Syringomyelia and Syringobulbia

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The term syringomyelia designates a disorder that is characterised by the formation of cavities in the spinal cord or the brainstem (syringobulbia). Typically, syringomyelia is associated with an underlying pathological condition that is marked by a disturbance in the circulation of cerebrospinal fluid. Spinal tumours, trauma, congenital malformations in the cranio-cervical junction and degenerative changes in the spine can all result in syringomyelia. The syrinx contains a fluid resembling cerebrospinal fluid and can appear at the centre or the periphery of the spinal cord too. Although there are various pathophysiological models to explain the occurrence of a syrinx, syringomyelia may be understood as the consequence of a chronic interstitial oedema, where the extracellular fluid cannot flow out of the spinal cord, resulting in the formation of an intramedullary cyst. The term hydromyelia refers to a congenital variant of syringomyelia where the central canal remains open, a condition that persists in approx. 1–5 % of the population predominantly in the thoracic spinal cord (approx. 70 % of cases); hydromyelia is covered with an ependymal layer. Unlike syringomyelia, hydromyelia is very rarely accompanied by pathological swelling of the spinal cord [24].

43.1 Clinical Presentation

The ‘syrinx’ (Greek: pipe) develops in a cranial or caudal direction from the area of pathology, which means that either the upper or lower

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portion of the syrinx is in close contact with the pathological area. In terms of direction, in the Chiari malformation, the syrinx is almost always oriented in a descending direction, but in post-traumatic syringomyelia, it is frequently ascending. First, one must determine using imaging procedures whether the syrinx communicates with the fourth ventricle or the subarachnoid space and whether there are any obstructions to flow in the foramen magnum, such as downward displacement of the cerebellar tonsils resulting from a Chiari malformation. The most important diagnostic procedure for determining whether a syrinx communicates with the subarachnoid space or the fourth ventricle is the MRI examination [6, 32]. Using different relaxation times and gadolinium contrast enhancement, it is possible to differentiate syrinx formation in the context of neoplasia from a syrinx that is caused by a disturbance in cerebrospinal fluid pulsation. To accomplish this, an EKG-triggered MRI procedure (known as a cardiac-gated MRI or a CINE-MRI) [21] also proves helpful as it displays the pulsation of cerebrospinal fluid in dynamic form and shows the focal disturbances of the cerebrospinal fluid. If the CINE-MRI suggests the possibility of focal arachnopathy, then a focused post-myelogram CT can be very helpful for showing the arachnopathy again in axial cross-sectional images. This may also make it possible to demonstrate flow of contrast medium into the syrinx [2]. Electrophysiological testing (electromyography, somatosensory evoked potentials) can also be helpful for demarcating the level and extent of the lesion as well as quantifying the functional deficit, since evidence of neuronal damage on electrophysiology often occurs prior to clinical symptoms. In this regard, we should mention specially fractionated SEP examinations, measurement of central motoric latency [16] and detection of silent periods for demonstrating changes in the lateral spinothalamic tract [23].

The initial neurological symptom is usually caused by the underlying illness and not by the syrinx. Accordingly, a deliberate and careful history is critically important for identifying the actual symptomatology. Clinically, abnormalities tend to occur only after a prolonged course, and they primarily involve protopathic sensory disturbances due to irritation of the spinothalamic tracts in the area of their segmental crossing. The protopathic sensory disturbance with maintained tactile sensation as well as maintained sense of vibration and position is referred to as ‘sensory dissociation’. Circumscribed muscular atrophy and paresis may occur as a result of nuclear lesions. As the syrinx increases in volume, elevated pressure causes damage to the long spinal nerve tracts resulting primarily in lower extremity paresis and spasticity with disturbances of bladder and bowel function. In the case of syringobulbia, pressure-related nuclear cranial nerve lesions may result in unilateral atrophy of the tongue, facial hypoesthesia or analgesia, as well as various types of nystagmus.

43.1.1 Aetiology

The pathophysiological mechanisms for the occurrence of syringomyelia are determined by the specific underlying condition. Theories to explain the pathogenesis include inflammatory ependymal processes, vascular causes and hydrodynamic disturbances. Persistent pressure differences between the spinal subarachnoid space and the central canal may lead to syringomyelia. This may result from subarachnoid blockage as occurs with an arachnoid cyst, which can then lead to an influx of CSF via the fourth ventricle and the obex. Alternatively, the pressure difference may arise across the spinal cord through what are known as the extracellular Virchow-Robin spaces [19]. Greitz [12] emphasise the importance of subarachnoid adhesions in the pathogenesis of syringomyelia. They suggest that subarachnoid adhesions induce a Venturi effect (also referred to as Bernoulli’s law), which causes acceleration in cerebrospinal fluid pulsation, leading to passive suction-related swelling of the spinal cord with accumulation of extracellular fluid and, in this way, can lead to the de facto formation of a syrinx.

43.1.2 Syringomyelia in Spinal and Cerebral Malformations

Syringomyelia and syringobulbia are associated with a variety of different developmental abnormalities of the nervous system. The developmental disorders that predispose to syringomyelia are typically classified as neurulation abnormalities (e.g. meningoceles, meningoencephaloceles and meningomyeloceles), which in turn are frequently associated with Chiari malformations [14]. About 75 % of patients with type 1 Chiari malformations develop syringomyelia [1]. An important pathogenetic mechanism in the development and progression of syringomyelia in these cases is tonsillar herniation with subsequent disturbances of cerebrospinal fluid pulsation at the cranio-cervical junction [17]. Due to pathological variations in size of the posterior cranial fossa, the basilar invagination, or the Dandy-Walker malformation, can also result in disturbances of cerebrospinal fluid circulation and thus can cause syringomyelia [13]. The accompanying syringomyelia typically affects the cervical spinal cord, but can also occur more distantly at any other site along the spinal cord.

43.1.3 Syringomyelia in Spinal Neoplasia

In every case of syringomyelia, the presence of an intramedullary tumour must be suspected until proven to the contrary. Nearly half of all intramedullary tumours, haemangioblastomas and ependymomas in particular, have cystic components that may present in the form of a syrinx [10]. The cystic fluid typically has a particularly elevated protein level and thus can be usually differentiated from other forms of syrinx by means of neuroradiological imaging.

43.1.4 Post-traumatic Syringomyelia

Trauma to the spinal cord, ischaemic infarcts, postmeningitic changes and spontaneous intramedullary haemorrhage, transverse myelitis and radiation necrosis can all potentially lead to the development of syringomyelia. In addition, syringomyelia may develop after a longer symptom-free interval following 'trivial' spinal cord trauma that initially involved no neurological deficit [26]. One should suspect traumatic syringomyelia whenever a patient develops new secondary neurological symptoms after spinal cord trauma. Spinal instability, compression of the spinal cord and the tethered cord syndrome are other potential causes of syringomyelia. In large case series, the incidence had been reported at approx. 3 % [9], while on long-term follow-up observation, the incidence could also be over 51 % [4]. The occurrence of late abnormalities of this kind has not yet been explained in detail. Arachnoid scarring may be produced by various mechanisms including inflammatory changes to the arachnoid membrane (bacterial, viral, chemical) or secondary to blood constituents after a subarachnoid haemorrhage. Mechanical irritation from trauma or degenerative changes in the spine such as scoliosis, kyphosis or instabilities may lead to circumscribed, local arachnoid scarring with subsequent disturbances in cerebrospinal fluid circulation, which in turn may trigger the formation of a syrinx. Often there are arachnoid cysts that arise distal to the syrinx, and they have an additional compressive effect on the spinal cord. After direct haemorrhage (haematomyelia) resulting from trauma, a cavity may be created as the blood residues are absorbed and from the development of reactive gliosis in the area of the haemorrhagic cyst. Arachnoid changes are of particular relevance in cases of subclinical trauma [11] (Figs. 43.1 and 43.2).

43.1.5 Hydromyelia

As a result of the increased use of diagnostic imaging procedures in recent years, there have been larger numbers of patients diagnosed with filiform enlargement of the central spinal canal. Sometimes, these patients are quickly given the diagnosis of syringomyelia and thereby stigmatised. In these cases, it is useful to perform

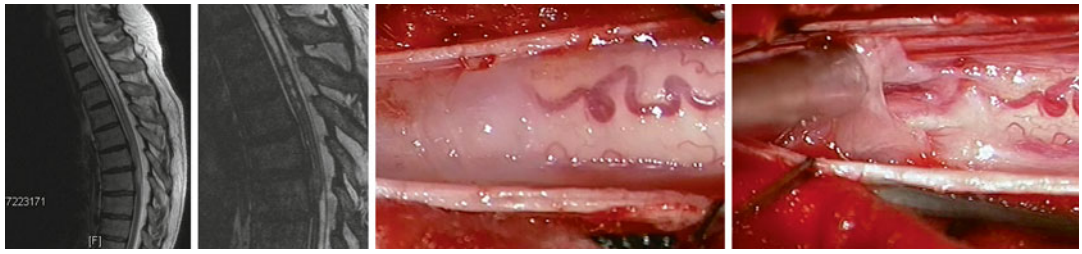

Fig. 43.1 Syringomyelia from an arachnoid cyst. MR-T2-weighted sagittal image with no obvious pathology seen (*left*); MR-3D-true-fisp-weighted sagittal image (CISS sequence) with visualisation of an arachnoid cyst compressing the spinal cord and ascending syringomyelia (*middle*). Intra-operative photo of CSF disturbance by the subarachnoid cyst and its resection (*right*)

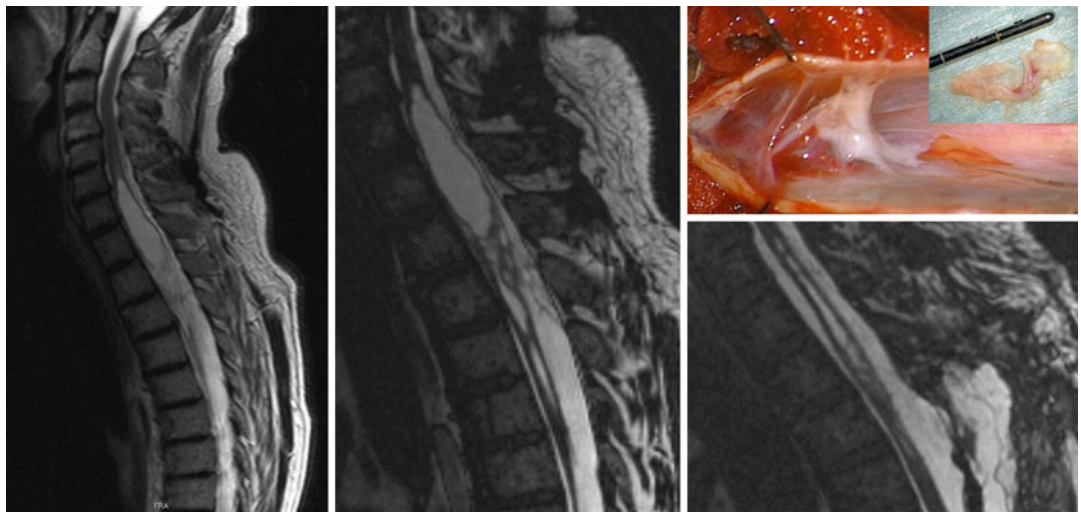

Fig. 43.2 Post-traumatic syringomyelia. MR-T2-weighted sagittal image with extensive syringomyelia (*left*). MR-3D-true-fisp-weighted image reveals post-traumatic tethering leading to CSF disturbance and consecutive syringomyelia (*middle*) [22]. Intra-operative visualisation of the thickened arachnoid membranes leading to a post-traumatic tethered cord with consecutive syringomyelia

a careful diagnostic evaluation in order to differentiate between true early-stage syringomyelia and a congenital enlargement of the central canal or terminal ventricle [24]. True hydromyelia does not have a progressive clinical-neurological course and does not change on serial imaging and hence does not constitute a pathological finding.

43.2 Clinical Course

In the pre-MRI era, the prevalence of syringomyelia was estimated to be about 8.4 per 100,000 [28] patients treated in a neurological clinic, but since the introduction and widespread use of MRI, this estimate has risen to 24.5 per 100,000 [16]. In those forms of syringomyelia associated with cerebral malformations, symptoms are often quite delayed and typically occur in the third to the fifth decades of life (the most frequently associated cerebral defect: Chiari I malformation). Frequently, the initial symptom is pain and paraesthesia in the arms [34] and, subsequently, nuclear atrophy. Spasticity primarily occurs in the lower extremities. With a progressive course, the expansion of the cavities results in paresis and atrophy, often unilateral at first, and scoliosis due to paresis of the paravertebral musculature.

In children, scoliosis is often the predominant initial clinical symptom of an underlying syringomyelia. Neurological abnormalities only develop after certain period has elapsed. In these cases, one clinical sign indicating the presence of a syrinx may be unilaterally diminished or absent abdominal skin reflexes [35]. The syrinx is predominantly located in the cervical or upper thoracic spinal segments. However, rostral extensions to the brainstem (syringobulbia) or caudal extension to the sacral segments is not unusual.

The spontaneous course of the illness depends upon the underlying pathology. In cases with tonsillar herniation or a mass in the area of the posterior cranial fossa, persistent disturbances in cerebrospinal fluid circulation may result in progressive syringomyelia, which can only be alleviated by relieving the disturbed CSF drainage in the posterior fossa. Likewise, in the case of spinal tumours, the clinical course of the secondary syrinx arising from the tumour depends upon treatment of the mass. Post-traumatic, post-inflammatory and post-chemical syrinxes frequently show an unpredictable course. Overall, approx. 60 % of patients with syringomyelia have a chronic and progressive course, while 25 % have a fluctuating course with periods of progression and periods when symptoms remain stable. Only approx. 15 % of patients have no progression in their deficits [31]. Generally, it is not possible to predict the course of the illness without knowing the individual aetiology. Case reports repeatedly remind us that any physical manoeuvres involving a sudden increase in intraspinal or intracerebral pressure may precipitate sudden deterioration. These would include vigorous sneezing or straining. Clinically, this often results in increased pain or the initial occurrence of pain. Secondary problems may also occur from infections, autonomic dysregulation and trophic disturbances. In most cases restoration of free cerebrospinal fluid flow leads to stabilisation of existing neurologic symptoms. In some cases, existing neurological deficits may even improve, although once neuropathy has been present for a long time, it will tend to persist. In cases of the Chiari malformation, symptoms caused by brainstem compression will often regress, whereas symptoms directly caused by the syrinx may remain unchanged.

43.2.1 Therapeutic Principles

Syringomyelia should be regarded as a symptom of increasing arachnoid scarring from different possible aetiologies, with resultant disturbances in spinal fluid circulation. In many cases neurosurgical intervention to restore free cerebrospinal fluid flow leads not only to a collapse of the syrinx but can also stop progression of the clinical symptoms of syringomyelia, while in favourable cases, this may even result in clinical improvement. In this regard, the size of a syringomyelia does not correlate directly with the intensity of clinical symptoms. For tumours compressing the spinal cord, neurosurgical intervention or radiotherapy may sometimes become necessary. Conservative treatment primarily includes physical therapy and the symptomatic treatment of spasticity as well as pain management.

Since syringomyelia almost always has a very slowly progressive although chronic course, initial treatment is with symptomatic, medical and physical therapies. However, younger patients with post-traumatic syringomyelia without neurological deficit (e.g. secondary to whiplash injury) may benefit from early surgical intervention to prevent neurological deficits from ever having a chance to develop. A longer course of the illness (more than 2 years) is generally regarded as an adverse predictor for symptomatic improvement following neurosurgical intervention. The paresis and atrophy that may be present following a longer course generally do not regress, even after successful microsurgical arachnolysis. Neurosurgical intervention is indicated even if other symptoms have been chronically present for many years, if clinical or neuroradiological follow-up shows progression of the syrinx, especially in the cervical area, with increasing loss of sensorimotor function or distinct dissociative sensory disturbances in the upper extremities. In addition, surgical intervention with decompression of the cysts may partially relieve the accompanying pain and may halt progression. However, there have been no controlled prospective studies regarding the various forms of surgical treatment for syringomyelia. Larger case report series and retrospective

studies have typically included a range of different neurosurgical interventions.

Relatively good evidence exists for both the pathophysiological mechanisms and clinical improvement after surgical treatment of the Chiari malformation with depressed tonsillar position by means of suboccipital decompression and, if necessary, reconstruction of the posterior cranial fossa with expansile duraplasty [3, 15, 18]. The question whether bony decompression of the posterior cranial fossa is an adequate treatment for tonsillar herniation or whether it is necessary to perform tonsillar coagulation remains a matter of dispute. Excessive expansion of the foramen magnum can lead to recurrence with herniation of larger portions of the cerebellum. During decompression it is important to be careful that a sufficient opening is created in the scarred arachnoid membranes to assure free passage of cerebrospinal fluid from the fourth ventricle. When treating patients with a Chiari malformation, it is important to be aware of the frequent coexistence of a bony anomaly affecting the craniocervical junction such as the Klippel-Feil malformation, basilar invagination or C1 subluxation. These congenital abnormalities may result in cranio-cervical instability after standard surgical treatment that may necessitate further surgical intervention.

After adequate neuroradiological demonstration of a focal arachnopathy, confirmed posttraumatic syringomyelia should be treated by surgical exploration. Preoperatively, the patients should be told clearly that the planned microsurgical arachnolysis is intended to stop the clinical-neurological progression of the condition, but that they should not anticipate regression of already existing deficits. After performing a laminectomy and opening the dura, the surgeon performs a microsurgical inspection with lysis of the arachnoid adhesions and, electively, may transect the denticulate ligament. Next, an allogeneic expansion duraplasty is performed to prevent recurrent adhesions in this area. In many cases, restoring free circulation of the cerebrospinal fluid results in the extensive collapse of the syrinx associated with clinical stabilisation [25].

Spinal cordectomy may be indicated following trauma with complete transection of the spinal cord if tension on the cord caused by scarring results in the cranial extension of the syringomyelia (traumatic tethered cord). This procedure may serve to prevent ascending tetraplegic symptoms that can result from such extension.

If no focal arachnopathy can be demonstrated after intensive neuroradiological workup (MRI with FLAIR, T1/T2 ± gadolinium-weighted images, cardiac-gated CINE-MRI for revealing CSF pulsation with additional 3D-CISS sequences) [22] but the syringomyelia appears to be clinically and neurologically progressive, the placement of a cysto-subarachnoid shunt may be indicated as a measure of last resort [16]. Multiple drainage procedures of various kinds performed in the past often resulted in only short-term success. Long-term results with clinical improvement of symptoms over time had been reported in up to 80 % of patients [20]. Based upon later reports suggesting the frequent occurrence of shunt dysfunction with recurrence of the syrinx in up to 50 % of surgically treated cases, as well as major scarring of the spinal cord at the insertion site, this therapeutic procedure could no longer be justified [8, 27, 29, 30].

43.2.2 Conservative Management

Treatment-resistant pain frequently dominates the clinical symptomatology in the treatment of syringomyelia and syringobulbia. After unsuccessful surgical treatment or if there is no access to surgical treatment, a multimodal pain medication trial should be initiated, preferably consisting of tricyclic antidepressants, carbamazepine or gabapentin. At the same time, psychotherapeutic and behavioural treatment approaches should also be offered. Especially in children with neurulation disorders and an associated syrinx, severe scoliosis may dominate the early symptomatic picture (19 %) [7]. If there is marked malpositioning, it is sometimes necessary to perform surgical and orthopaedic spinal interventions under continuous neuromonitoring. Basically, all children with idiopathic scoliosis

should undergo an MRI to rule out a syrinx as the cause of their scoliosis. One indication of the presence of a syrinx is significantly diminished or absent superficial abdominal reflexes. It is interesting that the scoliosis may stabilise or even improve if the syrinx is treated early (under the age of 10) with suboccipital decompression [29]. Depending upon the nature and the extent of autonomic dysfunction, it may be necessary to use sympatholytic or sympathomimetic medications, and in the case of arthropathy, it is sometimes necessary to treat with calcitonin. If there is marked spasticity, there are appropriate medical treatments available along with supportive treatment using physiotherapeutic exercises.

43.2.3 Obsolete Forms of Treatment

Radiation therapy for syringomyelia, which used to be performed for the treatment of pain, cannot be supported by a single major study. The intervention of implanting a syringo-subarachnoid shunt cannot be considered as a form of primary treatment either. Besides the fact that shunt treatment does not remedy the disturbance in cerebrospinal fluid circulation, implantation requires myelotomy, and long-term follow-up studies have demonstrated the frequent occurrence of a wide range of complications and secondary damage (~50 %). Shunt implantation may be complicated by shunt failure, dislocation, tethered cord and arachnoid scarring that causes new and additional iatrogenic disturbances in spinal fluid circulation [5, 33].

References

1. Anderson NE, Willoughby EW, Wrightson P (1986) The natural history of syringomyelia. Clin Exp Neurol 22:71–80
2. Arlien-Soborg P, Kjaer L, Praestholm J (1993) Myelography, CT, and MRI of the spinal canal in patients with myelopathy: a prospective study. Acta Neurol Scand 87(2):95–102
3. Asgari S, Engelhorn T, Bschor M, Sandalcioglu IE, Stolke D (2003) Surgical prognosis in hindbrain related syringomyelia. Acta Neurol Scand 107(1): 12–21
4. Backe HA, Betz RR, Mesgarzadeh M, Beck T, Clancy M (1991) Post-traumatic spinal cord cysts evaluated by magnetic resonance imaging. Paraplegia 29(9):607–612
5. Batzdorf U, Klekamp J, Johnson JP (1998) A critical appraisal of syrinx cavity shunting procedures. J Neurosurg 89(3):382–388
6. Brugieres P, Iffenecker C, Hurth M, Parker F, Fuerxer F, Idy-Peretti I et al (1999) Dynamic MRI in the evaluation of syringomyelic cysts. Neurochirurgie 45(Suppl 1):115–129
7. Dobbs MB, Lenke LG, Szymanski DA, Morcuende JA, Weinstein SL, Bridwell KH et al (2002) Prevalence of neural axis abnormalities in patients with infantile idiopathic scoliosis. J Bone Joint Surg Am 84-A(12): 2230–2234
8. Edgar R, Quail P (1994) Progressive post-traumatic cystic and non-cystic myelopathy. Br J Neurosurg 8(1):7–22
9. El Masry WS, Biyani A (1996) Incidence, management, and outcome of post-traumatic syringomyelia. In memory of Mr Bernard Williams. J Neurol Neurosurg Psychiatry 60(2):141–146
10. Evans A, Stoodley N, Halpin S (2002) Magnetic resonance imaging of intraspinal cystic lesions: a pictorial review. Curr Probl Diagn Radiol 31(3):79–94
11. Freund M, Aschoff A, Spahn B, Sartor K (1999) Posttraumatic syringomyelia. Rofo 171(6):417–423
12. Greitz D (2006) Unraveling the riddle of syringomyelia. Neurosurg Rev 29:251–264
13. Guille JT, Miller A, Bowen JR, Forlin E, Caro PA (1995) The natural history of Klippel-Feil syndrome: clinical, roentgenographic, and magnetic resonance imaging findings at adulthood. J Pediatr Orthop 15(5):617–626
14. Iskandar BJ, Hedlund GL, Grabb PA, Oakes WJ (1998) The resolution of syringohydromyelia without hindbrain herniation after posterior fossa decompression. J Neurosurg 89(2):212–216
15. Klekamp J, Batzdorf U, Samii M, Bothe HW (1996) The surgical treatment of Chiari I malformation. Acta Neurochir (Wien) 138(7):788–801
16. Klekamp J, Samii M (2002) Syringomyelia: diagnosis and treatment. Springer, Berlin/Heidelberg/New York
17. Levine DN (2004) The pathogenesis of syringomyelia associated with lesions at the foramen magnum: a critical review of existing theories and proposal of a new hypothesis. J Neurol Sci 220(1–2):3–21
18. Mariani C, Cislaghi MG, Barbieri S, Filizzolo F, Di Palma F, Farina E et al (1991) The natural history and results of surgery in 50 cases of syringomyelia. J Neurol 238(8):433–438
19. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ (1994) Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. J Neurosurg 80(1):3–15
20. Pillay PK (1991) The coperitoneal shunting for syringomyelia. J Neurosurg 75(5):835–836
21. Pujol J, Roig C, Capdevila A, Pou A, Marti-Vilalta JL, Kulisevsky J et al (1995) Motion of the cerebellar

tonsils in Chiari type I malformation studied by cine phase-contrast MRI. Neurology 45(9):1746–1753
22. Roser F, Ebner FH, Danz S, Riether F, Ritz R, Dietz K, Naegele T, Tatagiba MS (2008) 3D-constructive interference in steady-state (CISS) magnetic resonance imaging in syringomyelia: advantages to conventional imaging. J Neurosurg (Spine) 8:429–435
23. Roser F, Ebner FH, Liebsch M, Dietz K, Tatagiba MS (2008) A new concept in the electrophysiological evaluation of syringomyelia. J Neurosurg (Spine) 8:517–523
24. Roser F, Ebner FH, Sixt C, Hagen JM, Tatagiba MS (2010) Defining the line between hydromyelia and syringomyelia. A differentiation is possible based on electrophysiological and magnetic resonance imaging studies. Acta Neurochir (Wien) 152(2):213–219
25. Roser F, Tatagiba M (2007) Posttraumatic syringomyelia. J Neurosurg (Spine) 6(1):193–195
26. Rossier AB, Werner A, Wildi E, Berney J (1968) Contribution to the study of late cervical syringomyelic syndromes after dorsal or lumbar traumatic paraplegia. J Neurol Neurosurg Psychiatry 31(2): 99–105
27. Schaller B, Mindermann T, Gratzl O (1999) Treatment of syringomyelia after posttraumatic paraparesis or tetraparesis. J Spinal Disord 12(6):485–488
28. Schliep G (1978) Syringomyelia and syringobulbia. In: Vinken P, Bruyn G (eds) Handbook of clinical neurology. Elsevier, Amsterdam, pp 225–327
29. Sengupta DK, Dorgan J, Findlay GF (2000) Can hindbrain decompression for syringomyelia lead to regression of scoliosis? Eur Spine J 9(3):198–201
30. Sgouros S, Williams B (1995) A critical appraisal of drainage in syringomyelia. J Neurosurg 82(1):1–10
31. Sixt C, Riether F, Will BE, Tatagiba MS, Roser F (2009) Evaluation of quality of life parameters in patients who have syringomyelia. J Clin Neurosci 16(12):1599–1603
32. Tanghe HL (1995) Magnetic resonance imaging (MRI) in syringomyelia. Acta Neurochir (Wien) 134(1–2):93–99
33. Van Calenbergh F, van den Bergh WM (1993) Syringo-peritoneal shunting: results and problems in a consecutive series. Acta Neurochir (Wien) 123(3–4):203–205
34. Van den BR, Hoorens G, Van Calenbergh F (1990) Syringomyelia: a retrospective study. Part I: clinical features. Acta Neurol Belg 90(2):93–99
35. Zadeh HG, Sakka SA, Powell MP, Mehta MH (1995) Absent superficial abdominal reflexes in children with scoliosis. An early indicator of syringomyelia. J Bone Joint Surg Br 77(5):762–767