

Visual Consequences of Chiari Malformation

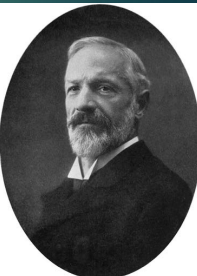
2018 VICTORIA CONFERENCE
JAMES KUNDART OD MED FAAO FCOVD-A
PACIFIC UNIVERSITY COLLEGE OF OPTOMETRY
FINANCIAL DISCLOSURE: NOTHING TO DISCLOSE

<https://www.conquerchiari.org/index.html>
<http://www.ajnr.org/content/34/5/919.long>

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Learning Objectives

- ▶ Which cause of Chiari malformation is most common? What are the presenting symptoms?
- ▶ Which cause of Chiari malformation is not evident with imagining, but may have ocular signs and symptoms?
- ▶ What are the differential diagnoses for Chiari syndrome?
- ▶ What are the surgical and non-surgical treatments for Chiari malformation?



https://en.wikipedia.org/wiki/Hans_Chari

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Evidence of Chiari in Europe Dates to 16th-17th Centuries



<https://www.ncbi.nlm.nih.gov/pubmed/25192779>

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Definition of Chiari Malformation



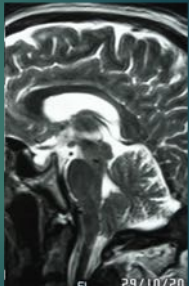
- ▶ Chiari malformation is traditionally defined as the **cerebellar tonsils being located 3mm-5mm or more below the foramen magnum** as measured on an MRI

<https://www.conquerchiari.org/documents/presentations/OVERVIEW%20Presentation.pdf>

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Problems with the Traditional Chiari Definition

- ▶ Some people have large herniations with no symptoms
- ▶ Others have only small herniations, but are severely symptomatic



<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>

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Alternative Definitions: Twining and Chamberlain Lines



<http://stroke.ahajournals.org/content/31/9/2062>

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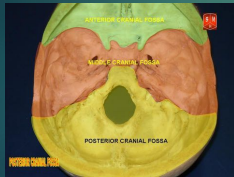
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Causes of Chiari: Small Posterior Fossa



- ▶ This is the default "anatomical determinism" theory
- ▶ When this occurs, **transient but recurrent hydrocephalus can result**
- ▶ In these cases, surgery may be the best option

https://en.wikipedia.org/wiki/Posterior_cranial_fossa

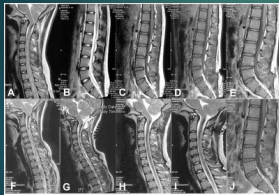


Causes of Chiari: Connective Tissue Disorders

- ▶ "There is one published large study that has looked at the association between type 1 Chiari and EDS
- ▶ This was undertaken by Milhorat et. al. in 2007 at The Chiari Institute in New York
- ▶ They looked at 2813 patients with a known diagnosis of Chiari malformation, Type I
- ▶ **They found that 357 (12-13%) had features of EDS"**



Causes of Chiari: Tethered Cord (?)



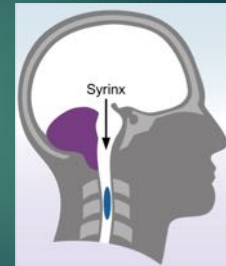
- ▶ "Tethered cord is a relatively new entity, medically speaking, and as such there is still quite a bit of controversy surrounding it
- ▶ **There is even occult tethered cord, since the tethering is not always apparent on MRI**
- ▶ A second area of controversy involves the relationship, if any, between tethered cord and Chiari and/or syringomyelia", or syringes

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5738018/>



Syrinxes and Syringomyelia

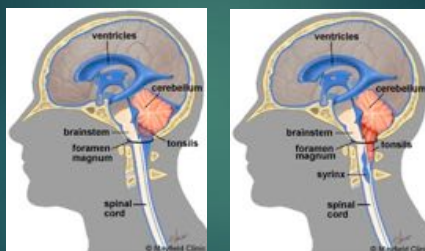
- ▶ A syrinx is a fluid-filled cyst in the spinal cord
- ▶ This causes spinal cord edema, compromising neuronal function
- ▶ Permanent nerve damage can result, but **they are easily missed with MRI of the head only**



<https://www.conquerchiari.org/documents/presentations/OVERVIEW%20Presentation.pdf>



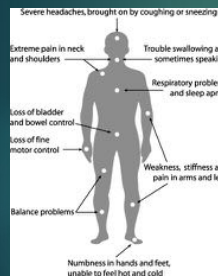
Normal vs. Chiari Cerebellum & Syrinx



http://www.mayfieldclinic.com/PE-Chiari.htm#_VXiR486VzPA



Spinal Cord Syrinxes and Chiari Symptoms

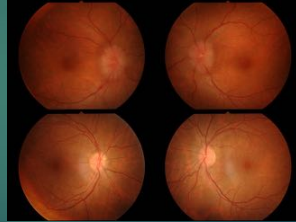


<http://www.mayfieldclinic.com/PE-Chiari.htm>



How Common is Chiari? Mimicking by IIH

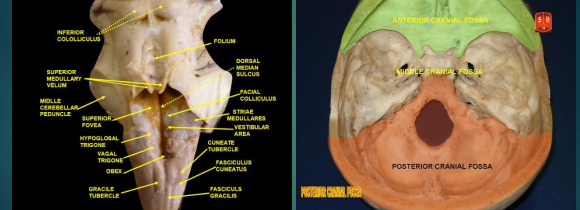
- ▶ The US Association of Neurological neurological surgeons performs 10K Chiari surgeries per year
- ▶ **Chiari may be as common as 1:1000 patients in the US.** Also...
- ▶ "Cerebellar tonsil position in patients with IIH was significantly lower than that in age-matched controls. **oftentimes peg-like, mimicking Chiari**"



<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>



Chiari Commonalities: The Low Obex and Small Posterior Fossa

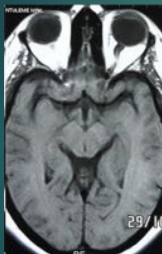


<https://en.wikipedia.org/wiki/Obex>

https://en.wikipedia.org/wiki/Posterior_cranial_fossa



Gender and Ethnic Trends in Chiari Malformation



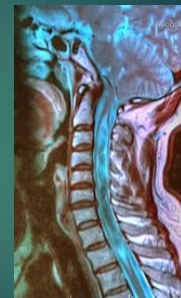
- ▶ Chiari is often diagnosed in young adults
- ▶ Female patients get Chiari more often than male patients
- ▶ It affects all ethnicities and body mass indices
- ▶ **Retrobulbar optic nerve tortuosity is common and often asymptomatic, as shown here**

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>



Three Types of Chiari

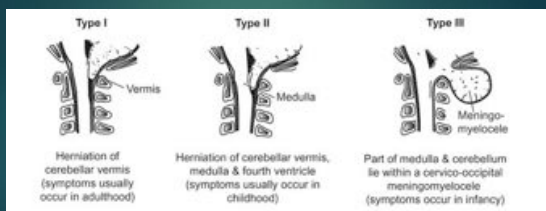
1. Chiari Malformation, Type I
2. Arnold-Chiari Malformation
3. Chiari Malformation, Type III



https://en.wikipedia.org/wiki/Chiari_malformation



Three Types of Chiari

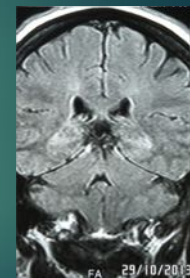


Wong, page 170



Classical Chiari Type I

- ▶ Mid-sagittal section MRI is the best way to diagnose the structural component of Chiari malformation, but depends on where the foramen magnum is placed
- ▶ Sometimes, coronal MRI is the best way to see **cerebellar tonsillar herniation (right)**

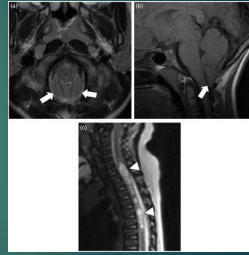


<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>



Congenital Chiari Type I

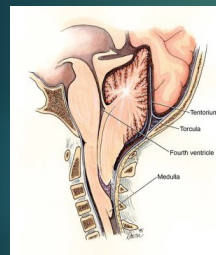
- ▶ (a) Axial T2W MRI shows crowding at the level of the foramen magnum with herniation of the cerebellar tonsils (arrow)
- ▶ (b) Sagittal T1W MRI shows caudal herniation of the **"peg shaped" cerebellar tonsils** (arrow)
- ▶ (c) Sagittal T2W MRI of a different patient shows **associated syringomyelia** (arrowheads)



<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4757284/>



Arnold-Chiari (Type II)

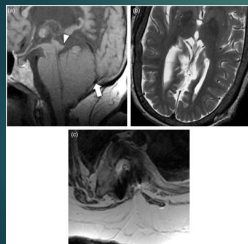


- ▶ Much less common than Type I, this birth defect leads to fourth ventricle hydrocephalus
- ▶ Expect childhood-onset with more severe symptoms
- ▶ **Surgical intervention is highly likely**
- ▶ Type II is the only time this malformation is properly called Arnold-Chiari

<http://www.fluidsbarrrierscns.com/content/5/1/2/figure/F2?highres=y>



Congenital Arnold-Chiari, Type II



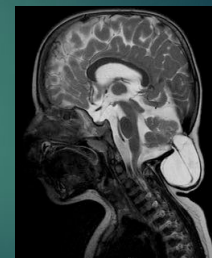
- ▶ Chiari II malformation. (a) Sagittal T1W MRI shows a **small posterior fossa** with a low torcular insertion (arrow) and **tectal beaking** (arrowhead)
- ▶ (b) Axial T2W MRI fails to demonstrate the cerebellum between the occipital lobes
- ▶ (c) T2W MRI shows spinal dysraphism*, or a neural tube defect

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4757284/>



Chiari, Type III

- ▶ This terrible birth defect is characterized by cerebellar herniation outside the skull cavity, called an **encephalocele**
- ▶ This happens when the neural tube does not close during the first trimester of gestation
- ▶ **Various teratogens like arsenic can cause this usually fatal condition**



<http://radiopaedia.org/articles/chiari-ii-malformation>



Chiari Type III is Congenital



- ▶ These patients are often born with paraplegia due to the CNS myelomeningocele
- ▶ This non-ambulatory patient may also be nonverbal as **the cerebellum controls coordination of the tongue as well as the eyes and limbs**

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4757284/>



Visual Symptoms of Chiari

Symptom	%
Headache	98
Dizziness	84
Difficulty sleeping	72
Weakness in arms/hands	69
Neck pain	67
Numbness/tingling in arm, hands	62
Fatigue	59
Nausea	58
Shortness of breath	57
Blurred Vision	57
Tinnitus	56
Difficulty swallowing	54
Leg weakness	52

- ▶ Blurred vision
- ▶ Diplopia and **strabismus**
- ▶ Nystagmus
- ▶ Photophobia
- ▶ **Visual field defects**

<https://www.conquerchiari.org/documents/presentations/SYMPTOMS%20Presentation.pdf>



Ocular Motility in Chiari

Table 13-1 Disturbances in Eye Movements in the Chiari Syndrome

Downbeat nystagmus (occasionally with a torsional component), worse on lateral gaze,^{154,207,203,704,802,804,846} and worse with convergence
 Divergence nystagmus¹⁵⁴
 Convergence nystagmus¹⁵⁷
 Horizontal nystagmus (unidirectional, present with eyes in central position (Figure 11-5))
 Periodic alternating nystagmus^{15,246}
 Gaze-evoked nystagmus¹⁵
 Rebound nystagmus including torsional rebound¹⁷⁷
 Seesaw nystagmus¹⁵⁶
 Impaired pursuit, especially downward (and VOR cancellation)^{155,846,848}
 Impaired OKN with slow build-up of eye velocity in response to a constant-velocity stimulus¹⁵⁴
 Esotropia,^{15,153,154,155,157} abducens paralysis¹⁷⁴
 Convergence spasm¹⁷³
 Divergence paralysis¹⁵⁷
 Skew deviation accentuated or alternating on lateral gaze,⁸⁴² with dissociated vertical nystagmus¹⁵⁷
 Saccades occasionally dysmetric, though usually accurate⁸⁴²
 Internuclear ophthalmoplegia^{15,155,824}
 Increased VOR gain¹⁵⁵
 Shortened VOR time constant and impaired tilt-suppression of post-rotatory nystagmus¹⁹¹
 Positional nystagmus¹⁵¹

Leigh & Zee, 5th edition



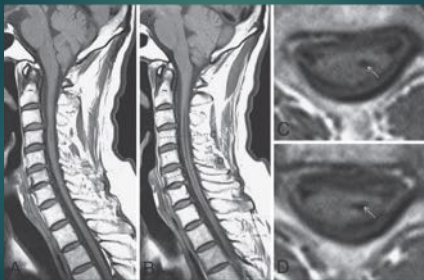
Chiari Malformation, Diplopia, and Strabismus

- Mild cases of Chiari may result in intermittent diplopia
- Often, this presents as **esotropia at far of the divergence insufficiency type**
- When this occurs, suspect CN VI palsy



<https://timroot.com/ophthobook/>

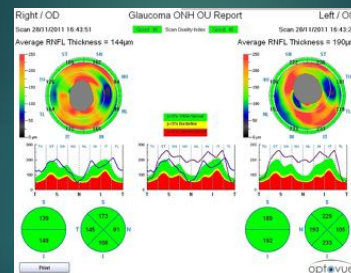
Why Pursuits and Saccades Are Affected in Chiari



<https://www.ncbi.nlm.nih.gov/pubmed/22864131>



OCT of 52 YOF with IIH (?) and Chiari Malformation

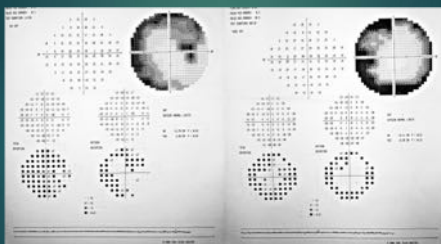


Notice the absent optic cup and nerve fiber layer thickening

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>



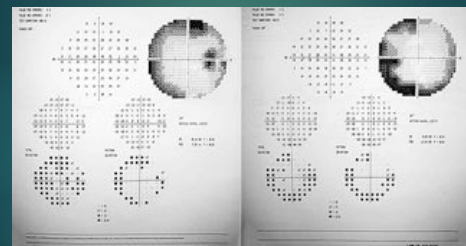
Baseline Visual Fields in Chari, Type I: Expected?



<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>



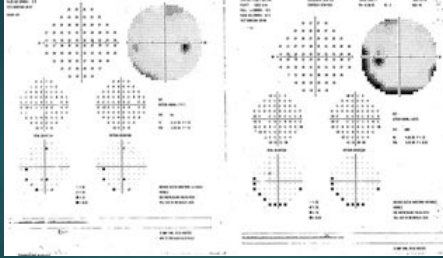
1-Month Post Tx Visual Fields in Chari, Type I



<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>



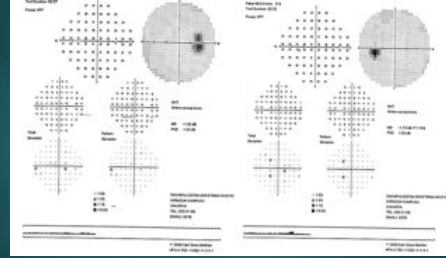
3-Months Post Tx Visual Fields in Chari, Type I



<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>



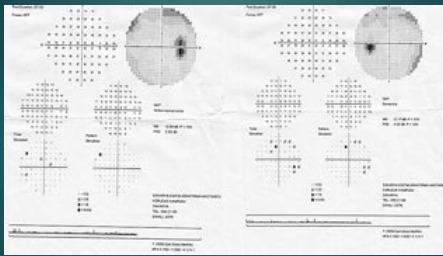
6-Months Post Tx Visual Fields: Notice Blind Spot OS



<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>



12-Months Post Tx Visual Fields in Chari, Type I



<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4054396/>



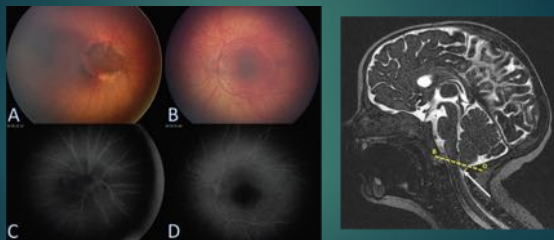
Differential Diagnoses That Mimic Chiari

Set of Symptoms	Conditions with similar symptoms
Headache Dizziness Weakness in arms/hands Numbness in arms/hands Leg Weakness Fatigue Tinnitus	Multiple Sclerosis Fibromyalgia Chronic Fatigue Lupus Migraines Carpal Tunnel ALS (Lou Gehrig's)

<https://www.conquerchiari.org/documents/presentations/DIAGNOSIS%20Presentation.pdf>



Chiari Obscured by Morning Glory Disc



<https://www.ncbi.nlm.nih.gov/pubmed/24802674>



Treatment: Syringes & Surgery

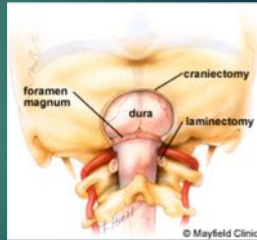
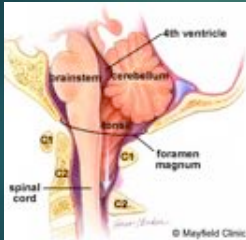
A 2004 survey of neurosurgeons (Schjman) found a wide range of thinking on when to operate for Chiari.

Case #	Symptoms/Diagnosis	% Would Operate
1	7 yr old with no symptoms 12 mm tonsils- no syrinx	8
1A	w/ 2mm wide syrinx	28
1B	w/ 8mm wide syrinx	75
2	9 yr old with headaches 10 mm tonsils- no syrinx	46
2A	w/ 2mm wide syrinx	64
2B	w/ 8mm wide syrinx	90
3	11 yr old w/progressive scoliosis 12 mm tonsils- no syrinx	58
3A	w/ small syrinx	85
3B	w/ 6mm wide syrinx	97

<https://www.ncbi.nlm.nih.gov/pubmed/14966661>



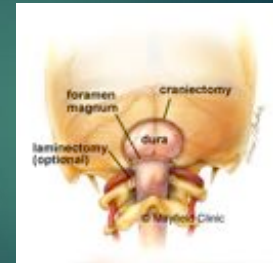
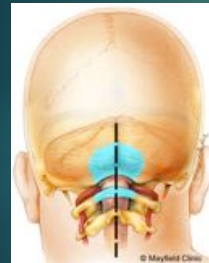
Surgical Treatments: Posterior Fossa Decompression



<http://www.mayfieldclinic.com/PE-Chiari.htm>



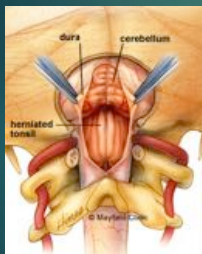
Craniectomy for Chiari



http://www.mayfieldchiaricenter.com/chiari_surgery.php



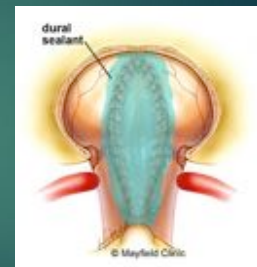
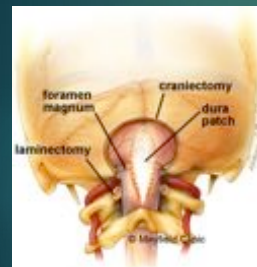
Opening the Dura in Chiari



http://www.mayfieldchiaricenter.com/chiari_surgery.php



Dura Patch for Chiari



<http://www.mayfieldclinic.com/PE-Chiari.htm#VXiR486VzPA>

http://www.mayfieldchiaricenter.com/chiari_surgery.php



Surgical Time Course for Posterior Fossa Decompression



Surgery: ~ 4 hrs.



ICU: One night

<https://www.conquerchiari.org/documents/presentations/TREATMENT%20Presentation%20.pdf>



Surgical Time Course for Posterior Fossa Decompression



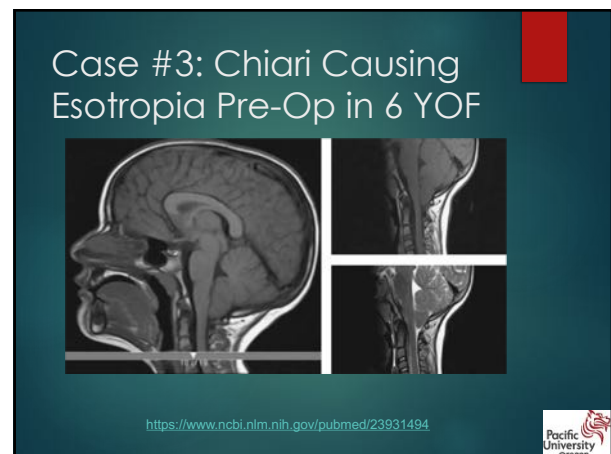
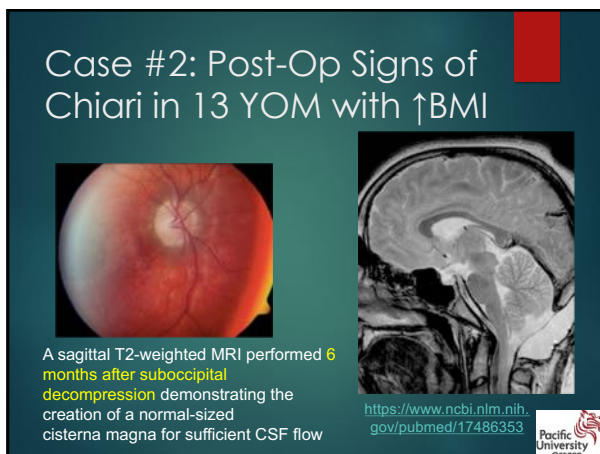
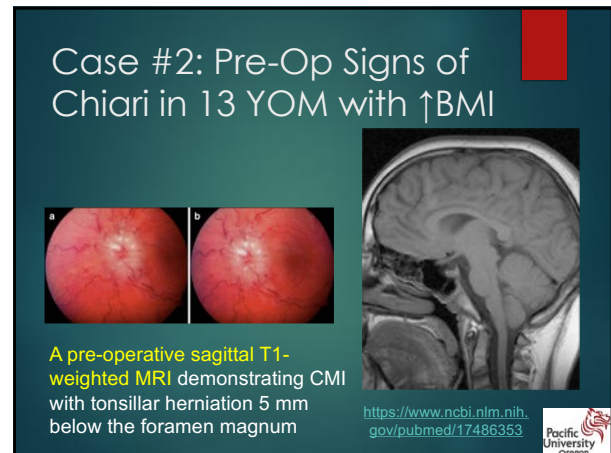
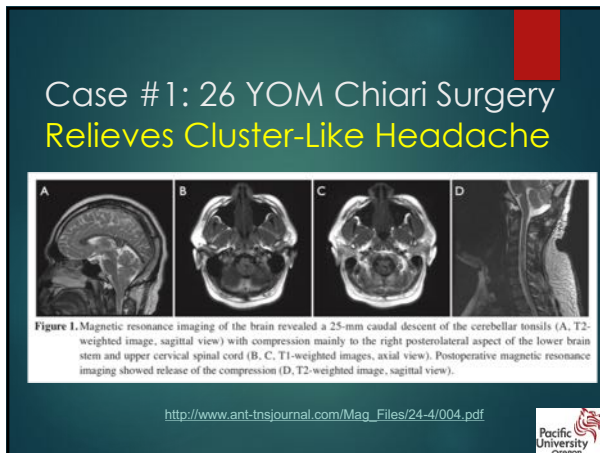
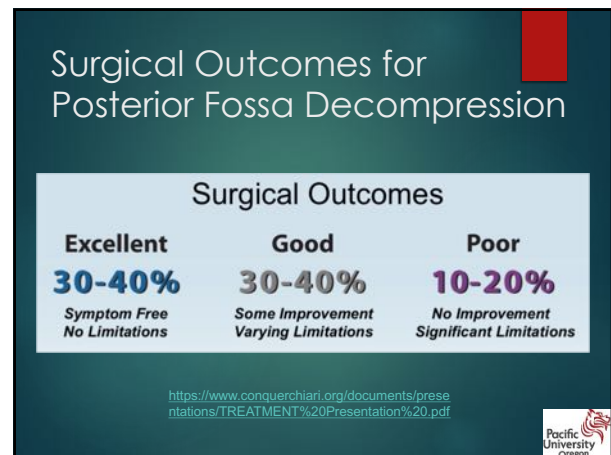
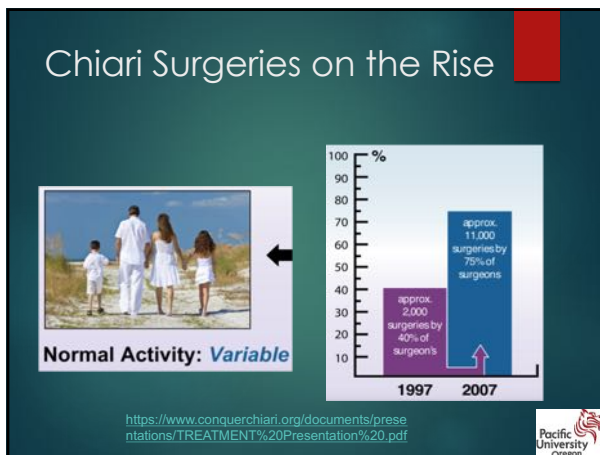
Hospital: 3-4 days



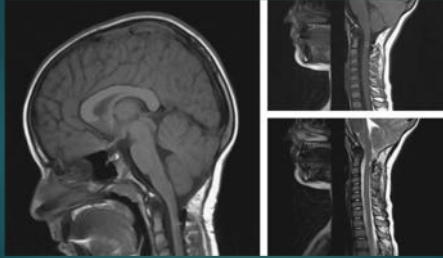
Rest at home: 3 wks to

<https://www.conquerchiari.org/documents/presentations/TREATMENT%20Presentation%20.pdf>





Case #3: Chiari Causing Esotropia Post-Op in 6 YOF



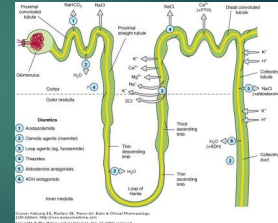
<https://www.ncbi.nlm.nih.gov/pubmed/23931494>



Non-Surgical Chiari Tx: Acetazolamide and Lasix



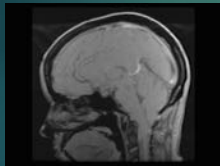
<https://www.onlineclinic.co.uk/images/product/acetazolamide-blister-pack-l.jpg>



<https://i.pinimg.com/originals/e7/80/67/e780675385d88cd1fe507bc885c5072e.jpg>



Future Directions in Chiari



GIF Animation of CSF flow in Chiari

<http://www.mayfieldclinic.com/PE-Chiari.htm>

- ▶ Researchers are looking for a new way to measure Chiari severity
- ▶ Focus areas include advanced MRI and engineering techniques to quantify cerebrospinal fluid (CSF) flow, crowding and compliance



Questions? Thank you!



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Reading and References

- ▶ For more information, see the excellent *Eye Movement Disorders* by Agnes Wong (2007, see chapter 10 on the cerebellum)

