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Syringomyelia: a practical, clinical concept for classification

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Abstract
Background The term syringomyelia describes many pathogenetically different disorders, and a variety of attempts to group these based on different criteria have been proposed in the literature. As a consequence a lack of consensus regarding classification and terminology exists. This inconsistency extends to the ICD-10 classification of diseases in regards to syringomyelia (G95.0) and hydromyelia (Q06.4). We propose a new unifying concept for classification that also incorporates diagnostics and treatment.
Methods The PubMed online database was used to gain a general overview of the existing pathogenetic theories in relation to syringomyelia. Illustrative cases at our department were included and similar cases of the literature were found using the PubMed database. All material was reviewed with main focus on the classification and terminology used.
Results Despite syringomyelia (G95.0) and hydromyelia (Q06.4) existing as independent ICD-10 entities, we have shown that the use of classifying terminology for fluid-filled cavities in the spinal cord is indiscriminate and inconsistent. Even though a general agreement on the believed pathogenetic mechanism exists, and the general treatment methods are used in accordance with this mechanism, the terminology fails to function as a simple and universal link between theory and treatment.
Conclusions We propose a new causal concept for an ICD classification with syringomyelia (G95.0) as the only describing terminology, thus abandoning the use of hydromyelia (Q06.4). Syringomyelia is divided into five subgroups according to the associated pathologies. The classification is based on applied diagnostics and serves as a clinical guidance for treatment.

Keywords Syringomyelia · Hydromyelia · Syrinx · Classification · Treatment · Diagnostics

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Introduction

The term syringomyelia describes many pathogenetically different disorders, and a variety of attempts to group these based on different criteria have been proposed in the literature. A popular broad grouping is a classification based on the assumed pathogenesis and association with other disorders [1–3]. Other proposed classifications are based on syrinx fluid composition, central canal communication between syrinx and the fourth ventricle, or the microanatomical localisation of the syrinx [4–7].

A given terminology is usually derived from the believed underlying pathogenetic theory, which in turn determines the choice of treatment. Even when restricting the subject to hindbrain-related syringomyelia, very different pathogenetic theories have been proposed—and when the same terms are used indiscriminately between different sets of theories, a terminological confusion is created. The inconsistency extends to the ICD-10 classification of diseases in which syringomyelia (G95.0) and hydromyelia (Q06.4) exist as two

separate entities despite the lack of consensus in definition. As a consequence, a selection of treatment methods have been used over the years, e.g. syringostomy, syringosubarachnoidal shunting, syringopleural shunting, syringoperitoneal shunting, sectioning of the filum terminale (terminal ventriculostomy), foramen magnum decompression with/without obex plugging and/or duraplasty, and percutaneous syrinx aspiration. All of these treatment methods have had varying outcome success [8].

The purpose of this paper is to give a thorough review of the major theories in this subject and, furthermore, the final aim is to investigate whether theory, terminology and treatment purposefully and meaningfully serve each other both in the literature and the clinic.

Methods and materials

The PubMed online database was used to gain a general overview of the existing pathogenetic theories in relation to "syringomyelia" and "hydromyelia". PubMed was also used to match syringomyelia cases with relevance to our own clinical cases.

Our own cases were found by searching the internal patient database at the Department of Neurosurgery at Copenhagen University Hospital, Rigshospitalet, over a 5-year period from 2005 to 2010. Informed consent from the included cases was obtained. ICD-10 codes of relevant disorders were combined with either syringomyelia (G95.0) or hydromyelia (Q06.4).

Results

Over the years, the multifarious nature of syringomyelia has caused a need to classify the disorder into smaller and more homogeneous groups. Based on pathogenesis it has been proposed to divide the disorder into three subgroups, which all fit under the term syringomyelia [3]:

1. Syringomyelia as a result of changed cerebrospinal fluid (CSF) flow dynamics related to hindbrain disorders, e.g. Chiari malformation, Dandy-Walker Syndrome, arachnoiditis or osseous abnormalities.
2. Syringomyelia as a result of intramedullary tissue damage caused by haemorrhage or infarction.
3. Syringomyelia as a result of intramedullary tumour with secretory capabilities.

Group 1, which forms the core for this review, accounts for more than 70 % of all cases of syringomyelia and thus makes up the largest of the syringomyelia entities [3]. Most of the effort to uncover the exact pathogenetic mechanism has focused on this relationship. Since syringomyelia in this group is associated with disorders that all change the conditions in the subarachnoidal space (SAS), a CSF-related hydrodynamic way of thinking has been applied to describe the pathogenesis. In support of this concept, the syrinxes in this group frequently contain fluid with a composition similar to CSF. This is unlike the two other groups, which show a varied composition of syrinx contents, including haemorrhagic or other proteinaceous fluids [4, 5].

Theory and terminology

The proposed theories for the pathogenesis of syringomyelia associated with hindbrain disorders (above-mentioned group 1) can be divided into three subgroups based on how the syrinx fluid is believed to have infiltrated the spinal cord. We suggest the following designations (Fig. 1 and Table 1):

1. Classic communicating
2. Transmedullary infiltration
3. Parenchymal formation

Table 1 shows a summary of the theories and the scientific background of their origin. It is noticed that different theoretical schools each represent an era. The classic communicating theories originate in an age where contrast X-ray and intrathecal pressure measurements were state of the art. The theories included in the transmedullary infiltration concept are born and cultivated when more advanced imaging techniques like intraoperative ultrasound and cine-magnetic resonance imaging (MRI) gain a footing. The latest theories are founded on parenchymal formation and are mainly based on studies of the literature and mathematical modelling.

Classic communicating

This theoretical subgroup is the oldest, and the name originates from the belief that a syrinx formation is due to an anatomical communication between the syrinx and the fourth ventricle. Gardner is instigator of this school. His original theory builds on a missing embryonic opening of the foramina of the fourth ventricle resulting in a dilated central canal in the spinal cord. The closed foramina direct an intraventricular CSF pulse wave created by the systolic blood filling of the choroid plexus towards the obex of the central canal, where the systolic "water hammer" effect and herniating structures through foramen magnum act as a one-way pump and valve. Gardner designates this dilation hydromyelia, like a spinal analogue of hydrocephalus. In comparison, syringomyelia describes a condition where a dilation of the central canal (hydromyelia) secondarily ruptures the ependyma and spreads as a diverticulum into the parenchyma parallel to the central

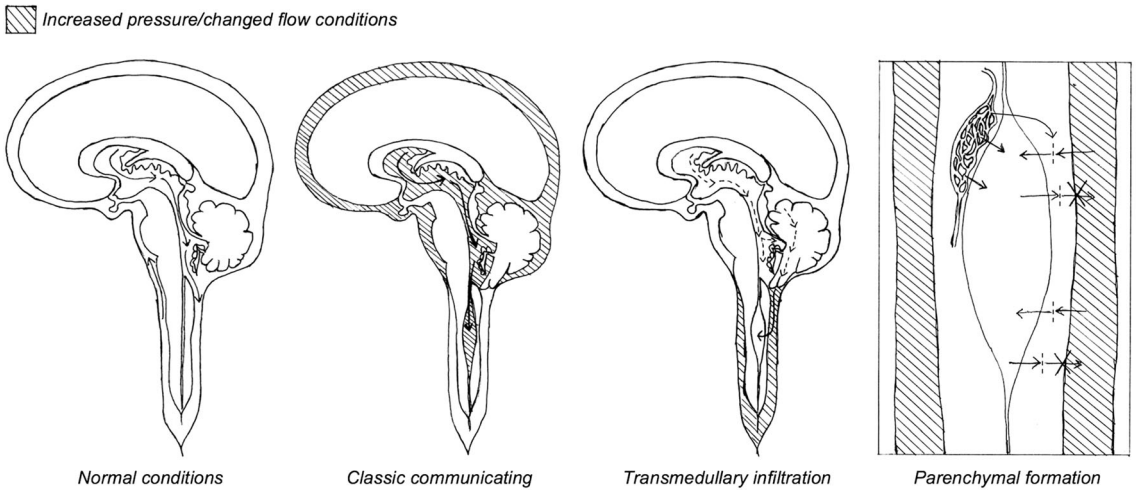

Fig. 1 Illustration of the believed methods of spinal cord fluid infiltration in the different theoretic subgroups. The *arrows* represent the direction of fluid flow

canal. This diverticulum/rupture into the tissue is termed a syrinx [4, 9]. According to Gardner Chiari malformation and Dandy-Walker Syndrome are directly caused by the foraminal atresia, and both conditions are individually responsible for sustaining a foraminal obstruction leading to hydromyelia and syringomylia.

Williams also supports the idea of a pulse wave forcing CSF through the central canal and he uses the subdividing term "communicating syringomyelia" with reference to the believed anatomical communication between the cystic spinal cord dilation and the fourth ventricle—regardless of whether the pathology is a dilatation of the central canal alone (hydromyelia) or whether it is a secondary rupture into the parenchyma [7]. A dilation spreading into the parenchyma is termed a syrinx, and syringomyelia designates a condition with longitudinal cavities in the spinal cord [7, 10].

Williams describes several phenomena contributing to the pathogenesis of syringomyelia, and, in contrast to Gardner, he does not consider the systolic pulse wave of a magnitude capable of explaining the formation of a spinal cavity. According to Williams, Valsalva-like manoeuvres (coughing, sneezing, etc.) produce an intracranial venous congestion, resulting in a spinal displacement of CSF due to the Monro-Kellie doctrine and a higher subarachnoidal compliance spinally. Patho-anatomical conditions, e.g. Chiari malformation, prevents the normal physiological displacement of CSF into the spinal SAS, for which reason it is forced via a "non-physiological" route through the obex into the central canal [7]. Furthermore, if there are structures blocking the foramen magnum, a craniospinal pressure dissociation is produced at the end of a Valsalva manoeuvre, where intracranial pressure (ICP) is still high and spinal SAS pressure has returned to normal. Due to the foramen magnum obstruction, CSF has nowhere else to go but into the central canal assisted by the lower spinal pressure, a phenomenon designated "suck" by Williams. Contributing to the maintenance of a syrinx is longitudinal fluid motions inside the cavity itself, e.g. during coughing, which Williams terms "slosh" [10].

Contrary to Williams, du Boulay supports Gardner's hypothesis of a systolic pulse wave, the only problem being syrinx formation without atresia of the fourth ventricle's foramina. Instead of only one pulse wave constituting the "water hammer", du Boulay advocates for two successive intracranial CSF pulse waves, a ventricular one and a cisternal one. The former corresponds to Gardner's systolic pulse wave and the latter is started by the first pulse wave exiting the foramina of the fourth ventricle. Under normal anatomical conditions, this secondary cisternal CSF pulse wave propagates into the spinal SAS. If a Chiari malformation obstructs the foramen magnum, CSF is forced via the fourth ventricle into the central canal. Furthermore during each systole the cerebellar tonsils function as a one-way valve, milking CSF unidirectionally in cranial to caudal direction through the narrow cranial part of the central canal. Du Boulay uses the term syrinx as description of the cystic CSF dilation in the spinal cord and syringomyelia designates the clinical syndrome. Hydromyelia is used synonymously with syringomyelia [11].

Transmedullary infiltration

As a deeper understanding of the diversity of syringomyelia as a disease complex evolved, it became clear that an anatomical communication between the syrinx and the fourth ventricle only exists in a minority of the cases—a fact that even the originator of the term "communicating syringomyelia", Williams, later acknowledged [3, 8, 12]. Ball and Dayan [13] used mathematical calculations to reject the hypothesis of a CSF pulse wave being of a magnitude capable of forming a syrinx by entering an open central canal. Instead, they proposed a theory based on the fact that the syrinx wall frequently

Table 1 Pathogenetic theories

Group	Authors, year	Method	Terminology	Treatment
Classic communicating	Gardner and Angel, 1958; Gardner, 1965	X-ray with contrast autopsy study	Hydromyelia: *pure dilation of the central canal, condition with a pure dilation of the central canal* Hydrocephalomyelia: *embryonic dilation of ventricles and the central canal* Syringobulbia: *cavity originating from the fourth ventricle but not communicating* via *the central canal* Syringomyeli: *condition with diverticulum from the central canal* Syrinx: *diverticulum from the central canal* True syrinx: *diverticulum from and parallel to the central canal totally lacking ependyma*	FMD with open dura (or synthetic duraplasty) + opening of foramen of Magendie + obex plugging + C1/C2 laminectomy
	Williams, 1969–1993[a]	Original theory: X-ray with contrast Pneumoencephalography Pressure measurement Modified theory: CT with contrast MRI Ultrasound Myelography	Classical syringomyelia: *as "communicating syringomyelia"* Communicating syringomyelia: *syrinx communicating with fourth ventricle* Hydromyelia: *dilation of the central canal with intact ependyma, included under "communicating syringomyelia"* Syringobulbia: *syrinx in the brainstem* Syringohydromyelia: *as "communicating syringomyelia"* Syringomyelia: *general term used in relation to pathogenetic different cystic formations in the spinal cord* Syrinx: *cavity in the spinal cord*	Primarily treatment of hydrocephalus by ventricular shunting, secondarily FMD with dissection of the arachnoid (and cerebellar tonsils if necessary), preferably no duraplasty, direct syrinx shunting only makeshift solution and preferably extrathecally then
	du Boulay et al., 1974	X-ray with contrast in monkeys Myelography Ventriculography Angiography	Hydromyelia: *as syringomyelia* Syringomyelia: *clinical syndrome with cystic formation in the spinal cord* Syrinx: *cyst in the spinal cord*	No treatment method mentioned
Transmedullary infiltration	Ball and Dayan, 1972	X-ray with contrast Autopsy study Mathematical model Intraoperative ultrasound	Communicating syringomyelia: *syrinx communicating with fourth ventricle* Syringobulbia: *syrinx in the brainstem* Syringomyelia: *syrinx in the spinal cord* Syrinx: *cavity*	No treatment method mentioned

Table 1 (continued)

Group	Authors, year	Method	Terminology	Treatment
	Oldfield et al., 1994		Communicating syringomyelia: *All CSF containing syrinxes, regardless of CSF infiltration route*	FMD with suboccipital craniectomy + C1/C2 laminectomy + dural graft, if no communication between the syrinx and the fourth ventricle is demonstrable further intra-arachnoid surgery (e.g. syringostomy) is unnecessary
		Phase-contrast MRI	Syringomyelia	
		Phase-contrast cine-MRI	Syrinx	
		Myelography		
		Ventriculography		
	Heiss et al., 1999	MRI	Syringomyelia: *condition with cystic formation in the spinal cord*	Suboccipital craniectomy + C1/C2 laminectomy + duraplasty with intact arachnoid, in case of heavy arachnoiditis dissection of this, syrinx shunting only makeshift solution
		Phase-contrast cine-MRI	Syrinx	
		Pressure measurement		
		Cardiac-gated ultrasound		
Parenchymal formation	Klekamp, 2002	Study of the literature	Hydromyelia Syringomyelia: *cystic cavity in the spinal cord containing ECF or CSF* Syrinx: tubular cavity	"Improvement of CSF flow", untethering
	Levine, 2004	Study of the literature	Hydromyelia: *syrinx communicating with fourth ventricle, intact ependyma, associated with hydrocephalus*	Primarily FMD without obex plugging or opening of foramen of Magendie, dissection of the arachnoid only in case of heavy arachnoiditis, alternatively syrinx shunting, in case of hydrocephalus also ventricular shunting, intermittent cervical halter traction
		Mathematical model	Syringobulbia Syringomyelia: *syrinx not communicating with fourth ventricle, ruptured ependyma* Syrinx: *tubular cavity in the spinal cord*	
	Koyanagi and Houkin, 2010	Study of the literature	Syringomyelia Syrinx	Primarily FMD, alternatively syringosubarachnoidal shunting

CSF cerebrospinal fluid, *ECF* extracellular fluid, *FMD* foramen magnum decompression

[a] Williams modified and developed his theory over the years

exhibits a great amount of small arteries and veins with a hyperplastic adventitia. A Valsalva-like manoeuvre performed by a patient with a foramen magnum obstruction, e.g. Chiari malformation, causes the spinal subarachnoidal pressure to rise due to congestion of the epidural venous plexus with no possibility of craniospinal pressure equalisation. The increased pressure forces CSF into the spinal parenchyma via the perivascular space (Virchow-Robin space), which becomes dilated, and by accumulation of CSF from these perivascular entrances to the spinal parenchyma a syrinx is formed. The syrinx may only secondarily rupture the ependyma, thus dilating the rudimentary central canal. Ball and Dayan use the term syrinx as the description of a cavity, while syringomyelia designates this as being located in the spinal cord. The term "communicating syringomyelia" is also used and refers to Williams' anatomical definition with communication between the syrinx and the fourth ventricle.

Oldfield uses the term "communicating syringomyelia" in a much broader sense than Williams. According to Oldfield, the term does not refer solely to syrinxes with a major anatomical communication to the fourth ventricle via the central canal, but instead describes all cavities containing CSF, no matter what the entrance route of CSF might be [8]. His theory is built on the same principle as Ball and Dayan's hypothesis with transmedullary CSF infiltration through the perivascular space, but Oldfield does not consider varying venous conditions responsible for the force exerted externally on the spinal cord. Instead Oldfield states that the regular systolic congestion of the brain creates a cranial CSF pulse wave that because of foramen magnum obstruction, e.g. Chiari malformation, is prevented from dissipating into the larger and more compliant spinal SAS. As a result, the force of the wave is transferred to the obstructing structures in the foramen magnum, e.g. the cerebellar tonsils, which thus are rhythmically compressed caudally in a piston-like motion creating a spinal CSF pulse wave that in time will force CSF into the parenchyma in the same fashion advanced by Ball and Dayan [8, 13]. Furthermore, the syrinx fluid will be forced to move longitudinally inside the spinal cord, similar to Williams' "slosh"-effect. Oldfield's theory can be viewed as a combination of the classic communicating theories by Gardner, Williams and du Boulay, and Ball and Dayan's transmedullary theory [4, 10, 11, 13].

A study by Heiss, including cine-MRI, pressure measurements and others, supports Oldfield's theory and, amongst other things, it shows that compliance of the spinal SAS is significantly reduced in patients with Chiari malformation compared with normal controls [12]. This supports that changed flow and pressure conditions in proximity to foramen magnum could exert a potent effect on the spinal cord at patho-anatomical circumstances. Heiss uses the term syringomyelia to describe a condition with a cystic formation in the spinal cord.

Parenchymal formation

Studies simultaneously measuring the pressure of the syrinx and the spinal SAS have shown an equal or even higher pressure in the syrinx [2, 14, 15]. This result contradicts the transmedullary theoretical movement and several authors have attempted to advance theories dealing with conflicting facts like this.

The latest trend in pathogenetic thinking is that spinal fluid-filled cavities are primarily the result of blood plasma ultrafiltration rather than CSF being pushed or sucked into the spinal parenchyma or central canal. These theories are all primarily based on thorough studies of the literature and theoretical mathematical modelling, thus in principle they are not directly based on new clinical or experimental data.

Klekamp suggests that a syrinx forms because of a changed equilibrium between CSF and the intramedullary extracellular fluid (ECF). According to Klekamp, a balanced exchange between CSF and ECF takes place through the perivascular space in the spinal cord under normal conditions. In this way the theory tries to bridge the concepts of transmedullary and parenchymal fluid formation. Klekamp's theory must be classified as parenchymal since he states that it is ECF and not CSF that accumulates in the formation of syringomyelia, even though such a division of the fluid origin is rather arbitrary due to the proposed communication between the two. An argument could be that CSF is unable to progressively infiltrate the spinal cord against the higher syrinx pressure. Conditions favouring a medullary flow of fluid, such as an obstruction of CSF flow caused by Chiari malformation or a tethered cord causing changed spinal cord movements, either of which resulting in increased CSF flow resistance, will in time lead to syrinx formation. An interstitial oedema is created and, depending on local flow resistance, ECF accumulates in the parenchyma or the central canal. Klekamp uses syrinx to describe a tubular cavity, and designates the condition syringomyelia, defined as a cystic cavity in the spinal cord containing fluid either identical to or being similar to CSF or ECF, having ependymal lining or not. An aggravation of the condition is believed to happen by the "slosh" effect [2].

According to Levine's theory, syringomyelia is a tubular cavity in the spinal cord not communicating with the fourth ventricle (non-communicating) and, most frequently, spreading out in the parenchyma, thereby not being fully covered by ependyma. The term syrinx is similarly used to describe a cavity in the spinal cord. Hydromyelia, on the other hand, designates a cavity in open connection with the fourth ventricle (communicating) and is, strictly speaking, just a dilation of the central canal with intact ependyma and, most frequently, it is explainable by a simultaneous hydrocephalic condition. Levine advances a pathogenetic theory that is built on craniospinal pressure dissociation in the SAS caused by a patho-anatomical blocking of the foramen magnum. CSF in

the SAS and the blood in the venous system of the neuroaxis can under normal conditions be considered as two columns of fluid in hydrostatic equilibrium. In the case of the foramen magnum being obstructed by a Chiari malformation, the CSF fluid column is divided in two, while the venous column still represents one continuous unit. Thus, pressure dissociation between the SAS and the venous system is created that also greatly influences the transmural pressure of the venous system. Cranially to the foraminal blocking, this tends to compress the venous microcirculation, while caudally the consequence is venous dilation. The size of the transmural pressure displacement varies with physical activity and causes mechanical stress on the vessels, which eventually destroys the blood-spinal cord barrier resulting in the leakage of an ultrafiltrate of the blood, eventually forming a syrinx [15].

Koyanagi and Houkin's theory is based on a description of the posterior spinal veins, which lacking a pial covering are situated directly in the SAS [14]. As the subarachnoidal compliance is reduced in case of Chiari malformation, compliance of these veins is reduced as well. The posterior spinal veins are of great importance to the diastolic return of blood to the heart, but due to the reduced compliance the vessel dilation and thus the return of the blood are impeded. As a result, Starling's equilibrium is disrupted and an intramedullary oedema, a syrinx, is formed, either in the parenchyma or in the central canal. Koyanagi and Houkin designate all kind of cavities/syrinxes in the spinal cord syringomyelia, regardless of communication with the fourth ventricle or degree of ependymal lining. Based on studies showing that tracer and contrast substances introduced into the subarachnoidal CSF can be found later in the medullary tissue, a possible exchange between the subarachnoidal CSF, medullary ECF and syrinx fluid is also suggested.

Illustrative cases

Our cases from the Department of Neurosurgery at Copenhagen University Hospital, Rigshospitalet, Copenhagen are summarised in Table 2. All chosen four cases had a hindbrain disorder in addition to their syringomyelia. The hindbrain disorders were: Dandy-Walker cyst or other cystic dilation of the fourth ventricle (cases 1, 3 and 4) and Chiari malformation (cases 2 and 4). Thus, one out of the four cases had more than one hindbrain disorder (case 4). The three patients with cystic malformations in the posterior fossa all presented with supratentorial hydrocephalus (cases 1, 3 and 4). Our cases illustrate the complexity, varied terminology and multifarious treatment methods.

We matched our own cases with cases from the literature in order to compare terminology and treatment (Table 3). Similar to our own cases, all chosen cases from the literature had a hindbrain disorder in addition to syringomyelia.

Treatment methods

As abnormal CSF flow across the foramen magnum is common to all theories, the general aim of treatment is to establish normal CSF flow exactly here. The proposed treatment methods are summarised in Tables 1, 2 and 3. They may be divided into:

1. Treatment of associated supratentorial hydrocephalus
   - Shunting/shunt revision (cases 1 and 3) [16]
   - Endoscopic third ventriculostomy [17]
2. Decompression of hindbrain disorder
   - Foramen magnum decompression (case 2) [18, 19]
   - Shunting/shunt revision/endoscopic fenestration of cystic fourth ventricle or cyst in posterior fossa (cases 3 and 4) [19–21]
3. Direct decompression of syrinx
   - Direct syrinx shunting (case 3)
   - Removal/decompression of spinal subarachnoidal obstruction (cases 1 and 3)

Logically, decompression of the foramen magnum is directly aimed at the primary pathology, and is recommended by most authors [8–10, 12, 14, 15]. The bony decompression consists of suboccipital craniectomy with or without upper cervical laminectomy. This may include one or more of the following procedures to further improve CSF flow: duraplasty, opening of the arachnoid, widening of the foramen of Magendie or tonsillar resection. Obex plugging has been abandoned as a remedy to prevent the "water hammer" effect. If hydrocephalus coexists, endoscopic third ventriculostomy or ventricular shunting could be the first-line treatment. In these cases, decompression of the ventricular system could remove the downward pressure towards the foramen magnum and thus indirectly disimpacts the cerebellar tonsils (Fig. 2) [10, 15]. A syrinx may coexist with spinal pathology without hindbrain abnormality. In these cases the syrinx may resolve by surgical correction of the spinal disease, e.g. by untethering. If this is inefficient, additional syrinx shunting may be employed [22, 23]. Direct syrinx shunting (subarachnoidal or extrathecal) is considered a secondary solution, or at best an alternative to decompression, since the CSF flow obstruction is not treated [10, 12, 14, 15].

Terminology

In the medical records of our own cases it was generally observed that the word *syrinx* consequently was

Table 2 Our cases

Case	Gender	Present age	Aetiology	Terminology	Treatment	Result
1	F	14 years	DWS Hydrocephalus Syringomyelia (holocord)	Hydromyelia Syringomyelia Syrinx	Revision of VP shunt	Improvement of headache and physical strength in the legs after a couple of months, unchanged syrinx size
					Untethering of spinal cord, removal of lumbosacral lipoma, lowering of valve pressure	General improvement, syrinx is considerably collapsed after 5 months
2	F	15 years	CM Syringomyelia (almost holocord)	Hydromyelia Syrinx formation	FMD with duraplasty	General improvement of extremity symptoms after 3 months, unchanged syrinx size
3	F	15 years	Cystic fourth ventricle	Arachnoid cyst	Decompression of syrinx	General improvement
			Hydrocephalus	Cyst		
			Spina bifida Syringomyelia (Th1–Th7)	Cystic formation Cystic dilation Hydromyelia	Syringosubarachnoidal shunting and decompression of syrinx	No specific information
				Intradural cyst Liquor cyst	Spinal cord untethering	Improvement of spasticity, worsening of bladder problems
				Spinal cyst		
				Syringomyelia Syrinx	Revision of syringosubarachnoidal shunt	Pain reduced
				Syrinx formation	Shunting of cystic fourth ventricle, decompression of syrinx	Immediate improvement of symptoms, but later relapse
					Decompression of syrinx	No apparent effect
					Osseous cervical stabilisation	Pain reduced
					Revision of hydrocephalus shunt system	Smaller syrinx, but in general aggravated neurofunctions
4	F	24 years	CM DWS Hydrocephalus Syringomyelia (obex-Th3)	Cervical syringobulbia Syrinx Syrinx formation	Revision of posterior fossa cyst shunt	After 2 months significant reduction of syrinx size, improvement of gait after 6 months, continued narrow and unchanged syrinx 2 years later, patient doing well

CM Chiari malformation, *DWS* Dandy-Walker syndrome, *ETV* endoscopic third ventriculostomy, *FMD* foramen magnum decompression

used to describe a cavity or a cyst in the spinal cord and, furthermore, that *syrinx* and *cyst* were used synonymously. The term *syrinx formation* was also frequently used and it describes the same as *syrinx*. The terms *hydromyelia* and *syringomyelia* were used indiscriminately regardless of communication with the fourth ventricle or not. Other constructed and "imaginative" terminology was also used (Table 2).

In the literature, the terminological use of "communicating" and "non-communicating" syringomyelia is associated with the classic communicating theories [4, 7, 11, 17, 21]. Apart from this, there seems to be no consensus in the literature regarding the definition of syringomyelia versus hydromyelia—the hybrid term "syringohydromyelia" is even used (Table 3).

Discussion

In spite of syringomyelia (G95.0) and hydromyelia (Q06.4) existing as independent ICD-10 entities, we have shown that the use of classifying terminology for fluid-filled cavities in the spinal cord is indiscriminate and inconsistent. Syringomyelia is in the ICD-10 category of "Other diseases of the spinal cord", while hydromyelia is classified under "Other congenital malformations of the spinal cord". In our observations, if any clinical distinction is made at all, the most prevalent classification is based on anatomical features (e.g. fourth ventricle communication or ependymal rupture) rather than congenital causality. Given these confounding circumstances, we therefore question the need and feasibility of sustaining two separate ICD-10 entities. We find it justifiable to propose

Table 3 Cases of the literature

Authors, year	Gender	Age	Aetiology	Terminology	Treatment	Results
Hammond et al., 2002	M	39 years	DWS Hydrocephalus Syringomyelia (C3-Th4)	Syringomyelia Syrinx	Revision of posterior fossa cyst shunt	Headache resolved, Dandy-Walker cyst reduced, syrinx size significantly decreased
Lucchetta et al., 2009	M	18 years	CM Syringomyelia (C1-D11)	Syringomyelia Syrinx	FMD with partial C1/C2 laminectomy and suboccipital craniectomy with duraplasty	All symptoms gone, large reduction of syrinx size
Marin et al., 2010	F	61 years	Posterior fossa arachnoid cyst Syringomyelia (C1-Th10)	Syringomyelia Syrinx	Endoscopic fenestration of cyst	Recovery of diplopia and anaesthesia in right leg, paraesthesia still present
					Posterior fossa craniotomy with further fenestration of cyst	No improvement
					FMD with duraplasty and removal of subarachnoid adhesions	Distinct improvement of all symptoms, syrinx size halved
McLone, 2000	Child	?	CM Hydrocephalus Myelomeningocele Syringomyelia (holocord)	Holocord hydromyelia Hydromyelia	Shunt revision	Distinct reduction of syrinx size
Mohanty et al., 2005	M	20 years	CM Hydrocephalus Syringomyelia (cervicothoracic region)	Communicating syringomyelia Non-communicating syringomyelia Syringomyelia Syrinx	ETV	General improvement of symptoms incl. improved gait and resolved headache, 2 years after a reduction of tonsillar herniation and a collapsed syrinx is observed, residual ventriculomegaly still present
	M	40 years	CM Hydrocephalus Syringomyelia (cervicomedullary)	Communicating syringomyelia Non-communicating syringomyelia Syringomyelia Syrinx	ETV	Cough-induced headache improved, 1 year after a reduction of tonsillar herniation and a collapsed syrinx is observed, residual ventriculomegaly still present though, patient is without any symptoms
Richter and Pincus, 2006	F	9 months	DWS Hydrocephalus Syringomyelia (C4-Th7)	Communicating syringohydromyelia Non-communicating syringohydromyelia Non-communicating syrinx Syringohydromyelia Syrinx	Shunting of posterior fossa cyst	Satisfyingly decompressed Dandy-Walker cyst, child is developing normally, 15 months old a totally collapsed syrinx is observed

CM Chiari malformation, *DWS* Dandy-Walker Syndrome, *ETV* endoscopic third ventriculostomy, *FMD* foramen magnum decompression

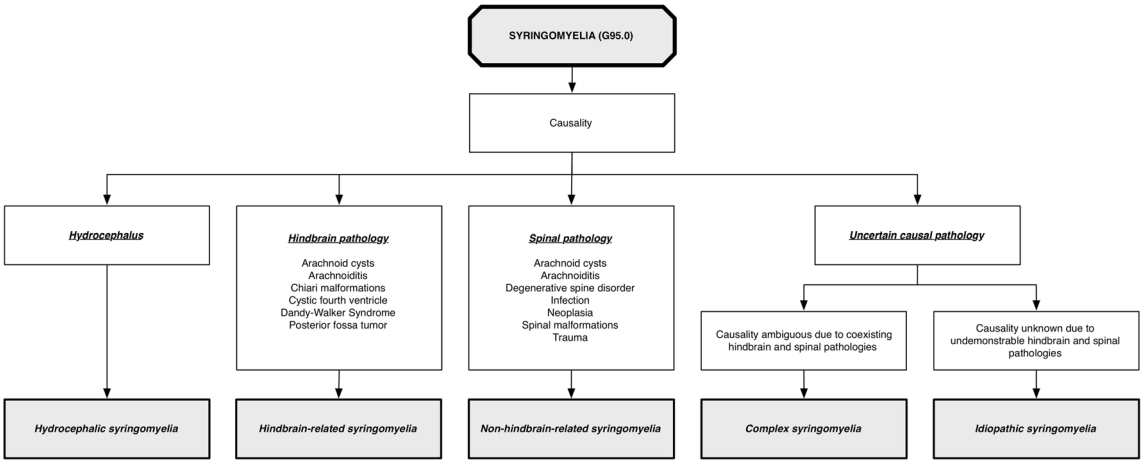

Fig. 2 A new causal ICD classification of syringomyelia (G95.0). Hydromyelia (Q06.4) is abandoned as classification. The first subgroup, hydrocephalic syringomyelia, describes the cases of syringomyelia with coexisting hydrocephalus, where treatment of the hydrocephalus by itself resolves the syrinx. Hindbrain-related syringomyelia and non-hindbrain-related syringomyelia describe the cases where there is a proven hindbrain or spinal pathology. Two additional subgroups, idiopathic syringomyelia and complex syringomyelia, respectively cover syringomyelia with unknown or ambiguous causality, i.e. indemonstrable or coexisting hindbrain and spinal pathology

a general ICD-10 reclassification in regards to fluid-filled spinal cord cavities even if the focus of our review is hindbrain-related syringomyelia; firstly, the lack of consensus in definition extends also to non-hindbrain related syringomyelia; secondly, hindbrain-related syringomyelia constitutes the majority of cases by far [3]. We propose that syringomyelia (G95.0) is used as the one and only classification for all fluid-filled cavities in the spinal cord regardless of associated pathology (Fig. 2). Consequently, we propose that hydromyelia (Q06.4) is abandoned as classification. We support our simplifying and unifying proposal by pointing out that there is general agreement on a pathogenetic mechanism involving

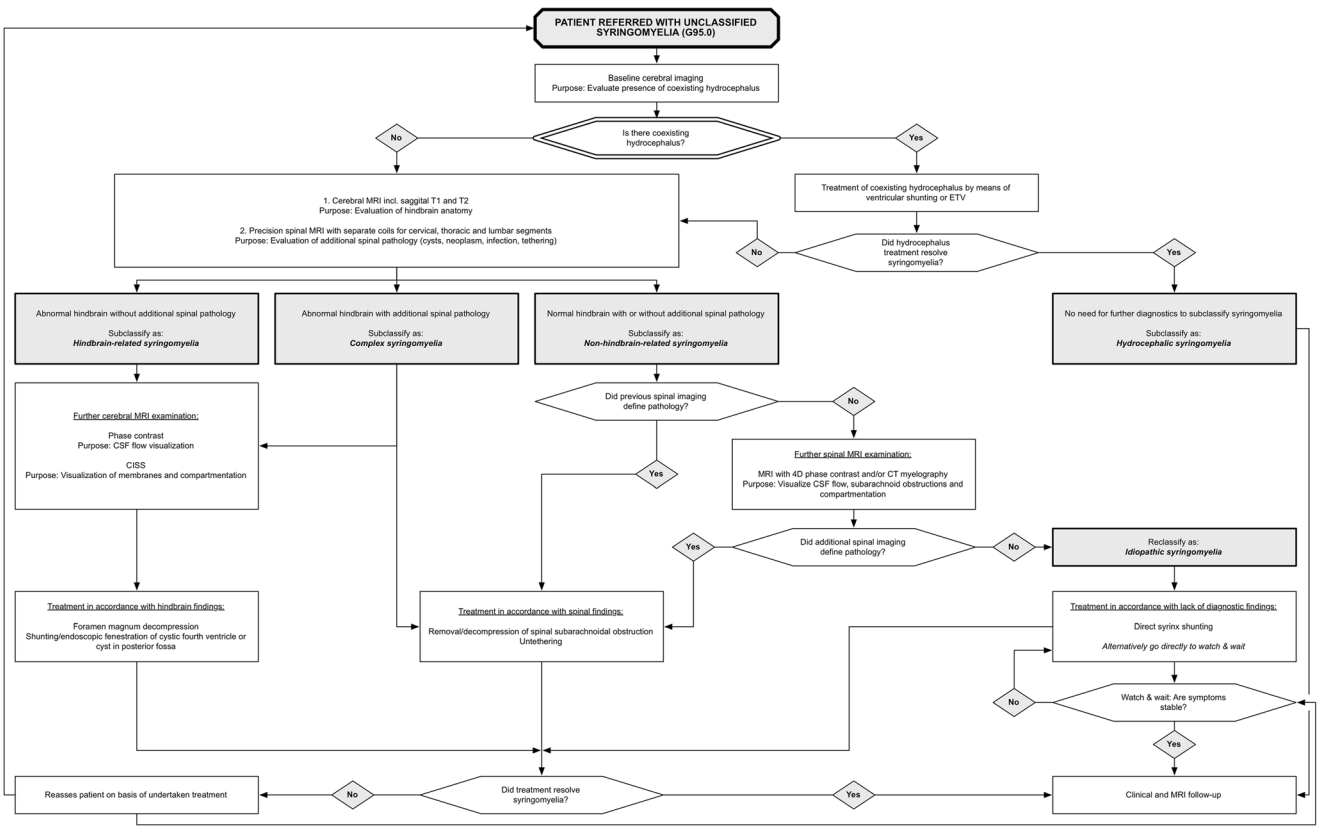

Fig. 3 Clinical algorithm for treatment of syringomyelia guided by our proposed causal ICD classification. Start from the *top*, "Patient referred with unclassified syringomyelia (G95.0)"

abnormal CSF flow across the foramen magnum and a general agreement on normalisation of this as the primary treatment, regardless of the terminology used. Thus, theory and treatment are well-defined and independent of classification. On this account it is clear that only one entity is needed. Keeping syringomyelia over hydromyelia is the most sensible option since syringomyelia is more commonly used in both the literature and the clinic, and syrinx and syringomyelia accompany each other logically.

Semantically, syrinx should be used as a noun to describe a fluid-filled cavity within the spinal cord and syringomyelia should describe the resulting disease concept. Thus, the two are part of the same pathophysiological entity.

In a previous attempt to sort out the terminological mess, Roser defined hydromyelia and syringomyelia based on radiological and electrophysiological criteria combined with the patient's symptoms [24]. Thus he forced a new definition to suit the existing terminology of hydromyelia and syringomyelia. Hydromyelia was classified as a subgroup of "idiopathic syringomyelia" with no neurological deficits except diffuse pain, a lack of electrophysiological alterations and no intraspinal CSF flow disturbing pathology. We do not find that such a classification justifies sustaining two different ICD-10 entities, since, according to this, hydromyelia does not per se constitute a separate disease, but only exists as a subgroup of syringomyelia with mild or no symptoms.

We also propose a new causal classification of syringomyelia that could serve as the basis for a new subdivision of the ICD classification. Our classification divides syringomyelia into five subgroups according to the associated pathologies: hydrocephalus, hindbrain pathology and spinal pathology (Fig. 2).

Thus, a new ICD classification could be based on these five subgroups, each with a separate subdividing diagnosis code. With this classification we also propose that the use of "communicating" and "non-communicating" syringomyelia is abandoned in the same way as hydromyelia since the use is just as inconsistent and of no consequence to the chosen treatment method.

We, furthermore, propose a clinical algorithm for diagnostics and treatment that sensibly incorporates the new classification (Fig. 3).

It would not make much sense to propose a new classification if this could not be applied easily and usefully in the clinic. It is important to remember that a given classification ideally should serve as a helpful guide for treating the patient, and not simply exist for the sake of the classification itself. In case of syringomyelia with hydrocephalus, where the syringomyelia is clinically and/or radiologically sufficiently corrected by treating the hydrocephalus, there is no need for further diagnostics. In cases where there is no hydrocephalus and in cases where treatment of coexisting hydrocephalus does not result in sufficient clinical or radiological improvement, further subclassification can guide the treatment.

Conclusions

A given treatment method originates from the believed pathogenetic theory, and the terminology ought to be the coherent link between the two. Comparison and analysis across publications, e.g. in meta-analyses, is of high value, particularly in relatively rare disorders like syringomyelia. This is impossible to take full advantage of if there is no consensus in classification and resulting terminology. Furthermore, an unambiguous classification provides the basis for a safe and unmistakable handling of patients between interdisciplinary colleagues in the clinic. We have proposed a new ICD classification that can be readily used as a standardising and easily accessible, clinically logical, diagnostic tool that also guides the clinical treatment.

Conflicts of interest None.

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