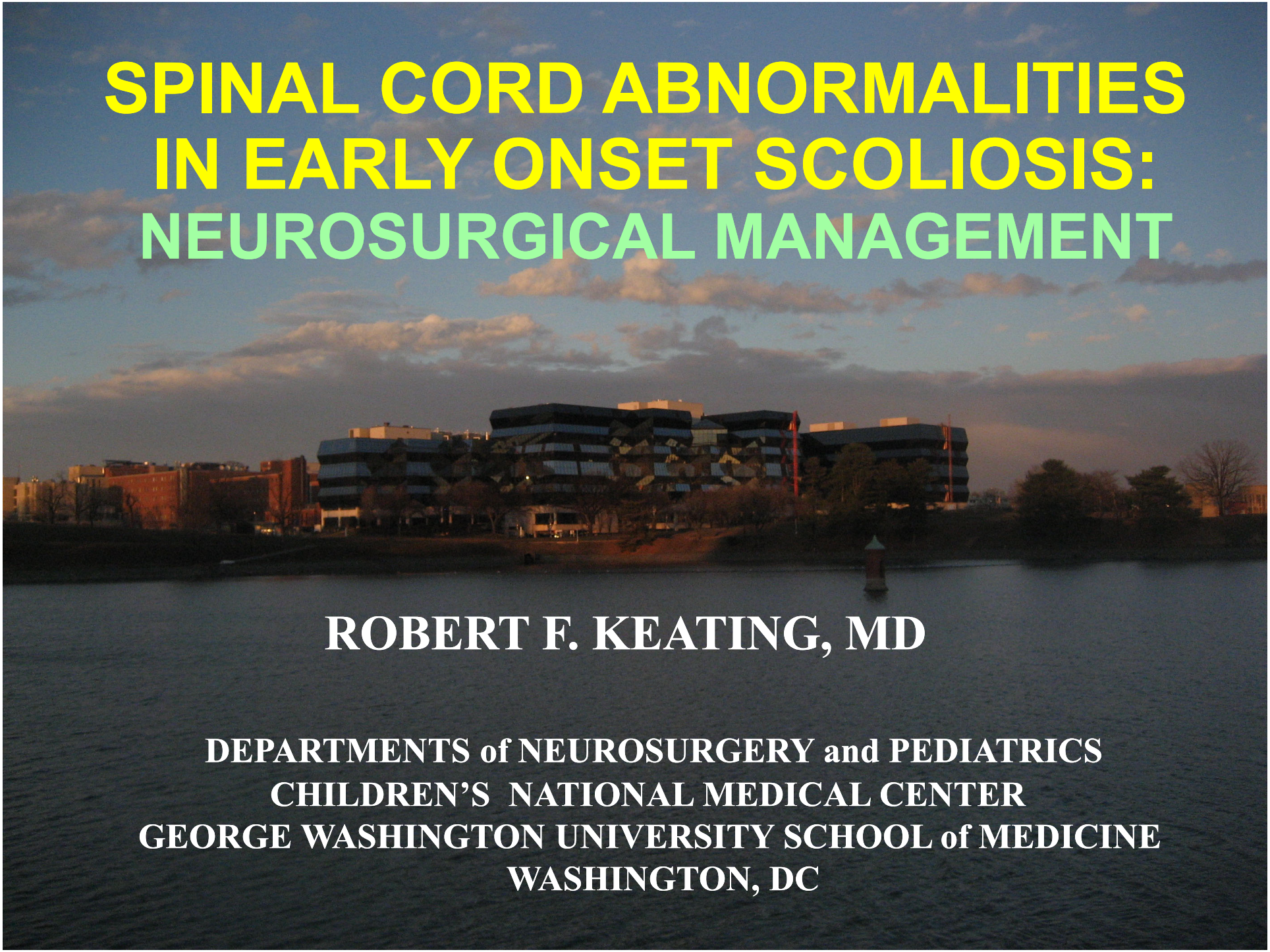

SPINAL CORD ABNORMALITIES IN EARLY ONSET SCOLIOSIS: NEUROSURGICAL MANAGEMENT

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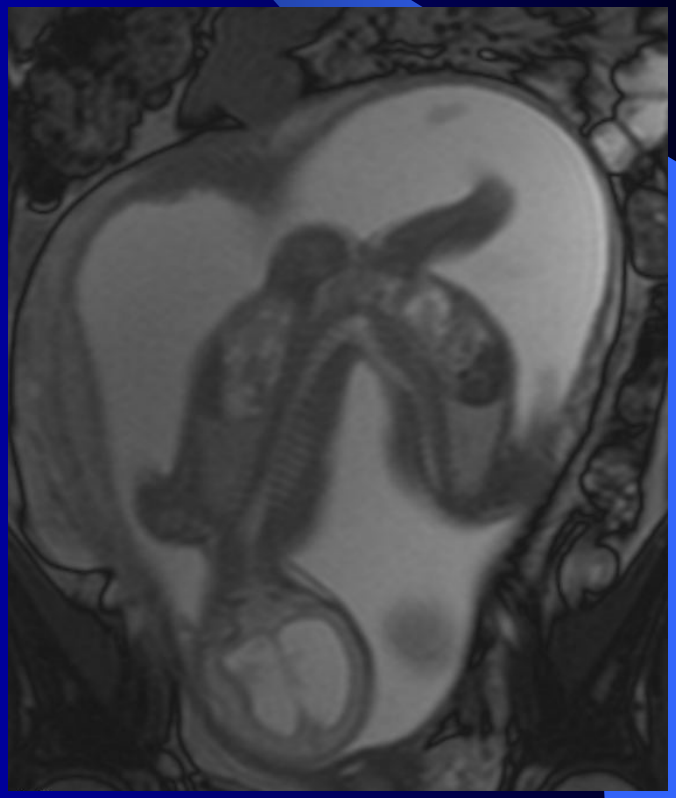
SCOLIOSIS

ETIOLOGY

NEUROGENIC

- SPINAL DYSRAPHISM / TETHERED CORD
  - MYELOMENINGOCELE
  - LIPOMYELOMENINGOCELE
  - SPLIT CORD MALFORMATIONS
  - DERMAL SINUS TRACT
  - ANORECTAL MALFORMATION
- SPINAL CORD TUMOR
- HYDROMYELIA
  - CHIARI
  - TETHERED CORD
  - POST TRAUMATIC / HEMORRHAGIC
  - IDIOPATHIC

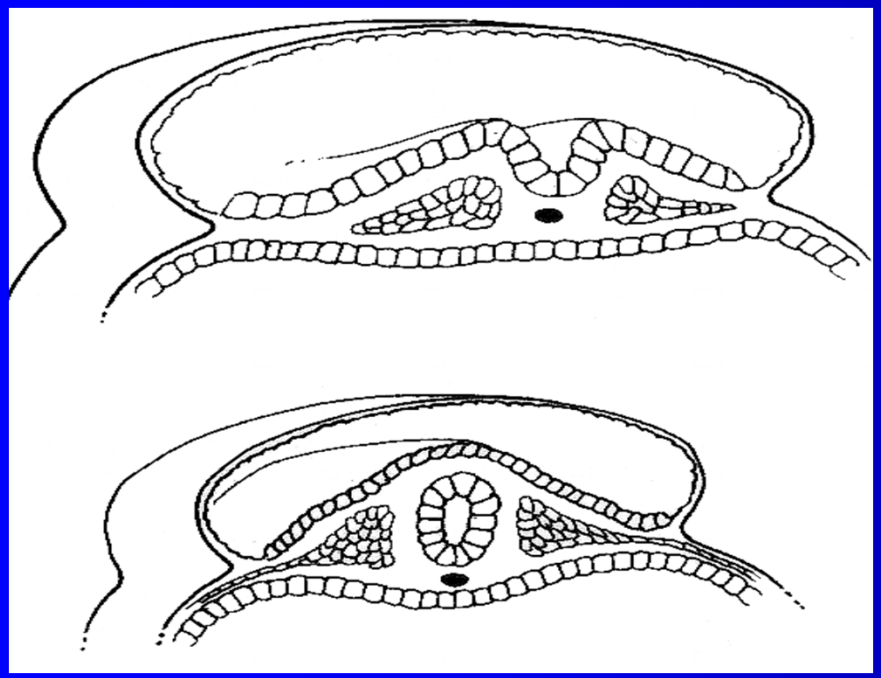


SPINAL DYSRAPHISM

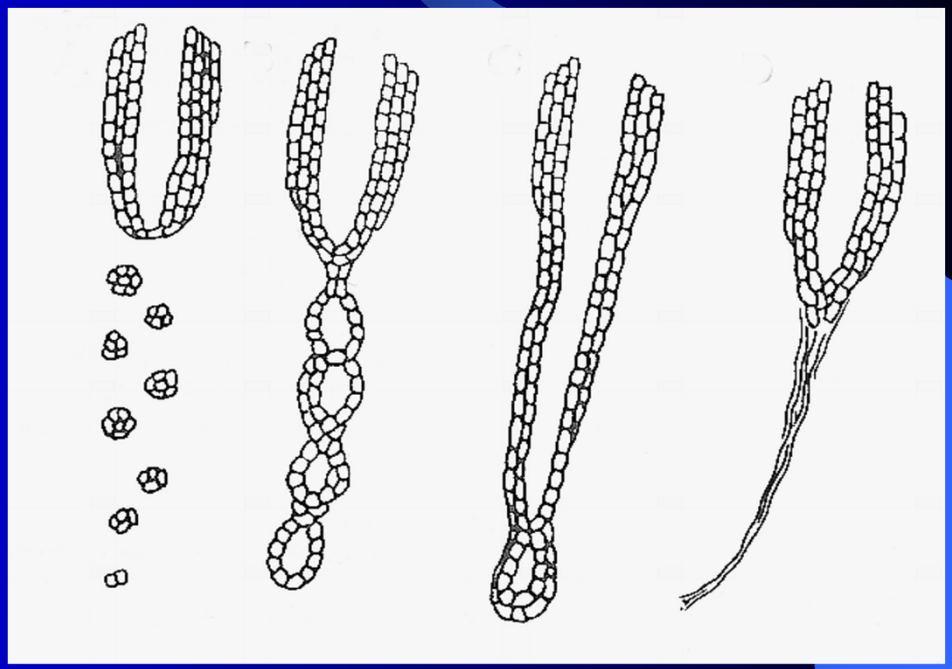
- MYELOMENINGOCELE
- LIPOMYELOMENING.
- SPLIT CORD MALFORM.
- FILUM DYSGENESIS
- DERMAL SINUS TRACT
- SACRAL DYSGENESIS
- INTRAMED CYSTS
- ANORECTAL MALFORMA

SPINAL DYSRAPHISM

EMBRYOGENESIS

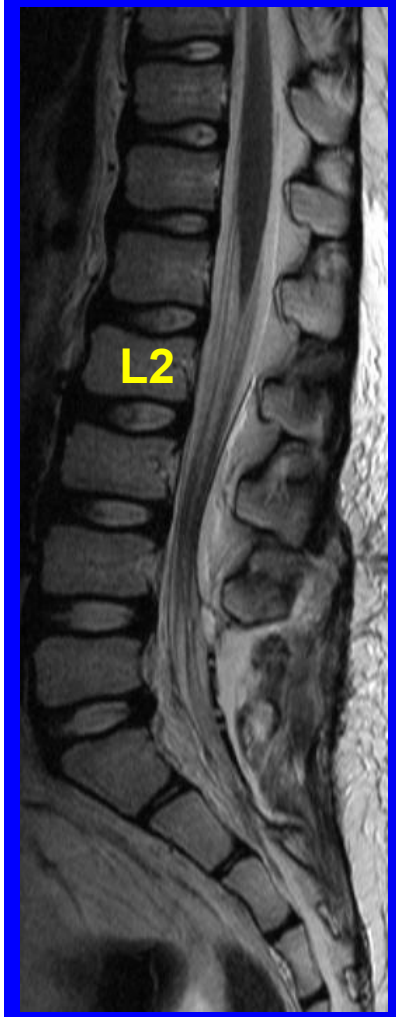


Formation of neural tube



Caudal Regression

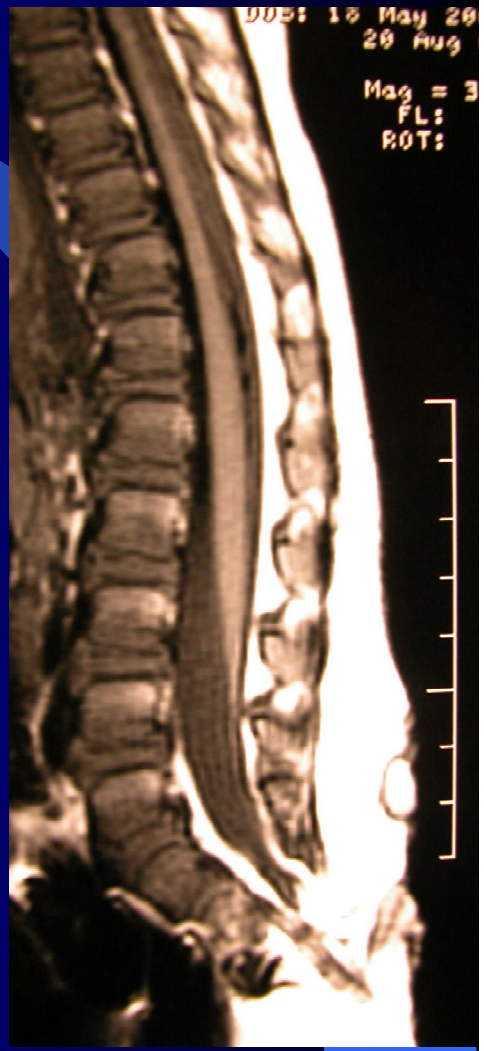
TETHERED SPINAL CORD



DEFINITION

AN ABNORM. LOW LYING CONUS WHOSE NL ASCENT IS TETHERED SECONDARY TO A MULTITUDE OF DYSRAPHIC CONDITIONS.

RADIOGRAPHIC DX DOES NOT ALWAYS CORRELATE w/ CLINICAL PRESENTATION.

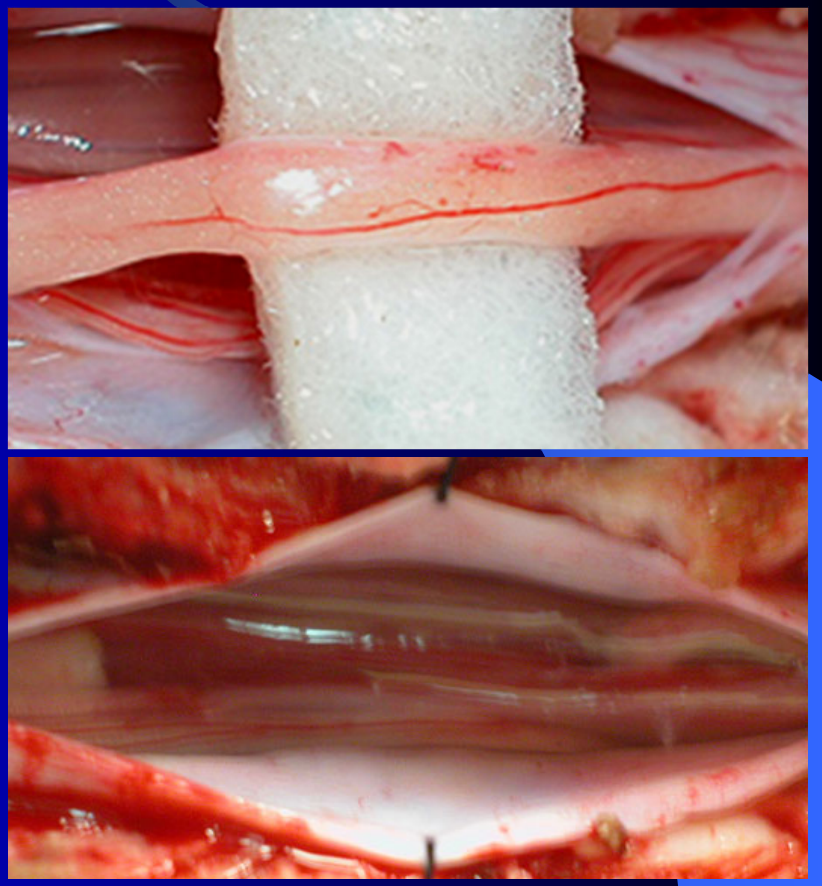


CLINICAL PRESENTATION

- CUTANEOUS MANIFESTATIONS
  - 30% TETH. PTS --> SKIN LESIONS
- LE ASYMMETRY
- NEURO SYMPTOMS
- SCOLIOSIS
- ONSET DURING GROWTH SPURTS

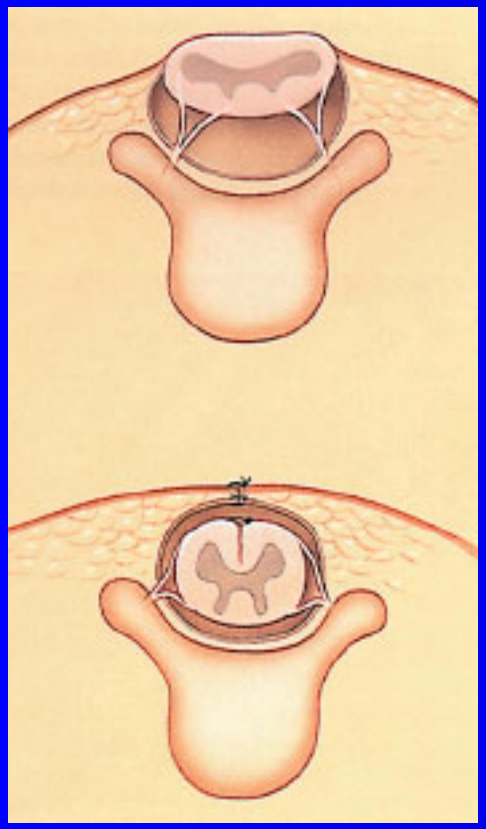
TREATMENT

- FUNDAMENTAL SHIFT IN PHILOSOPHY
  - PROPHYLACTIC
  - R:B RATIO ADVANTAGE
- OBJECTIVE-->RELEASE THE CORD
  - MYELOMENING.--> FREE ADHESIONS
  - DIASTEMATO-->REMOVE SPURR/ADHESIONS
  - THICKENED FILUM-->CUT
  - LIPOMYELO.--> RESECT LIPOMA/ FREE ADHESIONS



SPINAL DYSRAPHISM

MYELOMENINGOCELE



-MYELOMENINGOCELE
-MENINGOCELE
-MYELOCYSTOCELE
-MYELO MANQUE

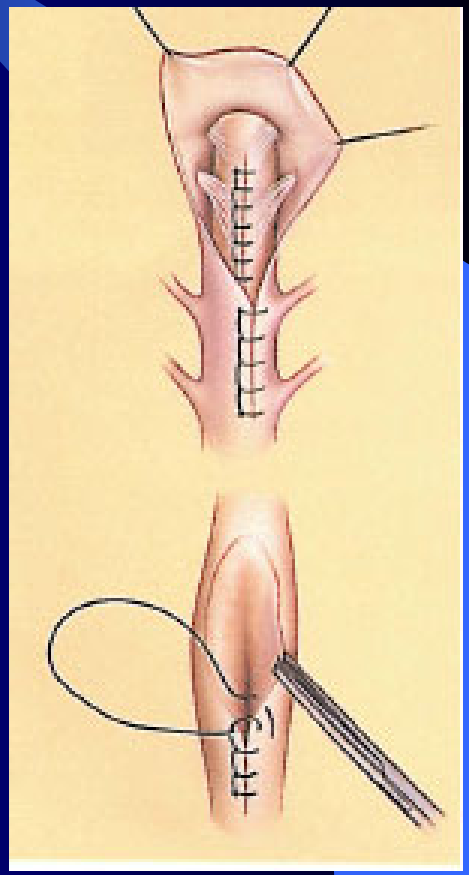
MYELOMENINGOCELE

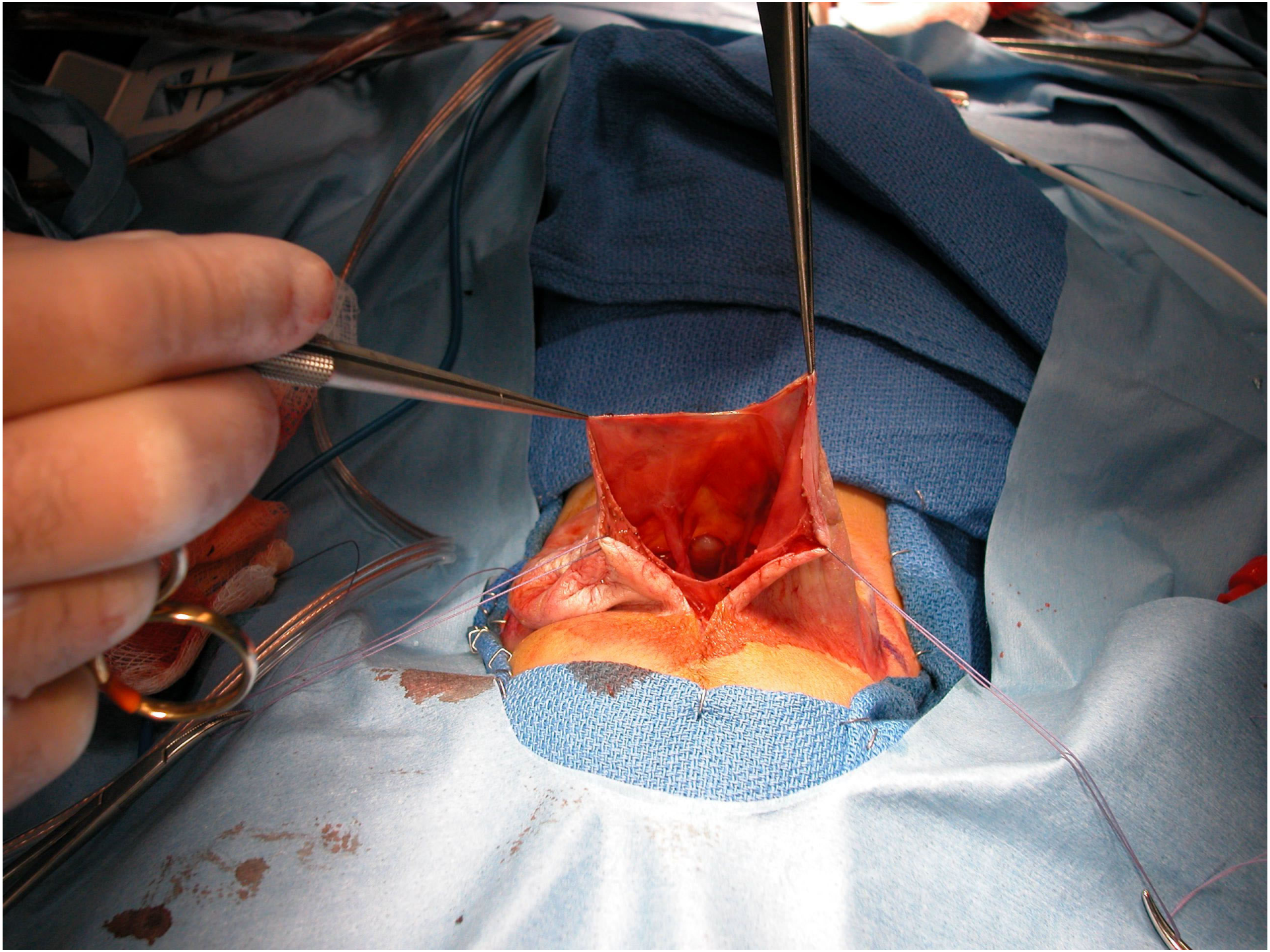
INCIDENCE

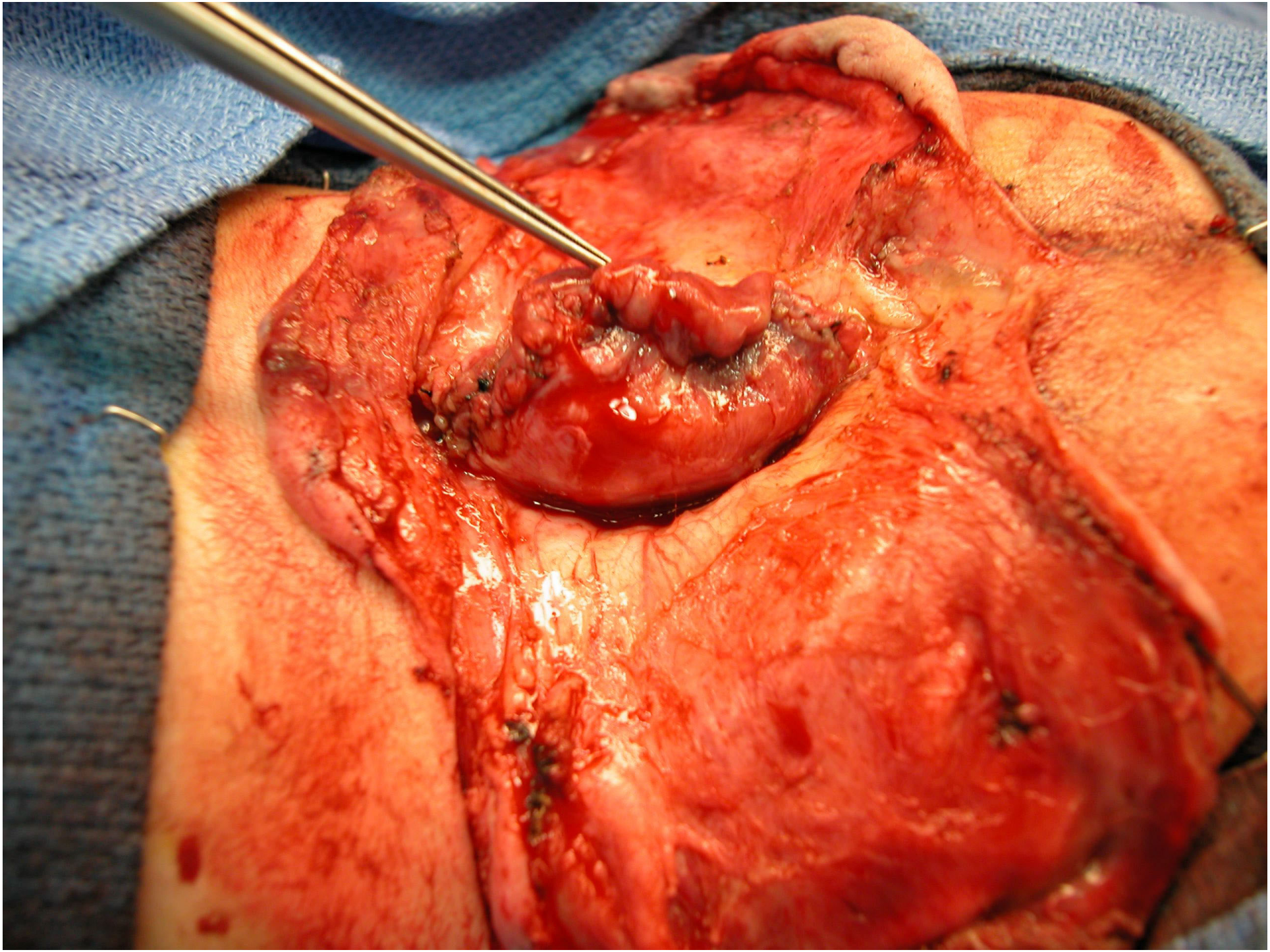
- APPEARS TO BE DECREASING IN US
  - 5.9 / 100,000 BIRTHS (‘84), 3.2 IN ‘92, 2.9 ‘03 (sweden)
  - IMPROVED MATERNAL NUTRITION…FOLATE
  - BETTER PRENATAL SCREENING / DIAGNOSIS
  - SELECTIVE TERMINATION OF PREG.

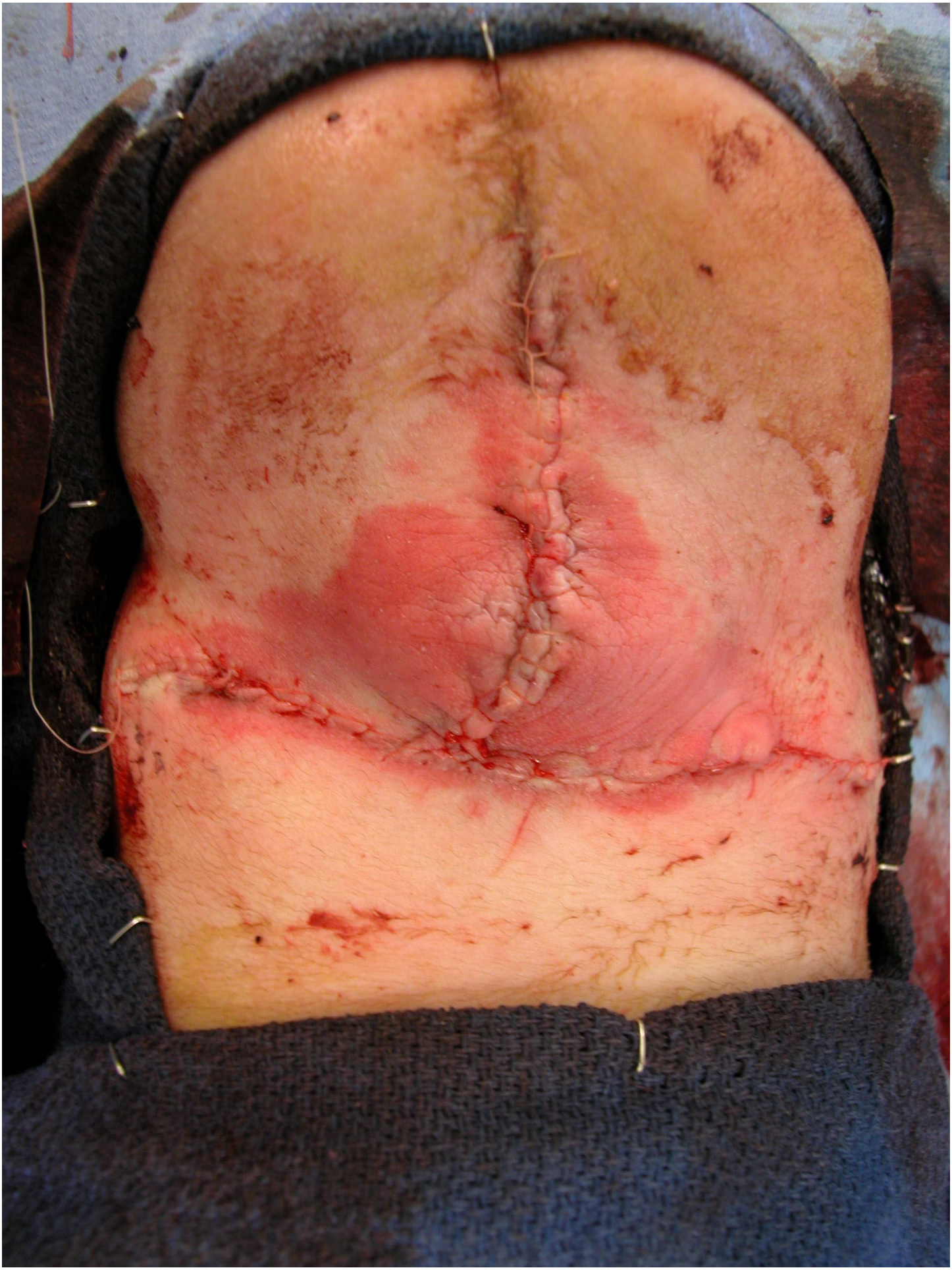
SURGICAL OBJECTIVES

- PROTECTION / PRESERVATION EXPOSED NEURAL TISSUE
- IDENTIFICATION OF PLACCODE
- RECONSTR. OF NEURAL TUBE
- UNTETHER CORD IF NECESS.
- DURAL CLOSURE
- ADEQ. FASCIAL / SQ CLOSURE





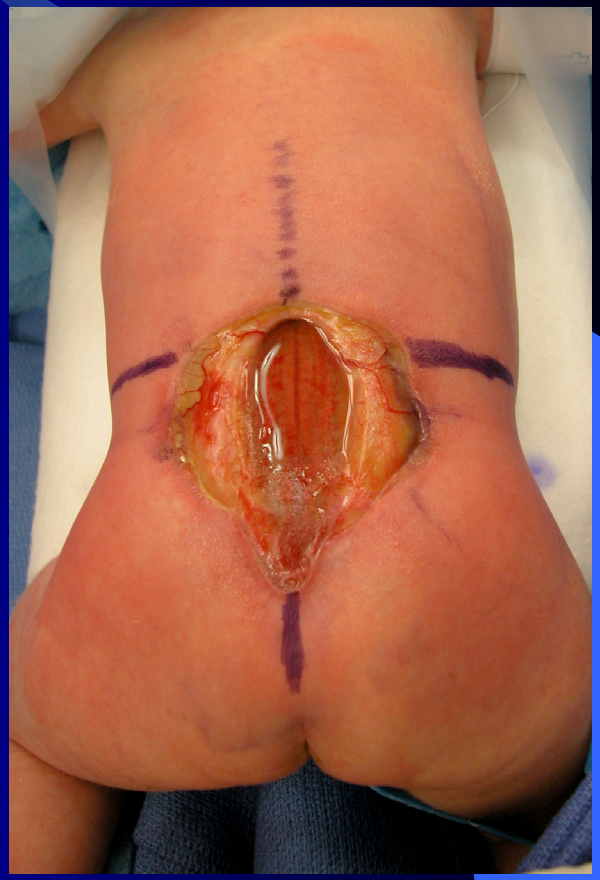




SCOLIOSIS

MYELOMENINGOCELE

- KYPHOSIS
  - ? DUE TO UNOPPOSED ABD MUSC IN UTERO
    - POST ELEMENTS FREQ DEFICIENT
  - ANT ELEMENTS OFTEN WEDGED
  - APEX USUALLY AT L1-2
- LORDOSIS
  - COMMONLY SEEN IN MMC PTS
  - OFTEN COMPENSAT. OFFSET. INSTAB.
    - PELVIC OBLIQ.
    - DISLOCATED HIP



SCOLIOSIS

MYELOMENINGOCELE

CONGENITAL

- UNCOMMON (20%)
- DEFORMITY PRESENT AT BIRTH
- FREQ. ASSOCIATED KYPHOSIS
- VTB ANOMALIES (ANTERIOR)
  - hemivertabrae
  - fused anterior elements
- UNILAT BARS
- OFTEN RIGID--> EARLY, SLOW PROGRESSION

SCOLIOSIS

DEVELOPMENTAL

- MORE COMMON
- MECH. INSTAB DUE TO MUSC PARALYSIS / VTB (POST) ABNL
- LESS RIGID than CONGEN CURVES
- CURVES INCREASE with UPRIGHT POSTURE / GRAVITY, TETHERING
- NEURO IMPAIRMENT DUE TO HYDRO-MYELIA / TETHERED CORD

MYELOMENIGOCELE

SCOLIOSIS

MYELOMENINGOCELE

- PREV. RELATED TO NEURO LEVEL
  - THOR. / UPPER LUMB. (95%) ---> SCOLI
  - LUMBAR / SACRAL (40%) ---> SCOLIOSIS
- SUSPECT TETHERED CORD
  - IN PTS <6 YRS w/ LUMBAR LESION and SCOLIOSIS
  - CURVES INCREASING > 10-12 degrees / yr

SCOLIOSIS

TETHERED CORD

REIGEL et al., Ped Neurosurg,1994;20:30.

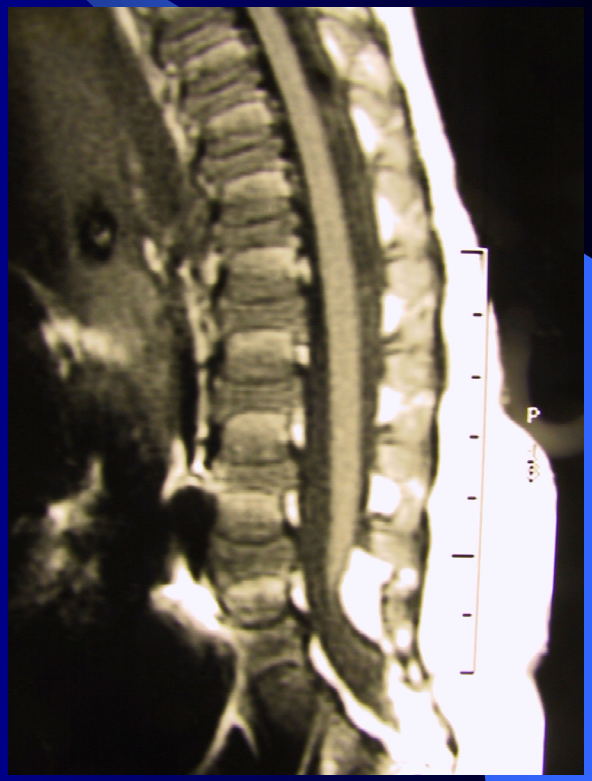
262 pts w/ tethered cd release med records reviewed over 20 years (2369 serial spine films--Cobb mm)

McLONE et al. Ped NSurg 1990/1991;16:8-13.

30 pts with prior MM repair with scoliosis as presenting sign for tethered cord.

LIPOMYELOMENINGOCELE

- TRANSITIONAL, DORSAL, FILUM
- NO HYDRO / CHIARI
- FILUM OFTEN INVOLVED
- OFTEN NL NEURO except B/B
  - FREQUENTLY ASYM LE
  - CUTANEOUS LESIONS ~ 60%

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TR:466
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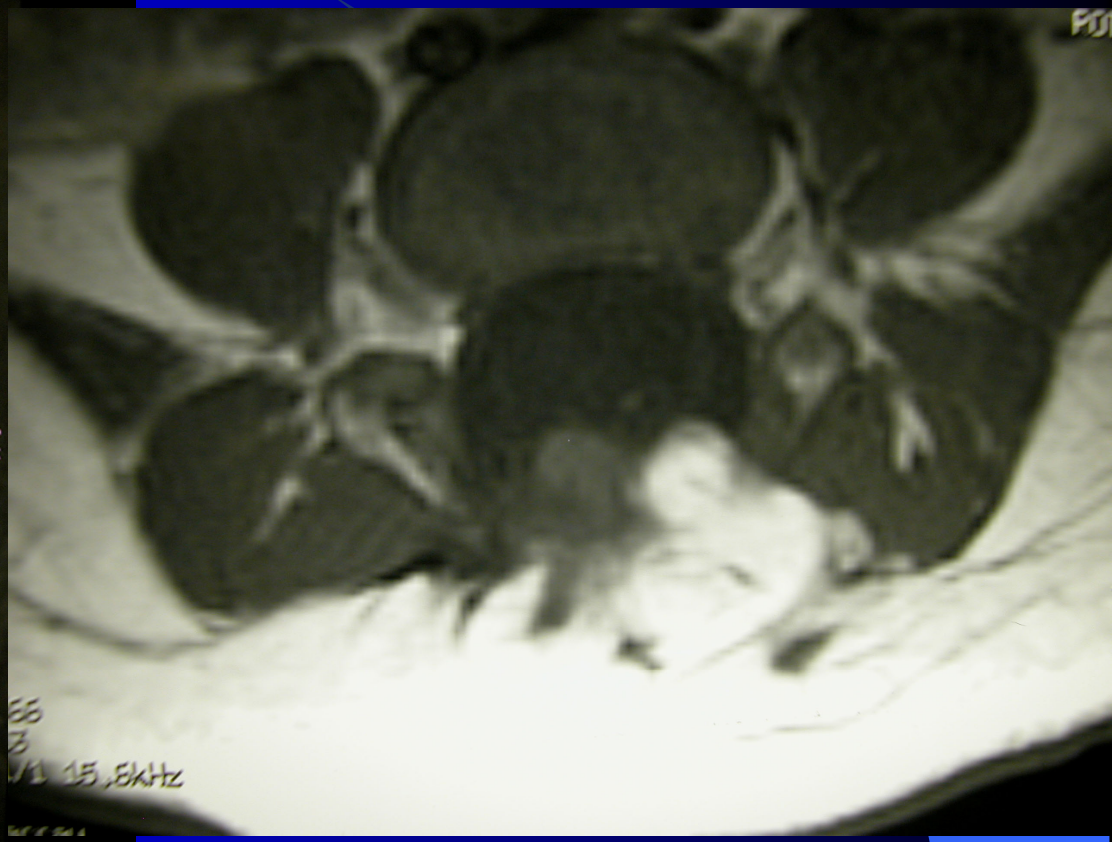
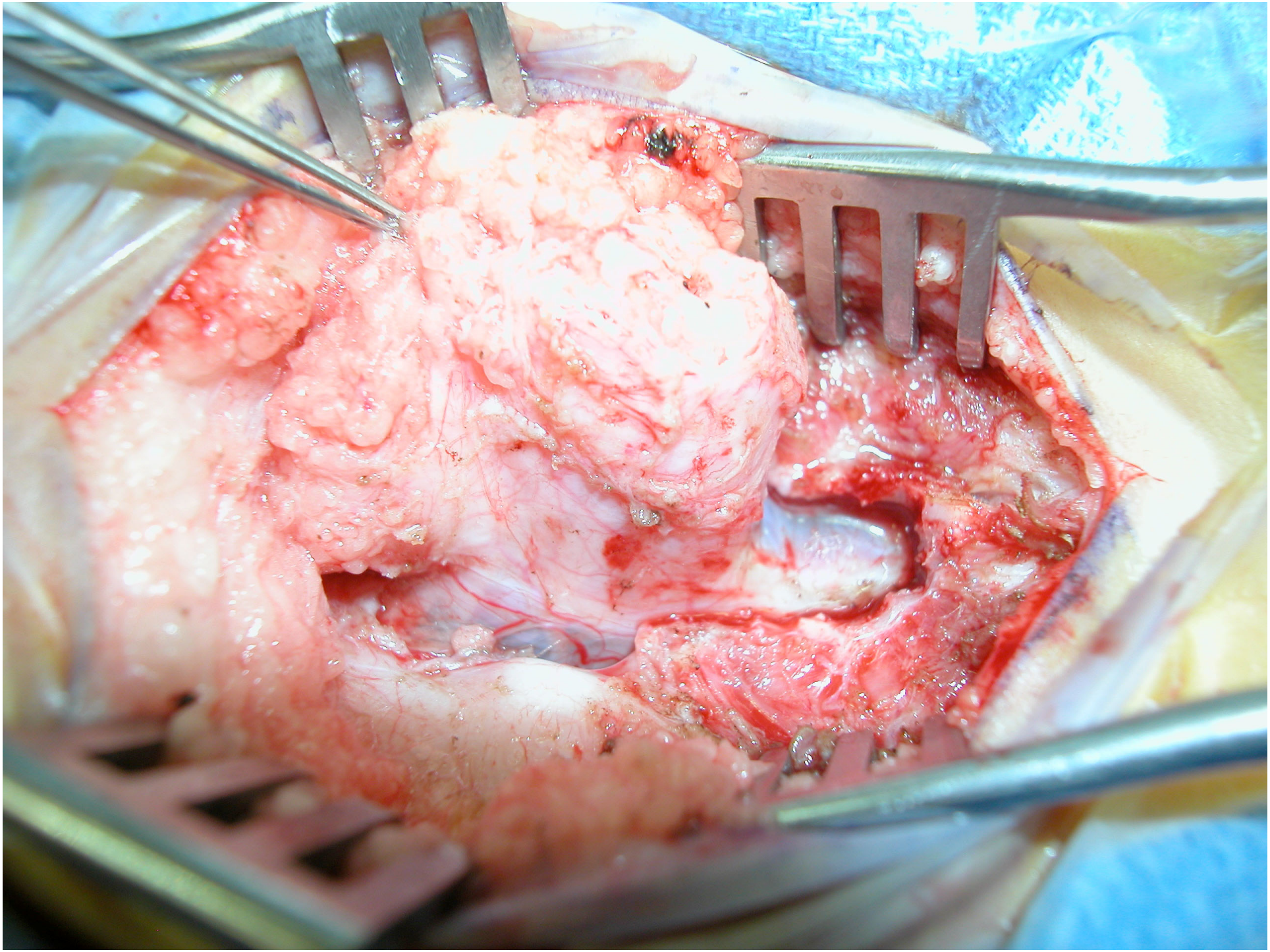
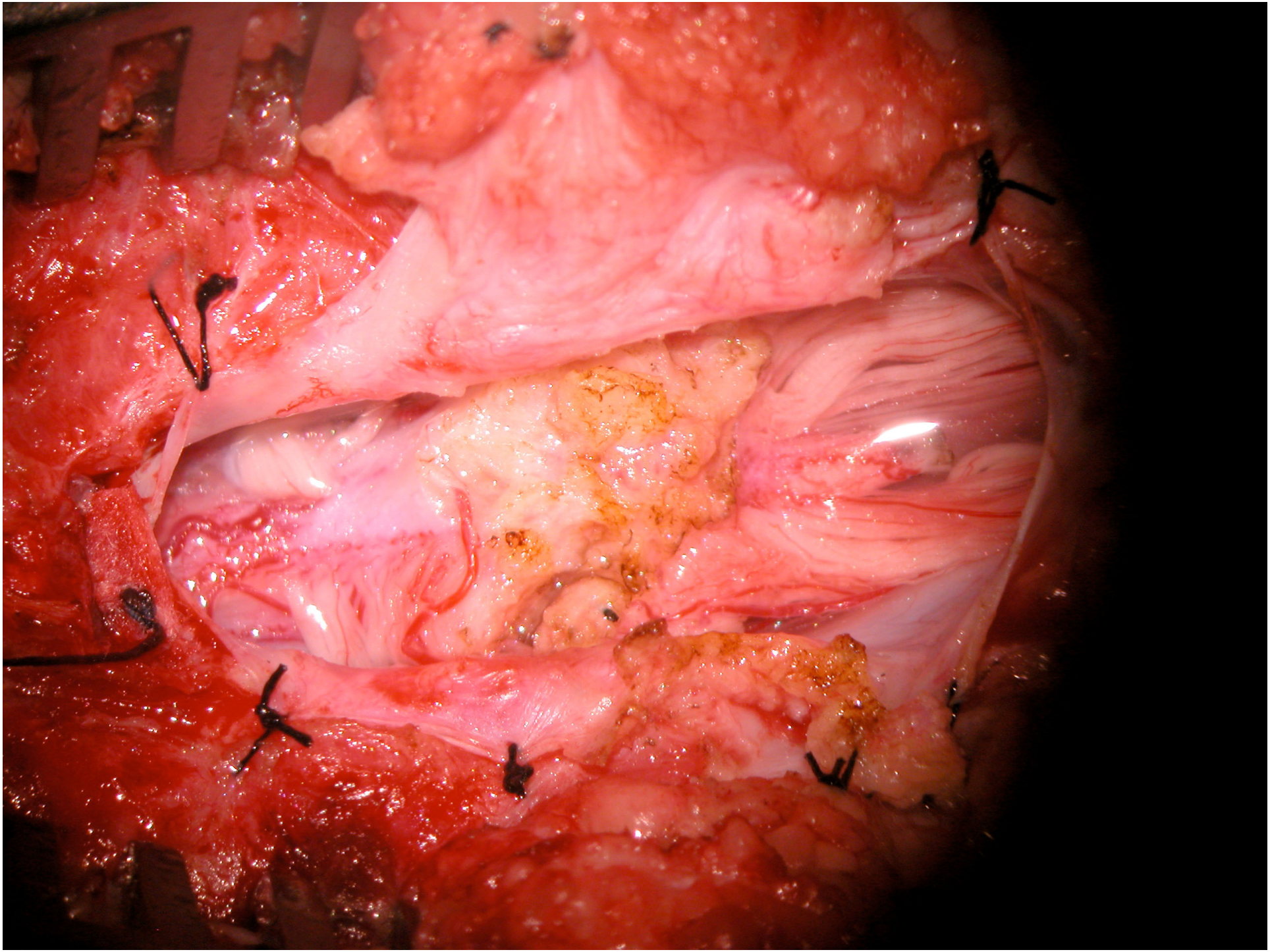
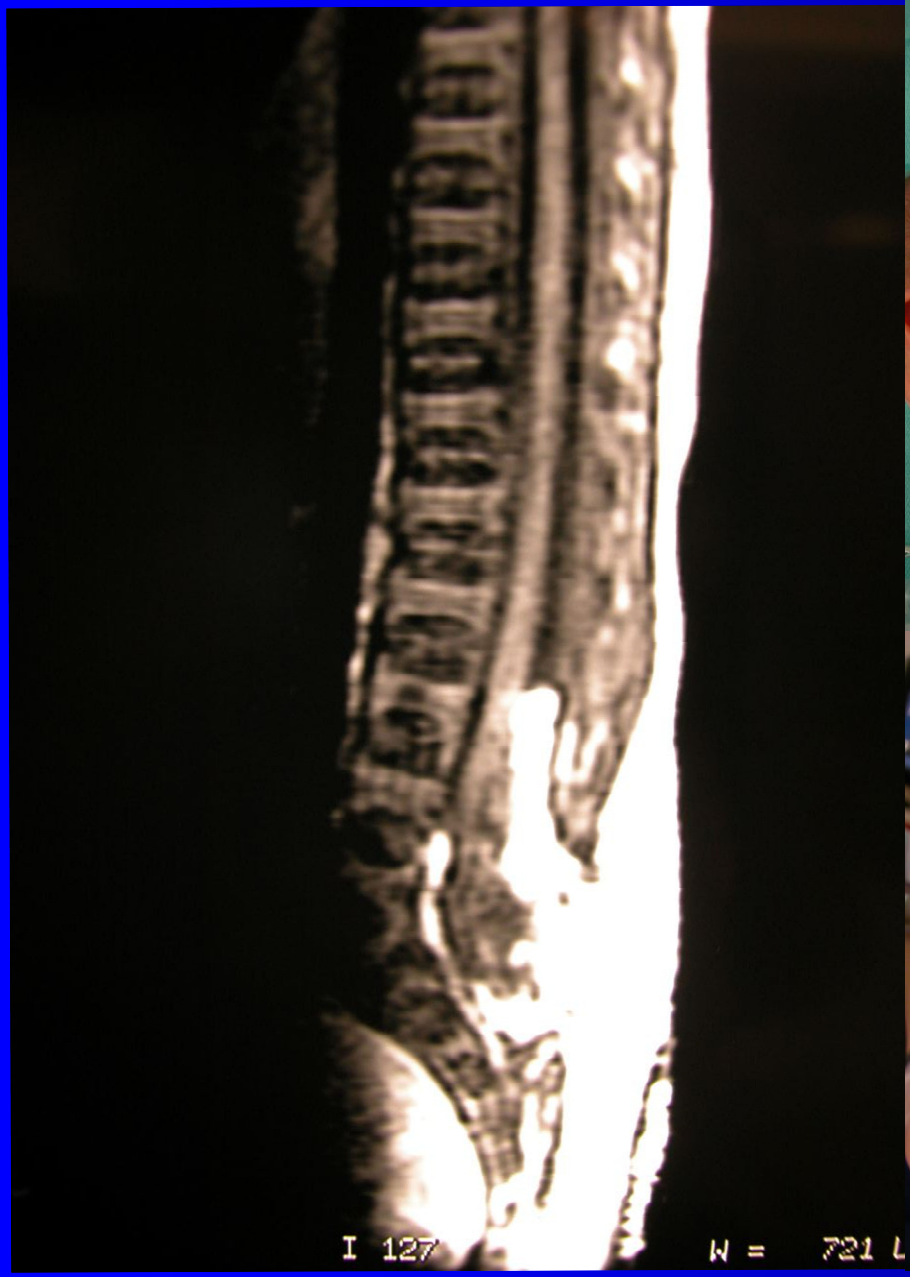
EC:1/1 15.6kHz


15.6kHz

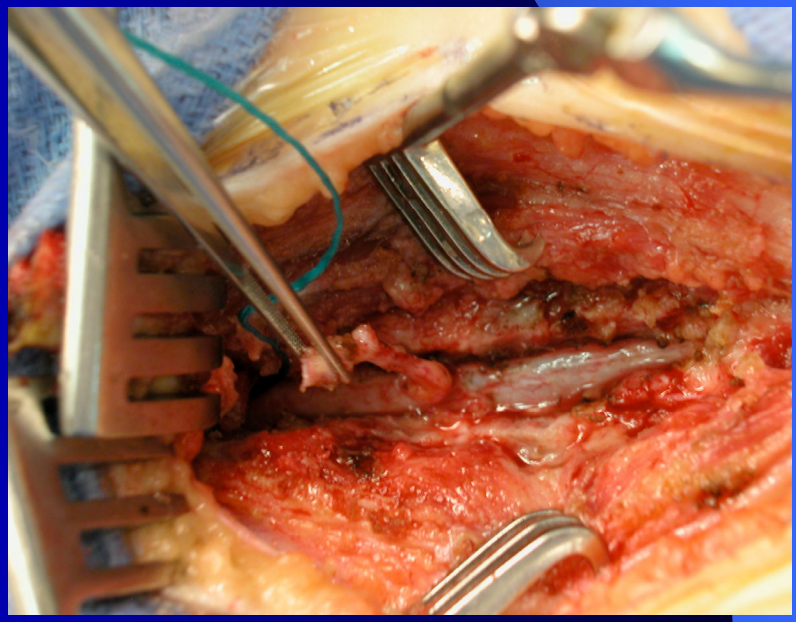


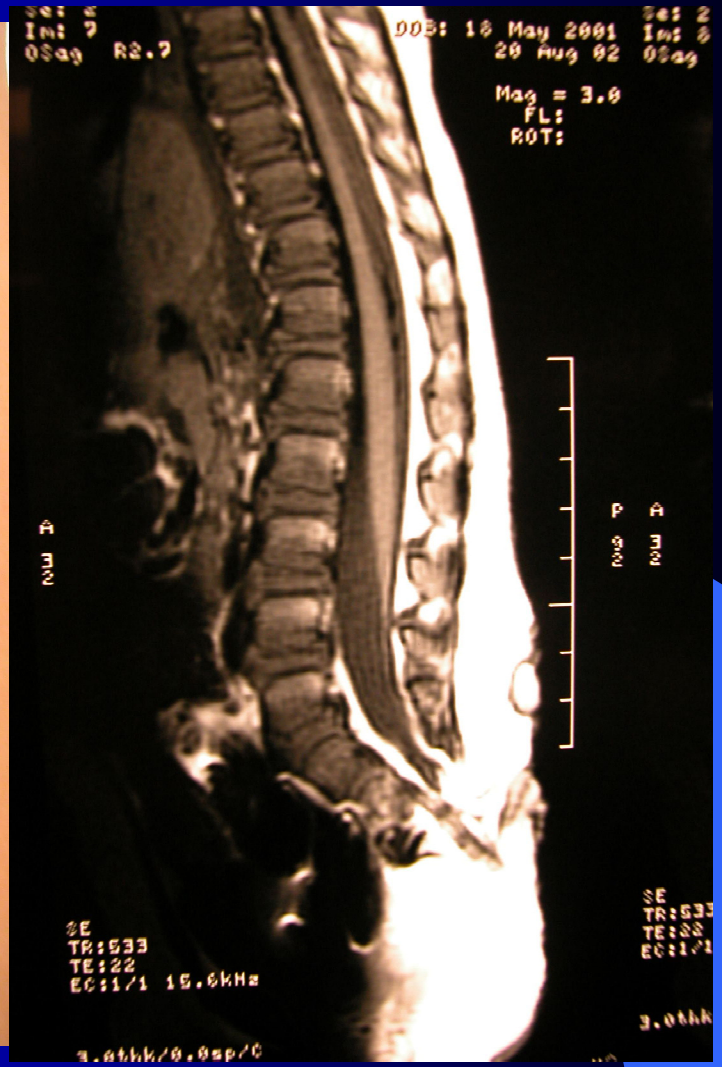



I 127
W = 721

DERMAL SINUS TRACTS

- FREQ. COMMUNICATE WITH CORD
- MAY PRESENT W/ REP. MENINGITIS
- OFTEN ASSOC. W/ LIPOMAS / HEMANGIOMAS/ DERMOIDS

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Img: 7
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003: 16 May 2001
20 Aug 02
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TR:533
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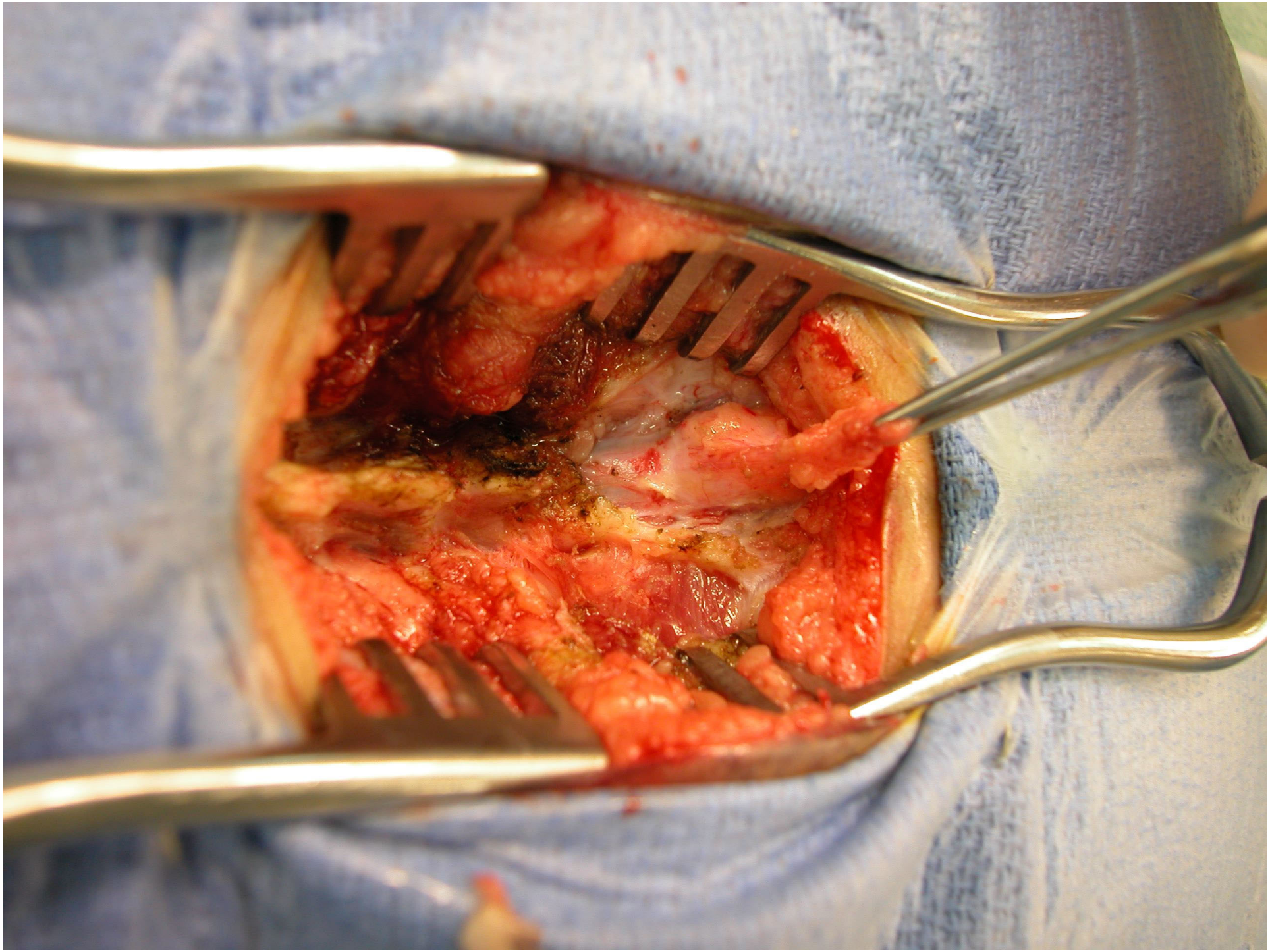
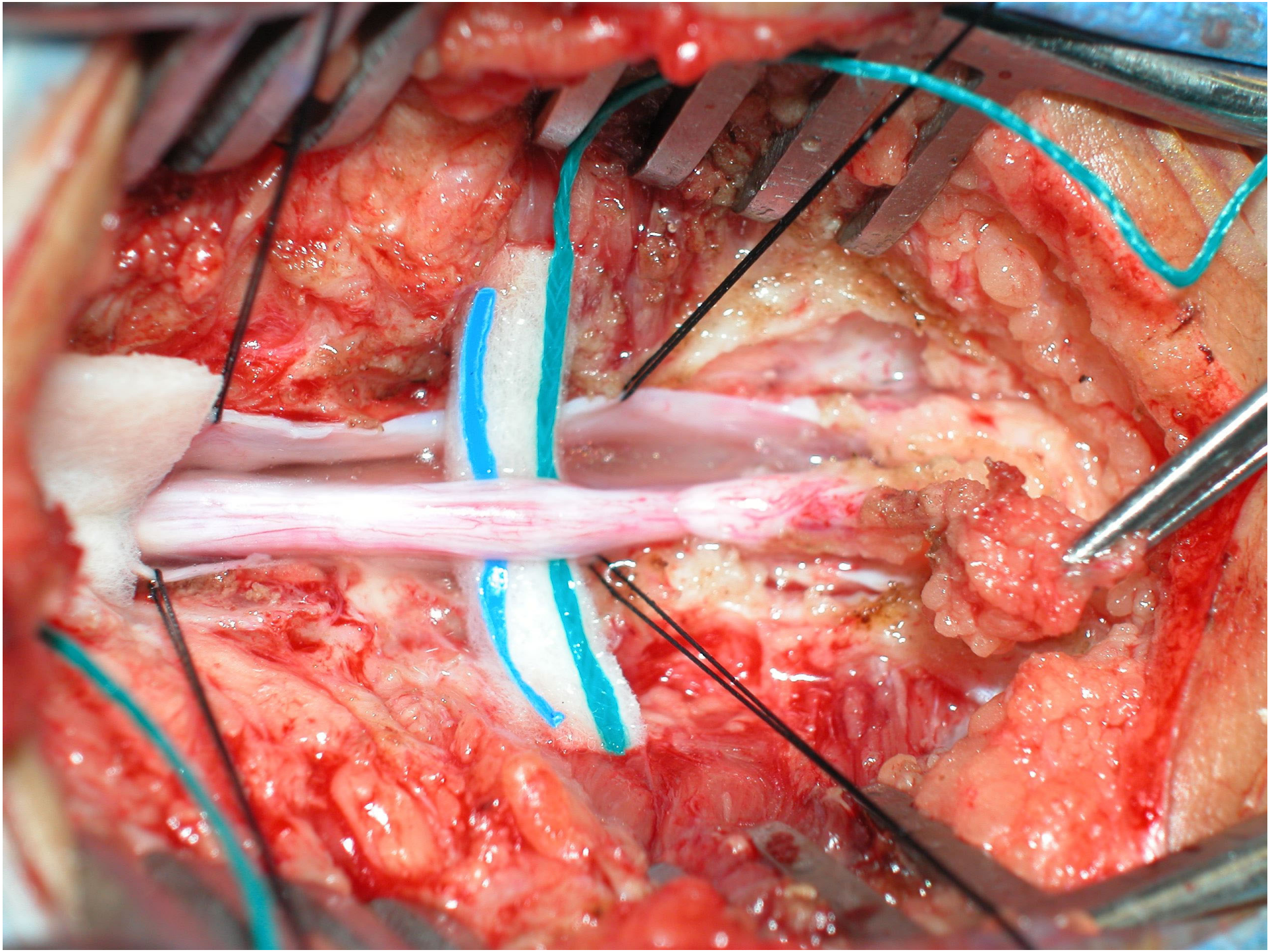
BC:1/1 15.6kHz





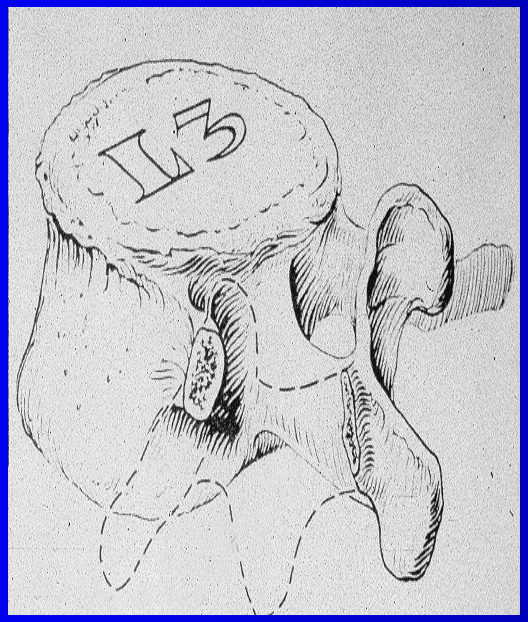
SPLIT CORD MALFORMATIONS

- INTRADURAL MIDLINE MASS, COMPOSED OF BONE, CARTILAGE OR FIBROUS TISSUE w/ ASSOC. DURAL SLEEVE, WHICH MAY RESTRICT CORD MOVEMENT
- FREQ. ASSOC. w/ HYPERTROPHIED FILUM
- HEMICORD OR TRUE SPLIT CORD AT DEFECT
- CORD USUALLY REUNITES AT LOWER LEVEL

-TYPE 1 (DIASTEMATOMYELIA)

-TYPE 2 (DIPLOMYELIA)

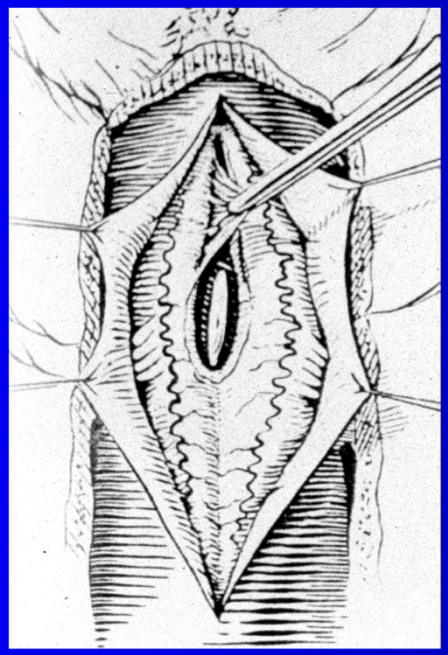
TYPE I SCM
DIASTAMATOMYELIA

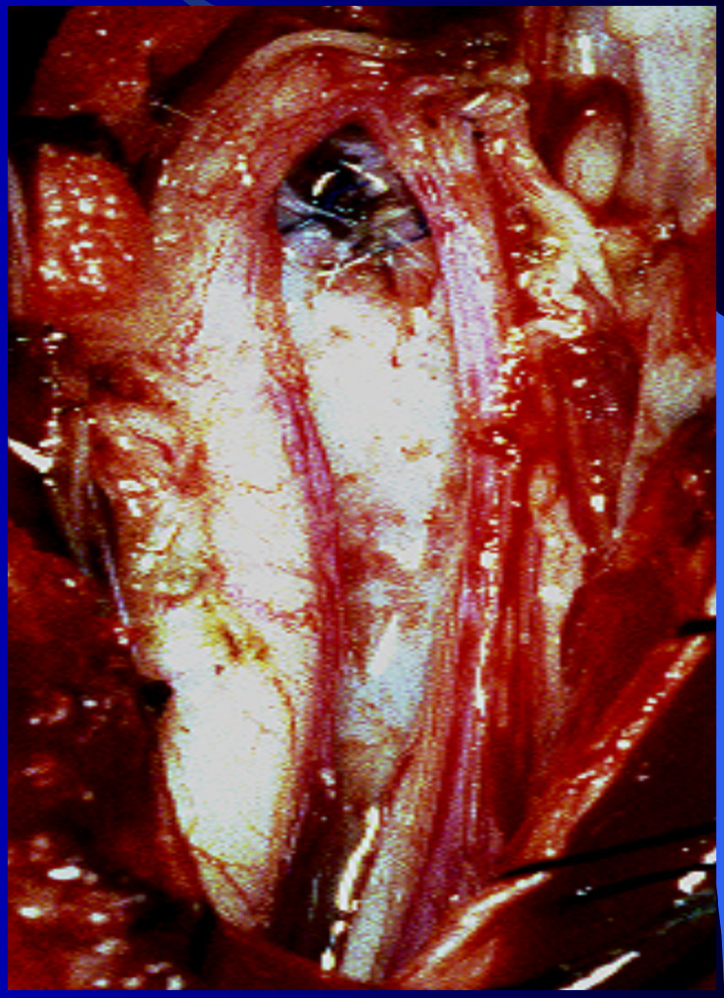


TYPE II SCM
DIPLOMYELIA

SURGICAL TREATMENT

CASE EXAMPLE



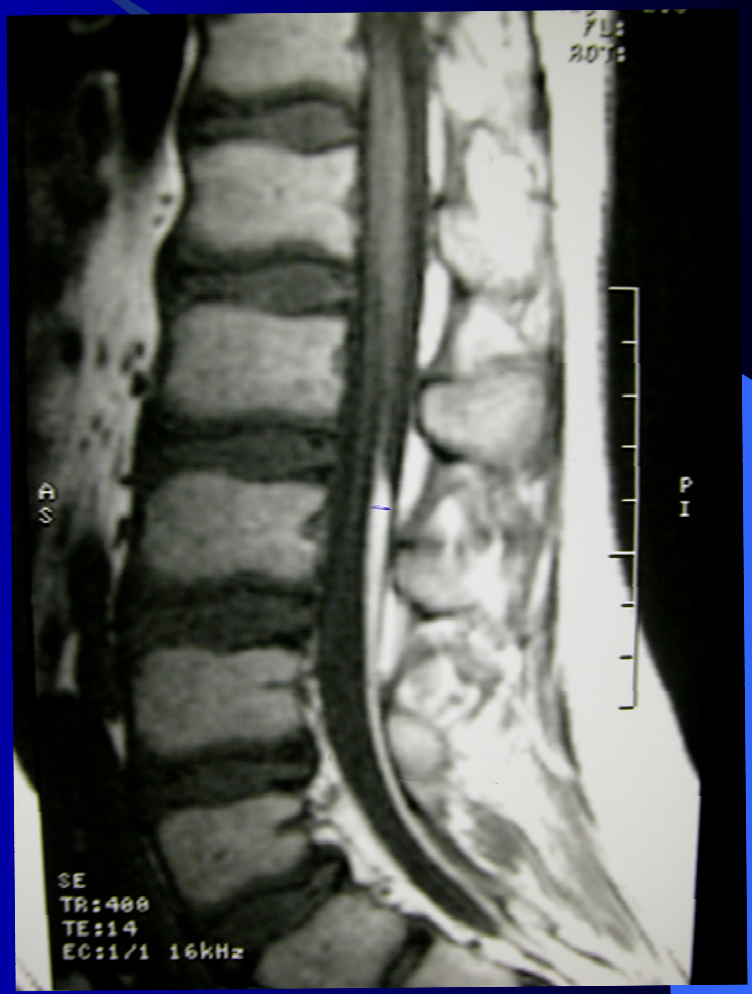


FILUM DYSGENESIS

-HYPERTROPHIC FILUM

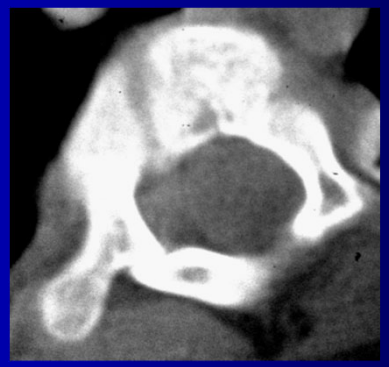
-LIPOMATOUS FILUM

- -ANORECTAL MALFORM
- -DERM SINUS TR
- -ARTHROGYRPOSIS
- -SYNDROMIC (SOTO),ETC.



ANORECTAL MALFORMATIONS

- 1 / 5,000 LIVE BIRTHS
- CLOACAL EXSTROPHY
- VATER COMPLEX
  - VTB ANOMALIES
  - ANORECTAL ANOM.
  - TE FISTULA
  - RENAL
  - CARDIAC
- IMPERFORATE ANUS



ANORECTAL MALFORMATIONS

ETIOLOGY OF TETHERING

- **35% of ARM pts----> tethered**

  Golonka NR, Haga LJ, Keating RF et al, J Ped Surg,2002.

  - **FILUM LIPOMA**
  - **HYPERTROPHIC FILUM**
  - **LIPOMYELOMENINGOCELE**
  - **DERMAL SINUS TRACT**

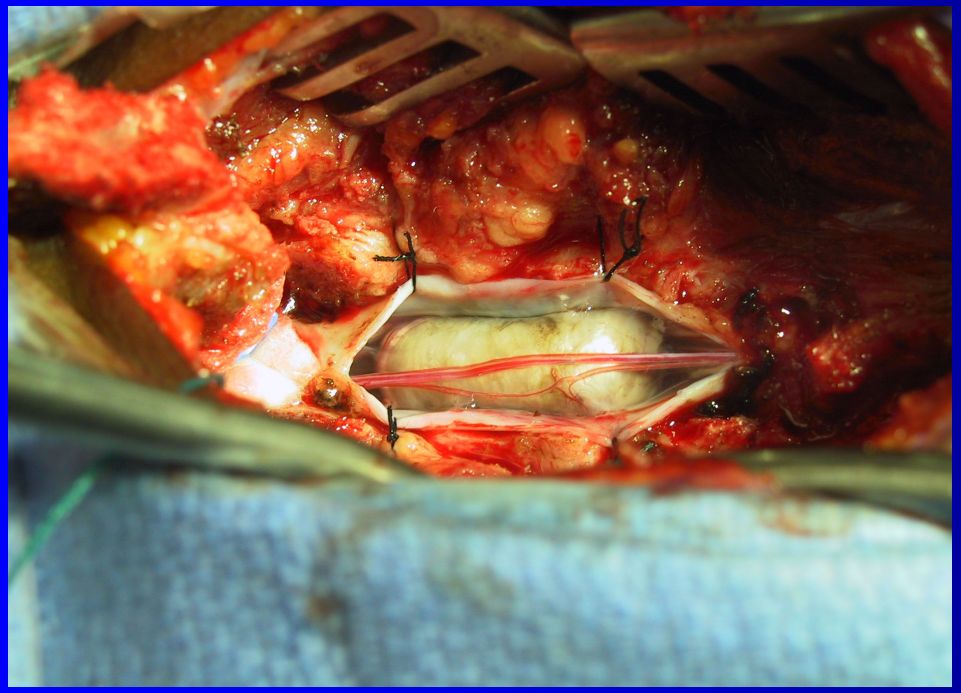
ECTODERMAL INCLUSIONS

-DERMOID / EPIDERMOID

-TERATOMA

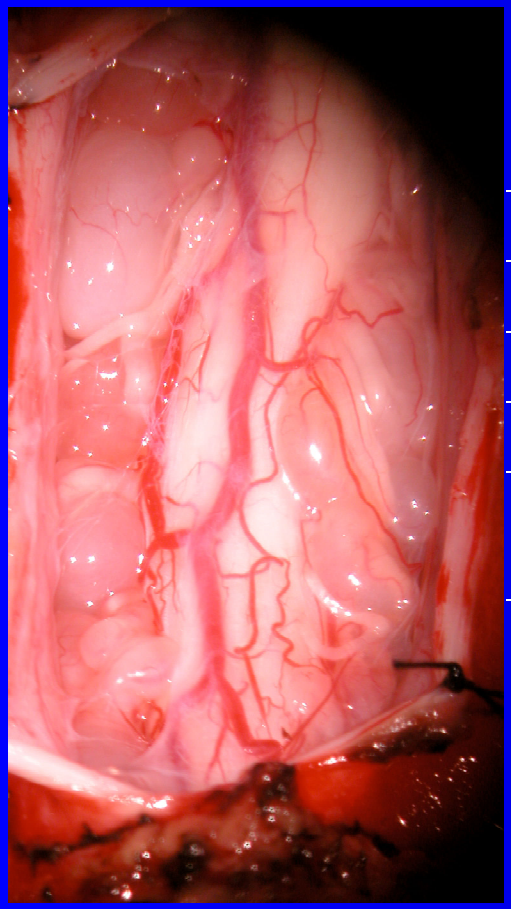
-LIPOMATOUS FILUM

-DERMAL SINUS TRACT



SCOLIOSIS

TUMOR



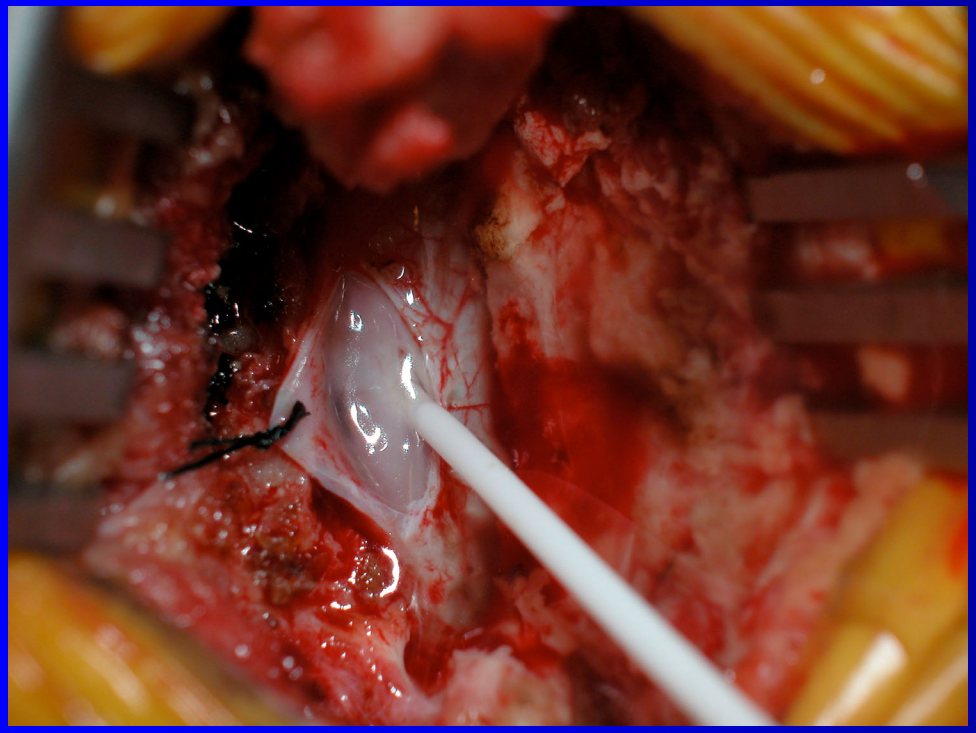
- UNCOMMON
- SCOLIOSIS MAY BE ONLY SIGN
- ASSOCIATED SYRINX EARLY SIGN
- PAIN / TEMP, QTT EARLY SIGNS
- IF SUSPECT NEURO CHANGES --> GAD + MRI
- 25-33% SP. CD TUMOR --> ASSOC SCOLI
  - Boldrey et al.,Arch Neurol Psych, 1949;61:528.
  - Hayden JW,Ped Clin N Am, 1967;14:611.

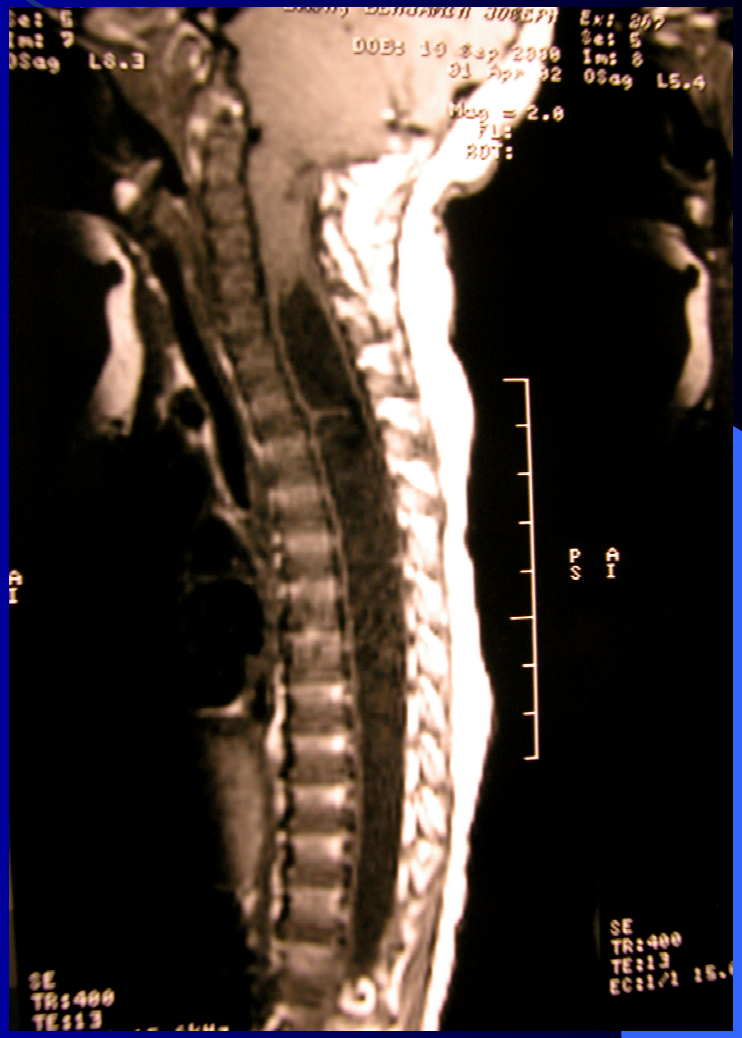
INTRAMEDULLARY CYSTS

-HOLOCORD SYRINX

-NEURENTERIC CYSTS

-RESP. SYNCYTIAL CYSTS



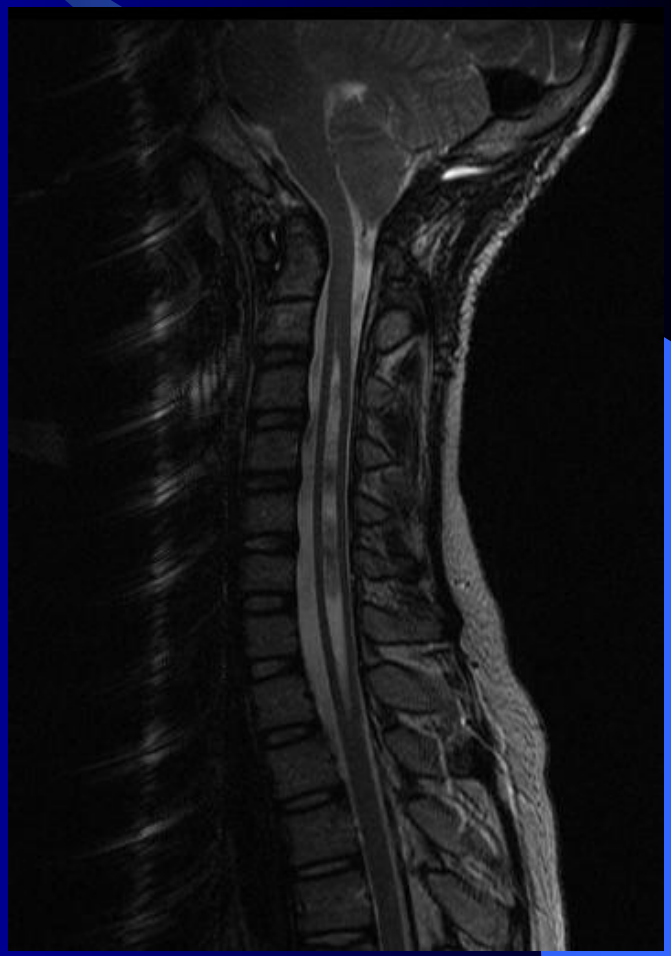


SCOLIOSIS

ETIOLOGY

HYDROMYELIA

- COMMONLY ASSOCIATED w/ SCOLIOSIS
- USUALLY CERVICAL / THORACIC OR HOLOCORD
- CHIARI, TETH.,POST-TRAUMA, TUMOR, IDIOPATHIC (10%)
- SCOLIOSIS ACCOMPANYING SYRINGOMYELIA 25-62%



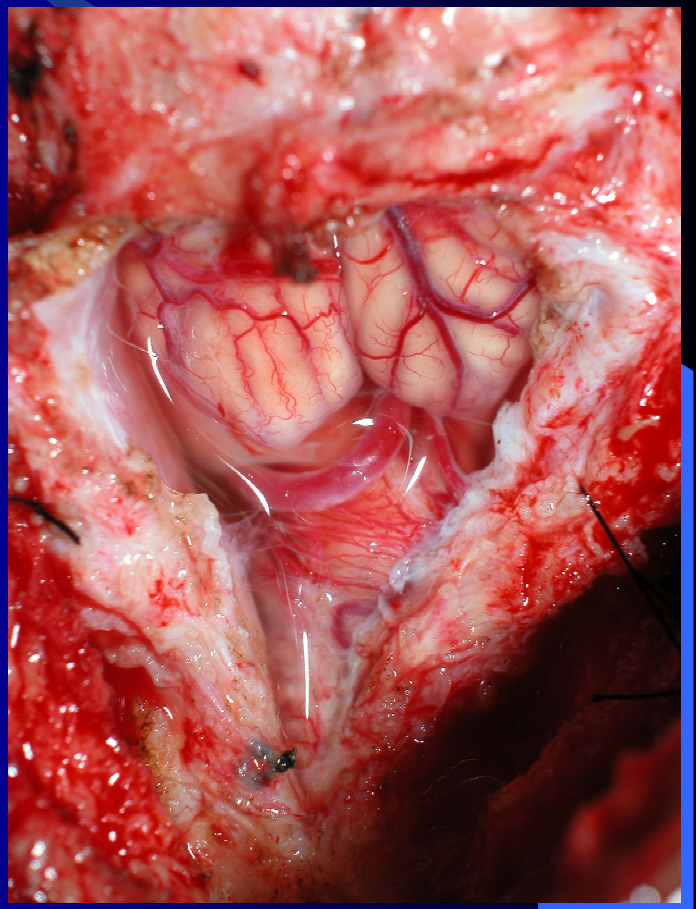
SCOLIOSIS

ETIOLOGY

HYDROMYELIA

SUSPICIONS RAISED BY:

- LG CURVE < 10 YRS
- ASSOCIATION WITH TORTIC / PAIN / NEURO CHG
- L CURVES MORE CONSPIC. ???
- DOUBLE PRIMARY CURVES



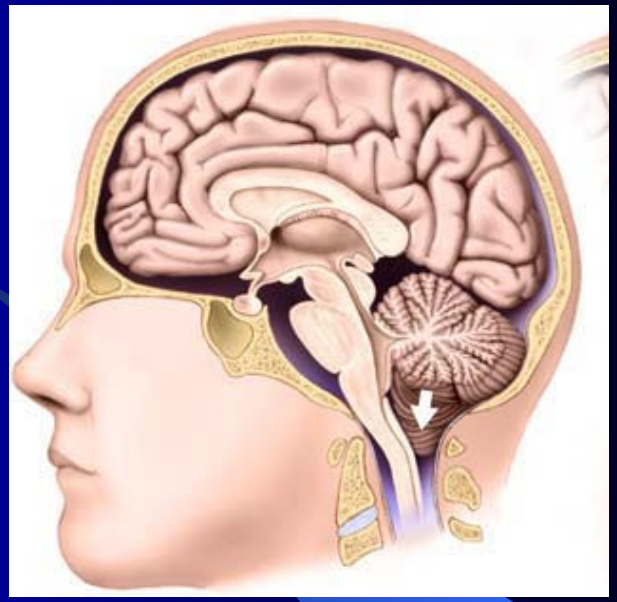
Syringomyelia

Etiology

- Chiari
- Tethered Cord
- Post-infectious
- Post-inflammatory
- Tumor
- Hydrocephalus
- Idiopathic

Introduction

- Chiari malformation type I is characterized by descent of cerebellar tonsils below the foramen of magnum



Chiari 0

Normal position tonsils with characteristic symptoms and hydromyelia

Chiari 0.5

Tonsillar descent < 4mm with S/S/syrinx

Chiari 1

Chiari 1.5

Tonsillar descent > 4mm w/ hindbrain herniation
No evidence of spina bifida

Chiari 2

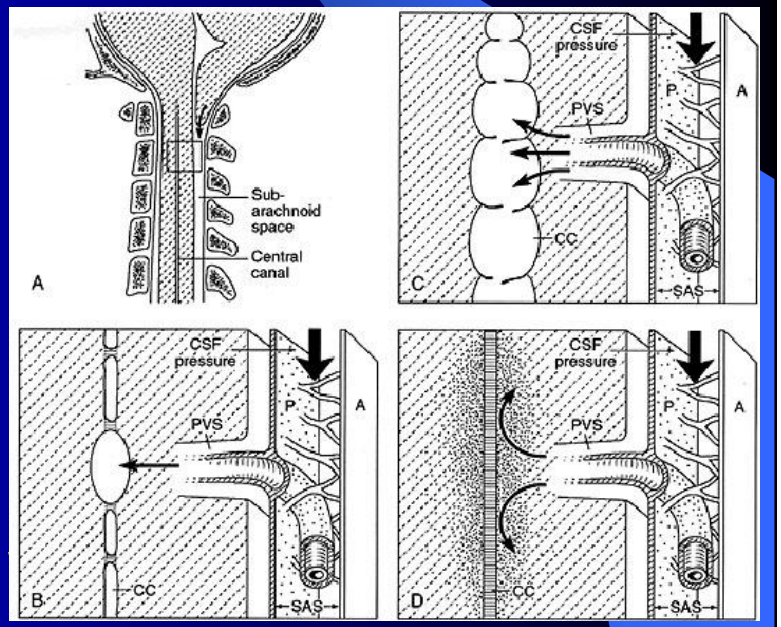
Vermian / Hindbrain herniation w/ S.B.

? Chiari 1.75

Tubbs, Oakes et al. The pediatric Chiari I malformation: a review. Childs Nerv Sys. 2007 Nov;23(11):1239-50. Epub 2007 Jul 18

PATHOPHYSIOLOGY

- **Heiss JD et al.** J Neurosurg 1999 Oct;91(4):553-6
  - Transcompartmental Pressure Differential
  - Int. occulsion of for. Magnum via pistoning tonsils leads to differential pressure gradients during systole / diastole, in turn promoting propagation of CSF caudally through the central canal.
  - May see spinal cord swelling prior to development of syrinx
    Levy, Heiss, Kent et al. J Neurosurg 2000 Jan;92(1

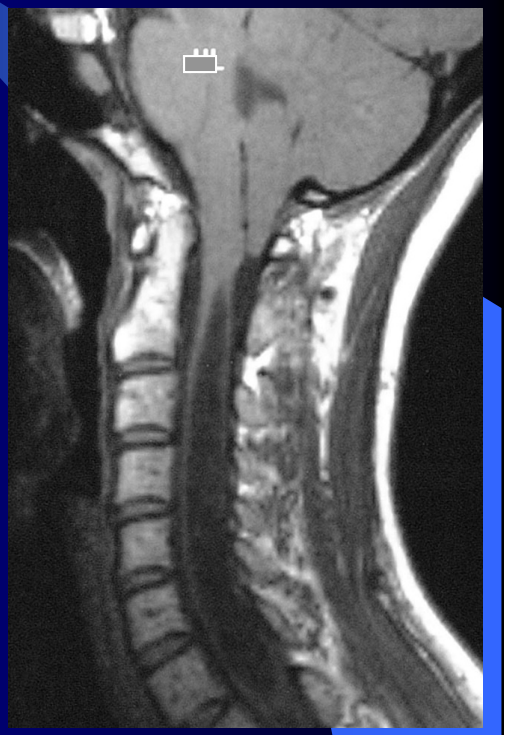


Chiari malformation

Presentation

Symptoms (age dependent / variable)

- Headache (occipital > vertex> frontal)
  - Incrsed w/ Valsalva / exertion/ crying/ cough
- Neck pain / stiffness
- Extremity paresth./ weakness/numbness
- Balance / coordination / gait difficulties
- Swallowing / aspir. / phonation challenges
- Visual changes (diplopia)
- Apneic episodes / night-time dry cough
- ? Fatique / decreased stamina
- Fill in the blank……..



Chiari malformation

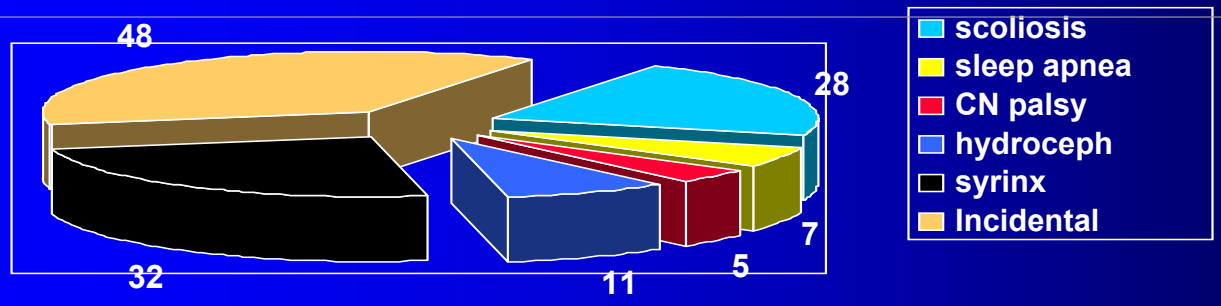
Presentation

Signs (age dependent)

- Restricted cerv. motion (esp. hyperext)
- Oculomotor paresis (ie. 6th n palsy)
- Weakness, numbness, gait / bal. changes
- Scoliosis
- Atrophy of intrinsics
- Hypertonicity / spasticity

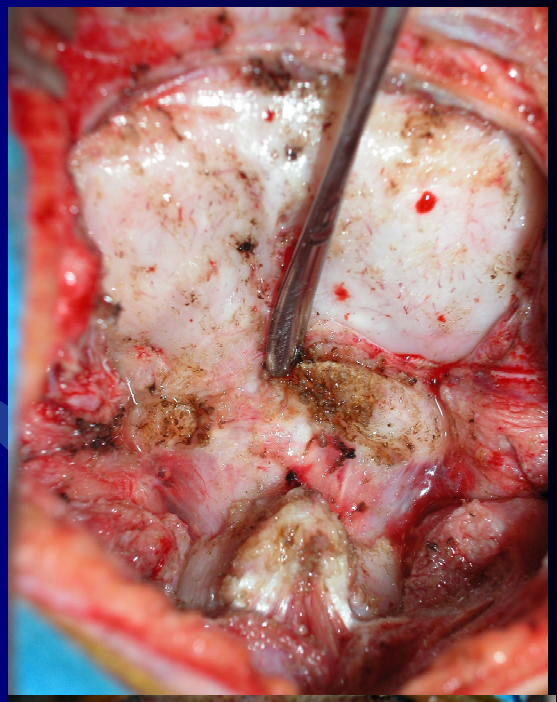
CNMC Chiari Experience

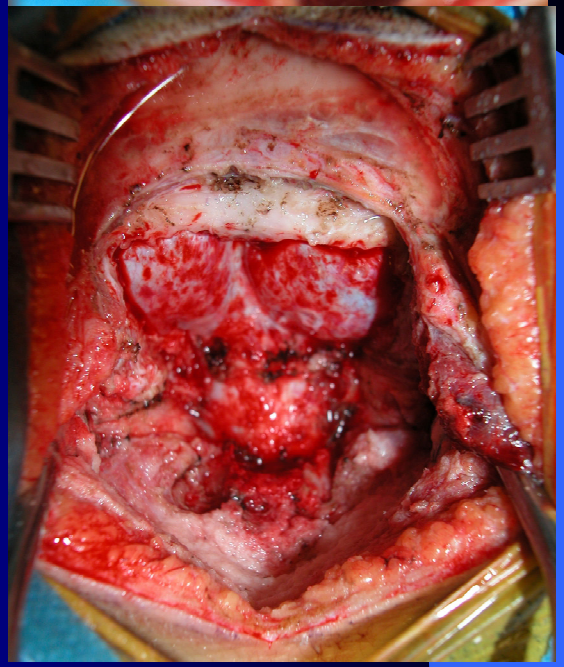
- **Presenting signs (110 pts)**
  - Syrinx 37 34%
  - Incidental 32 29%
  - Scoliosis 29 26%
  - Hydrocephalus 11 10%
  - Sleep apnea 7 6%
  - CN (6th n) 5 4%



CNMC Chiari Experience

- Surgical Therapeutics
  - 31/110 (28%) PF bony decomp., C1 lami
    - Intraop Us 22/110
      - 9/22 had bony decomp only after iUS
      - No repeat surg to date (5-36m, X=16.3)
      - 8/9 had imprvd sx (1 syrinx/ 1 6th n palsy)
  - 79/110 (72%) PF decompression, C1 lami, duraplasty
    - PFD, C1 lami, duraplsty, 4th vent stent
      - 15 cases
        - 2 secondary operations

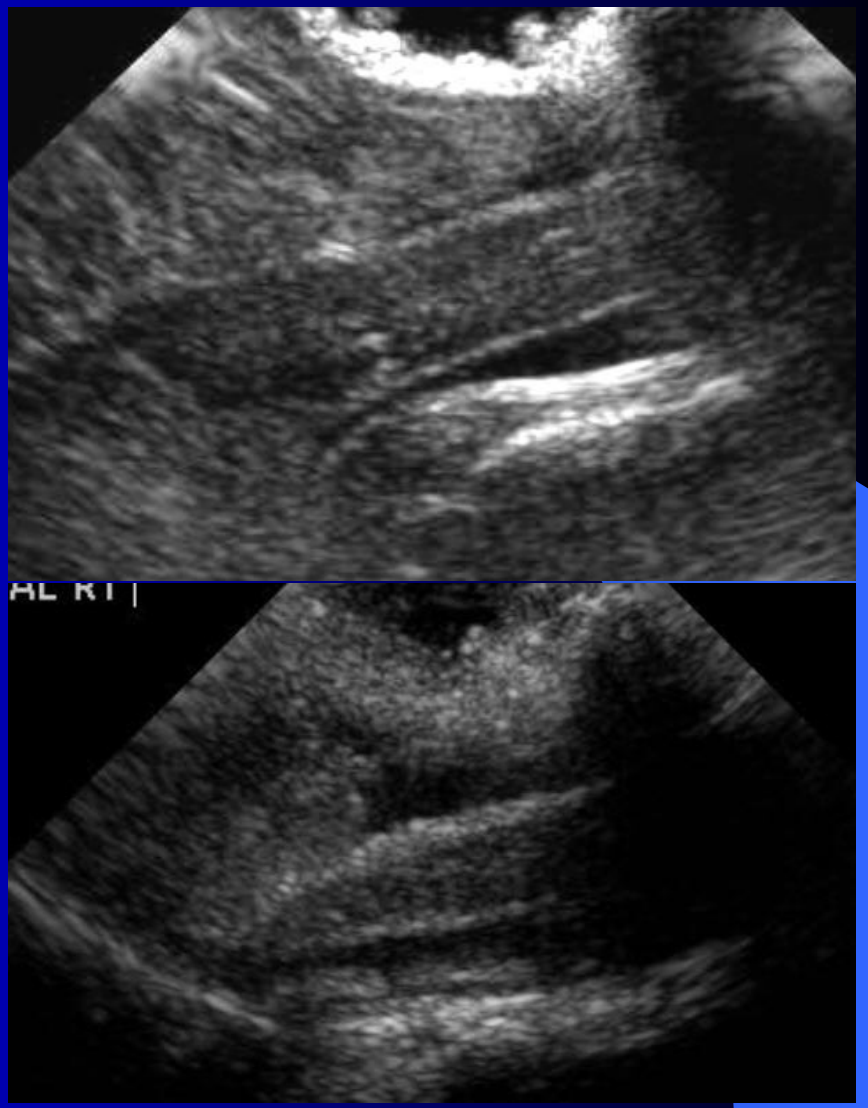




CNMC Chiari Experience

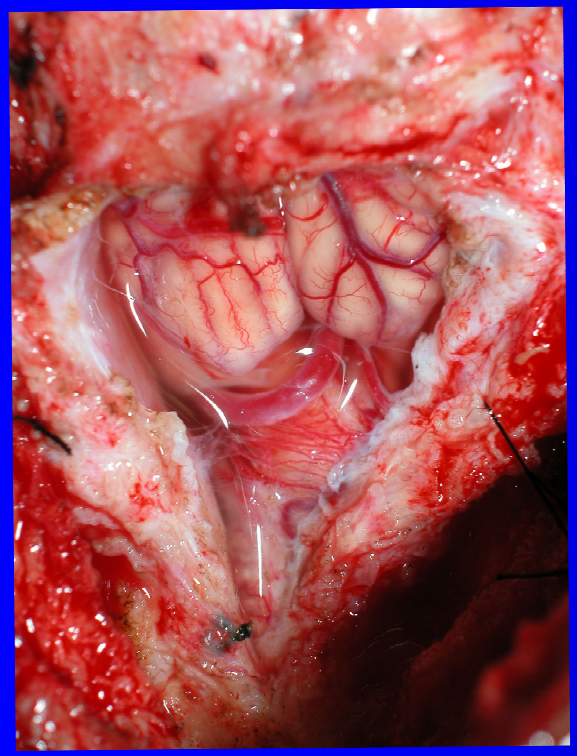
Surgery

- C1 lami, Occip. Decompr.
- Intraoperative Ultrasound
  - Decision point for open duraplasty
- Real-time evaluation of:
  - Tonsillar “pistoning”
  - Level of tonsills
  - Adequacy of CSF space

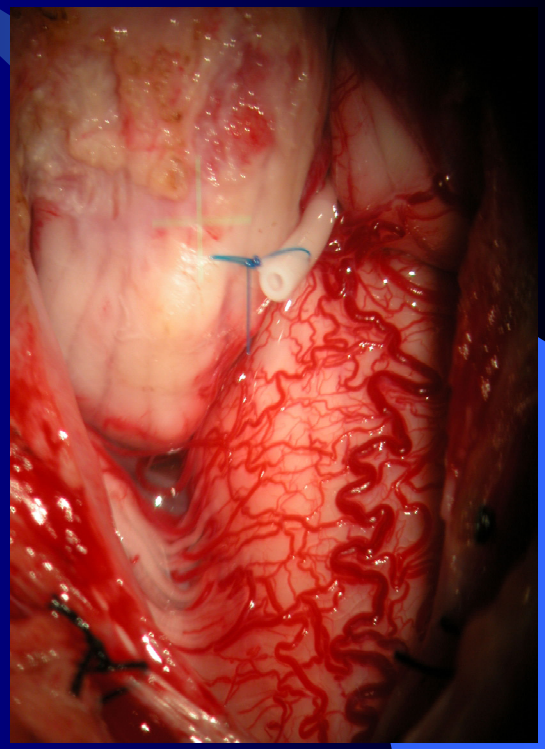


CNMC Chiari Experience

Surgery



- Open decompression Duraplasty
- Arachnoid integrity
- Lysis of adhesions
- CSF outflow eval.
  - Tonsillar resection
  - 4th vent stent
  - Obex obliteration

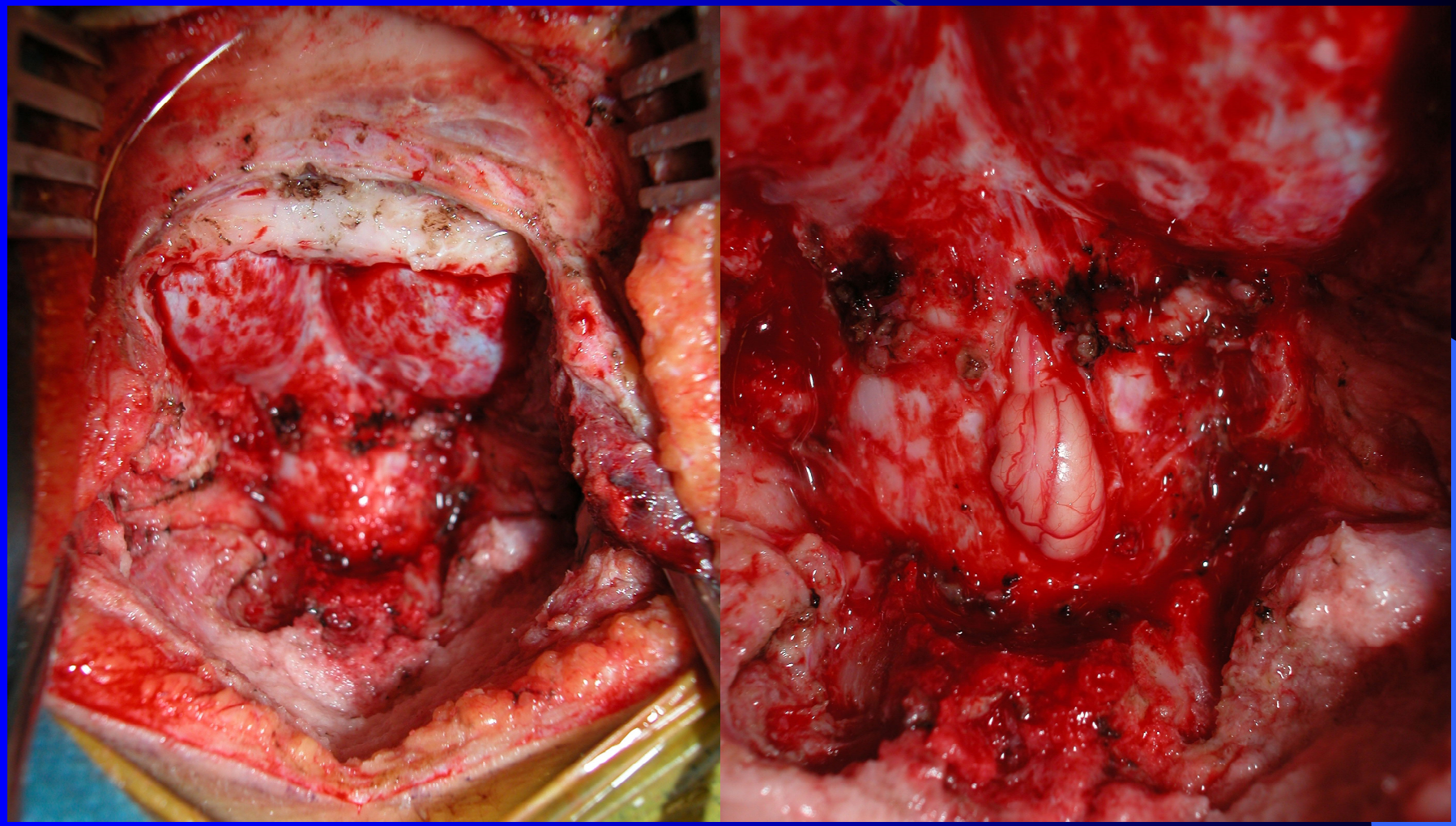


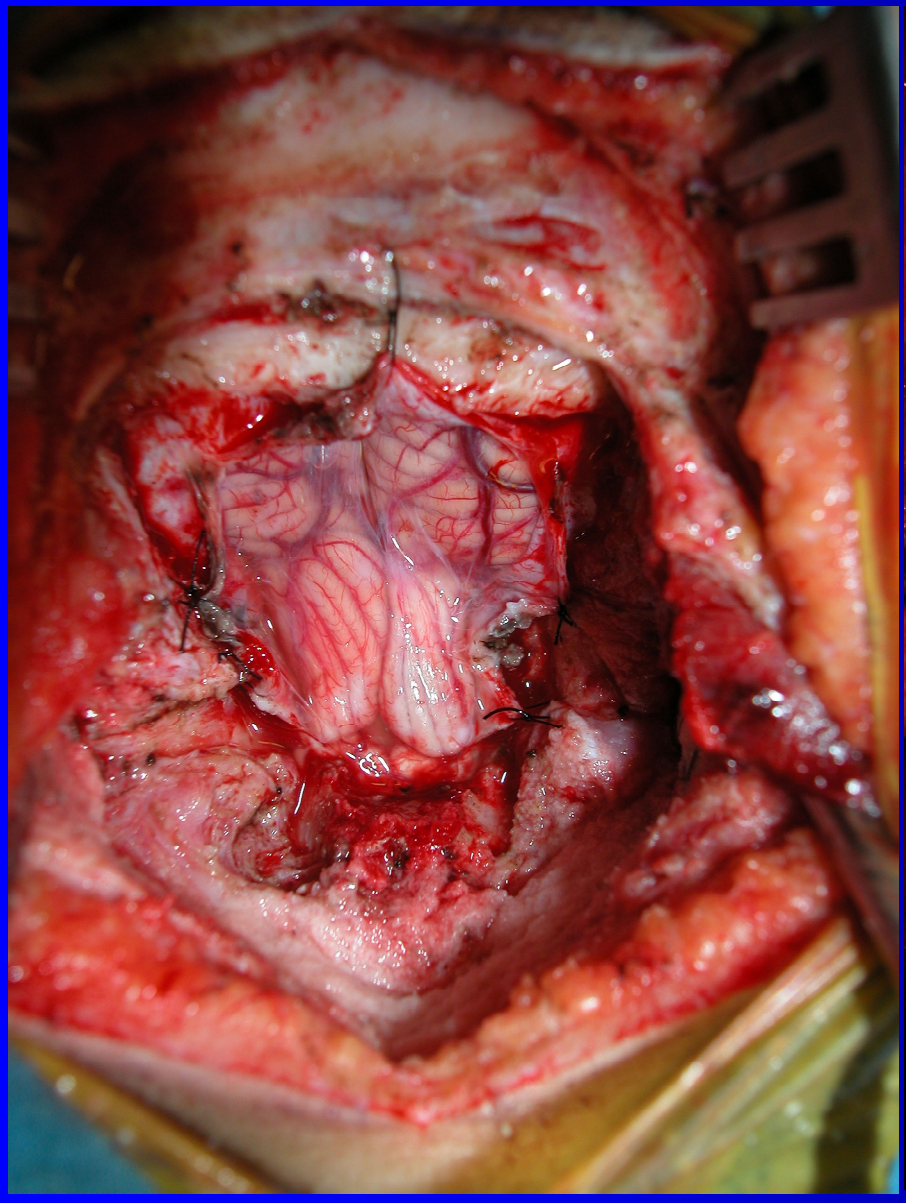
S.M.

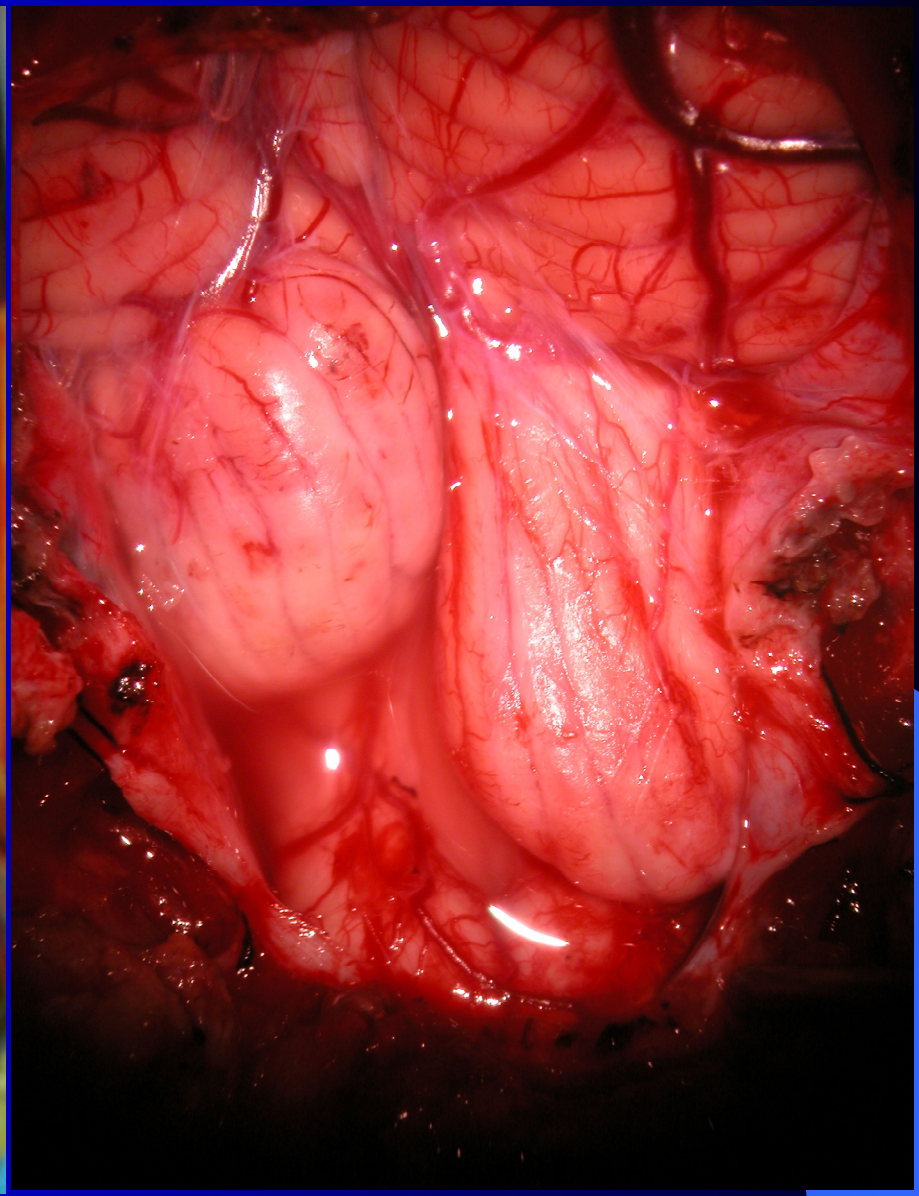
PROGRESSIVE HERNIATION

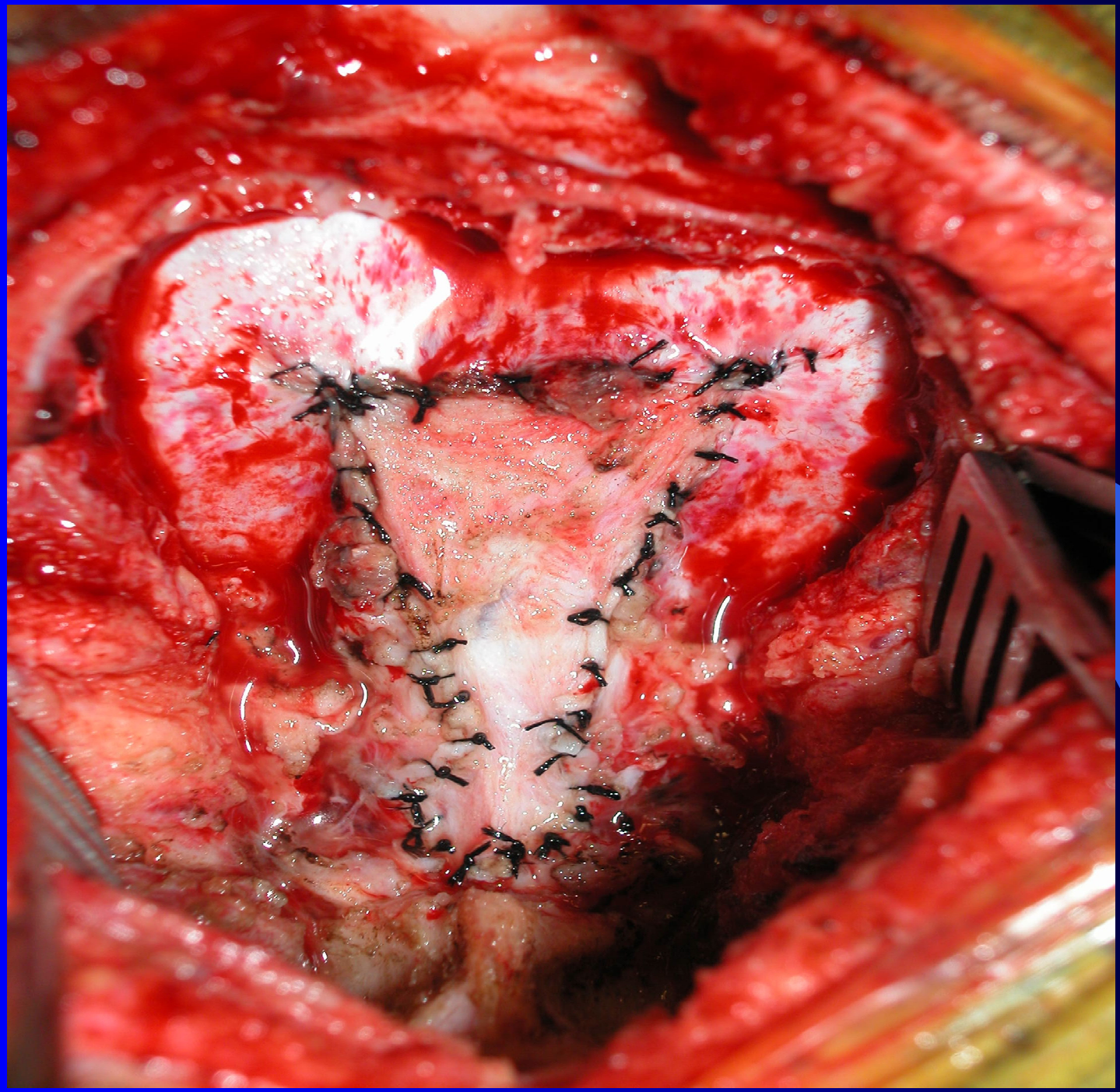
DOB:7/21/87

- Underwent PFC/ C1 lami / duraplasty 10/1/04



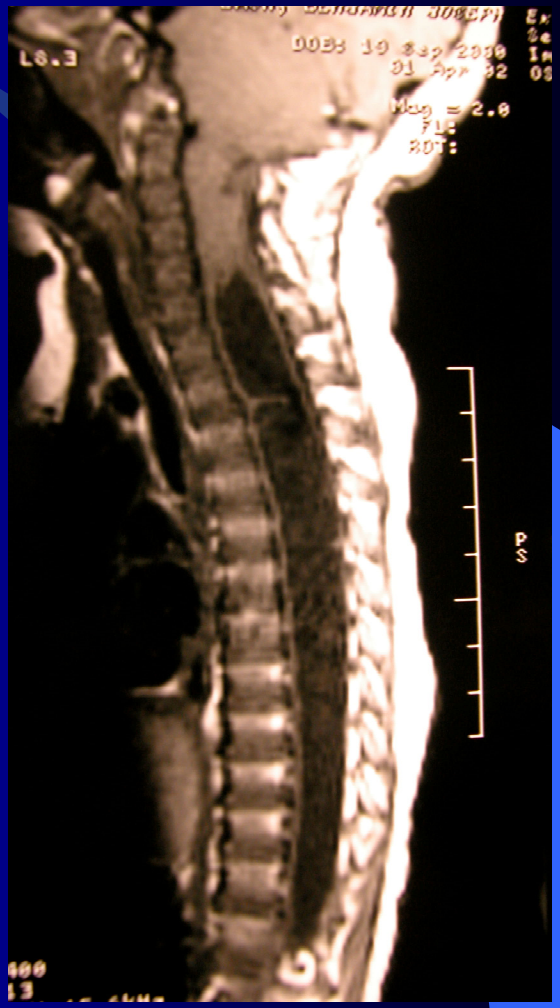






CNMC Chiari Experience

- Syringomyelia
  - 37 /110 34%
  - Size
    - 9 minimal (< 25% cord diameter)
    - 8 moderate (25-50% cord diameter)
    - 10 large (> 50% cord diameter)
  - Location
    - 23 cervical
    - 5 C-T
    - 7 Thoracic
    - 20 Holocord



CNMC Chiari Experience

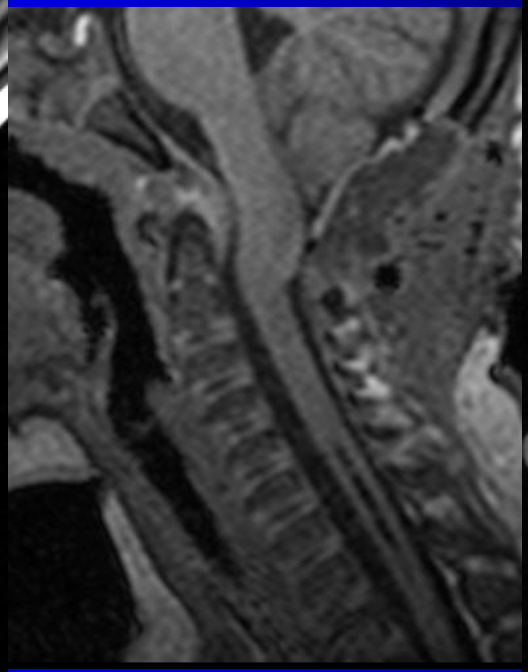
Syringomyelia

Outcome	Imprvd	Stable	Worse
• Minimal (n=9)	3	3	3?
• Moderate (n=8)	4	3	1
• Large (n=10)	7 *	3	-
• ----------			
Total n=27	14	9	1/3?

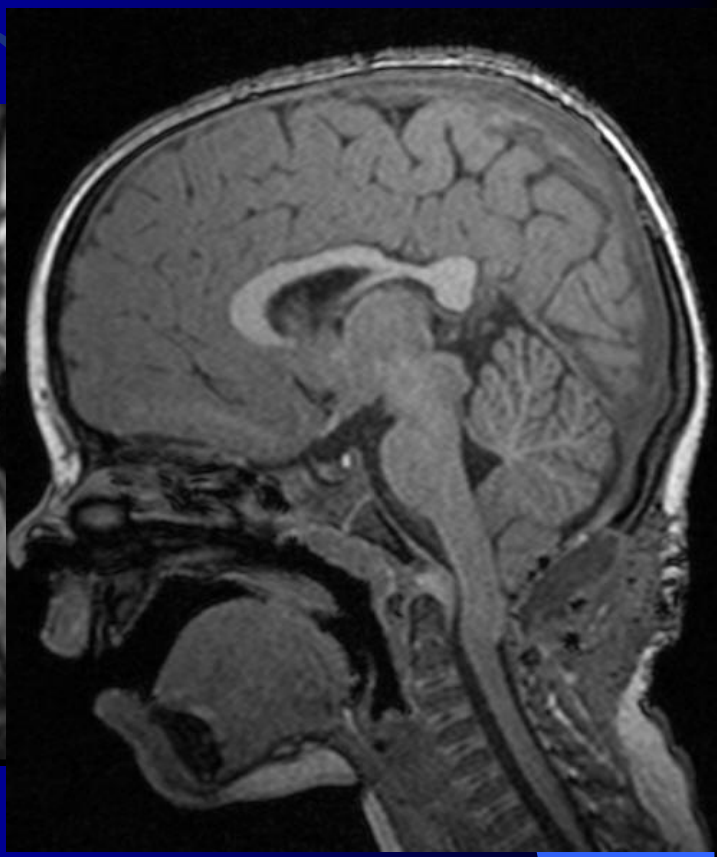
*4 pts eventually needed CSF diversion

Post-op syrinx resolution

preop



3 m postop



9 m postop

Syringomyelia

Etiology

- Chiari
- Tethered Cord
- Post-infectious
- Post-inflammatory
- Tumor
- Hydrocephalus
- Idiopathic

Syringomyelia Etiology

- **Tethered Cord**
  - **Innumerable accounts of teth cd and term syringomyelia**

Brophy et al. '89	lipo, tight filum	24%
Taviere et al. '89	lipo	25%
Gupta et al. '90	lipo	11%
	scm	38%
Tripathi et al.'92	scm	38%
	tight filum	35%
	lipo	15%

Syringomyelia

Tethered cord

175 patients with tethered cord (1997-2002)

16 patients (8 M : 8 F)

No hydro/chiari/SB

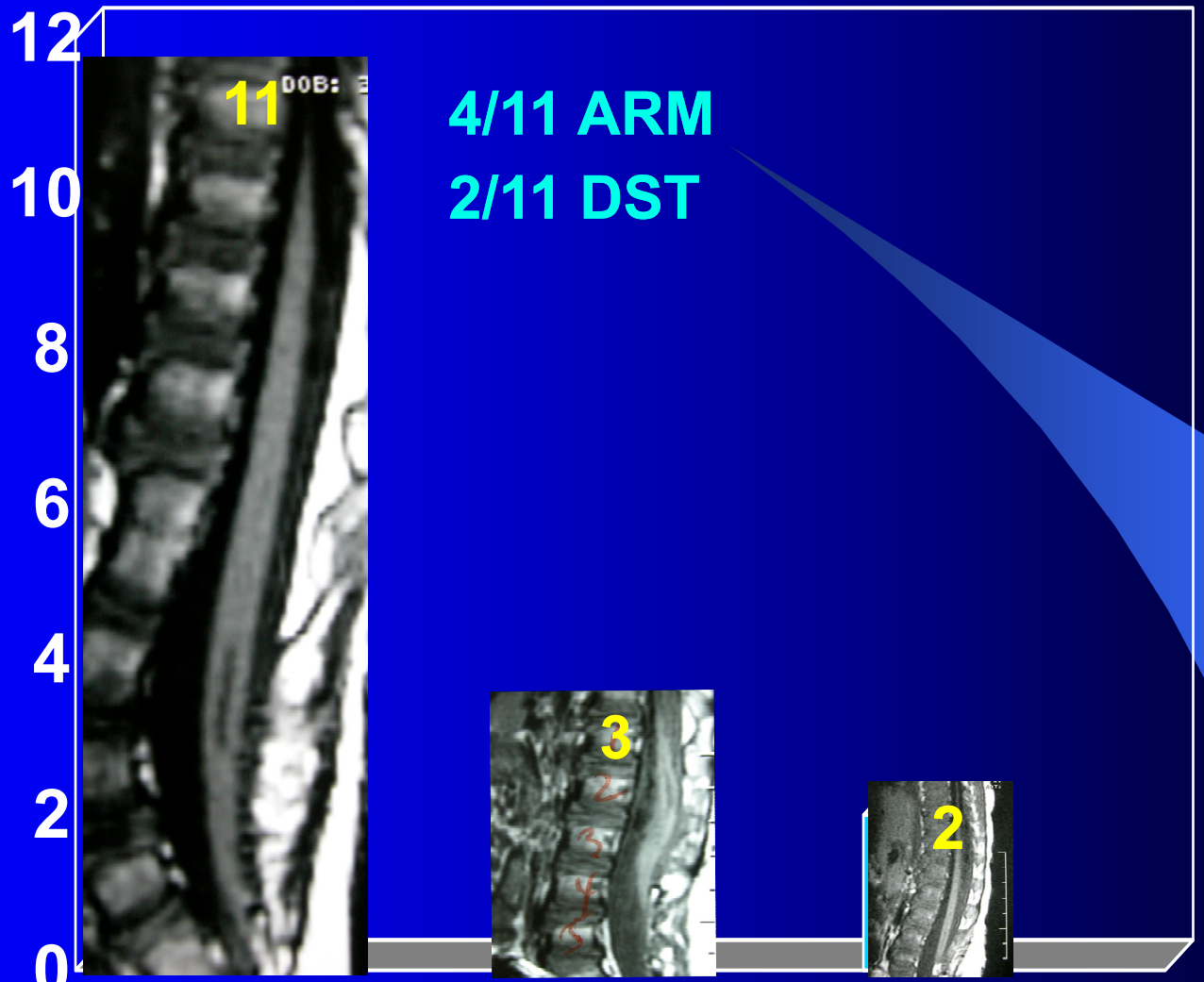
Age

3m-17y, X=41.4m

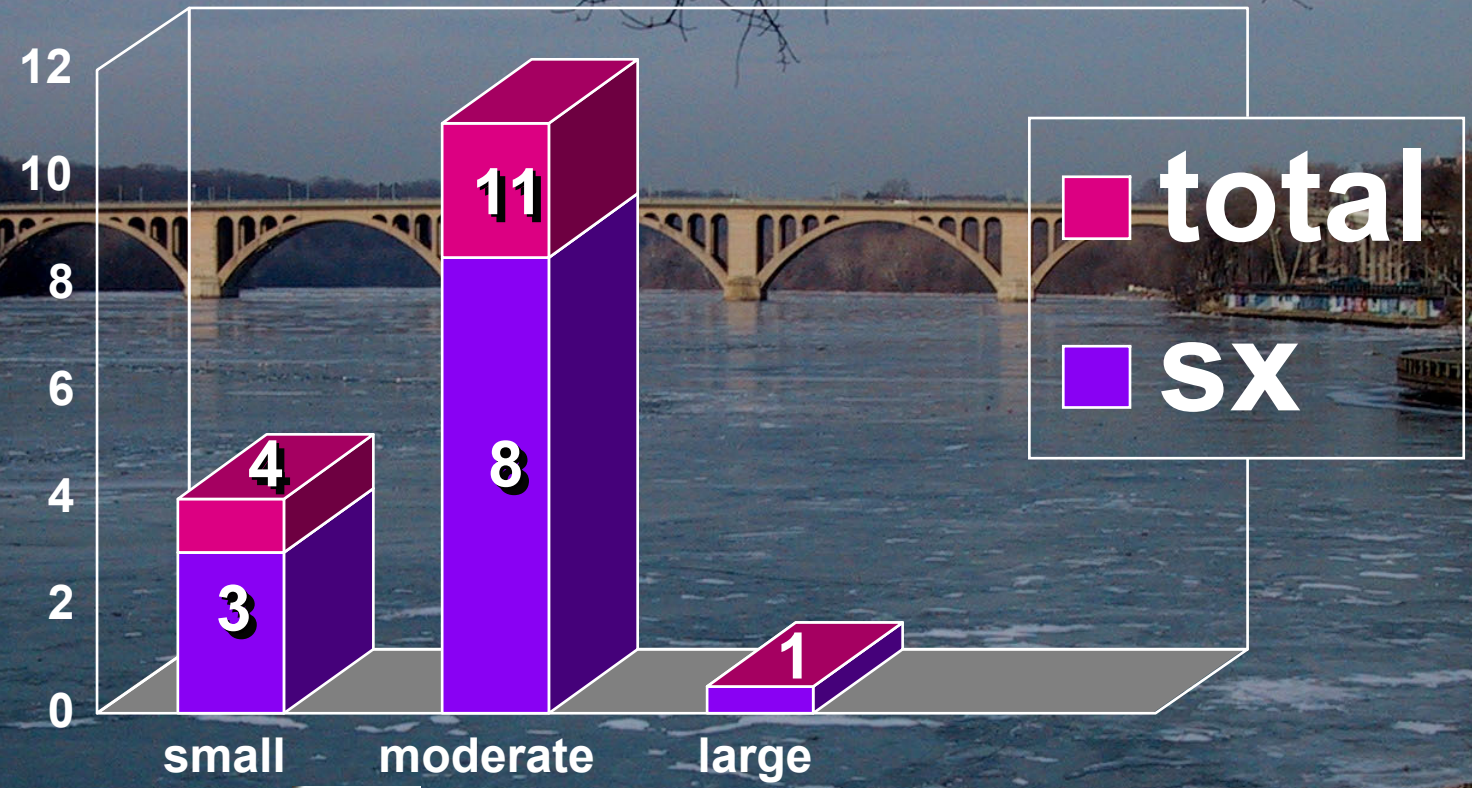
Follow-up

3m-60m, X=20.6

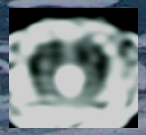
ETIOLOGY OF TETHERING



SYRINX INDEX



Small index < 25%

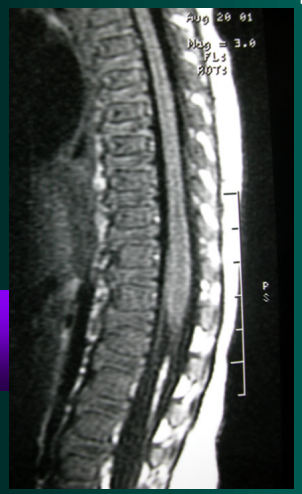


Moderate index 25-50%

Large index >50%

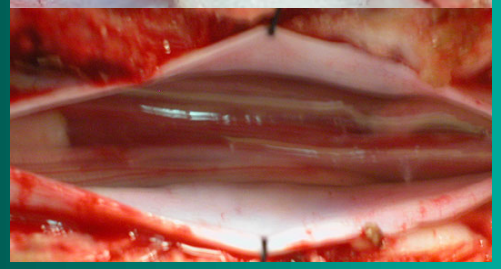
Clinical Example Filum Dysgenesis

SK: FT male born w/prom. sacral dimple, nl exam
US-->sl low(L2/3) cord, terminal syrinx
MRI-->conus L3,+lipo, mod sized syrinx(indx 25-50%)

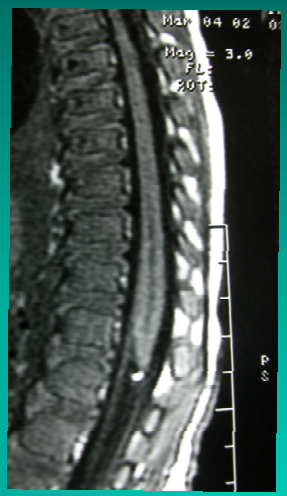


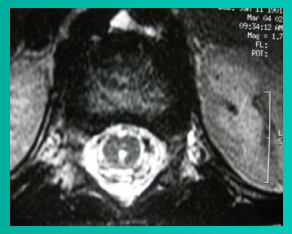
Preop

Filum sectioned @ 9m. Syrinx untouched

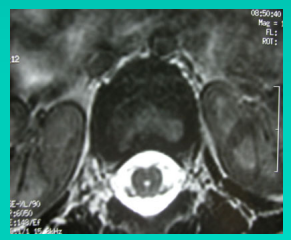


Intraop





5m post

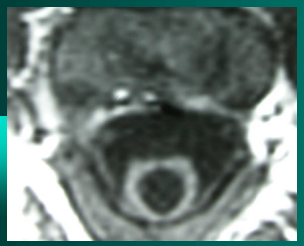


13mpost

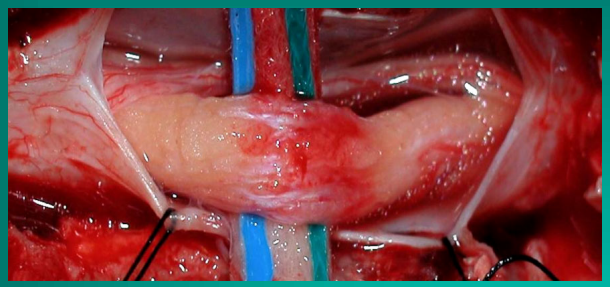
Clinical Example

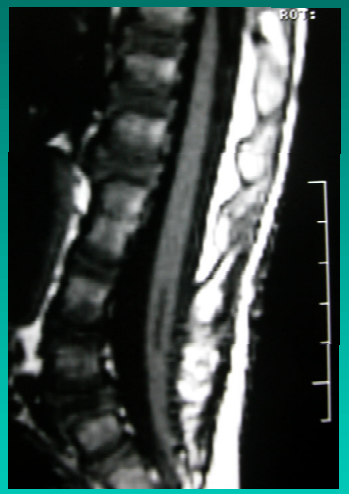
Filum/ARM

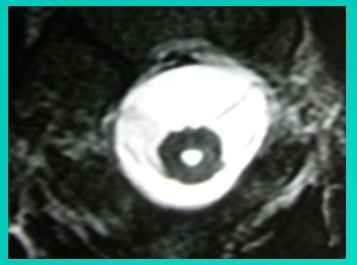
JK: 7m male born w/ imp anus, vtb anom, asym RLE
MRI-->L5/S1 conus,lg syrinx, hypertrophic filum



Preop



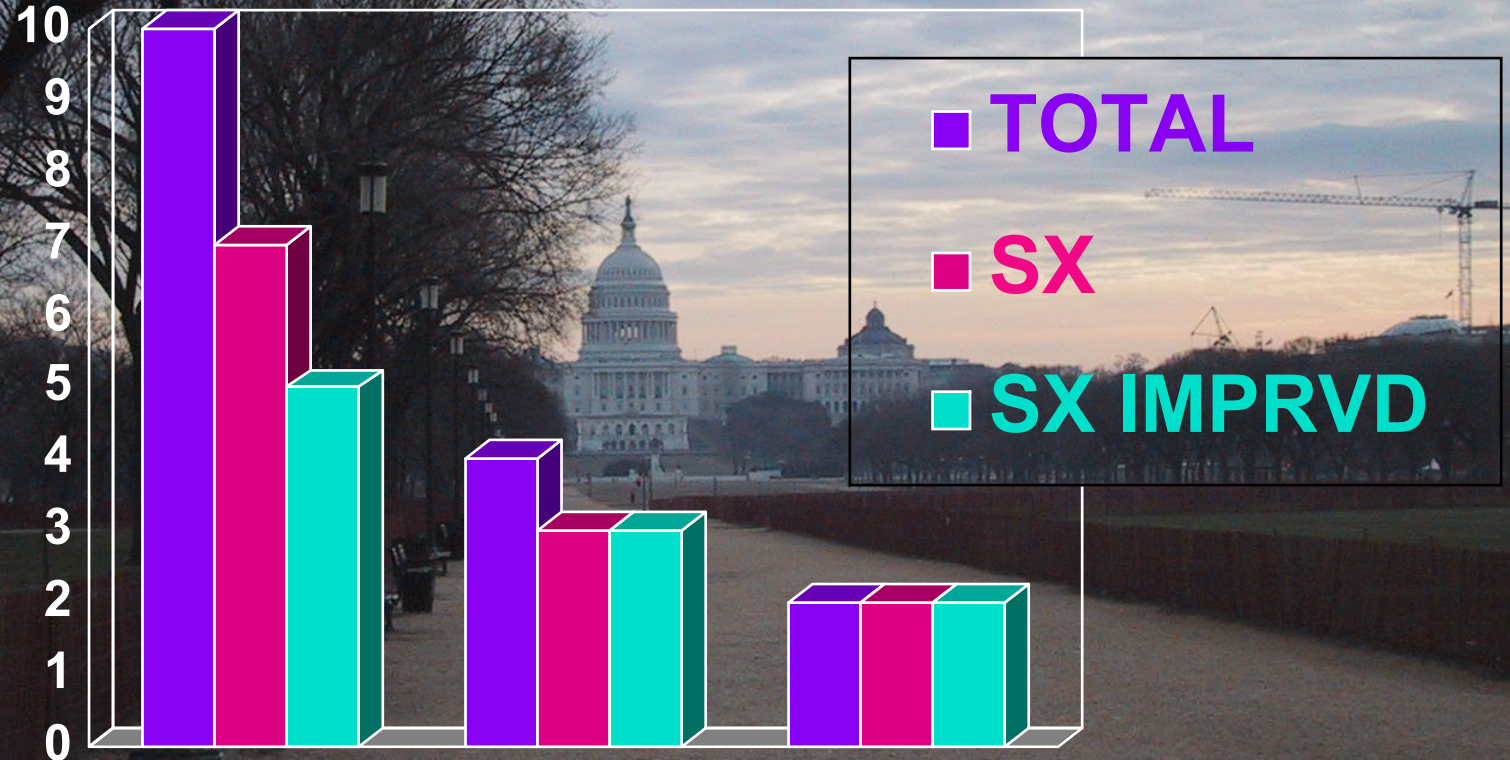




11m postop

TREATMENT

SYMPTOMS



3-156m,
X=63m

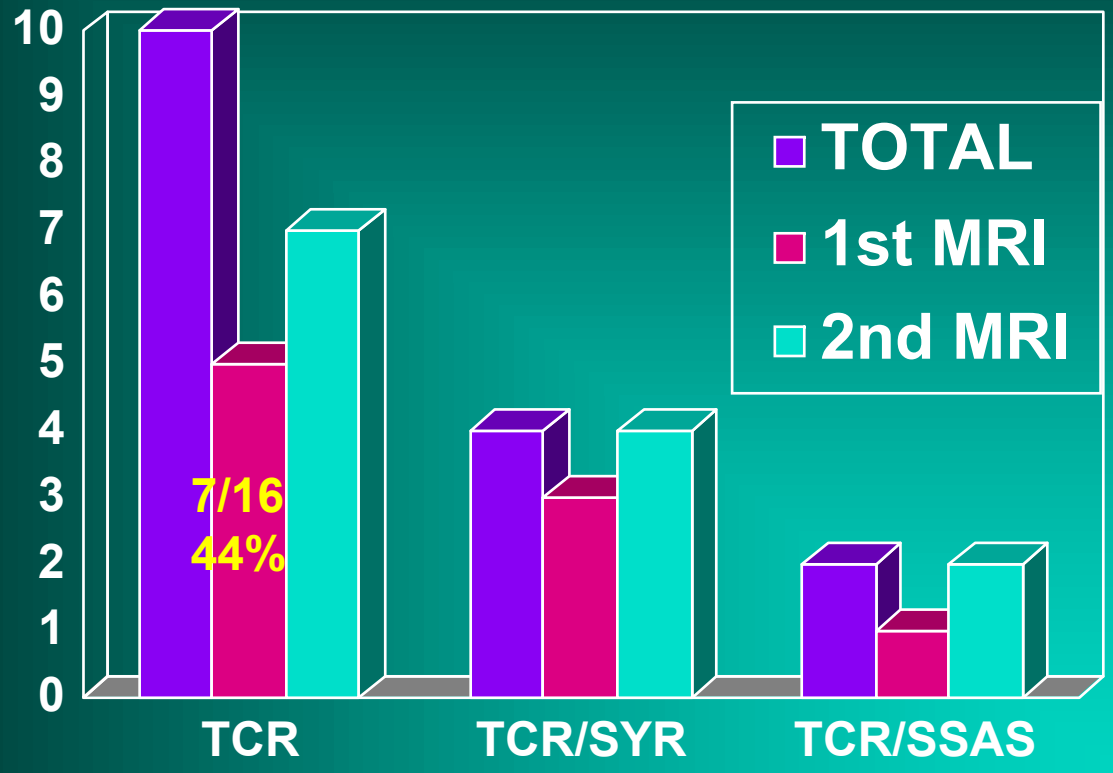
5-204m
X=35.4m

15-17m
X=16m

Age: Sx

TREATMENT

↓ SYRINX SIZE (81%)



1st MRI done 2-7m, X=4m
2nd MRI done 13-26m, X=15m

SCOLIOSIS

CONGENITAL

INCIDENCE OF INTRASPINAL ANOMALIES

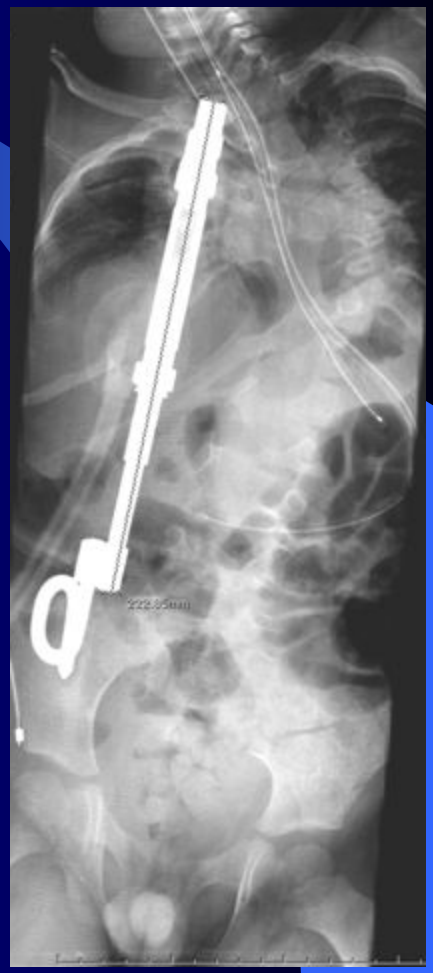
- HISTORICALLY 5-18%
  - McMaster MJ, Occult intraspinal anomalies and congenital scoliosis,J Bone Joint Surg Am, 1984
- MRI--> 53%
  - Nokes et al,Ped Rad, 1987;164:791.
  - 28 pt,X=9y,9/16 struc +, 6/12 nonstruc +
- MRI --> 13-38%
  - Bradford et al, J Ped Ortho, 1991;11:36.
  - 16/42 congen scoli pt, 1y-45y --> +
    - 10% with syrinx
  - Pahys J et al., 13% idiopathic infantile scoli--> intraspinal anomalies (majority requiring neurosurgery)

SCOLIOSIS

RADIOLOGY

MRI SCREENING VITAL

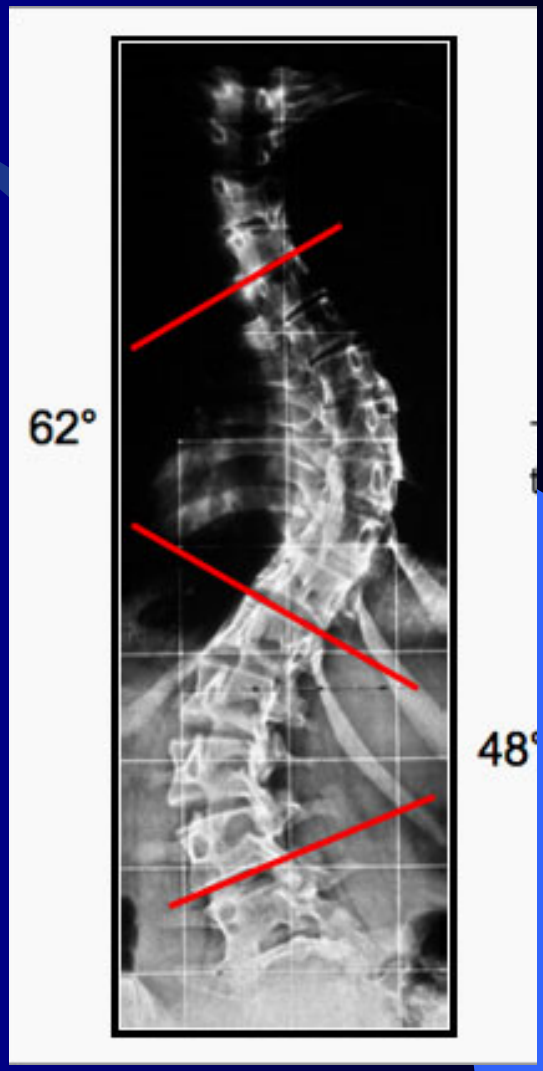
- TO DEFINE CORRECTABLE CAUSES/ PREVENT PROGRESSIVE NEURO LOSS
  - Tethered cord, hydromyelia, tumor
- TO EVAL. CONDITIONS THAT SHOULD BE TX PRIOR TO SCOLIOSIS TX (to offer stabilization / improvement of scoli)
  - tethered cord / diastematomyelia/ ARM
- TO PREVENT NEUROLOGIC INJURY AT TIME OF INSTRUMENTATION
  - tethered cord, intraspinal tumor



SCOLIOSIS

RECOMMEND MRI

- PTS w/ NEURO CHANGES
  - PTS C/O atypical BACK OR LEG PAIN
- PTS w/ NEUROCUT. STIGMATA
- PTS UNDERGOING SPINE STABILIZ for EARLY ONSET SCOLI
- RAPIDLY CHANGING CURVES
  - >10-12 DEGREES / YR




THANK YOU
R
MP
Page: 1 of 1
Compressed 27:1
IM: 1