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Syringomyelia Following Spinal Cord Injury

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Key Points

Shunting of the syrinx cavity improves pain, motor function and sensory loss in some SCI patients post syringomyelia.

Untethering improves spasticity and motor and sensory loss post SCI-related syringomyelia.

Subarachnoid-subarachnoid bypass may improve motor and bladder functioning post SCI-related syringomyelia.

Cordectomy may improve motor and sensory function post SCI-related syringomyelia.

Cordectomy may improve quality of life post SCI-related syringomyelia.

Embryonic tissue transplantation may destroy syringomyelia cysts and improve sensory loss.

Further research is needed to determine the potential benefits of cellular therapy for the treatment of syringomyelia.

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1 Executive Summary

Post-traumatic syringomyelia refers to the formation of an intramedullary cyst filled with cerebrospinal fluid (CSF) within the spinal cord (Brodbelt & Stoodley, 2003). Though uncommon, its impact can be devastating following spinal cord injury (SCI). It can be seen as early as two months after injury, or many years later (Vernon, Silver, & Symon, 1983).

Epidemiology of Syringomyelia After Spinal Cord Injury

Syringomyelia occurs in approximately 2% of individuals with SCI (Klekamp & Samii 2002). There is a 22% higher incidence of syringomyelia found at autopsy than those presenting clinically (Vannemreddy et al. 2002). No relationship has been reported between the level of SCI and the likelihood of developing syringomyelia, however, an increased risk of post-traumatic syringomyelia has been reported in complete SCI individuals (Vannemreddy et al. 2002; Kramer & Levine 1997).

Pathophysiology and Clinical Presentation of Syringomyelia After Spinal Cord Injury

The pathophysiology of syringomyelia following SCI is not completely understood. The most supported theory is William's "Cranial-Spinal Pressure Dissociation Theory" which involves formation of the cavity and its enlargement and extension (Williams et al. 1981). Most commonly, it is thought to be asymptomatic. When it expands or compresses surrounding nerve tracts, is can cause radicular pain, gait ataxia, sensory disturbance, dysesthesias and motor weakness (Brodbelt & Stoodley 2003; Klekamp & Samii 2002; Kramer & Levine 1997; Lyons et al. 1987). As syringomyelia progresses, reduction in sensation and increased spasticity may be seen (Carroll & Brackenridge 2005). Progression is usually slow in most patients, with the clinical presentation remaining static for many years (Mariani et al. 1991).

How is Syringomyelia Diagnosed?

- Magnetic Resonance Imaging (MRI) is currently the diagnostic test of choice for syringomyelia.
- Myelography Enhanced Computed Tomography (CT-myelography). A significant improvement over plain CT, as it is able to show swelling and fixation of the cord and localized CSF flow obstruction (Dworkin & Staas 1985; Klekamp & Samii 2002).
- Virtual Endocopy (VE) by Computer Tomography allows non-invasive exploration of the spinal canal in all directions and can provide information regarding the extent of stenosis, which plain CT cannot provide. Ultrasonography is useful in localizing syrinxes, determining the safest place to open the dura, and facilitating optimal shunt placement during surgery (Brodbelt & Stoodley 2003).
- Intraoperative Somatosensory Evoked Potentials have limited value in assessing for syringomyelia, however may be used during surgery to prevent neurological damage.

Management Options for Syringomyelia

Medications	Used to manage presenting symptoms, but they do not treat the syringomyelia itself.
Surgery	Only recommended for patients with neurological deterioration or intractable pain (El Masry & Biyani 1996; Klekamp & Samii 2002). Commonly reported in the treatment of syringomyelia in SCI patients.
Shunting	Can be performed using syringoperitoneal, syringopleural, syringosubarachnoid, or ventriculoperitoneal shunts. Shunting improves pain, motor function and sensory loss in some SCI patients with syringomyelia (Karam et al. 2014; Ushewokenze et al. 2010; Schaan & Jaksche 2001; Falci et al. 1999; Lee et al. 2000; Lee et al. 2001; Hess & Foo 2001; Hida et al. 1994).High rate of shunt failure has been observed.
Untethering	Used to prevent or revise neurological or orthopedic sequelae. Untethering improves motor and sensory loss (Falci et al. 2009; Falci et al. 1999; Lee et al. 2000; Lee et al. 2001), and improves spasticity in more patients with syringomyelia than shunting (Lee et al. 2000; Lee et al. 2001).
Subarachnoid–Subarachnoid Bypass (S-S Bypass)	A new type of surgical technique for posttraumatic syringomyelia that may improve motor and bladder functioning (Hayashi et al. 2013).
Cordectomy	Used to manage spasticity, pain and improve neurological dysfunction; An invasive and irreversible procedure, it is only considered when other options have been exhausted (Gautschi et al. 2011).
Neural Tissue Transplantation	A novel treatment for syringomyelia that involves transplantation of neural tissue alone or in conjunction with surgical unthethering or cyst shunting. Embryonic tissue transplantation along with drainage, untethering and shunting may obliterate syringomyelia cysts and improve sensory loss (Falci et al. 1997; Wirth III et al. 2001). However, further investigation is required.

Gaps in the Literature

Causes of Syringomyelia: The exact incidence and etiology (tumor, infection, trauma, chiari malformation, etc.) is uncertain.

Signs and Symptoms: Symptoms of syringomyelia are variable and may include any of the following: radicular pain, gait ataxia, sensory disturbance, dysesthesias and motor weakness, reduction in sensation, and increased spasticity. These differ from the classic symptoms seen in syringomyelia patients.

Management: Indications for each treatment modality are limited. Therefore, determining when to consider untethering, bypass, etc., can become a challenge. Furthermore, complications of treatment modalities are sparce in the literature.

Prognosis: To our knowledge, there is no published literature that specifically outlines the prognosis of syringomyelia following SCI, nor is there evidence suggesting recurrence risk after each treatment.

2 Introduction

Post-traumatic syringomyelia is a term used to describe the formation of an intramedullary cyst (syrinx) filled with cerebrospinal fluid (CSF) within the spinal cord (Brodbelt & Stoodley, 2003). Though uncommon, its impact can be devastating following spinal cord injury (SCI). It can be seen as early as two months after injury, or many years later (Vernon, Silver, & Symon, 1983). The typical clinical features of syringomyelia are motor and sensory deficits which correlate to the level of the syrinx (Davidson, Rogers & Stoodley, 2018).

2.1 Epidemiology

Syringomyelia occurs in approximately 2% of individuals with SCI (Klekamp & Samii 2002). No relationship has been reported between the level of SCI and the likelihood of developing syringomyelia. Incidence rates are similar in individuals with either tetraplegia or paraplegia (Brodbelt & Stoodley 2003; Klekamp & Samii 2002; Ko et al. 2012). However, an increased risk of post-traumatic syringomyelia has been reported in complete SCI individuals (Vannemreddy et al. 2002; Kramer & Levine 1997). Among individuals with complete SCI, the occurrence of syringomyelia has been found to be more frequent in those with residual spinal deformity and or spinal canal compromise (Jamous et al., 2021).

2.2 Pathophysiology

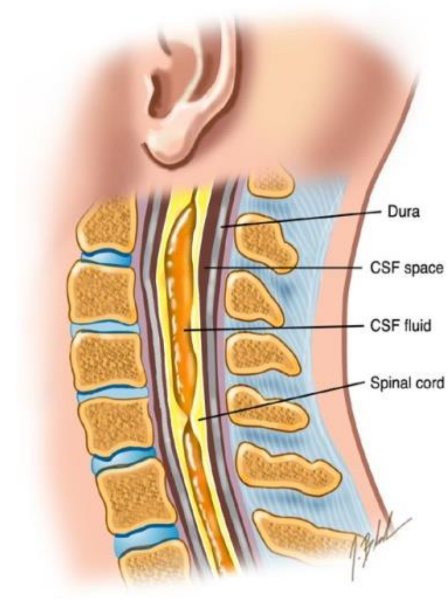

Figure 1. Cerebral spinal fluid filled cord (Elliot, 2008a)

The pathophysiology of syringomyelia following SCI is not completely understood. The most supported theory is William's "Cranial-Spinal Pressure Dissociation Theory" which involves formation of the cavity and its enlargement and extension (Williams et al. 1981). Initially, at the site of SCI, a cavity forms after liquefaction of cord tissue or hematoma (Biyani & El Masry 1994; Williams et al. 1981). The liquefaction and cyst formation at the site has been linked to microinfarcts and the release of cellular enzymes (Williams et al. 1981; Kao & Chang 1977). Cyst formation results in partial obstruction of cerebral spinal fluid movement, creating a pressure gradient between the intracranial space and spinal space (Sharma et al. 2006). The second phase, cyst enlargement and extension, is the result of this pressure gradient which has been linked to two mechanisms affecting fluid dynamics, "slosh" and "suck" (Biyani & El Masry 1994; Williams et al. 1981). The "slosh" is due to increased epidural venous pressure and CSF movement which are triggered by everyday activities such as coughing and sneezing (Williams 1992). This pressure causes areas of structural weakness in the cord leading to proximal and distal extension of the syrinx. The second mechanism "suck" is the result of a partial subarachnoid block. As the fluid is initially forced up due to increased epidural venous pressure, it returns slowly creating a pressure gradient across the partial subarachnoid block with negative pressure caudal to it (Biyani & El Masry 1994). This contributes to the syrinx formation and progression.

2.3 Clinical Presentation and Natural History

The classic symptoms of syringomyelia (i.e., suspended sensory loss, segmental weakness and burning) are often not present in individuals with SCI. Many individuals may lack symptoms in general or present with nonspecific symptoms that may be attributed to other complications of SCI such as spasticity, autonomic dysreflexia or neuropathic pain. Most commonly, symptoms include radicular pain, gait ataxia, sensory disturbance, dysesthesias or motor weakness (Brodbelt & Stoodley 2003; Klekamp & Samii 2002; Kramer & Levine 1997; Lyons et al. 1987). As syringomyelia progresses, reduction in sensation and increased spasticity may be seen (Carroll & Brackenridge 2005). Progression is usually slow in most patients, with the clinical presentation remaining static for many years (Mariani et al. 1991). According to a recent population-based study by Chen et al. (2020), the survival rate at 10-year follow-up after syringomyelia diagnosis in individuals with SCI was approximately 68.6%, which was comparable to that of individuals with SCI who did not have syringomyelia. Among individuals with syringomyelia post SCI, age ≥ 60 and <30 years, spinal cord or spinal canal operations within 1 year after syringomyelia diagnosis, history of pneumonia, and history of coronary heart disease were identified as risk factors for long-term mortality.

3 Diagnosis and Monitoring

3.1 Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) is currently the diagnostic test of choice for syringomyelia as it is able to detect fluid movement, syrinxes, and other abnormalities (Brodbelt & Stoodley 2003). MRI also has an important role in planning and monitoring outcomes of treatments. T_1-weighted (T_1W) and T_2-weighted (T_2W) images can be obtained from a MRI and allow for image contrast between different types of tissue (Enzmann 1991). Phase contrast with MRI can identify obstruction of the subarachnoid space and demonstrate

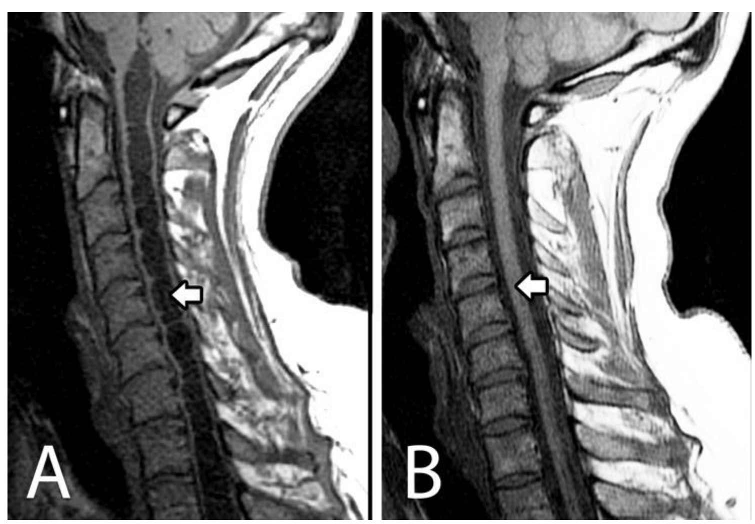

Figure 2. A) Abnormal cerebral spinal fluid due to syringomyelia. B) Normal cerebral spinal fluid flow post-surgery.

normalization of CSF flow following surgery (as seen in Figure 2; Brodbelt & Stoodley 2003). However, MRI is limited in its ability to differentiate posttraumatic syringomyelia from myelomalacia or tumor-associated syringomyelia (Biyani & El Masry 1994).

3.2 Myelography Enhanced Computed Tomography

Myelography-enhanced computed tomography (CT-myelography) of the spinal cord is a significant improvement over plain CT in the diagnosis of syringomyelia. Images from plain CT are considered unreliable due to imaging distortions of the surrounding bone (Klekamp & Samii 2002). CT-myelography is able to show swelling and fixation of the cord and localized CSF flow obstruction (Dworkin & Staas 1985; Klekamp & Samii 2002). As water soluble contrast accumulates in the cyst, CT-myelography can show the syrinx itself (Aubin et al. 1981). However, 10-50% of syrinxes may still be missed using this tool; therefore, MRI remains the diagnostic tool of choice (Brodbelt & Stoodley 2003).

3.3 Virtual Endoscopy by Computer Tomography

Improvements in CT technology have resulted in better three-dimensional imaging. Advanced computer graphics hardware and software have enabled the visualization of organs both inside and out. Virtual endoscopy (VE) is “a realistic 3D intraluminal simulation of tubular structures that is generated by post-processing of CT data sets” (Kotani et al. 2012, p. E752). Using this technology, surgeons can noninvasively explore the spinal canal in all directions. VE can provide information regarding the extent of stenosis, which plain CT cannot provide. It is useful for diagnosis, preoperative planning, and postoperative assessing.

3.4 Ultrasonography

Ultrasonography is usually confined to the intraoperative setting. It is useful in localizing syrinxes, determining the safest place to open the dura, and facilitating optimal shunt placement during surgery (Brodbelt & Stoodley 2003).

3.5 Intraoperative Somatosensory Evoked Potentials

Since the spinal cord is usually already significantly damaged, intraoperative somatosensory evoked potentials (SSEP) are of limited value in assessing for syringomyelia. However, it may be used during surgery to prevent neurological damage if action potentials can be obtained.

4 Management

Medications can be used to manage presenting symptoms but they do not treat the syringomyelia itself. Surgery is the only intervention which directly treats and resolves the syringomyelia. Surgery is only recommended for patients with neurological deterioration or intractable pain (El Masry & Biyani 1996; Klekamp & Samii 2002), although its use is commonly reported in the treatment of syringomyelia in SCI patients.

4.1Shunting

Shunting of the syrinx cavity can be performed using syringoperitoneal, syringopleural, syringosubarachnoid, or ventriculoperitoneal shunts. Most shunting procedures also involve laminectomies and duraplasties prior to the insertion of shunts (Figure 3).

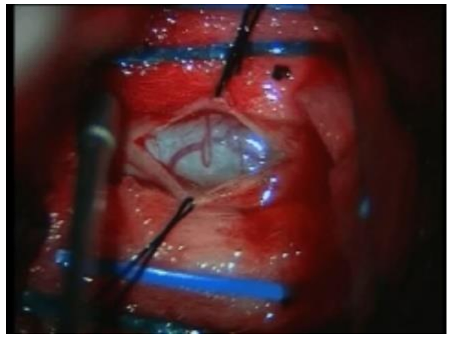

Figure 3. Drainage of syrinx followed by shunt insertion. (Elliot 2008b)

Table 1 Shunting

Author Year Country Research Design Score Total Sample Size	Methods	Outcome
Schaan & Jaksche 2001 Germany Cohort N=30	**Population:** Gender: males=21, females=9; Level of severity: complete=24, incomplete=6. **Intervention:** Patients with syringomyelia were divided into 3 groups: Group 1 (n=18) received single or multiple shunting procedures; Group 2 (n=5) received shunting procedures before surgical creation of a pseudomeningocele; Group 3 (n=7) was treated only with the surgical pseudomeningocele. **Outcome Measures:** Sensory and motor deficit, Pain, Syringobulbia.	1. There were no significant differences in outcomes between groups. 2. Prior to shunting (n=18) 15 had sensory deficits, 13 had motor deficits, 14 had pain and two had syringobulbia. Post-surgery: a. Sensory deficits improved in five, deteriorated in two, and were unchanged in eight patients. b. Motor deficits improved in five, deteriorated in two and were unchanged in four patients. c. Pain improved in five, deteriorated in four, and was unchanged in five patients. d. Syringobulbia improved in one and deteriorated in the second patient. 3. Prior to shunting and pseudomeningocele (n=5) five had sensory and motor deficits, one had pain and one had syringobulbia. Post-surgery: a. Sensory deficits improved in four and deteriorated in one patient. b. Motor deficits improved in three, deteriorated in one and was unchanged in one patient. c. Pain and syringobulbia improved in both patients. 4. Prior to pseudomeningocele only (n=7) seven patients had sensory deficits, six had motor deficits, three had pain, and one had syringobulbia. Post-surgery:

		a. Sensory deficits improved in six and were unchanged in one patient. b. Motor deficits improved in two, deteriorated in one, and were unchanged in three patients. c. Pain and syringobulbia improved in all patients.
Davidson, Rogers & Stoodley, 2018 Australia Pre-Post N=41	**Population:** Mean Age: 42 yr; Gender: males=19, females=22; Injury etiology: SCI Trauma=13, Arachnoiditis=4, Chiari=5, Other=19; Localization of syrinx: cervical=11, cervicothoracic=5, cervicolumbar=3, thoracic=18, thoracolumbar=2. **Intervention:** Syrinx to subarachnoid shunt. **Outcome Measure:** Reduction, stabilization, or recurrence of syringomyelia, self-reported quality of life (SF-12), clinical symptoms.	1. Ninety percent (n=32) of patients experienced a reduction in syringomyelia size at 3 mo follow-up. Post-operatively, eight patients had clinically stable syringomyelias, while one patient's condition was deemed clinically worse. 2. There were no significant differences in SF-12 post-surgery for physical (p=0.64) or mental (p=0.74) health. 3. The majority of patients had improved clinical symptoms (n=32) on pain, posterior column sensory loss, motor weakness, myelopathy, spasticity, and bladder dysfunction. Eight patient's symptoms remained stable, while one patient had an increased incidence of spasticity.
Tassigny et al., 2017 Belgium Pre-Post N=17	**Population:** Mean age: 43.3 yr; Gender: males=10, females=7; Injury etiology: SCI-trauma=5, arachnoid cyst=2, tumor=1, Chiari malformation=4, unknown=7; Localization of syrinx: cervical=1, cervicothoracic=5, thoracic=1, thoracolumbar=2, pan-medullaris=4, multiple=4. **Intervention:** Placement of myringotomy tube with drain. **Outcome Measure:** Syringomyelia clinical status, radiological status (shrinkage), reoperation.	1. Three patients experienced syrinx disappearance, the clinical status of eight improved, five stabilized, and one patient experienced clinically worse progression of syringomyelia. 2. Between one and three days post-operatively, an MRI was conducted to determine shrinkage. Eight patients experienced complete shrinkage, three almost complete, four a reduction is size, and two achieved stable size. 3. A total of three patients required recurrent surgery, one "surgical exploration" and two for a syringoperitoneal shunt.
Karam et al. 2014 Canada Pre-Post N=27	**Population:** Mean age: 40 yr; Gender: males=24, females=3; Injury etiology: MVA=16, Bicycle/Tractor/AirplaneAccidents=3, Falls & All-terrain Vehicles=3, Motorcycle Accidents=2; Severity of Injury: AIS A=14, C=3. D=10. **Intervention:** Sixteen patients underwent a shunting procedure, 14 of which received	1. Amongst the 16 patients who received a shunt only, 10 required revision surgery whereas only three of the 11 patients who received a shunt plus duraplasty required revisions; the difference between the groups was non-significant (p=0.0718). 2. No improvements in ASIA score from admission were reported. 3. Sequential MRI scans were available for 11 of the patients with a significant

	a syringo-subarachnoid procedure and two received a syringo-pleural procedure. 11 patients underwent duraplasty including seven patients who received shunting and lysis of adhesions. **Outcome Measures:** Odom Score, Level of syrinx.	correlation being found between clinical improvement assessed by the Odom Score and a reduction in the size of the syrinx ($p<0.001$). 4. Size of syrinx reduced from an initial 12±6 levels to 6±7 levels at follow-up.
Ushewokunze et al. 2010 UK Pre-Post N=40	**Population:** Mean age: 32 yr; Gender: males=38, females=2. **Intervention:** A laminectomy with the creation of a cerebrospinal fluid conduit and shunting. **Outcome Measures:** Adverse events, Stabilization rate.	1. Overall, 27 patients reported a stabilization of symptoms while 13 patients experienced a deterioration of symptoms. 2. At 6mo follow-up, a reduction in the size of syrinx was reported in 21 out of 33 patients. 3. Twenty-three patients required no further surgery while 17 patients underwent further surgery for deteriorating symptoms. 4. The most common early complications included pain (n=5), neurological deficit (n=4), wound infection (n=4), and CSF leak (n=2).
Li et al. 2021 USA Case Series N=34 (pre- and post- operative MRI for 24 patients)	**Population:** Mean age: 44 yr; Gender: males=15, females=9; TI=116; Severity of injury: AIS A=12, B=0, C=2, D=9, E=1. **Intervention:** Participants underwent surgical treatment consisting of syrinx fenestration, lysis of adhesions, and duraplasty. 21 of the patients underwent laminectomies, 21 patients underwent intradural lysis of adhesions, 17 underwent cyst fenestration, and 19 underwent duraplasty. In four patients, pre-existing syrinx shunts were removed; in two others, syrinx shunts (syringo-pleural and -subarachnoid) were placed. **Outcome Measures:** Syrinx length (measured in spinal segments), syrinx axial dimension (defined as the product of maximum syrinx transverse and antero-posterior dimensions), motor, sensory, bladder/bowel, and overall functional status.	1. The majority (15 of 24) off participants had cervical SCI. 2. The average syrinx length, measured in spinal segments, was similar when comparing pre- and post-operative MRIs (5.5 and 5.4 spinal segments, respectively). 3. Syrinx axial dimension was decreased in 16 of the patients postoperatively and stable or increased in the other eight. 4. Of the 24 patients, 23 remained clinically stable in regards to motor, sensory, and bowel/bladder function. 5. One patient, whose surgical treatment involved syringo-subarachnoid shunt placement, experienced neurologic deterioration; in this patient, motor function decline was noted postoperatively. 6. The change in syrinx size did not correlate with clinical outcomes.

Hess & Foo 2001 USA Case Series N=8	**Population:** Level of injury: T=2, C=5, L=1; Level of severity: AIS A/B=7, C=1; Mean time since injury: 10 yr. **Intervention:** Charts of patients who received shunts were assessed. **Outcome Measures:** Outcome of shunting, Complications.	1. A significant reduction in pain was reported by >80% of patients post-surgery; improvement in strength (n=6) and sensory (n=2) was also reported. 2. At follow-up, four patients had shunt failure resulting in neurologic decline, while two developed a new syrinx. 3. In one patient a new cavity was found with MRI, while the original remained decompressed.
Lee et al. 2001 USA Case Series N=45	**Population:** Mean age: 45.6 yr; Gender: males=30, females=15. **Intervention:** Records of patients who underwent surgical treatment for posttraumatic syringomyelia were assessed. Patients were divided into three groups: Group 1 underwent untethering only, Group 2 underwent shunting only, and Group 3 underwent both untethering and shunting. Patients were followed up to assess treatment efficacy. **Outcome Measures:** Improvement in symptoms, Magnetic Resonance Imaging (MRI), Complications.	1. There were no significant differences in outcomes between groups. 2. Patients in the surgical untethering group: a. Demonstrated improvement in motor and spasticity symptoms in the majority of patients (60% and 58%, respectively). b. Experienced one treatment failure and two complications. c. Revealed cyst re-accumulation at one yr follow-up. 3. The shunt only group experienced one complication and three treatment failures; 60% of patients in this group experienced improvement in gait followed by sensory (57%) and motor (54%). 4. Among those who underwent both untethering and shunting, 33% had clinical recurrence, one experienced CSF leak, and 50% showed improvement in motor symptoms.
Lee et al. 2000 USA Case Series N=34	**Population:** Mean age: 43.2 yr; Gender: males=23, females=11. **Intervention:** Records of patients who underwent surgical treatment for posttraumatic syringomyelia were assessed. Patients underwent laminectomies and a syringosubarachnoid shunt was inserted. Patients were divided into three groups: Group 1 underwent untethering only, Group 2 underwent shunting only, and Group 3 underwent both untethering and shunting. Patients were followed up to assess treatment efficacy.	1. At follow-up (>1 yr), 26 patients' resolution of one or more of the presenting symptoms was achieved post operatively; two patients experienced deterioration of motor function. 2. A decrease in spasticity was the most common improvement in patients who underwent untethering only (67%), followed by motor functioning (57%) and sensory loss (50%); this group experienced one treatment failure and two complications. 3. Improvement in gait was seen in the highest number of patients from the shunt only procedure group (60%), followed by motor (50%) and sensory loss (50%); in this group, two treatment

	Outcome Measures: Improvement in symptoms, Complications.	failures and two complications occurred. 4. Patients who underwent untethering and shunt procedures did not experience clinical reoccurrence; motor (67%) and gait (50%) improved in patients in this group.
Falci et al. 1999 USA Case Series N=59	**Population:** Mean age: 26 yr; Gender: males=49, females=10; Level of severity: AIS A=53, B=1, C=4, D=1. **Intervention:** All patients underwent spinal untethering and if a spinal cyst was present a lumbo-peritoneal shunt tube was placed along the length of the cyst. **Outcome Measures:** Pinprick, Motor and light touch scores, MRI findings, Somatosensory evoked potentials.	1. Participants with no previous surgery showed a significant increase in light touch (+2.38), pinprick (+3.88) and motor scores (+1.47) post-surgery. 2. Participants who had previous surgery had a decrease in touch, pinprick and motor score, although it was minimal (0.7, 0.8, and 0.5, respectively). 3. At 2wk post-surgery, MRI showed decreased cyst size or complete collapse. 4. Somatosensory evoked potentials were improved in amplitude compared to baseline; latency of 2 milliseconds or greater was observed in 27 patients.
Ronen et al. 1999 Israel Case Control N=10	**Population:** Mean age: 31.3 yr; Gender: males=10, females=0; Level of injury: C=5, L=4; Level of severity: incomplete=5, complete=5. **Intervention:** Charts of patients with syringomyelia were reviewed. Patients were divided into two groups: patients receiving rehabilitation only and patients receiving rehabilitation and shunting. **Outcome Measures:** Functional and neurological outcome.	1. Four out of five patients in the shunt surgery and rehabilitation group showed functional and neurological deterioration; the fifth patient remained unchanged. 2. Patients in the rehabilitation only group remained unchanged except for one who showed significant functional improvement without any change in neurological status.
Hida et al. 1994 Japan Case Series N=14	**Population:** Mean age: 48 yr; Gender: males=10, females=4; Level of injury: C=5, T=5, L=4. **Intervention:** Charts of patients who underwent syringosubarachnoid (n=6), syringoperitoneal (n=4), and ventriculoperitoneal (n=1) shunts were assessed. **Outcome Measures:** Neurological, motor, Sensory functioning, Shunt malfunction.	1. Neurological amelioration was obtained in all patients. 2. Of the nine patients with motor function difficulty, eight improved. 3. Sensory disturbance and relief of local pain or numbness improved in all patients. 4. Malfunction was reported in three of four syringoperitoneal shunts and in the one ventriculoperitoneal shunt.

In all of the studies, no shunting procedure was found to be superior to another. Schaan and Jaksche (2001) in a cohort study assessed the efficacy of syringomyelia treatment in three groups of patients: Group One received various shunts only, Group Two received shunting followed by surgical creation of a pseudomeningocele, and Group Three was treated with the pseudomeningocele only. The study found improvement in sensory and motor deficits, pain and syringobulbia in all three groups. However, more patients experienced greater pain post-surgery in the shunting only group than the other two groups. It should be noted that although groups did improve on some of the outcome measures, none of the groups were significantly different from each other.

Falci et al. (1999) demonstrated that untethering and shunt tube placement among individuals without prior surgery can significantly improve light touch, pinprick and motor scores. Two case series (Lee et al. 2001; Lee et al. 2000) further examined untethering and shunting treatment in individuals with syringomyelia. In the study by Lee et al. (2001), patients were divided into three groups: untethering only, shunting only, and untethering and shunting. Improvement in motor and sensory functioning was observed in all three groups, although the groups did not significantly differ from each other. In the first group, untethering only, improvement in spasticity was more common; while the shunting only group found gait improvement to be the most common. These results are also supported by Lee et al. (2000) earlier case series which found the same outcome measures were improved by the same corresponding intervention. Furthermore, shunting alone has been reported to significantly improve pain (Hess & Foo 2001; Hida et al. 1994; Davidson, Rogers & Stoodley, 2018), strength (Hess & Foo 2001), motor function (Hida et al. 1994; Davidson et al., 2018) and sensation (Hess & Foo 2001; Hida et al. 1994) in patients with syringomyelia, although, a high rate of shunt failure (36-50%) has been reported (Hess & Foo, 2001; Hida et al. 1994). Another case series by Li et al. (2021) investigated the radiographic and clinical outcomes following various surgical treatments, including syrinx fenestration, adhesion lysis, and duraplasty, in 24 individuals with syringomyelia following traumatic SCI. In four of the patients, pre-existing syrinx shunts were removed, and in two others, syrinx shunts were placed. It was found that all but one patient remained clinically stable regarding motor, sensory, and bowel/bladder function post surgery. The authors noted that the only patient who experienced deterioration in motor function post surgery had undergone syringo-subarachnoid shunt placement. Due to the lack of clarity in reporting post-surgical outcomes for each type of surgical intervention, as well as the small sample size, findings should be interpreted with caution (Li et al., 2021).

One case control study (Ronen et al. 1999) reviewed charts of patients receiving either rehabilitation only or rehabilitation and shunting for syringomyelia. The study found 80% of patients in the shunting and rehabilitation group experienced functional and neurological deterioration, while patients in the rehabilitation group remained either unchanged or improved. One must be careful when drawing conclusions from such a study because allocation to either group was dictated by receiving the treatment, which presumably was given to those patients already deteriorating or those were considered at a higher risk of deterioration.

Ushewokunze et al. (2010) studied the adverse events after laminectomy and shunting and reported a reduction in syrinx size among 21 of 40 patients and a stabilization of symptoms among 27 of 40. However, symptoms deteriorated for 13 individuals including pain, increased neurological deficits, infection and CSF leakage. In 17 individuals, a second surgical procedure was required to improve deteriorating symptoms.

Only one Pre-Post Study examined the use of a myringotomy tube (Tassigny et al., 2017). Of the 17 participants in the study, only one experienced clinically worse progression of the syrinx. Additionally, between days 1-3 post-surgery 15 patients experienced shrinkage of the syrinx, with eight of those being complete (Tassigny et al., 2017).

A Pre-Post study examining just shunting (Davidson et al., 2018) found that shunting alone was enough to reduce the size of the syrinx in up to 90% of patients. Although the majority of patients improved on clinical symptoms there were no significant differences between groups for quality of life. Finally, a study by Karam et al. (2014) revealed that patients who received both shunting and duraplasty were less likely to require reoperations (3/11 patients) compared with patients who received a shunt only with 10 of 16 requiring revisions and reoperations. However, this contrast was not statistically significant. Overall, 14 patients (52%) experienced an improvement in symptoms, 10 (37%) remained stable whilst three (11%) reported a progression of symptoms without improvement. In addition, a reduction in syrinx size was reported which correlated significantly with clinical improvement as measured by Odom Score. Only one patient experienced complications post-surgery with pain and dysphagia reported; however, it was later revealed the patient had developed another syrinx larger than before. The patients received another shunt and duraplasty which resulted in improvements reported at three-month follow-up.

Conclusions

There is level 2 evidence (from one cohort, four pre-post, and five case series studies; Karam et al. 2014; Ushewokenze et al. 2010; Schaan & Jaksche 2001; Falci et al. 1999; Lee et al. 2000; Lee et al. 2001; Hess & Foo 2001; Hida et al. 1994; Tassigny et al. 2017; Davidson et al. 2018) that shunting improves pain, motor function and sensory loss in some SCI patients with syringomyelia; however, a high rate of shunt failure has been observed; these findings are tempered by level 4 evidence (from one pre-post study; Karam et al. 2014) that reported minimal clinical improvement post shunting with or without duraplasty.

Key Points

Shunting of the syrinx cavity improves pain, motor function and sensory loss in some SCI patients post syringomyelia.

4.2 Untethering

Spinal cord tethering is commonly seen in patients with syringomyelia. A tethered spinal cord occurs when scar tissue forms and holds the spinal cord to its surrounding soft tissue membrane and dura. It has significant effects on spinal cord movement, CSF flow and extracellular fluid flow resulting in mobility issues and intramedullary pressure changes when exerting certain movements (Klekamp & Samii 2002). Untethering of the spinal cord is used to prevent or revise neurological or orthopedic sequelae.

Table 2 Untethering

Author Year Country Research Design Score Total Sample Size	Methods	Outcome
Falci et al. 2009 USA Pre-Post N=362	**Population:** Mean age: 40.5 yr; Level of injury: C6=163, C6-T1=83, T1=116; Level of severity: AIS A=229, B=36, C=41, D=54, E=2. **Intervention:** Surgical treatment for spinal cord untethering. **Outcome Measures:** Asia Impairment Scale (AIS) sensory and motor scores, Sensory and motor changes, Subjective report of changes post-surgery.	1. Sixty percent of the patients found an improvement in spasticity, 77% found an improvement in hyperhidrosis and 47% reported an improvement in neuropathic pain. 2. Most patients (86.5%) required only one surgery. 3. Progressive myelopathy regarding sensory and motor functions was arrested for an average of 3.3-3.4 yr post-surgery. 4. 89% of patients reported an arrest in loss of sensory and/or motor function post-surgery. 5. Return of function was reported in 46% of the patients.
Lee et al. 2001 USA Case Series N=45	**Population:** Mean age: 45.6 yr; Gender: males=30, females=15. **Intervention:** Records of patients who underwent surgical treatment for posttraumatic syringomyelia were assessed. Patients were divided into three groups: Group 1 underwent untethering only, Group 2 underwent shunting only, and Group 3 underwent both untethering and shunting. Patients were followed up to assess treatment efficacy. **Outcome Measures:** Improvement in symptoms, Magnetic Resonance Imaging (MRI), Complications.	1. There was no significant difference in outcomes between groups. 2. Patients in the surgical untethering group: a. Demonstrated improvement in motor and spasticity symptoms in the majority of patients (60% and 58%, respectively). b. Experienced 1 treatment failure and 2 complications. c. Revealed cyst re-accumulation at 1 yr follow-up. 3. The shunt only group experienced one complication and three treatment failures; 60% of patients in this group experienced improvement in gait followed by sensory (57%) and motor (54%). 4. Among those who underwent both untethering and shunting, 33% had clinical recurrence, one experienced CSF leak, and 50% showed improvement in motor symptoms.
Lee et al. 2000 USA Case Series	**Population:** Mean age: 43.2 yr; Gender: males=23, females=11. **Intervention:** Records of patients who underwent	1. At follow-up (>1 yr), 26 patients had resolution of one or more of their pre-operative symptoms; two patients experienced deterioration of motor function.

Author Year Country Research Design Score Total Sample Size	Methods	Outcome
N=34	surgical treatment for posttraumatic syringomyelia were assessed. Patients underwent laminectomies and a syringosubarachnoid shunt was inserted. Patients were divided into three groups: Group 1 underwent untethering only, Group 2 underwent shunting only, and Group 3 underwent both untethering and shunting. Patients were followed up to assess treatment efficacy. **Outcome Measures:** Improvement in symptoms, Complications.	2. A decrease in spasticity was the most common improvement in patients who underwent untethering only (67%), followed by motor functioning (57%) and sensory loss (50%); this group experienced one treatment failure and two complications. 3. Improvement in gait was seen most frequently in the shunt only procedure group (60%), followed by motor (50%) and sensory loss (50%); in this group, two treatment failures and two complications occurred. 4. Patients who underwent untethering and shunt procedures did not experience clinical reoccurrence; motor (67%) and gait (50%) improved in patients in this group.
Falci et al. 1999 USA Case Series N=59	**Population:** Mean age: 26 yr; Gender: males=49, females=10; Level of severity: AIS A=53, B=1, C=4, D=1. **Intervention:** All patients underwent spinal untethering and if a spinal cyst was present a lumbo-peritoneal shunt tube was placed along the length of the cyst. **Outcome Measures:** Pinprick, Motor and light touch scores, Magnetic Resonance Imaging (MRI) findings, Somatosensory evoked potentials.	1. Participants with no previous surgery showed a significant increase in light touch (+2.38), pinprick (+3.88) and motor scores (+1.47) post-surgery. 2. Participants who had previous surgery had a decrease in touch, pinprick and motor score, although it was minimal (0.7, 0.8, and 0.5, respectively). 3. At 2 wk post-surgery, MRI showed decreased cyst size or complete collapse. 4. Somatosensory evoked potentials were improved in amplitude compared to baseline; latency of 2 milliseconds or greater was observed in 27 patients.

In a pre-post trial, Falci et al. (2009) reported that spinal cord untethering resulted in improvement in neuropathic pain, sensory and motor functions. Falci et al. (1999) found untethering and shunt tube placement resulted in a significant improvement in light touch, pinprick and motor scores in SCI patients without previous syringomyelia surgery. Two case series (Lee et al. 2001; Lee et al. 2000) assessed the result of untethering only, shunting only, and untethering and shunting in a group of SCI individuals with syringomyelia. Both studies found that more patients in the untethering only group had improved spasticity than patients in the other groups. Motor function and sensory loss improvement was common in all three groups. However, the shunting group only demonstrated more improvement in gait than the untethering group.

Conclusions

There is level 4 evidence (from one pre-post and three case series studies; Falci et al. 2009; Falci et al. 1999; Lee et al. 2000; Lee et al. 2001) that untethering improves motor and sensory loss.

There is level 4 evidence (from two case series studies; Lee et al. 2000; Lee et al. 2001) that untethering improves spasticity in more patients with syringomyelia than shunting.

Key Points

Untethering improves spasticity and motor and sensory loss post SCI-related syringomyelia.

4.3 Subarachnoid–Subarachnoid Bypass

A new type of surgical technique for posttraumatic syringomyelia has been described in the literature, subarachnoid-subarachnoid bypass (S-S Bypass). Hayashi et al. (2013) hypothesized that reconstruction of the subarachnoid channels could re-establish CSF flow and therefore correct the underlying issue causing syrinx formation. In general, the S-S Bypass technique is accomplished by surgical laminectomy at the level of trauma, followed by a midline dural opening made under a microscope. One or two silicone tubes are inserted into the cephalic and caudal ends of the normal subarachnoid space; after a watertight dural closure, Bypass tubes are laid in the subdural space (Hayashi, 2013).

Table 3 Subarachnoid-Subarachnoid Bypass

Author Year Country Research Design Score Total Sample Size	Methods	Outcome
Hayashi et al. 2013 Japan Pre-Post N=20	**Population:** Mean age: 47.3 yr; Gender: males=19, females=1; Level of injury: thoracic=11, lumbar=5, cervical=4; Level of severity: complete=11, incomplete=9. **Intervention:** All patients underwent a laminectomy at the level of trauma with ventricular drainage tubes	1. Overall, 12 patient demonstrated clinical improvement, four remained stable and four showed deterioration of symptoms. Of the four who reported worsening symptoms, two improved after additional shunting but one reported no change. 2. There was a significant reduction in mean syrinx length from pre-surgery to post-surgery (p<.01).

Author Year Country Research Design Score Total Sample Size	Methods	Outcome
	inserted into the cephalic and caudal ends of the subarachnoid space. Bypass tubes were also inserted into the subdural space. Outcome Measures: Frankel Score (neurologic status), AISA motor score, Klekamp system (bladder function), Syrinx length.	3. A significant correlation was found between clinical outcome and change in the syrinx size whereby those who syrinx was reduced experienced clinical improvement (p=.01). 4. No significant correlation was found between preoperative and postoperative scores for either the ASIA Motor Score (59.6 versus 60.8 respectively) or Klekamp system for bladder function (1.1 versus 1.0 respectively).

A single pre-post study has assessed the effectiveness of S-S Bypass in 20 individuals (mean age=47.3 years, 19 males) with SCI-related syringomyelia (Hayashi et al. 2013). The mean time since SCI was 126 months (range 2-336 months) and they were followed up, on average, for 48.2 months (range 12-93 months). Post-surgery, 12 patients showed improvements, four remained stable, and four showed signs of deterioration. Three of the four patients who demonstrated deterioration underwent a shunt replacement; two improved and one remained unchanged. There was no significant correlation between ASIA scores at baseline and follow-up. Finally, no patient experienced a CSF leak that needed treatment (Hayashi et al. 2013). The authors conclude that S-S Bypass is not only an effective method in treating syringomyelia but that it may be associated with better clinical results than those of other surgical interventions (e.g., shunts, cordectomy). Hayashi et al. (2013) state that S-S Bypass can be conducted without myelotomy therefore reducing the risk of neurological damage and does not usually require performing arachnoid lysis which avoids the possibility of scarring. Although there is a risk of re-scarring or re-tethering, the bypass tubes prevent the obstruction of CSF flow caused by re-scarring (Hayashi et al. 2013). However, a potential methodological concern of this study was the use of a subjective grading approach to patient improvement, stabilization and deterioration. Further investigation from multiple studies is required to make conclusions as to its clinical effectiveness.

Conclusions

There is level 4 evidence (from one pre-post study; Hayashi et al. 2013) that subarachnoid-subarachnoid bypass may improve motor and bladder functioning post SCI-related syringomyelia.

Key Points

Subarachnoid-subarachnoid bypass may improve motor and bladder functioning post SCI-related syringomyelia.

4.4 Cordectomy

Cordectomy has been shown to be a useful procedure in the surgical treatment of syringomyelia. It can be used to manage spasticity, pain and improve neurological dysfunction. However, since it is an invasive procedure and irreversible, it is only considered when other options have been exhausted (Gautschi et al. 2011).

Table 4 Cordectomy

Author Year Country Research Design Score Total Sample Size	Methods	Outcome
Gautschi et al. 2011 Switzerland Pre-Post N=17	**Population:** Median age: 48.3 yr; Gender: males=15, females=2. **Intervention:** Patients received a spinal cordectomy. **Outcome Measures:** Quality of life using the EuroQol (EQ), Short Form-36 (SF-36).	1. Overall EQ scale improved from 42 points to 67 points (p=0.006) postoperatively. 2. The mental health score on the SF-36 improved significantly (p=0.01), whereas physical health score improved from 34.1 to 55.3 post-surgery (p=0.057). 3. The majority of patients (16/17) reported that they would undergo the operation again.
Ewelt et al. 2010 Switzerland Pre-Post N=15	**Population:** Mean age: 52 yr; Level of severity: AIS: A=7, B=8. **Intervention:** Patients received a spinal cordectomy. **Outcome Measures:** ASIA motor and sensory function, Visual Analogue Scale (VAS), Adverse events, Spasticity.	1. ASIA Scores: eight stabilized, four had improved motor and sensory scores, and one reported a progressive deterioration. 2. Pain: 10 patients stabilized and two improved. 3. Spasticity: nine patients stabilized, two improved, and four deteriorated. 4. No complications relating to surgery were reported.

Two pre-post studies examined the spinal cordectomy procedure on individuals with syringomyelia post SCI. Ewelt et al. (2010) found that cordectomy resulted in stabilization and improvement of motor and sensory function in 14 out of 15 patients. The study also reported improvement in pain and spasticity following the procedure. No significant adverse events were reported. Gautschi et al. (2010) reported a significant improvement in the quality of life of individuals who underwent the cordectomy procedure and high levels of subjective satisfaction.

Conclusions

There is level 4 evidence (from one pre-post study; Ewelt et al. 2010) that cordectomy improves motor and sensory function post SCI-related syringomyelia.

There is level 4 evidence (from one pre-post study; Gautschi et al. 2011) that cordectomy improves quality of life of individuals post SCI-related syringomyelia.

Key Points

Cordectomy may improve motor and sensory function post SCI-related syringomyelia.

Cordectomy may improve quality of life post SCI-related syringomyelia.

4.5 Neural Tissue Transplantation

A novel treatment for syringomyelia involves transplantation of neural tissue alone or in conjunction with surgical unthethering or cyst shunting. Embryonic spinal cord grafts have been shown to help repair structure and function of the spinal cord in experimental studies with an 80-90% survival rate (Houle & Reier 1988; Reier et al. 1988; Akesson et al. 1998). These grafts are used to fill the syrinx cavity and minimize cystic deformations in patients with progressive posttraumatic syringomyelia (Falci et al. 1997).

Two case reports (Falci et al. 1997; Wirth III et al. 2001), examined the use of embryonic tissue transplantation in the treatment of syringomyelia in patients post SCI. The studies involved untethering, cyst drainage and implantation of embryonic fetal tissue of SCI patients. Both studies reported collapse of cyst in the transplantation region and improvement in sensation. Improvement in spasticity was also observed, although it was short lived. Wirth III et al. (2001) demonstrated improvement in bladder functioning post-surgery, while Falci et al. (1997) showed significant improvement in deafferentation pain. At seven-month follow-up, MRI images showed no reoccurrence of the cyst in the transplantation region; however, improvement of secondary complications were not maintained (Falci et al. 1997). Further investigation using studies with more subjects is required to make conclusions as to its clinical effectiveness.

Conclusions

There is level 5 evidence (from two case reports; Falci et al. 1997; Wirth III et al. 2001) that embryonic tissue transplantation along with drainage, untethering and shunting may obliterate syringomyelia cysts and improve sensory loss.

Key Points

Embryonic tissue transplantation may destroy syringomyelia cysts and improve sensory loss.

4.6 Cellular Therapies

Cellular therapy is a relatively new treatment option, which contributes to significant advancements in technology and genetic research (Vaquero, Hassan, Fernandez, Rodriguez & Zurita, 2017). Cell transplantation with mesenchymal stromal cells via injection into the site of the syrinx shows potential in the future treatment of syringomyelia without surgery (Vaquero et al. 2017).

Table 5. Cellular Therapies

Author Year Country Research Design Score Total Sample Size	Methods	Outcome
Oraee-Yazdani et al., 2021 Iran Pre-Post N=11	**Population:** Mean age: 29.09±9.41 yr; Gender: males=9, females=2; Level of injury: cervical=4, thoracic=7; Severity of injury: ASIA A=11; Time since injury: ≥ 3 and ≤ 12 mo. **Intervention:** The patients received an intrathecal autologous combination of Mesenchymal stem cell (MSCs) and Schwann cells (SCs); and were followed up for 12 months. **Outcome Measures**: American Spinal Injury Association's (ASIA) sensory-motor scale, spinal cord independence measure (SCIM-III), subjective changes and adverse events (AE) (assessed by a checklist developed by the authors); electromyography (EMG), nerve conduction velocity (NCV), magnetic resonance imaging (MRI), and urodynamic study (UDS) were conducted for all the patients at the baseline, 6 mo, and 1 yr post-intervention.	1. Light touch AIS score alterations were approximately the same as the pinprick changes (11.6 ± 13.1 and 12 ± 13, respectively) in 50% of the cervical and 63% of the lumbar-thoracic patients, and both were more than the motor score alterations (9.5 ± 3.3 in 75% of the cervical and 14% of the lumbar-thoracic patients). 2. SCIM III total scores (21.2 ± 13.3) and all its sub-scores ("respiration and sphincter management" (15 ± 9.9), "mobility" (9.5 ± 13.3), and "self-care" (6 ± 1.4)) had statistically significant changes after cell injection (ps<0.05). 3. The most notable positive, subjective improvements were in trunk movement, equilibrium in standing/sitting position, the sensation of the bladder and rectal filling, and the ability of voluntary voiding. 4. Safety evaluation revealed no systemic complications, and radiological images showed no neoplastic overgrowth, syringomyelia, or pseudo-meningocele.

Author Year Country Research Design Score Total Sample Size	Methods	Outcome
Vaquero et al., 2018a Spain Pre-Post N=6	**Population:** Mean age: 39 yr; Gender: Males=6; Injury etiology: SCI-trauma=6; Mean time since injury: 13.7 yr; Level of severity: AISA-A=3, AISA-B=2, AISA-D=1; Lesion location: D5=2, D3=1, D4=1, D8=1, L1=1. **Intervention:** Cell therapy medicament (NC1, PEI number 12-141), developed by the Spanish Agency of Medicament and Health Products. The solution was injected into the syrinx over the course of one month. **Outcome Measures:** Alteration to genome of expanded cells, ASIA, SCI Functional Rating Scale of the International Association of Neurorestoratology (IANR-SCIFRS), Visual Analog Scale (VAS), Penn and modified Ashworth scale (MAS), Geffner scale, and neurogenic bowel dysfunction scale (NBD).	1. No genome alterations were detected during the cell expansion process. 2. Pin prick scores on the ASIA measure improved (p=0.06), this effect was only observed at 6 mo follow-up. 3. Scores on the IANR-SCIFRS (spinal cord function) increased at 3 mo follow-up (p=0.06), and 6 mo follow-up (p=0.06). 4. There were no significant differences in VAS score post-injection (p=0.25), although patients self-reported a decrease in neuropathic pain. 5. There were no significant differences in levels of spasticity or spasms experienced by patients post-injection (MAS, p=0.50). 6. The Geffner scale (bladder dysfunction) showed no significant differences post-injection (p=0.25). 7. The NBD scale showed no significant differences post-injection (p=0.12), although four patients observed an improvement.

To our knowledge, only one study has examined the use of cellular therapies for the treatment of syringomyelia in the SCI population. Vaquero et al. (2018) had a small patient sample of six, however they were able to observe strong improvements in spinal cord function at three- and six-months post-injection. Unfortunately, in terms of outcomes measuring bladder and bowel dysfunction no significant differences were reported. More studies are required on the use of cellular therapy as a potential non-surgical treatment for syringomyelia before any substantial conclusions can be drawn.

Cellular therapies have been used to improve various functional outcomes in individuals with SCI (Oraee-Yazdani et al., 2021). A recent study by Oraee-Yazdani and colleagues (2021) assessed the safety of intrathecal co-transplantation of autologous bone marrow mesenchymal and schwann cells in patients with subacute traumatic complete SCI. Radiological images revealed no presence of syringomyelia at 6-month and 12-month post-intervention.

Conclusions

There is level 4 evidence (from two pre-post studies; Vaquero et al., 2018a; Oraee-Yazdani et al. 2021) that mesenchymal cell therapy may be effective in improving spinal cord function post-SCI.

There is level 4 evidence (from one pre-post study; Oraee-Yazdani et al., 2021) that intrathecal co-transplantation of autologous bone marrow mesenchymal and schwann cells does not result in adverse events, such as syringomyelia, in individuals with subacute traumatic complete SCI.

Key Points

Further research is needed to determine the potential benefits of cellular therapy for the treatment of syringomyelia.

5 Summary

There is level 2 evidence (from one cohort, four pre-post, and five case series studies; Karam et al. 2014; Ushewokenze et al. 2010; Schaan & Jaksche 2001; Falci et al. 1999; Lee et al. 2000; Lee et al. 2001; Hess & Foo 2001; Hida et al. 1994; Tassigny et al. 2017; Davidson et al. 2018) that shunting improves pain, motor function and sensory loss in some SCI patients with syringomyelia; however, a high rate of shunt failure has been observed; these findings are tempered by level 4 evidence (from one pre-post study; Karam et al. 2014) that reported minimal clinical improvement post shunting with or without duraplasty.

There is level 4 evidence (from one pre-post and three case series studies; Falci et al. 2009; Falci et al. 1999; Lee et al. 2000; Lee et al. 2001) that untethering improves motor and sensory loss.

There is level 4 evidence (from two case series studies; Lee et al. 2000; Lee et al. 2001) that untethering improves spasticity in more patients with syringomyelia than shunting.

There is level 4 evidence (from one pre-post study; Hayashi et al. 2013) that subarachnoid-subarachnoid bypass may improve motor and bladder functioning post SCI-related syringomyelia.

There is level 4 evidence (from one pre-post study; Ewelt et al. 2010) that cordectomy improves motor and sensory function post SCI-related syringomyelia.

There is level 4 evidence (from one pre-post study; Gautschi et al. 2011) that cordectomy improves quality of life of individuals post SCI-related syringomyelia.

There is level 5 evidence (from two case reports; Falci et al. 1997; Wirth III et al. 2001) that embryonic tissue transplantation along with drainage, untethering and shunting may obliterate syringomyelia cysts and improve sensory loss.

There is level 4 evidence (from one pre-post study; Vaquero et al., 2018a) that mesenchymal stromal cell therapy may be effective in improving spinal cord function post-SCI.

There is level 4 evidence (from one pre-post study; Oraee-Yazdani et al., 2021) that intrathecal co-transplantation of autologous bone marrow mesenchymal and schwann cells does not result in adverse events, such as syringomyelia, in individuals with subacute traumatic complete SCI.

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Abbreviations

CSF	cerebrospinal fluid
CT-myelography	myelography-enhanced computed tomography
EQ	European quality of life
MRI	magnetic resonance imaging
SCI	spinal cord injury
SF-36	Short Form-36
S-S Bypass	subarachnoid-subarachnoid bypass
SSEP	somatosensory evoked potentials
VE	virtual endoscopy