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Chiari Malformation

Patient education guide · Brain & Spine Injury and Other · Madison Neurology LLC

What is it?

A Chiari malformation (type I in adults) is a structural condition in which the lowest part of the cerebellum (the tonsils) extends down through the skull's base opening, crowding the space where the brain meets the spinal cord. Many Chiari malformations are incidental findings causing no symptoms; symptomatic ones cause pressure headaches, neck pain, and a distinctive set of neurological complaints.

How does it happen?

The posterior fossa (the skull's back compartment) is too small for its contents — a developmental anatomical mismatch. Symptoms arise when the descent blocks normal cerebrospinal fluid flow at the skull base. Most cases are present from birth but become symptomatic (or discovered) in adulthood; trauma or strain sometimes unmasks symptoms.

Common symptoms

- Pressure headache at the back of the head, worsened by coughing, straining, or laughing ('Chiari cough headache') — the signature symptom
- Neck pain
- Balance problems, dizziness, and unsteadiness
- Numbness, tingling, or weakness in the hands; fine-motor clumsiness
- Swallowing difficulty, hoarseness, or sleep apnea in some; syringomyelia (a fluid cavity in the cord) causing cape-like sensory loss

How is it diagnosed?

MRI shows the tonsillar descent (5 mm or more below the foramen magnum, though the number alone doesn't determine symptoms) and checks for syringomyelia. CSF flow studies help in borderline cases.

The crucial clinical question is whether symptoms match the anatomy — many 5-10 mm descensions are incidental and need nothing.

How is it treated?

Asymptomatic Chiari needs no treatment — periodic monitoring only. Symptomatic Chiari with matching anatomy is treated with posterior fossa decompression surgery: removing a small section of skull (± the C1 arch and opening the covering) to restore CSF flow — highly effective for cough headache (relief in 80%+), and often improving balance and preventing syrinx progression. Headaches without the typical features are first managed medically.

What does the future hold?

Incidental Chiari never causes problems for most people. For symptomatic cases, decompression reliably relieves the pressure headache and stabilizes syringes; nerve symptoms improve variably. A minority need reoperation. Choosing an experienced Chiari surgeon meaningfully affects outcomes.

Questions patients commonly ask

Q: My MRI showed Chiari — do I need surgery?

A: Not if it's asymptomatic — many are incidental findings; surgery is for symptoms matching the anatomy, especially the classic cough headache.

Q: What is a syrinx?

A: A fluid-filled cavity within the spinal cord that Chiari flow obstruction can create; decompression usually stops its growth and often shrinks it.

Q: Why do I get a headache when I cough?

A: Coughing spikes CSF pressure, and the crowded skull base can't buffer it — that specific pattern is Chiari's calling card.

National support groups & resources

- **Bobby Jones Chiari & Syringomyelia Foundation** — bobbyjonescsf.org
- **Hydrocephalus Association** — hydroassoc.org
- **National Organization for Rare Disorders** — rarediseases.org — 1-800-999-6673

(Organization contact details were current at the time of compilation; please verify online.)

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