DEFINITION
Chiari (kee-AR-ee) I malformation (CM) is a congenital or acquired (rare) malformation in which the back compartment of the skull is too small, resulting in crowding of neurological tissues. The lower part of the cerebellum, called the cerebellar tonsils, hangs down through the opening at the bottom of the skull, preventing cerebrospinal fluid (CSF) from flowing freely into the spinal canal.

CAUSES
Congenital or primary CM (the majority of CM cases): A birth defect, possibly from small skull cavity during fetal development *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 a mass in the brain, 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

DIAGNOSTIC TESTS
MRI of the brain and spine will indicate whether or not a patient has CM or any other abnormality

Special test called a cine-MRI (or cine MRI CSF flow study) can be performed to help improve the certainty of a diagnosis

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

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 to a later date.

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.

OUR MISSION: To advance knowledge through research and to educate the medical, allied sciences, and lay community about Chiari malformation, syringomyelia and related conditions.