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PIONEERS IN NEUROLOGY

Hans Chiari (1851–1916)

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Hans Chiari will most be remembered for his 1891 paper, *Ueber Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns*, that described what is now regarded as the Chiari malformations [5]. Chiari (Fig. 1) was born on 4 November 1851 in Vienna. He came from a family of physicians and his father, Johann Baptist Chiari (1817–1854) was a prominent gynecologist who worked in Vienna and Prague and is credited with describing prolactinomas [1]. Chiari's brother, Ottokar, 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]. He was hired as a prosector [3] at the Vienna Institute, which was renowned for its knowledge and research under the control of Rokitansky. In 1875, he completed medical school and Rokitansky retired. Richard Ladislaus Heschl (1824–1881) succeeded Rokitansky as head of Pathological Anatomy in Vienna [1] and Chiari assisted him until Heschl's death in 1881 [1]. In Vienna, Chiari developed a reputation for his teaching skills, his attention to detail and cataloging. In 1878, he habilitated in pathological anatomy in Vienna and 4 years later he became extraordinarius at the German University in Prague. One year later, he was appointed ordinarius and superintendent of the pathological–anatomical museum in Prague [3].

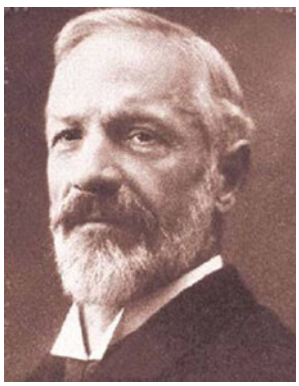

Fig. 1 Hans Chiari

Most of Chiari's accomplishments occurred while he was in Prague. For example, in 1877, he was noted as the first to describe the features 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, the so-called Budd–Chiari syndrome [1]. Such a syndrome had been described before but never explained to any extent. Also he studied the relationship between carotid artery plaques and thrombosis and published on the classical history of pathology. Chiari had a special interest in large glands and wrote extensively on the autodigestive capacity of the pancreas [1]. For the heart, he described connections (Chiari's network) between the Thebesian (valve of the coronary sinus) and Eustachian (valve of the inferior vena cava) valves. Chiari's name is also attached to the symptoms associated with aortoesophageal fistula after foreign

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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 probably described the first and only authentic case of traumatic pneumocephaly prior to roentgenography. 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. Interestingly, he implicated sneezing as a precipitating factor. Chiari also made important contributions on pituitary adenomas and in 1912, developed a novel transnasal approach to these lesions [4]. In 1888, he observed that syringes usually communicate with the central canal of the spinal cord. It was in 1891 in the journal *Deutsche Medizinische Wochenschrift* and later in 1896 that Chiari first published his works regarding hindbrain malformations.

Chiari's type 1 malformation was first described by him in a 17-year-old woman who died of typhoid fever and suffered from hydrocephalus but had "no symptoms referable to the cerebellum or medulla." Her malformation was described 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 [5]. In 1894, Julius Arnold (1835–1915) described a malformation in neonates whereby the fourth ventricle and cerebellum herniated through the foramen magnum while sparing the medulla [6]. Arnold studied under Rudolf Virchow (1821–1902) and Nikolaus Friedreich (1825–1882) in Heidelberg, later becoming Professor of Anatomy [1]. Chiari's type 2 malformation was similar to Arnold's case and was described as a "displacement of parts of the cerebellum and elongated fourth ventricle, which reach into the cervical canal" [7]. 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 canal" [7]. This type of malformation had been described earlier by the Scottish physician John Cleland in 1883 [8], who called it the "basilar impression syndrome." Cleland 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 three-fourths of an inch in length, lying in the fourth ventricle" [8]. 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" [8]. Cleland's work had little impact on the scientific community's attempt at better understanding these malformations of the hindbrain [7]. Chiari believed these malformations were secondary to hydrocephalus [7]. In 1907, the Chiari type 2 malformation was renamed the Arnold–Chiari malformation by Schwalbe and Gredig, from Arnold's laboratory [7]. However, the first physician to describe hindbrain herniation was Jean Cruveilhier (1791–1874), in his atlas produced between 1829 and 1842 [1]. Chiari described a single example of the most severe malformation of the hindbrain observed by Cruveilhier, now type 3; cervical spina bifida, with a partially absent tentorium cerebelli and prolapse of the fourth ventricle and cerebellum into the cervical canal, associated with hydromyelia [6]. In Chiari's type 4 malformation there was no hindbrain herniation but cerebellar hypoplasia [9]. In 1896, Chiari described an additional 63 cases of congenital hydrocephalus with an associated type 1 malformation in 14/63 and a type 2 malformation in 7/63 [10].

In 1906 and as a result of tensions within the Habsburg Empire, he left Prague (as head of the university and professor extraordinarius and superintendent of the pathological-anatomical museum) and moved to the University of Strasburg (German territory at the time) as ordinarius of pathological anatomy [3, 10]. On 6 May 1916, Hans Chiari passed away due to a throat infection [1, 9]. Chiari's classifications of hindbrain herniation remain widely accepted by clinicians. This prolific writer published approximately 180 papers between the years 1876 and 1916 and was always very careful to give credit to the discoveries of others.

References

1. Pearce JMS (2003) Fragments of neurological history. Imperial College Press, London, pp 395–398
2. Bejjani GK (2001) Definition of the adult Chiari malformation: a brief historical overview. Neurosurg Focus 11:1–8
3. Howard JM, Hess W (2002) History of the pancreas: mysteries of a hidden organ. Plenum Publishers, New York, pp 183–184
4. Greenblatt SH (1997) A history of neurosurgery in its scientific and professional contexts. American Association of Neurological Surgeons, Park Ridge
5. Chiari H (1891) Ueber Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns. Dtsch Med Wochenschr 17:1172–1175
6. Chiari H (1896) Ueber Veranderungen des Kleinhirns, des Pons und der Medulla oblongata Infolge von congenitaler Hydrocephalie des Grosshirns. Denkschriften Kais Akad Wiss Math-Naturw 63:71–116
7. Carmel PW, Markesbery WR (1972) Early descriptions of the Arnold–Chiari malformation: the contribution of John Cleland. J Neurosurg 37:543–547

8. Cleland J (1883) Contribution to the study of spina bifida, encephalocele, and anencephalus. J Anat Physiol London 17:257–291
9. Ashwal S (1990) Founders of child neurology. Norman Publishing: San Francisco, 190 pp
10. Koehler PJ (1991) Chiari's description of cerebellar ectopy (1891) with a summary of Cleland's and Arnold's contributions and some early observation on neural-tube defects. J Neurosurg 75:823–826