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RESEARCH ARTICLE

UPPER LIMB MUSCULOSKELETAL COMPLICATIONS IN SYRINGOMYELIA: A REVIEW

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Abstract

Syringomyelia is a long-lasting disease, characterized by the formation of a syrinx, a tubular hole, or cyst in the spinal cord. The syrinx tethers and destroys the surrounding neural tissue, particularly within the cervical spinal cord, responsible for the upper limbs. As a result, these patients typically have various musculoskeletal issues related to the shoulder, arm, and hand. Such complications include muscle atrophy, joint deformity, neuropathic (Charcot) joints, contractures, and indirect spinal deformities (scoliosis). Loss of protective sensation and motor neuron involvement are the main contributors in these phenomena. Early in disease, musculoskeletal manifestations can occur and sometimes precede profound neurologic impairment. Early recognition of such complications is the key to diagnosis and a meaningful care. This review focuses on the pathophysiology, clinical manifestations and musculoskeletal complications of the upper limb in syringomyelia, with specific attention to orthopaedic relevance. This review identifies a lack of focused synthesis on upper limb musculoskeletal complication s in syringomyelia and highlights the need for early multidisciplinary intervention.

Introduction: -

Syringomyelia is a rare disease characterized by the appearance of a fluid-filled cavity, named syrinx, within the spinal cord [1]. The syrinx slowly expands over months to years, affecting more and more neural parenchyma. It is the cervical and upper thoracic spinal cord that is mainly affected, and this part of the spinal cord is what governs movement and feeling in the upper limbs [2].

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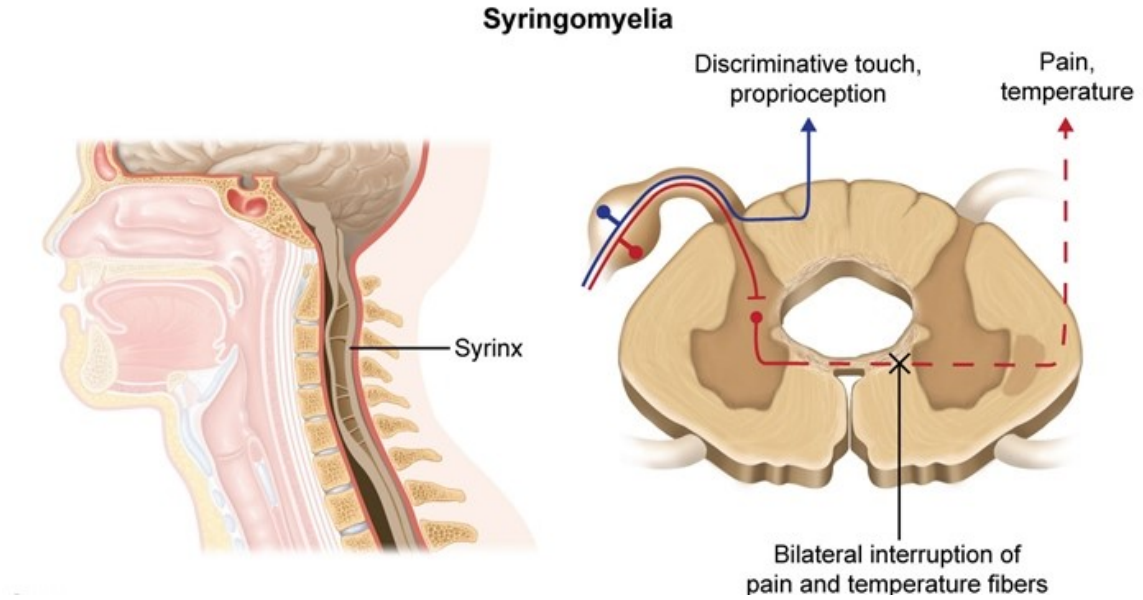
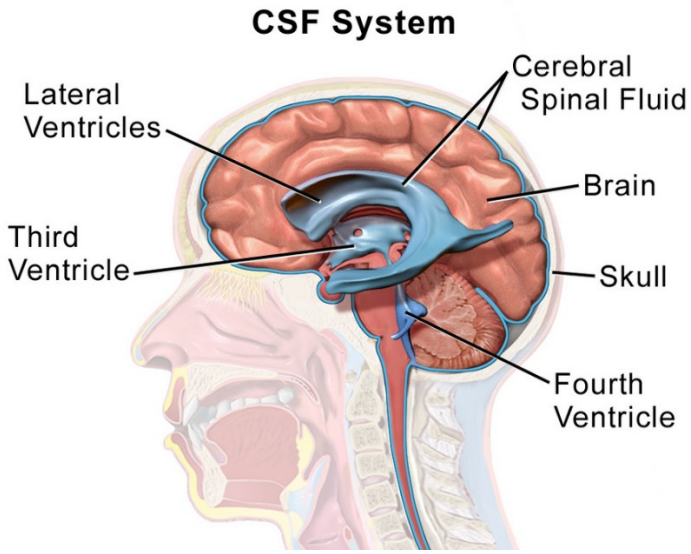


Image Reference: https://medical.uworld.com/library/neurology/762/syringomyelia/

Syringomyelia can arise from a number of underlying disorders, such as Chiari malformation, spinal tumors, trauma, infections, and congenital malformations [3]. Among these etiologies, Chiari type I malformation is by far the most common associated disorder. Disturbance of Cerebrospinal fluid (CSF) flow results in the development of the syrinx within the spinal cord.



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The neurological behavior characterized by syringomyelia is well-known and descried in the literature as sensory loss, pure motor deficits and autonomic failure. But the disease is also responsible for significant musculoskeletal sequelae, specifically in the upper limbs [4]. These are effects on the musculoskeletal system and stem from damage to the anterior horn cells, sensory tracts and autonomic nerve fibers.

Musculoskeletal problems frequently seen in individuals with huntington's disease include: Muscle wasting of the hand

1. Wasting of the hand muscles
2. Joint deformities
3. Neuropathic (Charcot) joints
4. Contractures
5. Scoliosis

These complications may result in a disastrous impact on patient's upper-limb function and quality-of-life. In some of these cases syringomyelia and its orthopedic manifestations might even precede any neurological symptoms initially for years [5]. Although the neurological aspects of syringomyelia have been studied extensively, consolidated literature on upper limb musculoskeletal complications and their clinical impact is almost non-existent. This void hinders early detection and focused rehabilitation. Therefore, this review aims to systematically analyze the upper limb musculoskeletal manifestations associated with syringomyelia and highlight their relevance in clinical and rehabilitative practice.

Literature Review:-

The neurological and orthopaedic features of syringomyelia have been described in a number of clinical and experimental investigations. The disease was described in the 19th century by Charles-Prosper Ollivier d'Angers, who first reported the existence of fluid-filled cavities inside the spinal cord [6]. It was later found that syrinx enlargement results in progressive destruction of spinal cord structures for sensory and motor function [7]. The loss of sensation to pain and temperature in syringomyelia was noted in studies by many authors and has been explained by involvement of the spinothalamic tract. Several other investigations report on the associated orthopaedic problems. It was reported that neuropathic arthropathy of shoulder is a frequent complication in syringomyelia [8]. This is because the patient loses protective pain sensation and continues to use the affected joint despite a series of injury.

Wasting of the hand is also an established manifestation of the disease. The anterior horn cells of the cervical region of the spinal cord are affected and their gradual loss causes wasting of the small muscles of the hand and claw hand deformity [9]. Studies have also reported that scoliosis is commonly found in patients with syringomyelia, particularly in children and teenagers [10]. Sometimes scoliosis may precede other neurological signs for a long time and can make the doctor search for the presence of a syrinx. In summary, the evidence suggests that musculoskeletal disease is prevalent and clinically significant in syringomyelia.

Pathophysiology: -

The pathophysiology of musculoskeletal disorders in syringomyelia is fundamentally dependent on the progressive syrinx distention within the spinal cord [1].

With the expansion of the syrinx, there is compression of the following key neural components: -

- Anterior horn cells
- Spinothalamic tract
- Posterior columns
- Autonomic nerve fibers

Lesions of these structures will produce a mixed motor, sensory, and autonomic neuropathy.

Motor Dysfunction: -

The destruction of anterior horn cells produces flaccid weakness and wasting. As a result, muscle wasting is progressive, primarily involving the internal muscles of the hand [9].

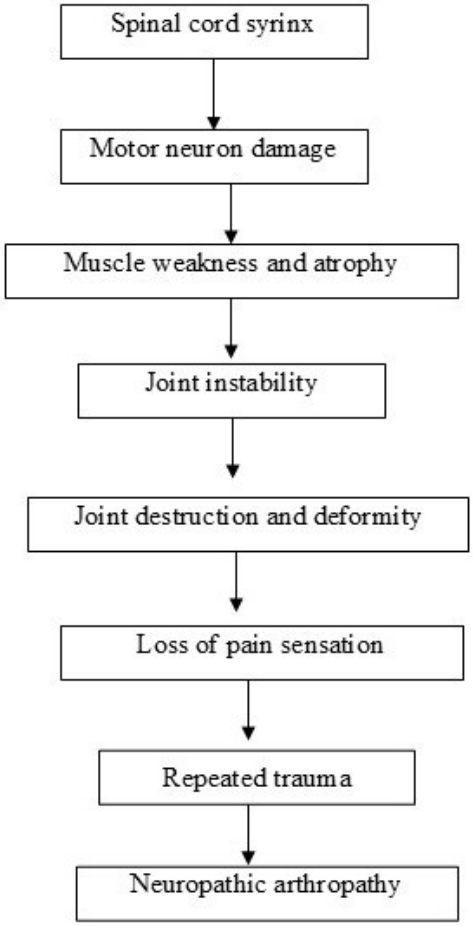
Sensory Dysfunction: -

Retrograde degeneration of the spinothalamic tract results in loss of pain and temperature in the classic "cape-like distribution" involving the shoulders and upper extremities [11]. Because they can't feel pain normally, they may injure their joints and not even realize it.

Autonomic Dysfunction: -

Involvement of the autonomic nerves may cause impaired circulation to bones and joints. This can lead to bone loss, joint destruction and neuropathic arthropathy [12]. The additive effect of these mechanisms thus causes a progressive upper limb musculoskeletal disorder.

Figure: Musculoskeletal Effects of Syringomyelia on the Upper Limb: Concept Diagram



Musculoskeletal Complications: -

Muscle Atrophy: -

Muscle Atrophy muscle atrophy is a very early and frequent finding in syringomyelia [9].Intrinsic muscular atrophy is now the main feature of the disease, affecting not only the small muscles of the hand (interossei, lumbricals).

Patients often develop:

- Weak grip strength
- Difficulty with fine motor movements
- Wasting of hand muscles

This muscle weakness may over time lead to deformities such as clawing of the fingers.

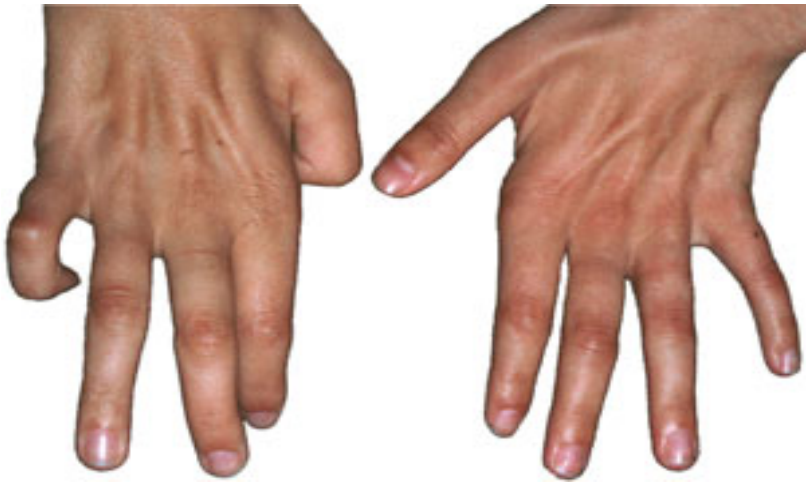


Image Reference:https://neuros.net/en/over-all_syringomyelia/

Joint Deformities:-

Joint deformities result from muscle imbalance, weakness, and chronic injury to joints [13]. Frequently involved joints are:

- Shoulder joint
- Elbow joint
- Wrist joint
- Small joints of the hand

Such deformities can cause limitation of joint motion and functional disability.

Neuropathic (Charcot) Joint:-

Neuropathic joint disease is also called a Charcot joint, and is a serious orthopedic complication of syringomyelia [8]. It is induced by the collapse of protective mechanisms to prevent joint destruction from minor trauma.
The most frequently involved joint in syringomyelia is the shoulder.

Clinical features are:-

- Joint swelling
- Instabilità (coll.) [Instability]
- Bone fragmentation
- Joint destruction
- Little pain despite extensive structural damage

Radiographic examination may reveal bone resorption, dislocation of the joint, and the formation of osteophytes.

Contractures:-

Contractures arise when muscles and the adjacent connective tissue (such as ligaments and joint capsules) become tight and shortened with a consequent inability to move the joint through a full range of motion [14].

In syringomyelia, contractures arise as a result of:

- Persistent muscle weakness
- Deformities of the joints
- Limited range of motion

Typical examples are of a flexion contracture of the fingers, wrist and elbow.

Diagnosis:-

Diagnosis of syringomyelia is largely clinical with supportive imaging.
Clinical Features

Clinical manifestations include:-

- Loss of pain and temperature sensation
- Muscular weakness
- Hand muscle wasting
- Joint deformities

Imaging: -

Magnetic resonance imaging (MRI) is the key diagnostic modality to visualize the syrinx in the spinal cord [16]. MRI may also reveal related abnormalities, including Chiari malformation or spinal neoplasms.

Table 1: Summary of Musculoskeletal Complications

Complication	Pathophysiology	Common Location	Clinical Features
Muscle Atrophy	Damage to anterior horn cells	Intrinsic hand muscles	Weak grip, claw hand
Joint Deformities	Muscle imbalance and repeated trauma	Shoulder, elbow, wrist	Instability, deformity
Neuropathic (Charcot) Joint	Loss of protective sensation and autonomic dysfunction	Shoulder, elbow	Swelling, bone destruction
Contractures	Muscle shortening due to weakness and immobility	Elbow, wrist, fingers	Reduced range of motion
Scoliosis	Asymmetric spinal muscle weakness	Thoracic spine	Lateral spinal curvature

Table 2: Summary Table of Key Research on Syringomyelia and Related Complications

Author	Year	Objective	Aim	Technology / Method Used	Accuracy / Outcome
StatPearls Publishing	2023	To provide overview of Syringomyelia	To explain causes, symptoms, and management	Literature review, clinical data, MRI use	Helpful for understanding diagnosis and treatment; no exact accuracy given
Greenberg	2020	To provide neurosurgery guidelines	To guide diagnosis and treatment	Clinical and surgical reference	Useful for management decisions
Barkovich	2019	To explain neuroimaging findings	To improve imaging diagnosis	Advanced MRI imaging	Highly useful for detecting syrinx
Deng et al.	2017	To review orthopedic manifestations	To explain bone and joint complications	Literature review, imaging	Showed importance of early detection
Tubbs et al.	2015	To study pediatric Syringomyelia	To explain features in children	Clinical study and imaging	Helps in early pediatric diagnosis
Strahle et al.	2015	To study scoliosis with Syringomyelia	To explain pathogenesis and management	Clinical study, MRI	Early diagnosis helps manage spinal deformity
Adams et al.	2014	To explain neurological disorders	To provide clinical knowledge	Clinical neurology review	Widely used reference for diagnosis
Panda et al.	2012	To report Charcot Joint shoulder case	To highlight rare presentation	Case report, X-ray/MRI	Imaging confirmed diagnosis
Tubbs et al.	2011	To review pathogenesis and	To explain causes and treatment	Literature review, neurosurgical data	Improved understanding of treatment approaches

		management	options		
Nacir et al.	2009	To report neuropathic arthropathy in Syringomyelia	To show joint complications	Clinical exam and imaging	Confirmed link between syringomyelia and joint damage
Brower	2008	To explain Charcot Joint	To describe imaging findings	Radiological imaging techniques	Imaging is key for diagnosis
Klekamp	2002	To study surgical treatment	To evaluate outcomes of surgery	Surgical data and follow-up	Surgery improves symptoms in many patients
Batzdorf	1991	To discuss diagnosis and treatment concepts	To improve clinical management	Clinical evaluation and imaging	Helped in better diagnosis and treatment planning
Milhorat et al.	1995	To study syringomyelia with Chiari I malformation	To describe clinical and radiological findings	Clinical study, MRI imaging	MRI useful in identifying syrinx and Chiari malformation
Barnett et al.	1973	To review syringomyelia cases	To analyze symptoms and outcomes	Case review and clinical observation	Provided clinical pattern understanding
Williams	1972	To study pathogenesis of Syringomyelia	To explain how syrinx forms in spinal cord	Theoretical and clinical observations	Provided early understanding of disease mechanism

Fig: Graph: Year vs Research Focus in Syringomyelia Studies

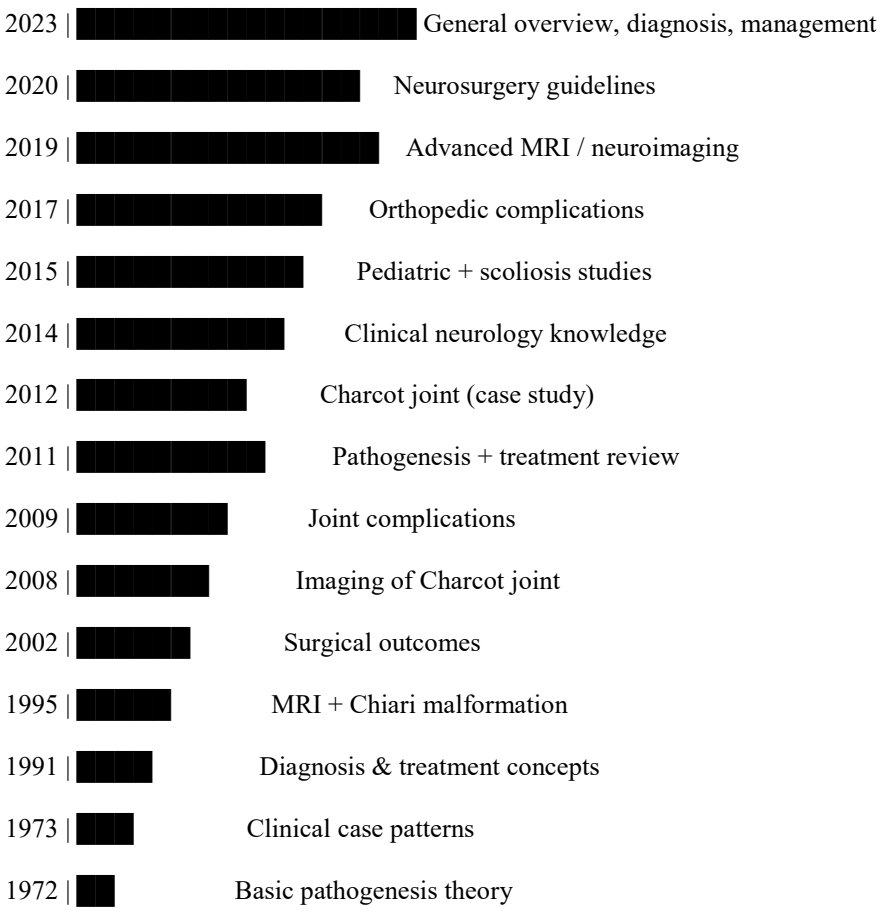


Table-to-Graph Summary; -

Time Period	Main Research Focus
1970–1980	Disease mechanism (pathogenesis)
1990–2000	Diagnosis and MRI development
2000–2010	Surgical treatment and imaging
2010–2020	Complications (joint, scoliosis)
2020–Present	Advanced imaging, guidelines, management

Management: -

The treatment for syringomyelia depends on the size and cause of the syrinx and the severity of symptoms.
Treatment with surgery the purpose of surgery is to restore CSF flow and decompress the syrinx. Also, to avoid adhesions, bleeding and complications.

Popular procedures include: -

- Posterior fossa decompression
- Syrinx shunting
- Resection of tumor (if exists)

Nonoperative Management: -

Conservative management may include:

- Physiotherapy
- Occupational therapy
- Orthopedic support
- Pain control

A staged treatment approach can be used in post-polio syndrome and may prevent progression of musculoskeletal impairments.

Discussion: -

The musculoskeletal system is commonly affected in syringomyelia, with manifestations that can be severe and that may present early, prior to more obvious neurological findings. Among these, intrinsic muscle atrophy of the hand, joint deformities, and neuropathic arthropathy of the shoulder are frequently encountered and represent significant sources of disability. These are the result of motor neuron loss (with dissociated sensory loss and autonomic dysfunction), progressive joint instability, and recurrent unperceived trauma. The clinical rationale of these observations is their implication as potential early diagnostics sign, particularly in individuals with unknown cause of upper limb deformities or weakness. From a rehabilitative standpoint, early intervention with PT is necessary to avoid permanent contractures and loss of function. Specific interventions including muscle strengthening, range of motion, splinting, and joint protection, may make a meaningful difference in patient outcomes and quality of life.
Despite the existing literature, further focused research is merited to investigate the rate of progression of musculoskeletal complications and to establish standardized rehabilitation protocols. Further research should address this deficiency by investigating multidisciplinary management, combining the expertise of neurology, orthopaedics and rehabilitation for the best interests of the patient.

Conclusion: -

Syringomyelia is a neurodegenerative disease with severe complications of the upper limbs musculoskeletal system, such as muscle atrophy, joint deformities, neuropathic arthropathy and contractures. These symptoms are based on the combined motor, sensory, and autonomic dysfunction. Prompt identification of such features is crucial for diagnosis and management. A multidisciplinary team approach comprising of neurologists, orthopaedic surgeons and rehabilitation physicians is necessary to maximise functional outcome and quality of life.

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