**Definition**
Chiari (kee-AR-ee) I malformation (CM) is a congenital or acquired (rare) abnormality in which the back compartment of the skull is too small, resulting in crowding of nerve tissues. The lower part of the cerebellum, called the cerebellar tonsils, protrudes down through the opening at the bottom of the skull, preventing cerebrospinal fluid (CSF) from flowing freely into the spinal canal and may put pressure on nerve structures including the brainstem.

**Causes**
Congenital or primary CM (the majority of CM cases): A smaller than normal posterior compartment of the skull (posterior fossa) present from birth. Some children who are born with CM may not show symptoms until adolescence or adulthood, if at all.

Acquired or secondary CM (rare): Caused later in life by increased volume of the brain (mass, enlarged fluid spaces) or excessive drainage of spinal fluid from the spine.

Primary CM is much more common than secondary CM. CM II only occurs in patients with spina bifida.

**Treatment**
Currently, the only effective treatment is surgery.

Aim of surgery: Return cerebrospinal fluid circulation as close to normal as possible, thus relieving symptoms; correction of the impaction or compression of the brainstem by the descending cerebellar tonsils.

**Important note:** The decision to proceed with surgery should be carefully based on symptoms and neurological findings. If there is any doubt about the significance of the imaging results, tests should be repeated and surgery should be deferred.

**Common Signs & Symptoms**
- Headache
- Neck pain
- Choking
- Ringing in ears
- Dizziness
- Imbalance
- Weakness in limbs

**Diagnostic Tests**
MRI of the brain and spine will indicate whether or not a patient has CM or any other abnormality.

Three components for appropriate diagnosis and treatment of CM:
- patient’s history of specific characteristic symptoms
- examination that shows signs consistent with CM
- head and spine MRI demonstrating characteristic anatomy of CM

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