

The Chiari Malformation

Nobuhito Morota, M.D.

Department of Neurosurgery
Kitasato University School of Medicine, Sagamihara, Japan

Abstract

History of the Chiari malformation (CM) goes back to 17th century when Tulp, a Dutch physician first described hindbrain herniation associated with myelomeningocele (MMC) in 1641. Solo hindbrain herniation without MMC was first reported by Langhans in 1881, followed by Chiari's classification into 3 types in 1890.

Once regarded as an uncommon disease, the CM has currently grown as one of most commonly recognized craniovertebral junction anomalies in the field of neurosurgery due to recent advancement of MRI. Published papers about the CM reached nearly 300 last year, a long leap from the status only a few papers had been published each year before 1980. However, controversies still remain surrounding CM type 1 about its pathophysiology, surgical indications and the best choice of surgical procedures. Those problems are even deeper when it comes to infants and toddler since the number of patient is limited.

Considering these backgrounds, it is intended to enhance comprehensive understanding about the CM, especially its epidemiology, natural history, diagnosis, and pathophysiology of CM type 1. In addition, based on my personal experience, detailed description of standard and modified surgical procedures together with their indication, and management of associated syringomyelia would be presented.

Although the same-named CM, CM type 2 is caused by pressure gradient between cranial and spinal spaces due to CSF leakage from the MMC in uterus. Its surgical indication and the selection of surgical procedure are another issue for controversy. The authors' policy of managing symptomatic CM type 2 and surgical procedure for choice, which is based on its pathophysiology and MRI findings, and called "upper cervical laminoplastic decompression" is fully described in the lecture.