The Chiari Malformations

R. Shane Tubbs
Mehmet Turgut
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Editors

Second Edition

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Foreword

While the brain malformations that carry Hans Chiari's name continue to be listed on the National Institutes of Health (NIH) Website under *Genetic and Rare Diseases Information Center*, we now recognize that the low-lying cerebellar tonsil that has long been pathognomonic of Chiari I is a common, albeit age-dependent, anatomical variant. This and other observations came to light since publication of the first edition of this book, and have contributed to refueling the diagnostic and therapeutic controversies associated with these entities, with debates on what combination of anatomical and clinical features we should consider pathological, and when and how we treat them. Concurrently, investigations into the diagnosis and surgical management of hindbrain hernias have shifted from an exploratory twentieth century period to an analytical twenty-first.

Alongside other pediatric neurosurgical disciplines, the Chiari field was defined and developed by a group of inspired young neurosurgeons—now hailed as pioneers and senior authorities—who believed children deserved their own surgical specialists, and proceeded, among others, to categorize and define the various field disciplines and their surgical management.

As the reigns of the specialty were gradually passed down from mentor to mentee, the new generation began to shepherd the field into an era of science, quality improvement, and technology. Consequently, the observations and suppositions of the past are now being scrutinized and analyzed through the lens of evidence-based medicine; the diagnostic and surgical challenges are being steadfastly vanquished by previously unimaginable technology; and the pathophysiology is being unraveled using the scientific method.

A necessary complement has been the acquisition of additional expertise and the bridging of professions. Accordingly, the Chiari research network, much of which is represented here, now comprises an amalgam of experts in the basic sciences, biostatistics and epidemiology, genetics, engineering, and others, as well as pediatric neurosurgeons, some of whom have also procured supplemental training in these areas. The result is a comprehensive 52-chapter second edition, in which gray-haired and

emerging world authorities on the anatomical, physiological, radiological, clinical, and surgical features of the Chiari malformations and related disorders join hands to report on the history, present-day status, and future prospects of this ripening field.

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Preface

During the last 50 years the Chiari malformations in both children and adult populations have been the topic of much controversy in the literature. Nowadays, the increased use of magnetic resonance imaging has shown that Chiari malformations may be much more common than previously believed. Nevertheless, the optimal choice of the surgical intervention itself is even more controversial, ranging from craniovertebral decompression alone to posterior cranial fossa decompression with duraplasty and cerebellar tonsillar resection.

Since the publication of our first edition on the Chiari malformations, additional topics have been identified that necessitate conversation. These include chapters on additional embryological and anatomical topics, ventral compression, craniosynostosis, diagnostics, advanced imaging, what to do with the so-called "benign" Chiari malformation, experimental models, and predictive and treatment analyses. This has increased the number of germane topics by 20 chapters. As in the first edition, this book includes chapters from leaders in the field. We hope that neuroscientists and clinicians at all levels will find this book to be a useful resource.

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Part I

History and Classifications

1 A History of the Chiari Malformations

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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) (Fig. 1.1) described a myelodysplastic individual and may have referred to hindbrain herniation [1, 2].

In 1829, the French pathologist and anatomist Jean Cruveilhier (1791–1874) (Fig. 1.2) of Paris also described a patient born with myelomeningocele in whom "…the considerably dilated cervical region contained both the medulla oblongata and the corresponding part of the cerebellum, which was elongated and covered the fourth ventricle, itself enlarged and elongated" [3]. What was described by Cruveilhier is now called the Chiari type II malformation. However, Cruveilhier's observation of the malformation occurred

Fig. 1.1 Nicolaes Tulp

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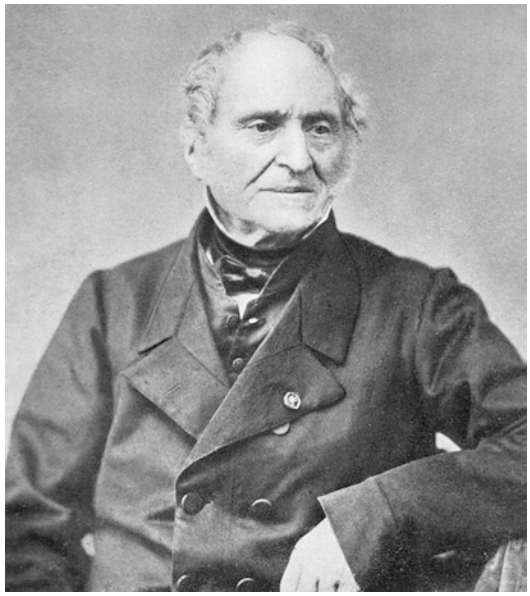


Fig. 1.2 Jean Cruveilhier

55 years before it was definitively described by Arnold and Chiari. In fact, some have used the term "Cruveilhier-Cleland-Chiari malformation" to describe this subset of patients with hindbrain herniation [3]. John Cleland published on spina bifida in 1883 [4]. The patient described by Cruveilhier was a child with a myelomeningocele who succumbed from sepsis secondary to meningitis [3]. His description on autopsy revealed bony anomalies of spina bifida with the associated split cord malformation as well as the variations in the posterior cranial fossa and cerebellum that are now known as a Chiari type II malformation. Two other cases with similar findings were noted by Cruveilhier, leading him to conclude that spina bifida occurred secondarily to a developmental abnormality. Further observations by Cruveilhier revealed clinical cases that involved a sac covering the myelomeningocele. There were no dangerous findings associated with the child until the sac was opened. When the sac was punctured, the child would end up with one or more of the following symptoms: fever, convulsions, infections, paraplegia, sepsis, and seizures, with death usually occurring in several hours to days.

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 (Fig. 1.3) as "pyramidal tumors." Langhans was born September 28, 1839, in Usingen (Nassau), Germany, and studied under Henle in Göttingen and von Recklinghausen in Berlin [5]. He was also a student under such names as Virchow (Fig. 1.4), Trauber, and Frerichs [6]. He served as assistant to von Recklinghausen until 1867 [6]. 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. Langhans with the physician Sahli and the surgeon Kocher formed a triumvirate, which made the medical school at Bern famous [7].

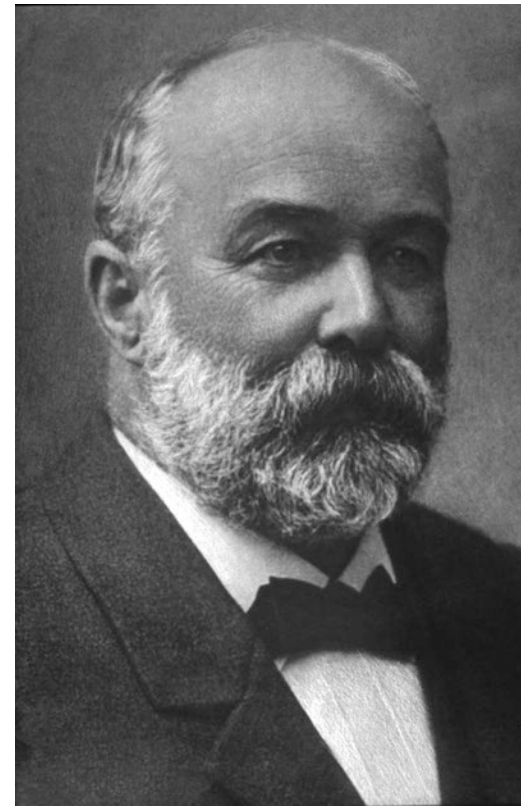


Fig. 1.3 Theodor Langhans

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 [8]. For example, he speculated that pathology at the foramen magnum resulted in syrinx formation

Fig. 1.4 Rudolph Virchow

[9]. The following is a translation of excerpts of Langhan's publication *Über Höhlenbildung im Rückenmark in Folge Blutstauung* [8]:

> 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 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. [8]

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 [8]. 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 [10–13]. Chiari (Fig. 1.5a) 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 [10]. 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. Chiari was hired as a prosector 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, and Chiari assisted him until Heschl's death in 1881. 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 [10].

Most of Chiari's accomplishments occurred while he was in Prague. For example, in 1877,

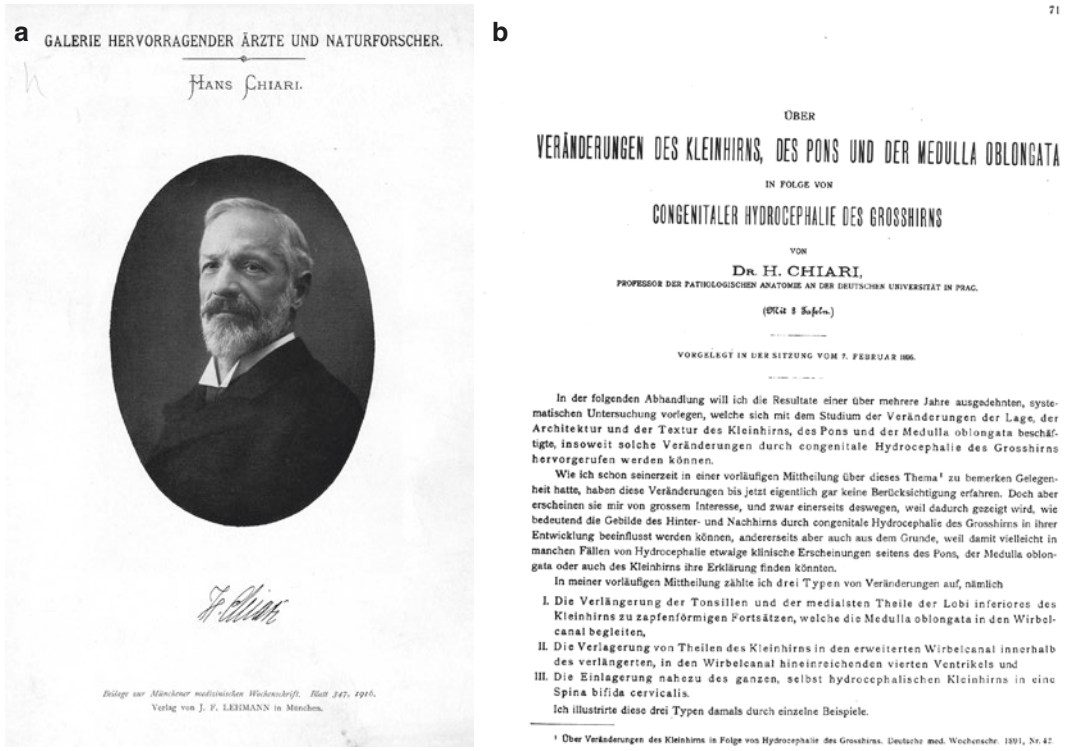


Fig. 1.5 (**a**) Hans Chiari. (**b**) First page of Chiari's seminal 1891 publication

Chiari was noted as the first to describe the features of a choriocarcinoma [10]. 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 [10]. 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 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 [14]. 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 syrinxes usually communicate with the central canal of the spinal cord. It was in 1891 in the journal *Deutsche Medizinische Wochenscriff* [11] and later in 1896 (*Über Veränderungen des Kleinhirns, des Pons und der Medulla oblongata in Folge von congenitaler Hydrocephalie des Großhirns* (*Changes in the Cerebellum, Pons, and Medulla Oblongata due to Congenital Hydrocephalus of the Cerebrum*)) [12] (Fig. 1.5b) that Chiari first published his works regarding hindbrain malformations (Fig. 1.6a–h). Chiari's type I malformation was first described by him in a 17-year-old woman who died of typhoid fever

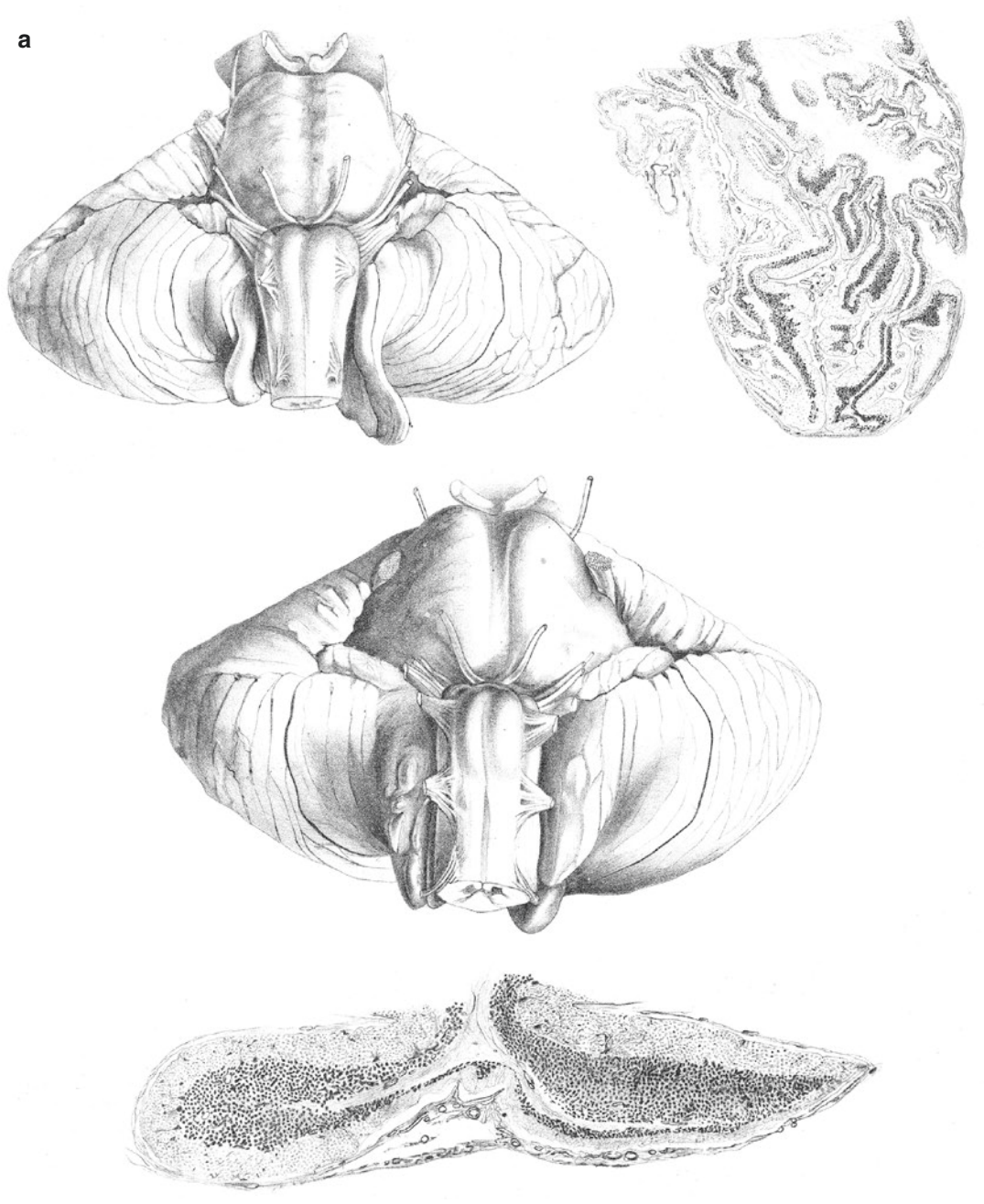
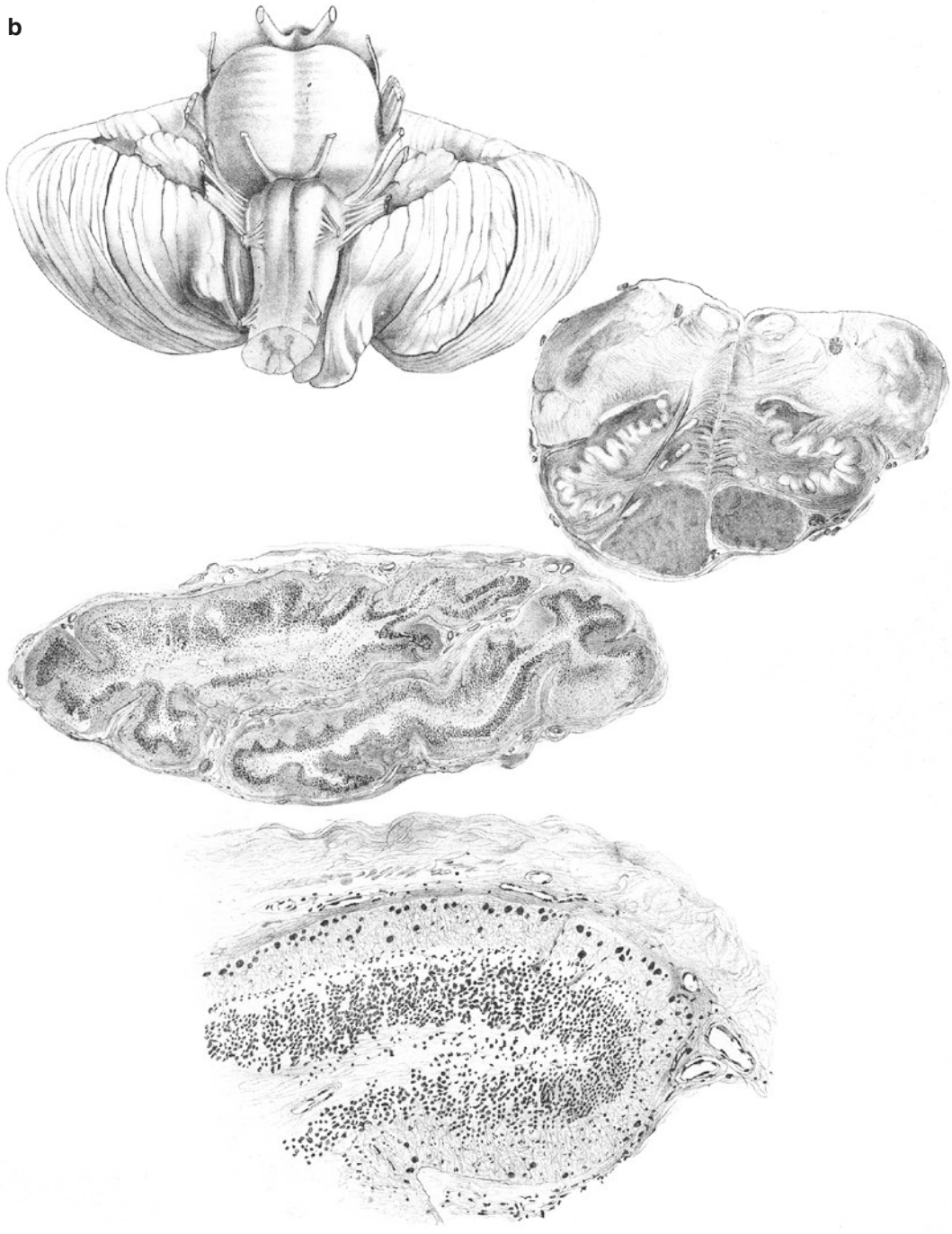


Fig. 1.6 (**a–h**) Drawings from Chiari's descriptions of the hindbrain hernias noting the findings at autopsy that would propagate in what are known today as the "Chiari malformations"



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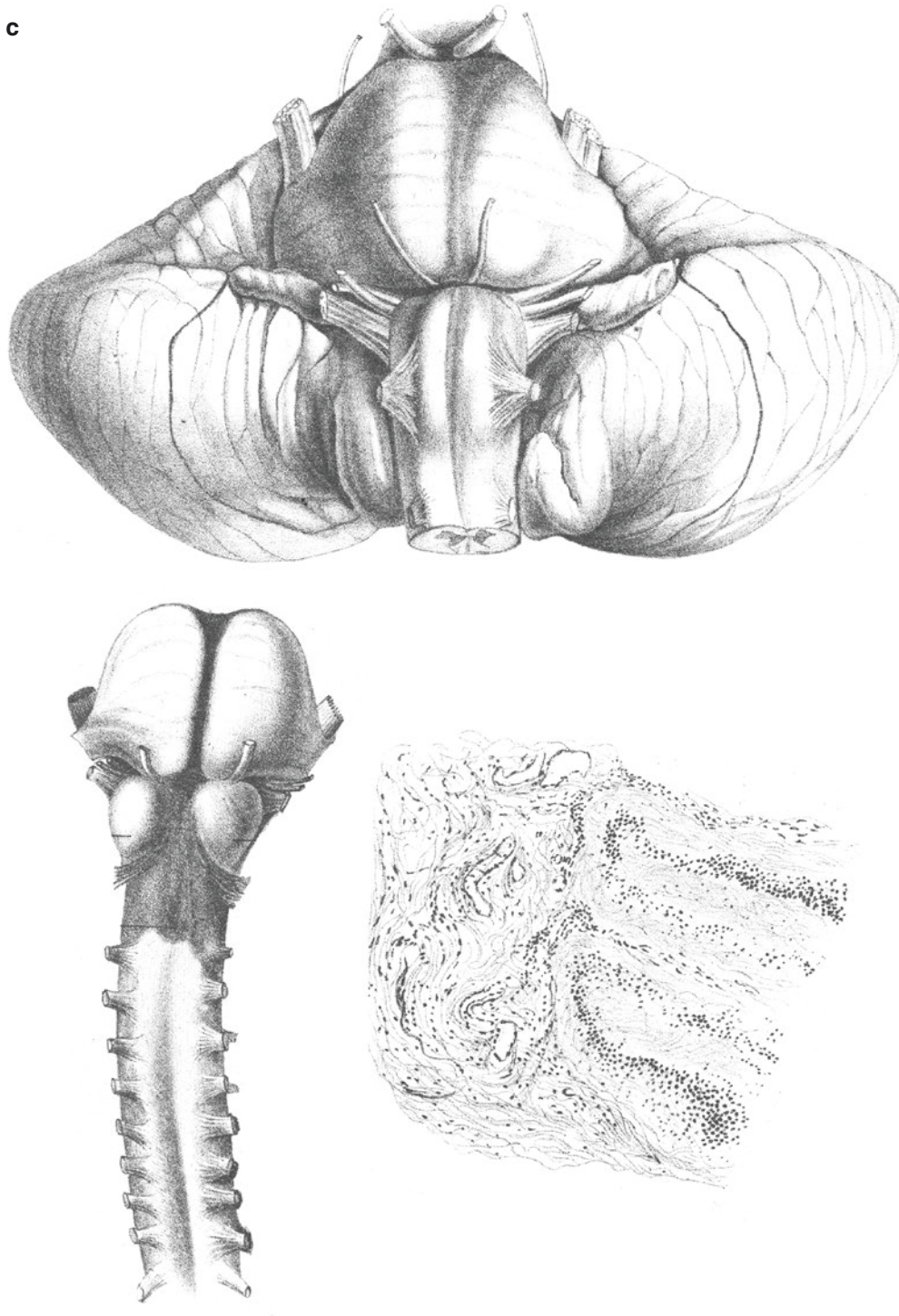

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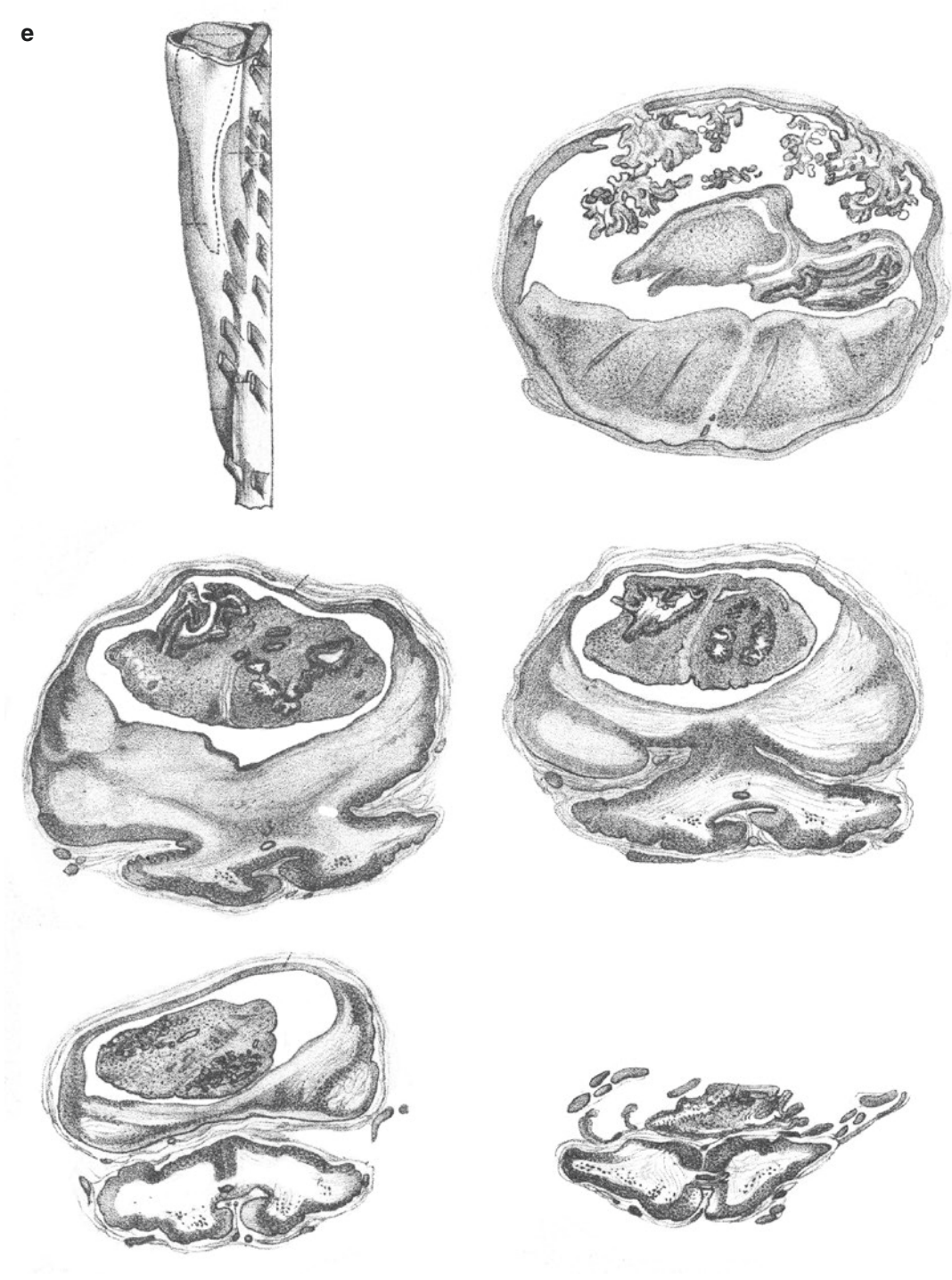

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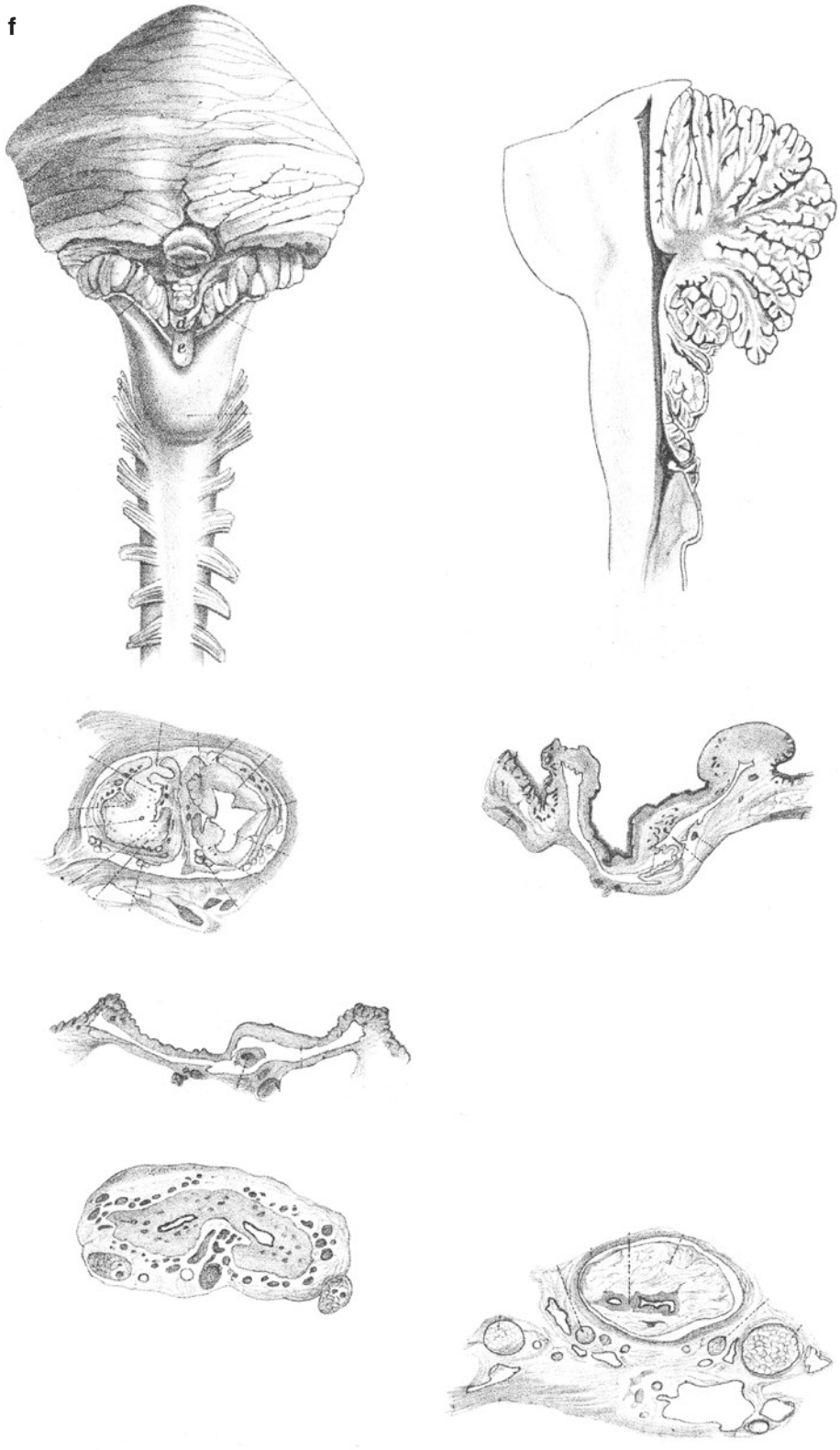

Fig. 1.6 (continued)

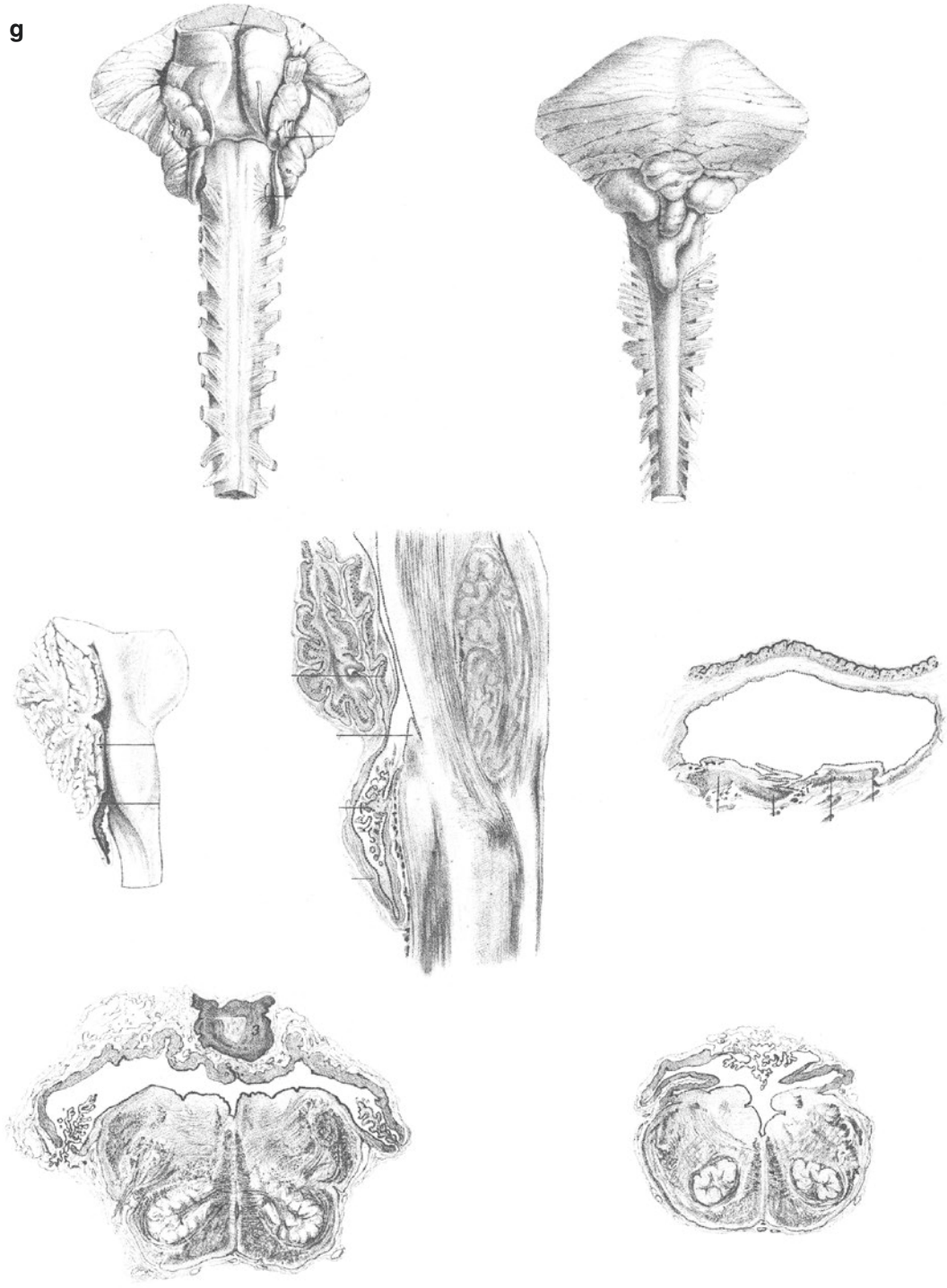

Fig. 1.6 (continued)