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CLASSICS IN PEDIATRIC NEUROSURGERY

Hans Chiari (1851–1916)

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Abstract
Introduction Hans Chiari will ever be remembered in neurosurgery for his paper (Ueber Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns) describing what is now regarded as the Chiari malformations in 1891.
Contributions In 1899 and in conjunction with British internist George Budd, he described hepatic vein thrombosis now known as Budd–Chiari syndrome. Among his other accomplishments, he studied the relationship between carotid artery plaques and thrombosis. Chiari is also remembered by the eponym Chiari's network, which is a network of fine fibers extending from the Thebesian to Eustachian valves in the heart. In 1883, he demonstrated a fistulous connection between a pneumatocele in the frontal lobes and the ethmoid sinuses in a patient who died of meningitis following rhinorrhea and thus first indicated a mechanism to explain meningitis in this context. Chiari also made significant contributions with his observations of pituitary adenomas. It is the works of Hans Chiari on which we base many of our current understandings of the pathology of the nervous system.

Keywords History · Craniocervical junction · Brain · Neurosurgery

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Introduction

Hans Chiari (Fig. 1) was born on November 4, 1851 in Vienna and wrote his first landmark paper (Ueber Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns) describing what is now regarded as the Chiari malformations in 1891 [2, 4]. Hans Chiari came from a family of physicians. His father, Johann Baptist Chiari, was a prominent gynecologist working in Vienna and Prague, credited with describing prolactinomas [10]. His brother, Ottokar Chiari, was a rhinolaryngologist [2]. Chiari studied medicine in Vienna, assisting one of the most revered pathologists at the time, Karl Rokitansky (1804–1878), at the Vienna Institute of Pathology [1]. Chiari went on to become a prosector [8], in what was at the time the center of medical advancement and knowledge, under the control of Rokitansky. Chiari's completion of medical school in 1875 coincided with Rokitansky's retirement. Richard Heschl (1824–1881) succeeded Rokitansky as Head of Pathological Anatomy in Vienna [1]. Chiari continued to assist Heschl until Heschl's death in 1881 [1]. While in Vienna, Chiari developed a reputation for his teaching skills and his attention to detail, laying the groundwork for his achievements later in his career. After Heschl's death, Chiari left Vienna for a pathology professorship at the German University in Prague [8].

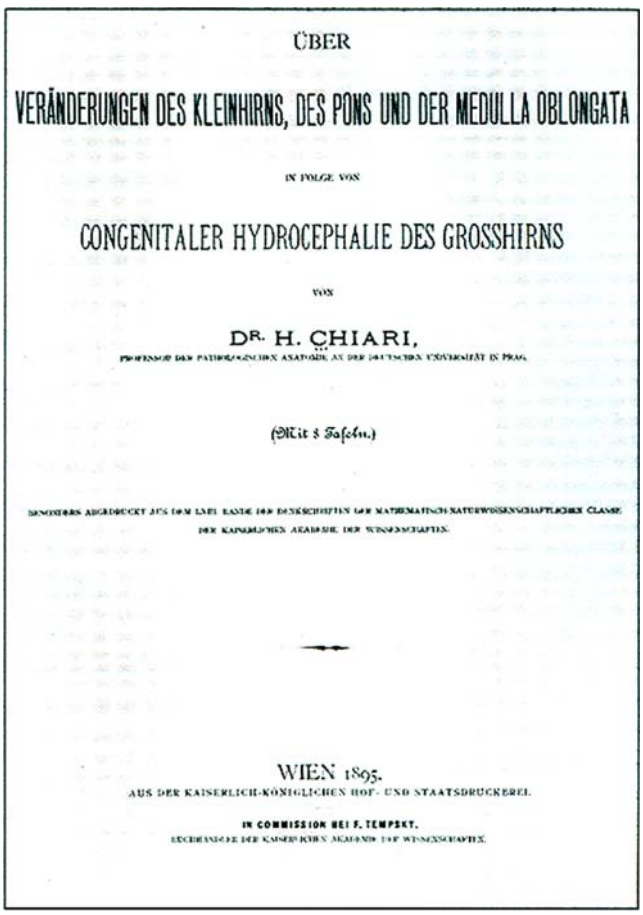
Fig. 1 Photograph of Hans Chiari (1851–1916)

Early contributions

Most of Chiari's accomplishments occurred while in Prague. In 1877, Chiari was noted for being the first to give an account of a choriocarcinoma [1]. In 1899 and in conjunction with British internist George Budd (1808–1882), Chiari provided a clinical and pathological explanation of hepatic vein thrombosis (Budd–Chiari syndrome) [1]. Prior to Chiari, Budd–Chiari syndrome had been described but never explained. Among his other accomplishments, he studied the relationship between carotid artery plaques and thrombosis [11] and published on the classical history of pathology. Chiari also had extensive publications on the autodigestive capacity of the pancreas [10] and described a network of fine fibers extending from the Thebesian to Eustachian valves in the heart. Chiari's name is also attached to the symptoms associated with aortoesophageal fistula after foreign body ingestion or gunshot wound. His most well-known work was in the field of neurology, where he described malformations of the brainstem and cerebellum. In 1883, he demonstrated a fistulous connection between a pneumatocele in the frontal lobes and the ethmoid sinuses in a patient who died of meningitis following rhinorrhea and thus first indicated a mechanism to explain meningitis in this context. Chiari also made significant contributions with his observations of pituitary adenomas [7]. It was between 1891 and 1896 that Chiari first published his works regarding hindbrain malformations.

Descriptions of hindbrain hernias

Chiari's type 1 malformation was limited to the cerebellum and described by him as a "peg-like elongation of tonsils and medial divisions of the inferior lobes of the cerebellum into cone-shaped projections which accompany the medulla oblongata into the spinal canal," while sparing the medulla [4]. In 1894, Julius Arnold (1835–1915) also described a malformation in neonates whereby the fourth ventricle and cerebellum herniated through the foramen magnum while sparing the medulla (Carmel 1972). Arnold studied under Rudolf Virchow (1821–1902) and Nikolaus Friedreich (1825–1882) in Heidelberg, later becoming professor of anatomy [10]. Chiari's type 2 malformation was described as a "displacement of parts of the cerebellum and elongated fourth ventricle, which reach into the cervical canal" [3]. Chiari later refined his description of type 2 malformations to include greater hindbrain involvement, as a "displacement of part of the lower vermis, displacement of the pons and displacement of the medulla oblongata into the cervical canal and elongation of the fourth ventricle into the cervical


ÜBER

VERÄNDERUNGEN DES KLEINHIRNS, DES PONS UND DER MEDULLA OBLONGATA

IN FOLGE VON

CONGENITALER HYDROCEPHALIE DES GROSSHIRNS

VON

DR. H. CHIARI,

(Mit 8 Tafeln.)

WIEN 1895.

AUS DER KAISERLICH-KÖNIGLICHEN HOF- UND STAATSDRUCKEREI

IN COMMISSION BEI F. TEMPSKY.

Fig. 2 Title page of Chiari's second paper (1895) describing hindbrain hernias

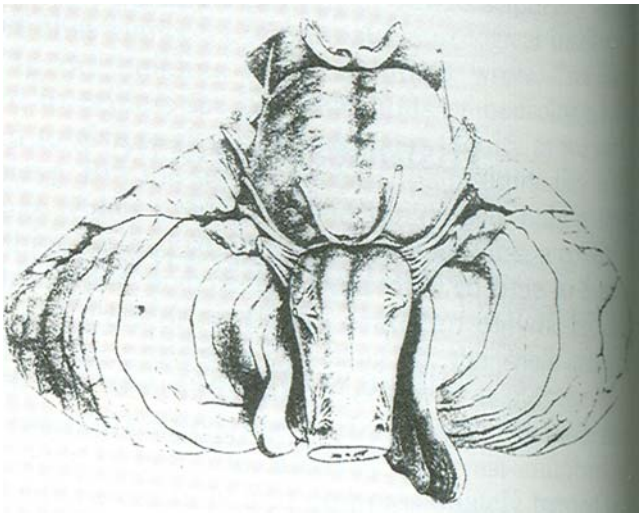


Fig. 3 Drawing of Chiari's first case from his 1896 publication (Ueber Veranderungen des Kleinhirns, des Pons und der Medulla oblongata Infolge von congenitaler Hydrocephalie des Grosshirns) illustrating the "Chiari 1 malformation"

canal" [3]. This type of malformation was described earlier by the Scottish physician John Cleland in 1883 [6] who called it the "basilar impression syndrome." Cleland noticed the malformation from autopsies and described it as the "inferior vermiform process, which extends up so far that what appears to be the pyramid touches the corpora quadrigemina, while the uvula looks backward and the laminated tubercle hangs down from an exaggerated velum posticum, as an appendix 3/4 of an inch in length, lying in the fourth ventricle" [6]. Cleland argued that the malformation resulted from primary dysgenesis of the brainstem and that "hydrocephalus was obviously of much later origin, when the different parts of the brains were already formed" [6]. Though it preceded Chiari's, Cleland's work had little impact on the scientific community's attempt at better understanding these malformations [3]. Chiari believed these malformations were from raised intracranial pressure causing herniation, because all the cases he observed also had hydrocephalus [3] (Fig. 2). In 1907, the Chiari type 2 malformation was renamed the Arnold–Chiari malformation by Schwalbe and Gredig while working in Arnold's laboratory [3]. However, the first physician recognized for describing this type of hindbrain herniation was Jean Cruveilhier (1791–1874), who described the malformation more than 50 years before in his atlas [10]. Chiari described one example of the most severe malformation, type 3—cervical spina bifida, whereby there is a partially absent tentorium cerebelli with prolapse of the fourth ventricle and cerebellum into the cervical canal, associated with a hydromyelic cavity communicating with the fourth ventricle [5]. His type 4 malformation had no degree of hindbrain herniation and consisted of only cerebellar hypoplasia, which Chiari also attributed to hydrocephalus [10]. In 1896, Chiari described an additional 63 cases of congenital hydrocephalus with an associated type 1 malformation in 14 of 63 and a type 2 malformation in seven of 63 [5] (Fig. 3).

Conclusion

In 1906, Chiari left Prague (as head of the university and professor extraordinarius and superintendent of the Prague pathological–anatomical museum) as a result of tensions within the Habsburg Empire to work at the University of Strasbourg, France [8, 9]. On May 6, 1916, after an accomplished career, Hans Chiari passed away due to a throat infection [1, 10]. Chiari's classifications of hindbrain herniation remain widely accepted by clinicians. This prolific writer published more than 170 writings between the years 1876 and 1916.

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