TITLE: Treatment of Hydromyelia in Adults: A Review of Clinical Effectiveness and Guidelines

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CONTEXT AND POLICY ISSUES:

Hydromyelia occurs when the central canal (center of the spinal cord) is abnormally enlarged which can lead to the accumulation of cerebrospinal fluid (CSF).\(^1\)\(^,\)\(^2\) CSF normally flows around the spinal cord and brain.\(^3\) In hydromyelia, the result of the accumulation of fluid is a cystic cavity (syrinx) that is connected to the fourth ventricle of the brain.\(^2\)\(^,\)\(^4\) The syrinx may expand and elongate over time destroying the spinal cord and nerves.\(^4\) This results in a variety of symptoms such as scoliosis, pain, muscle weakness, loss of sensitivity to hot and cold, and headaches.\(^1\)\(^,\)\(^5\) Hydromyelia is usually seen in infants and children with hydrocephalus or birth defects like Chiari malformation II.\(^2\) Chiari malformation II is associated with a form of spina bifida; with Chiari malformation II, there is a malformed cerebellar herniation and often a malformed brain stem.\(^6\)

It has been reported that the term hydromyelia can be used interchangeably with syringomyelia\(^2\)\(^,\)\(^4\) as it can be difficult to distinguish between the two conditions.\(^4\) Syringomyelia is defined as a condition in which a patient has a fluid-filled cavity (syrinx) in the spinal cord, not necessarily at the fourth ventricle of the brain.\(^1\)\(^-\)\(^3\) It has also been reported that hydromyelia can be included (as a subset) within the condition syringomyelia.\(^1\) One study stated that about 10.0\% of syringomyelia cases are with fourth ventricle communication; which can be interpreted to mean that up to 10.0\% of syringomyelia cases are in fact, hydromyelia.\(^1\)

In syringomyelia, the syrinx is a closed cavity that expands and elongates over time, destroying the spinal cord and eventually may affect the nerve fibers that carry information from the brain and resulting in similar symptoms as hydromyelia.\(^3\) There are two major forms of syringomyelia. First, and in most cases, syringomyelia occurs in adults with Chiari malformation I (80.0\% to 85.0\% of syringomyelia cases\(^6\)); the symptoms often start to appear between the ages of 25 and 40.\(^3\) Chiari malformation I is a congenital malformation involving a smaller than average

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Cerebellar tonsillar descent which may cause a disruption to the flow of CSF such that the CSF enters into the spinal cord and forms a cavity. The second major form of syringomyelia develops as a complication from trauma, meningitis, hemorrhage, arachnoiditis, or an intramedullary tumour. When syringomyelia is a result of a complication, the syrinx develops in the segment of the spinal cord that has been damaged by one of these conditions. In the case of trauma, the cavity develops in the damaged segment of the spinal cord and symptoms may appear months or years after the injury.

Incidence of syringomyelia is approximately 8.4 cases per 100,000. No incidence or prevalence rates were reported for hydromyelia. Diagnosis for both hydromyelia and syringomyelia can be made through magnetic resonance imaging (MRI).

Surgery is often the treatment option for patients with hydromyelia when the child is experiencing mild to moderate neurological deficit. Treatment also involves shunting in order to drain the cyst. Treatment may not be required if there are no symptoms, the cyst is not progressing, or in elderly patients. For syringomyelia, surgery is the treatment of choice in patients who are symptomatic and in patients with large cystic cavities. The most common surgical treatment is decompression to the enlarged area of the cerebellar tonsils to restore normal flow to CSF. If the patient has syringomyelia without the Chiari malformation II, then the cavity itself may be decompressed. If there is a tumour, there will likely be a surgery to remove the tumour that is causing the syringomyelia.

It is unclear how to manage patients with hydromyelia or syringomyelia if surgery is not an option. There is a lack of information, protocols, and treatment algorithms available for non-surgical management of hydromyelia. This report was undertaken to seek evidence for non-surgical management of these disorders.

**RESEARCH QUESTIONS:**

1. What non-surgical treatment strategies exist for adult patients with hydromyelia?
2. What physiotherapy treatment is available for adult patients with hydromyelia?
3. What evidence-based guidelines exist for treating adults with hydromyelia?

**METHODS:**

A limited literature search was conducted on key health technology assessment resources, including OVID Medline, OVID Embase, EBSCOhost CINAHL, The Cochrane Library (Issue 1, 2009), University of York Centre for Reviews and Dissemination (CRD) databases, ECRI, EuroScan, international health technology agencies, and a focused Internet search. Results include articles published between 2004 and February 2009, and are limited to English language publications only. No filters were applied to limit the retrieval by study type.

HTIS reports are organized so that the higher quality evidence is presented first. Therefore, health technology assessment reports, systematic reviews, and meta-analyses are presented first. These are followed by randomized controlled trials, controlled clinical trials, observational studies, and evidence-based guidelines.
Due to the lack of clarity between hydromyelia and syringomyelia, literature on both disorders was included.

**SUMMARY OF FINDINGS:**

Two case reports were identified that focused on treating patients with syringomyelia. No relevant health technology assessments, systematic reviews, meta-analyses, randomized controlled trials, controlled clinical trials, or evidence-based guidelines were retrieved.

**Observational studies**

Patel *et al.* published a case report in 2005 on a patient with posttraumatic syringomyelia. The patient (23 years old) was experiencing pain in the left arm (shoulder to fingers) after a knee surgery with epidural. This had been the patient’s second knee surgery in 18 months. As a child, the patient had sustained multiple head trauma. After MRI scans, the patient was diagnosed with two syrinxes. Surgery was not an option for this patient; it was not stated why. The patient was treated with an antiepileptic and an anticonvulsant to treat painful neuropathy. Other treatments included an anti-inflammatory diet and a five week course of spinal manipulation therapy. After five months of treatment, MRI scans showed that both syrinxes had decreased in length and width. No adverse effects were observed. No conclusions regarding effectiveness of therapy were stated in the report.

Haas *et al.* published a case report in 2005 regarding non-surgical treatment of a 53 year old patient with syringomyelia. The patient presented with intractable pain and a spinal cord syrinx, diagnosed after MRI. The patient had nine years prior fallen 9 feet and landed on the head, upper back, and neck. No further information was provided. In 1995, the patient had decompression surgery that relieved the symptoms. The patient was treated using the “Clinical Biomechanics of Posture” protocol (a chiropractic protocol). The patient was treated 26 times in a three week period. Pain was reduced by 50.0% and posture improved (as evidenced by lateral cervical radiographs) from a 10 degree cervical lordosis (sway back) to a 30 degree lordosis. At one-year follow up, the patient showed stable improvement in both the reported pain level and the lordosis. The authors concluded that structural rehabilitation may have a positive effect on symptoms for patients with syringomyelia. There was no statement regarding size or condition of syrinx.

**Limitations**

There is an absence of rigorous literature for non-surgical treatment of hydromyelia and syringomyelia. Furthermore, there appears to be confusion in the literature as to the definitions of these medical conditions and whether they are interchangeable. This makes it difficult to summarize the recent evidence published on hydromyelia.

Furthermore, the vast majority of the literature focused on surgically treating syringomyelia with decompression, shunting, and laminectomy techniques. The study designs were overwhelmingly narrative reviews, case reports, and small retrospective case series.

Only two case reports on non-surgical treatment of patients with syringomyelia as a result of trauma were retrieved. The location was not at the fourth ventricle and can thus be concluded the cases were not hydromyelia.
No literature was found that identified other physical therapy strategies for treating hydromyelia or syringomyelia. No literature was found that provided guidelines or recommendations for non-surgically treating hydromyelia or syringomyelia.

CONCLUSIONS AND IMPLICATIONS FOR DECISION OR POLICY MAKING:

No literature on treatment that used the terminology “hydromyelia” was retrieved. When the literature was expanded to include syringomyelia, two case reports were found that focused on non-surgical treatment of syringomyelia. Both case reports used chiropractic care, in addition to other treatments, and found improvement in pain. It is important to note that because of the study design and multiple interventions, it is not possible to draw a conclusion about the cause and effect of manipulation therapy and therapeutic benefit. In addition, these findings may not be generalizable to a syrinx in the fourth ventricle, as would be seen in hydromyelia. Furthermore, no rigorous evidence was identified to support any non-surgical treatment of hydromyelia.

No other physical therapy treatments were mentioned in the literature. In addition, no information on evidence-based guidelines or clinical guidelines were retrieved. Because of the limited evidence identified, it is difficult to draw conclusions about non-surgically treating patients with hydromyelia. However, it should be noted that an absence of evidence does not imply an absence of effect.

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