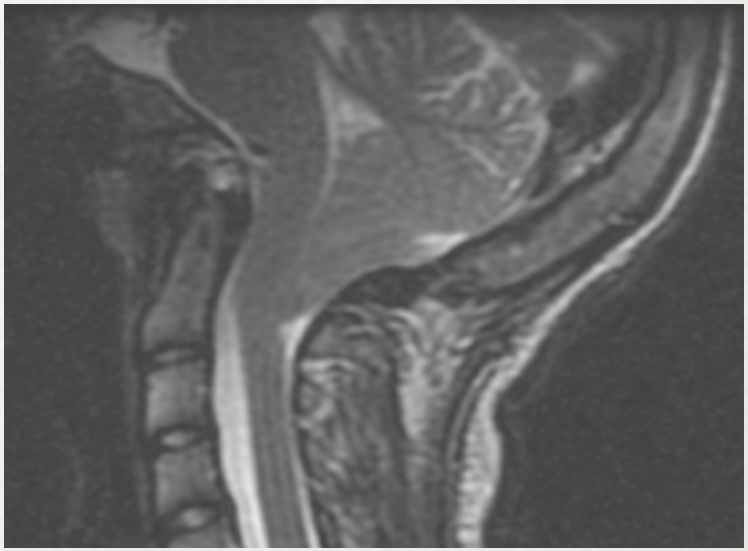
COMPLEX CHIARI MALFORMATIONS: RECOGNITION AND MANAGEMENT STRATEGIES



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CESME, TURKEY, NOVEMBER 16, 2012

CHIARI I

2004
3 YEAR OLD BOY
SUBOCCIPITAL HEADACHES
SWALLOWING DIFFICULTY

2010
9 YEARS OLD
HEADACHES GONE
DOING WELL IN SCHOOL

SURGERY: SOD, TONSILLAR SHRINKING AND DURAPLASTY

CHIARI 1.5

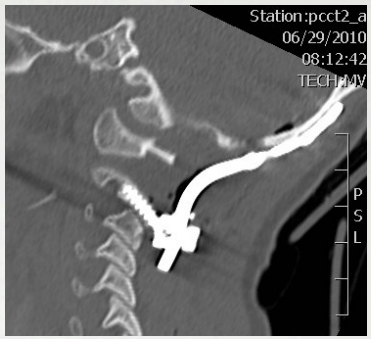
2005
2 YEARS OLD
"DEVELOPMENTAL DELAY"

2007
4 YEARS OLD
"IMPROVED"

2009
6 YEARS OLD
DROOLING, SNORING

SURGERY: SOD, TONSILLAR SHRINKING AND DURAPLASTY

CHIARI 1.5



2009
PRE-OP CT

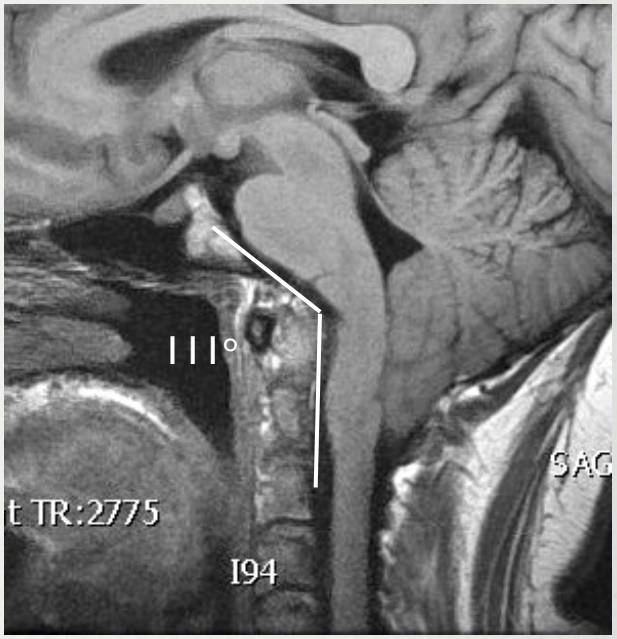
2010
POST-OP CT

SURGERY: RE-DO CHIARI, POSTERIOR O-C2 FUSION WITH ODONTOID REDUCTION

COMPARISON

3 YEAR OLD BOY

2 YEAR OLD GIRL



21 YEAR OLD GIRL

TONSILLAR AND BRAINSTEM HERNIATION (CHIARI1.5)

ANTERIOR BRAINSTEM COMPRESSION (PBC2)
MEDULLARY KINK
CRANIOCERVICAL ANGULATION

CHIARI MALFORMATIONS

CHIARI 0--SYRINGOMYELIA WITHOUT HINDBRAIN HERNIATION

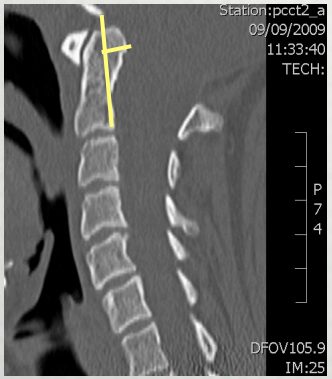
CHIARI1--TONSILLAR HERNIATION >5 MM BELOW FORAMEN MAGNUM, USUALLY WITH PEGGED TONSILLAR TIPS AND CROWDING AT THE CVJ

CHIARI 1.5--TONSILLAR, BRAINSTEM AND 4TH VENTRICLE HERNIATION

ANTERIOR BRAINSTEM COMPRESSION (pBC2)

Neurosurgery. 1999 Mar;44(3):520-7; discussion 527-8.
Ventral brain stem compression in pediatric and young adult patients with Chiari I malformations..
Grabb PA, Mapstone TB, Oakes WJ.

"COMPLEX" CHIARI MALFORMATIONS



TONSILLAR AND BRAINSTEM HERNIATION (CHIARI 1.5)

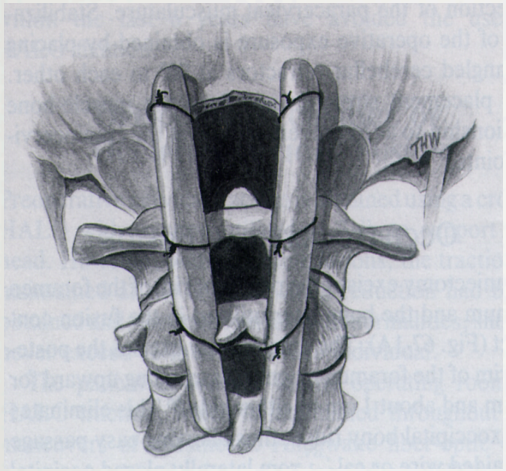
ANTERIOR BRAINSTEM COMPRESSION

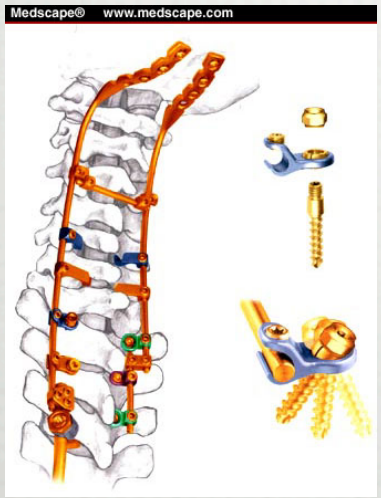
MEDULLARY KINK

CRANIOCERVICAL ANGULATION

SYRINGOMYELIA AND SCOLIOSIS?

THE EVOLUTION OF COMPLEX CHIARI MALFORMATION MANAGEMENT





1) GARDNER WELLS TRACTION
TRANSORAL ODONTOID RESECTION
POSTERIOR O-C RIB GRAFT FUSION
HALO APPLICATION

2)TRANSORAL ODONTOID RESECTION
POSTERIOR O-C2 INTRUMENTATION AND FUSION

3)POSTERIOR O-C2 INTRUMENTATION
ODONTOID REDUCTION AND FUSION
ENDOSCOPIC TRANSNASAL ODONTOID RESECTION
(IF NEEDED)

PCMC "COMPLEX" CHIARI EXPERIENCE:

- 1995 to 2010
- 210 consecutive Chiari patients operated
- 168 met criteria for Chiari I
- 42 met criteria for "Complex Chiari" either by MRI findings or clinical description
- Complete film set available for 101 patients
- These 101 patients formed our study group
- Analyzed for risk factors determining need for occipitocervical fusion

PCMC "COMPLEX" CHIARI EXPERIENCE:

- 64 Chiari I
- 37 "Complex Chiari"

11 UP-FRONT O-C FUSIONS
3 TOR

8 REOPERATIONS FOR O-C FUSION
0 TOR

0 REOPERATIONS FOR
O-C FUSION

INDICATIONS FOR FUSION/REDUCTION

"Complex Chiari"

PLUS

Bulbar symptoms

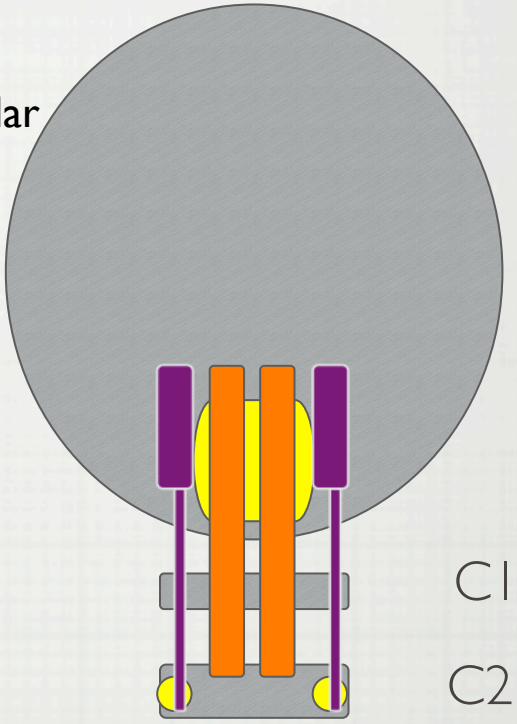
Myelopathy

Severe headaches

Progressive or unresolved syrinx

SURGICAL TECHNIQUE

- SOD and C1 laminectomy or Redo-Chiari exploration with tonsillar shrinking and duraplasty
- Bilateral C2 (C3) pars screws
- O-C2 Rod-plate construct
- +/- Odondoid reduction
- Rib graft x 2
- Cable and maxillofacial screw fixation
- DBX



J Neurosurg. 2004 Nov;101(2 Suppl):189-95.

Treatment of basilar invagination associated with Chiari I malformations in the pediatric population: cervical reduction and posterior occipitocervical fusion.
Kim LJ, Rekate HL, Klopfenstein JD, Sonntag VK.

FOLLOW-UP

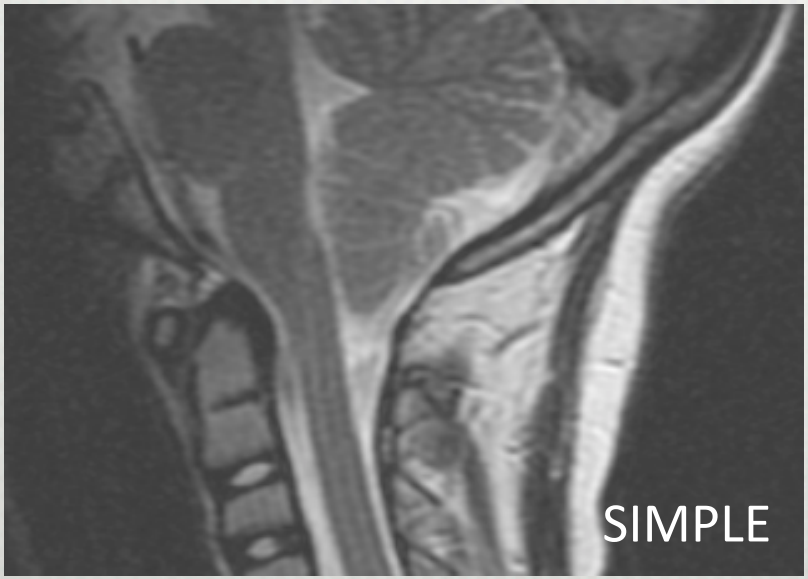
- All 19 patients have had successful arthrodesis
- Complete follow-up data is forthcoming, but all post-fusion patients have had improvement of their pre-operative symptoms, oftentimes dramatically
- ALL Syrinxes and scoliosis have stabilized or improved

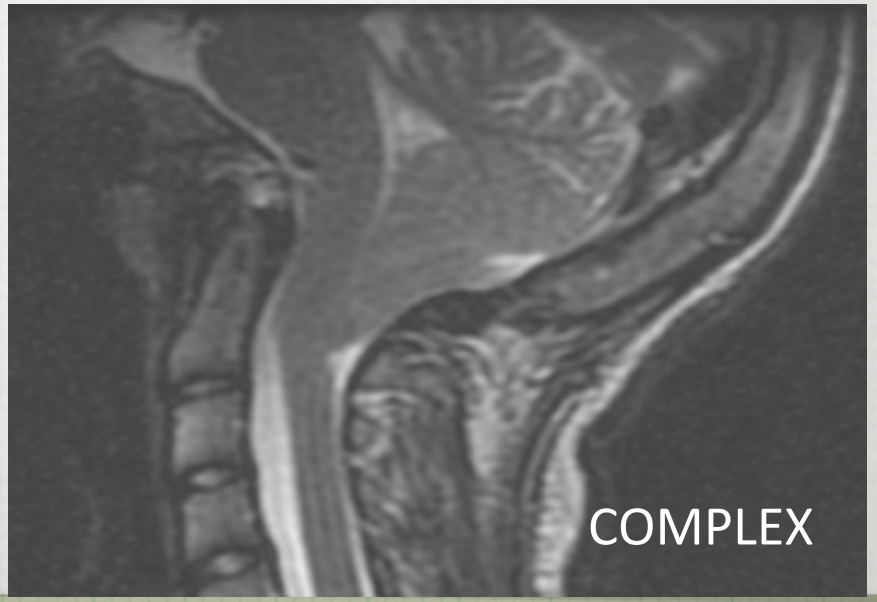
HYPOTHESIS

- CHIARI I MALFORMATION ACTUALLY REPRESENTS A SPECTRUM OF DISEASE.

- PATIENTS WHO FAIL TO RESPOND TO SIMPLE DECOMPRESSION OFTEN HAVE COMPLEX ANOMALIES OF THE CVJ & O-C INSTABILITY REQUIRING FUSION.

CERTAIN RISK FACTORS MY ALLOW EARLY IDENTIFICATION AND IMPROVED MANAGEMENT OF "COMPLEX" CHIARI PATIENTS.





STUDY DESIGN

- IRB-APPROVED REVIEW OF CLINICAL AND RADIOGRAPHIC DATA IN 101 CHILDREN UNDERGOING SURGERY FOR CHIARI MALFORMATION BETWEEN 1995-2010 AT PRIMARY CHILDREN'S MEDICAL CENTER.

- PATIENTS WITH CHIARI 2 MALFORMATION WERE EXCLUDED.

POSSIBLE CLINICAL RISK FACTORS:

age at surgery
length of follow-up
requirement for reoperation
gender
secondary diagnosis

possible radiographic risk factors:

SCOLIOSIS
CHIARI TYPE (1 OR 1.5)
TONSILAR DESCENT
VENTRAL COMPRESSION (PBC2)
SYRINGOMYELIA
MEDULLARY KINK
CLIVUS-AXIS ANGLE (CXA)
BASILAR INVAGINATION

✦ *UNIVARIATE & MULTIVARIATE REGRESSION (COX PROPORTIONAL HAZARDS) ANALYSES USING TIME TO FUSION AS PRIMARY OUTCOME.*

RADIOGRAPHIC PARAMETERS

CHIARI 1.5

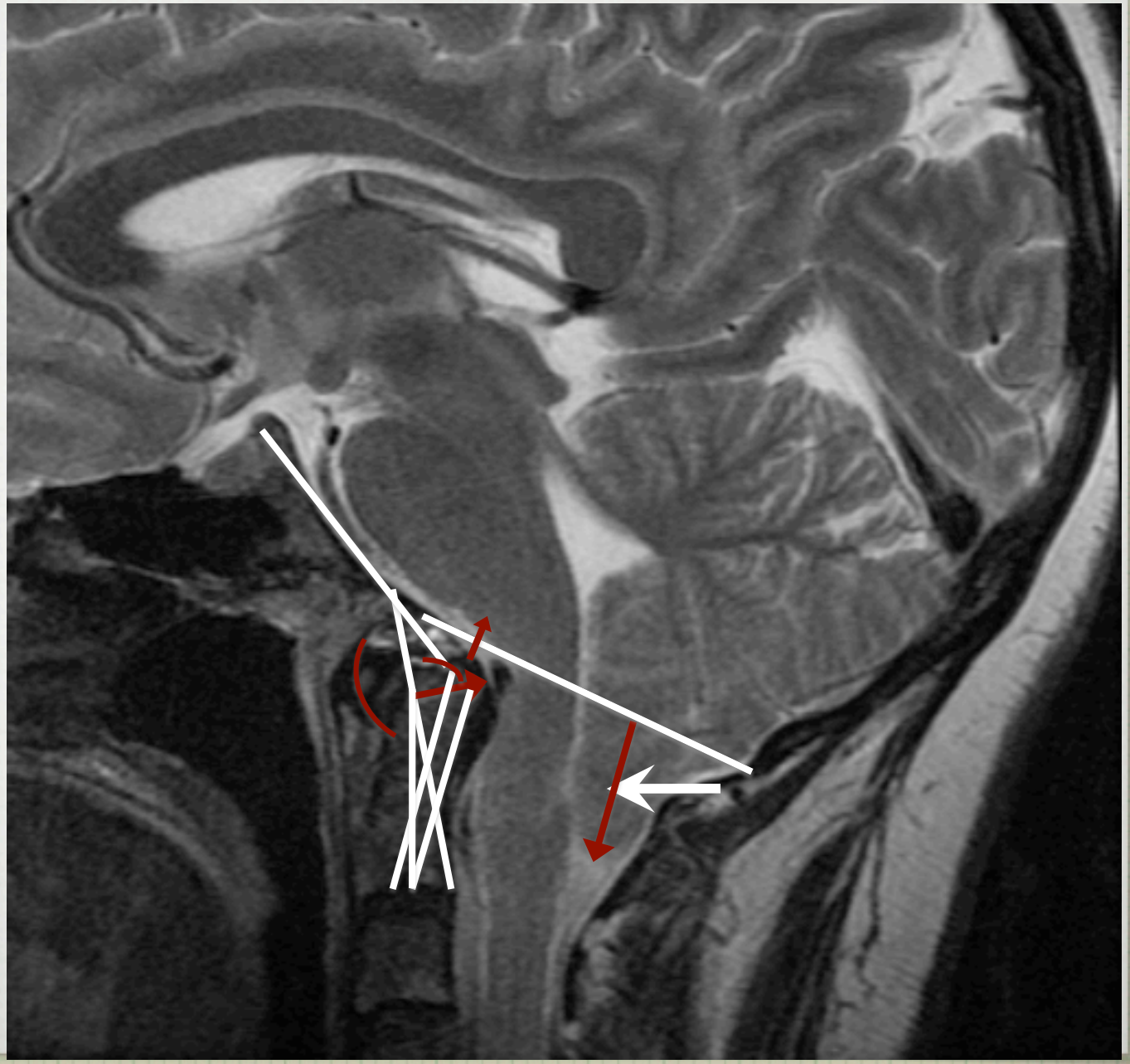
PBC2

CXA

TONSILAR DESCENT

RETROFLEXED ODONTOID

BASILAR INVAGINATION



PATIENT DEMOGRAPHICS

variable	*decompression*	*fusion*	*p value*
# pts	**82**	**19**	
male	46 (56%)	8 (42%)	NS
mean age	8.7	11.1	0.08
range (SD)	0.7 - 16.8 (5.1)	1.9 - 21.9 (6.5)	
mean f/u	2.2	2.6	NS
range (SD)	0.1 - 7.8 (1.9)	0.1 - 9.3 (2.7)	

UNIVARIATE ANALYSIS

variable	decompression	fusion	univariate p value
Chiari 1.5	18 (22%)	18 (95%)	<0.001
Chiari 1	64 (78%)	1 (5%)	
scoliosis	20 (24%)	2 (11%)	NS
no scoliosis	62 (76%)	17 (89%)	
syrinx	42 (51%)	9 (47%)	NS
no syrinx	40 (49%)	10 (53%)	
kink	21 (26%)	15 (79%)	<0.001
no kink	61 (74%)	4 (21%)	
retroflexed	30 (37%)	13 (68%)	0.01
no retroflexion	52 (63%)	6 (32%)	
basilar invagination	0 (0%)	7 (37%)	<0.001
no BI	82 (100%)	12 63%)	

UNIVARIATE ANALYSIS

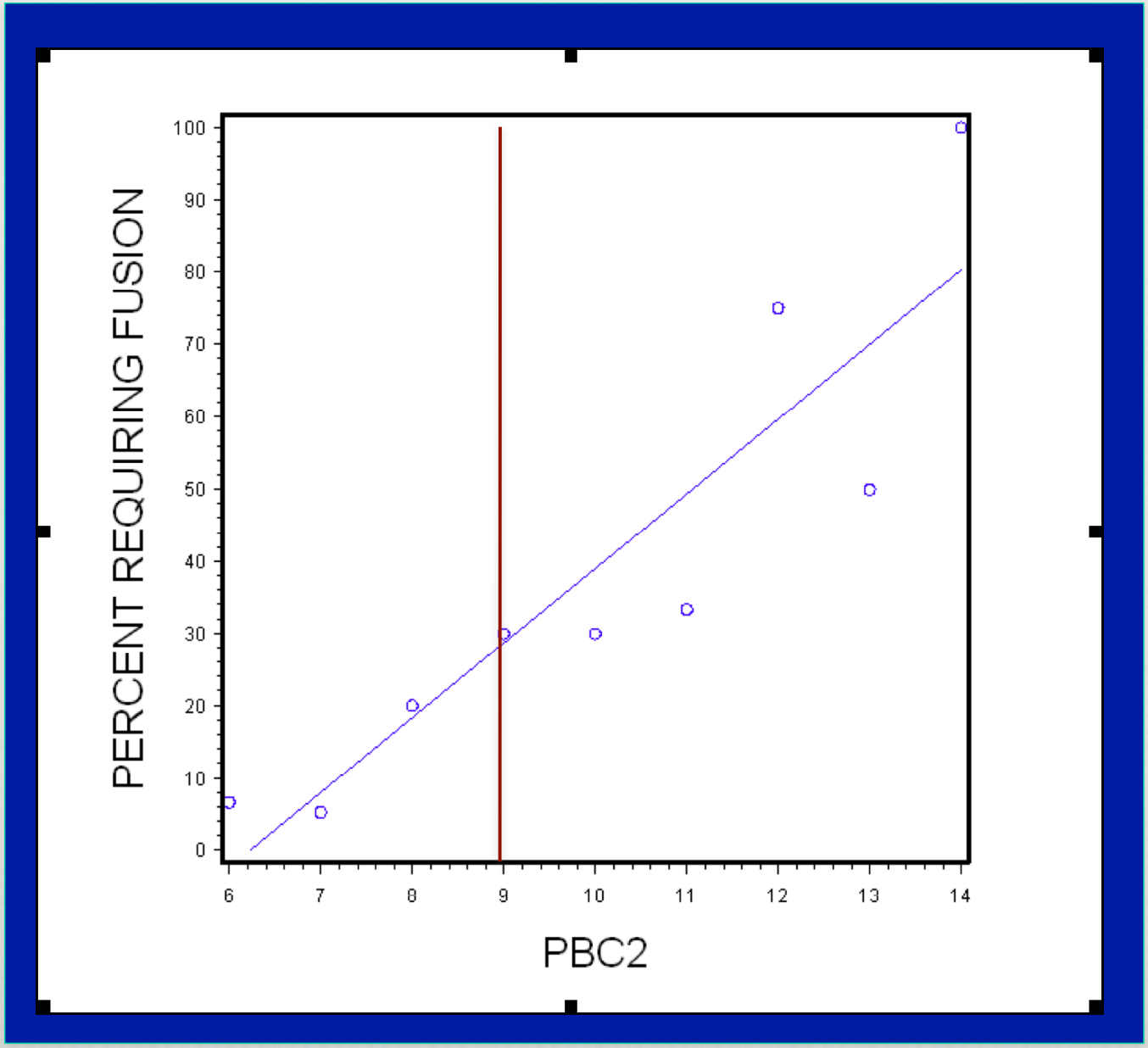
variable	decompression	fusion	univariate p value
pBC2			
mean (SD)	7.2 (2.1)	10.2 (2.3)	<0.001
pBC2 ≥ 9	20 (24%)	14 (74%)	<0.001
pBC2 < 9	62 (76%)	5 (26%)	
tonsilar descent			
mean (SD)	13.0 (5.1)	16.3 (6.5)	0.02
tonsils > 15	24 (30%)	9 (47%)	NS
tonsils ≤ 15	57 (70%)	10 (53%)	
CXA			
mean (SD)	141.1 (14.6)	115.2 (17.2)	<0.001
CXA < 125	9 (11%)	15 (79%)	<0.001
CXA ≥ 125	70 (89%)	4 (21%)	

MULTIVARIATE ANALYSIS

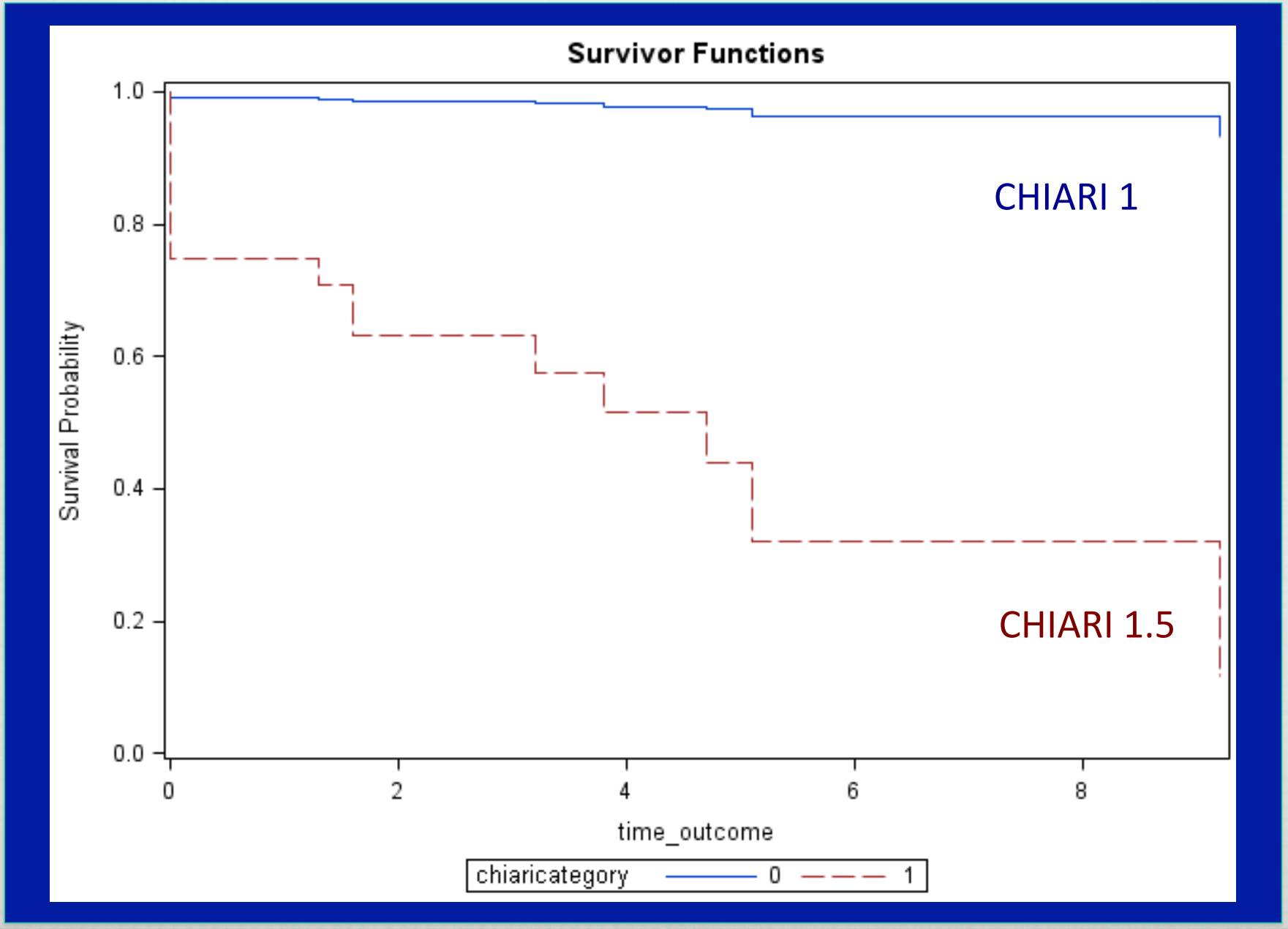
variable	univariate p value	hazard ratio (95% CI)	multivariate p value
Chiari 1.5	<0.001	**14.7 (1.8 - 122.5)***	<0.001
Chiari 1			
kink	<0.001		NS
no kink			
retroflexed	0.01		NS
no retroflexion			
basilar invagination	<0.001	**9.8 (2.2-44.2)**	<0.001
no BI			
CXA			
mean (SD)	<0.001		<0.001
CXA < 125	<0.001	**3.9 (1.2 - 12.6)***	<0.001
CXA ≥ 125			
tonsilar descent			
mean (SD)	0.02		NS
tonsils > 15	NS		NS
tonsils ≤ 15			
pBC2			
mean (SD)	<0.001		
pBC2 ≥ 9	<0.001	***co-linear with CXA***	
pBC2 < 9			

* neither Chari type or CXA violated the proportional hazards assumption

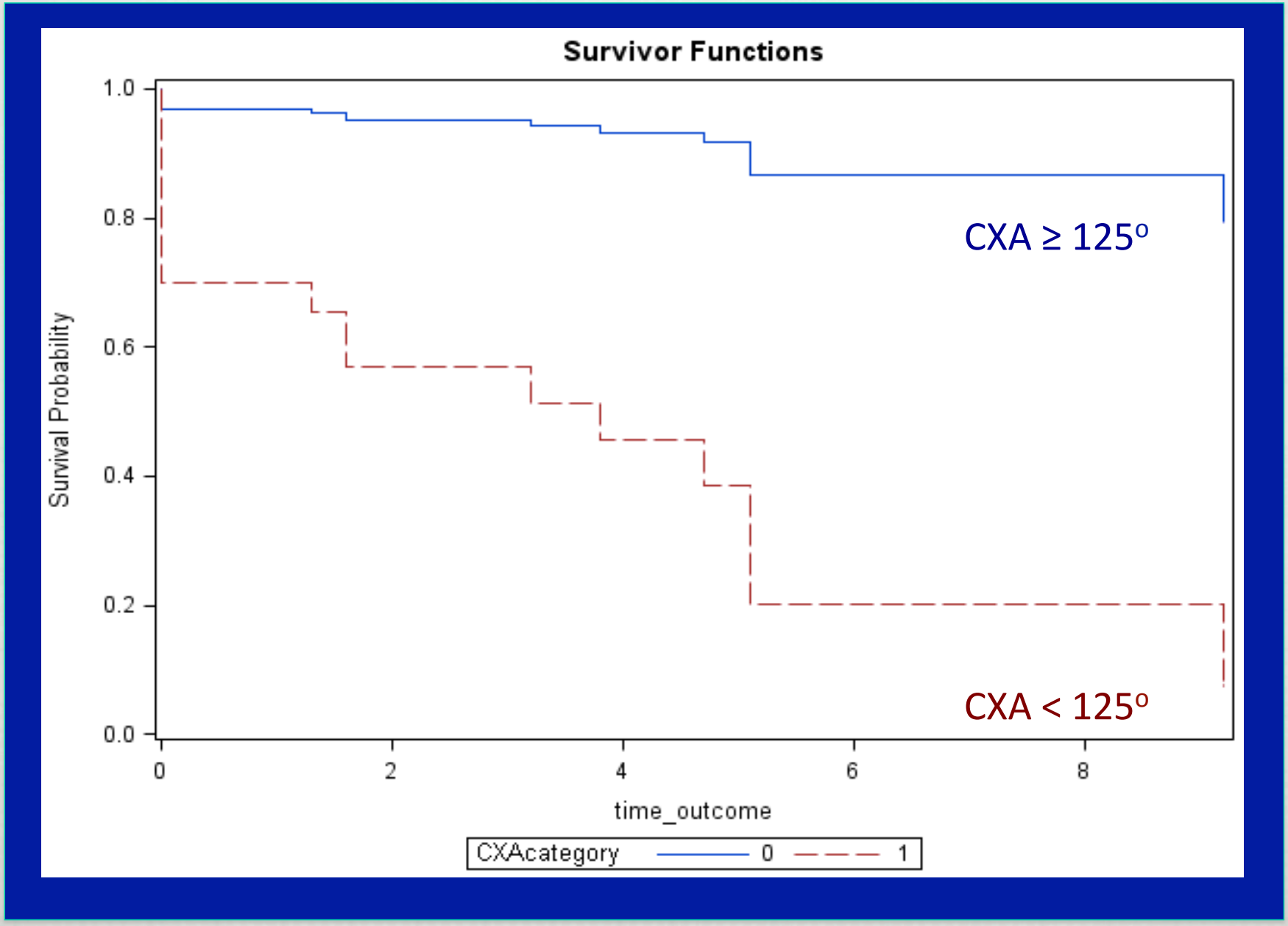
PBC2 > 9


PERCENT REQUIRING FUSION
100
90
80
70
60
50
40
30
20
10
0
6
7
8
9
10
11
12
13
14
PBC2

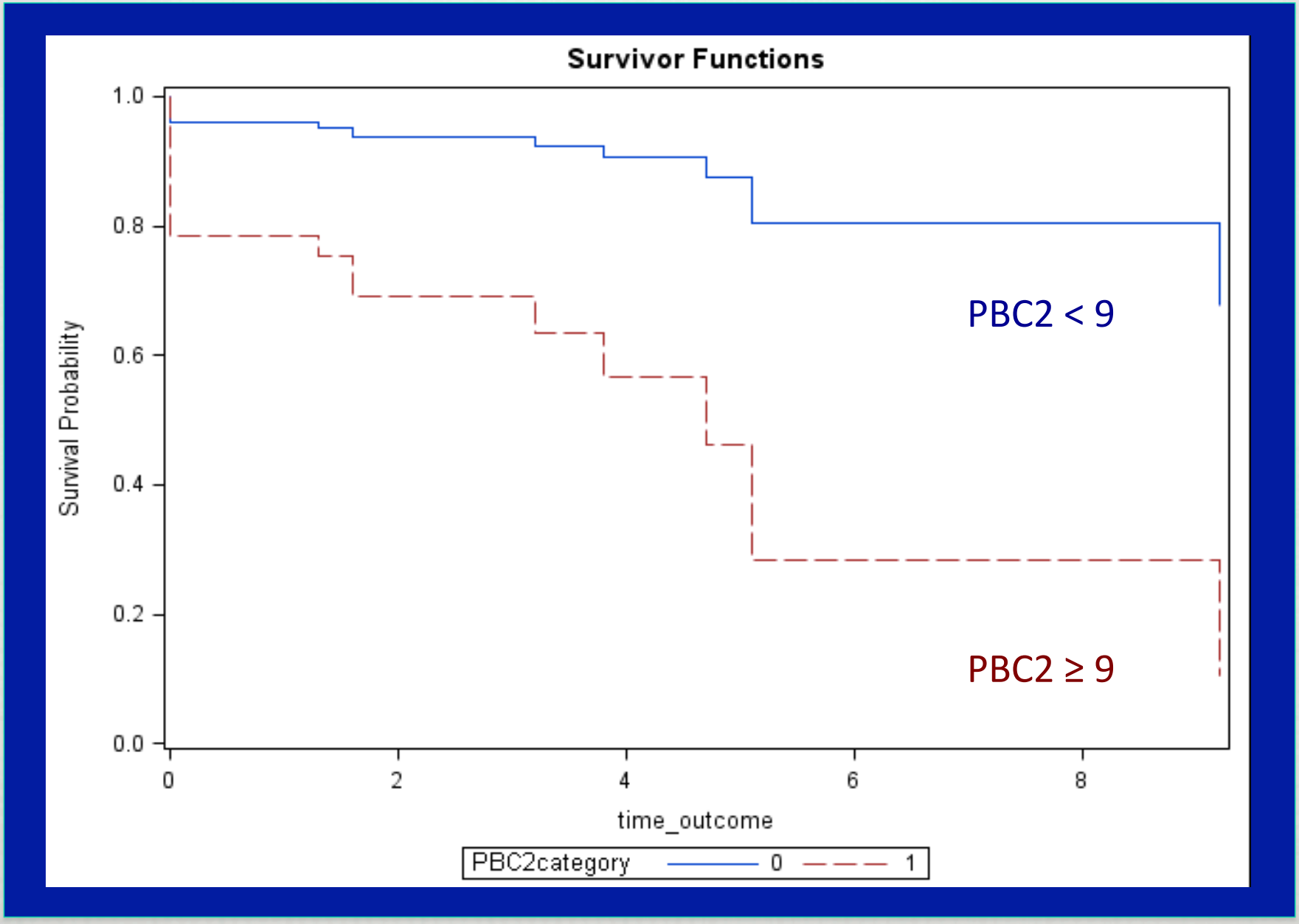
TIME TO FUSION: CHIARI TYPE



CLIVUS-AXIS ANGLE



PBC2 (VENTRAL COMPRESSION)

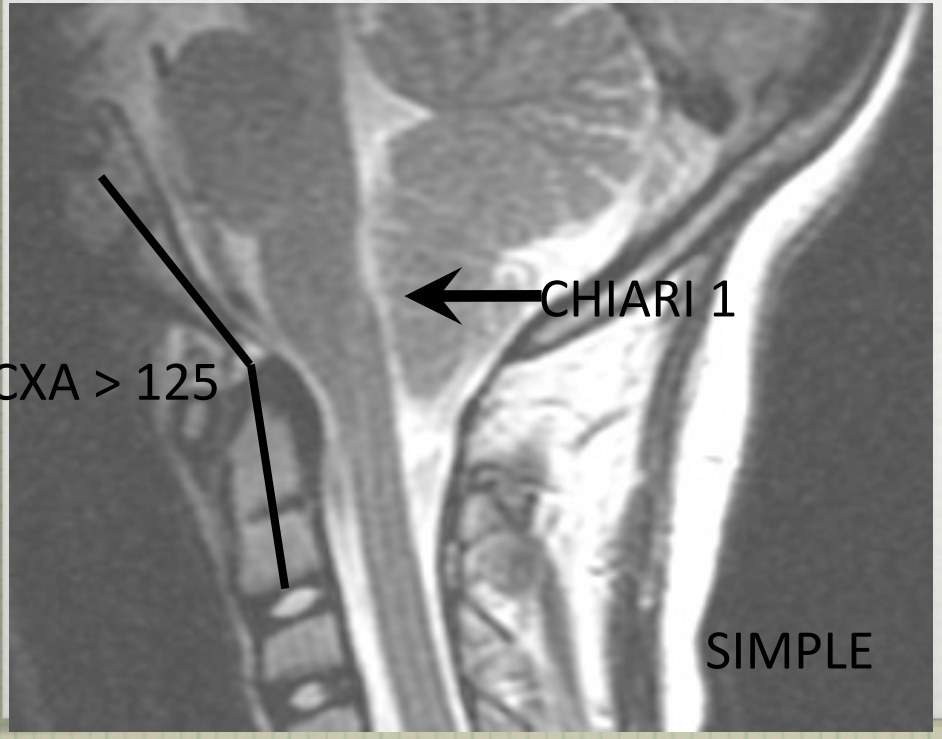


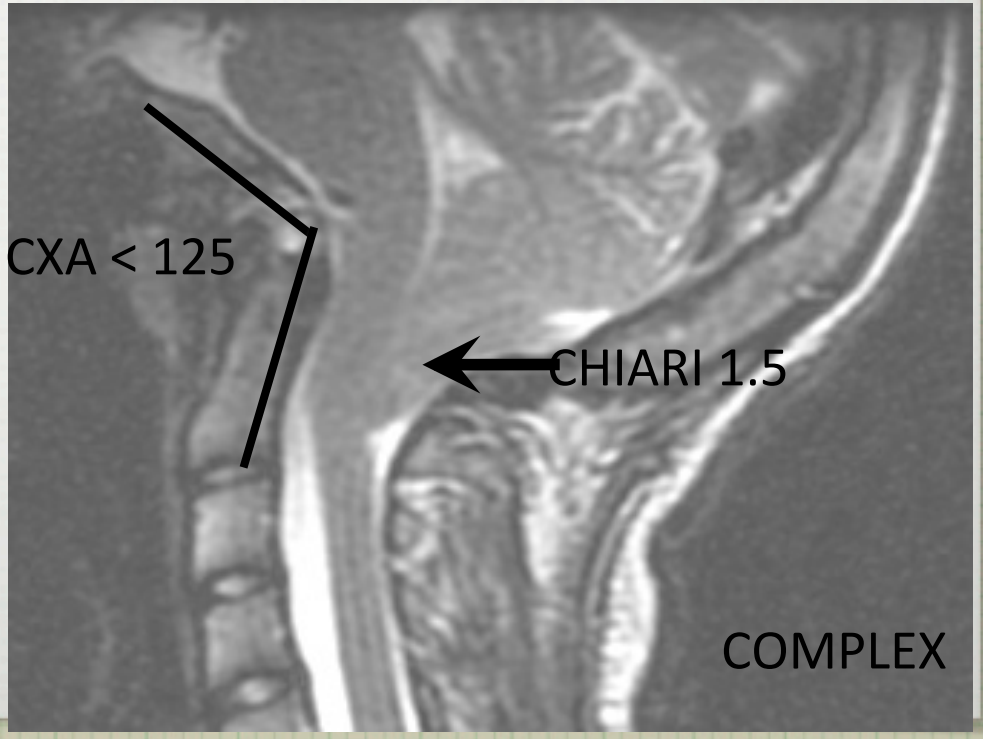
RESULTS

HIGH RISK: CHIARI 1.5 & CXA < 125° 83.3% fusion

INTERMEDIATE RISK: CHIARI 1.5 OR CXA < 125° 13% fusion

LOW RISK: CHIARI 1 & CXA > 125° 1.7% fusion

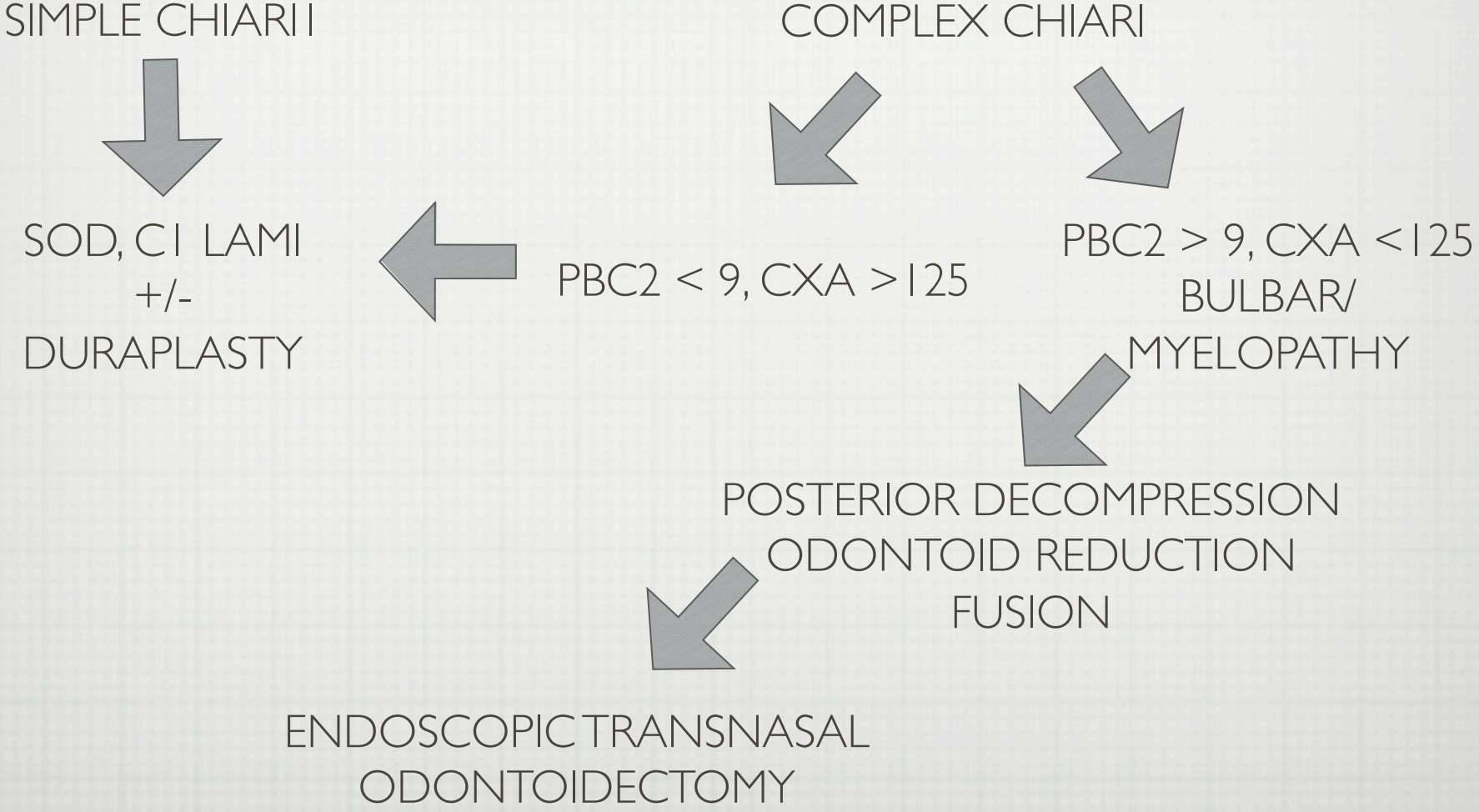


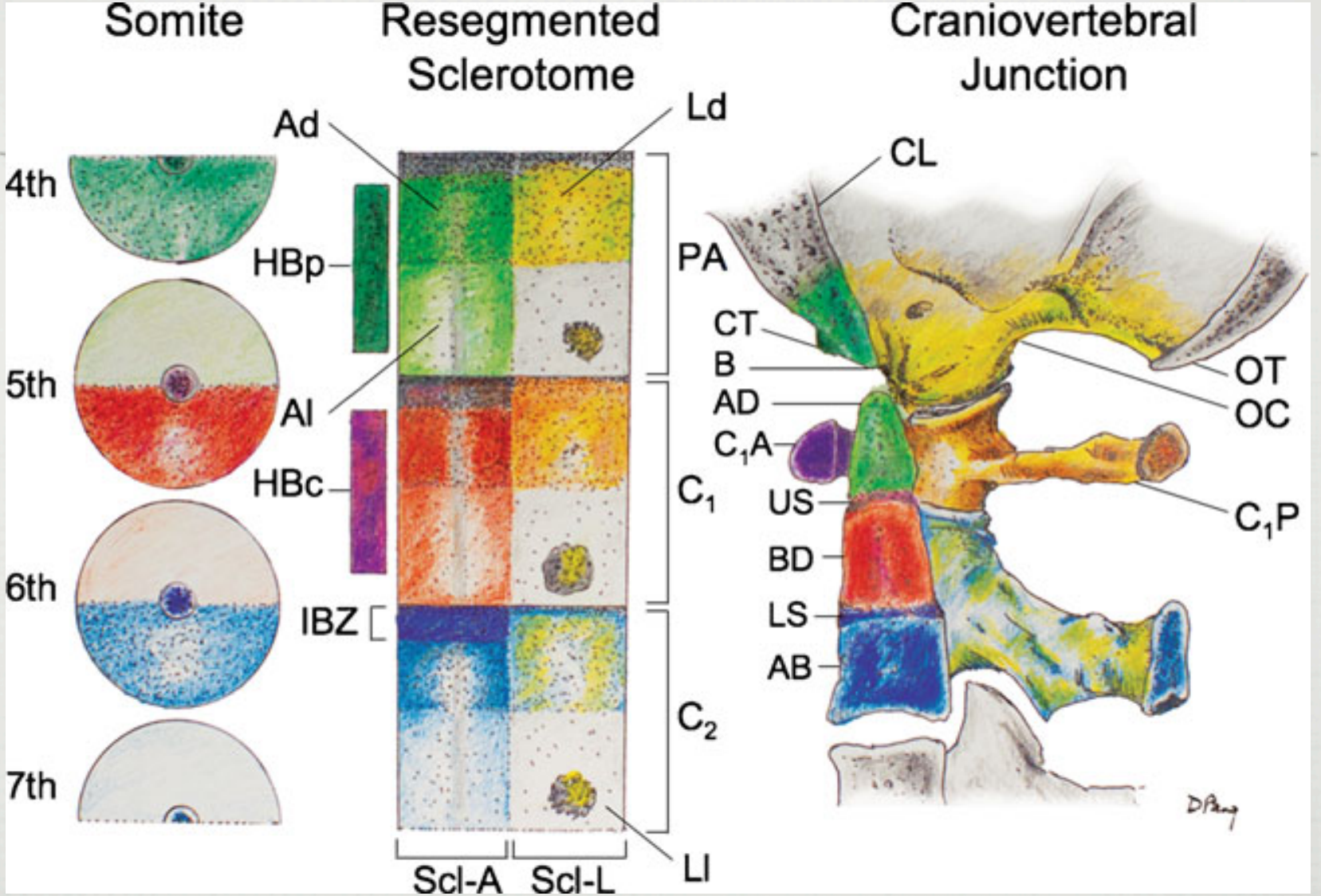


WHERE DOES THIS LEAVE US?

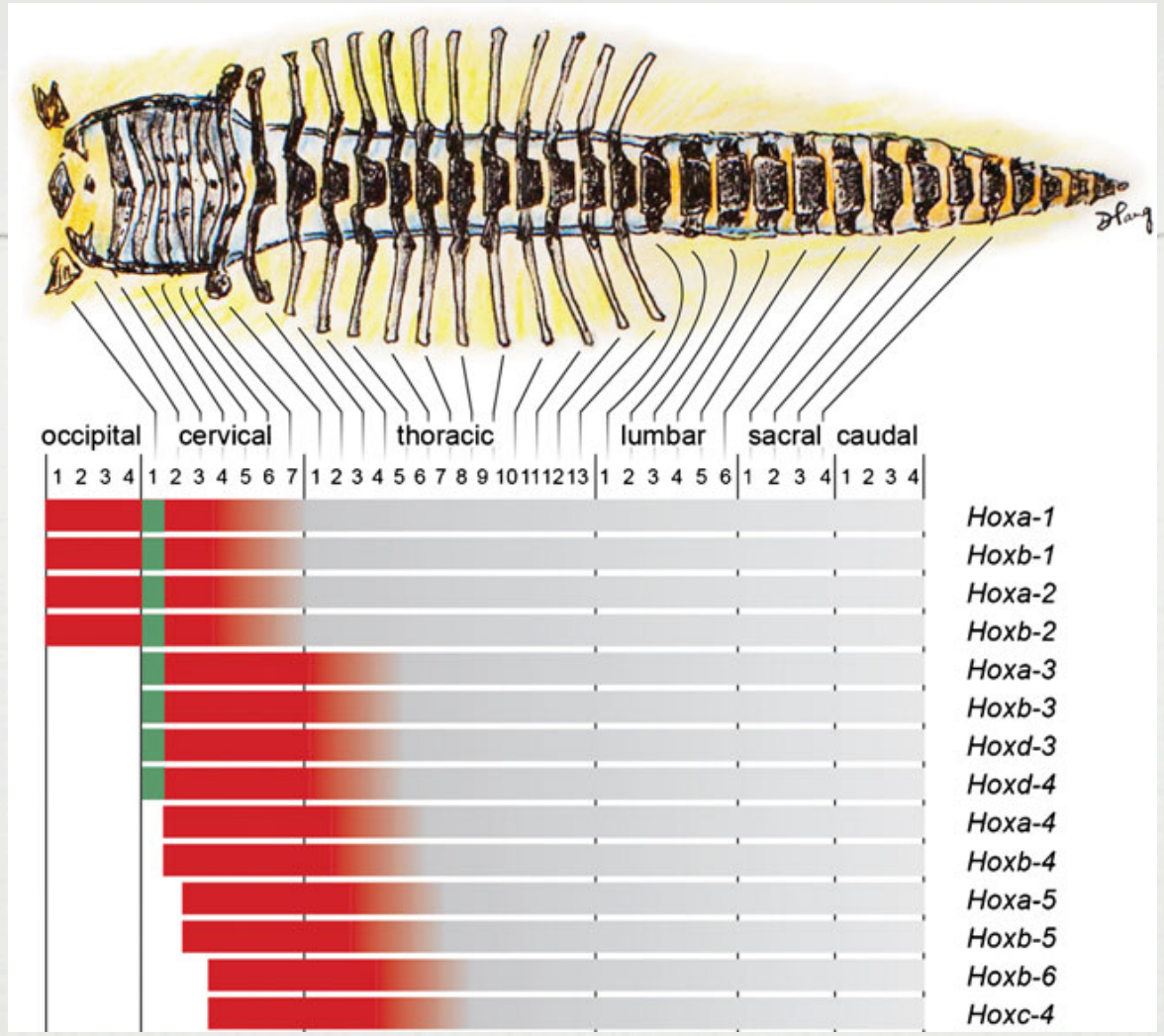
- Patients with "Complex" Chiari malformation are very challenging
- Multiple issues must be recognized and pursued:
- Symptom management
- Biomechanical stress leading to failure
- Syringomyelia and scoliosis
- Avoidance of excessive procedures
- With modern techniques, can or should this condition be managed with one posterior procedure??
- Where does odontoidectomy fit in?

MANAGEMENT ALGORITHM





Embryology and bony malformations of the craniovertebral junction
Dachling Pang & Dominic N. P. Thompson Childs Nerv Syst (2011) 27:523–564



HOX GENES CONTROL THE POSITIONAL IDENTITY OF PREVERTEBRAL SEGMENTS

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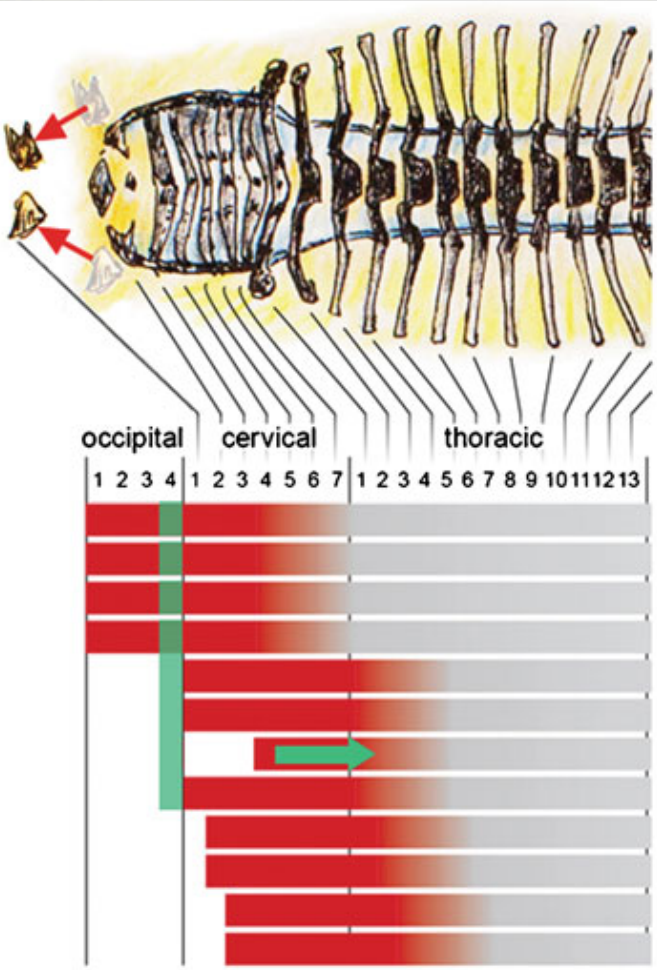

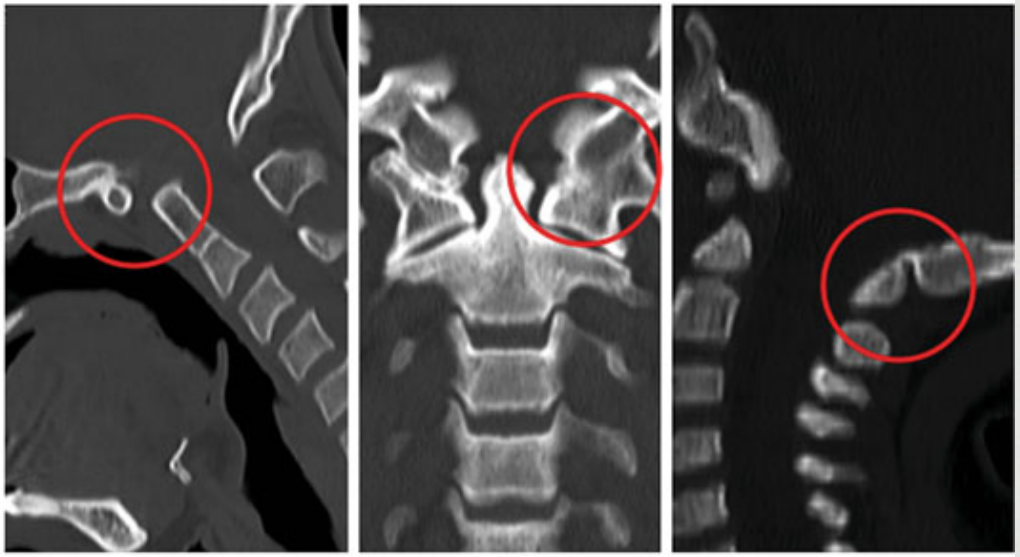

HOX D-3 MUTATION LEADS TO PARTIAL LOSS OF C1 EXPRESSION DOMAIN

THE C1 SCLEROTOME "BEHAVES" LIKEAN OCCIPITAL SCLEROTOME

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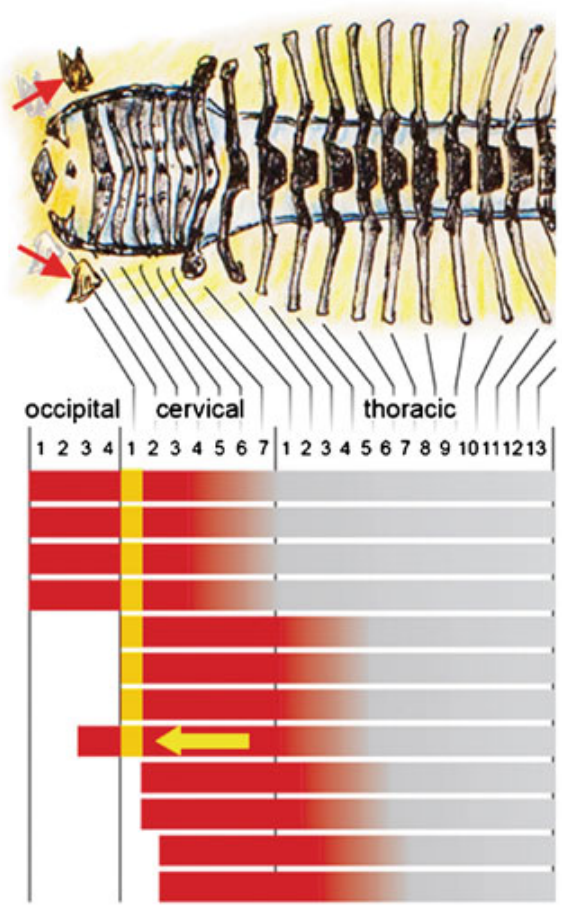

HOX D-3 MUTATION LEADS TO "GAIN OF FUNCTION" IN FOURTH OCCIPITAL SCLEROTOME

THE OCCIPUT "IMITATES" THE C1 SEGMENT

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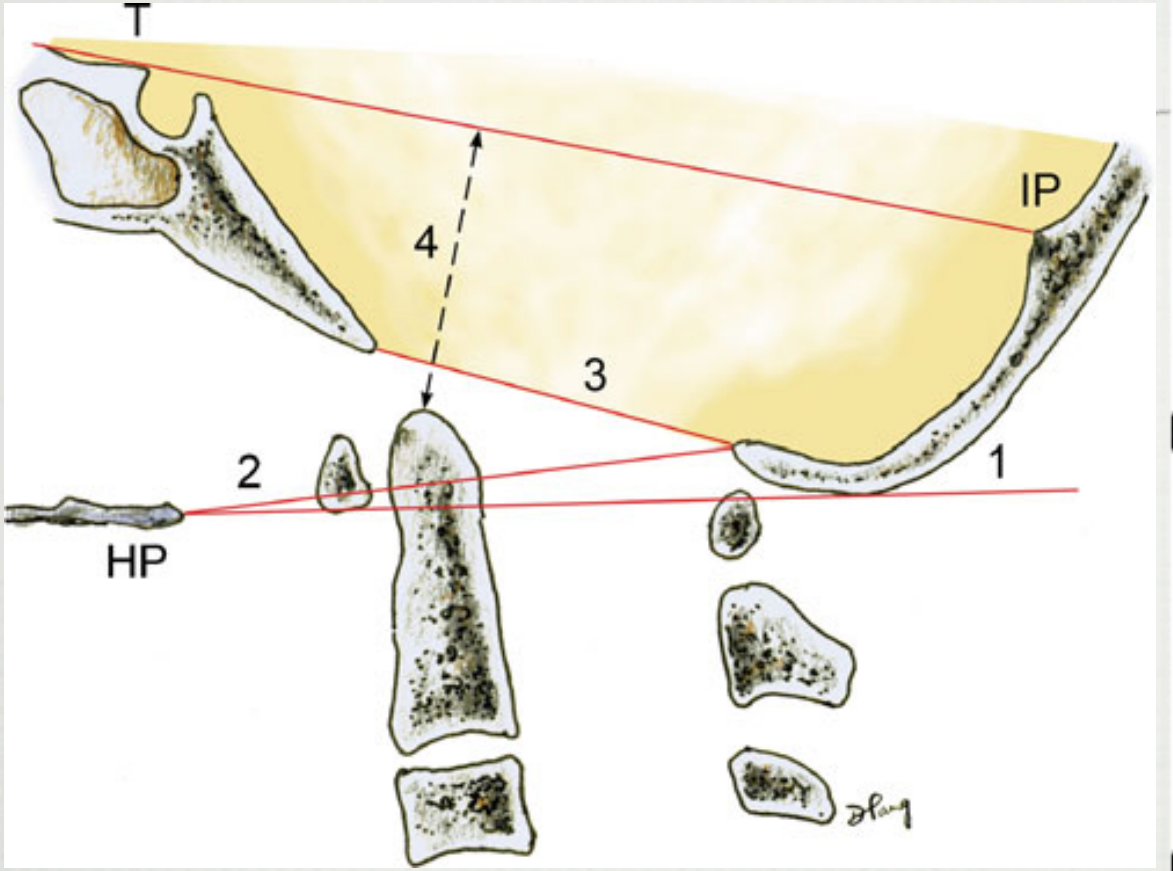

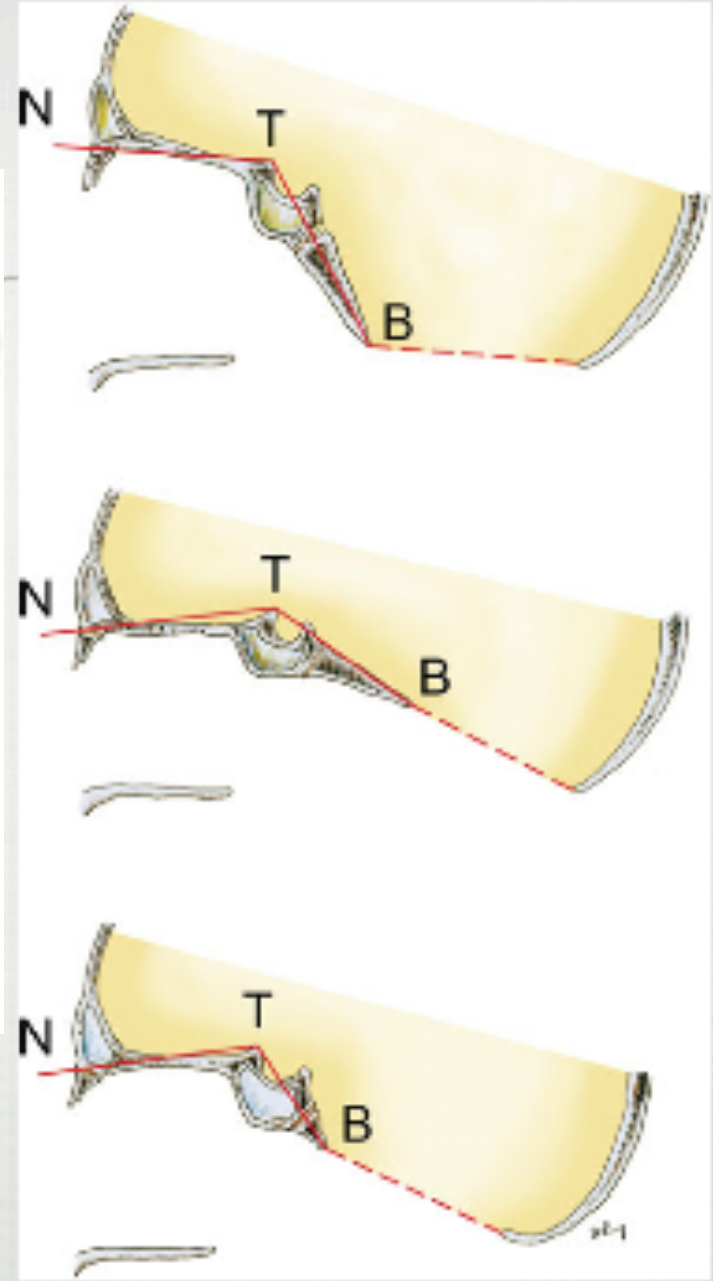

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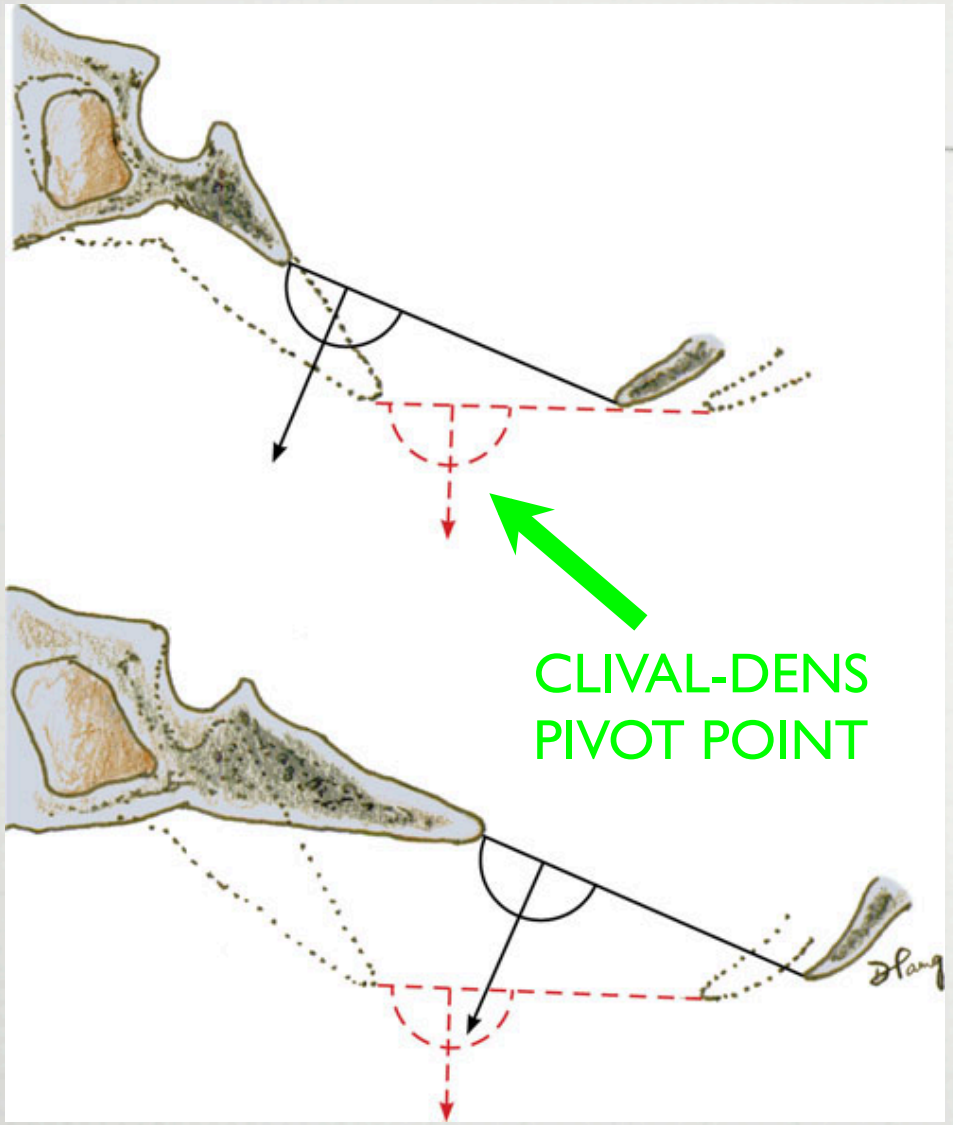

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BIOMECHANICAL HYPOTHESIS

"COMPLEX" CHIARI WITH RISK FACTORS
(CHIARI 1.5 AND CXA < 125°)

↓

DORSAL DECOMPRESSION AND RELEASE
OF POSTERIOR TENSION BAND

↓

CRANIAL SETTLING AND/OR ACCENTUATION OF THE
FORWARD BENDING MOMENT OF THE CLIVAL-DENS PIVOT POINT

↓

FORWARD FOLDING OF THE CRANIOCERVICAL ANGLE

↓

PROGRESSIVE BRAINSTEM COMPRESSION AND WORSENING
SIGNS AND SYMPTOMS

CHIARI I MALFORMATIONS

HYDRODYNAMIC IMBALANCE

+/- SMALL POSTERIOR FOSSA VOLUME

"COMPLEX" CHIARI MALFORMATIONS

GENETICALLY-DRIVEN SKULL BASE MORPHOLOGY

CLIVAL-CERVICAL RELATIONSHIP
ODONTOID RETROFLEXION
BASILAR INVAGINATION

BIOMECHANICAL STRESS AND POTENTIAL FAILURE

PRE-OP ⇨ SLOW

POST-OP ⇨ RAPID

HYDRODYNAMIC FACTORS