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Shoulder arthropathy secondary to syringomyelia: systematic review

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- *Purpose:* The purpose of this study was to collect and evaluate clinical and radiological evidence on shoulder neuroarthropathy (NA) in syringomyelia (SM) that may support the management and treatment of patients with this condition.
- *Materials and methods:* This systematic review is based on the analysis of reports available in PubMed, Embase, Cochrane Database of Systematic Reviews and Cochrane Central Register of Controlled Trials using the following keywords: syringomyelia, neuroarthropathy, Charcot joint and shoulder degeneration. Thirty-nine publications were found presenting case reports or case series meeting our criteria. Pooled data included a group of 65 patients and 71 shoulders with NA secondary to SM.
- *Results:* The most commonly reported symptoms were range of motion (ROM) limitation, weakness, swelling, pain and dissociated sensory loss. NA is usually monolateral and concerns only the shoulder. The average active shoulder ROM was flexion −59.2° (S.D. 37.9), internal rotation −29.8° (S.D. 22.6) and external rotation −21.1° (S.D. 23.6). Most of the patients (75%) presented with complete or nearly complete proximal humerus degeneration, while the degree of glenoid preservation varied. Fifty-two neuroarthropathic shoulders were treated conservatively with physiotherapy, anti-inflammatory medication and splinting. Eighteen patients were treated by surgical intervention.
- *Conclusion:* Shoulder NA due to SM is a devastating and progressive condition, and its course is often unpredictable. Patients with unexplained shoulder degeneration should be evaluated for SM, especially if there are additional neurological symptoms. Conservative treatment usually reduces shoulder pain without improving ROM. For select patients, shoulder arthroplasty may be a better option for restoring function.

Keywords

- shoulder neuroarthropathy
- Charcot shoulder
- syringomyelia

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Introduction

Shoulder neuroarthropathy (NA) is a rarely described, devastating disease that causes progressive degeneration of joint structures, persistent inflammation and severe limitation of shoulder function (1, 2). The joints of the upper limb are much less often affected by the process of NA (3). The shoulder is the most frequent which accounts for only 5% of all Charcot joint locations (4). Syringomyelia (SM) is a rare and slowly progressive disease characterised by the formation of a fluid-filled cavity called a syrinx, which destroys the spinal cord and causes typical neurological symptoms (Fig. 1) (1, 5). A syrinx is most commonly situated in the cervical and thoracic segments (5). It typically coexists with type 1 Chiari malformations (6). At the same time, SM is reported to be the main cause of the Charcot joint in the upper limbs (3, 7, 8). This may bring overlapping neurological symptoms to the clinical picture. Patients suffering from SM might experience a variety of additional problems, such as headaches, weakness, dissociated sensory disorders and neuropathic pain (1, 9). Less frequent problems include balance disorders, spasticity and urinary disorders (10). Of patients with a diagnosis of SM, 20–30% develop shoulder NA (11). For many of them, shoulder problems are the first clear symptom of SM, prompting them to visit a physician. Although the pathophysiology of arthropathy associated with SM is not clear, two different theories of the causes of arthropathy have been proposed. The first one links the disrupted autonomical system, which provokes vascular disorders, necrosis and joint degeneration. The attributed joint degeneration is a result of repeated injuries due to loss of pain sensation (11, 12, 13).

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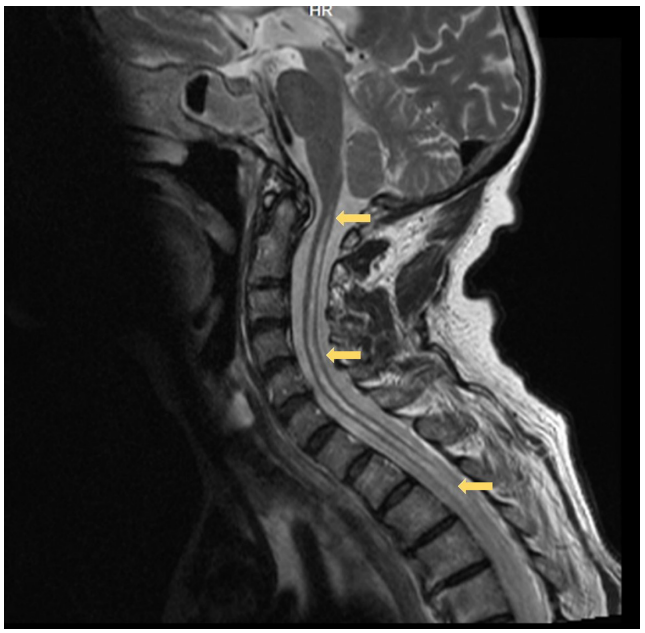

Figure 1
Cervical spinal cord MRI with syrin.

There is currently very little evidence in the literature related to shoulder arthropathy due to SM. All data come from a few case reports and small case series (4, 5, 8, 9, 10, 11). We have developed a particular interest in this condition, reporting probably the largest series so far (11). Therefore, the aim of this systematic review is to collect all available clinical and radiological evidence on shoulder arthropathy in SM. The secondary aim is to support the management of patients with this condition.

Materials and methods

This systematic review is based on an analysis of reports available in PubMed from 1966 until 2021, Embase from 1980 until 2021, Cochrane Database of Systematic Reviews and Cochrane Central Register of Controlled Trials using the following keywords: syringomyelia, neuroarthropathy, Charcot joint and shoulder degeneration. For the purpose of this systematic review, the following inclusion criteria were applied:

1. Shoulder arthropathy based on radiographic evaluation (at least H1 or G1 in the neuropathic gleno-humeral degeneration (NGH) classification) (Figs. 2, 3, 4 and 5) (14);
2. SM confirmed by magnetic resonance imaging (MRI) or myelography of the spinal cord; and
3. Available clinical data.

Thirty-nine publications have been found presenting case reports or case series meeting our criteria. Pooled data included a group of 65 patients and 71 shoulders with NA secondary to SM (Fig. 6).

Clinical and radiological data were analysed in the following categories:

1. Epidemiological factors: age, gender distribution, the affected side, time from diagnosis of SM to onset of symptoms of arthropathy;
2. Clinical symptoms: loss of sensation, weakness, limitation of range of motion (ROM), swelling and additional neuroarthropathic joints; the analysis included average active ROM in the shoulder;
3. Radiological evaluation: length of the syrinx in the spinal cord in MRI, Arnold-Chiari malformation, the advancement of shoulder degeneration (based on NGH classification – Table 1) (14);
4. Treatment: conservative (physiotherapy, medication, splinting, radiotherapy) vs operative – treatment effectiveness. An improvement of ROM of at least 20° in flexion and/or internal rotation (IR) and/or external rotation (ER) or information from the author that there has been an improvement was recorded as well as information on complete or partial pain relief.

Statistical analysis was performed using IBM SPSS Statistics 26.0. Basic descriptive statistics for quantitative variables were calculated using the programme. Logistic or polynomial regression analysis was performed for nominal variables and linear regression analysis for quantitative variables. The level of significance was $\alpha = 0.05$.

Logistic regression analysis was performed to determine whether cavity length in MRI had an impact on the

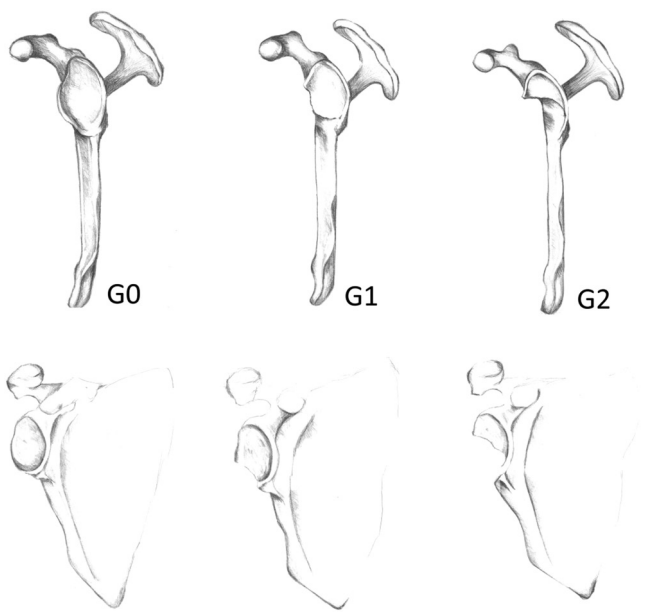

Figure 2
NGH classification – G stages.

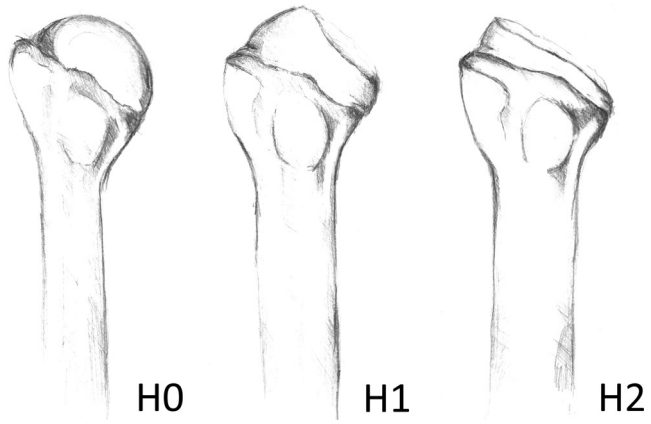

Figure 3
NGH classification – H stages.

frequency of individual symptoms such as pain, swelling, weakness and sensory loss.

To determine whether the length of the cavity in MRI affects the ROM in the shoulder, a linear regression analysis was performed using the insertion method. The following variables were taken into account as explained variables in the models: flexion, IR and ER.

In order to determine the effect of cavity length in MRI on the stage of G and H advancement, logistic regression analysis (for H) and polynomial regression (for G) were performed.

Fisher's exact test analysis was performed to compare the groups of patients with different degrees of H and G in terms of the occurrence of pain, swelling, weakness and ROM limitations.

We compared patients with different degrees of H and G in terms of flexion, IR and ER by performing the Mann–Whitney *U* test for H and the Kruskal–Wallis test for G. In order to standardise the measurement of IR and facilitate its statistical evaluation, IR was presented or converted to degrees (15).

To compare average ROM before and after conservative treatment, a Student *t* test was performed.

The significance level was $\alpha = 0.05$. Statistical analyses were performed using IBM SPSS Statistics 25.0.

Results

Epidemiological factors

Basic epidemiological data were calculated based on all pooled cases – 65 patients (75 arthropathic shoulders).

The average age of the analysed patients was 51.1 years (S.D. 15.8; min 22, max 76). Women were slightly more represented (1.2:1). The shoulder NA was almost irrelevant to the side (1.03:1). The vast majority of patients had unilateral NA (59 patients, 90.8%).

Other epidemiological data were available for 57 patients.Forty-one patients (63.1%) first sought medical advice related to SM due to shoulder complaints. The average time to obtain diagnosis of NA related to SM since this group first reported symptoms was 37 months (S.D. 63.1 months; min 1 week, max 20 years). Among the remaining patients, SM was diagnosed earlier and shoulder disorders appeared after an average of 11.6 years (S.D. 9; min 1; max 34).

Clinical symptoms

In 55 patients (84.6%), only the shoulder was affected by NA. Six had bilateral shoulder involvement. Thirteen cases suffered from accompanying arthropathy in other joints: elbow, five patients; hand, four patients; elbow and wrist, two patients; wrist, one patient and elbow and hand, one patient.

The most frequently reported symptoms within the affected shoulder were:ROM limitation, 67 out of 68 shoulders (98.5%); shoulder weakness, 62 out of 69 shoulders (89.9%); dissociated sensory loss, 53 out of 64 shoulders (82.8%) swelling, 55 out of 70 shoulders (78.6%) and pain, 49 out of 70 shoulders (70%).

There have been some additional, less frequent symptoms: opposite extremity weakness, 25 patients; balance disorders (loss of balance, unsteadiness), 6 patients; lower extremities weakness, 3 patients; lower

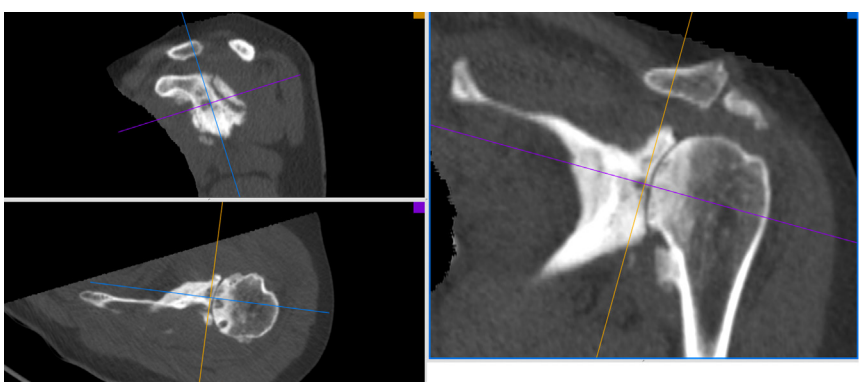

Figure 4
Preoperative CT scans of the shoulder with NA (G1H1).

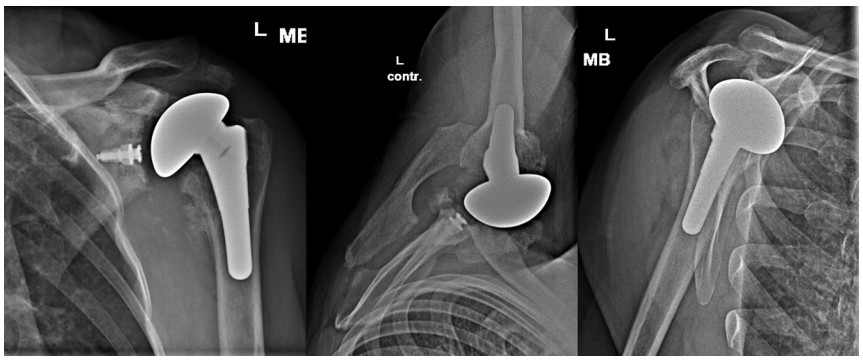

Figure 5
Postoperative X-ray after TSA.

extremities spasticity, 3 patients and urinary disorders (urinary incontinence and bladder dysfunction), 3 patients.

Subjective shoulder symptoms were reported in all pooled cases, whereas accompanying problems were reported in 63 cases. ROM data were available in 51 pooled cases.

Average active ROM was as follows: flexion, 59.2° (s.d. 37.9; min 10, max 180); IR, 29.8° (s.d. 22.6; min 0, max 90) and ER, 21.1° (s.d. 23.6, min -30, max 90).

Radiological evaluation

The average length of the syrinx in spinal cord MRI was 9.17 vertebral segments (+/−5.45), based on 48 reports. Additionally, 28 patients (43.08%) had Arnold-Chiari malformation, based on 65 reports.

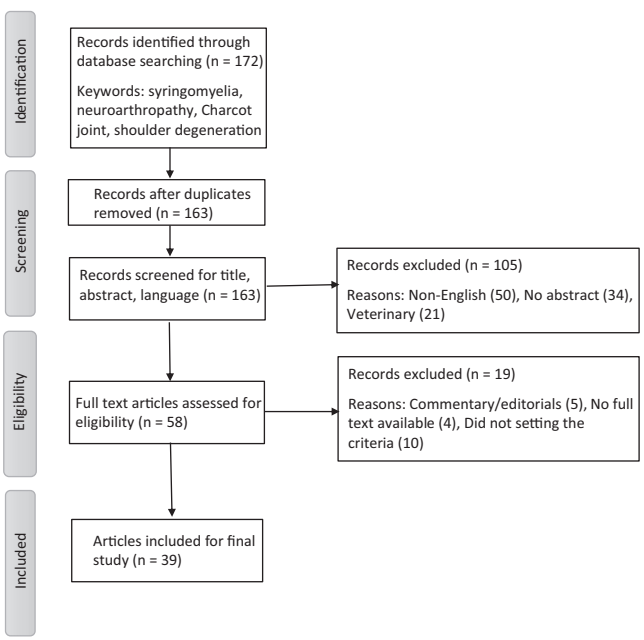

Figure 6
Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines flowchart illustrating the search strategy.

Radiological reports of shoulder X-rays were available for 62 cases, whereas X-ray images were available for 47 cases. Based on the latter distribution of degeneration, the progression was calculated (Table 2). Most of the patients (75%) presented with complete or nearly complete proximal humerus degeneration (H2), while the degree of glenoid preservation varied.

Correlations

The analysis did not show any significant influence of the length of the cavity as seen on MRI on the occurrence of individual symptoms: pain, swelling, weakness and sensory disturbance.

However, there was a significant negative relationship between the length of the cavity in MRI and shoulder flexion ($\beta = -0.48$; $P = 0.004$) and IR ($\beta = -0.51$; $P = 0.006$).

ROM limitations occurred in all patients regardless of grade, either G or H. Weakness was significantly more frequent in patients with the highest H grade (H2) than in patients with a lower grade (H1), with the occurrence of 88% and 54%, respectively ($P = 0.028$). Similarly, for G grade, weakness occurred in all examined patients with the highest-grade G (G2) and was significantly more frequent than in patients with G1 and G0 (65% and 25%, respectively; $P < 0.001$).

Furthermore, the swelling was significantly more frequent in patients with G1 than in patients with G0 (100% vs 50%, $P = 0.021$). This was also observed in 82% of the G2 cases but did not reach statistical significance.

The analysis showed no significant differences in flexion, IR or ER among patients with grades H1 and H2. Differences between grades G0, G1 and G2 in terms of the analysed variables were also not significant.

Treatment

Fifty-two neuroarthropathic shoulders were treated conservatively with physiotherapy, anti-inflammatory medication and splinting. The effect of conservative treatment on shoulder ROM was as follows: no change, 64.3% of patients; improvement, 32.1% and deterioration,

Table 1 NGH classification.

G (glenoid)	**H** (proximal humerus)
0 – No X-ray changes of glenoid	0 – No X-ray changes of proximal humerus
1 – Partial glenoid degeneration	1 – Partial proximal humerus degeneration (tuberosities preserved)
2 – Total glenoid degeneration	2 – Total proximal humerus degeneration (tuberosities not preserved)

3.6%. The effect on pain was as follows: pain reduction, 64.3%, no change, 28.6% and worsening, 7.1%. Data on the effectiveness of conservative treatment on ROM and pain were available in 44 cases. However, precise shoulder ROM of only 18 patients was reported after conservative treatment. After conservative treatment, the average ROM change was as follows: flexion from 68.6° to 84.4° ($P-=0.007$), IR from 30.2° to 47.6° ($P=0.002$) and ER from 29.7° to 31.2° ($P=0.33$). ER was statistically insignificant.

Eighteen patients were treated by surgical intervention. Shoulder arthroplasty was performed on 12 shoulders in 11 patients, including 1 patient bilaterally (Table 3). Four neuroarthropathic shoulders were treated with hemiarthroplasty resulting in three patients with pain relief and improvement of ROM and one without any ROM change and the same level of pain. One patient received humeral head resurfacing, resulting in ROM improvement and pain relief. Shoulder arthroplasty with rotator cuff repair was performed in three shoulders, all with ROM improvement and pain relief. Four shoulders were treated with reverse shoulder arthroplasty (RSA), all with good final results: two without any complications and two with some complications, but without the need for revisions. In both cases, significant functional improvement remained. In one case, baseplate screw loosening was noted. Another case experienced immediate shoulder dislocation, which was effectively treated conservatively. Shoulders remained stable with very good function for over 5 years of follow-up. One case was reported with multiple revisions following the initial total shoulder arthroplasty. The procedure resulted in instability and several additional procedures (sequentially revision hemiarthroplasty, followed by RSA and revision RSA). Due to the persistent instability of the prosthesis, the implants were finally removed. As expected, the ultimate result was poor.

We could not make any calculations on functional measures in surgically treated patients, since our data were incomplete.

Two patients qualified for shoulder arthrodesis, but the results were not reported. Three patients with infected shoulders required a synovectomy after primary conservative treatment.

Table 2 Number of patients in each classification group.

	H0	H1	H2
G0	0	3	1
G1	0	3	14
G2	0	5	18

Discussion

Our data clearly show that shoulder NA due to SM is a devastating condition, and its course is often unpredictable. The clinical picture and the course of SM are highly variable. In the majority (63%), shoulder problems are the first characteristic manifestation of SM. Yet it usually takes a very long time before a proper and definitive diagnosis is made. On the other hand, cervical SM may exist without any typical symptoms for unpredictable periods of time before the shoulder is affected. The main neurological symptoms of SM are headaches, sensory disorders (dissociated sensory loss, paraesthesia, causalgia), dysfunction of the sphincters (urinary or defecation disorders), muscle weakness or atrophy, spasticity, balance disorders and progressive scoliosis (6). Perhaps as many as 30% of patients with diagnosed SM in the cervical spine will develop shoulder arthropathy in the future (9, 16). However, it is still unknown what triggers shoulder destruction in patients with SM. Loss of proprioceptive function might be the factor that initiates a chain of shoulder microtrauma and leads to progressive joint arthropathy. Additionally, loss of pain perception may cause a prolonged, clinically silent course of degeneration. Most of the analysed patients were diagnosed with dissociative sensory disturbances within the affected shoulder. This is one of the most characteristic symptoms of SM, and it should always prompt diagnostics in this direction (17). It is generally asymmetrical and most often affects the area of the shoulder with NA. It is also puzzling why only some patients with a syrinx develop shoulder arthropathy. Additional factors such as smoking, alcohol or dyslipidaemias are associated with NA (18). As was clearly demonstrated in the study, syrinx length correlates with loss of shoulder function. Other syrinx features, such as location, may also influence neurological disturbance in the spinal cord.

Shoulder NA secondary to SM occurs in different age groups, and it does not have any gender predilection. Most often, arthropathy was asymmetric, which is characteristic of SM. An additional joint was rarely involved; in those rare cases, it was mainly the elbow, and it usually affected the same upper limb.

Nearly all patients had some degree of shoulder ROM limitation and weakness. Eighty-four per cent were not able to raise the arm to shoulder level. The majority had typical swelling related to inflammation. Swelling in the shoulder is generally a rare phenomenon, so it usually

Table 3 Results of shoulder arthroplasty in NA secondary to SM.

Reference	Case details		Symptoms	Preoperative ROM	NGH classification	Type of endoprosthesis	Postoperative ROM	Follow-up, years
	Age, years	Sex						
Crowther & Bell (26)	47	F	Pain, swelling, sensation disorders	100/?/?	G1H1	Resurfacing	140/?/50, no pain	2 years
Ueblacker *et al.* (22)	62	F-right	Pain, weakness	30/10/0	G1H2	RSA	110/80/45, no pain	2 years
Ueblacker *et al.* (22)	62	F- left	Pain, weakness	20/15/0	G1H2	RSA	110/80/70, loosening of the screw – screw replacement; no pain	2 years
Matsuhashi *et al.* (7)	54	F	Pain, swelling, weakness, sensation disorders	45/30/−20	GH2H1	TSA+RC repair	110/80/20, pain relief	10 years
Matsuhashi *et al.* (7)	55	M	Pain, swelling, weakness	15/50/15	G1H2	TSA+RC repair	80/50/20, pain relief	8 years
Matsuhashi *et al.* (7)	64	F	Pain, swelling, weakness	45/30/0	G1H2	TSA+RC repair	120/70/20, pain relief	8 years
Snoddy *et al.* (23)	67	M	Pain, swelling, weakness	50/15/−30	-	Partial, TSA, RSA	20/0/0, infection - removal of the endoprosthesis	1 year
Schoch *et al.* (2)			Pain, weakness, sensation disorders	20/15/45	-	Partial	20/30/0, symptoms like preoperative	10.4 years
Schoch *et al.* (2)			Pain, weakness, sensation disorders	30/70/0	-	Partial	100/75/20, no pain	11 years
Schoch *et al.* (2)			Pain, weakness, sensation disorders	80/45/10	-	Partial	150/70/90, no pain	5.2 years
Wawrzyniak *et al.* (10)	76	F	Swelling, weakness, sensation disorders	20/20/0	G1H2	RSA	140/45/40, no pain	4 years
Wawrzyniak *et al.* (10)	58	F	Pain, swelling, sensation disorders	60/50/0	G1H2	RSA	90/50/40, no pain	2.5 years

NA, neuroarthropathy; ROM, range of motion; RSA, reverse shoulder arthroplasty; SM, syringomyelia; TSA, total shoulder arthroplasty; RC, rotator cuff; NGH classification, classification of neuropathic gleno-humeral degeneration (14).
?:internal rotation and external rotation were unknown.

prompts diagnostics towards tumour or rheumatoid arthritis. Twenty-one patients had a biopsy to exclude a tumour when seeking a diagnosis for the cause of arthropathy. Each biopsy gave results indicating severe inflammation, which clearly caused swelling. Recurrent deterioration of function and increased swelling are also characteristic of the sinuous course of the disease (19). The most surprising observation was very commonly reported shoulder pain, despite neuropathy disrupting spinal sensory tracks. Possible reasons for pain in SM include disorders of sensory balance due to a disjunction between the anterolateral and dorsal columns of the spinal cord or abnormal levels of neurotransmitters, such as substance P (20, 21).

We recently published the results of the analysis of shoulder X-rays for this condition and showed that in most patients, the destruction of the proximal humerus is complete or almost complete, while the glenoid bone loss is more variable. We have also proposed and validated a new radiological classification for neuropathic shoulder degeneration concerning the glenoid and proximal humerus condition (14). Surprisingly, there was no clear correlation between degrees of ROM loss and radiological advancement on either humeral or glenoid bone loss. However, some trends show that ROM deteriorates as joint destruction progresses. Other factors might be affecting ROM, especially considering overlapping neurological disorders, extent of the cord involvement or clinical shoulder symptoms such as swelling or pain. Clinical symptoms were, however, correlated with the degree of glenoid or proximal humerus loss.

Most of the patients with this condition have been treated conservatively, mainly with anti-inflammatory drugs, splinting and physiotherapy. Often, such treatment reduces shoulder pain and swelling, but the greater challenge is regaining ROM. After conservative treatment, only about one-third of patients experienced an improvement in shoulder motion. In each case of improvement in ROM, the pain also subsided or decreased. The data on ROM were too small to make a thorough analysis of the improvement after conservative treatment. Therefore, we could anticipate significant improvement in pain and swelling with time, with some form of physiotherapy and anti-inflammatory medication. However, most patients will not improve their shoulder ROM. For this reason, surgical treatment with different shoulder arthroplasty designs has been attempted to restore shoulder biomechanics. Due to recurrent inflammation in the shoulder and the uncertain course of joint destruction in SM, there are still many doubts regarding the indication for arthroplasty. Additional contraindications are not uncommon in this group of patients and disqualify them from surgical treatment. Still, there have been some promising results from shoulder

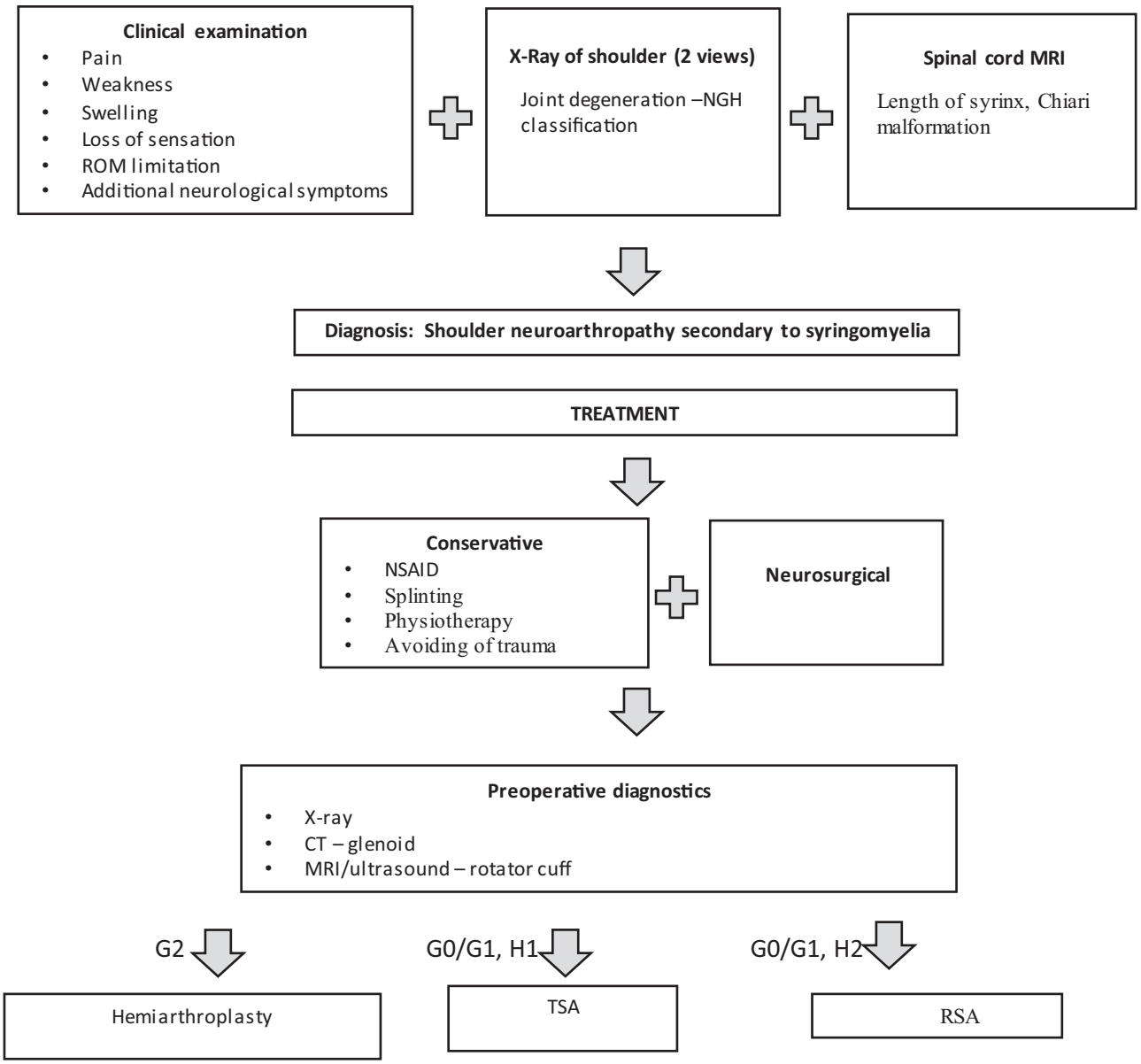

Figure 7
The algorithm to help guide the management of patients with shoulder NA in SM.

replacement with both reverse and anatomical designs (either partial or total) (2, 8, 11, 12).

Hemiarthroplasty has been used with some success. However, in most cases, rotator cuff failure probably occurs due to the high inflammation and destruction of the shoulder joint. Matsuhashi *et al.* presented three patients treated with anatomical total shoulder replacement with simultaneous rotator cuff repair who experienced pain relief and significant ROM improvement (8). However, RSA is currently the gold standard for the treatment of cuff-deficient shoulders. It has been performed in four cases with promising results (11, 22).

It is crucial to note that in severe bone loss implantation of glenoid, a base plate may not be possible. We believe that early detection is the key to successful treatment. NGH classification may be helpful, with G1 showing a relatively preserved glenoid and enough bone stock to insert the implant. It is important to remember that long-term results are lacking. Another question remains as to whether shoulder arthroplasty with debridement of inflammation stops further bone loss and degeneration. Snoody *et al.* describe the patient as repeatedly requiring subsequent operations due to a loosening of an endoprosthesis (23). Undoubtedly, recurrent inflammation is a big challenge

here. Therefore, before deciding to treat the surgical shoulder with NA, we should use neurosurgical treatment options and seek to improve shoulder function with conservative treatment. Additionally, it is important not to perform arthroplasty in the face of increasing inflammation.

The available literature shows that effective neurosurgical treatment often stops the progression of the syrinx (7, 12). Therefore, before making a decision about shoulder surgery, a neurosurgical assessment is necessary (24, 25).

After analysing the most common clinical symptoms, useful diagnostic tests and various treatment options, we created an algorithm to help guide the management of patients with shoulder NA in SM (Fig. 7).

The main limitation of our study is the variety of analysed articles, both in terms of data availability and presentation. This condition is so rare that we have to rely on case reports and case series, which makes it difficult to create a homogeneous group with specific clinical data.

Conclusion

Shoulder NA due to SM is a devastating and progressive condition with weakness, ROM limitation, swelling and pain. Overlapping neurological symptoms, such as dissociated sensory loss, often make treatment difficult. Every patient with unexplained shoulder degeneration should be evaluated for SM. Both conservative and surgical treatments may be a good solution, especially for reducing symptoms such as pain and swelling. However, endoprosthesis should be considered in order to regain shoulder function.

ICMJE conflict of interest statement
The authors declare no conflict of interest. Przemysław Lubiatowski is an associate editor on the editorial board of EFORT Open Reviews. Przemysław Lubiatowski was not involved in the review or editorial process for this paper, on which he is listed as an author.

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