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Capsulorraphy with Achilles allograft augmentation for shoulder instability in patients with Ehlers-Danlos syndrome

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Background: Surgical management of shoulder instability in patients with Ehlers-Danlos syndrome (EDS) remains challenging secondary to the pathologic nature of their connective tissue. Allograft reconstruction of the shoulder capsule in EDS has the potential to increase stability by providing healthier connective tissue. The purpose of this study was to report the surgical technique and outcome of open capsulorraphy and augmentation of the anterior capsule with an Achilles tendon allograft in patients with shoulder instability in the setting of EDS.
Methods: Five shoulders (4 patients) with EDS and severe anteroinferior or multidirectional instability underwent open capsular shift combined with Achilles allograft augmentation of the anterior capsule. Patients were evaluated for pain, motion, recurrent instability, subjective shoulder value, American Shoulder and Elbow Surgeons score, complications, and reoperations. The mean follow-up time was 3.6 years (range, 2-5 years).
Results: Shoulder stability was restored in 4 of 5 (80%) shoulders. At the final follow-up, the mean subjective shoulder value and American Shoulder and Elbow Surgeons scores were 84 and 77.3, respectively. One shoulder developed recurrent posterior instability after an injury 1.6 years after the index procedure. The mean pain visual analog scale was 7 preoperatively and 2 at the most recent follow-up. Before surgery, all patients reported the use of narcotic pain medication, whereas at the most recent follow-up, only the one patient who had experienced recurrence reported moderate pain. Except for the shoulder that required revision surgery for posterior shoulder instability, there were no complications or other reoperations.
Conclusion: Open capsulorraphy with Achilles allograft augmentation improved stability and pain in 4 of 5 shoulders with instability in the setting of EDS. In this small case series of patients with EDS, Achilles tendon allograft augmentation was safe and effective as a primary or revision surgical procedure for anterior shoulder instability. A larger patient cohort with longer follow-up is needed to confirm these findings.
Level of evidence: Level IV; Case Series; Treatment Study

Keywords: Ehlers-Danlos syndrome; joint hypermobility; shoulder; anterior inferior instability; Achilles tendon allograft

Ehlers-Danlos syndrome (EDS) is a hereditary heterogeneous connective tissue disorder. It is divided into 6 main types with 13 subtypes. In most types, EDS results from mutations in collagen encoding genes. EDS can cause a variety of musculoskeletal symptoms. A majority of complaints, especially in the Ehlers-Danlos hypermobility type,

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concern the shoulder.[16] Chronic arthralgia and instability oftentimes result in a poor Health-Related Quality of life index.[10] Instability in patients with EDS typically is managed with physical therapy directed to strengthening of the deltoid, rotator cuff, and periscapular muscles.[2,11] Most surgeons consider surgery a contraindication in this patient population, although attempted surgical correction of shoulder instability in EDS has been reported.[14,16]

Open or arthroscopic capsulorraphy for shoulder instability in patients with EDS is controversial due to the patient's inherent abnormal collagen properties, which leads to early plastic deformation of the capsule, failure, and recurrent instability.[3] Various alternative procedures have been described, including capsular augmentation with allograft and glenohumeral arthrodesis.[1,3,12,13,17] Capsulorraphy with allograft augmentation is particularly appealing, because it provides an opportunity to substitute abnormal EDS collagen with normal allograft collagen. Reported outcomes with allograft augmentation are limited to isolated case reports.[3,12]

We have adopted an anterior capsular shift, with or without labral repair augmented with Achilles tendon allograft reconstruction of the anterior capsule for patients with anterior shoulder instability in the setting of EDS, after a failure of nonoperative treatment with physical therapy. Because this procedure is only offered after failure of a good program of conservative treatment, the number of procedures performed to date is limited. The purpose of this study was to describe the surgical technique for capsulorraphy with Achilles allograft augmentation for anterior instability in patients with EDS and to report the outcome obtained in a small cohort of patients.

Methods

This is a retrospective case series of open anterior capsulorraphy augmented with an Achilles tendon allograft to reconstruct the anterior capsule, which was performed between 2010 and 2016 at 2 institutions in 5 shoulders (4 patients with EDS) in the setting of anteroinferior instability. A retrospective chart review was performed to record demographic and clinical data, complications, and reoperations. All patients were also contacted at the most recent follow-up to specifically assess their shoulder pain, stability, and overall function.

Patients

All patients included in this study were female, with a mean age at the time of Achilles allograft augmented capsulorraphy of 28.8 years (range, 21-42 years, Table I). They all had been diagnosed with EDS type III (hypermobility). The diagnosis of EDS had been performed by a geneticist according to specific EDS diagnostic criteria. All patients reported incapacitating shoulder instability since adolescence. All shoulders remained unstable despite an adequate program of physical therapy for several months, including rotator cuff, deltoid, and scapular stabilizing exercises. Four of the 5 shoulders had undergone 1 or more prior surgical procedures, which had failed to restore shoulder stability. The number and type of prior surgeries in the cohort are summarized in Table I. All patients complained of numerous subluxation or dislocation episodes occurring daily. Physical examination demonstrated multidirectional laxity and anterior apprehension with positive relocation. Radiographs and magnetic resonance imaging had been obtained in all shoulders. None of the patients had evidence of anterior glenoid bone loss.

Surgical technique

Surgery was performed with the patient in the semi–beach chair position on a shoulder table. A deltopectoral approach was performed in all cases. Exposure of the joint capsule was performed through a subscapularis tenotomy. Surgical separation of the subscapularis from the underlying anterior capsule using sharp dissection is particularly difficult in patients with EDS. The subscapularis-capsule plane was dissected from distal to proximal to take advantage of the more muscular nature of the subscapularis inferiorly. Next the subscapularis tendon was tagged with sutures and divided vertically leaving a 0.5 to 1 cm cuff laterally for later repair.

The thin anterior capsule was then detached circumferentially from the humeral neck, tagged with sutures and divided in a T fashion from lateral to medial aiming at the mid-equator of the glenoid. A Fukuda retractor was used to retract the humeral head posteriorly. The anterior labrum was inspected for the presence of a tear (Bankart lesion). Two or 3 suture anchors loaded with a heavy nonabsorbable suture were inserted into the anterior glenoid rim, and the anterior labrum and capsule were repaired using a horizontal mattress configuration. The end of these sutures was left uncut after knot tying, because they were used for fixation of the Achilles allograft medially.

The Achilles tendon allograft was then opened. The narrower distal end of the Achilles allograft was fixed to the medial glenoid

Table 1 Characteristics of the 5 shoulders included in this study

Patient	Sex	Age (yr)	Type	Side	Dominance	Prior surgeries	Follow-up time (yr)
1-right	F	29	I/III	R	R	1*	4.9
1-left	F	26	I/III	L	R	2†	2.5
2	F	26	II	R	R	1*	3.7
3	F	21	II	R	R	2‡	4.7
4	F	42	III	L	R	0	2.0

F, female; *R*, right; *L*, left.
* Open capsular shift.
† Bankart repair followed by capsular shift.
‡ Bankart repair followed by revision Bankart repair and capsular shift.

neck with the same sutures that had been used for repair of the anterior labrum and capsule (Fig. 1). The Fukuda retractor was then removed from the joint, and a shifting repair of the native anterior capsule was performed, as described by Neer and Foster,[15] with the arm in approximately 30° of abduction and 30° of external rotation. The inferior capsular flap was advanced superolaterally and repaired with multiple sutures. The superior capsular flap was then advanced inferolaterally over the inferior capsular flap and repaired. Lateral traction was then applied to the Achilles tendon allograft to determine the ideal length, and the allograft was fixed laterally with multiple nonabsorbable sutures. The subscapularis tenotomy was then repaired anatomically with multiple interrupted sutures, including sutures in the interval region between the anterior leading edge of the supraspinatus and the superior edge of the subscapularis. The rest of the closure was routine.

Postoperative management

After surgery, all patients used a shoulder immobilizer for 6 weeks. After 6 weeks, active assisted range of motion exercises in elevation, external rotation, and internal rotation were initiated along with isometric scapular strengthening. Rotator cuff and deltoid strengthening were initiated at week 10. All patients were recommended to avoid rapid progression through range of motion exercises to decrease the possibility of overstretching of the soft tissues and recurrence.

Patient evaluation

Patients were evaluated at regular intervals (6 weeks, 3 months, 6 months, 1 year, and yearly thereafter). The mean follow-up time was 3.6 years (range, 2-5 years, Table I). At the most recent follow-up, shoulders were evaluated for pain using a visual analog scale (VAS), subjective shoulder value,[7] motion, stability, and American Shoulder and Elbow Surgeons scores.

Results

Tables I and II summarize the outcomes for all 5 shoulders (4 patients) included in the study. Overall, capsulorraphy with Achilles allograft augmentation led to improvements in shoulder stability and pain. The mean VAS for pain improved from 7 preoperatively to 2 at the most recent

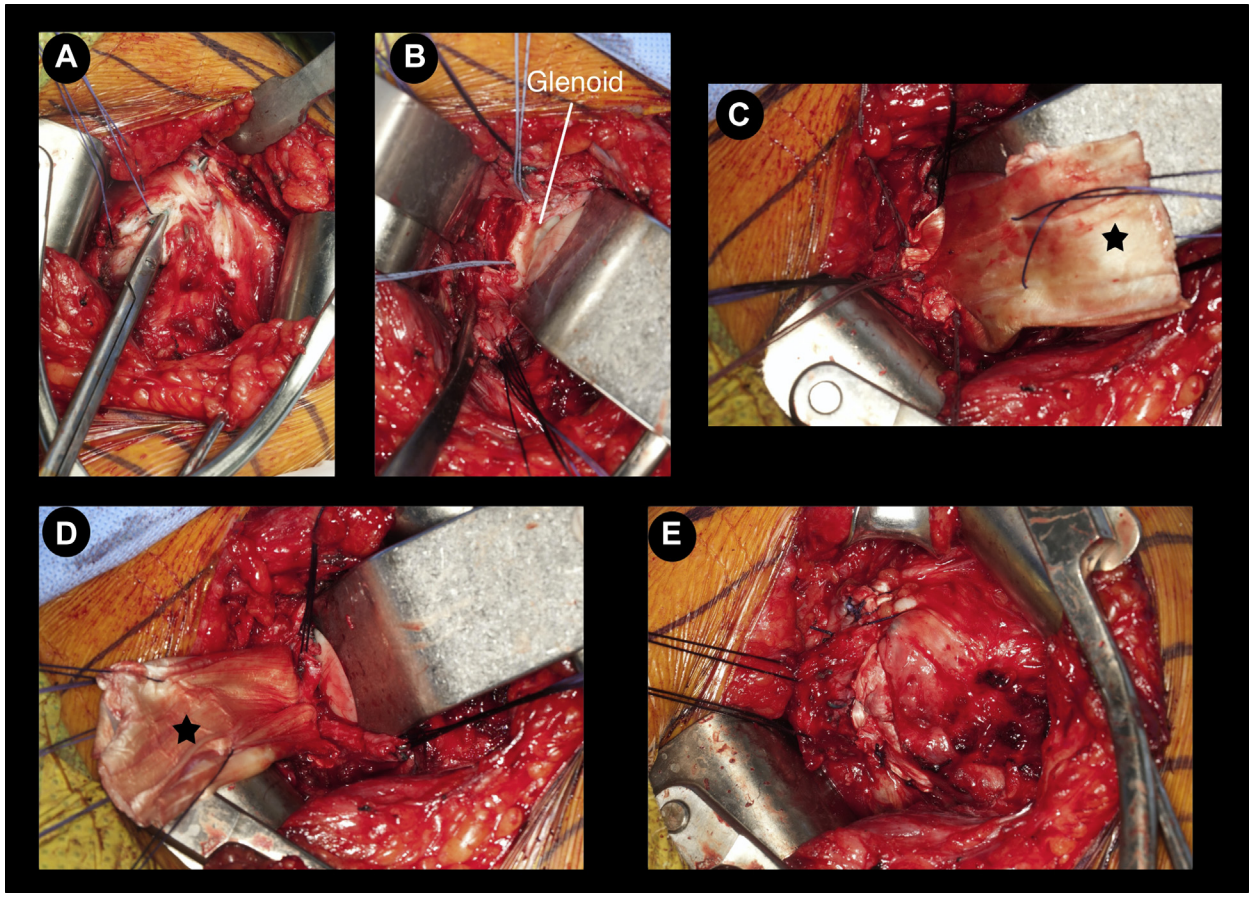

Figure 1 Surgical technique for capsulorraphy with Achilles allograft augmentation in patients with Ehlers-Danlos syndrome (left shoulder shown). (**A**) Dissection of the plane between the subscapularis and anterior capsule is best performed from inferior to superior. (**B**) Anchors placed into the glenoid to repair the labrum, reattach the capsule, and attach the medial aspect of the allograft. (**C**) Achilles allograft secured medially. (**D**) Capsular flaps prepared for capsular shift under the allograft. (**E**) Lateral fixation of the Achilles allograft before repair of the subscapularis tenotomy. The star symbol indicates the location of the Achilles tendon allograft.

Table 2 Outcomes for the 5 shoulders included in this study

Patient	VAS pain		FE		ER		IR		Stable	SSV	ASES	Reoperation
	P	F	P	F	P	F	P	F				
1-right	3	2	180	180	90	70	T4	T10	Yes	90	85	No
1-left	8	2	10	175	70	70	GT	T10	Yes	90	85	No
2	8	2	180	170	100	40	T4	L1	Yes	80	83	No
3	8	6	150	150	70	40	T4	T9	No	70	48	Yes
4	8	0	140	180	100	60	T7	T12	Yes	90	100	No

VAS, visual analog score; *FE*, forward elevation; *ER*, active external rotation; *IR*, active internal rotation; *SSV*, subjective shoulder value; *ASES*, American Shoulder and Elbow Surgeons score; *P*, preoperative; *F*, follow-up; *GT*, level of greater trochanter.

follow-up. Before surgery, all patients reported the use of narcotic pain medication, whereas at the most recent follow-up, only 1 patient reported moderate pain (patient 3, most recent VAS for pain 6); this patient had sustained a reinjury 1.6 years after the index anterior capsule Achilles allograft augmentation procedure, developed recurrent unidirectional posterior instability, and underwent surgery for augmentation of the posterior capsule with an Achilles tendon allograft. At the most recent follow-up, the mean subjective shoulder value and American Shoulder and Elbow Surgeons scores were 84 and 77.3, respectively. Except for the shoulder that required revision surgery for posterior shoulder instability, there were no complications or other reoperations (Fig. 2).

Discussion

Shoulder instability in patients with EDS can become extremely incapacitating. Physical therapy is the main treatment modality recommended for shoulder instability in EDS.[2,11] However, when a well-structured program of

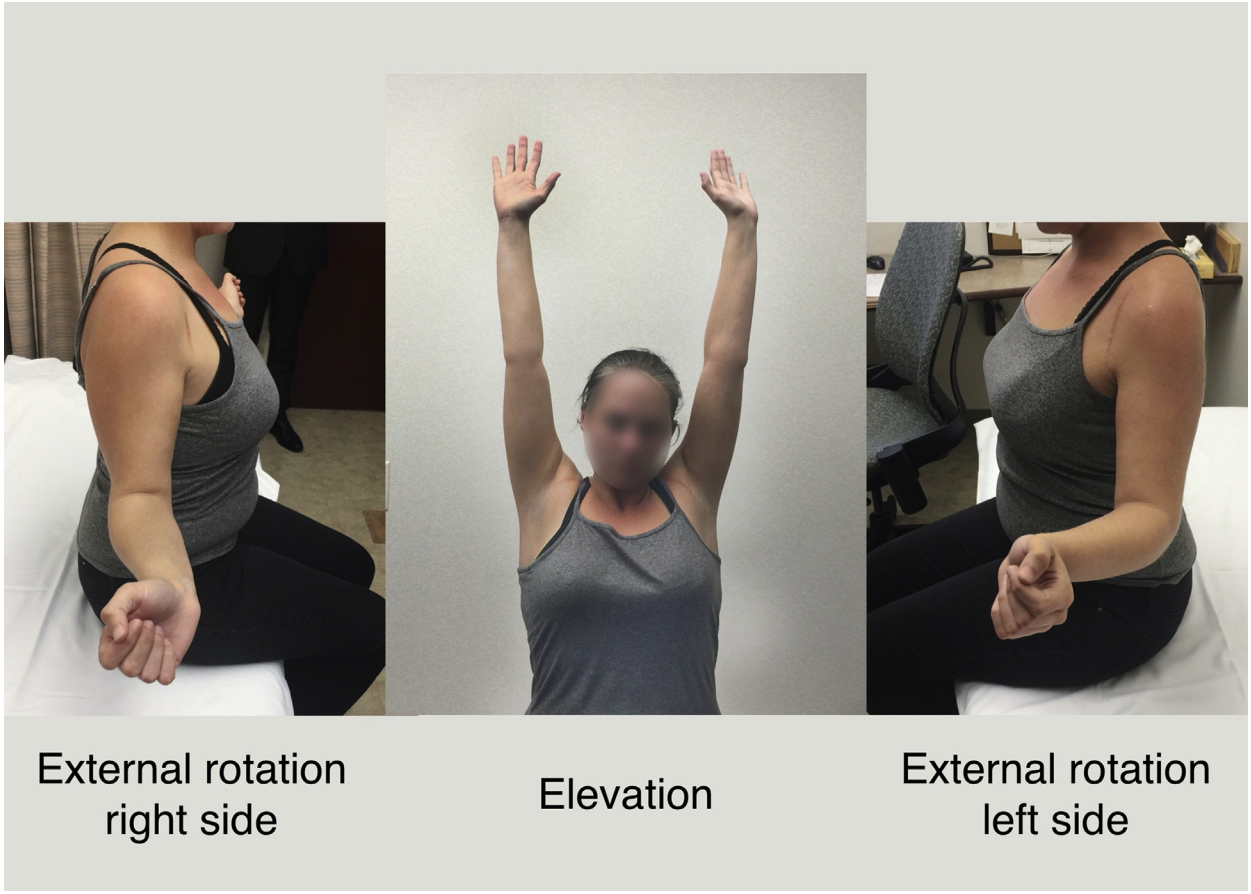

Figure 2 Postoperative range of motion 1 year after bilateral capsulorraphy with Achilles allograft augmentation in a patient with Ehlers-Danlos syndrome.

nonoperative management remains unsuccessful, surgery is indicated. The abnormal laxity of the soft tissues and predisposition for plastic deformation in patients with EDS has translated into high reported failure rates after conventional open or arthroscopic shoulder procedures.[9,10,13,16] As such, many patients with debilitating shoulder instability in the setting of EDS are told that surgery is not an option for them. The results of our study indicate that capsulorraphy augmented with allograft tendon can restore shoulder stability in patients with EDS. However, these results are limited by the small sample size and short-term follow-up.

Several procedures have been described for surgical management of shoulder instability in EDS.[1,3,12,13,17] The prevailing consideration when analyzing these studies is that EDS tissue will plastically deform over time and lead to a relapse in symptoms.[3] Vavken et al[17] described a case series of 18 open inferior capsular shifts in 15 patients with generalized hyperlaxity or EDS. At a mean follow-up of 7.5 years, 87% had improvement of stability and reduction of pain, and 64% of the patients were able to return to sports. However, only 47% reported no further episodes of instability and 7% complained of moderate or severe pain. Although one case of shoulder instability in EDS has been reported after arthroscopic capsular plication combined with anterior and posterior bone augmentation,[16] shoulder instability in EDS does not seem to be associated with substantial glenoid bone loss. In fact, many consider EDS a relative contraindication to bone procedures, such as transfer of the coracoid process, especially when there is an element of voluntary instability.[6]

The use of allograft tendon represents an attractive alternative for shoulder instability in EDS. Allograft from donors without EDS provides ''healthy'' collagen. Allograft and augmented autograft reconstruction have been reported for lower extremity procedures in patients with EDS.[4,18] Allograft reconstruction of a symptomatic anterior cruciate ligament (ACL) tear has been reported in 2 patients with EDS. One allograft ACL reconstruction was performed using an Achilles tendon allograft.[4] The other ACL reconstruction was performed using an ipsilateral autograft ipsilateral hamstring tendon combined with a ligament augmentation and reconstruction system (LARS) ligament. These 2 patients were followed for 2 years, and both reported participation in regular preinjury activities, including pivoting sports. Reconstruction of a ruptured patellar tendon using a bone-patellar tendon-bone allograft was also reported in 1 patient with EDS, although the follow-up for the patient was only 6 months because the patient died of an unrelated aortic dissection.[8]

Chaudhury et al were first to report the use of Achilles allograft augmentation for shoulder instability in the setting of EDS.[3] They reported the case of a single patient who underwent augmentation of both the anterior and the posterior capsule in both shoulders, with a successful outcome. The shoulders included in our study underwent a capsular shift combined with augmentation of the anterior capsule, which was enough to restore stability in 4 of 5 shoulders. Karpyshyn et al described the use of an acellular dermal allograft for reconstruction of the posterior capsule in a patient with EDS.[12] However, they did not report the result of the procedure in terms of stability or overall outcome. Dewing et al[5] reported on 20 shoulders with severe anterior capsular deficiency stabilized using either tibialis anterior or semitendinosus allograft; 5 of the patients (10 shoulders) included in this study were diagnosed with EDS. Several patients with EDS reported return of instability, but the majority did not undergo further suegery.

Our study is not without limitations. This is a small series with relatively short follow-up. In addition, advanced imaging was not obtained during the evaluation of these shoulders to determine the degree of healing or incorporation of the allograft. The strengths of the study include the use of the exact same technique in all 5 shoulders and reporting of the outcome in a series of shoulders, as opposed to a single case report.

Conclusion

In this small case series, open capsulorraphy with Achilles allograft augmentation improved stability and pain in 4 of 5 shoulders with instability in the setting of EDS. Our results show that at midterm, Achilles tendon allograft augmentation is effective as a primary or revision surgical procedure for anterior shoulder instability in patients with EDS. Longer follow-up is needed to determine the durability of this procedure.

Disclaimer

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References

1. 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;7:e541-5. https://doi.org/10.1016/j.eats.2018.01.011
2. Burkhead WZ Jr, Rockwood CA Jr. Treatment of instability of the shoulder with an exercise program. J Bone Joint Surg Am 1992;74:890-6.
3. 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:e1-5. https://doi.org/10.1016/j.jse.2011.10.033
4. Choi J, Naito K, Curry EJ, Li X. Anterior cruciate ligament reconstruction with achilles tendon allograft in a patient with Ehlers-Danlos

syndrome. Orthop J Sports Med 2018;6:2325967118785170. https://doi.org/10.1177/2325967118785170
5. Dewing CB, Horan MP, Millett PJ. Two-year outcomes of open shoulder anterior capsular reconstruction for instability from severe capsular deficiency. Arthroscopy 2012;28:43-51. https://doi.org/10.1016/j.arthro.2011.07.002
6. Domos P, Lunini E, Walch G. Contraindications and complications of the Latarjet procedure. Shoulder Elbow 2018;10:15-24. https://doi.org/10.1177/1758573217728716
7. Gilbart MK, Gerber C. Comparison of the subjective shoulder value and the Constant score. J Shoulder Elbow Surg 2007;16:717-21. https://doi.org/10.1016/j.jse.2007.02.123
8. Iacono V, Cigala F, Fazioli F, Rosa D, Maffulli N. Reconstruction of chronic patellar tendon tear with allograft in a patient with Ehlers-Danlos syndrome. Knee Surg Sports Traumatol Arthrosc 2010;18:1116-8. https://doi.org/10.1007/s00167-009-1022-7
9. Iannotti JP, Antoniou J, Williams GR, Ramsey ML. Iliotibial band reconstruction for treatment of glenohumeral instability associated with irreparable capsular deficiency. J Shoulder Elbow Surg 2002;11:618-23. https://doi.org/10.1067/mse.2002.126763
10. Johannessen EC, Reiten HS, Lovaas H, Maeland S, Juul-Kristensen B. Shoulder function, pain and health related quality of life in adults with joint hypermobility syndrome/Ehlers-Danlos syndrome-hypermobility type. Disabil Rehabil 2016;38:1382-90. https://doi.org/10.3109/09638288.2015.1102336
11. Johnson SM, Robinson CM. Shoulder instability in patients with joint hyperlaxity. J Bone Joint Surg Am 2010;92:1545-57. https://doi.org/10.2106/JBJS.H.00078
12. Karpyshyn J, Gordey EE, Coady CM, Wong IH. Posterior glenohumeral capsular reconstruction using an acellular dermal allograft. Arthrosc Tech 2018;7:e739-45. https://doi.org/10.1016/j.eats.2018.03.011
13. Legato JM, O'Connell M, Fuller DA. Shoulder arthrodesis. J Orthop Trauma 2018;32(Suppl 1):S4-5. https://doi.org/10.1097/BOT.0000000000001212
14. Levy HP. Hypermobile Ehlers-Danlos syndrome. In: Adam MP, Ardinger HH, Pagon RA, Wallace SE, Bean LJH, Stephens K, et al., editors. Seattle, WA: GeneReviews(R); 1993.
15. Neer CS II, Foster CR. Inferior capsular shift for involuntary inferior and multidirectional instability of the shoulder. A preliminary report. J Bone Joint Surg Am 1980;62:897-908.
16. Rombaut L, De Paepe A, Malfait F, Cools A, Calders P. Joint position sense and vibratory perception sense in patients with Ehlers-Danlos syndrome type III (hypermobility type). Clin Rheumatol 2010;29:289-95. https://doi.org/10.1007/s10067-009-1320-y
17. 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;25:907-12. https://doi.org/10.1016/j.jse.2015.10.010
18. Williams J, Hutt J, Rickman M. Anterior cruciate ligament reconstruction in Ehlers-Danlos syndrome. Case Rep Orthop 2015;2015:160381. https://doi.org/10.1155/2015/160381