COMPLEX CHIARI MALFORMATIONS: RECOGNITION AND MANAGEMENT STRATEGIES

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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
SURGERY: SOD, TONSILLAR SHRINKING AND DURAPLASTY

2005
2 YEARS OLD
“DEVELOPMENTAL DELAY”

2007
4 YEARS OLD
“IMPROVED”

2009
6 YEARS OLD
DROOLING, SNORING

CHIARI 1.5
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

CHIARI I--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)

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


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

![HYPOTHESIS](image)

- 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 MAY ALLOW EARLY IDENTIFICATION AND IMPROVED MANAGEMENT OF “COMPLEX” CHIARI PATIENTS.

![SIMPLE](image)

![COMPLEX](image)
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

<table>
<thead>
<tr>
<th>variable</th>
<th>decompression</th>
<th>fusion</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td># pts</td>
<td>82</td>
<td>19</td>
<td>NS</td>
</tr>
<tr>
<td>male</td>
<td>46 (56%)</td>
<td>8 (42%)</td>
<td>NS</td>
</tr>
<tr>
<td>mean age</td>
<td>8.7</td>
<td>11.1</td>
<td>0.08</td>
</tr>
<tr>
<td>range (SD)</td>
<td>0.7 - 16.8 (5.1)</td>
<td>1.9 - 21.9 (6.5)</td>
<td></td>
</tr>
<tr>
<td>mean f/u</td>
<td>2.2</td>
<td>2.6</td>
<td>NS</td>
</tr>
<tr>
<td>range (SD)</td>
<td>0.1 - 7.8 (1.9)</td>
<td>0.1 - 9.3 (2.7)</td>
<td></td>
</tr>
<tr>
<td>variable</td>
<td>decompression</td>
<td>fusion</td>
<td>univariate p value</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------------</td>
<td>----------</td>
<td>--------------------</td>
</tr>
<tr>
<td><strong>Chiari 1.5</strong></td>
<td>18 (22%)</td>
<td>18 (95%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>Chiari 1</strong></td>
<td>64 (78%)</td>
<td>1 (5%)</td>
<td></td>
</tr>
<tr>
<td><strong>scoliosis</strong></td>
<td>20 (24%)</td>
<td>2 (11%)</td>
<td>NS</td>
</tr>
<tr>
<td><strong>no scoliosis</strong></td>
<td>62 (76%)</td>
<td>17 (89%)</td>
<td></td>
</tr>
<tr>
<td><strong>syrinx</strong></td>
<td>42 (51%)</td>
<td>9 (47%)</td>
<td>NS</td>
</tr>
<tr>
<td><strong>no syrinx</strong></td>
<td>40 (49%)</td>
<td>10 (53%)</td>
<td></td>
</tr>
<tr>
<td><strong>kink</strong></td>
<td>21 (26%)</td>
<td>15 (79%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>no kink</strong></td>
<td>61 (74%)</td>
<td>4 (21%)</td>
<td></td>
</tr>
<tr>
<td><strong>retroflexed</strong></td>
<td>30 (37%)</td>
<td>13 (68%)</td>
<td>0.01</td>
</tr>
<tr>
<td><strong>no retroflexion</strong></td>
<td>52 (63%)</td>
<td>6 (32%)</td>
<td></td>
</tr>
<tr>
<td><strong>basilar invagination</strong></td>
<td>0 (0%)</td>
<td>7 (37%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>no BI</strong></td>
<td>82 (100%)</td>
<td>12 (63%)</td>
<td></td>
</tr>
</tbody>
</table>
## Univariate Analysis

<table>
<thead>
<tr>
<th>Variable</th>
<th>Decompression</th>
<th>Fusion</th>
<th>Univariate p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>pBC2</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean (SD)</td>
<td>7.2 (2.1)</td>
<td>10.2 (2.3)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>pBC2 ≥ 9</td>
<td>20 (24%)</td>
<td>14 (74%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>pBC2 &lt; 9</td>
<td>62 (76%)</td>
<td>5 (26%)</td>
<td></td>
</tr>
<tr>
<td><strong>Tonsilar Descent</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean (SD)</td>
<td>13.0 (5.1)</td>
<td>16.3 (6.5)</td>
<td>0.02</td>
</tr>
<tr>
<td>Tonsils &gt; 15</td>
<td>24 (30%)</td>
<td>9 (47%)</td>
<td>NS</td>
</tr>
<tr>
<td>Tonsils ≤ 15</td>
<td>57 (70%)</td>
<td>10 (53%)</td>
<td></td>
</tr>
<tr>
<td><strong>CXA</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean (SD)</td>
<td>141.1 (14.6)</td>
<td>115.2 (17.2)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>CXA &lt; 125</td>
<td>9 (11%)</td>
<td>15 (79%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>CXA ≥ 125</td>
<td>70 (89%)</td>
<td>4 (21%)</td>
<td></td>
</tr>
</tbody>
</table>
## MULTIVARIATE ANALYSIS

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

* neither Chari type or CXA violated the proportional hazards assumption
PBC2 > 9
TIME TO FUSION: CHIARI TYPE

Survivor Functions

CHIARI 1

CHIARI 1.5

Survival Probability vs. time_outcome
CLIVUS-AXIS ANGLE

Survivor Functions

Survival Probability

CXA ≥ 125°

CXA < 125°

time_outcome

CXAc category 0 1
PBC2 (VENTRAL COMPRESSION)

Survivor Functions

Survival Probability

PBC2 < 9

PBC2 ≥ 9

time_outcome

PBC2category 0 1
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

SIMPLE CHIARI I

SOD, C1 LAMI
+/- DURAPLASTY

COMPLEX CHIARI

PBC2 < 9, CXA > 125

PBC2 > 9, CXA < 125
BULBAR MYELOPATHY

POSTERIOR DECOMPRESSION
ODONTOID REDUCTION
FUSION

ENDOSCOPIC TRANSNASAL ODONTOIDECTOMY
Thus, the neural arch of the vertebra is derived from the caudal-lateral part of a single somite. However, labelling and transplantation experiments of half-somites have repeatedly demonstrated that each vertebral body is made up of cells from the axial zones of two adjacent somites. Although the exact boundary of individual somite participation is not known, the juxta-positioning of axial and lateral sclerotomal components.

**Fig. 14**
Formation of the human craniovertebral junction. Sclerotomal primordia and their vertebral phenotypes are colour-matched. During resegmentation, the caudal half of the fourth somite (fourth occipital somite) and rostral half of the fifth somite combine to form the proatlas sclerotome (PA). Derived from the proatlas are: the axial zones (Ad and Al) which become the basion (B) of the basioccipital or clivus (CL) and the apical segment of the dens (AD); the lateral dense zone (Ld) becomes the exoccipital comprising the occipital condyle (OC), and lateral rim and opisthion (OT) of the foramen magnum; the proatlas’ hypochordal bow (HBp) forms the ventral clival tubercle (CT). The C1 resegmented sclerotome (C1) comes from adjacent halves of the fifth and sixth somites. Derived from the C1 sclerotome are: the axial zones form the basal segment of the dens (BD); the lateral zone forms the posterior atlantal arch (C1P); the hypochordal bow (HBc) forms the anterior atlantal arch (C1A). The C2 resegmented (C2) sclerotome comes from the sixth and seventh somites. From the C2 sclerotome: the axial zone forms the C2 vertebral body (AB); the lateral zone forms the neural arch of C2 vertebra. The intervertebral boundary zone (IBZ) between the proatlas and C1 sclerotome forms the upper dental synchondrosis (US) and the IBZ between the C1 and C2 sclerotomes forms the lower dental synchondrosis (LS).

**Fig. 15**
The severance line, which results in final cellular separation of the skull from the cervical spine, runs through the original resegmentation fronts of the adjacent somites 4 and 5, corresponding to the junction between the basion and apical segment of the dens in the axial proatlas, and between the exoccipital, or future occipital condyle, and the lateral mass of C1, derived from the lateral portion of the C1 resegmented sclerotome.
Embryology and bony malformations of the craniovertebral junction


The cranial region of the axial sclerotome of the proatlas soon fuses with the other three axial occipital sclerotomes to become the basion of the basioccipital, but its most caudal portion, probably derived from the first cervical somite (somite 5), forms the anlage for the apical segment of the dens. Late in resegmentation, a boundary zone appears between this apical dental centrum and the loosely cellular prevertebra of the basioccipital, and the former soon detaches from the basioccipital and eventually becomes joined to the basal segment of the dens to complete the dental pivot (see below) (Fig. 14).

Herein lies the most unique feature of the transitional zone of the CVJ between somites 4 and 5: unlike other IBZs that form intervertebral discs, downstream activity of the proatlas’ IBZ includes a physical severance of cells from the immediately adjacent loose perichordal zone of the basioccipital. The severance line appears to go through the original resegmentation fronts of the adjacent somites 4 and 5, so that the final cellular separation occurs right through the junction between the basion and the apical segment of the dens, both derived from the axial portion of the proatlas, which in turn comes from a combination of the caudal half of somite 4 and the rostral half of somite 5 (Fig. 15). This action, no doubt mediated by special cleavage genes, not only allows the skull to become independent from the vertebral column, but also the separation of the primordium for the apical dens from the basiocciput and final installation of the axis–dens assembly.

Fig. 20  Expression domains of Hox genes lined up with the mouse embryonic vertebral column. Only the anterior expression boundary (in red) is important, and since multiple genes have the same anterior expression boundary along the prevertebral axis, each prevertebral segment has its own combination of Hox gene expression domains (Hox code). For example, the Hox code for C1 is Hoxa-1, Hoxb-1, Hoxa-2, Hoxb-2, Hoxa-3, Hoxb-3, Hoxd-3, Hoxa-5, Hoxb-5, Hoxb-6, and Hoxc-4 (designated by the vertical green bar).

HOX GENES CONTROL THE POSITIONAL IDENTITY OF PREVERTEBRAL SEGMENTS

Embryology and bony malformations of the craniovertebral junction
HOX D-3 MUTATION LEADS TO PARTIAL LOSS OF C1 EXPRESSION DOMAIN

THE C1 SCLEROTOME “BEHAVES” LIKE AN OCCIPITAL SCLEROTOME

Embryology and bony malformations of the craniovertebral junction
HOX D-3 MUTATION LEADS TO “GAIN OF FUNCTION” IN FOURTH OCCIPITAL SCLEROTOME

THE OCCIPUT “IMITATES” THE C1 SEGMENT

Embryology and bony malformations of the craniovertebral junction
Embryology and bony malformations of the craniovertebral junction

Embryology and bony malformations of the craniovertebral junction
BIOMECHANICAL HYPOTHESIS

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

↓

DORSAL DECOMPRESSSION 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 \(\rightarrow\) SLOW
POST-OP \(\rightarrow\) RAPID

HYDRODYNAMIC FACTORS