

Hindbrain Herniation: A Review of Embryological Theories

R. Shane Tubbs¹, Mohammadali M. Shoja², Mohammad R. Ardalan²,
Ghaffar Shokouhi³, Marios Loukas⁴

¹ Department of Cell Biology and Section of Pediatric Neurosurgery,
University of Alabama at Birmingham, Alabama, USA

² Tuberculosis and Lung Disease Research Center, Tabriz University
of Medical Sciences, Tabriz, Iran

³ Department of Neurosurgery, Tabriz University of Medical Sciences, Tabriz, Iran

⁴ Department of Anatomical Sciences, St. George's University, Grenada

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SUMMARY

Herniation of the hindbrain outside of the setting of intracranial hypertension, trauma, and brain tumors is an uncommon phenomenon with estimated incidence of less than 1%. In the late 1890's, Hans Chiari, a German pathologist, classified hindbrain herniation into three forms. This classification was then extended to include six types. We reviewed the current literature for the proposed embryological theories as well as the potential genetic mutations/syndromes associated with the hindbrain herniation or Chiari malformation. The review is illustrated by a unique cadaver with Chiari type I malformation (i.e. herniation of the cerebellar tonsils through the foramen magnum). Finally, it seems that no single theory could explain all forms of the Chiari malformation, and that this malformation might be a heterogeneous entity.

INTRODUCTION

In the early 1890's, Dr. Hans Chiari, professor of pathological anatomy at the German University in Prague, used autopsy specimens to describe congenital anomalies of the hindbrain later termed the Chiari malformations (Oakes and Tubbs, 2004). The Chiari malformations appeared to be separate anatomical entities, all of which involved the hindbrain; three of the four major classes consist of a type of hindbrain herniation, while the fourth (type IV) consists of cerebellar hypoplasia with no hindbrain herniation (Oakes and Tubbs, 2004; Tubbs and Oakes, 2004; Tubbs et al., 2003a). Type I Chiari malformation is specifically defined as greater than 5 mm of herniation of the cerebellar tonsils through the foramen magnum,