
Tonsillar herniation below foramen magnum (mm)	11.1 ± 5.4	5.5 ± 3.9, p*<.0001	0.3 ± 1.0, p**<.0001
Pointed tonsils	15 (75%)	1 (5%), p*<.0001	0 (0%), p**<.0001
Small cerebellar sulci	17 (85%)	0 (0%), p*<.0001	0 (0%), p**<.0001
Foramen magnum CSF pathway			
Ventral AP diameter (mm)	1.3 ± 0.7	3.2 ± 1.6, p*<.0001	6.2 ± 1.8, p**<.0001
Dorsal AP diameter (mm)	0± 0.1	13.4 ± 6.9, p*<.0001	5.2 ± 2.6, p**<.0001
Syrinx to 4th ventricle connection	1 (5%)	----------	----------
Ventriculomegaly	4 (20%)	4 (20%)	0 (0%)

Significant when compared to Chiari I Before Surgery, paired t-test* and unpaired t-test**

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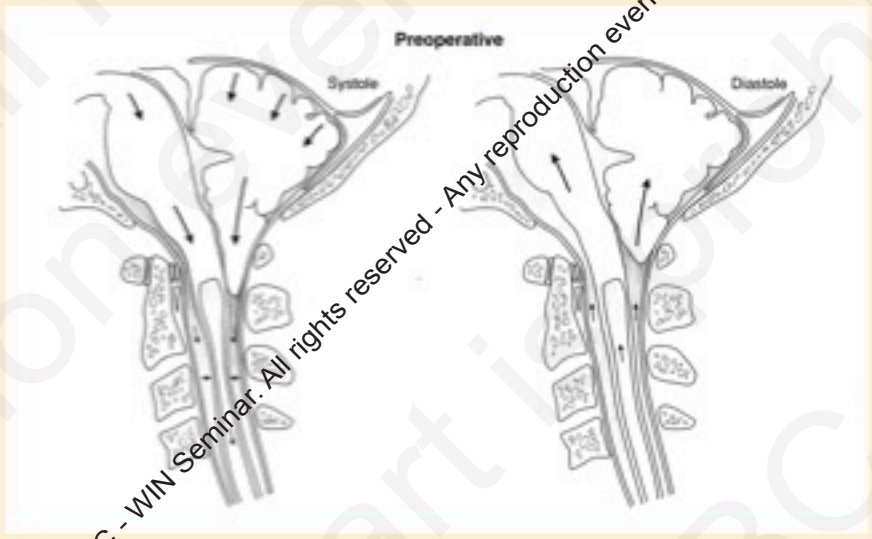
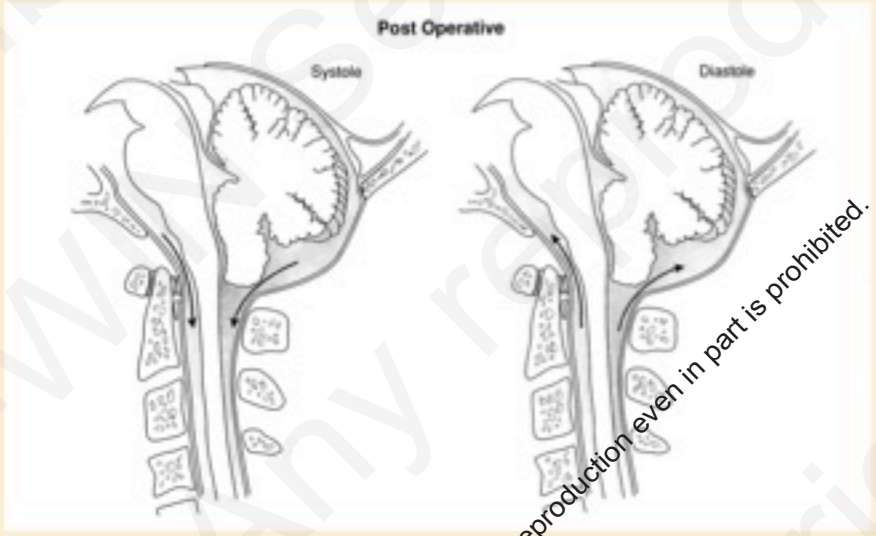
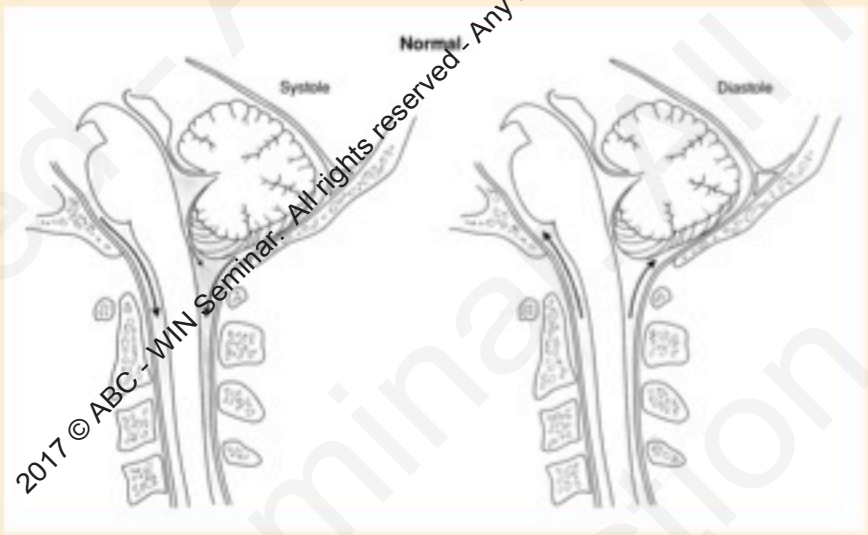
Effective Decompressive Surgery







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Conclusions

- The study results were consistent with the theory of pathophysiology of the Chiari I-type of syringomyelia
- Partial block at the foramen magnum remains the principle cause of syrinx progression
- Treatment should be directed at correcting the partial block at the foramen magnum

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MRI Resolution of Syringomyelia is a Slow Process after Craniocervical Decompression

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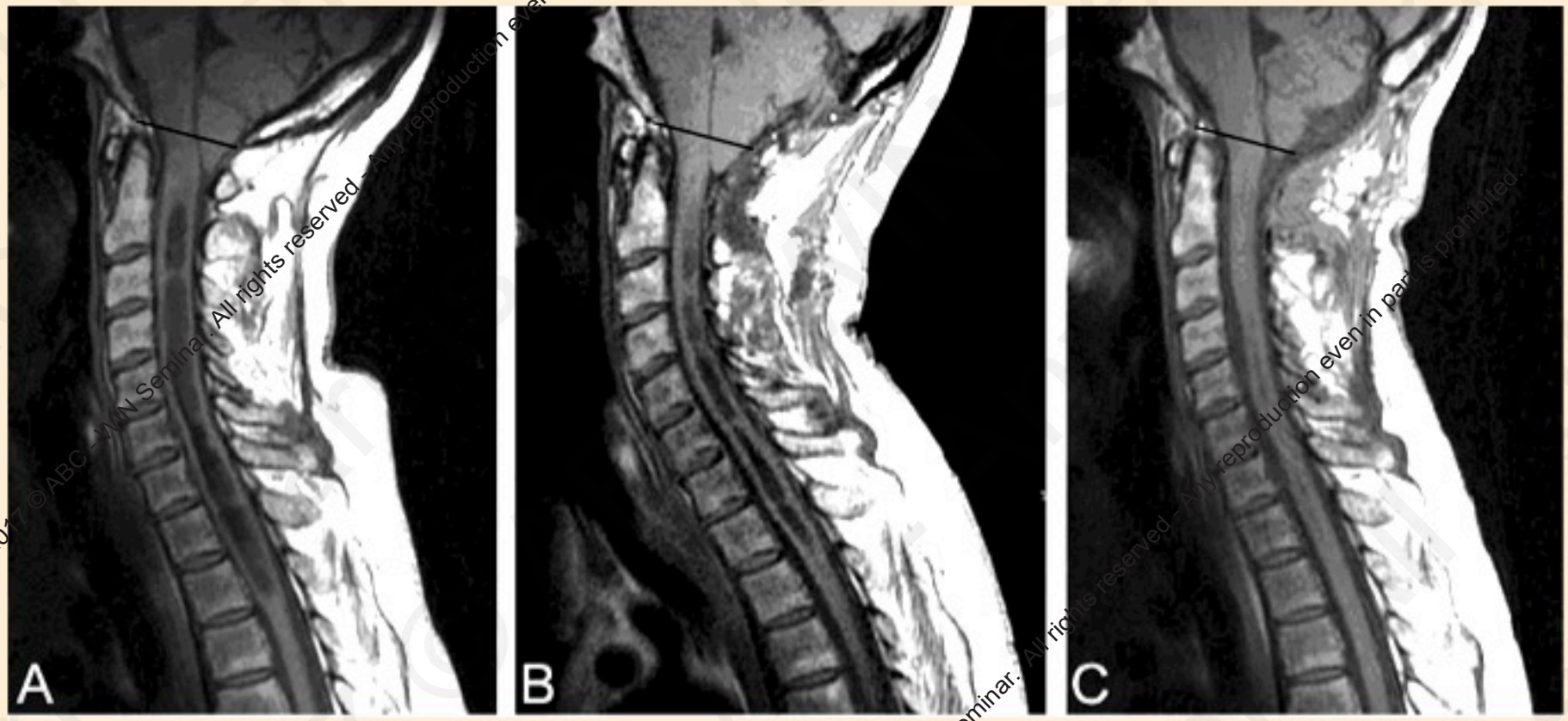
Response of Cervical Syrinx to Craniocervical Decompression



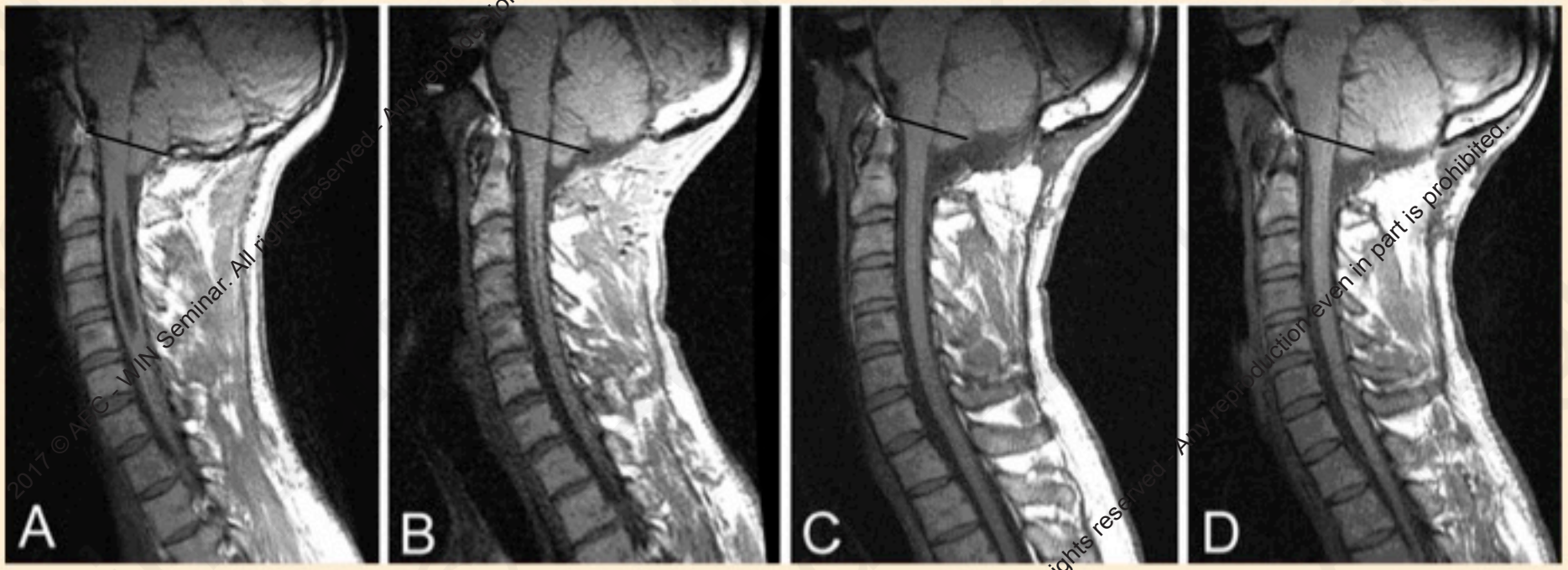
Before Surgery

1 Week after Surgery

3 Months after Surgery

Modified from Wetjen NJ, Heiss JD, Oldfield EH: J Neurosurg, 2008

Cisterna Magnum Present after Surgery-- Tonsils Ascend and Become Rounded



Before Surgery

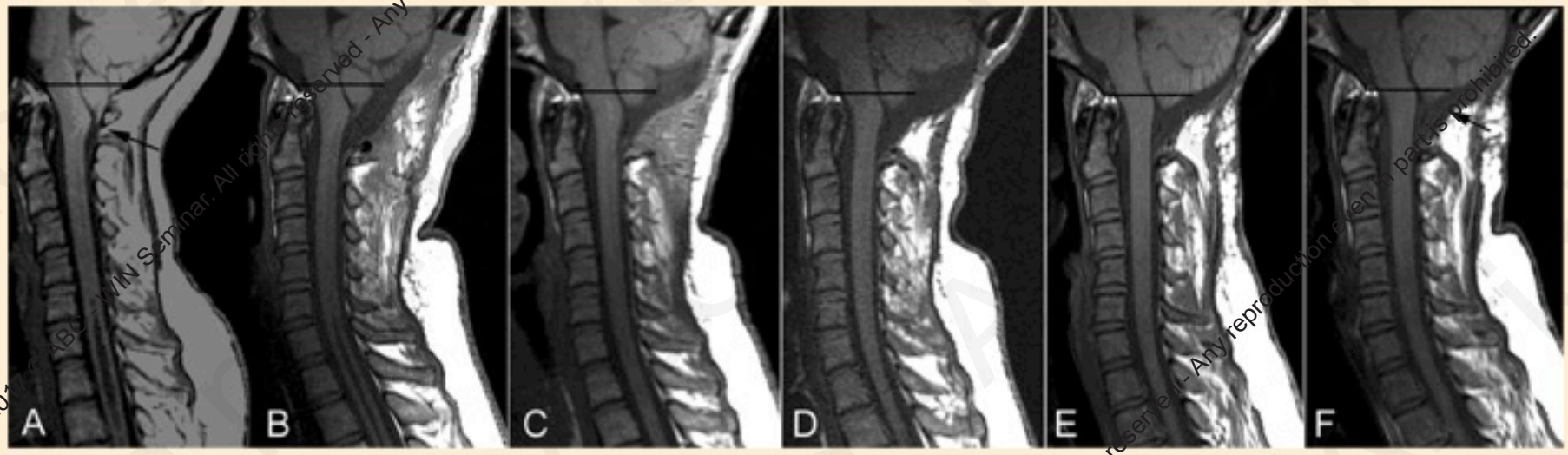
3 Months after Surgery

1 Year after Surgery

3 Years after Surgery

Modified from Wetjen NJ, Heiss JD, Oldfield EH: J Neurosurg, 2008

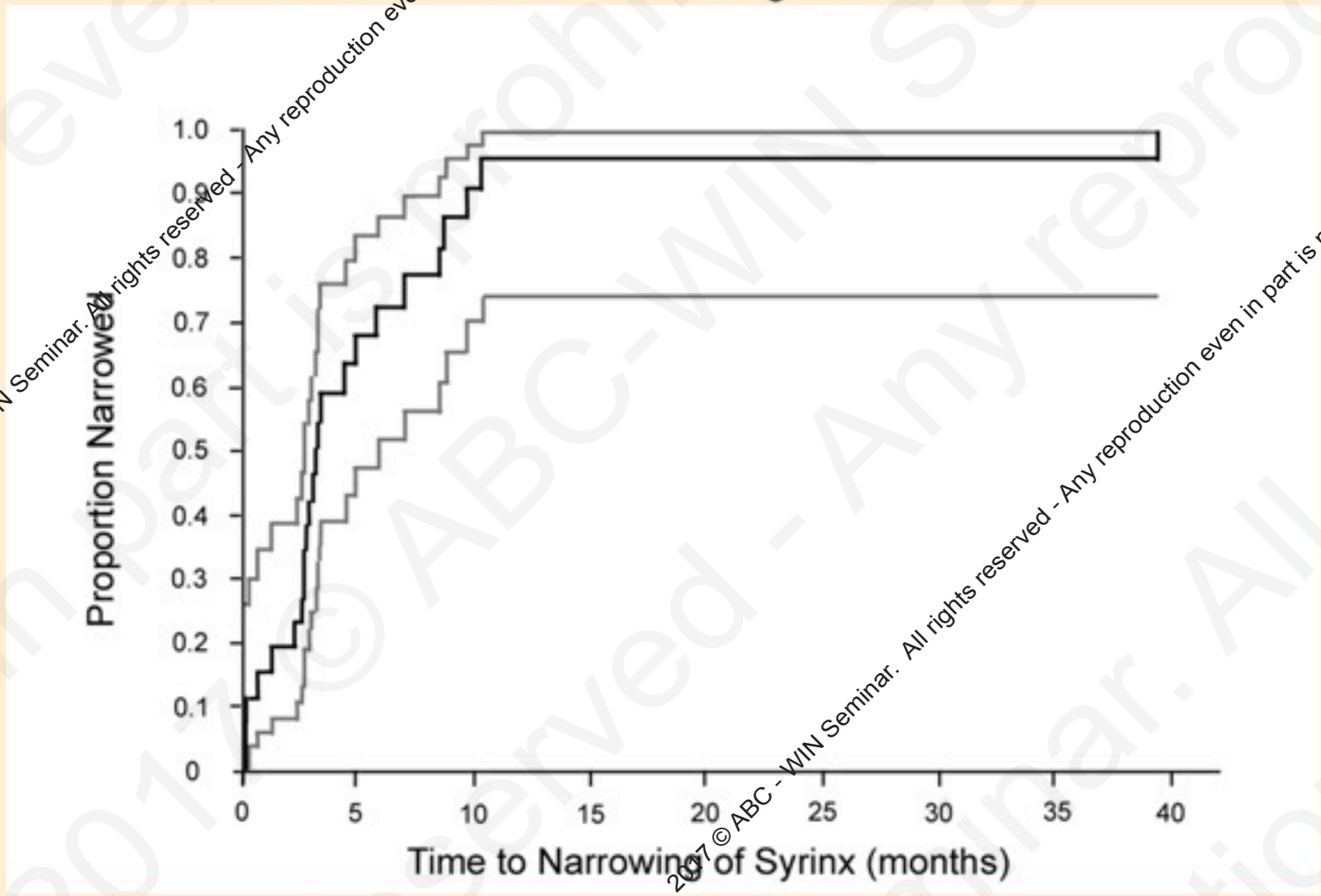
Resolution of Hindbrain Deformity after Surgery



Before Surgery	1 Week after Surgery	3 Months after Surgery	1 Year after Surgery	3 Years after Surgery	5 Years after Surgery

Modified from Wetjen NJ, Heiss JD, Oldfield EH: J Neurosurg, 2008

Period Before Syrinx Diameter Narrowed by 50%



So-Called Idiopathic Chiari I Malformation Results from Decreased Size of the Posterior Fossa

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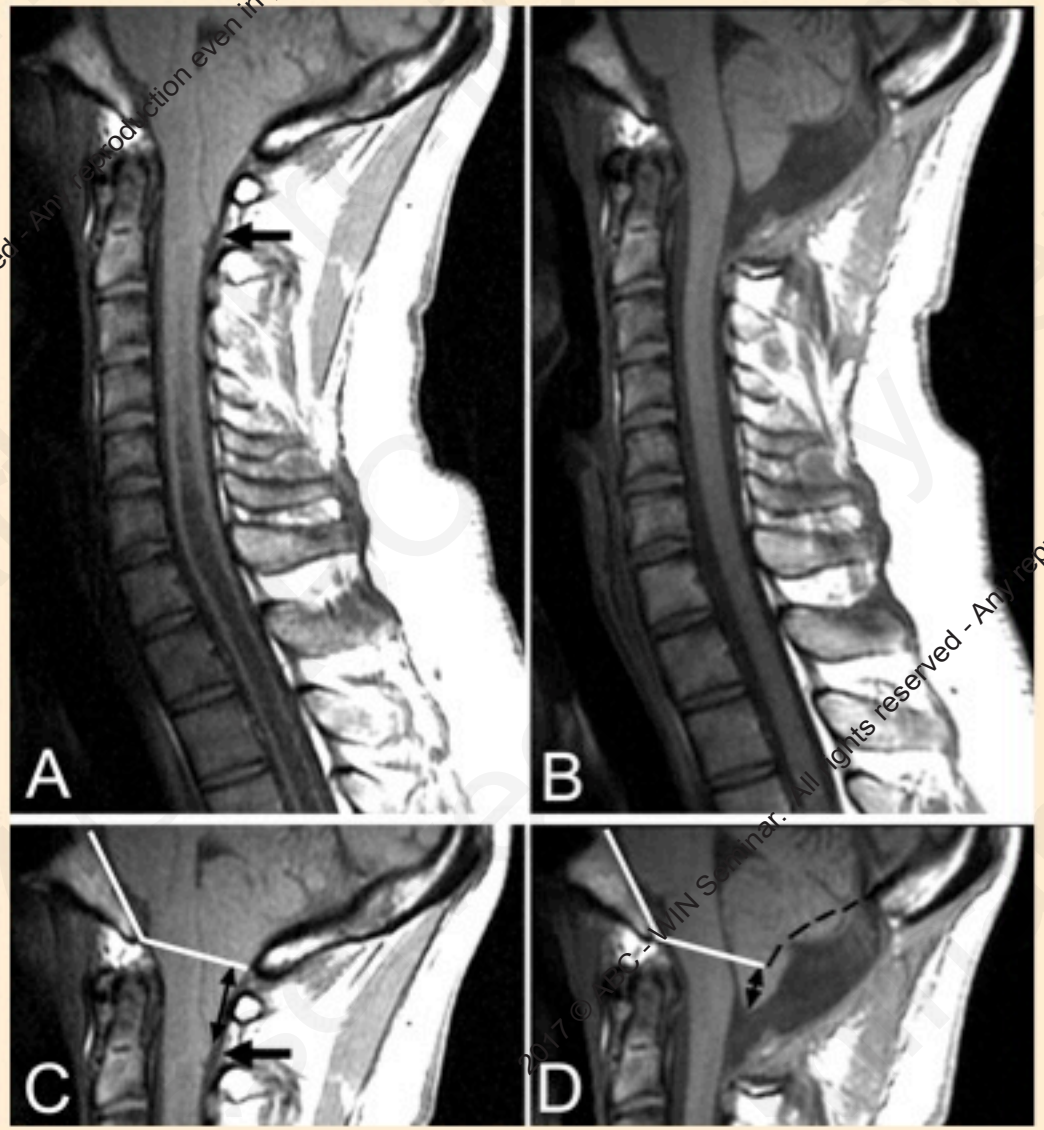
Response of Syrinx to Craniocervical Decompression

Before Surgery

6 Months after Surgery

Successful Decompression Relieves the CSF Obstruction and Hindbrain Compression and Reduces Syrinx Size



Anatomic Changes After Successful Decompression

Feature	Preop (N = 48)	1 wk Postop† (N = 42)	3–6 mos Postop (N = 48)	Control Group (N = 18)
tonsillar ectopia (mm)	12.3 ± 5.1	9.0 ± 4.1 (p < 0.0001)‡	6.0 ± 3.3 (p < 0.0001)‡	0.3 ± 1.0
tonsillar shape				
round	9	16	48	18
pointed	39	26	0	0
presence of cervicomedullary protuberance				
yes	34	22	0	0
no	14	20	48	18
bulbopontine sulcus distance superior to basion (mm)	9.5 ± 2.6 (p < 0.0001)§	9.9 ± 2.5 (p < 0.02)‡ (p < 0.0001)§	10.7 ± 2.3 (p < 0.0001)‡ (p < 0.0007)§	13.6 ± 2.8
AP width of CSF pathway (mm)				
ventral	1.7 ± 0.8 (p < 0.0001)§	1.9 ± 0.9 (p < 0.0001)§	2.7 ± 0.9 (p < 0.0001)‡ (p < 0.0001)§	6.2 ± 1.8
dorsal	0.08 ± 0.3 (p < 0.0001)§	6.1 ± 4.3 (p < 0.0001)‡	12.0 ± 4.5 (p < 0.0001)‡ (p < 0.0001)§	5.2 ± 2.6
clivus length (mm)	38.6 ± 3.4 (p < 0.0001)§	NA	NA	43.2 ± 3.5
basiocciput length (mm)	19.7 ± 3.3 (p < 0.0001)§	NA	NA	26.3 ± 4.4
supraocciput length (mm)	40.1 ± 4.0	NA	NA	41.5 ± 4.4

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Lack of MRI Resolution of Syringomyelia Indicates Persistent Obstruction of the CSF Pathways at the Foramen Magnum that Can Usually be Corrected with Craniocervical Decompression

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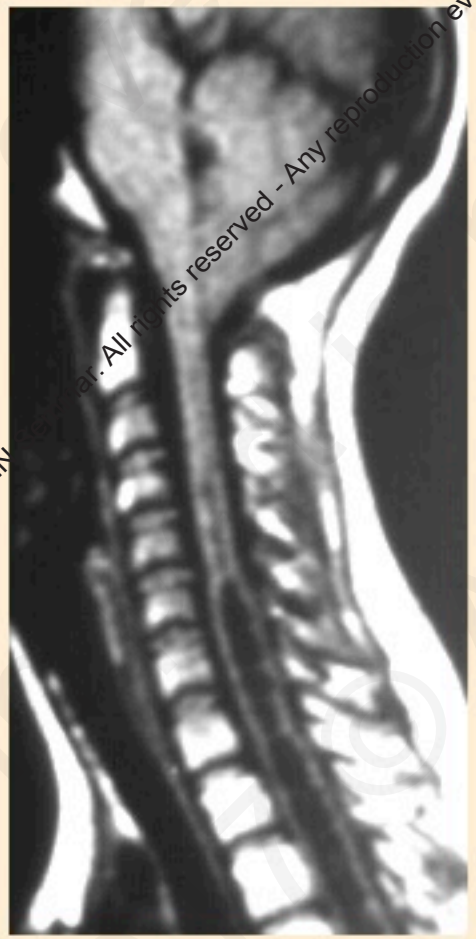
Persistent Syringomyelia in 16 patients After Craniocervical Decompression

Symptom or sign	After 1st Surgery—Syrinx Persisted			
	Absent	Mild	Moderate	Severe
Headache	6	2	6	2
Dysesthetic Pain	4	4	8	0
Subjective Weakness	5	6	0	5
Sensory Loss Reported	2	0	9	5
Impaired Ambulation	5	6	2	3
Weakness By Examination	7	4	3	2
Atrophy	12	1	1	2
Spasticity	10	1	2	3
Ataxia	6	7	0	3
Sensory Loss By Examination	2	3	5	6

Persistent Syringomyelia after Craniocervical Decompression Usually Resolves after Re-Exploration Surgery



Before 1st Surgery

After 1st Surgery

After 2nd Surgery

Persistent Chiari I & Syringomyelia

Before 1st Surgery

1 Year After 1st Surgery

Failure to Open CSF Pathways at the Foramen Magnum

- Pseudomeningocele and Adhesions 4
- Extradural Band and Adhesions 2
- Intradural Adhesions Alone 4
- Not enough bone removed 6

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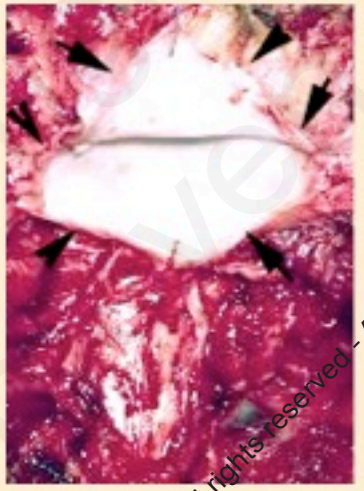
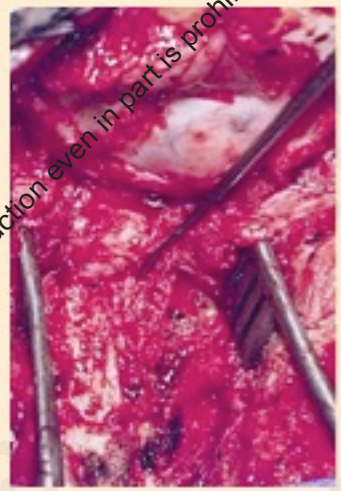
Failure to Open CSF Pathways at the FM

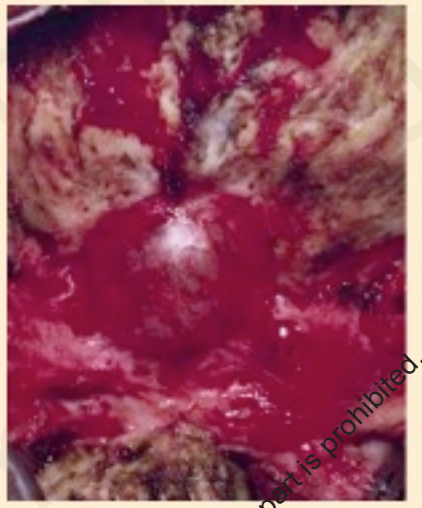


Pseudo-meningocele



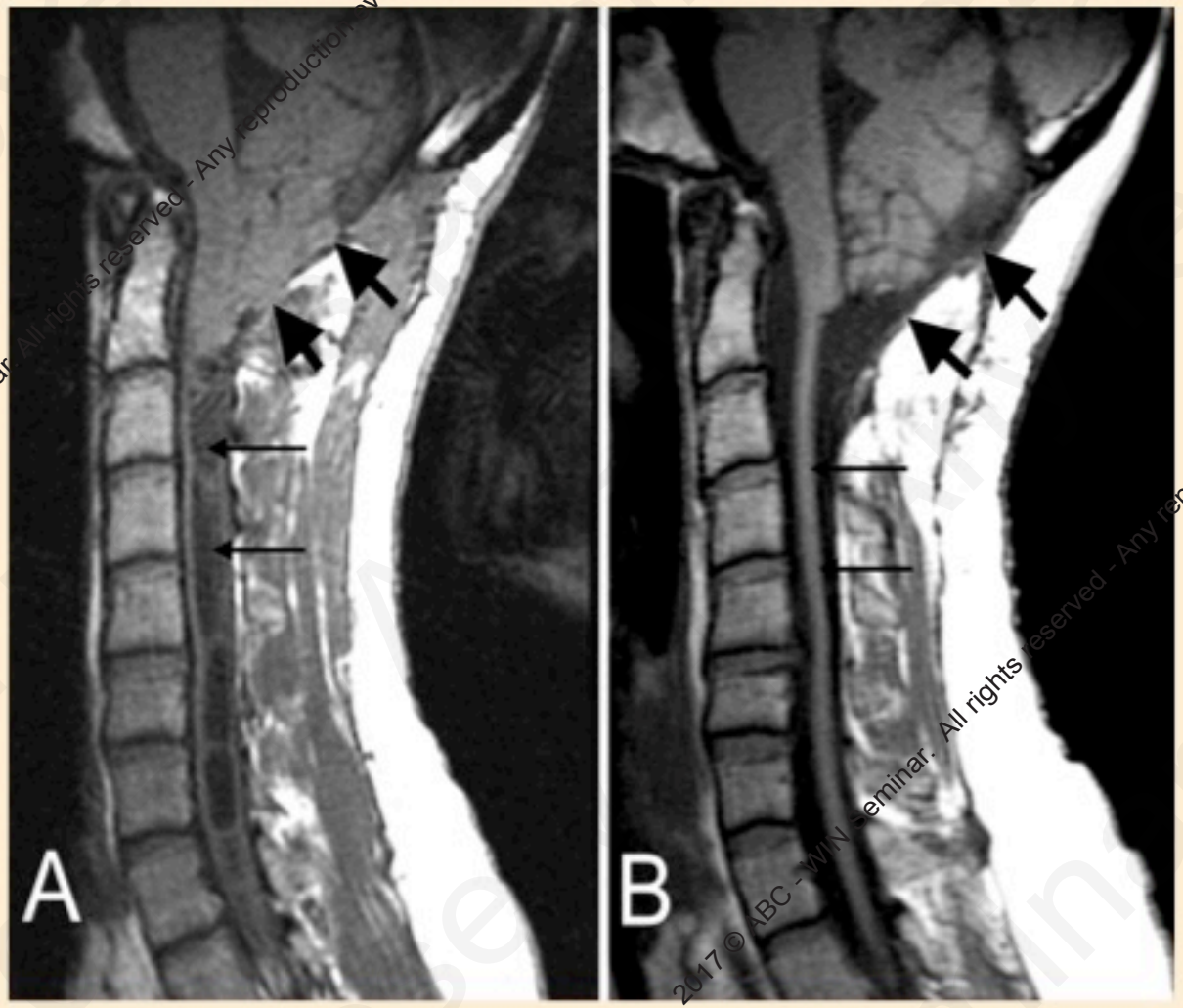
Extradural Band

Adherent Graft

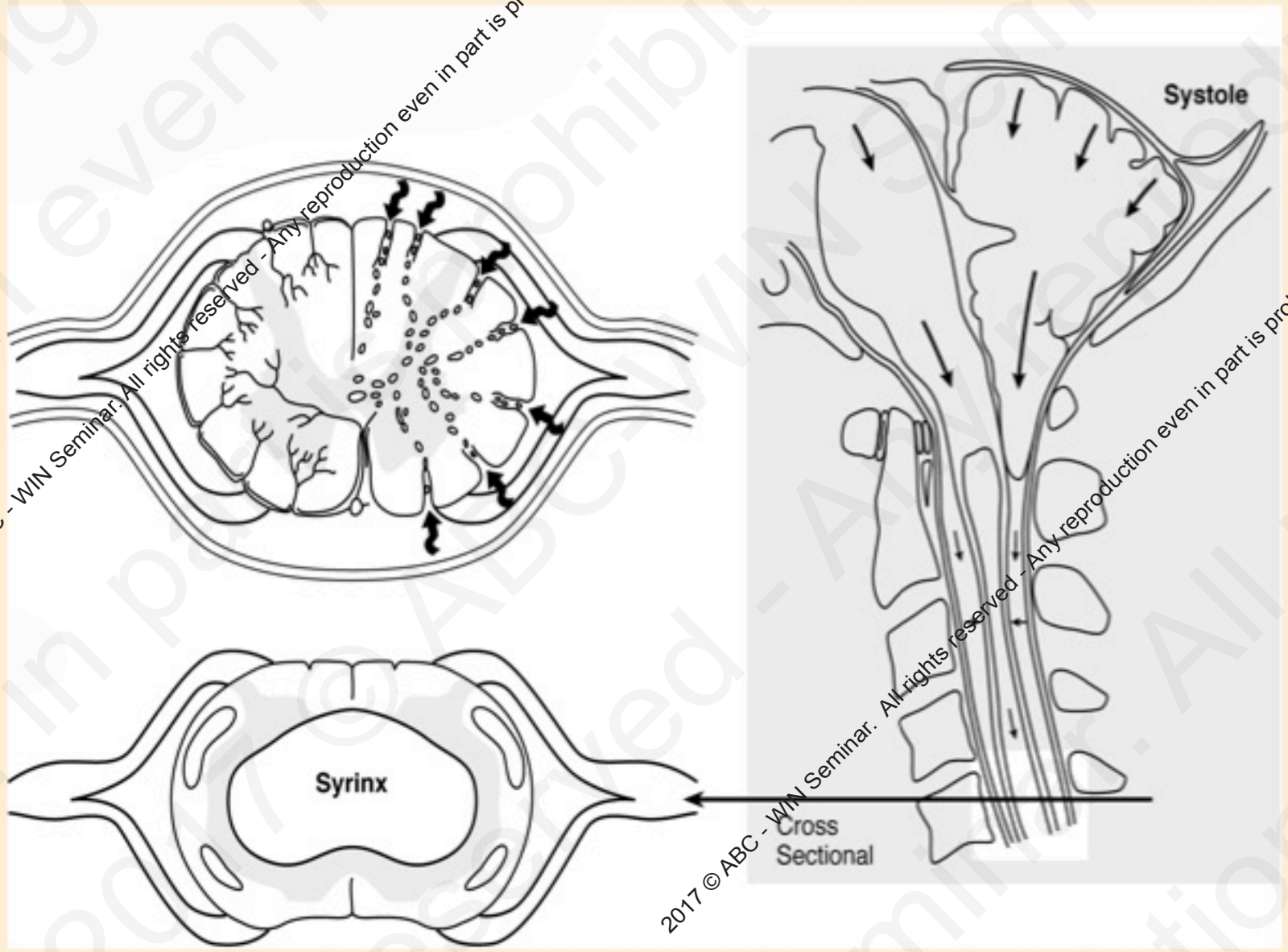


Small Craniectomy

Second Craniocervical Decompression Surgery Resolves Chiari I and Syringomyelia



Chiari I Syrinx Development



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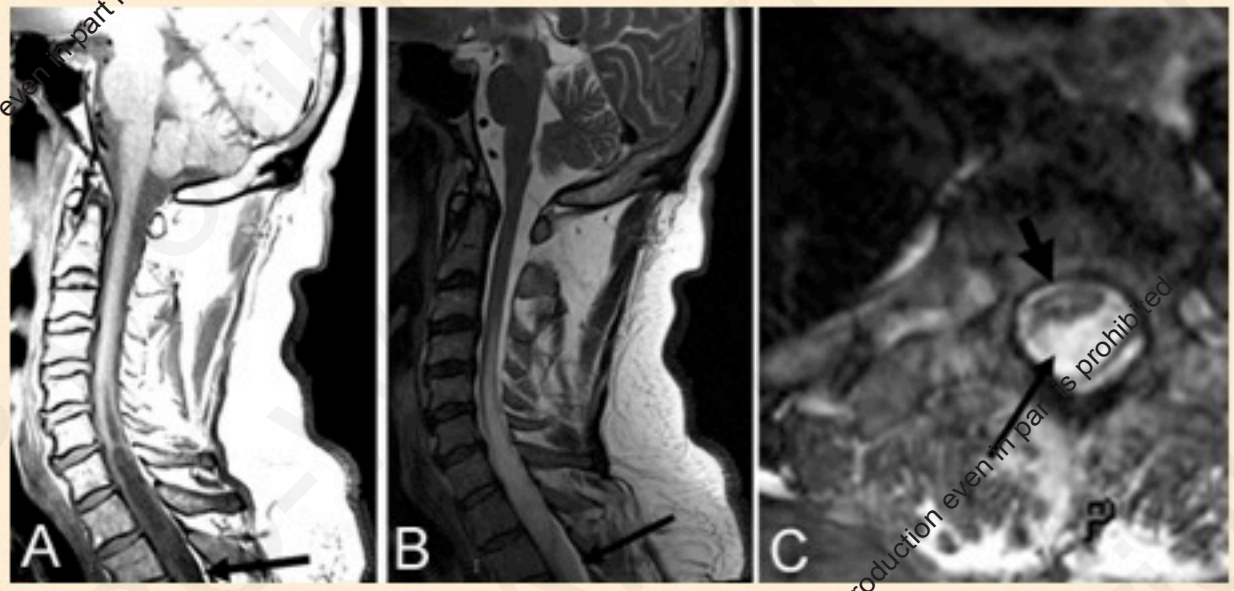
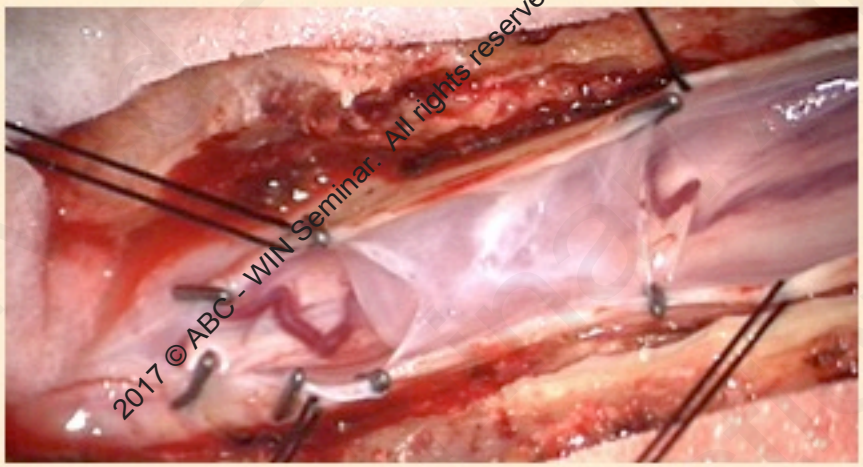
Primary Spinal Syringomyelia

- Associated with spinal lesions and trauma
  - Arachnoid scarring
  - Arachnoid cysts
  - Epidural compression
    - Disc herniation
    - Tumor
    - Spinal deformity
- Ascending level of spinal cord malfunction
- Prevalence 1:50,000
- Standard Rx
  - Laminectomy and duraplasty to restore flow across site of blockage
  - Syrinx shunt placement as a last resort

56 yo with ascending sensory-motor deficit and syringomyelia



Arachnoid cyst associated with a syrinx



Hypothesized Pathophysiology

Syrinx formation

SAS block obstructs pulsatile CSF flow

↓

Shortened spinal canal

↓

Decreased compliance

↓

Increased cervical pulse pressure

↓

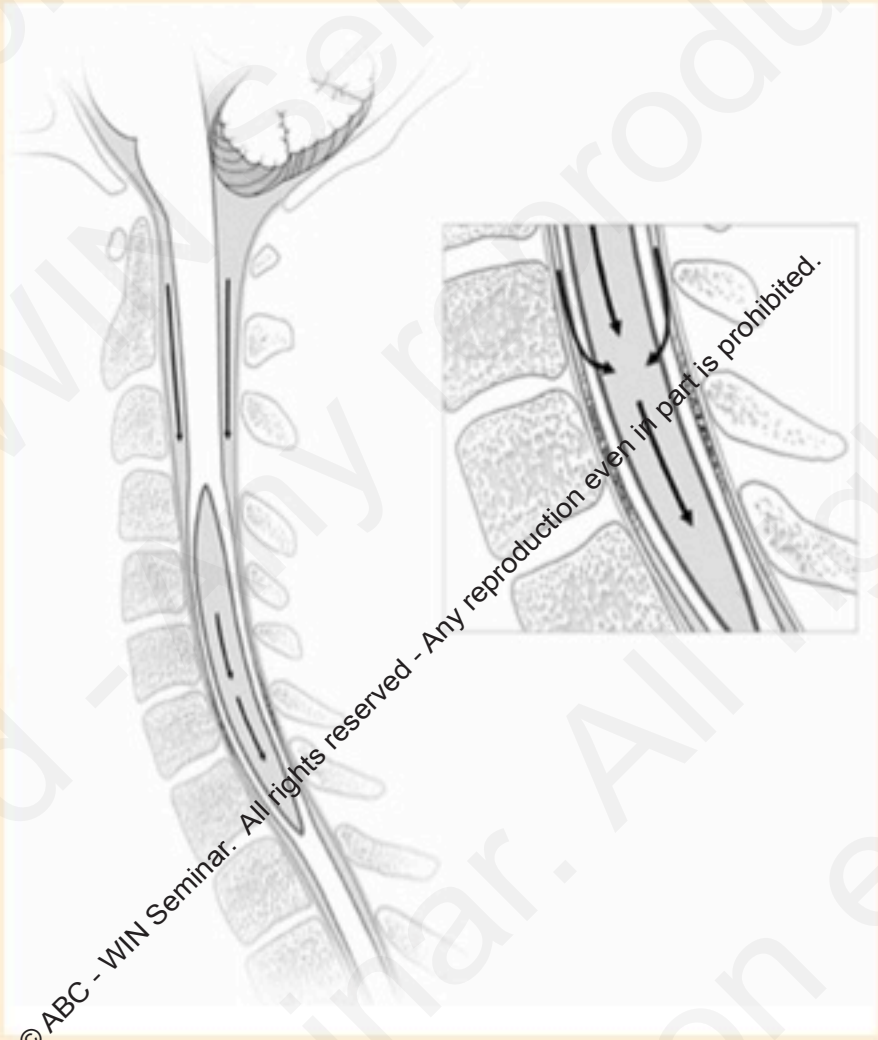
CSF enters spinal cord

↓

Cord fluid coalesces into syrinx

↓

Pulsation extends syrinx



Obstructed CSF flow leading to syrinx formation

Study Population

- Primary Spinal Syringomyelia: 36 patients
  - Post-traumatic: 18
  - Post-meningitic: 2
  - Postsurgical scarring: 1
  - Pantopaque: 1
  - No previous event: 14
- Previous spinal surgery: 22 patients
  - Previous syringomyelia surgery 11
- Age at surgery: 43.3 ± 9.1 years
- Follow-up time: 4.3 ± 3.2 years

Patient presentation before surgery

Clinical signs and symptoms before surgery

Symptom/Sign	No. of patients	%
sensory deficit	36	100
subjective weakness	29	81
dysesthetic pain	24	67
spasticity	22	61
ambulation		
normal (0)	19	53
slow and unsteady gait (1)	8	22
walking with aid (2)	1	3
nonambulatory (3)	8	22
ASIA score		
75-100	28	78
50-74	2	5
<50	6	17

MRI findings before surgery

Syrinx Size

Parameter	Measurement
syrinx	
diameter (mm)	7.1 ± 2.8
length (cm)	17.3 ± 12.2
length (levels)	9.1 ± 5.2

Syringomyelia as a late sequela of thoracic fracture-dislocation

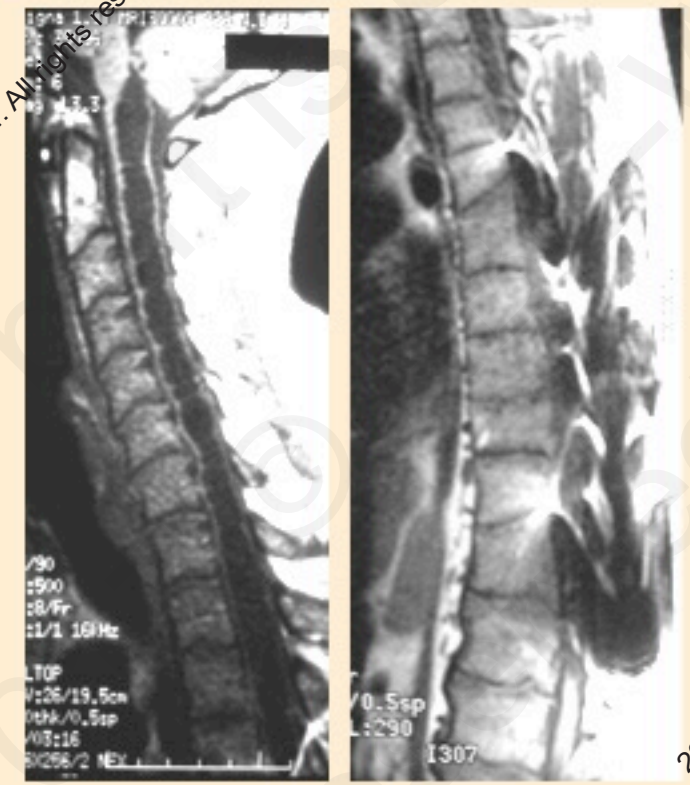
Post-infectious syringomyelia

Myelo-CT

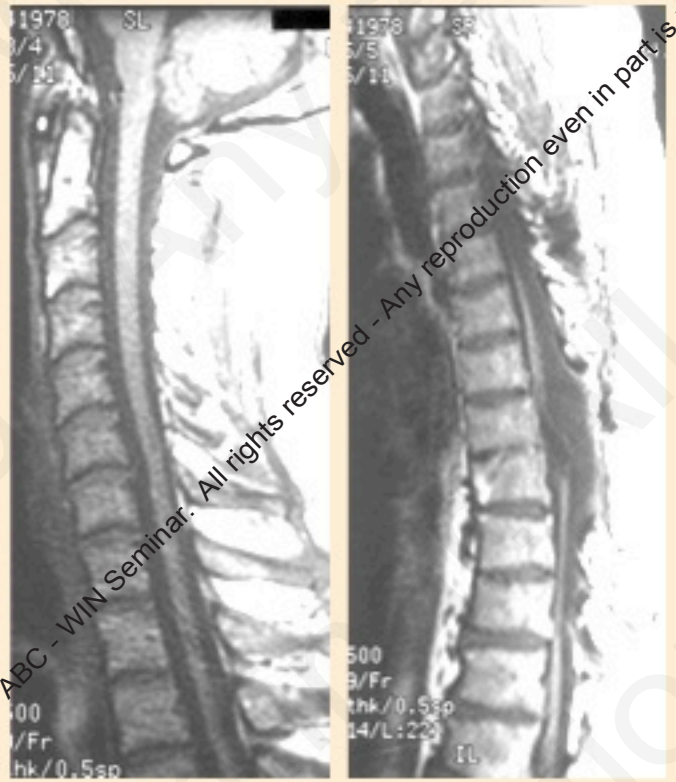
Intraoperative Ultrasonography

MRI findings after surgery

	Before Surgery	After Surgery	p Value
syrinx			
diameter (mm)	7.1 ± 2.8	3.6 ± 3.8	<0.0001
length (cm)	17.3 ± 12.2	11.2 ± 12.4	<0.0002
length (levels)	9.1 ± 5.2	6.1 ± 5.4	<0.0001



Before Surgery



After Surgery

Clinical outcome after surgery (n = 36)

Symptom/Sign			
No. of patients (%)	Initial	3 months	4.3 yr. μ
sensory deficit	36 (100%)	33 (92%)	33 (92%)
subjective weakness	29 (81%)	22 (61%)	22 (61%)
dysesthetic pain	24 (67%)	21 (58%)	22 (61%)
spasticity	22 (61%)	19 (53%)	18 (50%)
ambulation			
normal (0)	19 (53%)	17 (47%)	13 (36%)
Slow, unsteady gait (1)	8 (22%)	9 (25%)	6 (17%)
walking with aid (2)	1 (3%)	3 (8%)	3 (8%)
nonambulatory (3)	8 (22%)	7 (20%)	14 (39%)
ASIA score			
75-100	28 (78%)	28 (78%)	25 (69%)
50-74	2 (5%)	4 (11%)	6 (17%)
<50	6 (17%)	4 (11%)	5 (14%)

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Clinical results vs. radiographic and physiologic findings

- Good Long-Term Clinical Outcome (n = 28)
  - Decrease in syrinx size
    - Diameter (mm)
      - Before surgery: 7.1 ± 2.9
      - After surgery: 2.0 ± 1.9
    - Length (cm)
      - Before surgery: 14.4 ± 11.4
      - After surgery: 6.5 ± 8.0
  - Normal pressure transmission (mmHg/sec)
    - Before surgery: 5.2 ± 3.6
    - After surgery: 5.9 ± 2.8
    - Normal: 6.3 ± 2.0

- Poor Long-Term Clinical Outcome (n = 8)
  - Syrinx size unchanged or larger
    - Diameter (mm)
      - Before surgery: 7.1 ± 2.8
      - After surgery: 7.3 ± 4.6
    - Length (cm)
      - Before surgery: 23.7 ± 11.9
      - After surgery: 22.1 ± 13.9
  - Impaired pressure transmission (mmHg/sec)
    - Before surgery: 1.8 ± 1.7
    - After surgery: 2.4 ± 2.6
    - Normal: 6.3 ±2.0

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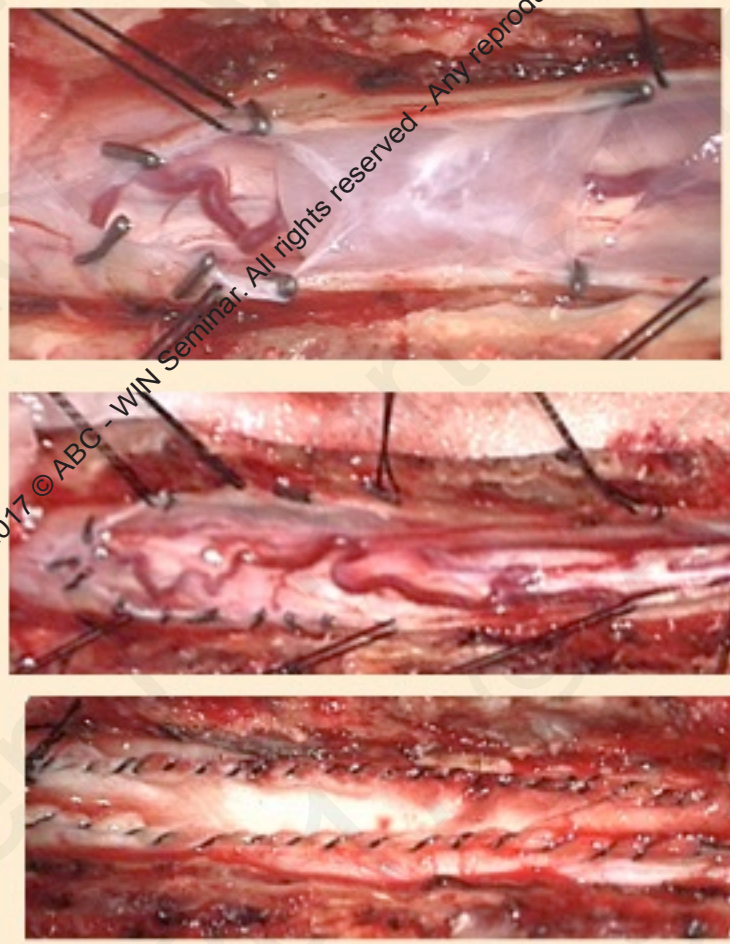
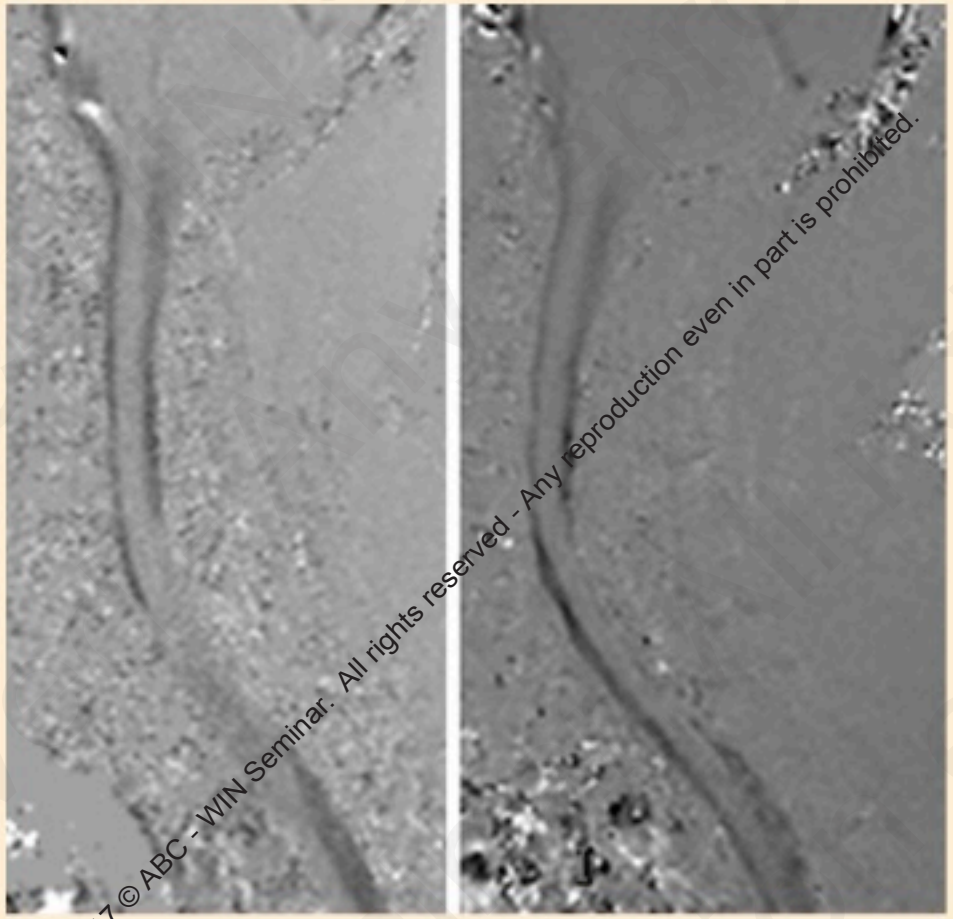
Surgical relief of the obstruction of the spinal subarachnoid space



Phase-Contrast MRI

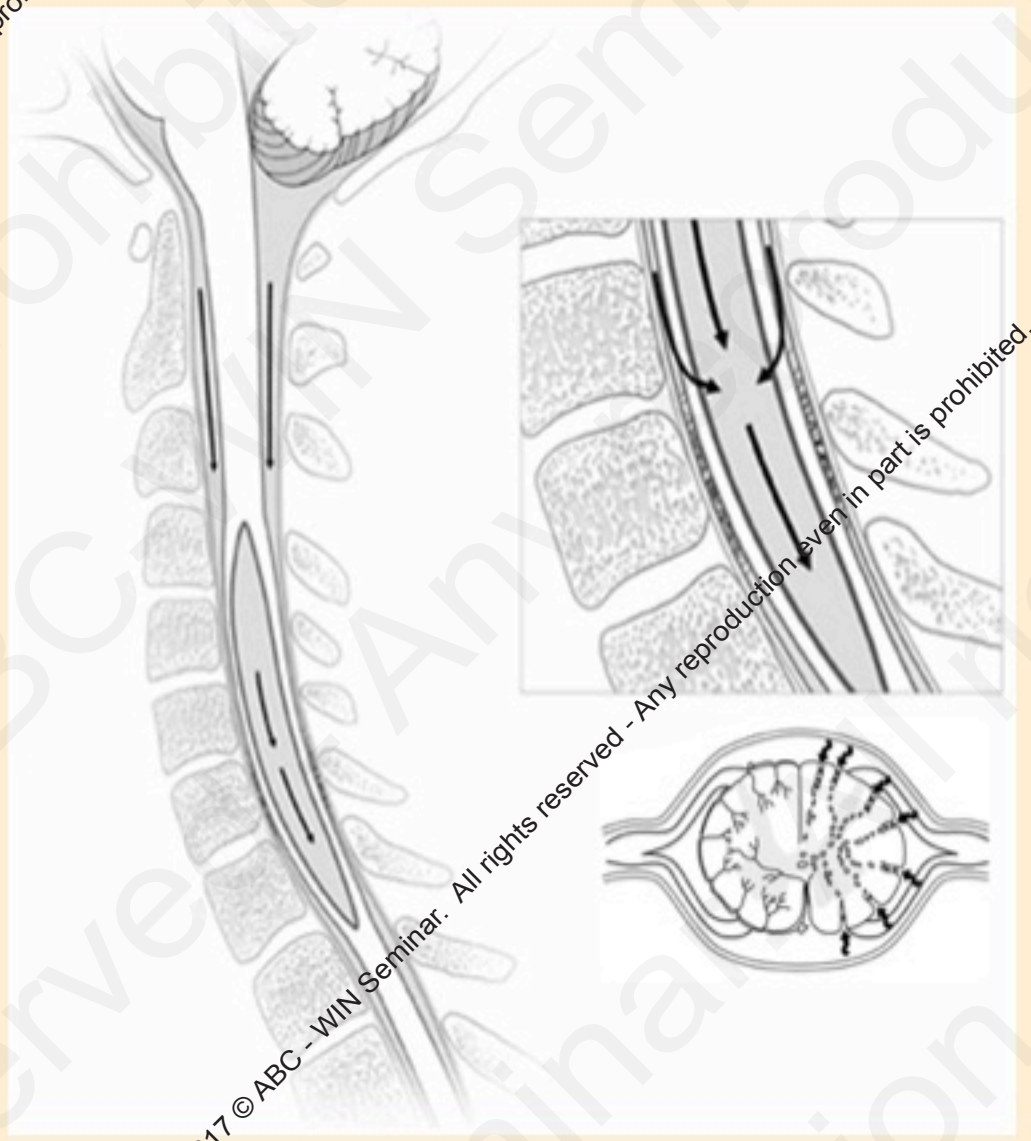


Before Surgery After Surgery

Opening the CSF obstruction resolves syringomyelia by reversing its mechanism of formation

Before Surgery

After Surgery



Primary Spinal Syringomyelia

- Focal Arachnoiditis or Trauma
  - Laminectomy and duroplasty usually opens the spinal subarachnoid space and resolves syringomyelia
  - Relief of myelographic block is the best determinant of long-term clinical outcome after laminectomy and duroplasty

- Extensive Arachnoiditis
  - Laminectomy and duroplasty often ineffective in opening the spinal subarachnoid space and treating syringomyelia; syrinx shunting is usually necessary to drain the syrinx

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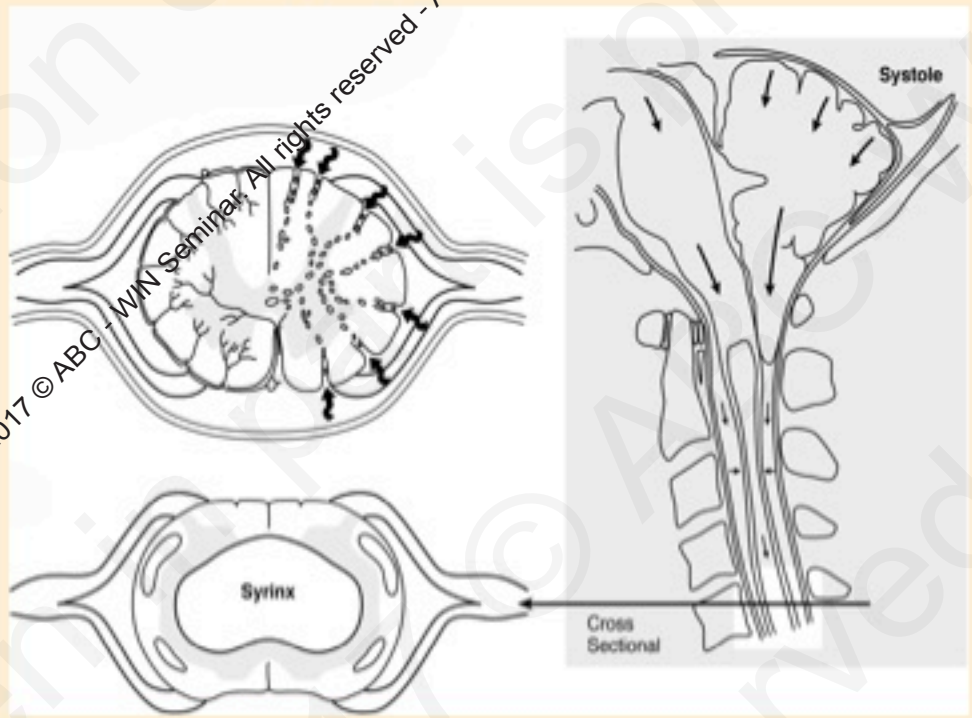
Unifying Theory of Syringomyelia Development

Chiari I



Primary Spinal

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- Bioengineering
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