

Chiari Malformation Classification

According to the type of [herniation](#) of the brain tissue displaced in the [spinal canal](#), and the characteristics of the anomalies of the development of the brain or the spine.

Among Chiari malformations, Arnold-Chiari I malformation, otherwise known as the Chiari type I malformation, is the more common variant. It is characterized by the caudal descent of the cerebellar tonsils through the foramen magnum. However, there is no accepted universal definition amongst clinicians as the malformation shows incongruity in clinic radiological correlation. Some authors, for example, have recently proposed to define the Chiari type I malformation as 'Chiari syndrome,' while the term malformation should be reserved for types II-III. Clinically, Chiari malformation type I can be asymptomatic ('hindbrain hernia') or manifests itself in late childhood or adulthood with tussive headaches and focal neurological manifestations. On the other hand, type II, III, and IV are congenital and clinically important. The nosographic collocation of type 0 or 0.5 Chiari malformation or 'Chiari like' (symptoms without tonsillar herniation), type 1.5 Chiari malformation (intermediate between type I and II), and complex Chiari is controversial and not universally accepted ¹⁾.

Variants of the originally described Chiari malformations are termed Chiari 0, Chiari 1.5, and Chiari 3.5. Each has distinct anatomical characteristics and some of these are extremely rare and incompatible with life (e.g. Chiari 3.5). Chiari IV malformation has been further clarified. Some physicians might be unfamiliar with the newer classifications of Chiari malformations because these conditions are rare or even unique. Furthermore, care is needed in using the term "Chiari IV malformation", which must be consistent with Chiari's original description, i.e. an occipital encephalocele containing supratentorial contents ²⁾.

[Chiari Malformation Type 0](#)

[Chiari Malformation Type 1](#)

[Chiari Malformation Type 1.5](#)

[Chiari Malformation type 2](#)

[Chiari Malformation Type 3](#)

[Chiari Malformation Type 3.5](#)

[Chiari Malformation Type 4](#)

¹⁾

Kular S, Cascella M. Chiari I Malformation. 2020 Jul 4. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 Jan-. PMID: 32119496.

²⁾

Azahraa Haddad F, Qaisi I, Joudeh N, Dajani H, Jumah F, Elmashala A, Adeeb N, Chern JJ, Tubbs RS. The newer classifications of the chiari malformations with clarifications: An anatomical review. Clin Anat. 2018 Apr;31(3):314-322. doi: 10.1002/ca.23051. Epub 2018 Feb 20. PMID: 29344999.

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