

# Acquired Chiari Malformation

Acquired [Chiari Malformation](#) was first described by Hoffman and Tucker, in [1976](#), as a late complication of [lumboperitoneal shunt](#) <sup>1)</sup>.

Unlike the “classic” [Chiari Malformation type 1](#), Acquired Chiari Malformation (ACM) is defined by the herniation of [cerebellar tonsils](#) subsequent normal [hindbrain](#) in prior brain imaging, and it is not related to [craniovertebral junction malformations](#).

[Chiari malformation](#) could be also “iatrogenic” and then called “acquired” [Chiari I](#) malformation. <sup>2) 3)</sup> In fact, the term acquired [Chiari malformation](#) (ACM) was used by Payner et al. in the presence of the herniation of [cerebellar tonsils](#) subsequent to the documentation of a normal [hindbrain](#) in prior brain imaging <sup>4)</sup>. It was mainly described after [cerebrospinal fluid shunting](#), first after [lumboperitoneal shunting](#), and lately recognized after [supratentorial shunting](#) <sup>5) 6) 7) 8)</sup>.

## Epidemiology

Chumas et al. reviewed a series of 143 patients to determine the incidence of ACM and found that the incidence of hindbrain herniation may be as high as 70% in asymptomatic patients with LP shunts. Five of these patients became symptomatic and required Chiari decompression. During their follow-up period, which is about 5.7 years, there was one shunt-related death due to unsuspected tonsillar herniation <sup>9)</sup>.

## Pathogenesis

According to Johnston et al., the pathogenesis of [syrinx](#) formation may be explained by the [hydrodynamic theory](#), as the lumbar shunt can be responsible of the disorders of CSF circulation. Johnston et al. also reported one patient having syrinx formation before ACM suggesting that “Chiari malformation may follow rather precede syrinx formation” in some cases <sup>10)</sup>

## Treatment

[Acquired Chiari Malformation Treatment](#).

## Case reports

A sneeze caused acute left arm pain in a 36-year-old woman with a lumboperitoneal (LP) shunt that had been placed 3 years earlier for relief of headaches caused by pseudotumor cerebri. Numbness progressed up the left arm, neck, and back of the head and finally into the left face along with weakness of the hand and arm. Magnetic resonance imaging (MRI) and computed tomography revealed new tonsillar herniation and a large eccentric syrinx extending from C2 to T6. The functioning LP shunt was clamped, and a ventriculoatrial shunt was placed. Pain lessened and motor

function improved slightly. MRI revealed complete resolution of the syrinx and resolution of the tonsillar herniation <sup>11)</sup>.

## References

<sup>1)</sup>

Hoffman HJ, Tucker WS. Cephalocranial disproportion. A complication of the treatment of hydrocephalus in children. Childs Brain. 1976;2(3):167-76. PubMed PMID: 971636.

<sup>2)</sup>

Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y. Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. J Neurosurg. 1997 Jan;86(1):40-7. PubMed PMID: 8988080.

<sup>3)</sup>

Welch K, Shillito J, Strand R, Fischer EG, Winston KR. Chiari I "malformations"—an acquired disorder? J Neurosurg. 1981 Oct;55(4):604-9. PubMed PMID: 7277007.

<sup>4)</sup> <sup>8)</sup>

Payner TD, Prenger E, Berger TS, Crone KR. On the pathogenesis of syringomyelia: A review. J R Soc Med. 1994;73:798-806.

<sup>5)</sup>

Caldarelli M, Novegno F, Di Rocco C. A late complication of CSF shunting: acquired Chiari I malformation. Childs Nerv Syst. 2009 Apr;25(4):443-52. doi: 10.1007/s00381-008-0760-z. Epub 2008 Dec 5. PubMed PMID: 19066913.

<sup>6)</sup> <sup>9)</sup>

Chumas PD, Armstrong DC, Drake JM, Kulkarni AV, Hoffman HJ, Humphreys RP, Rutka JT, Hendrick EB. Tonsillar herniation: the rule rather than the exception after lumboperitoneal shunting in the pediatric population. J Neurosurg. 1993 Apr;78(4):568-73. PubMed PMID: 8450330.

<sup>7)</sup>

Di Rocco C, Velardi F. Acquired Chiari type I malformation managed by supratentorial cranial enlargement. Childs Nerv Syst. 2003 Dec;19(12):800-7. Epub 2003 Oct 28. PubMed PMID: 14586634.

<sup>10)</sup>

Johnston I, Jacobson E, Besser M. The [acquired Chiari malformation](#) and syringomyelia following spinal CSF drainage. Acta Neurochir 1998;140:417-28.

<sup>11)</sup>

Sullivan LP, Stears JC, Ringel SP. Resolution of syringomyelia and Chiari I malformation by ventriculoatrial shunting in a patient with pseudotumor cerebri and a lumboperitoneal shunt. Neurosurgery. 1988 Apr;22(4):744-7. PubMed PMID: 3374786.

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