



## Rx FOR SUCCESS

## Arnold-Chiari Malformation

Arnold-Chiari malformation [Chiari malformation (CM)] is a congenital defect at the base of the brain. The cerebellum (part of the brain which controls balance) and the brain stem (which controls respiratory and heart functions) are pushed down into the spinal canal. This herniation of the brain sometimes blocks the flow of fluid (cerebrospinal fluid (CSF)) leading to increase CSF pressure. This elevated pressure may cause enlarged brain ventricles (hydrocephaly) or syringomyelia (cavity or “syrinx” the cervical spinal cord).

CM is often asymptomatic or it can present with headache and/or cerebellar problems, such as vertigo, imbalance, and in-coordination. If syringomyelia is present, signs and symptoms depend on the severity of the spinal cord compression by a growing syrinx.

There are four types. Type I CM is most common and least severe. It can be an incidental finding on a brain scan done for other reasons (such as migraine headache). Brain herniation is mild. Usually, there are no symptoms and no clinical findings. However, hydrocephalus and/or syringomyelia can occur with Type I. Type II CM causes more severe brain herniation, resulting in significant neurologic problems. Hydrocephalus and myelomeningocele (protuberance of membrane and spinal cord through a hole in the vertebral bone) are usually present (with or without syringomyelia). Type III and Type IV CM are most severe and rare.

Due to considerable risk, preventive surgery is not indicated. Surgery is only done for symptomatic lesions. The goal of surgery is to decompress nerve tissue and restore the free flow of CSF. Surgical procedures vary but may include removal of the back of the skull and back of the first few neck vertebrae. This creates more space for the cerebellum and brainstem. A tube is sometimes placed in a syrinx to shunt fluid to the peritoneal or chest cavity. Surgery may or may not relieve symptoms and has not been shown to improve mortality. Surgery is unlikely to reverse severe neurological defects (e.g. paralysis), and such applicants are uninsurable.

The most likely scenario that will be encountered in underwriting is the incidental, asymptomatic (no more than mild headache) Type 1 CM without a syrinx or hydrocephaly. No treatment or follow-up is deemed necessary by the AP. The applicant has no bladder problems due to nerve damage, mobility problems, or cognitive impairment.

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Possible ratings are given in the table below.

CHIARI MALFORMATION	
Incidental <b>Type I</b> OR <b>Type I</b> with no syringomyelia and/or hydrocephaly and no posterior fossa decompression surgery	0
<b>Type I</b> with syringomyelia and/or hydrocephaly (with or without shunt), but no posterior fossa decompression surgery <ul style="list-style-type: none"> <li>• Best case hydrocephalus</li> <li>• Hydrocephalus with shunt</li> <li>• Best case syringomyelia</li> <li>• Others</li> </ul>	0 Table C Table E Decline
After posterior fossa decompression surgery: <ul style="list-style-type: none"> <li>• <math>\geq 1</math> year after surgery.</li> <li>• All signs and symptoms have resolved (other than mild headache).</li> </ul>	
Type I	Rate as above
<b>Type II</b>	Table F
Others	Decline

For example, client had a CT of the head after trauma. Chiari I malformation was noted. Client is asymptomatic. He would not be rated.

*To get an idea of how a client with a history of Arnold-Chiari Malformation would be viewed in the underwriting process, use the Ask "Rx"pert Underwriter on the next page for an informal quote.*

**Ask “Rx”pert Underwriter (Ask Our Expert)**

After reading the *Rx for Success* on Arnold-Chiari Malformation please feel free to use the Ask “Rx”pert Underwriter for an informal quote.

Producer \_\_\_\_\_ Phone \_\_\_\_\_ Fax \_\_\_\_\_  
 Client \_\_\_\_\_ Age/DOB \_\_\_\_\_ Sex \_\_\_\_\_

If your client has had a diagnosis of Arnold Chiari malformation, please answer the following questions and send the most recent CT or MRI reports.

**1. When was the diagnosis made? (Date)**

\_\_\_\_\_

**2. When was the surgery completed? (Date)**

\_\_\_\_\_

**3. Please note type of surgery.**

Ventricular shunt       Decompression       Other

**4. Please note current neurologic status and/or symptoms.**

Headache	<input type="checkbox"/> Yes	<input type="checkbox"/> No	Breathing problems	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Paralysis	<input type="checkbox"/> Yes	<input type="checkbox"/> No	Dizziness/fainting	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Muscle weakness	<input type="checkbox"/> Yes	<input type="checkbox"/> No			

**5. What are the current symptoms if any?**

\_\_\_\_\_

**6. Is there a history of any other neurological conditions?**

Yes. Please give details. \_\_\_\_\_  
 No

**7. Is your client on any medications?**

Yes. Please give details. \_\_\_\_\_  
 No

**8. Has your client smoked cigarettes in the last 12 months?**

Yes     No

**9. Does your client have any other major health problems (e.g., cancer, etc.)?**

Yes. Please give details. \_\_\_\_\_  
 No