
The Chiari Malformations: A Historical Context

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Abstract

It was not until the late nineteenth century when reports of hindbrain herniation (Chiari malformations) began to be published. However, even earlier reports can be found in the extant literature. These include a report of hindbrain herniation in association with myelomeningocele by Tulp (1593–1674) in 1641. Cleland and Arnold would also report cases of hindbrain herniation found in patients with myelomeningoceles. Probably the first description of hindbrain herniation in the absence of myelodysplasia was made by Langhans in 1881. However, it was Chiari, 10 years later, who would classify and further our knowledge of these embryologic derailments. Penfield, Gardner, and Van Houweninge Graftdijk would each further our understanding of these malformations via surgical intervention. The evolution of surgery for hindbrain herniation is indebted to pioneers such as those described herein. Our current understanding and treatment of these embryological derailments are based on years of observation and surgical trial and error.

Early Descriptions of Hindbrain Herniation

Although a thorough study of hindbrain herniations associated with spina bifida aperta would not take place until the late nineteenth century, rare reports are found in earlier literature. For example, in *Observationes Medicae*, published in 1641, the famous Dutch physician and anatomist Nicolaes Tulp (1593–1674) described a myelodysplastic individual and may have referred to hindbrain herniation [2, 7]. In 1829, Jean Cruveilhier (1791–1874) of Paris also described a patient born with myelomeningocele in whom “... the considerably dilated cervical region

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contained both the medulla oblongata and the corresponding part of the cerebellum, which was elongated and covered the fourth ventricle, itself enlarged and elongated [7].”

Probably the first description of hindbrain herniation in the absence of myelodysplasia and what would become known as Chiari I malformation was described by Theodor Langhans as “pyramidal tumors.” Langhans was born September 28, 1839, in Usingen (Nassau) Germany, and studied under Henle in Göttingen and von Recklinghausen in Berlin (see Fig. 2.1) [20]. He was also a student under such names as Virchow, Trauber, and Frerichs [6]. He served as assistant to von Recklinghausen until 1867 [24]. He was later made professor ordinarius in Giessen and then moved to Switzerland in 1872 where he was appointed professor and chair of Pathological Anatomy in Bern succeeding Klebs [23]. Langhans with the physician Sahli and the surgeon Kocher formed a triumvirate, which made the medical school at Bern famous [6, 30, 36].

In his 1881 publication *Über Höhlenbildung im Rückenmark in Folge Blutstauung* (regarding cavity creation in the spinal cord as a consequence of obstruction to blood flow), Langhans made many observations and hypotheses that were far ahead of his time [3]. For example, he speculated that pathology at the foramen magnum resulted in syrinx formation [25]. The following is a translation of excerpts of Langhan’s publication *Über Höhlenbildung im Rückenmark in Folge Blutstauung* [3]:

In the case, which first brought to my attention the necessity to look for cavity formation in the spinal cord following a change in the cerebellar cavity, I could not find a cause for the increase in pressure; but great pressure on the pons and medulla oblongata from above was indeed apparent. Upon dissection of the cerebellum, nothing was of note except for an obvious/significant development of both tonsils, which protruded down in the form of two symmetrical pyramidal tumors and pushed the medulla oblongata in a frontal direction at almost a right angle.

The formation of the cavities, according to my observations, was connected to other changes in the central nervous system, more specifically to changes in the cerebellar cavity, which must have impeded the circulation to a great extent. The increase in pressure in the cerebellar cavity will

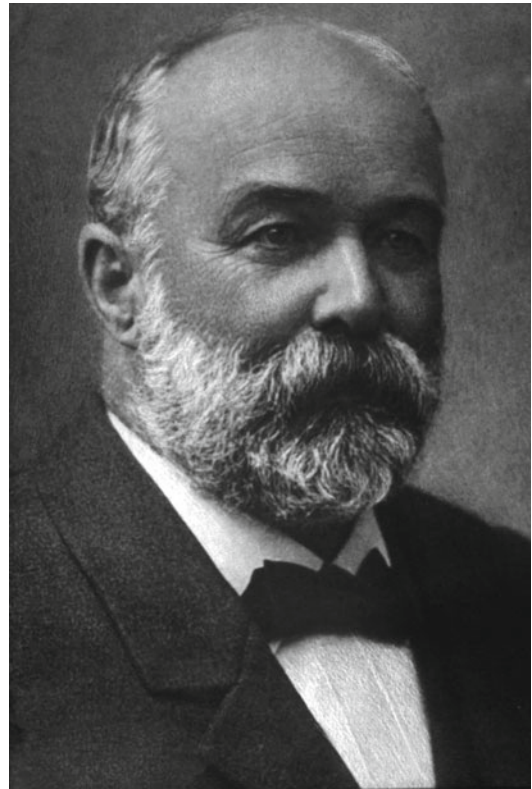


Fig. 2.1 Theodor Langhans

hinder or greatly impede the outflow of blood and cerebral spinal fluid.” “In all cases, the ventral part of the spinal cord is affected and if at all, only a small portion of the dorsal part. The cavities do not start in the medulla oblongata at the calamus scriptorius or in the upper 1–2 cm of the spinal cord.” “The direction in which the central canal extends is constant – to the side and posteriorly. In my opinion, the decisive factor for this is the consistency of the white matter. The cavity creation starts there where the increased pressure, which exists in the cerebellar cavity stops and, therefore, a diverticulum can only occur toward the area of less pressure.” “According to my theory, a diverticulum is more likely to occur than a widening of the central canal, because the development of the diverticulum in the dorsal part meets less resistance than a central expansion [3].

These descriptions are striking for several reasons including Langhans first describing pathologic tonsillar ectopia and hypothesizing that this obstruction at the foramen magnum results in development of syringomyelia. Additionally, the fact that syringomyelia normally does not include

the first segment of the cervical cord was clearly recognized by Langhans [22]. Lastly, Langhans realized that fluid accumulation within the spinal cord could occur via dilation of the central canal or outside of this region.

Hans Chiari will most be remembered for his 1891 paper *Ueber Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns* (concerning changes in the cerebellum due to hydrocephalus of the cerebrum) that described what is now regarded as the Chiari malformations [31–33]. Chiari (see Fig. 2.2) was born on November 4, 1851, in Vienna. Chiari came from a family of physicians and his father, Johann Baptist Chiari (1817–1854), is credited with describing prolactinomas [1]. 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 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, Chiari 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 1878, Chiari 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].

Most of Chiari's accomplishments occurred while he was in Prague. For example, in 1877, Chiari 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]. Prior to Chiari, such a syndrome had been described but never explained to any extent. Among his other accomplishments, Chiari studied the relationship between carotid artery plaques and thrombosis. Chiari's name is also attached to the symptoms associated with aortoesophageal fistula after foreign body ingestion or gunshot wound. In 1883, Chiari probably described the first and only



Fig. 2.2 Hans Chiari

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, Chiari implicated sneezing as a precipitating factor for this pathogenesis. Chiari also made significant contributions with his observations of pituitary adenomas and, in 1912, developed a novel transnasal approach to lesions of the pituitary gland [4]. Of note, Schloffer, who first performed a transsphenoidal pituitary operation in Innsbruck, Austria, examined pituitary adenomas from specimens that he obtained from Chiari in Prague.

In 1888, Chiari 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 hind-brain malformations. Chiari's type I 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 single myelodysplastic infant without hydrocephalus 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 II 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 II 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]. In 1907, the Chiari type II malformation was renamed the Arnold-Chiari malformation by Schwalbe and Gredig while working in Arnold’s laboratory [21]. Although little attention was given to the posterior fossa abnormalities in this report, Arnold’s students seized this opportunity to immortalize their professor by affixing the moniker “Arnold-Chiari malformation” to this condition [2, 34]. In the end, however, it was the significant contributions of Chiari that shed the most light on these forms of hindbrain herniation; thus, now referring to them as Chiari malformations is appropriate.

Chiari type II malformation was described earlier by the Scottish physician John Cleland in 1883 [8] 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 hang down from an exaggerated velum posticum, as an appendix three fourths of an inch in length, lying in the fourth ventricle” [8, 28]. 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, 35]. Though it preceded Chiari’s, Cleland’s work had little impact on the scientific community’s attempt at better understanding these malformations of the hindbrain [7, 26]. Chiari believed these malformations were due to hydrocephalus [7]. Chiari described one example of the most severe malformation of the hindbrain seen observed by him, type III; 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 [6]. Chiari’s type IV malformation had no degree of hindbrain herniation and consisted of cerebellar hypoplasia, which Chiari also attributed to hydrocephalus [9]. In 1896, Chiari described an additional 63 cases of congenital hydrocephalus with an associated type I malformation in 14/63 and a type II malformation in 7/63 [10].

In 1906 and as a result of tensions within the Hapsburg Empire, Chiari left Prague (as head of the university and professor extraordinarius and superintendent of the Prague pathological-anatomical museum) to travel to the University in Strasbourg, France, where he was appointed ordinarium of pathological anatomy [3, 10]. On May 6, 1916, after an accomplished career, Hans Chiari passed away due to a throat infection [1, 9]. 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 [27].

Many years later, in 1935, Russell and Donald [12], at the London Hospital, described ten additional pathological specimens of hindbrain herniation.

Surgical History of the Chiari Malformations

Although many have attributed the first successful patient series to Gardner in the 1950s, earlier attempts at surgical decompression were attempted [15]. In the late 1930s, Wilder Penfield

in Montreal commented “the anomaly [Chiari malformation] may present itself as an unexpected clinical problem to be dealt with by the neurosurgeon.” In 1938, Penfield and Coburn [11] reported a 29-year-old woman with loss of hearing and weakness on the right side of the face. Her history included removal of a thoracic “spina bifida” in infancy. On physical examination, she was noted to have nystagmus, absence of the right corneal reflex, truncal ataxia, and decreased peripheral reflexes. The patient underwent posterior cranial fossa exploration with the authors not considering hindbrain herniation in their differential. Later, they stated:

In retrospect, it seems that we should have suspected the Arnold-Chiari malformation. Instead, a suboccipital craniotomy was carried out, with a tentative diagnosis of a tumor of the acoustic nerve bilaterally [11].

Unfortunately, the patient of Penfield and Coburn never regained consciousness and died 2 months later. At autopsy, the authors identified a Chiari II malformation and hydrocephalus. These authors suggested that in the future, the cerebellar tonsils be left intact and the posterior margin of the foramen magnum be removed with the posterior elements of C1 and C2 [11].

Not known to many is that 8 years prior to the publication of Penfield and Coburn, the Dutchman Cornelis Joachimus Van Houweninge Graftdijk (1888–1956) (see Fig. 2.3) operated a patient, in 1930, with myelomeningocele and ventriculogram-proven hindbrain herniation who had rapid head growth [29]. This contribution was published in his thesis for a Doctorate of Medicine entitled *Over Hydrocephalus* (About Hydrocephalus) [16]. With surgery, he attempted to relieve CSF obstruction at the craniocervical junction by redundant cerebellar tissue. Although his patient died, this report marks the first known attempt at surgical correction of a hindbrain herniation. Van Houweninge Graftdijk said:

I decided to try widening the space through which the brain had herniated in order to allow for better flow of CSF. [16]

The idea that displacement of the foramina of the fourth ventricle into the upper end of the spinal canal might precipitate hydrocephalus, and

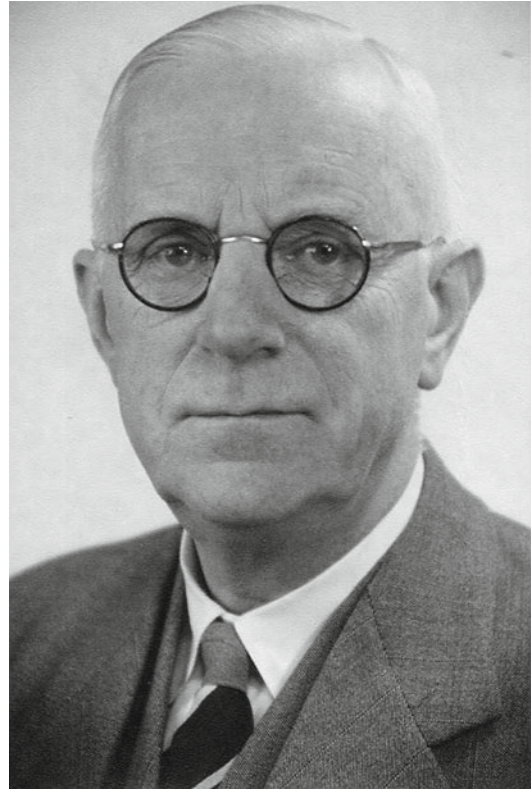


Fig. 2.3 Cornelis Joachimus Van Houweninge Graftdijk

that the whole of the anomaly might act as a valve, was first expressed by Van Houweninge Graftdijk in 1932 [16]. For this case, part of the occipital bone and posterior elements of the first two vertebrae were removed. Unfortunately, the patient developed fever on postoperative day 2 and 84 days after the operation; the bladder was perforated so that on day 98 postoperative, the patient died.

Van Houweninge Graftdijk also presented evidence that fluid could flow more readily from the spine to the head than vice versa [18]. It was in children with spina bifida that he concluded that the pressure within the meningocele was lower than the pressure within the cranium [19]. Conversely, Russell and Donald [12] commented on Van Houweninge Graftdijk’s theory but hypothesized that hindbrain hernias did not cause hydrocephalus in these patients. Interestingly, in 1935, these authors stated:

If hydrocephalus, either congenital or postoperative, were due to such a malformation, then

air injected by the lumbar route would collect in the ventricles and not in the sulci upon the cerebral convexities. Such a result would be not only of academic interest but of clinical importance. It would point to the desirability of decompressing the spinal cord at the foramen magnum to facilitate the circulation of fluid in the leptomeningeal spaces. Such an operation has not yet been carried out. [12]

However, these authors, in a footnote, stated that while their above-mentioned paper was in press, their attention was drawn to the writings of Van Houweninge Graftdijk. Van Houweninge Graftdijk also surmised that cerebrospinal fluid can readily escape in an upward direction from the vertebral canal into the ventricles or cerebral meningeal spaces but has difficulty in passing from the ventricles down into the vertebral canal. To address such issues surgically, he excised redundant cerebellar tissue and/or bone over the posterior surface of the malformation as previously mentioned [12, 16]. Van Houweninge Graftdijk also postulated that caudal traction theory by the myelomeningocele is responsible for “pulling” the hindbrain caudally thus resulting in a Chiari II malformation [19].

Cornelis Joachimus Van Houweninge Graftdijk was born in Giessendam, Holland. His father was a family doctor in Giessendam, and his brother died young as the result of hydrocephalus. Although his brother and sisters were given the family surname “Graftdijk,” Cornelis was given the name “Van Houweninge Graftdijk” as his parents did not want this surname to die out. Cornelis graduated from the University of Leiden, became a physician in 1913, and, following the advice of his teacher Professor Korteweg, became a ship doctor so that he could gain some “practical knowledge.” In 1914, he studied surgery with Prof. Zaaijer for 5 years then began his own practice in Leiden at the hospital Diaconessenhuis. He maintained his affiliation with Prof. Zaaijer at the University Hospital in Leiden and, in addition to practicing general surgery, treated pediatric patients with hydrocephalus. This interest probably arose from his concern over his older brother who had hydrocephalus. Van Houweninge Graftdijk trained doctors in surgery and, for many years, was “chef de clinique” of the Department of Surgery at University Hospital.

In the early 1930s, Van Houweninge Graftdijk wrote his dissertation [16] on hydrocephalus and on June 21, 1932, he obtained his doctor’s degree cum laude. Soon after, Prof. Zaaijer left the university and was replaced by Prof. Suermondt who had no interest in hydrocephalus. It is not clear why Van Houweninge Graftdijk did not succeed Prof. Zaaijer but as a result of this, he left University Hospital and was not able to continue working on hydrocephalus. He continued to practice at the hospital Diaconessenhuis. He became chairman of the Dutch Association of Surgery, and in 1940, he wrote a book *Heelkunde voor Den Medicus Practicus* [17], a textbook of surgery for family doctors.

In 1938, McConnell and Parker [8] published their results of posterior fossa decompression for Chiari I malformation in five patients. Two of these patients had successful outcomes. In 1945, Bucy and Lichtenstein [2] reported successful decompression for a Chiari I malformation of a 40-year-old woman without hydrocephalus, and in 1948, Chorobski and Stepien [3] operated a woman with life-altering Valsalva-induced headache and Chiari I malformation that had full resolution of her symptoms. Most remembered for his direct approach to the hindbrain was Gardner. In 1957, he and Goodall [6] reported their efforts at surgically addressing syringomyelia by decompressing the hindbrain and sealing off the hypothetical communication between the syrinx and fourth ventricle in 17 patients. They reported improvement in 13, decline in 3, and death in 1. In their series, some patients had improvement of preoperative symptoms. The publication of Gardner and Goodall appears to have resulted in the widespread adoption of posterior fossa decompression for hindbrain herniation with larger reports following over the next few decades [1, 5, 7, 9, 10, 13]. Finally, it was Gardner in the 1950s that showed in a large series of patients that decompression of the hindbrain herniation in patients with syringomyelia often improved symptoms in patients. This report led to widespread adoption of this method of treatment. In fact, a PubMed search of the terms Chiari malformation and surgery yielded roughly 1,500 publications between 1950 and 2011 [14].

The evolution of surgery for hindbrain herniation is indebted to pioneers such as those described herein. Our current understanding and treatment of these embryological derailments are based on years of observation and surgical trial and error.

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