Nomenclature

20

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Contents

The terminology of Chiari malformation and syringomyelia is variable and at times controversial. Individual specialists can be quite dogmatic over which name or expression is correct. In this appendix the nomenclature and classification of syringomyelia and Chiari malformations is reviewed briefly and arguments for and against the various terms are presented. A more detailed historical review is given in Chap. 1.

20.1 Spinal Cord Cavitation

One of the controversies surrounding this subject is whether spinal cord cavities, containing fluid identical with or closely resembling cerebrospinal fluid, should be referred to as syringomyelia, hydromyelia, syringohydromyelia or hydrosyringomyelia. The term syringomyelia was first used by Charles-Prosper Ollivier d'Angers (1796–1845) in '*De la moelle épinière et de ses maladies*'[1] and subsequently in the more comprehensive work '*Traité des maladies de la moelle épinière contenant l'histoire anatomique, physiologique et pathologique de ce centre nerveux chez l'homme*'[2] (Ollivier d'Angers 1824, 1827). He derived the term from the Greek 'syringo' meaning tube or pipe and 'myelio' referring to the spinal marrow, and he used it to describe a tubular

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[1] Translated as 'On the spinal cord and its diseases'

[2] Translated as 'Treatise on the Spinal Marrow and its Diseases—anatomy, functions and general considerations on its diseases'

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fluid-filled cavity in the spinal cord, most often associated with spina bifida (Grossmann et al. 2006; Mortazavi et al. 2011; Walusinski 2012). Schüppel used the term hydroamyelus in 1865 to describe a dilatation of the central canal (Schuppel 1865). The finding of spinal cord cavities, apparently separate from the central canal and surrounded by gliosis, led to a proposal by Simon, in 1875, that hydromyelia be used to describe central canal dilation and distension and that the term syringomyelia be reserved to describe cavities and cystic conditions independent of the central canal (Newton 1969; Simon 1875). In 1876, Leyden concluded that hydromyelia and syringomyelia were identical conditions (Newton 1969; Leyden 1876), but Kahler and Pick supported a distinction between hydromyelia and syringomyelia. They made the observation that a hydromyelia is lined by ependyma, whereas glial cells form the wall of syringomyelia cavities (Newton 1969; Kahler and Pick 1879). Hans Chiari himself found that, in 45 out of 75 well-documented cases of the so-called syringomyelia, the cavity was, in all probability, connected to the central canal (Newton 1969; Chiari 1888). This observation and the difficulty in differentiating between hydromyelia and syringomyelia, by radiological, clinical or pathological means, led some to use the combined terms syringohydromyelia or hydrosyringomyelia to describe a cavity which is partially lined by ependyma but which also extends into the spinal cord substance (Hogg et al. 1998). Thus, some clinicians argue that the term syringomyelia should apply to a glial-lined cavity separate from the central canal, that hydromyelia be reserved for central canal dilation still lined by ependyma and that the term syringohydromyelia is correct for a cavity involving a dilated central canal that is partially lined by ependyma. However, the distinction is easily

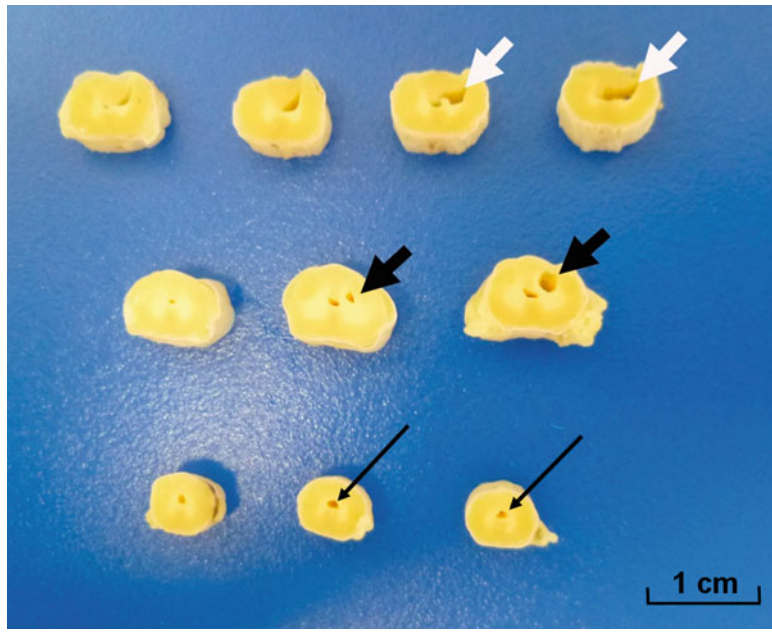

Fig. 20.1 Hydromyelia, syringomyelia or syringohydromyelia? In these sections of the spinal cord from a dog, it can be seen that some segments appear to have a syringohydromyelia (*top row*, short white arrows), some a separate syringomyelia cavity (*middle row*, short black arrows) and some a hydromyelia (*bottom row*, long arrows). The ependyma is disrupted in all sections on microscopy, and the apparently separate syrinx cavity is actually connected to the central canal more caudally or cranially, i.e. it is not truly separate. It can be argued that, without the aid of post-mortem and microscopic examination, it is impossible to categorise cavities on the basis of ependymal lining and central canal connection. It can also be argued that categorisation is completely unnecessary from a clinical point of view (Picture courtesy of Dr Fernando Constantino-Casas)

Table 20.1 Should syringomyelia be used in preference to syringohydromyelia?

Yes
1. Syringomyelia is already overwhelmingly used in preference to syringohydromyelia,[a,b] especially by those authors considered expert in this field
2. The classification of syrinx cavity by central canal connection and ependymal lining is unworkable (Fig. 20.1). Concurrent involvement of the central canal and disruption of the ependymal limiting is almost universal, so only one term is actually required
3. It is correct from a historical point of view. Ollivier d'Angers originally defined syringomyelia as a fluid-filled tubular dilatation within the spinal cord
4. Adding 'hydro' and an extra two syllables is superfluous and complicates a word that is already difficult for patients to understand, pronounce or remember
No
1. Syringohydromyelia may be defined as the coexistence of hydromyelia (fluid contained within the ependyma of the central canal) and syringomyelia (fluid forming a cavity within the white matter). Using the term syringohydromyelia makes it clear to the reader that a syrinx also involves the central canal
2. Differentiation between hydromyelia and syringomyelia is difficult to make by radiological, clinical or pathological methods. Use of the combined term, syringohydromyelia (or hydrosyringomyelia), acknowledges this reality
3. Adding a component, meaning 'water' or 'liquid', to a 'tube' in the 'marrow' makes it clear that this is a fluid-containing cavity, emphasising that it is a disorder of CSF circulation

[a]A PubMed search performed on 24th June 2012 revealed 136 articles when the key word 'syringohydromyelia' was used. In comparison 3,584 articles were found using the keyword 'syringomyelia'
[b]Online searches performed on 8th July 2012 in Wikipedia, Encyclopaedia Britannica, Oxford Dictionary, American Heritage Dictionary, Collins Dictionary and Merriam-Webster failed to find a listing for syringohydromyelia. Search results returned either 'not found' or defaulted to 'syringomyelia'

Table 20.2 Should we use hydromyelia to describe a small central dilation?

Yes
1. There is a need for a term to describe this MRI appearance. An ependymal-lined distension or dilation of the central canal has already been defined as hydromyelia; therefore, it is the most appropriate term to use
No
1. Many clinicians use the term inappropriately when syringomyelia would be more correct[a]
2. Using the term central canal dilation or distension is descriptive and less open to misinterpretation
3. Before using this term a clearer definition is required, e.g. the size of central canal dilation. In veterinary medicine central canal dilation with a transverse width of two millimetres or more is categorised as syringomyelia. If less than two millimetres, then it is described as central canal dilation (Knowler et al. 2011)
4. Use of the term hydromyelia implies pathology when it is not clear that this is always the case. By analogy, ventricular dilation (or ventriculomegaly) does not necessarily equate with clinically active hydrocephalus

[a]A PubMed search performed on 14th July 2012 using the criteria *hydromyelia* [*Title*] revealed 78 publications; however, only one was found (Jinkins and Sener 1999) which used the term hydromyelia to describe a central canal dilation. The remainder were referring to an extensive syrinx

blurred (Fig. 20.1). The simpler term syringomyelia is favoured by an increasing majority especially as recent post-mortem and experimental studies have suggested that the ependyma is disrupted following only minor central canal dilatation (Hu et al. 2011; Radojicic et al. 2007). The arguments for and against the terms syringomyelia, hydromyelia and syringohydromyelia are represented in Tables 20.1 and 20.2.

20.2 Abnormalities of the Craniovertebral Junction

The first known illustration of hindbrain herniation was in an early nineteenth-century anatomy atlas by Jean Cruveilhier (Cruveilhier 1829; Pearce 2000). There was also a brief description of a single case in a paper on encephalocoele and spinal bifida by John Cleland in 1883 (Cleland

Table 20.3 Classification of Chiari malformations

Chiari malformation	Original description by Chiari (Batzdorf 2001b)	Modern description
Type I	Elongation of the cerebellar tonsils and the medial part of the inferior cerebellar lobes into cone-like projections, which accompany the medulla into the spinal canal	Volumetrically small posterior cranial fossa with hindbrain overcrowding (Milhorat et al. 1999) The classic radiographic description is tonsillar herniation of at least 3 mm below the foramen magnum
Type II	Displacement of portions of the vermis and also of the pons and medulla into the spinal canal and elongation of the fourth ventricle into the spinal canal	Downward displacement of the cerebellar vermis, brainstem and fourth ventricle, associated with a myelomeningocoele (Geerdink et al. 2012)
Type III	Displacement of virtually the entire hydrocephalic cerebellum into a cervical spinal bifida	Hindbrain herniation into a high cervical or occipital encephalocoele (Castillo et al. 1992)
Type IV	Hypoplasia in the region of the cerebellum without displacement of portions thereof into the spinal canal	Obsolete term describing cerebellar hypoplasia unrelated to the other Chiari malformations
Type 0		Overcrowding of the posterior fossa with abnormal brainstem anatomy (posterior pontine tilt, downward displacement of the medulla, low-lying obex) but with normally placed cerebellar tonsils (Markunas et al. 2012)
Type 1.5		Cerebellar and brainstem herniation through the foramen magnum. Similar to type II malformation but not associated with spinal dysraphism (Tubbs et al. 2004)
Canine		Disparity in volume between the caudal cranial fossa and its contents so that the cerebellum and brainstem are herniated into or through the foramen magnum

1883). It is, however, Hans Chiari (1851–1916), a pathologist working in Prague, who is honoured with the medical eponym because he provided the first detailed description of four types of cerebellar pathology, observed during post-mortem studies of infants with congenital hydrocephalus (Chiari 1891, Chiari 1896). Descriptions of the various types of Chiari malformation have, however, evolved since (Table 20.3). Chiari was careful to acknowledge the work of others, and in his conclusion in the second paper, he noted the observations of Cleland and also of Arnold, who had described an infant with spina bifida and hindbrain herniation (Arnold 1894). Two loyal students of Arnold, Schwalbe and Gredig, subsequently inserted his name when describing four patients with the type II malformation (i.e. in association with myelomeningocoele) coining the term Arnold-Chiari malformation (Schwalbe and Gredig 1907). Consequently, some refer to the spectrum of disorders of hindbrain herniation as Arnold-Chiari syndrome (Bejjani 2001), albeit to the indignation of others (Solt 2011; Sarnat 2007). The majority now use the term Chiari malformation to describe hindbrain herniation. The arguments for and against the terms hindbrain herniation, Chiari malformation and Arnold-Chiari malformation are presented in Tables 20.4 and 20.5.

20.3 Communicating and Noncommunicating Syringomyelia

Syringomyelia and hydromyelia were traditionally classified into communicating and noncommunicating types (Milhorat et al. 1995). The term communicating indicated the presence of a connection between the syringomyelic cavity and the fourth ventricle. Communicating syringomyelia, or hydromyelia, is associated with hydrocephalus, especially of the fourth ventricle and conditions which cause obstruction to the outflow of this chamber, for example, post-meningitic or posthaemorrhagic hydrocephalus or complex hindbrain malformations such as

Table 20.4 Should hindbrain herniation be used in preference to Chiari malformation?

Yes
1. In common with most eponymous terms, Chiari malformation tells us nothing about the nature of the condition
2. There are many causes of hindbrain herniation including cranial constriction, spinal cord tethering, cranial settling, intracranial hypertension, intraspinal hypotension, basilar impression/invagination and craniosynostosis. Nomenclature should identify the underlying subgroup and cause, for example, 'hindbrain herniation secondary to intracranial hypertension'
3. Using eponymous terms for diseases is to be discouraged especially when there is a simple alterative
No
1. The eponym Chiari malformation is by far the most commonly used term[a]
2. Some cases with overcrowding of the posterior fossa and disrupted CSF flow (Chiari 0) do not actually have a hindbrain herniation
3. The classification of various Chiari malformations is commonly used and well understood. For example, the term 'Chiari type II' is much simpler to say or read than 'hindbrain herniation associated with spinal myelomeningocoele'

[a]A PubMed search performed on 14th July 2012 using the criteria (*Chiari* [*Title*]) *not Budd* [*Title*]), *not Arnold* [*Title*], revealed 1,422 publications. A search using the criteria *Chiari* [*Title*]) *and Arnold* [*Title*] revealed 509 publications. A search using the criteria *hindbrain herniation* [*Title*] revealed 22 publications

Table 20.5 Should the term Arnold-Chiari malformation be dropped?

Yes
1. Arnold's work does not merit an eponym, and it was an injustice that his name was added. Chiari provided a detailed and careful description of the pathology, whereas Arnold's observation in a single case was 'a ribbon of tissue that protruded through the foramen magnum'. Arnold did not even acknowledge previous work by Cleland and Chiari (Solt 2011; Sarnat 2007)
2. Arnold-Chiari is a confusing term because there is not universal agreement as to the definition. Some use it for all types of hindbrain herniation and others use it exclusively to refer to hindbrain herniation associated with a myelomeningocoele[a]
3. The eponym Arnold-Chiari malformation is not the most frequently used term[b]
4. Many patients think that Arnold-Chiari was one person
No
1. This term is useful to distinguish the type II malformation, i.e. hindbrain herniation associated with myelomeningocoele, and should be reserved exclusively for that
2. If we are going to use an eponymous term, then the contribution of this clinician should be recognised as well as that of Hans Chiari

[a]A PubMed search performed on 14th July 2012 using the criteria (*Chiari* [*Title*]) *and Arnold* [*Title*]) *and spina bifida* revealed 42 publications. A search using the criteria (*Chiari* [*Title*]) *not Arnold* [*Title*]) *not Budd* [*Title*]) *and spina bifida* however revealed 44 publications

[b]A PubMed search performed on 14th July 2012 using the criteria (*Chiari* [*Title*]) *not Budd* [*Title*]), *not Arnold* [*Title*], revealed 1,422 publications. A search using the criteria *Chiari* [*Title*]) *and Arnold* [*Title*] revealed 509 publications. A search using the criteria *hindbrain herniation* [*Title*] revealed 22 publications

Dandy-Walker malformation. It was at one time widely assumed that the pathogenesis of communicating syringomyelia is due to a water-hammer effect of cerebrospinal fluid from the fourth ventricle, being forced into the central canal (Williams 1980; Gardner 1965). Indeed this theory is still accepted by some authors (Hagihara and Sakata 2007). However, this theory was challenged, not least because communicating syringomyelia is sometimes seen with distal spinal cord pathology, such as caudal, cervical, spinal and intradural subarachnoid cysts (Yamashita et al. 2012) and thoracic spinal cord tumours (Mock et al. 1990). Communicating hydrocephalus, however, is now known to be present in fewer than 10 % of affected human patients, so this categorisation now tends to be disregarded (Batzdorf 2001a).

Noncommunicating syringomyelia describes a cavity that has developed caudal to a syrinx-free

Table 20.6 Are the terms communicating and noncommunicating syringomyelia obsolete?

Yes
1. These terms originate from a time, before the advent of magnetic resonance imaging, when our understanding of the pathogenesis of syringomyelia was different. Nowadays most consider the water-hammer theory unlikely to be correct because a communication between the central canal and fourth ventricle is uncommon in humans
2. These terms have fallen out of use already[a]. If they were of value to clinicians, then they would still be in common usage
No
1. Communicating syringomyelia may have a different pathogenesis and management and therefore should be considered separately. The preferred management is addressing the hydrocephalus, typically with a intraventricular shunt

[a]Noncommunicating syringomyelia is reputed to account for 90 % of cases (Batzdorf 2001a). However, a PubMed search performed on the 15th July 2012 using the keywords 'noncommunicating syringomyelia' revealed only 19 articles out of a possible 3,958 articles found using the keyword syringomyelia (0.005 %)

portion of spinal cord and which is associated with obstruction of the cerebrospinal fluid pathways, either at the foramen magnum, such as in Chiari type I malformation, or more distally, for example, spinal arachnoid webs or adhesions (Milhorat et al. 2001). A classification by communicating versus noncommunicating once again becomes inconsistent with a condition such as Chiari type I malformation, which is more commonly associated with noncommunicating syringomyelia (Milhorat et al. 1995), but can also cause a communicating syringomyelia (Lena et al. 1992). Arguments as to the usefulness or otherwise of these terms are represented in Table 20.6.

20.4 Subarachnoid Channels

The term spinal subarachnoid space is widely used in neurological literature and refers, of course, to the channels along which CSF flows. The word space has several dictionary definitions, but, in common usage, it implies that there is nothing there, e.g. a space between words in this sentence and a space between buildings and outer space. In reality, these channels, within the spinal canal, are filled with cerebrospinal fluid. They are in continuity with the basal cisterns inside the skull and the cerebral ventricles. Together, these CSF-filled channels provide, amongst other things, an essential mediator of compliance, within and between the cranial and spinal compartments. As an incompressible physical medium, the CSF transmits pulsatile energy waves, both arterial and venous, and these must have important roles to play in the development of syringomyelia cavities. If cerebral arteries were suspended in an empty space, then the energy of their pulsations would not be transmitted beyond their points of origin. There is, therefore, an argument for using the term subarachnoid channels, in preference to subarachnoid space.

20.5 Veterinary Terminology

In veterinary medicine similar arguments about nomenclature have occurred (Cappello and Rusbridge 2007). It is debatable whether the term Chiari malformation should be applied to the dog. The analogous canine condition, characterised by decreased volume of the caudal fossa and caudal displacement of the caudal cerebellar vermis into or through the foramen magnum, is very similar to the human condition (see Chap. 14). The condition in the dog, however, is inconsistent with the original historical description of Chiari malformation, not in the least because dogs do not have cerebellar tonsils. In veterinary medicine there is resistance to using eponyms, but they are not without precedent, especially where the name is a simple term, used to describe a complicated process, for example, Horner's syndrome or Wallerian degeneration. It is considered more correct to use an anatomical description, and early reports of the canine condition make reference to occipital hypoplasia with syringomyelia (Rusbridge et al. 2005; Rusbridge and Knowler 2003), although this term was dropped because it was realised that the pathogenesis of

the condition was likely to be more complex than simply a hypoplastic occipital bone(s). The term caudal occipital malformation syndrome was popular in the USA (Dewey et al. 2005, 2007) and is still used occasionally,[3] but it, too, is inappropriate, for similar reasons. In addition, if using an anatomical description, it should be accurate and the term 'caudal occipital' actually refers to the supraoccipital bone. There is also a reluctance to use the term hindbrain herniation because some veterinary surgeons believe that this is an embryological term, referring to the rhombencephalon. However, there is no other term used to describe the contents of the caudal (posterior) cranial fossa, and the exact reason for the reluctance to use the term hindbrain is unclear, especially when the word forebrain is used commonly. Equally, hindbrain herniation may not be appropriate because in many dogs there is not a marked hindbrain herniation but, rather, an overcrowding in the most caudal region of caudal cranial fossa, similar to Chiari 0 (Table 20.3). Eventually, a meeting of international experts decided that the term 'Chiari-like malformation' should be used to describe the canine condition (Cappello and Rusbridge 2007).

20.6 Singular and Plural

We refer to the medical instrument used for injecting or withdrawing fluid as a syringe. We would order a box of syringes for ward supplies. Yet, we use the term syrinx to refer to the vocal organ of a bird and syrinxes when using the plural of this word. When speaking of fluid-filled cavities within the spinal cord, therefore, it would seem logical to refer to more than one syrinx as syrinxes. To say that the correct plural is 'syringes' implies that the correct singular word for a cystic cavity inside the spinal cord is a syringe. In truth, we are dealing with two different words, albeit with a common etymological origin. They have, correspondingly, different plurals.

Notwithstanding these arguments and noting that some journals refuse the use of syringes as the plural of syrinx, the editors of this monograph have allowed both terms to be used, at the preference of individual authors.

20.7 Eponyms, Acronyms and Abbreviations

A large number of medical terms have their origins in the Latin or Greek languages. Unfortunately, teaching of these languages no longer forms part of a mainstream education. As a result, many new medical terms, introduced throughout the twentieth century and beyond, have taken the form of acronyms. Not uncommonly, such acronyms are uttered by individuals who may not know what the abbreviation actually stands for, even though the meaning of the expression may be clear, at least to those who use it. The abbreviation CSF, for example, will not need to be explained to most readers of this book but, when used for the first time in speaking to a patient, may produce a look of puzzlement.

Less widely used acronyms do require explanation at the outset, a convention demanded by most publishers of medical literature. A danger exists, however, that too many acronyms or abbreviations are introduced into a specialist article or book chapter. Such a practice may make it easier for the author to write the work, but it will very likely make it more difficult for the reader to understand. If the reader has to keep referring back a few pages, to be reminded of the meaning of a given acronym or abbreviation, then absorbing the meaning of the whole composition becomes more difficult. We have attempted to restrict this practice in this monograph, preferring to write expressions in full.

Patients do seem to like medical acronyms, presumably because they are easier to say than many medical tongue-twisters, such as

[3] A PubMed search performed on 14th July 2012 using the criteria (*Chiari* [*Title*]) *not Budd* [*Title*]) *and dog* revealed 25 publications. A search using the criteria *occipital hypoplasia* [*Title*]) *and dog* revealed 3 publications. A search using the criteria *caudal occipital malformation syndrome* [*Title*] revealed 2 publications.

Table 20.7 Abbreviations and acronyms

Acronym	Full expression
ACM	Arnold-Chiari malformation
CM	Chiari malformation
CM0	Chiari malformation type 0
CM1/CMI	Chiari malformation type I
CM2/CMII	Chiari malformation type II
CVD	Craniovertebral decompression
FMD	Foramen magnum decompression
HBH	Hindbrain hernia
PTSM	Post-traumatic syringomyelia
SCI	Spinal cord injury
SM	Syringomyelia

syringomyelia. Unfortunately there is not a great deal of consistency in the use of such terms (Table 20.7).

A curious, modern approach is to give peculiar names to newly discovered genes and proteins. An example is Sonic Hedgehog (Chap. 4), a name taken from a popular computer game of its time and given to a family of proteins involved in the development of the nervous system. This practice must reflect the fact that the modern research scientist is exposed to such recreational activities, whereas earlier generations of biologists might only have been exposed to their Latin and Greek studies.

The adoption of eponyms has been widespread in medicine for a long time. Such words usually honour the name of the person who first identified a condition or at least described it in any detail. Sometimes the name derives, instead, from a place or area where a disease was first discovered. Occasionally the name refers to a patient who suffered from the condition. The disadvantage of most eponyms is that they tell us little, if anything, about the nature of the condition to which they apply. The World Health Organisation prefers that eponyms be avoided, in favour of more descriptive terms. Thus, hindbrain hernia might be preferable to Chiari malformation. Unfortunately, common usage tends to resist such initiatives, and eponyms are likely to remain in widespread use in medical practice. For example, is it likely that we will abandon the eponym Parkinson's disease and readopt the expression paralysis agitans?

References

Arnold J (1894) Myelocyste, Transposition von Gewebskeimen und Sympodie. Beitr Pathol Anat 16:1–28

Batzdorf U (2001a) A brief history of syringomyelia. In: Tamaki N, Batzdorf U, Nagashima T (eds) Syringomyelia: current concepts in pathogenesis and management. Springer, Tokyo, pp 3–9

Batzdorf U (2001b) Treatment of syringomyelia association with Chiari I malformation. In: Tamaki N, Batzdorf U, Nagashima T (eds) Syringomyelia current concepts in pathogenesis and management. Springer, Tokyo, pp 121–135

Bejjani GK (2001) Definition of the adult Chiari malformation: a brief historical overview. Neurosurg Focus 11(1):E1. doi:10.3171/foc.2001.11.1.2

Cappello R, Rusbridge C (2007) Report from the Chiari-Like Malformation and Syringomyelia Working Group round table. Vet Surg 36(5):509–512. doi:10.1111/j.1532-950X.2007.00298.x

Castillo M, Quencer RM, Dominguez R (1992) Chiari III malformation: imaging features. AJNR Am J Neuroradiol 13(1):107–113

Chiari H (1888) Ueber die Pathogenese der Sogenannten Syringomyelie. Z Heilk 9:307

Chiari H (1891) Ueber Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns. Dtsch Med Wochenschir 42:1172–1175

Chiari H (1896) Ueber Veränderungen des Kleinhirns, des Pons and der medulla oblongata in Folge von genitaler Hydrocephalie des Grosshirns. Denkschr Akad Wiss Wien 63:71–116

Cleland J (1883) Contribution to the study of spina bifida, encephalocele, and anencephalus. J Anat Physiol 17:257–292

Cruveilhier J (1829) L'Anatomie pathologique du corps humain; descriptions avec figures lithographiées et coloriées; diverses alterations morbides don't le corps humain et susceptible. Bailliere, Paris

Dewey CW, Berg JM, Barone G et al (2005) Foramen magnum decompression for treatment of caudal occipital malformation syndrome in dogs. J Am Vet Med Assoc 227(8):1270–1275, 50–51

Dewey CW, Marino DJ, Bailey KS et al (2007) Foramen magnum decompression with cranioplasty for treatment of caudal occipital malformation syndrome in dogs. Vet Surg 36(5):406–415. doi:10.1111/j.1532-950X.2007.00286.x

Gardner WJ (1965) Hydrodynamic mechanism of syringomyelia: its relationship to myelocele. J Neurol Neurosurg Psychiatry 28:247–259

Geerdink N, van der Vliet T, Rotteveel JJ et al (2012) Interobserver reliability and diagnostic performance of Chiari II malformation measures in MR imaging-part 2. Childs Nerv Syst 28(7):987–995. doi:10.1007/s00381-012-1763-3

Grossmann S, Maeder IM, Dollfus P (2006) 'Treatise on the Spinal Marrow and its Diseases' (Anatomy,

functions and general considerations on its diseases) by Ollivier d' Angers CP (1796–1845). Spinal Cord 44(12):700–707. doi:10.1038/sj.sc.3101956

Hagihara N, Sakata S (2007) Disproportionately large communicating fourth ventricle with syringomyelia: case report. Neurol Med Chir (Tokyo) 47(6):278–281

Hogg J, Peterson A, El-Kadi H (1998) Imaging of cranial and spinal cerebrospinal fluid collections cerebrospinal fluid collections USA: the American Association of Neurological Surgeons Publications Group. Stuttgart, Germany and New York, USA: Thieme Publishing Group

Hu HZ, Rusbridge C, Constantino-Casas F et al (2011) Histopathological investigation of syringomyelia in the Cavalier King Charles spaniel. J Comp Pathol 146(2–3):192–201. doi:10.1016/j.jcpa.2011.07.002

Jinkins JR, Sener RN (1999) Idiopathic localized hydromyelia: dilatation of the central canal of the spinal cord of probable congenital origin. J Comput Assist Tomogr 23(3):351–353

Kahler O, Pick A (1879) Beitrag zur Lehre von der Syringo-und Hydromyelia. Vjschr Prakt Heilkd 142:20–41

Knowler SP, McFadyen AK, Rusbridge C (2011) Effectiveness of breeding guidelines for reducing the prevalence of syringomyelia. Vet Rec 169(26):681. doi:10.1136/vr.100062

Lena G, Boudawara Z, Genitori L et al (1992) 14 cases of communicating syringomyelia associated with Chiari I malformation in children. Neurochirurgie 38(5): 297–303

Leyden E (1876) Über Hydromyelus un Syringomyelie. Arch Pathol Anat Physiol 68:1–20

Markunas CA, Tubbs RS, Moftakhar R et al (2012) Clinical, radiological, and genetic similarities between patients with Chiari type I and type 0 malformations. J Neurosurg Pediatr 9(4):372–378. doi:10.3171/2011.12.PEDS11113

Milhorat TH, Capocelli AL Jr, Anzil AP et al (1995) Pathological basis of spinal cord cavitation in syringomyelia: analysis of 105 autopsy cases. J Neurosurg 82(5):802–812. doi:10.3171/jns.1995.82.5.0802

Milhorat TH, Chou MW, Trinidad EM et al (1999) Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. Neurosurgery 44(5):1005–1017

Milhorat T, Fox A, Todor DR (2001) Pathology, classification, and treatment of syringomyelia. In: Tamaki N, Batzdorf U, Nagashima T (eds) Syringomyelia: current concepts in pathogenesis and management. Springer, Tokyo, pp 10–30

Mock A, Levi A, Drake JM (1990) Spinal hemangioblastoma, syrinx, and hydrocephalus in a two-year-old child. Neurosurgery 27(5):799–802

Mortazavi MM, Rompala OJ, Verma K et al (2011) Charles Prosper Ollivier d'Angers (1796–1845) and his contributions to defining syringomyelia. Childs Nerv Syst 27(12):2155–2158. doi:10.1007/s00381-011-1416-y

Newton EJ (1969) Syringomyelia as a manifestation of defective fourth ventricular drainage. Ann R Coll Surg Engl 44(4):194–213

Ollivier d'Angers C-P (1824) De la moelle épinière et de ses maladies. Crevot, Paris

Ollivier d'Angers C-P (1827) Traité des maladies de la moelle épinière contenant l'histoire anatomique, physiologique et pathologique de ce centre nerveux chez l'homme. Crevot, Paris

Pearce JM (2000) Historical note. Arnold Chiari, or "Cruveilhier Cleland Chiari" malformation. J Neurol Neurosurg Psychiatry 68(1):13

Radojicic M, Nistor G, Keirstead HS (2007) Ascending central canal dilation and progressive ependymal disruption in a contusion model of rodent chronic spinal cord injury. BMC Neurol 7:30. doi:10.1186/1471-2377-7-30

Rusbridge C, Knowler SP (2003) Hereditary aspects of occipital bone hypoplasia and syringomyelia (Chiari type I malformation) in cavalier King Charles spaniels. Vet Rec 153(4):107–112

Rusbridge C, Knowler P, Rouleau GA et al (2005) Inherited occipital hypoplasia/syringomyelia in the cavalier King Charles spaniel: experiences in setting up a worldwide DNA collection. J Hered 96(7): 745–749. doi:10.1093/jhered/esi074

Sarnat HB (2007) Semantics do matter! Precision in scientific communication in pediatric neurology. J Child Neurol 22(11):1245–1251. doi:10.1177/0883073807307981

Schuppel (1865) Über Hydromyelus. Arch Heilk 6:289

Schwalbe E, Gredig M (1907) Ueber Entwicklungsströrungen des Kleihirns, Hirnstamms und Halsmarks bei Spina bifida (Arnold'sche und Chiari'sche Missbildung). Beitr Pathol Anat 40:132–194

Simon T (1875) Über Syringomyelia und Geshwulstbildung im Rückenmark. Arch Psychiatr NervenKr 5:120–163

Solt I (2011) Chiari malformation eponym- time for historical justice. Ultrasound Obstet Gynecol 37(2):250–251. doi:10.1002/uog.8876

Tubbs RS, Iskandar BJ, Bartolucci AA et al (2004) A critical analysis of the Chiari 1.5 malformation. J Neurosurg 101(2 Suppl):179–183. doi:10.3171/ped.2004.101.2.0179

Walusinski O (2012) Charles-Prosper Ollivier d'Angers (1796–1845). J Neurol 259(6):1255–1256. doi:10.1007/s00415-012-6424-7

Williams B (1980) On the pathogenesis of syringomyelia: a review. J R Soc Med 73(11):798–806

Yamashita T, Hiramatsu H, Kitahama Y et al (2012) Disproportionately large communicating fourth ventricle associated with syringomyelia and intradural arachnoid cyst in the spinal cord successfully treated with additional shunting. Case report. Neurol Med Chir (Tokyo) 52(4):231–234