Syringomyelia

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Introduction

Overview

In this article, the author reviews the early and late manifestations of syringobulbia, the differential diagnosis of syringomyelia and hydromyelia, and the 90% chance of long-term stabilization or improvement with surgical treatment in Chiari-related syringomyelia. Syringomyelia and associated conditions, such as arachnoiditis and the Chiari I anomaly, are now readily diagnosed by MRI. An associated Chiari anomaly may be overlooked by MRI in the supine position. Misdiagnosis may occur if sagittal imaging alone is used because sagittal MRI might overestimate the degree of tonsillar ectopia in Chiari I malformation. Posterior fossa decompression in Chiari I ameliorates denervation of paraspinal muscles, one of the essential elements in the pathophysiology of syringomyelia-related scoliosis. Patients with idiopathic syringomyelia (unrelated to Chiari I anomaly) have abnormally narrow upper and mid cervical spinal canal diameters. In posttraumatic syringomyelia, a disturbance in the removal of K ions in tissue surrounding the syrinx cavity may contribute to water accumulation in the injured spinal cord, leading to syrinx formation or exacerbation of the underlying pathology.

Key points

- MRI is the diagnostic modality of choice for syringomyelia and associated conditions, such as Chiari I anomaly or arachnoiditis, and for the differential diagnosis of syringomyelia and hydromyelia.
- Preventive measures for the development of syringomyelia include the careful use of epidural anesthesia and the avoidance of a traumatic or prolonged labor and maneuvers that may induce “craniospinal” dissociation, such as Valsalva maneuver.
- Pain is a frequent and disabling manifestation of syringomyelia and a persistent symptom in a significant percentage of patients despite successful drainage of the syrinx.
- Some patients show progressive neurologic deterioration and disabling pain despite proper surgical drainage of the syrinx and correction of the associated craniovertebral anomaly.

Historical note and terminology

Ollivier d'Angers first coined the name "syringomyelia" in 1824 (Ollivier d'Angers 1824), but the condition was first recognized by Stephanus in 1545 (Stephanus 1545). Schultze first described its clinical picture (Schultze 1882); its relation to Chiari malformation was first outlined by John Cleland (Cleland 1883). Abbe performed the first reported surgical drainage of a syrinx in 1891 (Abbe 1892), although claims were made that Horsley aspirated a syrinx in 1890 (Putnam and Warren 1899). The “hydrodynamic” theory of syringomyelia was proposed by Gardner (Gardner 1965) and modified by Williams' “craniospinal pressure dissociation” theory (Williams 1978).

The term “syringomyelia” is used to indicate the presence of a fluid-filled cavity within the spinal cord. "Hydromyelia" refers to dilatation of the central canal of the spinal cord: cavities partially or completely lined by ependymal cells. Syringomyelia indicates cavities that lie outside the central canal and do not have ependymal lining but may have partial connection with the central canal. “Communicating” syringomyelia refers to cavities with a direct communication with the fourth ventricle through the obex and is usually associated with hindbrain malformations. “Noncommunicating” syringomyelia refers to cavities without communication with the fourth ventricle and is usually secondary to trauma and tumors of the spinal cord but is also associated to hindbrain malformations.

Clinical manifestations

Presentation and course

Onset is usually insidious and rarely acute (Milhorat et al 2003), beginning between 25 and 40 years of age but ranging from infancy to 70 years. Initial manifestations include pain, numbness of the hands, stiffness of the legs, scoliosis (Godzik et al 2014), vertigo, oscillopsia, diplopia, dysphonia, dysphagia, laryngeal stridor, hiccups (Seki et al 2004), paroxysmal cervicobrachial cough-induced pain, sweating abnormalities, torticollis, drop attacks, and neurogenic arthropathy (Barnett et al 1973; Schliep 1978; Caplan et al 1990). The dissociated sensory loss is commonly first observed along the ulnar border of the hand and forearm, extending to the arm, upper part of the chest and back in a cape or half-cape distribution, uni- or bilaterally, and in the face following an “onionskin” distribution. All types of sensation may be lost in a limb due to involvement of the root entry zone. In advanced cases, compression of the spinothalamic tracts or the posterior columns give rise to long tract signs in the legs. Wasting and weakness first appear in the hands and progress to the forearm, arm, and trunk.{embed="pagecomponents/media_embed" entry_id="20612"} Hypotonia, areflexia, and fasciculations are commonly observed. Upper limb jerks may be preserved or brisk, or the Hoffman sign may be present due to the Chiari anomaly (Logue and Edwards 1981). Involvement of corticospinal tracts in advanced cases leads to spastic paraparesis. In the presence of lumbar syringomyelia, upper and lower motor neuron signs are found in the legs. Horner syndrome is present in 5% to 23% of cases and is (often) the only sign of the disease (Kerrison et al 2000). The incidence is probably higher because sympathetic denervation can be overlooked when the change in pupillary diameter is minimal. The diagnosis is best established by looking for subtle ptosis and miosis (Nogués et al 2010). Trophic changes in the hands include hyperkeratosis, scars from old burns, subcutaneous edema, or hematomas.{embed="pagecomponents/media_embed" entry_id="20613"}. Rarely, uniform enlargement of 1 upper limb associated with sympathetic hyperactivity can be observed. The presence of subcutaneous edema may give a juicy appearance to the digits and dorsum, the so-called "main succulente." (Contributed by Dr. Martín Nogués.)}| Neuropathic arthropathy may involve shoulder, elbow, and hand, separately or simultaneously. Rotatory or vertical nystagmus is the most common sign of syringobulbia. Others include an onionskin pattern of sensory loss in the face, palatal weakness, and tongue atrophy with fasciculations (Jonesco-Sisesti 1986; Delpirou et al 2001). The cavity may ascend to the pons or even the internal capsule (Barnett et al 1973).{embed="pagecomponents/media_embed" entry_id="20614"}

Advanced cases may have gastrointestinal (nausea, vomiting, epigastric pain, constipation, fecal incontinence), respiratory (dyspnea, stridor, central and obstructive sleep apnea, respiratory arrest) (Heidel et al 2002), and cardiovascular (orthostatic hypotension, postural tachycardia) (Nogués et al 2001) manifestations. Involuntary movements include spinal myoclonus, postural tremor, and focal dystonia (Nogués et al 1999a). In posttraumatic syringomyelia, onset of symptoms ranges from 3 months to 35 years after trauma and include an ascending sensory level, pain in the neck or arms, increased muscle weakness, and spasticity (Barnett et al 1973).

Pain is a major complaint in a significant proportion of syringomyelia patients and is probably due to disordered neuronal processing in the damaged dorsal horn.

Prognosis and complications

Untreated syringomyelia tends to run a slowly progressive course.{embed="pagecomponents/media_embed" entry_id="20615"} Nevertheless, it is known that, for some patients, syringomyelia improves or stabilizes without surgical treatment, and spontaneous resolution of syringomyelia has been observed more frequently using MRI (Tortora et al 2012). However, the mechanisms of the development of syringomyelia and its spontaneous resolution remain controversial (Kyoshima and Bogdanov 2003), and surgery has been reported as an effective and safe treatment of Chiari-related syringomyelia, with a 90% chance of long-term stabilization or improvement on average (Aghakhani et al 2009). Scoliosis is associated with larger maximum syrinx diameters (Godzik et al 2014), and treatment with posterior fossa decompression may lead to a reduction of the denervation of the paraspinal muscles (Sha et al 2017). Syringobulbia usually complicates syringomyelia, possibly through the “slosh” concept (Williams 1997). When intraspinal pressure is increased by straining or tightening of the abdominal muscles and cannot equalize intracranial pressure due to ectopia, this pressure is transmitted to the syrinx, the fluid cyst traveling upwards. Other long-term complications include neurogenic arthropathies, cervical spondylosis, central and obstructive sleep apnea, and sudden death (Nogués et al 1999b; Heidel et al 2002).

Although early decompression of Chiari I malformation with syringomyelia results in stabilization of scoliosis, surgery is required to improve the spinal deformity (Eule et al 2002; Bradley et al 2007). Postoperative relief of headaches and neck pain is observed in 83% of children with Chiari I malformation (Tubbs et al 2003). Even adequately operated patients may show subtle deterioration, most likely due to gliosis alongside the walls of the syringomyelic cavities, even though the syrinx is no longer distended. Bad prognosis is associated with symptoms of over 2 years duration, presence of ataxia, nystagmus, bulbar symptoms, muscle atrophy, dorsal column dysfunction, or a longer and deviated syrinx or narrowing of the subarachnoid space (Dyste et al 1989). Persistent dysesthetic pain can occur despite improvement or collapse of the syrinx on the postoperative MRI; it may remain the most disabling complaint for patients (Batzdorf 1988). Spontaneous drainage of the syrinx into the subarachnoid space may occur, usually in the context of an acute episode of local pain and neurologic deterioration (Bogdanov et al 2000).

Complications after foramen magnum decompression include spinal cord damage by neck hyperextension or hyperflexion during intubation, spinal cord ischemia due to arterial hypotension, CSF leakage with formation of a symptomatic pseudomeningocele, posterior fossa bleeding, infection, hydrocephalus, cerebellar ptosis, and severe brainstem compression that necessitates transoral odontoidectomy (Tubbs et al 2003).{embed="pagecomponents/media_embed" entry_id="20616"}{embed="pagecomponents/media_embed" entry_id="20617"} Complications of shunting procedures are malfunction, local hematoma, infection, or a collapsed syrinx.

Clinical vignette

A 19-year-old female noticed mild progressive weakness and numbness distally in upper extremities for the last 12 months. Birth history was normal, and there was no history of head injury. On examination she had left Horner syndrome, bilateral weakness and wasting of intrinsic hand muscles, an area of suspended dissociated sensory loss from C3 to T8 bilaterally, areflexia in upper limbs, and brisk lower limb reflexes. Hairline was rather low, and there was a mild thoracic scoliosis. Nerve conduction studies showed reduced compound motor action potential amplitudes in both median and ulnar nerves as well as normal sensory nerve action potentials. An MRI showed a Chiari I anomaly and an extensive bilateral cervicothoracic syrinx.{embed="pagecomponents/media_embed" entry_id="20618"}{embed="pagecomponents/media_embed" entry_id="20619"} The patient was treated with decompression of the foramen magnum and placement of 2 syringo-peritoneal shunts. She did well initially, but 6 months after surgery she developed progressively disabling headaches occurring specifically in the upright position or during periods of strenuous activity. An MRI showed further cerebellar descent, an abnormal angulation and indentation of the midportion of the medulla oblongata, and partial collapse of the syrinx.{embed="pagecomponents/media_embed" entry_id="20620"} Symptoms disappeared after further duraplasty of the posterior fossa and extraction of the shunts. The condition has remained stable during a follow-up period of 10 years.

Biological basis

Etiology and pathogenesis

Many familial cases of craniovertebral malformations including syringomyelia, syringomyelia with Chiari malformation, and simple Chiari malformation have been reported (Inoue et al 2003). It is likely that syringomyelia associated with Chiari I anomaly is genetically determined. The term “Chiari 1.5 malformation” entails Chiari I malformation in combination with brainstem herniation through the foramen magnum (Tubbs et al 2004). Many patients with syringomyelia were born after a difficult labor (Williams 1978). Syringomyelia may complicate basal or spinal arachnoiditis after bacterial meningitis, subarachnoid hemorrhage, tuberculosis, trauma, repetitive deceleration in skydivers (Wrobel and Taubman 2003), a CSF leak after brachial plexus avulsion (Scholsem et al 2008), idiopathic intracranial hypotension (Richards et al 2016), and reaction to radiopaque material, spinal anesthesia, or detergents. Exceptional cases of familial adhesive arachnoiditis associated with syringomyelia have been reported (Pasoglou et al 2014). Syringomyelia may be secondary to intra- or extramedullary spinal tumors and infratentorial tumors. Ependymomas and hemangioblastomas have a 50% incidence of associated syringomyelia. The following classifications based on etiologic factors have been proposed (Moufarrij and Awad 1997): (1) communicating syringomyelia, in which there is a demonstrable communication with the fourth ventricle by imaging studies; (2) blockage of CSF circulation (a) at the posterior fossa-craniovertebral junction level by a Chiari malformation, basal adhesive arachnoiditis, masses, basilar impression, or meningeal carcinomatosis, or (b) at the spinal level by tumors, arachnoid cysts, adhesive arachnoiditis, cervical spondylosis (Kimura et al 2004), or infectious masses; (3) injury to the

spinal cord tissue by trauma, radiation necrosis, infarction, hemorrhage, infection, associated to transverse myelitis, demyelinating disease, amyotrophic lateral sclerosis, or compressive myelopathy; (4) dysraphism; (5) intramedullary tumors; and (6) idiopathic syringomyelia.

In syringomyelia secondary to basal arachnoiditis as well as in a low percentage of syrinx associated to Chiari I anomaly, Gardner's “hydrodynamic theory” may be operative (Gardner 1965). CSF may be forced to enter through the obex into a patent central canal leading to communicating syringomyelia. In the majority of the Chiari I-associated cases, there is no demonstrable communication at the obex. The anatomical and physiological blocks to the CSF, which occur in response to brain expansion during cardiac systole, flow from the cranial to the spinal subarachnoid space and force the cerebellar tonsils into the partially enclosed spinal subarachnoid space (Oldfield et al 1994). Exaggerated spinal pulse pressures are generated, forcing movement of CSF from the subarachnoid space into the spinal cord through the Virchow-Robin spaces. Alternatively, these transmedullary pressure gradients may induce movement of the extracellular fluid normally produced within the spinal cord, particularly in narrow regions of the spinal CSF pathways. Abnormal accumulation of this interstitial fluid may constitute a “presyrinx” state, a potentially reversible alteration, seen in MRI as an enlarged spinal cord with parenchymal T1 and T2 prolongation before the appearance of syringomyelia, and which can manifest as an acute or chronic myelopathy (Goh et al 2008). These intramedullary high-signal lesions may vanish after decompression and fixation (Yurube et al 2009).

Syringomyelia in the absence of a Chiari anomaly may be due to an encroachment of neural structures in the posterior fossa, the so-called “tight cisterna magna.” Patients with this condition undergo craniocervical decompression with favorable results. It has been suggested that spinal cord cavities associated with tight cisterna magna should not be classified into an idiopathic group but rather into an organic group that is associated with foramen magnum lesions (Chang and Nakagawa 2003). Idiopathic syringomyelia have abnormally narrow upper- and mid-cervical spinal canal diameters, which may cause longitudinal pressure dissociation and transmural pressure gradients, leading to cavity formation (Struck et al 2016).

Extradural spondylotic compression of the spinal cord has been advocated as another etiology of syringomyelia (Butteriss and Rirchall 2006). A purely extradural decompression could be sufficient to induce regression of the medullary cavitation (Rebai et al 2002).

The involution of the central canal is the rule in man as opposed to animals, and its persistence may be genetically determined, explaining familial and idiopathic syringomyelia (Aboulker 1979).

In posttraumatic syringomyelia, necrosis and cysts resulting from fluid egress from damaged axons may develop at the site of spinal cord trauma. Alterations of aquaporin-4 expression or function may contribute to the fluid imbalance leading to syrinx formation or enlargement (Hemley et al 2012). A disturbance in the removal of K ions in tissue surrounding the syrinx cavity may contribute to water accumulation in the injured spinal cord, leading to syrinx formation or exacerbation of the underlying pathology (Najafi et al 2016).

Predisposing factors include increasing age, cervical and thoracic levels compared with lumbar, displaced fractures, and spinal instrumentation without decompression (Vannemreddy 2002). When syringomyelia is secondary to spinal arachnoiditis, a vascular mechanism may be responsible. In tumor-associated syringomyelia, neoplastic growth may interfere with blood supply to the spinal cord and cause ischemia, necrosis, and cavity formation. Alternatively, it may affect tissue fluid drainage through perivascular spaces. Edema may play a major role in syrinx development and maintenance as well.

Epidemiology"

A prevalence of 5.6 to 8.6 per 100,000 inhabitants has been reported in England (Brewis et al 1966). Proportional rates from several series range from 0.4% to 1% of cases admitted to neurologic clinics. Regional differences have been noted, with a higher prevalence in certain areas of Germany and Russia as well as a considerable proportion of familial cases (Schliep 1978). An autosomal dominant predisposition may be the primary factor in the appearance of familiar cases (Robenek et al 2006). Males and females are equally affected. Incidence of posttraumatic syringomyelia is 0.02% (Carroll and Brackenridge 2005).

Prevention

The avoidance of a traumatic and prolonged labor (including the use of forceps) may prevent the development of syringomyelia in predisposed newborns. Similarly, careful use of epidural anesthesia, avoiding entering into the subarachnoid space, may prevent the development of spinal arachnoiditis and syringomyelia. Finally, in the presence of a Chiari anomaly and disturbed CSF flow across the foramen magnum, avoidance of any maneuver that may induce "craniospinal" dissociation (such as excessive coughing, sneezing, or weight lifting) may prevent the enlargement of an established syrinx until appropriate decompression has been accomplished. Nevertheless, it is still controversial whether activity restriction is necessary in asymptomatic children with Chiari anomaly (Luciano 2011).

Differential diagnosis

In hydromyelia, neurologic exam and electrophysiological testing are normal, and the central canal is less than 6 mm in diameter on MRI (Roser et al 2010). Cervical spondylotic myelopathy can also present with lower motor neuron signs in upper limbs and upper motor neuron signs in lower limbs. Nevertheless, it is more common in the elderly, and there is no neurogenic arthropathy, trophic changes, or bulbar signs. On MRI there is narrowing of the spinal canal or cavitation. Motor neuron disease and progressive spinal muscular atrophy present with no sensory disturbances and a normal MRI (Visser et al 2002). In multiple sclerosis, dissociated sensory loss, trophic changes, and neurogenic arthropathies are rare manifestations, CSF shows high IgG and myelin-associated protein, and visual evoked potentials are usually abnormal. Noncommunicating syringomyelia may occur in multiple sclerosis patients with spinal cord pathology. It can be a subtle finding without clinical correlates (Weier et al 2008). Diabetes, leprosy, and hereditary sensory neuropathies may present trophic changes and dissociated sensory loss, but lower limbs are affected first, the distribution of the sensory abnormalities is in a glove and stocking pattern, and nerve conduction studies and nerve biopsy are abnormal. Spinal cord tumors may mimic syringomyelia, although pain is usually more severe, progression is more rapid, and there are no trophic changes or neurogenic arthropathies. An increased protein concentration in the CSF, an enlarged spinal cord, and enhancement of the lesion with gadolinium favors the diagnosis of an associated spinal cord tumor. Spinal syphilis is ruled out by the absence of dissociated sensory loss, increased CSF protein, and cell count with positive VDRL. Ulnar and median mononeuropathies and thoracic outlet syndrome rarely lead to trophic changes or neurogenic arthropathy. Sensory loss is localized to the distribution of the nerve affected. Nerve conduction studies confirm the entrapment, sensory nerve action potentials are abnormal, and MRI is normal.

Nevertheless, carpal tunnel syndrome and ulnar entrapment at the elbow are more common in syringomyelia patients than in the normal population. Filiform intramedullary cavities may be found, incidentally, in the dorsal spinal cord of normal young subjects; they are most likely due to persistence of the central canal of the spinal cord.

Diagnostic workup

MRI is the diagnostic modality of choice. T1-weighted images best delineate cord and syrinx morphology, whereas T2-weighted sequences best evaluate for associated conditions such as myelomalacia, gliosis, or tumor. In the presence of a Chiari I anomaly, axial and sagittal T1-weighted images of the cervical spine should be performed to evaluate for a cervical syrinx.{embed="pagecomponents/media_embed" entry_id="20621"} If the syrinx cavity extends caudal to the field of view, a separate examination of the thoracic spine should be performed as well to define the full extent of the central syrinx cavity. Intracranial neuroimaging should be done to rule out hydrocephalus. Tonsillar descent is considered abnormal when it is more than 5 mm below the foramen magnum, and occasionally, it can only be revealed in the sitting position during surgery, the so-called "up-and-down Chiari malformation" (Silva et al 2010). Most tonsillar herniations are asymmetrical, and most syringomyelia is eccentric (Deng et al 2014). CSF flow and peak systolic and diastolic velocities may be studied with dynamic MRI, these new techniques becoming more frequently used because there is little correlation between the degree of tonsillar descent and the presence or absence of syringomyelia. Additional anatomic measurements, such as posterior fossa size and geometry, are useful for the study of syringomyelia patients with Chiari malformation (Levy 2003). Caudal flow occurs normally during systole in the basal cisterns and in the spinal subarachnoid space. Following cardiac diastole, CSF motion reverses, flowing rostrally in the spinal canal. Flow dynamics assessed by cine phase contrast MRI could independently predict response to posterior fossa decompression (McGirt et al 2006). Following foramen magnum decompression, CSF should be identified anterior to the cervicomedullary junction and posterior to the tonsils on sagittal T1-weighted images. MR phase contrast imaging demonstrates pulsatile fluid motion in syrinx cavities in synchrony with the adjacent subarachnoid space, mainly in large cavities. Following decompression, syrinx flow decreases as the size of the cavity decreases (Levy 2003). Slit-like syrinx cavities may be found in patients studied with MRI for radicular pain, paresthesia, numbness, or muscle spasm. They probably do not represent true syringomyelia but rather remnants of

the central canal (hydromyelia) detected in a small percentage of adults. As hydromyelia does not represent a disease with an underlying pathology, no clinical or radiological progression has been seen.

Motor conduction velocity may be mildly reduced in upper limbs due to degeneration of the fastest conducting fibers. Normal sensory nerve action potentials indicate the preganglionic localization of the disorder. F-waves are prolonged in the presence of a cervical syrinx involving segments C8 and T1 (Rossier et al 1985).

Somatosensory evoked potentials help to assess dorsal column function during shunt placement. CSF composition is normal. Brainstem auditory evoked potentials (BAEP) are abnormal in a third of Chiari I malformation patients, but they do not add clinically relevant information in establishing which symptomatic patients with Chiari malformation should undergo surgical treatment. Protein levels are raised in the presence of an associated tumor. Plain skull x-rays may show signs of arrested hydrocephalus, basilar impression, or platybasia. Cervical spinal x-rays may show a wide cervical spinal canal, occipitalization of the atlas, and fusion of cervical vertebrae.

Management

The evidence that syringomyelia can spontaneously resolve makes the treatment strategy controversial (Kyoshima and Bogdanov 2003). Because of the slow progression of symptoms and that surgery may lead to a neurologic deficit, some centers advocate waiting for signs of neurologic deterioration or change in MRI findings before operating (Nishizawa et al 2001). Others, however, argue that deterioration may be sudden and that surgical intervention is warranted in symptomatic Chiari malformation when the condition is diagnosed. Nevertheless, there is general agreement that early correction of abnormal CSF dynamics is the best alternative. Indications for surgery include symptoms from foramen magnum compression, advancing syringomyelia, suboccipital and lower central neck pain, particularly provoked by Valsalva maneuver, cape-like dissociated sensory loss, dysesthesias, gait difficulty, nystagmus, upper or lower extremity weakness, cranial nerve dysfunction, and blurred vision (Logue and Edwards 1981). Surgery is not recommended in the following situations: (1) contraindications to surgery, such as advanced disease with respiratory failure and severe deficits and kyphoscoliosis; (2) patients in stable condition who progressed fairly rapidly in the early stage and then became stable; (3) patients with spontaneous cure, which may be observed in 1% of cases (Bogdanov et al 2000); (4) asymptomatic patients; and (5) a spindle syrinx in a patient with pain. Children in whom scoliosis is the only manifestation may be treated conservatively because spontaneous shrinkage of the cavity may occur.

Because shunting of the syrinx carries the risk of spinal injury, there is a strong tendency that surgical treatment of syringomyelia associated with the Chiari malformation should be directed at reconstructing normal CSF pathways. Most authors currently recommend bony decompression with opening and patching of the dura as the initial treatment. Plugging of the obex and exploration of the fourth ventricular outlet are becoming less common because there is rarely a functional communication between the fourth ventricle and the syrinx. Furthermore, this procedure may produce bradycardia, postoperative respiratory distress, or damage to the hypoglossal and vagal nuclei. In the uncommon case of a patient with a Chiari I malformation and extensive subarachnoid scarring, simple bone and dural decompression will not eliminate the obstruction of the free flow of CSF within the subarachnoid spaces and may not induce resolution of syringomyelia (Heiss et al 1999; Levy 2000). If hydrocephalus is present, it has to be treated first. Endoscopic third ventriculostomy provides a durable method of treatment for hydrocephalus associated with Chiari malformation (Hayhurst et al 2008). Section of the filum terminale is another useful strategy in cases with and without a Chiari anomaly (Royo-Salvador et al 2005).

When foramen magnum decompression is unsuccessful, shunting of the syrinx is appropriate, although the best type of shunting procedure to use remains unclear. Syringoperitoneal and syringopleural shunting place the distal catheter into a seemingly low-pressure extrathecal site. Some authors consider the preferred treatment to be syringosubarachnoid shunt through a dorsal root entry-zone myelotomy at the level of the largest part of the syrinx, in order to prevent cord damage and in order to obtain some relief of pain (Iwasaki et al 2000).

When syringomyelia is secondary to trauma or adhesive arachnoiditis, results are less promising; postoperative recurrence of adhesions is the rule. Techniques include decompressive laminectomy-adhesiolysis and augmentation duraplasty in arachnoiditis cases, ventriculoperitoneal shunt for hydrocephalus, cyst extirpation in pseudomeningeal cyst, and both anterior and posterior decompression-fusion in the case of postlaminectomy kyphosis (Lee et al 2002). Olfactory ensheathing cells show promise in preclinical animal models as a cell transplantation therapy for repair of the damaged spinal cord (Mackay-Sim et al 2008).

Outcomes

Untreated syringomyelia tends to run a slowly progressive course.|{picture:mns7.jpg}{caption:End-stage syringomyelia, natural history}{label:Sagittal MRI scan from a nonsurgical 70-year-old male patient with slowly progressive syringomyelia starting at 30 years of age.

Special considerations

Pregnancy

Epidural anesthesia may promote the formation of subarachnoid or intramedullary cysts in previously healthy pregnant women (Nogués et al 1992).

Anesthesia

General anesthesia is preferred to epidural or spinal anesthesia. Nitrous oxide and isoflurane have been used safely (Murayama et al 2001).

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**References especially recommended by the author or editor for general reading.

ICD and OMIM codes

ICD codes

ICD-9:
Syringomyelia and syringobulbia: 336.0

ICD-10:
Syringomyelia and syringobulbia: G95.0

OMIM numbers

Syringomyelia, isolated: 6700
CM1 with syringomyelia: 8420

Profile

Age range of presentation

19-44 years
13-18 years
06-12 years
45-64 years
02-05 years

Sex preponderance

male=female

Family history

family history may be obtained

Heredity

heredity may be a factor

Population groups selectively affected

none selectively affected

Occupation groups selectively affected

none selectively affected

Differential diagnosis list

hydromyelia
cervical spondylitic myelopathy
motor neuron disease
progressive spinal muscular atrophy
multiple sclerosis
diabetes
leprosy
hereditary sensory neuropathies
spinal cord tumors
spinal syphilis
ulnar and median mononeuropathies

thoracic outlet syndrome
carpal tunnel syndrome
ulnar entrapment

Associated disorders

Arnold-Chiari malformation
Chiari I malformation
Ependymomas
Hemangioblastomas
Horner syndrome
Morvan syndrome

Other topics to consider

Central neuropathic pain
Chiari malformation
Ependymoma
Hemangioblastoma
Horner syndrome
Intramedullary spinal cord metastatic tumors
Sacral agenesis
Sleep and neuromuscular and spinal cord disorders
Sleep disorders associated with neuromuscular and spinal cord disorders