

REVIEW ARTICLE

ACTIVE CONSERVATIVE MANAGEMENT OF PRIMARY SPINAL SYRINGOMYELIA: A SCOPING REVIEW AND PERSPECTIVES FOR AN ACTIVITY-BASED CLINICAL APPROACH

Line OLSSON, MSc^{1*}, Wolfram ANTEPOHL, MD, PhD^{2*} and Sophie L. RAVN, PhD^{1,3}*From the ¹Specialized Hospital for Polio and Accident Victims, Roedovre, Denmark, ²Department of Rehabilitation Medicine, Region Östergötland & Department of Medicine and Health, Linköping University, Sweden and ³Department of Psychology, University of Southern Denmark, Denmark*

Objective: This scoping review aimed to identify and synthesize existing research on active conservative management of primary spinal syringomyelia and associated symptoms and to discuss perspectives for clinical application using an activity-based approach.

Methods: PubMed, Embase, Scopus, and Web of Science were systematically searched for empirical studies of conservative management or therapies of adults with primary spinal syringomyelia from inception to April 2021. In addition, abstracts from relevant conferences were searched. Study characteristics and key findings were extracted, and findings descriptively synthesized.

Results: Of 1,186 studies screened, 7 studies met the eligibility criteria (4 single case studies and 3 cohort studies, a total of 90 individuals). The interventions were primarily physiotherapeutic, mostly by posture correction and exercises, and effects were alleviation of pain, improved physical function, improved activities of daily living and quality of life. Analysis of factors triggering symptoms and rationale for choice of intervention based upon these was limited.

Conclusion: Evidence of active conservative management of primary spinal syringomyelia and associated symptoms is limited. Many variations and limitations in the existing research limit the conclusions. High-quality research is needed to enable healthcare professionals to apply evidence-based active conservative interventions.

Key words: syringomyelia; syrinx; spinal cord injury; conservative management; activity; exercise; rehabilitation; treatment.

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Correspondence address: Line Olsson, The Specialized Hospital for Polio and Accident Victims in Denmark, Fjeldhammervej 8, 2610 Rødovre, Denmark. E-mail: lol@specialhospitalet.dk

* These authors contributed equally to this work.

LAY ABSTRACT

People with primary spinal syringomyelia and associated symptoms are often managed conservatively. In most cases, this means without active treatment. Pathophysiological knowledge and our own physiotherapeutic experience propose that the symptoms may be connected to specific active movements and activities of daily life. The aim of this study was to map existing evidence for active rehabilitation as a means of reducing symptoms and preventing syrinx progression. Only 7 low-quality research studies with different kinds of treatment were found. Mostly, physiotherapy was offered with exercises and focus on posture. Outcomes included alleviation of pain as well as improved physical function and quality of life. Unfortunately, analysis of symptom-inducing factors to guide the choice of intervention was limited. Future high-quality studies are needed to develop active conservative interventions for people with primary spinal syringomyelia.

Syringomyelia is a disorder characterized by development of a longitudinal cavity (syrinx), filled with cerebrospinal fluid (CSF) (1) within the spinal cord parenchyma or the central canal. It may occur as a phenomenon related to congenital malformation, such as Arnold-Chiari malformation (2) or dysraphism (3). Alternatively, syringomyelia may develop as a complication secondary to previous spinal trauma (post-traumatic syringomyelia), tumours, haemorrhages, infectious or inflammatory processes, or degenerative spinal disease, with or without prior symptoms of spinal cord injury (SCI) (1). We refer to these forms of syringomyelia as primary spinal syringomyelia (PSS), as proposed by Heiss et al. (4). Localization of a syrinx is possible at all levels of the spinal cord, and it may expand rostrally and/or caudally (5). However, it develops most frequently at the cervical and thoracic levels of the spinal cord and in cases of a complete SCI (6).

The estimated prevalence of syringomyelia in the general population ranges from 9 to 133 per 100,000

(7). Among individuals with SCI, an MR-diagnosed syringomyelia can be found in 21–28% of cases, with 1–9% being symptomatic (7). The most common and characteristic clinical signs are neuropathic pain, sensory disturbance, loss of motor function, spasticity, and fatigue (6). Symptoms and severity can vary greatly among patients, and there is no correlation between cavity size and magnitude of clinical features (6). Symptoms may occur from weeks, up to several decades after the original SCI, with a median of 15 years post-injury (6).

In terms of pathophysiology, there is no agreement about the direct or indirect mechanisms behind syrinx formation and progression, but an alteration of the physiological CSF circulation dynamics in the spinal subarachnoid space (spinal subarachnoid block) has been suggested as an underlying mechanism (4, 8). Thereby, increased CSF pressure with possible enlargement and extension of the syrinx may lead to compression of the surrounding nerve tissue over time, thus causing progressive myelopathy. Clinically, this corresponds to the fact that the Valsalva manoeuvre, as well as common activities, such as coughing, sneezing, and heavy lifting, increase intrathoracic and intra-abdominal pressure and may trigger symptoms (4, 9). While many theories agree that pressure changes are central, there is no agreement on how these pressure changes are conveyed to the syrinx cavity. One of the concepts frequently cited is the “slosh-and-suck theory” formulated by Williams (10). More recently, Greitz (11) put forward the “intramedullary pulse pressure theory”, highlighting the role of pulsations rather than pressure gradients. One common denominator in the pathogenesis of PSS appears to be arachnoiditis (often due to bleeding, infectious or inflammatory processes, or tumours) causing the formation of streaks of connective tissue in the subarachnoid space that impact CSF circulation and/or cause spinal cord distension, which then may trigger symptoms and expansion of the syrinx (12). Despite these theories, the underlying mechanisms for both the formation of a syrinx and the progression (and sometimes reduction) in syrinx volume are still a topic of ongoing research and debate (13).

Progressive neurological deterioration due to PSS may have life-long disabling consequences for many of the patients (6, 14). Most often, a conservative approach is offered as a first line-treatment after diagnosis, but in cases of increasing symptoms and rapid deterioration of neurological function, surgery is often the treatment of choice (1). Different surgical procedures exist, but syringes unfortunately reoccur in up to approximately 50% of patients after 5 years (1). Hence, alternative conservative treatments appear highly relevant. However, Kleindienst et al.

(1) concluded in a recent systematic review that no consensus regarding conservative treatment of post-traumatic syringomyelia was available. Forty-three of the patients managed surgically experienced clinical improvements and 50% had symptom stabilization. Among the patients managed conservatively, symptom stabilization was attained in 88% and 2% improved clinically with symptom reduction (1). In addition, more symptom stability was reported by Ronen et al. (15) for 5 patients receiving conservative treatment compared with 5 patients who underwent surgical treatment. Roy et al. (16) concluded in their review that conservative management was superior to surgical intervention in the setting of idiopathic or traumatic aetiology. In summary, this might indicate that conservative management might be as good as or better than surgery for selected patients, but no randomized controlled trial with comparison between those 2 patient groups is completed. Unfortunately, no descriptions of the content of such conservative interventions were provided in either of these papers (1, 16) leaving it unclear whether “conservative management” means “doing nothing” or consists of an actual active intervention. Hence, there is a great need to increase knowledge about active conservative interventions with focus on the exact content of interventions to aid both clinicians and researchers.

Activities of daily living and physical training for a patient with PSS may have a physiological impact on CSF pressure and syrinx and thereby the patient’s symptoms. One of the authors (LO) is a clinical physiotherapist with relevant clinical experience of conservative management of this patient group from an activity-based perspective. Clinically, this approach has shown positive effects in the form of stabilized or reduced symptom load and improved quality of life (QoL). It is essential to link anamnestic information and clinical analysis based on symptom provocation tests, and thereby plan the interventions according to the goals of rehabilitation. However, such an approach needs to be based on evidence.

Based on the outlined background, a scoping review of existing evidence on active conservative management of PSS, including the specific therapeutic content, is relevant. Therefore, the aims of the current study were: to identify and synthesize existing research on active conservative management of PSS and associated symptoms, focusing on content of intervention, measures, and results, and to discuss perspectives for a future activity-based approach.

METHODS

This study is a scoping review inspired by the Preferred Reporting Items for Systematic reviews

and Meta-Analyses extension for Scoping Reviews (PRISMA-ScR) Checklist (17). An internal protocol was produced, outlining the study method. Details are described below.

Search strategy

To identify potentially relevant material, 4 bibliographic databases were searched on 13 January 2019: PubMed, Embase (via Ovid), Scopus, and Web of Science. A search string was outlined with relevant search words combined with the Boolean logic operators “AND” and “OR”. The search string was developed by the authors and piloted and refined prior to the final search. The following search string was applied in all databases:

(chiropract* OR rehabilitati* OR conservative* OR ADL OR physiotherap* OR “occupational therap*” OR exercise* OR training OR “physical therap*”)

AND

(syringomyeli* OR syrinx OR hydromyeli* OR myelopathi* OR (SCI AND cyst) OR (“spinal cord injur*” AND cyst))

The screening strategy had a broad search string to explore a wide range of studies. The searches were carried out without date limitations. The final outputs from the different databases were exported to and combined in EndNote, where duplicates were manually removed. All unique studies were then transferred to Covidence, an online tool for carrying out systematic reviews. An updated search was performed on 6 April 2021, limited to literature published from 14 January 2019 to 6 April 2021. Records from the updated search were handled in the same manner as the original search. In addition to the systematic searches in bibliometric databases, several additional steps were carried out. First, the Physiotherapy Evidence Database (PEDro) was searched for the words “syrinx” and “syringomyelia” to determine if any potentially relevant studies came up. Secondly, different combinations of the search words were searched using Google and Google Scholar. Thirdly, the same approach was used for ResearchGate. These 3 additional steps were repeated several times until November 2021. Fourthly, the monograph “Syringomyelia – Diagnosis and Treatment” by Klekamp and Samii (18) was scanned for relevant publications. Fifthly, abstract books from relevant spinal cord injury conferences (NoSCoS 2017 and 2019 as well as ISCoS 2017, 2018, 2019, and 2020) were searched for potentially relevant abstracts. An attempt was made to contact all authors of such abstracts to ask for publication status of their data, if full articles could not be found. This approach was also carried out for potentially relevant abstracts identified in the systematic searches. Sixthly, all references in the included

studies and all identified reviews were searched. These steps were performed to determine if there was any literature not identified in the systematic searches.

Eligibility criteria

All search results underwent the same screening procedure using the same eligibility criteria. Studies that fulfilled the following criteria were included:

- Studies in adult individuals (≥ 18 years of age) with symptomatic PSS, i.e. syringomyelia neither related to Arnold Chiari I malformations nor congenital conditions (e.g. spinal dysraphism).
- Studies published in Danish, Dutch, English, French, German, Norwegian, Spanish, or Swedish.
- Studies conducted in samples receiving active conservative interventions or therapies for syringomyelia-related symptoms, defined as non-surgical and non-pharmacological management (e.g. physiotherapy or chiropractic intervention) potentially in combination with medication, but not in combination with surgical procedures. Of note, for this criterion to be fulfilled it was not enough to only mention “conservative management”. Rather, studies had to explicitly mention “active” or describe an intervention deemed active by LO and WA, such as rehabilitation or training. In cases of combined samples (e.g. a part of the sample receiving active conservative management and others surgery), only analyses in the relevant subsample were included.
- Peer-reviewed studies with original data (hence, no conference abstracts, reviews, editorials, or so forth).

Screening procedure

The screening procedure for database search results consisted of 2 steps using the above-outlined eligibility criteria. First, 2 authors (LO and WA) independently screened titles and abstracts of all unique studies. Disagreements and doubts were handled through discussion among them. In cases of doubt, studies were transferred to the next screening step. Then, the same authors independently evaluated the remaining studies using full texts. Again, cases of disagreement or doubt were resolved through discussion. Here, SR was included in some of the discussions. Reasons for exclusion at this point were registered. The remaining studies were the ones included in the scoping review.

Data extraction

Authors LO and WA worked together to extract the following data from the included studies in a predefined table containing the following information:

- Author(s), year of publication and country of origin
- Study design
- Study sample and sample size
- Level and cause of syrinx
- Clinical problem
- Study intervention
- Measures and results
- Key conclusions

Data synthesis

A descriptive and narrative synthesis of the included studies was performed to provide a full overview of the findings. This overview formed the basis for an in-depth discussion of the clinical perspectives and implications of the findings in the discussion section of the present paper.

RESULTS

Screening flow

The first searches yielded a total of 1,786 records, of which 990 were unique. A total of 78 papers were passed on to full-text screening, of which 7 studies fulfilled all eligibility criteria and were included. During full-text screening the main reasons for exclusion were wrong intervention type (no explicit active conservative intervention) and wrong publication type. The updated searches yielded a total of 311 additional hits across databases with 196 unique records. After screening, no additional studies were included based on the outlined eligibility criteria. The flowchart of the screening process is shown in Fig. 1.

Descriptive characteristics

All study samples were independent, and studies were of different designs. Three of the identified studies were retrospective cohort studies using clinical data, interviews, and questionnaires (15, 19, 20), while 4 were case reports (21–24). Sample sizes varied from a single case (the 4 case studies) to 5, 14, and 67 patients who were managed conservatively in the 3 cohort studies, respectively, which in addition also reported on surgically managed patients. Patients' age ranged from 18 to 80 years, and, for the 4 studies reporting on this (21–24), syrinx locations included all parts of the medulla, often spreading over several levels. While the 2 biggest studies had mixed populations in relation to underlying pathology (both PSS and others) (19, 20), the study by Ronen et al. (15) described 5 cases that all had PSS. Among the 4 single case studies, 1 described PSS related to a specific condition (Hirayama disease)

(21), and 3 referred to patients with PSS after trauma (22–24). The most frequently reported symptoms were pain, muscle weakness, and loss of sensation, as well as impaired activities of daily living (ADL) and lower QoL. The extracted descriptive information is succinctly summarized in Table I.

Study intervention content

The 7 studies varied in their description of intervention content. One case report (24) and 2 cohort studies (15, 19) merely presented the active conservative interventions as “multidisciplinary rehabilitation”, “physiotherapy”, or “training” without any descriptions of the content of these concepts as applied in the study setting. The 3 remaining case studies (21–23) provided a rather detailed description of the active conservative interventions performed, 2 categorized as physiotherapeutic (21, 22) and 1 as chiropractic treatment (23). A common feature in all 3 studies was a focus on posture corrections and training of muscle stability to avoid possible affection of the medulla and thus the syringomyelia symptoms. The intervention in Güler et al. (22) was also aimed at strength and balance, while Arooj et al. (21) also described intervention elements, such as avoidance of flexion, application of cervical collar, and training to improve muscle tone. The remaining cohort study by Smith et al. (20) referred to a variety of physiotherapeutic interventions, as reported by participants of their study. Participants had received either a single intervention or a combination of these.

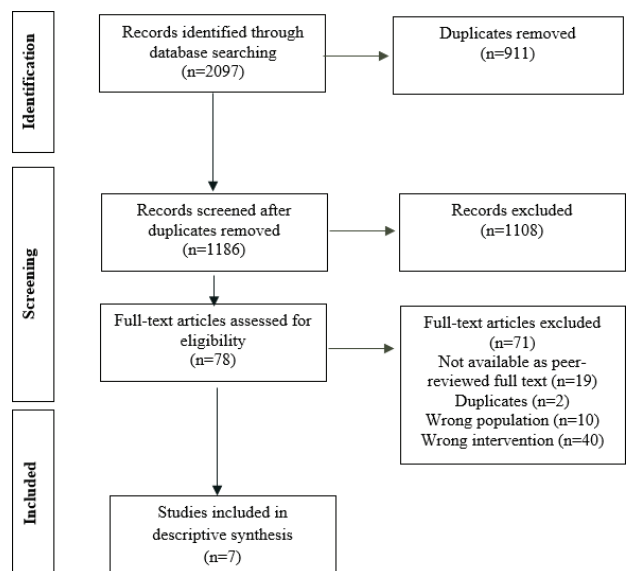


Fig. 1. Flowchart of the study selection process in total for searches 1 and 2. Two reviewers independently screened each paper against title and abstract. When no information was found to cause exclusion, each individual study was categorized and underwent full-text screening at the later stage.

Table I. Data extraction of the included studies

Author, year (ref)	Study Design (n)	Sex and age	Level and cause of syrinx	Clinical problem	Study intervention	Measures and results	Key conclusions
Ronen et al. 1999 (15)	Retrospective cohort study (n = 5) ^a	Men (100%), mean age 31 (range 14–58)	No description of levels of syrinx. Spinal traumas	Pain, numbness, sensory deficits, muscle weakness, decreased tendon reflexes, in-creased spasticity	Physical exercises without further description of content	Evaluation based on patient files showed neurological stability and functional improvement.	Conservative rehabilitation with training is recommended with clinical and MRI follow-up.
Sixt et al. 2009 (18)	Retrospective cohort study (n = 67) ^a	Men (100%), 14–74 year of age ^b	No description of levels of syrinx. Mixed population of AC, spinal trauma, spinal tumours/cysts, meningitis, scoliosis and others	Pain, numbness, gait ataxia, paresis, headache, vertigo, bladder disturbance, skeletal deformation, swallowing disorder	Physiotherapy without further description of content	Evaluation of QoL and functional disability by SF-36 and SDI showed no difference in QoL in summary scores (PCS, MCS & SDI) between surgical or active conservative treatment.	Diagnosis and treatment of the pathology is recommended to reduce expansion of the syrinx cavity. Maintenance of QoL and coping mechanisms is essential in both intervention groups to reduce symptom intensity and duration. Recommendation for physical and psychotherapy.
Smith et al. 2015 (19)	Retrospective cohort study (n = 14) ^a	Men (51%), 21–70+ years of age ^b	No description of levels of syrinx. Mixed population of congenital, idiopathic and primary spinal syringomyelia	Pain, weakness, and fatigue reported as the most frequent symptoms	Physiotherapy with stretching, hydrotherapy, exercises, massage, TENS, acupuncture as separate interventions or in combination.	Self-reported patient questionnaire, informed by SDI, Oswestry Index and NDI, showed beneficial effects of physiotherapy in a majority with symptom deterioration over time, in 64% of respondents. Semi-structured telephone interviews by categorical content analysis approach showed a beneficial effect of physiotherapy for symptom management and QoL in 75% of patients.	Physiotherapy has the capability to reduce symptoms and improve QoL, with pain relief as the most common treatment goal. More knowledge of syringomyelia by the physiotherapists and the patients is appropriate in the conservative management.
Arooj et al. 2013 (20)	Case report (n = 1)	A 25-year-old man	Syrinx C5–C7/Myelopathy	Increasing muscle weakness in both hands for 5 years	Physiotherapy by application of cervical collar and correction of posture, avoidance of neck flexion, exercises to improve muscle tone and grip.	<p>“No significant correlations between either physiotherapy and symptom progression (rs = 0.84; p = 0.57), surgical intervention and symptom progression (rs = 0.27; p = 0.06, or physiotherapy and pain (rs = 0.16; p = 0.29)</p> <p>Evaluation by dynamic MR showed that neutral cervical posture does not reveal any significant cord compression related to symptoms, while MR scan in flexed position does.</p> <p>Avoidance of neck flexion and cervical muscle tone stability reduce symptoms and cessation of muscle weakness progression</p>	Dynamic MR scan in flexion and extension may be essential in symptomatic patients to reveal cord compression in change of posture.
Guler-Uysal et al. 1996 (21)	Case report (n = 1)	A 62-year-old man	Syrinx T6–conus medullaris. SCI after traffic accident 25 years earlier	Aggressive physical symptoms with decline from walking to wheelchair dependence in 12 months	Physiotherapy with strengthening of abdominal and back muscles, posture corrections and standing balance training.	Results described as maintenance of good posture and achievement of gait function with aids after 4 months.	Follow up to check for a syrinx formation is recommended after vertebral fractures

(Continued)

Table 1. (contd.)

Haas et al. 2005 (22)	Case report (n = 1)	A 41-year-old man	Syrinx C2–C4. Fall 9 years earlier with surgical intervention shortly after	Cluster headaches, neck pain, thoracic pain, back pain, arm/hand numbness and knee pain	Chiropractic manipulative therapy with adjustment of abnormal postural position of cervical lordosis by thrust, traction and exercises over 3 weeks.	Follow-up MRI after 1 year showed improved cervical mobility. Clinical examination showed no strength reduction. Improvement of posture and overall symptoms. Pain scale showed 50% decrease in pain severity	Reduction of abnormal postures may have decreased the amount of stress and strain on tissues including the spinal cord. The flexed posture of the head and neck may have had implications in development and progression of the syrinx
Gill et al. 2014 (23)	Case report (n = 1)	An 80-year-old man	Syrinx T7–T9. Previous fall several years earlier	Progressive deterioration in mobility for 1 year	Multidisciplinary rehabilitation without further description of content	Evaluation described as pain reduction, increased mobility and returning to an independent lifestyle.	Repetitive straining is consistent with the hypothesis that Valsalva-like manoeuvres may increase subarachnoid fluid pressure and precipitate acute presentation of previously asymptomatic syringomyelia

MR; magnetic resonance; SCI: spinal cord injury; AC: Arnold Chiari; PCS: Physical Component Score; MCS: Mental Component Score; QOL: quality of life; SF-36: Short-Form 36-item; SDI: Syringomyelia Disability Index; TENS: transcutaneous electrical nerve stimulation; NDI (Neck Disability Index).

*Refers to the subsample that received an active conservative intervention.

^aRefers to sex and age of the total sample, data for the subsamples not available.

Study measures and results

Similar to the descriptions of intervention content, the descriptions of study measures and results also varied markedly across the included studies.

The 4 single case studies reported functional outcomes at impairment level with either stabilized (21, 22) or improved (21–24) motor function by strength and reduced pain (23, 24) as the main features. Among these, 3 studies (20–22) reported stabilized or improved posture, all indicating that this might be a reason for stabilized or improved function. For Haas et al. (23), the effects on pain and posture were stable at 1-year follow-up. However, it was unclear how these outcomes were measured in several studies (21–24). Haas et al. (23) evaluated outcomes as postural improvement by means of chiropractic manipulative therapy and patient exercises, both visually and by means of radiographs. On the activity level, improved gait and mobility were reported by Güler et al. (22) and Gill et al. (24). The latter also reported “return to an independent lifestyle” as an effect of their conservative intervention.

The 3 remaining studies (15, 19, 20) reported study results in different ways. In their 5 cases receiving conservative interventions, Ronen et al. (15) concluded neurological stability and functional improvements due to training without further description. Specifically, they reported stability in functional status in 4 patients, while 1 patient was significantly improved. However, it was not reported how this was evaluated. Only very general descriptions of outcome in terms of “improved”, “deteriorated”, or “unchanged” neurological function were provided. In the 2 cohort studies (19, 20), patient status (19) and perceived effectiveness (19, 20) were performed using a variation of self-report questionnaires focusing on functional status, QoL, and pain, among others, and semi-structured interviews collecting patients’ subjective descriptions.

The study by Sixt et al. (19) was not designed to evaluate an intervention, but rather to describe QoL in patients with syringomyelia in general. Nonetheless, they described a sample with 2 subsamples having received 2 different interventions (surgery and active conservative management) and concluded that patients’ QoL and functional disability as measured by the Syringomyelia Disability Index (SDI) and the subscales of the Short Form 36 (the physical and mental component scores) respectively, were independent of the choice of surgical or conservative treatment. Yet, an enhanced risk of complications and worsening of chronic symptoms related to surgical management was mentioned, but no specific results regarding the effect of the active conservative intervention were reported. However, the importance of maintenance of good physical and mental QoL as well as coping

mechanisms was essential in both intervention groups to facilitate reduction of symptom intensity and duration. The study by Smith et al. (20) is the only paper providing a more detailed analysis of the perceived effects of physiotherapy using patients' perception reported in questionnaires and interviews. Pain, followed by weakness and fatigue, was the most frequent symptom, which increased by prolonged activity, certain postures and positions. A majority of patients reported symptom deterioration over time, and pain reduction was the goal of rehabilitation in 59% of participants. Bi-variate, non-parametric analyses revealed no significant correlations between either physiotherapy treatment and symptom progression, surgical intervention and symptom progression, physiotherapy treatment and pain, nor surgical intervention and pain. The physiotherapeutic interventions perceived as most effective were stretching and hydrotherapy, resulting in reduction of pain and stiffness and in improvement of function. However, no description of the specific movements and exercises included in the treatment programs was available. The effectiveness of treatment was also reported to be hampered by limited knowledge about syringomyelia among the physiotherapists and limited information provided to the patients. Commonly for the 2 studies (19, 20), physiotherapy and surgery were perceived equally effective. Overall, physiotherapy resulted in symptom regression and improvement of QoL in a majority of the patients.

DISCUSSION

This scoping review identified and synthesized existing studies in individuals with PSS who received an active conservative intervention. Seven studies were included, which were heterogeneous in both method, therapeutic content and findings. These indicated that active conservative interventions, focusing on elimination of progressive syringomyelia-related symptoms, might lead to stability or improvement in physical function and activity, such as posture and pain, gait, ADL and QoL. Hence, it seems important to discuss some general issues across the studies in relation to clinical implications and perspectives for active conservative management of PSS.

Study intervention content

A general issue across the included studies was limited information on the exact content of the interventions. This limits comparability across studies, but, more importantly, it limits clinical applicability. The term "active conservative management" is a broad concept, potentially covering very different approaches. By defining active conservative management as a criterion for

eligibility, we found a limited number of studies that applied different modalities, such as physiotherapy with different treatments or exercises, chiropractic with manipulation, a combination of several of these interventions, or an interdisciplinary approach. In the included studies, the predominant approach was physiotherapeutic, with physical activities by the patients as well as manual treatments by the physiotherapists. Unfortunately, an in-depth description of the exact content was often not available.

Despite different interventions, a common approach in several of the studies was focus on posture alignment. Smith et al. (20) reported that the patients experienced aggravation of symptoms in certain postures and positions but also in prolonged activity. Osama et al. (25) hypothesized that a reduction in symptoms in a case of management of idiopathic syringomyelia also was due to reduction in postural deviations. This central aspect of posture is also highlighted by 3 of the case studies in the current review, potentially supporting a theory of optimizing the position of spine segments to avoid stress and strain on the spinal cord and/or the scar tissue after the initial trauma/inflammation. This has been suggested to contribute to growth and progression of the syrinx and thus possible symptoms related to it (25). Hence, the aspect of posture deserves distinct attention in future evaluations of a conservative therapeutic approach.

In several of the included studies, the management of syringomyelia also involved physical exercise to improve or maintain physical function. Physical exercise and weightlifting increase intrathoracic/intra-abdominal pressure and may trigger neurological symptoms in patients with syringomyelia (5). Balmaseda et al. (26) showed development of symptomatic syringomyelia 5 years after injury, directly related to heavy weightlifting exercises. This supports the possibility of triggering an acute presentation of previously asymptomatic syringomyelia after disease or trauma (27). Also, the increased intrathoracic pressure due to normal automatic reflexes as coughing and sneezing may increase symptoms in symptomatic syringomyelia patients (9, 27). Hence, there is a need for caution concerning the use of weightlifting exercises in these patients, which, in some individuals, might deteriorate the condition. If this form of training is used, patients should be carefully monitored for potential adverse effects.

Study measures and results

Improvement in QoL and ADL by means of symptom reduction and functional improvement is often a central goal for the patient in active conservative rehabilitation of syringomyelia. However, the tools

used for evaluation in the included studies addressed various aspects of function, activity and QoL. Sixt et al. (19) (although not formally evaluating an intervention) and Smith et al. (20) used the SDI, which was developed as a questionnaire specifically adapted for syringomyelia patients. The SDI is based on the Neck Disability Index (NDI), with inclusion of questions in relation to individual routine activities and physical function and not only in relation to neck disabilities (19, 20). This emphasizes the importance of describing the patient's perception of what kind of ADL triggers syringomyelia symptoms. This knowledge may guide clinical reasoning concerning the choice of treatment and evaluation of an intervention's effect. As shown in Table I, the clinical problems and symptoms presented in the studies varied widely, but the correlation between ADL and triggered symptoms was hardly described in the case reports. In general, in the studies with effect of exercise-based interventions, no description of the therapists' considerations and assessments concerning the load in training exercises in relation to the symptoms of the specific patient was provided. Neither was the potential correlation with an increase in CSF pressure in different exercises mentioned. This may reflect limited knowledge about the pathophysiological mechanisms among health professionals, as mentioned by Smith et al. (20) and constitutes a challenge to physiotherapists to select specific interventions and to be able to create strategies with the patient to reduce symptoms in daily activities.

Limited and low-quality evidence

In addition to the above points, there is a general issue of limited and low-quality evidence in this area. The current study revealed a very limited number of studies assessing an active conservative intervention in PSS. While this may, in part, be due to the criterion of the explicit description of an active approach in the current study, it indicates a lack of priority and a limited focus in the existing literature. As a part of this process, surgical techniques for management of the condition have been evolving continuously, while focus on active conservative management has been sparse. Furthermore, most of the included studies did not have randomized or other controlled designs and a limited number of participants, which affected the quality and applicability of the evidence. While single case and cohort studies without control groups hold merit, such designs are problematic in testing the effectiveness of health interventions. Study quality was not explicitly assessed or graded in the current study; thus the level of evidence provided by the studies included in this review must be considered low. Therefore, it is important to design and perform high-quality studies

in order to gain further knowledge about the effect of active conservative interventions in PSS.

Clinical experience and perspectives for a future approach

One of the authors' (LO) clinical experience of physiotherapeutic interventions shows that many patients typically report increased symptoms related to various ADL, such as hanging laundry, hoovering, carrying bags, or activities involving upper-extremity elevation both static and dynamic, but also by some physical training. Specific clinical examinations and tests have been central to analyse syringomyelia symptoms in different positions, movements, and activities, and to link these findings to changes in cerebral spinal fluid (CSF) pressure and spinal tissue stretching. Based on that analysis, the treatment has specifically focused on optimizing posture in activities, enhancing muscle stability of the spine, and assessing acceptable load in activities and training, commonly to avoid stress and strain of the spinal cord and thus CSF pressure disturbance. In clinical practice, the outcome of this activity-based approach has been effective in stabilizing and reducing symptoms for most patients. Furthermore, it is central to encourage the patient to be able to manage the condition and to achieve reduction of symptom intensity by relevant changes in daily activities, which also is recommended by Goetz et al. (28) as an interdisciplinary approach. Physiotherapy as an intervention requires knowledge of the pathophysiological components of syringomyelia as the basis for choosing a specific intervention to improve function and avoid symptom progression. Thus, patient education about the disease and empowerment are important factors.

Study limitations

Despite several positive elements of this study, some limitations should be taken into account. First, the number of studies included was potentially limited due to the search string. The search words could have included "management" and "therapy", which possibly would have resulted in more hits. Secondly, 19 abstracts deemed relevant for full-text assessment were not available in peer-reviewed article format, but were published as posters, a letter to the editor, and a book chapter, respectively. This criterion may therefore have prevented relevant information being included. Thirdly, several of the excluded studies dealt with congenital conditions, such as Arnold Chiari malformation, as well as with surgical management. Some of these may have contained information relevant even concerning active conservative approaches. The authors tried to omit this problem by reading those articles and could not find relevant information. Fourthly, there may

be some excluded studies that did address active conservative interventions but did not describe this explicitly. There may therefore be additional relevant evidence not included in the present work. Fifthly, in the assessment of 2 studies (19, 20), deviations from protocol were made, as they did not fulfil the outlined eligibility criteria. These studies had mixed samples, with limited possibility to differentiate who had PSS and who had received active conservative interventions or surgical interventions. This is a major issue, as these 2 studies therefore combined samples and interventions that did not fulfil the outlined eligibility criteria. Sixthly, included studies did not have to evaluate an intervention, but merely report on a sample that received an active conservative intervention, as was the case for Sixt et al. (19). This means the included studies are a broader reflection of existing studies in samples receiving an active conservative intervention rather than studies only assessing effects of an active conservative intervention. Seventhly, the *a priori* internal protocol was amended several times during the review, affecting the systematic approach. Eighthly, the studies were generally very different in study designs, interventions, and outcomes, which made comparability difficult. Finally, despite generally poor study quality in several of the included studies, we did not formally assess study quality and take this into account in the analysis.

CONCLUSION

This scoping review identified existing evidence concerning “active” conservative management of syringomyelia symptoms over 4 decades. Unfortunately, there is not much information about active conservative treatment available in the literature, albeit it is needed. The 7 studies found were very heterogeneous; more than half of them being case reports, which provided only anecdotal evidence. With this important point in mind, the studies indicated that physiotherapeutic treatment may be effective in reduction of symptoms, improvement in physical function, ADL and QoL for the patient. However, a specific description of the exact content of the interventions in relation to the pathophysiological mechanisms of CSF pressure and flow was lacking in all but 1 study (24). Therefore, additional research using appropriate study designs to reveal the causal relationship between intervention and outcome is essential, as is collaboration and a more unified approach among professionals working with active conservative management of patients with syringomyelia. Future research based on the results of this review and our own clinical experience should focus on providing an empirical and clinical basis for a structured and “active” conservative management of syringomyelia. It would be important to develop a specific clinical analysis of the symptomatic activities

in the individual patient and to use this central knowledge to design a specific activity-based intervention to reduce symptoms. Relevant research questions for future research could be:

- Can a thorough patient history combined with an examination based on pathophysiological concepts provide a sound basis for an individually adapted activity based conservative treatment approach?
- Can this approach improve or stabilize symptoms in patients with primary syringomyelia over time?
- Can patients thus be empowered and gain control over their symptomatology to optimize their QoL?
- Can patients not responding to this active conservative approach be identified as suitable candidates for surgery?
- Can this type of approach be applied to optimize rehabilitation and follow-up after surgery?

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Competing interests

The authors have no conflicts of interest to declare.

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Author contributions

LO and WA contributed equally to the work. They were responsible for designing the review protocol, screening potentially eligible studies, extracting and analysing data, conducting the narrative analysis, interpreting results, creating tables, and writing the manuscript. SLR participated in designing the review protocol and conducting the searches and contributed to writing the method section and provided continuous feedback on the manuscript.

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