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.
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# Abbreviations

<table>
<thead>
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<th>Abbreviation</th>
<th>Description</th>
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<tbody>
<tr>
<td>CSF</td>
<td>Cerebrospinal Fluid</td>
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<tr>
<td>CT-myelography</td>
<td>Myelography-enhanced computed tomography</td>
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<td>EQ</td>
<td>European Quality of Life</td>
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<td>MRI</td>
<td>Magnetic Resonance Imaging</td>
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<td>SCI</td>
<td>Spinal Cord Injury</td>
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<td>SF-36</td>
<td>Short Form-36</td>
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<td>S-S Bypass</td>
<td>Subarachnoid-subarachnoid bypass</td>
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<td>SSEP</td>
<td>Intraoperative somatosensory evoked potentials</td>
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<td>VE</td>
<td>Virtual Endoscopy</td>
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</table>
Syringomyelia Following Spinal Cord Injury

1.0 Introduction
Post-traumatic syringomyelia is a term used to describe 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). For example, Ko et al. (2012) reported that syringomyelia developed within five years after injury in 7.3% of patients.

1.1 Epidemiology
Syringomyelia occurs in approximately 2% of individuals with SCI (Klekamp & Samii 2002). Syringomyelia found at autopsy has a 22% higher incidence than those presenting clinically (Vannemreddy et al. 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). Lyons et al. (1987) reported that the development of syringomyelia occurred after a shorter interval in individuals with incomplete SCI than those with complete SCI; although, other studies have found no such relationship (El Masry & Biyani 1996; Ko et al. 2012; Vernon et al. 1982).

1.2 Pathophysiology
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, enlargement and extension, is the result of this pressure gradient. It has been linked to two mechanisms affecting fluid dynamics, ‘slosh’ and ‘suck’ (Biyani & El Masry 1994; Williams et al. 1981). The ‘slosh’ and ‘suck’ effects are due to increased epidural venous flow triggered by everyday activities such as coughing and sneezing (Williams 1992). Specifically, the ‘slosh’ is due to increased epidural venous pressure and CSF movement. 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.
1.3 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 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).

2.0 Diagnosis and Monitoring

2.1 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. T₁-weighted (T₁W) and T₂-weighted (T₂W) 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 normalization of CSF flow following surgery (as seen in Figure 2; Brodbelt & Stoodley 2003). MRI limited in its ability to differentiate posttraumatic syringomyelia from myelomalacia or tumor-associated syringomyelia (Biyani & El Masry 1994).

2.2 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).

2.3 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.

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

2.5 Intraoperative Somatosensory Evoked Potentials
Since the spinal cord is usually already significantly damaged, intraoperative somatosensory evoked potentials 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.

3.0 Management
Medications can be used to manage presenting symptoms but they do not treat the syringomyelia itself. 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.

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

Table 1 Shunting

<table>
<thead>
<tr>
<th>Author Year</th>
<th>Country</th>
<th>Research Design</th>
<th>Score</th>
<th>Total Sample Size</th>
<th>Methods</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Karam et al. 2014</td>
<td>Canada</td>
<td>Pre-Post</td>
<td>N=27</td>
<td>Population: Mean age: 40 yr; Gender: males=24, females=3; Injury etiology: MVA=16, Bicycle/Tractor/Airplane Accidents=3, Falls &amp; All-terrain Vehicles=3, Motorcycle Accidents=2; Severity of Injury: AIS A=14, C=3, D=10.</td>
<td>Intervention: 16 patients underwent a shunting procedure, 14 of which received a syringo-subarachnoid procedure and 2 received a syringopleural procedure. 11 patients underwent duraplasty including 7 patients who received shunting and lysis of adhesions. Outcome Measures: Odom Score, Level of syrinx.</td>
<td>1. Amongst the 16 patients who received a shunt only, 10 required revision surgery whereas only 3 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 correlation being found between clinical improvement assessed by the Odom Score and a reduction in the size of the syrinx (p&lt;0.001). 4. Size of syrinx reduced from an initial 12±6 levels to 6±7 levels at follow-up.</td>
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<td>Author Year</td>
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<td>Ushewokunze et al. 2010</td>
<td>UK</td>
<td>Pre-Post</td>
<td>N=40</td>
<td>Population: Mean age: 32 yr; Gender: males=38, females=2. <strong>Intervention:</strong> A laminectomy with the creation of a cerebrospinal fluid conduit and shunting. <strong>Outcome Measures:</strong> Adverse events, Stabilization rate.</td>
<td>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. 23 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).</td>
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<td>Hess &amp; Foo 2001</td>
<td>USA</td>
<td>Case Series</td>
<td>N=8</td>
<td>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. <strong>Intervention:</strong> Charts of patients who received shunts were assessed. <strong>Outcome Measures:</strong> Outcome of shunting, Complications.</td>
<td>1. A significant reduction in pain was reported by &gt;80% of patients post-surgery; improvement in strength (n=6) and sensory (n=2) was also reported. 2. At follow-up, 4 patients had shunt failure resulting in neurologic decline, while 2 developed a new syrinx. 3. In 1 patient a new cavity was found with MRI, while the original remained decompressed.</td>
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<td>Lee et al. 2001</td>
<td>USA</td>
<td>Case Series</td>
<td>N=45</td>
<td>Population: Mean age: 45.6 yr; Gender: males=30, females=15. <strong>Intervention:</strong> 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. <strong>Outcome Measures:</strong> Improvement in symptoms, Magnetic Resonance Imaging (MRI), Complications.</td>
<td>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 1yr follow-up. 3. The shunt only group experienced 1 complication and 3 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, 1 experienced CSF leak, and 50% showed improvement in motor symptoms.</td>
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<td>Schaan &amp; Jaksche 2001</td>
<td>Germany</td>
<td>Cohort</td>
<td>N=30</td>
<td>Population: Gender: males=21, females=9; Level of severity: complete=24, incomplete=6. <strong>Intervention:</strong> 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.</td>
<td>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 2 had syringobulbia. Post-surgery: a. Sensory deficits improved in 5, deteriorated in 2, and were unchanged in 8 patients. b. Motor deficits improved in 5, deteriorated in 2 and were unchanged in 4 patients.</td>
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<td>Author Year Country</td>
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<td>Lee et al. 2000 USA Case Series</td>
<td>Case Series</td>
<td>N=34</td>
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<td><strong>Population:</strong> Mean age: 43.2 yr; Gender: males=23, females=11.</td>
<td><strong>Intervention:</strong> 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.</td>
<td><strong>Outcome Measures:</strong> Improvement in symptoms, Complications.</td>
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<td>c. Pain improved in 5, deteriorated in 4, and was unchanged in 5 patients.</td>
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<td>d. Syringobulbia improved in 1 and deteriorated in the second patient.</td>
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<td>3. Prior to shunting and pseudomeningocele (n=5) 5 had sensory and motor deficits, 1 had pain and 1 had syringobulbia. Post-surgery:</td>
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<td></td>
<td>a. Sensory deficits improved in 4 and deteriorated in 1 patient.</td>
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<td>b. Motor deficits improved in 3, deteriorated in 1 and was unchanged in 1 patient.</td>
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<td>c. Pain and syringobulbia improved in both patients.</td>
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<td>4. Prior to pseudomeningocele only (n=7) 7 patients had sensory deficits, 6 had motor deficits, 3 had pain, and 1 had syringobulbia. Post-surgery:</td>
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<td>a. Sensory deficits improved in 6 and were unchanged in 1 patient.</td>
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<td>b. Motor deficits improved in 2, deteriorated in 1, and were unchanged in 3 patients.</td>
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<td>c. Pain and syringobulbia improved in all patients.</td>
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<tr>
<td>Falci et al. 1999 USA Case Series</td>
<td>Case Series</td>
<td>N=59</td>
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<td><strong>Population:</strong> Mean age: 26 yr; Gender: males=49, females=10; Level of severity: AIS A=53, B=1, C=4, D=1.</td>
<td><strong>Intervention:</strong> All patients underwent spinal untethering and if a spinal cyst was present a lumbo-peritoneal shunt</td>
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<td>1. At follow-up (&gt;1 yr), 26 patients resolution of one or more of the presenting symptoms was achieved post operatively; 2 patients experienced deterioration of motor function.</td>
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<td>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 1 treatment failure and 2 complications.</td>
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<td>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, 2 treatment failures and 2 complications occurred.</td>
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<td>4. Patients who underwent untethering and shunt procedures did not experience clinical reoccurrence; motor (67%) and gait (50%) improved in patients in this group.</td>
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<td>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.</td>
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<td>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).</td>
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</table>
A tube was placed along the length of the cyst.

**Outcome Measures:** Pinprick, Motor and light touch scores, MRI findings, Somatosensory evoked potentials.

<table>
<thead>
<tr>
<th>Author Year</th>
<th>Country</th>
<th>Research Design</th>
<th>Score</th>
<th>Total Sample Size</th>
<th>Methods</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ronen et al. 1999</td>
<td>Israel</td>
<td>Case Control</td>
<td>N=10</td>
<td></td>
<td>Tube was placed along the length of the cyst.</td>
<td>3. At two 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.</td>
</tr>
<tr>
<td>Hida et al. 1994</td>
<td>Japan</td>
<td>Case Series</td>
<td>N=14</td>
<td></td>
<td>Tube was placed along the length of the cyst.</td>
<td>1. 4 out of 5 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.</td>
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**Discussion**

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.

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. 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. Furthermore, shunting alone has been reported to significantly improve pain (Hess & Foo 2001; Hida et al. 1994), strength (Hess & Foo 2001), motor function (Hida et al. 1994) 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).
Further, a case study by Ghobrial et al. (2015) reported on a patient who had received T2 and T3 laminectomies followed by arachnoidolysis and syringopleural shunt placement. Despite immediate improvement in upper-extremity strength and myelopathy, the patient presented with a unilateral deltoid palsy. Electromyography and nerve conduction velocity conducted at a three-month follow-up revealed a C-5 palsy with a lesion in the C-5 nerve root. The cause for the palsy was speculated by Ghobrial et al. (2015) to be the result of critical traction on the C-5 nerve root after a positional change post-surgery with axial imaging revealing a dorsal shift at the C-4 and C-5 level. The authors acknowledged that such a consequence of syrinx decompression has not been reported in past literature; however, this could be due to the low prevalence of syringomyelia and spinal syrinx (Ghobrial et al. 2015).

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 who 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, neurological deficit, infection and CSF leakage. In 17 individuals, a second surgical procedure was required to improve deteriorating symptoms.

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, two pre-post, and five case series studies; Karam et al. 2014; Ushewokunze 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) 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.
3.2 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

<table>
<thead>
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<th>Author Year</th>
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<td>Falci et al. 2009 USA Pre-Post N=362</td>
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<td>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.</td>
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<td>1. 60% 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.</td>
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<td>Lee et al. 2001 USA Case Series N=45</td>
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<td>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.</td>
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<td>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 1 complication and 3 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, 1 experienced CSF leak, and 50% showed improvement in motor symptoms.</td>
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<td>Lee et al. 2000 USA Case Series</td>
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<td>Population: Mean age: 43.2 yr; Gender: males=23, females=11.</td>
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<td>1. At follow-up (&gt;1 yr), 26 patients had resolution of one or more of their pre-operative symptoms; 2 patients</td>
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Shunting of the syrinx cavity improves pain, motor function and sensory loss in some SCI patients post syringomyelia.
**Author Year Country Research Design Score Total Sample Size**

| 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. **Outcome Measures:** Improvement in symptoms, Complications. | 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 1 treatment failure and 2 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, 2 treatment failures and 2 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. | N=34 |  |

| 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. | Falci et al. 1999 USA Case Series N=59 |  |

**Discussion**

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.

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

3.3 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

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<td>Hayashi et al. 2013</td>
<td>Japan</td>
<td>Pre-Post</td>
<td>N=20</td>
<td>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 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), AIS motor score, Klekamp system (bladder function), Syrinx length.</td>
<td>1. Overall, 12 patient demonstrated clinical improvement, 4 remained stable and 4 showed deterioration of symptoms. Of the 4 who reported worsening symptoms, 2 improved after additional shunting but 1 reported no change. 2. There was a significant reduction in mean syrinx length from pre-surgery to post-surgery (p&lt;.01). 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).</td>
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Discussion

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.*

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<td>Gautschi et al. 2011</td>
<td>Switzerland</td>
<td>Pre-Post</td>
<td>N=17</td>
<td>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).</td>
<td>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.</td>
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<tr>
<td>Ewelt et al. 2010</td>
<td>Switzerland</td>
<td>Pre-Post</td>
<td>N=15</td>
<td>Population: Mean age: 52 yr; Level of severity: AIS: A=7, B=8. Intervention: Patients received a spinal cordectomy.</td>
<td>1. ASIA Scores: 8 stabilized, 4 had improved motor and sensory scores, and 1 reported a progressive deterioration. 2. Pain: 10 patients stabilized and 2 improved.</td>
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<td>Outcome Measures: ASIA motor and sensory function, Visual Analogue Scale (VAS), Adverse events, Spasticity.</td>
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**Discussion**

Two pre-post studies examined the spinal corpectomy procedure on individuals with syringomyelia post SCI. Ewelt et al. (2010) found that corpectomy 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 corpectomy procedure and high levels of subjective satisfaction.

**Conclusions**

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

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

**3.5 Neural Tissue Transplantation**

A novel treatment for syringomyelia involves transplantation of neural tissue alone or in conjunction with surgical untherethering 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 untherethering, 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.

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

4.0 Summary

Syringomyelia is a term used to describe the formation of an intramedullary cyst filled with cerebrospinal fluid within the spinal cord. The pathophysiology of syringomyelia following SCI is not completely understood. Magnetic resonance imaging is currently the diagnostic test of choice for diagnosing syringomyelia and is able to detect fluid movement, syringomyelia and other abnormalities.

There is level 2 evidence (from one cohort, one 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) 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.
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