JABŁOŃSKI, Jędrzej, NADOLNY, Filip, KANIA, Martyna, GRUBSKI, Damian, BARTKOWIAK, Hanna, ADAMOWSKA, Agnieszka, ZIARNIK, Kacper, and ŚNIATAŁA, Alicja. Surgical Treatment of the Glenohumeral Joint for Indications of Instability and Pain in Hypermobile Ehlers-Danlos Syndrome – Systematic Review. Quality in Sport. 2024;35:56292. eISSN 2450-3118.
https://dx.doi.org/10.12775/QS.2024.35.56292
https://apcz.umk.pl/QS/article/view/56292

The journal has been 20 points in the Ministry of Higher Education and Science of Poland parametric evaluation. Annex to the announcement of the Minister of Higher Education and Science of 05.01.2024. No. 32553.
Has a Journal's Unique Identifier: 201398. Scientific disciplines assigned: Economics and finance (Field of social sciences); Management and Quality Sciences (Field of social sciences).
Punkty Ministerialne z 2019 - aktualny rok 20 punktów. Załącznik do komunikatu Ministra Szkolnictwa Wyższego i Nauki z dnia 05.01.2024 r. Lp. 32553. Posiada Unikatowy Identyfikator Czasopisma: 201398.
Przypisane dyscypliny naukowe: Ekonomia i finanse (Dziedzina nauk społecznych); Nauki o zarządzaniu i jakości (Dziedzina nauk społecznych).

The authors declare that there is no conflict of interests regarding the publication of this paper.
Received: 21.11.2024. Revised: 05.12.2024. Accepted: 16.12.2024. Published: 16.12.2024.

Surgical Treatment of the Glenohumeral Joint for Indications of Instability and Pain in Hypermobile Ehlers-Danlos Syndrome - Systematic Review

Jędrzej Jabłoński
Regional Hospital in Poznań
Juraszow 7/19 60-479
jedrzejkosma@gmail.com
https://orcid.org/0009-0009-6204-407X

Filip Nadolny
University Hospital in Poznań
Przybyszewskiego 49, 60-355 Poznań
nadolnyfilip@gmail.com
https://orcid.org/0009-0000-6433-5975

Martyna Kania
University Hospital in Poznań
Przybyszewskiego 49, 60-355 Poznań
martyna.kania@outlook.com
https://orcid.org/0009-0006-4400-0258

Damian Grubski
Józef Struś Multispecialist Municipal Hospital
Szwajcarska 3, 61-285 Poznań
damianxgrubski@gmail.com
https://orcid.org/0009-0003-9501-9950

Hanna Bartkowiak
Józef Struś Multispecialist Municipal Hospital
Szwajcarska 3, 61-285 Poznań
hannabartkowiak22@gmail.com
https://orcid.org/0009-0000-6914-4908

Agnieszka Adamowska
Józef Struś Multispecialist Municipal Hospital
Szwajcarska 3, 61-285 Poznań
a.adamowska12@gmail.com
https://orcid.org/0009-0009-1977-2522

Kacper Ziarnik
Regional Hospital in Poznań
Juraszow 7/19 60-479
kacper.ziarnik@gmail.com
https://orcid.org/0009-0006-4676-3232

Alicja Śniatała
Regional Hospital in Poznań
Juraszow 7/19 60-479
ala.sniatala@gmail.com
https://orcid.org/0009-0003-8488-3268

ABSTRACT

Introduction

Ehlers-Danlos Syndrome (EDS) is a group of connective tissue disorders, the essence of which are abnormalities in the function of collagen. One type of EDS with an unknown genetic basis is the hypermobile form of EDS (hEDS). hEDS manifests mainly in musculoskeletal defects, as well as chronic pain. There is extremely high need to correctly diagnose and then correctly treat patients with hEDS. Initially conservative treatment is used, but if patients fail to achieve relief, surgical treatment is considered.

Aim of the study

The purpose of the authors was to gather all the information regarding hEDS in terms of surgical treatment. The authors focused mainly on finding the advantages and disadvantages of the various methods and also on collecting information on the indications for choosing the given surgical methods.

Materials and methods

The methodology of the literature search involved using the keywords: „hypermobile Ehlers-Danlos syndrome", „shoulder instability", „joint hypermobility", „chronic pain" and „Latarjet". The search terms were entered into the PubMed database. References include systematic reviews, but also clinical trials, as well as case reports.

Conclusion

Currently available literature does not offer enough information to undertake standardized management of patients with hEDS. Not only is there a lack of studies on groups of patients with hEDS, but also on large groups of patients without hEDS, making it impossible to choose the best method based on the available clinical trials. Moreover, through the lack of such studies, it becomes impossible to truly assess whether patients with hEDS have a higher rate of surgical failure and thus a higher risk of inappropriate management of a specific patient with hEDS.

Keywords: Hypermobile Ehlers-Danlos syndrome, shoulder instability, joint hypermobility, chronic pain, Latarjet

Introduction

Ehlers-Danlos syndrome is a genetically heterogeneous group of inherited connective tissue disorders, the essence of which are abnormalities in the structure and/or function of collagen, fibrillin and elastin.[1] The main disorders caused by this syndrome will be related to the gastrointestinal, cardiovascular and musculoskeletal systems.[2] One of the subtypes of EDS is hypermobile Ehlers-Danlos syndrome (hEDS), which will be the focus of this paper.[2] The EDS nosology has evolved over the past 30 years (1988 Berlin criteria,[3] 1998 Villefranche nosology,[4] 1998 Brighton criteria,[5] 2017 EDS classification[6]) and there are currently 13 subtypes of this disease.[2]

HEDS is probably the most common connective tissue disorder in the world.[1] Suspicion of hEDS arises when a patient reports musculoskeletal complaints, including hypermobility, subluxations and joint dislocations.[1] These complaints cause not only chronic and severe disability but also a significant reduction in quality of life and significantly negatively affect the mental health of EDS patients.[7] In our article, we attempted to gather all currently available information and reports on the effectiveness of surgical treatment of the shoulder joint for indications of instability in patients suffering from hEDS.

Epidemiology

The exact incidence of EDS is unknown, but some estimates suggest that the disease affects about 1 in 5,000 to 1 in 20,000 people all over the world.[2] No increased predisposition to the disease is found due to ethnicity.[7] It has been shown that the diagnosis of hEDS is made more often in women than in men, but in other types of EDS the prevalence is similar in males and females.[7]

Pathogenesis of EDS

The phenotypes of individual EDS subtypes, and thus the genes responsible for them, differ from one another.[7] Current efforts are focused on gathering common elements of the various pathomechanisms to create a universal model for this disease.[7] Historically, the first types of EDS with identified molecular bases were associated with defects in the primary structure, processing, or modification of fibrillar procollagen types I, III, and V6.[7] Interestingly, regarding the hEDS variant, there is still a lack of clear information about the gene and the exact molecular pathomechanism of this form of EDS.[8] Therefore, the diagnosis of hEDS is based on clinical criteria observed during physical examination, the presence of connective tissue disorders in family members of the diagnosed patient, and the exclusion of other potential

causes of joint hypermobility.[7] These criteria were revised by the International Consortium on Ehlers-Danlos Syndromes and Hypermobility Spectrum Disorders in 2017.[8]

It is worth noting that the shoulder joint is particularly susceptible to the complication of recurrent instability in hEDS due to its anatomy. The humeral head is constrained by a very shallow glenoid cavity, and the primary role in maintaining stability is played by soft tissue structures. This provides an exceptionally wide range of motion but also predisposes the joint to instability. Stability, which depends on a complex balance between static (related to the capsulo-labro-ligamentous complex) and dynamic (the rotator cuff and biceps tendons) soft tissue stabilizers, can be compromised by the insufficiency of these structures, as is the case in patients with hEDS.

Clinical Characteristics of hEDS

hEDS manifests with soft and hyperextensible skin, abnormal wound healing, easy bruising, and musculoskeletal defects.[2] One of the most common symptoms, and often the initial basis for diagnosis, is generalized joint hypermobility and joint instability, which can lead to subluxation or dislocation of the joint.[9] It's worth noting that these conditions may occur even after minor injuries or at rest.[9] They are considered one of the most burdensome features of hEDS for patients and also predispose them to osteoarthritis.[9]

Musculoskeletal symptoms in hEDS are often accompanied by neuropathic and nociceptive pain, a tendency for easy bruising, and atrophic scars.[9] Additionally, patients with hEDS experience a higher incidence of abdominal hernias, pelvic organ prolapse, ascending aortic dilation, mitral valve prolapse, swallow and phonation disorders, and functional bowel disorders compared to the general population.[9] They may also have a higher prevalence of chronic fatigue, irritable bowel syndrome, temporomandibular joint dysfunction, and sleep disturbances.[1] These symptoms significantly impact the mental health of patients with hEDS.[10] A strong association has been established between hEDS and anxiety disorders and depression[10], and there is increasing evidence linking hEDS to eating and neuro-developmental disorders.[10] The extensive symptoms, affecting both physical and psychological well-being, underscore the need for a multidimensional approach in hEDS treatment.[10] Therefore, in addition to pharmacological treatment, rehabilitation, and surgery, hEDS management includes psychotherapy and psychiatric care.[10]

Treatment Methods - Clinical Management of hEDS

EDS is not curable, and management depends on the subtype of EDS.[8] Ideally, a multidisciplinary care team should oversee the care of patients with hEDS, which includes a primary physician, geneticist, and specialists, particularly in orthopedics.[8] Currently, the scientific literature lacks clear evidence-based guidelines for managing patients with hEDS, which means that clinical decisions are primarily based on the experience of individual physicians.[8]

Regarding dysfunctions of the musculoskeletal system, physical therapy and rehabilitation tailored to the specific complaints and limitations of the patient, such as recurrent shoulder dislocations, play a key role.[8] The literature mentions low-resistance exercises, such as swimming, which strengthen core muscles and indirectly increase joint stability by reinforcing the shoulder girdle, thus reducing the frequency of dislocation episodes.[8] Exercises aimed at increasing muscle strength also aim to improve proprioception.[9] Throughout the treatment, the emphasis is primarily on improving movement patterns.[9] Orthopedic braces and splints are also utilized, allowing for rest of hypermobile joints and reducing the risk of dislocation during daily activities.[9] When conservative treatment fails, surgical options are considered, indicated for issues such as impingement disorders, dysplasias, and cord/nerve entrapment with neurological deficits.[9] An integral part of the clinical picture for patients with hEDS is chronic or acute pain, which is often quite severe.[11] This pain may be widespread or localized, for example, in the shoulder girdle.[11] It frequently serves as the initial symptom of hEDS and is a direct reason for patients seeking help.[11] Notably, there is currently a lack of clear guidelines regarding pain management and treatment strategies, primarily due to the insufficient number of studies on treatment methods for hEDS.[11]

The treatment of hEDS must be multidimensional, addressing not only pain management but also psychological support and treatment of musculoskeletal disorders, with rehabilitation and activity modification being the first line of intervention.[12] Rehabilitation aims to achieve dynamic stability that protects against dislocations during daily activities.[12] This is accomplished by improving scapulohumeral coordination, correcting underlying scapular dyskinesia, and strengthening the periscapular stabilizers, rotator cuff muscles, deltoid muscle, and scapular stabilizers.[12] Recurrent joint dislocations are reported by as many as 95% of patients, leading most to consider surgical treatment at some stage of their management.[12] It is important to emphasize that surgical treatment is significantly complicated by soft tissue insufficiency, which results in structural instability of the skin and susceptibility to vascular damage, as well as the occurrence of intraoperative bleeding and postoperative hematomas.[12]

Additionally, delayed wound healing has been observed, attributed to fibroblast dysfunction, which is associated with a higher risk of postoperative complications.[12]

Most surgical interventions for shoulder treatment in hEDS patients attempt to reduce capsular laxity by applying ligamentous or bony structures.[12] Surgical management of shoulder instability in this patient population typically involves either arthroscopic or open surgical techniques, each with its own unique set of advantages and considerations. It is worth noting that standard treatment methods, such as the Bankart repair or the Latarjet procedure, are ineffective in the long term for patients with shoulder instability due to hEDS, prompting the search for alternative surgical approaches.[13]

There are publications reporting positive outcomes from arthroscopic shoulder stabilization procedures, such as Bankart repairs and remplissage techniques, but they have only good short-term outcomes in patients with EDS.[13] However, the long-term durability of these procedures in this patient population jest questionable, as the inherent tissue laxity associated with the disorder may increase the risk of recurrent instability. Therefore, alternative surgical treatment methods are being sought, which will be presented in the following subsections.

Arthroscopic Capsular Plication

The first method to be described will be arthroscopic capsular plication which has emerged as a viable treatment option for shoulder instability in patients with hEDS. The available evidence suggests that arthroscopic capsular plication can be an effective treatment for shoulder instability in patients with hEDS.[14] In the literature, this method is described, among others, in the course of multidirectional shoulder instability in patients with hEDS, in a publication authored by Victor Housset and Geoffroy Nourissat.[15] The main goal of the procedure is to reduce a global volume of the glenohumeral joint to improve the postoperative rehabilitation by improving the proprioception.[15] Verification of the indications for this operative procedure is based on a physical examination of the patient and an evaluation of the degree of instability of the glenohumeral joint, which is both assessed through the anterior and/or posterior apprehension test, a sulcus sign, a Gagey sign and clinical signs of joint hyperlaxity included in the Beighton criteria.[15]

In order to perform the procedure, all attempts at conservative treatment, including but not limited to the use of orthoses and compression garments, should be conducted beforehand, as these can help manage pain.[15] Preoperative imaging studies are necessary: standard plain X-ray radiographs should be performed to rule out any bony changes, followed by an arthro-CT scan or arthro-MRI to exclude any potential capsular-labral complex lesions and to visualize a

patulous redundant inferior capsule, which allows for the identification of anatomical changes within the joint.[15]

Indications for this surgery include multidirectional shoulder instability persisting for six months after attempts at conservative treatment, the presence of hEDS and a decision made by a multidisciplinary team.[15] In contrast, contraindications for the arthroscopic capsular plication procedure include the presence of anterior and/or posterior capsular-labral lesions, significant bony defects on the humeral and/or glenoid sides, a voluntary component, and advanced arthritis of the glenohumeral joint.[15]

The main advantages of this surgical method are that it is less invasive than open procedures, provides optimal exploration of both the anterior and posterior capsular-labral complex, and does not involve aggression to the labrum or cartilage.[15] However, the primary disadvantages of arthroscopic capsular plication include the risk of losing part of the range of motion, particularly in external rotation, as well as higher costs compared to open procedures.[15] Unfortunately, the authors of the studies have shown that the failure rate of this operation is higher in patients with hEDS than in those without the condition.[15]

The authors indicate that the main goal of arthroscopic capsular plication is not only to improve shoulder function but also to alleviate the patient's pain, thereby enhancing their quality of life.[15] Alberta et al. demonstrated in their work that arthroscopic plication was effective in reducing anterior translation and external rotation, although there was a partial loss of range of motion.[16]

Positive results of treatment using arthroscopic capsulorrhaphy for multidirectional instability (MDI) in patients with hEDS were also reported in a publication by Galano et al.[17] In this study, they presented the case of a 16-year-old girl with hEDS who experienced a reduction in shoulder instability 21 months after undergoing arthroscopic capsular plication.[17]

Arthroscopic posterior bone block augmentation and Latarjet

Positive outcomes from treatment using bone block augmentation via an arthroscopic approach are reported by Daniel Grant Schwartz et al. in their publication on managing posterior shoulder instability in patients with hEDS and multidirectional instability (MDI). [18] The publication describes the implantation of a posterior bone block with an iliac crest bone graft in a patient diagnosed with hEDS and MDI.[18] The effectiveness of this method is evidenced by the authors' follow-up conducted 12 months post-surgery. Improvements were noted in the Walch-Duplay Score, indicating the degree of shoulder instability, which increased from 40 to 85, and in the Rowe Score assessing shoulder stability, motion, and function, which

rose from 15 to 80, indicating a favorable evaluation.[18] Although the iliac crest bone graft technique is typically performed using an open approach, attempts have also been made to carry out this procedure arthroscopically, as demonstrated by the aforementioned authors.

Further insights are provided by Armstrong et al., who describe an arthroscopic technique combining capsular plication with iliac crest autograft or distal tibial allograft.[19] The indication for this surgery was recurrent multidirectional shoulder instability, and the technique involves completely arthroscopic augmentation of bone grafts along the anterior and posterior margins of the glenoid.[19] The authors cite one of the advantages of the procedure as being less invasive compared to open techniques.[19] Additionally, this operation reduces soft tissue laxity through capsular repair and enhances shoulder stability using both anterior and posterior bone grafts.[19] Another benefit of this technique is that it preserves key anatomical structures, including the pectoralis minor muscle, the coracoid process, and the tendon of the subscapularis muscle.[19] However, the procedure is technically challenging due to difficulties in achieving the correct angle for screw placement, as the conjoint tendon remains intact [19]. The authors also note several drawbacks, including the risk of graft resorption and higher costs associated with harvesting allografts.[19] They also observe that when harvesting bone grafts from patients with hEDS, there may be local complications at the graft site, as this group is particularly prone to wound complications.[19] Currently, the literature lacks definitive studies indicating the superiority of one technique over another, and patients with hEDS present additional challenges.

Another surgical technique related to bone grafting and the glenohumeral joint is the Latarjet procedure, which is indicated for anterior instability in the glenohumeral joint.[20] The surgical technique involves harvesting a piece of the coracoid process along with the coracobrachialis muscle and the short head of the biceps, and implanting these structures in the area of the anterior rim of the glenoid.[20] The coracoid graft extends the articulating glenoid arc, reducing the risk of recurrent anterior dislocation of the glenohumeral joint and enhancing dynamic stability during abduction and external rotation.[20] Indications for this operation include failed shoulder stabilization procedures, loss of more than 20% of the anteroinferior glenoid bone, significant damage to the shoulder muscles and ligaments contributing to instability, engaging Hill-Sachs lesions, and recurrent instability in competitive athletes.[20] There are also contraindications for this procedure, including coracoid fracture, voluntary dislocations, and irreversible rotator cuff damage.[20]

The main drawback of the Latarjet procedure is that it only addresses anterior dislocations, while patients with hEDS typically present with multidirectional instability

(MDI), which may render this technique insufficient.[12] Furthermore, addressing anterior instability with this technique may lead to more pronounced posterior instability in patients with bidirectional anteroinferior instability.[12]

The effectiveness of methods involving bone grafts is also highlighted by Andrew Homere et al., who suggest that procedures for bone augmentation, which increase the articular surface of the shoulder and do not rely on stabilization through soft tissues, should be preferred for patients with hEDS.[21]

Bilateral anterior and posterior glenohumeral stabilization using Achilles tendon allograft augmentation or tibialis tendon allografts

The surgical technique known as stabilization using Achilles tendon allograft augmentation involves fixing the Achilles tendon allograft to the anterior glenoid rim, spanning from the 6 o'clock position to the 12 o'clock position, using previously placed suture anchors along with an additional anchor placed anterosuperiorly, which allows for the reconstruction of the anterior labrum.[22] This technique enables the replacement of the lax and scarred capsule with new collagenous tissue, which is more durable and reduces the risk of further shoulder dislocations.[22] Procedures based on allografts are considered salvage operations after previous surgical treatments have failed to yield improvement.[12] Chaudhury S. et al. discuss its effects by presenting a case of a 28-year-old woman with hEDS and a 10-year history of recurrent shoulder subluxations who underwent this procedure and reported successful outcomes three years post-operation.[22] Indications for this procedure include the ineffectiveness of conservative treatment followed by the failure of surgical treatments such as arthroscopic capsular plication, open capsular shift, and coracoid bone block procedures.[22]

The effectiveness of stabilization using Achilles tendon allograft raises doubts in the publication by MacDonald et al., where a total long-term failure rate was found in 11 out of 16 shoulders after an average of 27 months following the use of Achilles tendon allografts in patients with multidirectional shoulder instability.[23] In the group of 11 patients with operation failure, 6 failures were due to refractory pain and 5 failures were due to recurrent instability.[23] The average time from operation to allograft failure was 22 months, indicating that this method has poor long-term outcomes.[23] Another publication by Alcid et al. reports high effectiveness in utilizing allograft tendon with tibialis tendon allografts.[24] This publication presents the results of revision anterior stabilization in 15 patients, with 13 of them showing surgical success evaluated after a minimum of 2 years of follow-up.[24]

In a retrospective study on open Shoulder Anterior Capsular Reconstruction with tibialis anterior tendon allograft, which described 10 shoulders in 5 patients with hEDS, Dewing et al. reported a recurrence of shoulder instability in 60% of cases at a mean follow-up of 3.8 years.[25] Schoorl et al. presented the surgical outcomes of 4 patients, in which five shoulders with hEDS and severe anteroinferior or multidirectional instability were operated on.[26] The open capsular shift combined with Achilles allograft augmentation of the anterior capsule performed on these patients was evaluated for pain, range of motion, recurrent instability, subjective shoulder value, American Shoulder and Elbow Surgeons score, complications, and reoperations.[26] The average follow-up period was 3.6 years, and in four out of five cases, the procedure resulted in a lasting improvement in pain and shoulder stability.[26] The aforementioned literature sources indicate that allografts may be a treatment option for patients with hEDS and serve as an alternative salvage procedure to arthrodesis, especially for young or active patients.

Open capsular shift

In 1980, Neer and Foster described the open anterior-inferior capsular shift of the shoulder joint as a method for treating inferior and multidirectional shoulder instability.[27] This procedure involves detaching the capsule from the humeral neck to create two flaps, which are then proximally shifted to create an overlap, thereby reducing the excess volume of the capsule and strengthening its anterior portion.[27] In Neer and Foster's original study, only 1 out of 40 patients with MDI had unsatisfactory outcomes after more than 2 years of follow-up.[27] Pollock and colleagues applied the same technique in 49 patients, achieving stability in 96% of cases after an average follow-up of 5 years.[28] It is important to emphasize that although the results of this method were impressive, these studies did not include patients with hEDS, making it difficult to relate these findings to individuals with connective tissue disorders. This is because, in the case of hEDS, the joint capsule has less structural integrity, which may hinder the effectiveness of this procedure.

In the publication by Vavken et al., the results of open inferior capsular shift surgery performed for multidirectional shoulder instability in a group of 15 adolescents with hEDS, who did not improve with non-operative treatment, were presented.[29] The authors reported the outcomes of the open inferior capsular shift method after 7.5 years post-surgery using the American Shoulder and Elbow Surgeons score and an 11-point version of the Disabilities of Arm, Shoulder and Hand scale, with significant improvements noted in both scales.[29] Furthermore, Vavken et al. noted that as many as 64% of patients in this group returned to sports.[29]

A 270º capsulorrhaphy involving the anterior, inferior and posterior capsule

Broida et al. describe their own preferred technique for resupplying an unstable shoulder in a patient with hEDS. Namely, the authors of the publication promote a 270º capsulorrhaphy, involving the anterior, inferior, and posterior capsule. This technique uses absorbable PDS sutures without anchors, thanks to which not only tightening of the capsule occurs but as the sutures resorb, they cause an inflammatory reaction, which through scarring can further stiffen the joint capsule.[12]

Open and arthroscopic Bankart repair

Both the open and arthroscopic methods for treating Bankart lesions, which involve damage to the anteroinferior part of the glenoid labrum, are associated with good treatment outcomes, particularly in patients with glenoid labral tears. However, similar to arthroscopic capsular plication, poorer long-term results of such treatment are noted in patients with hEDS compared to those without.[12]

Glenoid osteotomy

In patients with hEDS who have a dysplastic or retroverted glenoid, an open wedge glenoid osteotomy may be considered.[12] Although there are no reported cases of this procedure in patients with hEDS, the correction of the glenoid alignment has yielded good results in healthy individuals with posterior shoulder instability. Patients with EDS and a dysplastic glenoid may benefit from osteotomy in combination with a procedure to reduce and reinforce the shoulder joint capsule.[12]

Thermal Capsulorraphy

Another surgical method mentioned in the literature is thermal-assisted capsulorrhaphy, which is promoted as a supposedly less invasive alternative to procedures such as capsular shift.[12] This method involves the arthroscopic delivery of radiofrequency energy through a monopolar or bipolar thermal probe to the synovial surface of redundant tissues such as the superior glenohumeral ligament, middle glenohumeral ligament, and anterior and posterior bands of the inferior glenohumeral ligament complexes.[30] This causes the collagen within the tissue to denature, leading to a lasting contraction of the joint capsule, which reduces capsular redundancy and consequently enhances the stability of the shoulder joint.[12] A characteristic feature of this method is that in the first few months after surgery, there is an impairment of

tissue biomechanical properties, which is associated with an initial increase in joint weakness, marking a significant drawback of this procedure.[12] Rolfes et al. conducted a systematic review comparing arthroscopic capsular plication to thermal capsulorrhaphy, which included four studies involving 112 shoulders with multidirectional instability treated via thermal capsular shrinkage.[31] The results of this study indicated that success rates ranged from 53% to 93%, with a cumulative successful return to activity in 81% of patients.[31]

In the publication by Galano et al., they indicate the high destructiveness of this method due to complications related to excessive heat, including chondrolysis and axillary nerve injury [17]. Massive chondrolysis, as a complication of thermal capsulorrhaphy, is also mentioned by other authors, which further highlights the scale of the problem and indicates that it is a complication considered to outweigh any potential benefits of performing this procedurę.[32, 33, 34, 35, 36]

The effects of treatment using this method are also reported by Aldridge et al., who performed the procedure bilaterally for bilateral shoulder multidirectional instability in a 9-year-old patient with hEDS, achieving improvement in the stability of both shoulder joints.[30] After two years of observation, the 9-year-old patient showed no instability in the left shoulder and only occasional subluxations in the opposite shoulder.[30] After two years of follow-up, Aldridge et al. re-evaluated the patient's quality of life, obtaining data indicating that the patient actively participated in cross-country running, swimming, and basketball.[30] She had no subluxations or dislocations in the left shoulder and reported an average of one subluxation every 4 to 6 weeks concerning the aforementioned right shoulder.[30] The symptoms accompanying these episodic subluxations were not as intense as before the operation, and the patient reported satisfaction and an improvement in her quality of life after the procedure.[30] For the authors of this publication, it was unclear why one arm experienced occasional instability despite both arms undergoing the same treatment protocols, however, it is assumed that this may be due to difficulties in replicating the same tissue reactions in different arms using the same thermal device.[30] This characteristic further undermines the effectiveness of thermal capsulorrhaphy and distances specialists and patients from choosing this method.

Chaudhury et al. stated that thermal capsulorrhaphy has a high failure rate and is not a recommended procedure for treating shoulder instability in patients with hEDS.[22] It is worth noting that there are currently no long-term controlled studies regarding MDI and thermal energy, which further works against this method.

Shoulder arthroplasty

Another method for treating unstable, painful shoulders in patients with hEDS is shoulder arthroplasty, which, along with arthrodesis described in the next subsection, is considered a salvage procedure used as a last resort in treating this condition in cases where patients experience frequent dislocations or have evidence of severe joint deterioration despite prior first-line surgical treatment.[12] In the publication by Broida et al., it is stated that this procedure should not be performed until all other conservative and surgical treatment options have been exhausted.[12] Skedros et al. described a case report of a patient with hEDS who underwent reverse total shoulder arthroplasty, but without satisfactory results, as the patient reported during follow-up visits that the surgery did not significantly improve shoulder function and only slightly reduced pain in that joint.[37] Additional reports on this method are provided by Rogers et al., who present the outcomes and complications of shoulder arthroplasty performed in a group of patients with hEDS and compare them with a corresponding group of patients without EDS.[38] This study included 10 patients with EDS who underwent 6 primary anatomical total shoulder arthroplasties and 4 reverse shoulder arthroplasties.[38] The mean follow-up was 60 months (range 25-97 months).[38] During follow-up visits, similar postoperative pain, range of motion, complications, and reoperations were noted in patients with EDS compared to controls.[38] Moreover, there was an improvement in pain levels measured by the visual analog scale (VAS) pain score (from 6.5 to 1.7, $P<.001$), as well as improvements in range of motion: elevation (from 96 to 138 degrees) and external rotation (from 36 to 57 degrees).[38] Complications were noted in three shoulders; in two cases, instability occurred, but it did not require reoperation, while one patient experienced an acromial fracture.[38] The conclusions from Rogers et al.'s study indicate that the group of patients with hEDS did not experience a higher number of complications compared to the control group, which included patients without collagen structure disorders.[38] However, it remains a fact that 30% of patients (3 out of 10) experienced complications, which represents a high risk that must be taken into account when choosing this particular surgical method.[38] Furthermore, to determine the actual effectiveness of shoulder arthroplasty, clinical studies on large groups of patients with hEDS would be necessary, which are currently lacking.

Shoulder arthrodesis

Another method described for treating glenohumeral joint instability and associated pain is shoulder arthrodesis. This procedure belongs to salvage procedures and is the last line of treatment for this condition in patients with hEDS, especially if arthritis is present.[19] It is used

in patients for whom previous conservative and surgical treatment methods have failed and who experience frequent dislocations or have evidence of severe joint deterioration.[12] Legato et al. describe a case report of a 25-year-old woman with a long history of left shoulder multidirectional instability in the setting of hEDS, in whom arthrodesis of this joint was performed.[39] The described patient underwent three previous surgeries to stabilize the shoulder but continued to suffer from pain and recurrent dislocations, which was the basis for the diagnosis of recurrent dislocations and the inclusion of salvage treatment.[39] The goal of shoulder arthrodesis is to obtain both glenohumeral and subacromial fusion with the shoulder positioned at 30 degrees of flexion, 30 degrees of abduction, and 30 degrees of internal rotation.[39] Once fixation is achieved, the glenohumeral and subchondral regions are compressed, and the empty spaces between the edges of the bony surfaces are filled with autologous bone from the previous cuts.[39] In postoperative observation, it was noted that the patient not only had no problems reaching her mouth with the operated limb but was also able to reach the top of her head and lumbar spine through scapulothoracic motion.[39]

Discussion

Treatment of hEDS must be multidirectional and must cover not only pain management, but also psychological support and, above all, treatment of musculoskeletal disorders, which are the primary cause of pain and of seeking help. Therapy in this population is complicated by the severe degree of instability, as well as the underlying connective tissue abnormalities of the joint. In addition to immediate pain management, physical therapy is the first-line treatment and should target dynamic kinetics, resting rotator cuff tone, and scapulothoracic mechanics.[12] However, unfortunately, often the specific nature of the disease, makes this management fail to bring relief to patients and it is necessary to implement stabilizing surgical treatment, which has a higher risk of failure than in patients without hEDS.[12] This treatment should focus on addressing capsular redundancy and reinforcement with allograft tissue. Only if these are not effective are bone block procedures considered.[12] If complaints persist and dislocations continue to recur, salvage treatment such as shoulder arthroplasty or arthrodesis is introduced.[12] However, it is worth noting that this is a simplified regimen and the optimal surgical treatment of shoulder instability in patients with hEDS has not yet been established in the literature.

The surgical interventions mentioned in this systematic review should be performed by an experienced surgeon who is familiar with patients with hypermobility and hEDS, as this is a group that is particularly at risk of failure and complications after surgery.[12] The crux of the problem in the topics described in our paper is that many of the surgical techniques mentioned

do not have studies on large groups of patients to test long-term efficacy in improving quality of life. Moreover, such studies are even more lacking within the initially small group of patients that constitute hEDS patients. This creates a significant problem and generates a great need for long-term studies on relatively large groups of patients with hEDS, as this is necessary in order to make comparisons between different surgical methods, comparing not only the methods themselves but also comparing their effectiveness between patients without underlying connective tissue disease and those suffering from hEDS. Due to the rarity of hEDS, there is a significant paucity of reports on both conservative and surgical treatment. Therefore, management should be individualized and based on research-proven findings, which are still lacking.

Future studies should aim to test whether the above-described procedures offer clinical benefit in patients with hEDS. Preoperative diagnosis of hEDS will allow informed consent to be obtained from patients regarding their higher level of surgical risk, as well as prepare the attending surgeon for the increased risk of postoperative complications that may occur in this patient population. Further research is needed on both orthopedic surgery in patients without hEDS and in patients with hEDS. Only studies on large groups of patients with long-term follow up can give scientifically validated data on the actual effectiveness of the various methods and be the basis for proposing the most effective methods to patients. This is extremely important, as proper management of unstable, painful shoulder joints will not only give patients relief from pain, but also reduce the risk of psychological complaints and many other possible complications in the course of hEDS.

Disclosure:

Author's contribution

Conceptualization: Jędrzej Jabłoński, Martyna Kania

Methodology: Hanna Bartkowiak

Software: Filip Nadolny

Check: Damian Grubski, Alicja Śniatała, Kacper Ziarnik

Formal analysis: Agnieszka Adamowska

Investigation: Jędrzej Jabłoński

Resources: Hanna Bartkowiak

Data curation: Martyna Kania

Writing - rough preparation: Filip Nadolny

Writing - review and editing: Kacper Ziarnik

Visualization: Damian Grubski

Supervision: Filip Nadolny

Project administration: Alicja Śniatała

Receiving funding: no specific funding.

All authors have read and agreed with the published version of the manuscript.

Funding Statement

This research received no external funding.

Institutional Review Board Statement

Not applicable.

Informed Consent Statement

Not applicable.

Data Availability Statement

Not applicable.

Acknowledgments

Not applicable.

Conflict of interest Statement

The authors deny any conflict of interest.

References:

1. Brad Tinkle, Marco Castori, Britta Berglund, Helen Cohen, Rodney Grahame, Hanadi Kazkaz et al. Hypermobile Ehlers–Danlos Syndrome (a.k.a. Ehlers–Danlos Syndrome Type III and Ehlers–Danlos Syndrome Hypermobility Type): Clinical Description and Natural History. Am J Med Genet Part C Semin MedGenet 175C:48–69. DOI 10.1002/ajmg.c.31538.

2. Stephanie Buryk-Iggers, Nimish Mittal, Daniel Santa Mina, Scott C. Adams, Marina Englesakis, Maxim Rachinsky et al. Exercise and Rehabilitation in People With Ehlers-

Danlos Syndrome: ASystematic Review. Arch Rehabil Res Clin Transl. 2022;4:100189. DOI: https://doi.org/10.1016/j.arrct.2022.100189.

3. Beighton P, De Paepe A, Danks, et al. International nosology of heritable disorders of connective tissue, Berlin, 1986. Am J Med Genet 1988;29:581–94.

4. Beighton P, De Paepe A, Steinmann B, Tsipouras P, Wenstrup RJ. Ehlers-Danlos syndromes: revised nosology, Villefranche, 1997. AmJMedGenet1998;77:31–7.

5. Grahame R, Bird HA, Child A, et al. The revised (Brighton 1998) criteria for the diagnosis of benign joint hypermobility syn drome (BJHS). J Rheumatol 2000;27:1777–9.

6. Malfait F, Francomano C, Byers P, et al. The 2017 international classification of the Ehlers-Danlos syndromes. Am J Med Genet Part C Semin Med Genet 2017;175:8–26.

7. Fransiska Malfait, Marco Castori, Clair A. Francomano, Cecilia Giunta, Tomoki Kosho, Peter H. Byers. The Ehlers–Danlos syndromes. Nat Rev Dis Primers. 2020 Jul 30;6(1):64. doi: 10.1038/s41572-020-0194-9. PMID: 32732924.

8. Malfait F, Francomano C, Byers P, Belmont J, Berglund B, Black J et al. The 2017 international classification of the Ehlers-Danlos syndromes. Am J Med Genet C Semin Med Genet. 2017 Mar;175(1):8-26. doi: 10.1002/ajmg.c.31552.

9. Hakim A. Hypermobile Ehlers-Danlos Syndrome. 2004 Oct 22 [updated 2024 Feb 22]. In: Adam MP, Feldman J, Mirzaa GM, Pagon RA, Wallace SE, Amemiya A, editors. GeneReviews. Seattle (WA): University of Washington, Seattle; 1993–2024.

10. Bulbena A, Baeza-Velasco C, Bulbena-Cabré A, Pailhez G, Critchley H, Chopra P et al. Psychiatric and psychological aspects in the Ehlers-Danlos syndromes. Am J Med Genet C Semin Med Genet. 2017 Mar;175(1):237-245. doi: 10.1002/ajmg.c.31544. Epub 2017 Feb 10. Erratum in: Am J Med Genet A. 2017 Dec;173(12):3241. doi: 10.1002/ajmg.a.38488.

11. Chopra P, Tinkle B, Hamonet C, Brock I, Gompel A, Bulbena A et al. Pain management in the Ehlers-Danlos syndromes. Am J Med Genet C Semin Med Genet. 2017 Mar;175(1):212-219. doi: 10.1002/ajmg.c.31554. Epub 2017 Feb 10.

12. Broida SE, Sweeney AP, Gottschalk MB, Wagner ER. Management of shoulder instability in hypermobility-type Ehlers-Danlos syndrome. JSES Rev Rep Tech. 2021 Mar 24;1(3):155-164. doi: 10.1016/j.xrrt.2021.03.002.

13. Dala-Ali B, Penna M, McConnell J, Vanhegan I, Cobiella C. Management of acute anterior shoulder dislocation. Br J Sports Med. 2014 Aug;48(16):1209-15. doi: 10.1136/bjsports-2012-091300. Epub 2012 Jul 21.

14. Caprise PA Jr, Sekiya JK. Open and arthroscopic treatment of multidirectional instability of the shoulder. Arthroscopy. 2006 Oct;22(10):1126-31. doi: 10.1016/j.arthro.2006.08.002.

15. Housset V, Nourissat G. Arthroscopic Capsular Plication for Multidirectional Shoulder Instability in Hypermobile Ehlers-Danlos Syndrome Patients. Arthrosc Tech. 2021 Nov 18;10(12):e2767-e2773. doi: 10.1016/j.eats.2021.08.024.

16. Alberta FG, Elattrache NS, Mihata T, McGarry MH, Tibone JE, Lee TQ. Arthroscopic anteroinferior suture plication resulting in decreased glenohumeral translation and external rotation. Study of a cadaver model. J Bone Joint Surg Am. 2006 Jan;88(1):179-87. doi: 10.2106/JBJS.D.02974.

17. Galano GJ, Soldano D, Kippe MA, Ahmad CS. Arthroscopic shoulder suture capsulorrhaphy in a patient with Ehlers-Danlos Syndrome. Curr Orthopaedic Pract 2008;19:589-93. doi: https://doi.org/10.1097/BCO.0b013e328313a9a4.

18. Schwartz DG, Goebel S, Piper K, Kordasiewicz B, Boyle S, Lafosse L. Arthroscopic posterior bone block augmentation in posterior shoulder instability. J Shoulder Elbow Surg. 2013 Aug;22(8):1092-101. doi: 10.1016/j.jse.2012.09.011. Epub 2013 Jan 20.

19. Armstrong MD, Smith B, Coady C, Wong IH. Arthroscopic Anterior and Posterior Glenoid Bone Augmentation With Capsular Plication for Ehlers-Danlos Syndrome With Multidirectional Instability. Arthrosc Tech. 2018 Apr 23;7(5):e541-e545. doi: 10.1016/j.eats.2018.01.011.

20. Getz CL, Joyce CD. Arthroscopic Latarjet for Shoulder Instability. Orthop Clin North Am. 2020 Jul;51(3):373-381. doi: 10.1016/j.ocl.2020.02.002. Epub 2020 May 5.

21. Homere A, Bolia IK, Juhan T, Weber AE, Hatch GF. Surgical Management of Shoulder and Knee Instability in Patients with Ehlers-Danlos Syndrome: Joint Hypermobility Syndrome. *Clin Orthop Surg*. 2020;12(3):279-285. doi:10.4055/cios20103.

22. Chaudhury S, Gasinu S, Rodeo SA. Bilateral anterior and posterior glenohumeral stabilization using Achilles tendon allograft augmentation in a patient with Ehlers-Danlos syndrome. *J Shoulder Elbow Surg*. 2012;21(6):e1-e5. doi:10.1016/j.jse.2011.10.033.

23. Peter B. MacDonald, Sheila McRae, MSc, Jeff Leiter. Achilles Allograft Stabilization of the Shoulder in Refractory Multidirectional Glenohumeral Instability. Department of

Orthopaedic Surgery University of Manitoba Faculty of Medicine Winnipeg, Manitoba, Canada; doi: 10. 1097/BTE.0b013e31816938da.

24. Alcid JG, Powell SE, Tibone JE. Revision anterior capsular shoulder stabilization using hamstring tendon autograft and tibialis tendon allograft reinforcement: minimum two-year follow-up. J Shoulder Elbow Surg. 2007;16(3):268-272. doi:10.1016/j.jse.2006.07.008.

25. Dewing CB, Horan MP, Millett PJ. Two-year outcomes of open shoulder anterior capsular reconstruction for instability from severe capsular deficiency. Arthroscopy. 2012;28(1):43-51. doi:10.1016/j.arthro.2011.07.002.

26. Schoorl TJ, Nguyen NTV, van Noort A, Alta TDW, Sanchez-Sotelo J. Capsulorraphy with Achilles allograft augmentation for shoulder instability in patients with Ehlers-Danlos syndrome. J Shoulder Elbow Surg. 2021;30(4):865-870. doi:10.1016/j.jse.2020.07.019.

27. Neer CS 2nd, Foster CR. Inferior capsular shift for involuntary inferior and multidirectional instability of the shoulder. A preliminary report. J Bone Joint Surg Am. 1980;62(6):897-908.
28. Pollock RG, Owens JM, Flatow EL, Bigliani LU. Operative results of the inferior capsular shift procedure for multidirectional instability of the shoulder. J Bone Joint Surg Am. 2000;82-A(7):919-928. doi:10.2106/00004623-200007000-00003.

29. Vavken P, Tepolt FA, Kocher MS. Open inferior capsular shift for multidirectional shoulder instability in adolescents with generalized ligamentous hyperlaxity or Ehlers-Danlos syndrome. J Shoulder Elbow Surg. 2016;25(6):907-912. doi:10.1016/j.jse.2015.10.010.

30. Aldridge JM 3rd, Perry JJ, Osbahr DC, Speer KP. Thermal capsulorraphy of bilateral glenohumeral joints in a pediatric patient with Ehlers-Danlos syndrome. Arthroscopy. 2003;19(5):E41. doi:10.1053/jars.2003.50161.

31. Rolfes K. Arthroscopic treatment of shoulder instability: a systematic review of capsular plication versus thermal capsulorrhaphy. J Athl Train. 2015;50(1):105-109. doi:10.4085/1062-6050-49.3.63.

32. D'Alessandro DF, Bradley JP, Fleischli JE, Connor PM. Prospective evaluation of thermal capsulorrhaphy for shoulder instability: indications and results, two- to five-year follow-up. Am J Sports Med. 2004;32(1):21-33. doi:10.1177/0095399703258735.

33. Good CR, Shindle MK, Kelly BT, Wanich T, Warren RF. Glenohumeral chondrolysis after shoulder arthroscopy with thermal capsulorrhaphy. Arthroscopy. 2007;23(7):797.e1-797.e7975. doi:10.1016/j.arthro.2007.03.092.

34. Levine WN, Clark AM Jr, D'Alessandro DF, Yamaguchi K. Chondrolysis following arthroscopic thermal capsulorrhaphy to treat shoulder instability. A report of two cases. J Bone Joint Surg Am. 2005;87(3):616-621. doi:10.2106/JBJS.D.02158.

35. Lubowitz JH, Poehling GG. Glenohumeral thermal capsulorrhaphy is not recommended--shoulder chondrolysis requires additional research. Arthroscopy. 2007;23(7):687. doi:10.1016/j.arthro.2007.05.001.

36. Petty DH, Jazrawi LM, Estrada LS, Andrews JR. Glenohumeral chondrolysis after shoulder arthroscopy: case reports and review of the literature. Am J Sports Med. 2004;32(2):509-515. doi:10.1177/0363546503262176.

37. Skedros JG, Phippen CM, Langston TD, Mears CS, Trujillo AL, Miska RM. Complex Scapular Winging following Total Shoulder Arthroplasty in a Patient with Ehlers-Danlos Syndrome. Case Rep Orthop. 2015;2015:680252. doi:10.1155/2015/680252.

38. Rogers T, Werthel JD, Crowe MM, et al. Shoulder arthroplasty is a viable option in patients with Ehlers-Danlos syndrome. J Shoulder Elbow Surg. 2021;30(11):2484-2490. doi:10.1016/j.jse.2021.03.146.

39. Legato JM, O'Connell M, Fuller DA. Shoulder Arthrodesis. J Orthop Trauma. 2018;32 Suppl 1:S4-S5. doi:10.1097/BOT.0000000000001212.