

Chiari Malformation Education Sheet



I **HAVE** Chiari



I **DON'T** have Chiari

Definition: *A serious neurological disorder where the bottom part of the brain, the cerebellum, descends out of the skull and crowds the spinal cord, putting pressure on both the brain and spine, and disrupting the normal flow of cerebrospinal fluid (CSF); also known as Arnold-Chiari, ACM, ACM I, ACM II, CM*

Types: *Type I is shown above; Type II also involves the brainstem and is associated with Spina Bifida*

Number of People Affected: *About 1 in 1,000 people*

Cause: *Most people are believed to be born with it; there is evidence that some cases have a genetic basis*

Symptoms:

- Many different symptoms; affects each person differently
- Most common is severe headaches; usually in the back of the head, and made worse by straining, coughing, sneezing, or bending over
- Other symptoms include balance problems, visual disturbances, and trouble swallowing
- Can develop at any age, but commonly become noticeable in childhood or a person's early 30's
- Size of the malformation is NOT related to severity of symptoms
- In some, a fluid-filled cyst can develop in the spinal cord, this is known as *syringomyelia*

Diagnosis:

- No single, objective test to say whether someone has symptomatic Chiari
- MRI (Magnetic Resonance Imaging) can show whether the cerebellum is crowding the spine
- cine MRI can show whether there is adequate CSF flow
- Diagnosis is made through a combination of MRI's, patient reported symptoms, a neurological exam, and possibly other tests
- Because of the wide range of symptoms, and the fact that many doctors are not aware of the condition, people often go years without a proper diagnosis

Treatment/Surgery:

- If symptoms are mild, they can be treated individually or just tolerated
- If symptoms are severe, getting worse, the nervous system is compromised, or if there is syringomyelia, surgery is usually recommended
- Goal of surgery, performed by a neurosurgeon, is to make more room around the cerebellum by removing part of the skull and spine
- Surgery usually takes 3-4 hours, and involves 3-4 days in the hospital; recovery can take several weeks to several months
- Doctors may not always agree on whether surgery is necessary

What To Expect:

- Precise data is not available, but up to 50% of people become symptom free after surgery; another 20%-30% improve significantly, and about 20% do not improve or get worse
- After what can be a long recovery, many people lead a normal, or near normal, life
- For those with residual symptoms, some lifestyle modifications may be required and Chiari will be a chronic condition
- People for whom initial surgery fails often undergo additional surgery, sometimes several
- Unfortunately, symptoms can come back, sometimes even years after surgery

CONQUER  CHIARI

For More Information Visit: www.conquerchiari.org