



# Is there a relationship between the extent of tonsillar ectopia and the severity of the clinical Chiari syndrome?

Dan Heffez MD; John Broderick MD; Michael Connor DO; Michael Mitchell MD; Joanna Galezowska; Jugal Ghorai PhD;

Ramin Golchini

The Wisconsin Chiari Center, The Milwaukee Neurological Institute and Columbia-St Mary's Hospital

## Introduction

Chiari 1 malformation is diagnosed when the cerebellar tonsils extend 5mm below the opisthion-basion line. Lesser degrees of tonsillar ectopia are often dismissed. However, the 5 mm threshold has never been validated as clinically meaningful.

**Objective:** to examine the correlation of the extent of tonsillar ectopia to the severity of the Chiari syndrome.

## Methods

Patients were grouped by extent of tonsillar ectopia on the mid-sagittal MRI image, (group 1: 0-<3mm, N=97, group 2: 3-5mm, N=148, and group 3: >5mm, N=183). Groups were compared with regard to demographics, neurological symptoms(51), neurological signs, extent of pain and response to HADS and sf-36 questionnaires. Observed differences were analyzed using one-way anova and Chi-square and paired analysis performed using Tukey's multiple comparison method for means, two sample Z test for proportions or Student t-test where appropriate. Logistic regression analysis related extent of tonsil position to the likelihood of given symptoms.

## Results

Mean age, female preponderance and trauma history within six months of symptom onset did not differ between the groups. Patients in group 1 were more symptomatic than those in groups 2 and 3 with regards to a number of symptoms, (p=0.04 – p=0.000). Regression analysis confirmed an inverse relationship between the extent of tonsillar ectopia and the odds of many of the symptoms. No symptom was more prevalent in Group 3 than in Groups 1 and 2. The pain score was greatest in group 1, (p=0.006). Prevalence of objective signs of myelopathy did not differ between groups with the exception of Hoffman sign which was more prevalent in group 1, (p=0.034). HADS and sf-36 scores did not differ between groups.

### Demographics

	Group 1	Group 2	Group 3
mean age	42.11	38.23	39.98
% Female	78.3	89.8	85.7
Hx of trauma (%)	70	60	54.6
Hx of trauma < 6 months (%)	23.7	22.2	19.2
mean ectopia	0.6 mm	4.2 mm	9.17 mm

### Linear Regression Analysis

Symptom	Coefficient of TE	P-value
Headache	+0.092	0.319
Occipital HA	-0.037	0.365
Retro-ocular pain	-0.092	0.000
Frontal HA	-0.886	0.000
Pressure HA	-0.085	0.007
Diplopia	-0.058	0.020
Vertigo	-0.032	0.151
Nausea	-0.102	0.000
Vomiting	-0.102	0.011
Irritable bowel	-0.052	0.039
Urinary urgency	-0.061	0.006
Urinary retention	-0.067	0.005
Facial pain	-0.097	0.000
Facial paresthesiae	-0.056	0.017
Dental pain	-0.053	0.024
Leg pain	-0.073	0.001
Leg numbness	-0.049	0.028
Leg paresthesiae	-0.061	0.007
Worsened by valsalva	+0.044	0.130
Worsened by neck extension	-0.049	0.045
Leg weakness	-0.055	0.018
Myelopathy	+0.005	0.795
Fibromyalgia	-0.114	0.002

A negative value of the coefficient implies an inverse relationship between tonsillar ectopia and the likelihood of a symptom. p-value <0.05 implies statistical significance.

### Learning Objectives

1) recognize symptomatic tonsillar ectopia, 2) optimize imaging of the foramen magnum, 3) understand Chiari from a neurological rather than anatomical perspective.

fig 1



Figure 1: Thin section T1 weighted post Gadolinium MRI image through the plane of the foramen magnum which shows compression and distortion of the brain stem, (BS), by the cerebellar tonsils, (T), and the vertebral arteries, (anterior and lateral to the brain stem). The PICA arteries are positioned between the tonsil and the brain stem. Cerebellar tonsils did not project below the opisthion-basion line on mid-sagittal MRI image.



### Neurological Signs

Sign	Group 1	Group 2	Group 3	p-value
	%	%	%	Chi-square
	N=97	N=148	N=183	
Romberg	23.9	14.8	15.3	0.13
tandem	18	8.9	12	0.112
position sense	11.4	7.5	15.6	0.079
dysmetria	3.0	6.0	5.4	0.565
disdiadoko	4.1	4.0	1.6	0.346
gag absent	16.8	23.5	25.7	0.266
<b>Hoffmann</b>	<b>29.5</b>	<b>16.4</b>	<b>18</b>	<b>0.034</b>
clonus	26	21.2	20.3	0.532
recruitment	8.2	7.4	10.9	0.732
inversion	37.1	37.1	34.4	0.845
DTR>=3 <sup>1</sup>	32	26.3	26.5	0.554
DTR= 2+ <sup>2</sup>	46	37.5	40.4	0.404

(1= at least one DTR was rated 3/4 or greater, 2= at least one DTR was rated 2+/<sup>4</sup> i.e. more brisk than normal but perhaps not quite 3/4. DTR's tested: flexor digitorum, brachioradialis, biceps, triceps, patellar, Achilles tendon).

### Conclusions

The extent of tonsillar ectopia does not correlate directly with the severity of the clinical Chiari syndrome and should not remain the principal diagnostic criterion on which to base treatment recommendations. A neurological paradigm based on cervical myelopathy should be considered.

### References:

Milhorat TH, Chou MW, Trinidad EM, et al: Chiari 1 malformation redefined: clinical and radiographic findings for 364 symptomatic patients. Neurosurgery, 1999, 44:1005-1017.

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fig 1

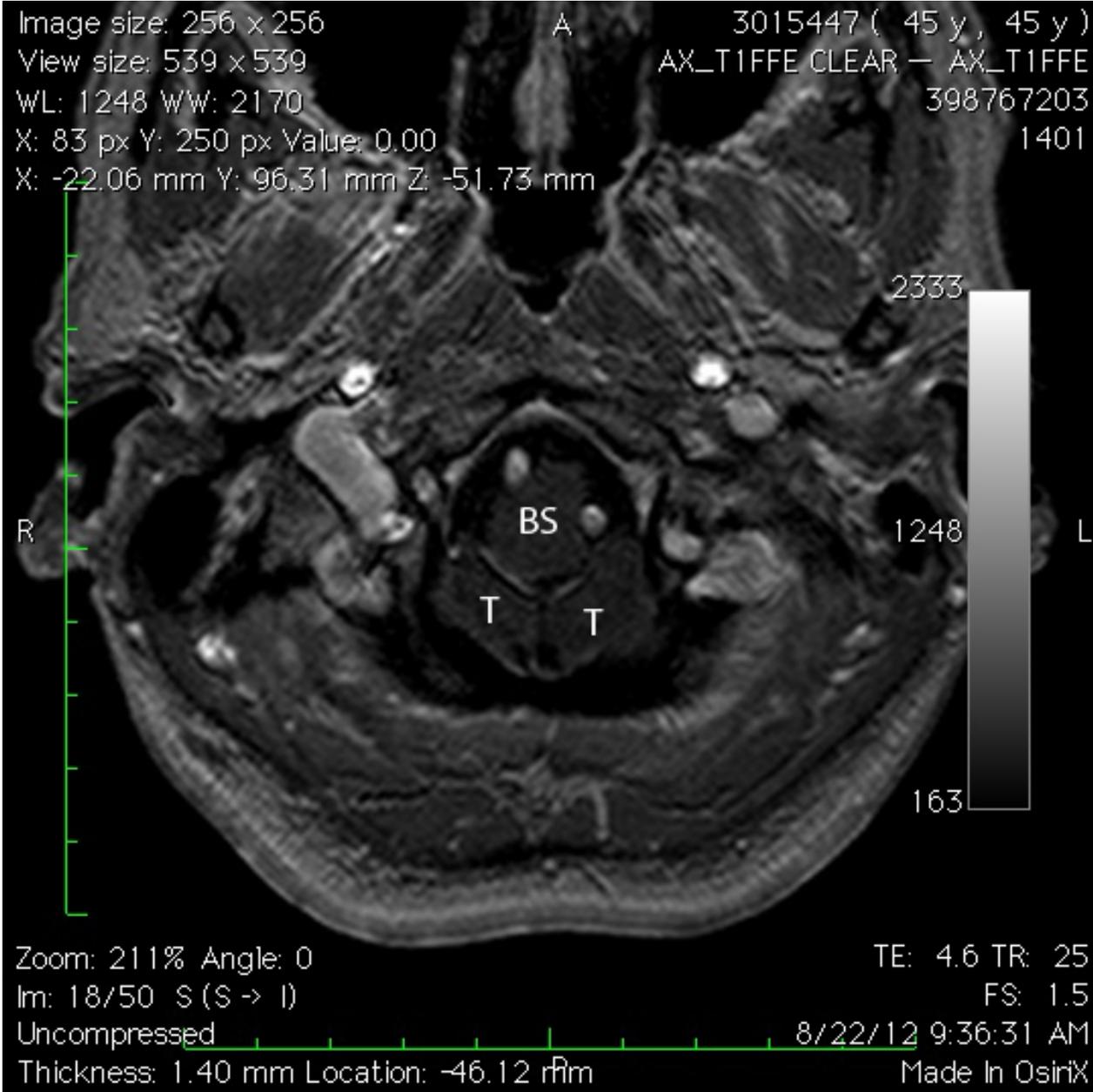


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Image size: 256 x 256

S

3015447 ( 45 y , 45 y )

View size: 539 x 539

SAG\_T1 SENSE — SAG\_T1

WL: 856 WW: 1488

398767203

X: 84 px Y: 248 px Value: 763.58

301

X: 12.26 mm Y: -46.38 mm Z: -108.84 mm



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