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Treatment of Multidirectional Shoulder Instability in Patients with Ehlers-Danlos Syndrome

An estimated fewer than 1 in 10,000 people in the world are affected by hypermobile Ehlers-Danlos syndrome. This is the most common of the thirteen known subtypes[1] of the rare connective tissue disorder, Ehlers-Danlos syndrome ("Ehlers Danlos Syndromes"). Often being misdiagnosed or not diagnosed at all, this disease is one that many medical professionals are unaware of. There is a lack of available literature pertaining to Ehlers-Danlos syndrome, which causes frustrations for patients experiencing a host of unexplainable symptoms, and medical providers alike, who are trying to provide adequate treatment for the multitude of symptoms in a variety of organ systems. Laura Kiesel, a contributor to the Harvard Health Blog, shared her story of being diagnosed with hypermobile Ehlers-Danlos syndrome, called "Ehlers-Danlos syndrome: A mystery solved." It started out with stories of her clumsiness - falling while walking on evenly paved ground, dropping and breaking dishes, and falling off of her bike into a swamp - resulting in a body constantly covered in scabs and bruises. Then, she says:

> I never connected this clumsiness to how my joints and tendons seemed as fragile as the glassware I sometimes shattered: ankles that twisted and sprained at the slightest misstep; wrists wrecked and inflamed for years from the first few attempts at downward dog during an introductory yoga class; a jaw that partially dislocated from the simple act of

> chewing on a tortilla chip. These incidences became less rare and more routine as time wore on, as well as more severe (Kiesel).

More recently Kiesel describes her body as being "besieged by an incessant, widespread, bone-deep aching pain." Physical therapists would ask her to tell them about the terrible accidents she had been through or rough sports she played, when in fact no trauma had occurred at all.

Finally in 2017, Kiesel was incredibly relieved to be diagnosed with hypermobile Ehlers-Danlos syndrome. Her geneticist said that on average it takes anywhere between 10 and 20 years for someone to be diagnosed with the rare disorder. Hypermobile Ehlers-Danlos syndrome is the only Ehlers-Danlos subtype that still has no known specific gene to test for, however it is still hereditary, which can be seen by patterns in other family members. The pain and recurrent injuries related to the disorder would explain the sensations she described, "my sacrum and hips felt as if they had been infused with bits of broken glass that ground against me as I walked and rubbed my soft tissue raw with too much sitting," which all now makes sense because of the knowledge of her abnormal connective tissues in her body. With the wide variety of symptoms from Ehlers-Danlos syndrome, the dots had all been connected with why she had been experiencing migraines, had recurrent skin rashes, issues with regulating body temperature, and even bladder and bowel problems (Kiesel).

Though Ehlers-Danlos syndrome can affect many different systems throughout the body associated with the symptoms Kiesel described, and also psychological symptoms such as anxiety and depression, as well as many other symptoms, this paper will be focusing on the orthopedic aspects of the shoulder joint in particular. Ehlers-Danlos syndrome often leads to multidirectional instability of the shoulder, which presents as dislocations or partial dislocations of the joint in multiple directions. Commonly known as being ‘double-jointed,’ the shoulder is able to come out of its socket and move more than the average person’s joint. Since Ehlers-Danlos syndrome is not widely understood, there are controversies related to affective and necessary treatments. Typically, physical therapy is the first method of treatment. However, because of the connective tissue abnormalities, this is not always sufficient. This is especially true when working with a physical therapist that does not understand the disorder or how to modify exercises to be beneficial for someone that is overly flexible and has loose joints. These characteristics of hypermobile Ehlers-Danlos patients make it difficult to treat, on top of the already challenging multidirectional instability. If physical therapy does not treat the loose shoulder joint, then surgery may need to be implemented. There is not a particular operative technique that is agreed upon to be the ‘best’ for treatment of multidirectional instability in someone with Ehlers-Danlos syndrome. It is a controversial topic because the abnormal connective tissues tend to stretch over time, which is not conducive to long-term success. Nonetheless, surgery still may be attempted if deemed necessary to provide some relief for the patient. Two studies of non-operative treatment methods and three studies of operative treatments will be discussed. For a patient with Ehlers-Danlos syndrome, physical therapy should be implemented first and can be sufficient in providing relief for those with more mild symptoms, but for more severe cases, surgery should be considered.

Joints are defined as any place where two bones meet, even if the bones are not able to move at that spot. They are named after the two bones that come into contact. The glenohumeral joint is where the glenoid cavity of the scapula conjoins with the head of the humerus[2]. A synovial joint, or diarthrosis, is any two bones that are divided with a thin encapsulated space that is filled with fluid, called synovial fluid, serving the purpose of lubricating the joint and allowing a generous range of motion compared to other types of joints. The parts of the bones that face each other are covered with a 2 to 3 mm layer of hyaline cartilage called articular cartilage, with the purpose of allowing joint movement by reducing friction. Between the articular cartilage surfaces of the two bones is the joint cavity, or articular cavity, which is a narrow space filled with synovial fluid. The texture of synovial fluid is viscous and slippery, and it almost eliminates friction during synovial joint movement so that the bones do not rub directly against one another. It is composed of large concentrations of albumin and hyaluronic acid, which work to provide nourishment to the articular cartilages and remove waste between joints. The joint is enclosed to keep the synovial fluid inside with a joint capsule, or articular capsule, that is made of connective tissue. It has a fibrous capsule on the outside that is a continuation of the periosteum from the two bones. On the inside is a synovial membrane with fibroblast-like cells that secrete the fluid and macrophages that remove capsular waste and debris. Articular capsules and ligaments have an abundance of lamellar corpuscles and sensory nerve endings that allow the brain to monitor positioning and movement of the joint.

The shoulder joint is a ball and socket synovial joint, which is one of two multiaxial joints in the body, the other being the hip. These joints allow the bones that they connect to move in multiple planes. The smooth hemispherical humerus head fits into the cuplike socket of the glenoid cavity of the scapula. The glenoid cavity has a fibrocartilage ring called the glenoid labrum around its edges. Some other names for the commonly known shoulder joint are the glenohumeral joint or the humeroscapular joint. The shoulder joint capsule is fairly loose and the glenoid cavity is shallow, which allows for more motion of the joint.

Stability in joints is due to a collaboration of muscles, tendons, and ligaments. Tendons and ligaments are called accessory structures in synovial joints[3]. Tendons serve as the most important joint stabilizers. They are either bands or cords of tough collagenous connective tissue that connect muscle to bone and therefore transfer muscular tension to the bone. Ligaments are also bands or cords of collagenous material, but the difference is in that they attach a bone to another bone for stability. The primary stabilization mechanism of the glenohumeral joint is the biceps brachii muscle[4] found on the anterior side of the upper arm. The biceps brachii tendon from the long head of the muscle progresses through the intertubercular groove in the humerus, and is then attached to the glenoid on its superior margin. Its purpose is to press the head of the humerus against the glenoid cavity. Other stabilizing muscles include the supraspinatus, infraspinatus, teres minor, and subscapularis. The rotator cuff is composed of the tendons of these four muscles that are fused to all sides of the joint capsule except the inferior side. The deltoid muscle is like a cap over the shoulder joint. There are five main ligaments that support the shoulder joint. Three of the five are the glenohumeral ligaments, specifically the inferior, middle, and superior glenohumeral ligaments, that are relatively weak. The coracohumeral ligament attaches at the coracoid process of the scapula and the greater tubercle of the humerus. The fifth is the transverse humeral ligament, which attaches to the greater and lesser tubercles of the humerus. It also makes a tunnel-like area for the tendon from the long head of the biceps brachii muscle to sit. Additionally, there are four bursae, which are fibrous sacs of synovial fluid that help to cushion muscles, allow tendons to slide smoothly over joints, and sometimes modify the direction that a tendon pulls on a muscle. These include the subdeltoid, subcoracoid, subacromial, and subscapular bursae.

Since the glenohumeral joint is a multiaxial ball in socket joint, there are many components of movement associated with it. The range of motion (ROM) is measured by the amount of degrees a joint can move. This is dependent on three factors: articular surface structure of the bones, strength and tautness of ligaments and articular capsules, and actions of the muscles and tendons. The shoulder joint is crossed by the tendons of the biceps brachii, which attach to the scapula, and stabilize the head of the humerus against the glenoid cavity. These structures working together is what prevents shoulder dislocation. There are multiple types of movement exhibited in the glenohumeral joint including flexion, extension, abduction, adduction, medial rotation, lateral rotation, and circumduction. Flexion[5, 6] is defined as the humerus moving anteriorly in the sagittal plane. Extension[5] is the exact opposite of flexion, the humerus moves posteriorly in the same plane. Abduction[6] is the arm moving away from the midline of the body in the coronal plane. Adduction[6] is the opposite, which is the arm moving back toward the midline of the body in the coronal plane. Medial rotation[6] is also known as internal rotation of the humerus, and lateral rotation is external rotation of the humerus. Lastly, circumduction is moving the humerus in a circle going forward, all the way above the head, around, and back down to the midline of the body.

The abnormal anatomy and physiology of Ehlers-Danlos syndrome concerns the connective tissues and collagen in the body. Connective tissues are found almost anywhere in the body including skin, muscles, tendons, ligaments, gums, blood vessels, organs, and more ("What Are the Ehlers-Danlos Syndromes?"). Collagen is one of the body's primary building materials.

It is made of a strong and fibrous protein that has the purpose of strengthening connective tissues as well as giving adequate flexibility where needed. The effects seen in Ehlers-Danlos syndrome patients come from either poor collagen strength or lacking enough collagen that is structurally normal (“Ehlers-Danlos Syndromes”). In the case of multidirectional shoulder instability, the focus will be mainly the tendons and ligaments that hold the joint in place. Ehlers-Danlos syndrome includes a broad range of subtypes that affect varying body parts based on the specific clinical criteria of each type. The overlapping general signs and symptoms of the 13 subtypes are joint hypermobility, which is when joints can move and stretch more than normal, skin hyperextensibility, which is skin that can stretch abnormally, and fragile tissues in the body. All of these are due to distinct connective tissue abnormalities for each different type of Ehlers-Danlos syndrome. Connective tissues are building materials in the body that provide strength and flexibility with strong proteins. Normally, it is able to stretch to a certain extent and then return back to its normal position. However, it is structurally different with Ehlers-Danlos syndrome. This leads to some or all of the connective tissues stretching past the normal limits, which causes damage to the affected area (“What Are the Ehlers-Danlos Syndromes?”).

People with Ehlers-Danlos syndrome commonly experience joint dislocations and subluxations[7]. This is a normal occurrence especially for hypermobility type Ehlers-Danlos syndrome. Shoulder dislocations occur when the humeral head comes out of the glenoid cavity. Structurally, synovial joints are the most complex of all the joints and also the most likely to develop dysfunctions that interfere with a good quality of life. Because the shoulder joint capsule is loose and the glenoid cavity is shallow, the stability of the joint is less than other joints (Saladin). A subluxation is just a partial dislocation of a joint. Both dislocations and subluxations happen when one has multidirectional shoulder instability. Multidirectional shoulder instability[8] is defined as the joint being pushed past its normal limits of motion so that it moves too much in multiple directions. If any of the strong tissues that hold the humerus and glenoid in place are weak, the ball and socket joint will not stay in the correct positioning. The “ball” of the humeral head will be able to move out of the glenoid “socket” due to weakness of the labrum cartilage covering the glenoid rim, the ligamentous capsule that encloses the entire joint and holds the humeral head in the glenoid socket, and the rotator cuff of four muscles and tendons between the humeral head and scapula. Humeral head translation is measured by how much the humerus moves in any specific direction. Causes of multidirectional shoulder instability include having loose joints, repeated overhead motions, many injuries, dislocations, or fracturing part of the glenohumeral joint. Symptoms include feeling as if the shoulder slips out of place, pain or discomfort during motion, shoulder weakness, feelings of grinding, popping, or catching of the joint, and problems using the affected arm (“Understanding Multidirectional Shoulder Instability”). The symptoms experienced by patients with both Ehlers-Danlos syndrome and multidirectional shoulder instability may seriously impact daily life, so treatment options include physical therapy and sometimes surgery if necessary.

Studies of physical therapy specifically for patients with hypermobile Ehlers-Danlos syndrome are extremely sparse. The following is a case report of a non-operative treatment method with the purpose of surveying the effectiveness of a stage-based exercise program in a patient with multidirectional shoulder instability and Ehlers-Danlos syndrome. The motive behind this stemmed from reading the results from a trial done for a stage-based rehabilitation program for multidirectional instability, but there were no patients with Ehlers-Danlos syndrome included in that study. For this case study, the subject is a 14-year-old female Ehlers Danlos Syndrome patient suffering with multidirectional shoulder instability from childhood and recurrent shoulder dislocation. Her bilateral instability caused problems that interfered with her daily living.

The basis of this study is a comparison of a stage-based program with traditional treatment with orthosis, which is the use of braces for support and alignment. Before intervention, her active range of motion including active flexion, passive flexion, and abduction, sulcus sign, and shoulder instability in each direction were measured. A sulcus sign[9] is an orthopedic evaluation of glenohumeral stability done by pulling the arm downwards, and it is positive when there is a visible depression in the shoulder greater than the width of a finger (Inferior Sulcus Test). Since her humeral head dislocated so easily, the passive range of motion and muscle strength could not be measured since the passive range of motion exercise caused dislocation. Also, a modified Rowe score was measured before the interventions to document her shoulder instability. Two interventions, labeled A and B, were implemented in three month intervals[10]. Intervention A was the first three months and was an exercise program with orthosis. The purpose was to reinforce the strength of the muscles that surround the shoulder joint, rather than functional training for the shoulder joint specifically. It included isometric shoulder exercises for rotator muscles, and scapula stabilizers using a TheraBand (Kitagawa et al). Isometric exercises are muscle contractions that have no change in the length of the muscle and an increase in tension, but there is no movement (Saladin). Five specific areas worked were shoulder abduction resistance exercises, internal and external rotation, extension, and flexion. The exercises caused dislocations, so the physical therapist assisted for stability. Intervention B was an implementation of the Watson program for multidirectional instability, which is an exercise program. The purpose of the Watson program to strengthen the rotator cuff and deltoid muscles to benefit scapular muscles. It has six stages to retrain control of the scapula before any exercises for the deltoid or rotator cuff are implemented.

General findings after 6 months indicated an increased active range of motion, no change in passive range of motion due to dislocation, positive sulcus sign, and improved shoulder stability. Furthermore, the patient's pain and discomfort decreased, and she had the ability to control both humeral heads during daily life activities. The results of both interventions indicate through measurements of the active and passive range of motion, sulcus scale, and Rowe score that the scapular motor control program would be effective for EDS hypermobile patients. Of the two, the Watson program for shoulder instability had a favorable impact on the patients with EDS. Another important finding states that reinforcing the strength of the muscles around the shoulder joint ensures the humeral head will remain properly in place when the shoulder moves. Even though passive range of motion was not increased, the study found that dynamic stability rather than static stability aids in the cooperation of shoulder muscles. Ultimately, focusing on both neuromuscular and muscle strengthening exercises would be most helpful to EDS patients with glenohumeral joint instability during shoulder movement (Kitagawa et al).

The second non-operative study exhibited is one of few to explore humeral head translation, both anterior-posterior and superior-inferior in patients with hypermobile Ehlers-Danlos syndrome and Hypermobility Spectrum Disorder. The study aims to determine if patients with these two different hypermobility disorders have a greater tendency to have excessive humeral head translations compared to non-hypermobile patients. Furthermore, it aims to determine if isometric pulley exercises benefit these hypermobile patients, and finally it examines what impact, if any, gravity loads have on humeral head translations. Multidirectional instability in shoulders is often treated with rotator cuff muscle strengthening exercises but this isn't conclusive in patients with hypermobile Ehlers-Danlos syndrome or Hypermobility Spectrum Disorder. The subjects for this study are 13 female patients with hypermobility type of Ehlers-Danlos syndrome and 14 with Hypermobility Spectrum Disorder, with a mean age of 35, all with multidirectional instability, compared to health subjects with a mean age of 34 years. Each subject had a Beighton score of 5.9 or higher. Healthy subjects in the study had a mean Beighton score of 1.5 (Spanhove). The Beighton score[11] is a hypermobility diagnostic test, which examines the knuckle of the pinky fingers, base of thumbs, elbows, knees, and spine. The maximum score is 9, meaning each of the joints meets the criteria for being hypermobile ("Assessing Joint Hypermobility").

With the use of ultrasound[12], shoulder instability, and more specifically how the humeral head moves with the glenoid, can be explored. To achieve results for this, measurements of the acromiohumeral and humeroglenoid distance were studied. The measure of the acromiohumeral distance was determined to be the shortest distance between the inferior acromion edge and the upper humerus edge. Additionally, the measured humeroglenoid distance is from the most posterior point of the humeral head to the posterior glenoid. The patients performed four isometric pulley exercises for this study which included external shoulder rotation, shoulder extension, shoulder flexion, and elbow extension. They were assessed during each exercise to measure the distances of both the acromiohumeral and humeroglenoid.

Findings[13] indicate that patients with mobility issues experienced significant levels of inferior translation while completing isometric external rotations and significant levels of superior translation while completing isometric shoulder extension, flexion, and elbow extension. Compared to the control group, hypermobile patients experienced more superior translation in both shoulder and elbow extension. Furthermore, they experienced more inferior translation than the control group when holding dumbbells, indicating they have a greater degree of humeral head translations than healthy patients. Isometric external rotation did not cause variances between the two groups perhaps because during those exercises, there is an activation of the posterior rotator cuff muscles resulting in greater humeral head translation in both groups. Elbow extensions resulted in the greatest superior translation for hypermobile patients because of the Triceps Brachii's important role in shoulder stabilization. Hypermobile Ehlers-Danlos and Hypermobility Spectrum Disorder patients who also experience multidirectional shoulder instability appear to have Biceps that cannot perform a stabilizing role. This further supports the findings that there is decreased Bicep Brachii activity in patients experiencing multidirectional shoulder instability. Results regarding use of dumbbells indicate that in hypermobile patients, stabilizing muscles cannot compensate for the dumbbell weight resulting in inferior translation which is also impacted by laxity of the capsular ligaments as well. This study could be implemented for hypermobile patients in physical therapy treatments due to results that signify the practicality of the exercises. It would be ideal for patients with more mild symptoms, since stabilizing the surrounding muscles is a preferred first method of treatment compared to surgery, which may need to be implemented once physical therapy studies do not prove successful for treatment (Spanhove).

Physical therapy is the traditional treatment method for patients with multidirectional instability, and is most often successful for treating patients without connective tissue disorders. However, Ehlers-Danlos syndrome adds an additional level of complexity on top of the already challenging multidirectional instability, which is why surgical procedures may need to be implemented for treatment. This study was published in 2003, and is a case report of a 9-year-old girl, who presents with Ehlers-Danlos syndrome and severely disabling multidirectional instability of the shoulders, as well as recurrent dislocations of the hips, and overall joint laxity. The patient history showed two and a half years of recurrent bilateral dislocations beginning at age 6. These dislocations initially stemmed from high energy sports like basketball and swimming, but gradually changed within a few months to affect her ability to do daily activities without experiencing dislocations.

The purpose of this case report is to restore the patient's ability to function comfortably in her everyday life, as well as test the viability of adding a new technique called a thermal capsulorrhaphy to normal treatment plans for children with Ehlers-Danlos syndrome and multidirectional instability. Until the time of this article, the typical treatment of joint laxity from Ehlers-Danlos syndrome was an open surgical capsulorrhaphy. Though this was an effective treatment method, it is invasive and the patients had significant morbidity afterwards. Instead of this, a newer method is thermal-assisted surgery that has shown great potential and a wide variety of applications. This technique is arthroscopic, generally meaning that it is minimally invasive and involves making small incisions in the affected area and inserting a camera called an arthroscope. Arthroscopic techniques are preferred usually over open techniques due to more consistent outcomes, being less invasive, shortening hospital stays, ability to go back to sports and activity more quickly, decreased loss of motion, and less pain. Prior to this procedure, the patient tried a year of nonoperative treatment with stabilizing braces, restricted activity, and physical therapy. This unfortunately was not successful due to the debilitating nature of her joint laxity (Aldridge et al).

Thermal capsulorrhaphy uses probes that emit radiofrequency thermal energy to tighten the ligaments and shoulder capsule. The thermal energy alters the structure of collagen in connective tissues, and decreases the size of the capsule, which reduces the possibilities of future dislocations since the humeral head can no longer move as much (“Thermal Capsulorrhaphy as a Treatment of Joint Instability”). The goal of this thermal capsulorrhaphy case report is to treat multidirectional instability, and also to implement new technology involving thermal energy released by radiofrequency probes or lasers. When examined before the surgery, the patient had no signs of scars or decreased size, clinically referred to as atrophy, of her deltopectoral girdle on either side. She had equal range of motion, both active and passive, of both joints. Her shoulder laxity was so severe that gravity and the weight of her arms alone was causing posterior glenohumeral subluxations[14].

During her arthroscopic procedure with thermal capsulorrhaphy, her gross anatomy appeared normal, including a normal biceps tendon, rotator cuff, superior glenohumeral ligament, inferior glenohumeral ligament, and middle glenohumeral ligament. However, she did have an unusually small labrum and a spacious capsule. The thermal probe was applied to the superior glenohumeral ligament, middle glenohumeral ligament, and anterior and posterior bands of the inferior glenohumeral ligament until enough tissue response was observed. This would appear as shortening of the ligaments and capsule due to the alteration of collagen structure.

Once the response was noticed, it would mean successful reduction of instability and tightening of the shoulder joint. Following the procedure, she was placed in a right shoulder spica cast for two months. Spica casts are used so that the motion of the shoulder joint is completely inhibited after operations. One month after the right cast was removed, the left shoulder was operated on with the same thermal capsulorrhaphy technique and then placed in a spica cast for two months for immobilization. After the casts were both removed, the patient was prescribed physical therapy to strengthen her rotator cuff, deltoid, and periscapular muscles, as well as working on range of motion since her shoulder was tightened.

The outcomes of this procedure were positive and promising for future patients with the same diagnoses. Upon following up with the patient two years later, she reported no subluxations or dislocations at all in the left shoulder and for the right shoulder, one subluxation every month to month and a half. Though experiencing infrequent right subluxations, she said that the symptoms were nothing like the pain she was enduring before the operations. Because of her treatment, she was able to participate in many sports including running track, swimming, and basketball, all of which require frequent movement of the shoulder joints. When discussed among professionals, this new technique shows promising results for the one patient, but they are unsure of how it would do if the technique was used on large amounts of patients. The reason for the patient's instability in one shoulder and not the other after the surgery is unclear. Some possibilities are that the thermal probe could produce different tissue responses in different shoulders, hand dominance of the patient, or follow through with the physical therapy exercises afterwards. Even though it did not work perfectly, she was still pleased with the results and it is common for a certain amount of shoulder laxity to be corrected with time and maturity. It is believed, overall, that thermal capsulorrhaphy should be a technique considered for young patients with Ehlers-Danlos syndrome and unsuccessful conservative therapy for correcting severe shoulder instability (Aldridge et al).

This next study is a case study on the effects of a bilateral glenohumeral stabilization anteriorly and posteriorly using an Achilles tendon allograft. Stabilization surgery is controversial for patients with Ehlers-Danlos syndrome if conservative options are unsuccessful in treating it. The goal of surgical glenohumeral stabilization procedures for Ehlers Danlos syndrome patients is to bring back normal restraints to the joints by minimizing the volume and lax nature of the shoulder capsule. Simple and traditional stabilization methods are usually ineffective. This is especially true for long-term stabilization because of the laxity of connective tissue and foundational structures. Because of this, to reinforce the shoulder joint it may be necessary to do an additional augmentation to add support to the joint. This procedure was done with the goal of replacing the scarred, lax capsule from multiple previous capsular repair failures, with new tissue that is strong and collagenous. The Achilles tendon allograft was used because of its ability to augment the loose shoulder capsule, since the tendon is strongly built and able to withstand more than the native shoulder tendons and ligaments. Previous reports stated success in using Achilles tendon allografts, as well as other robust structures with varied success like the Iliotibial band, tibialis tendon, and fascia lata grafts for shoulder stabilization procedures on patients with prior failed operations. The allograft technique is a good option for young or active patients because it is less invasive than arthrodesis and replacement surgeries, while still repairing the lax ligaments.

The observed patient is a 28-year-old woman with hypermobility type Ehlers-Danlos syndrome and multidirectional shoulder instability. She reported having ten years of recurrent subluxations of the left shoulder, as well as pain during driving, carrying heavy loads, and sometimes experienced pain. Her history includes multiple open surgical procedures for multidirectional instability of her left shoulder, which were all unsuccessful because she still had symptoms that arose a shortly after each procedure. After all the failed attempts, the patient requested that she receive another stabilization procedure to fix her recurrent instability. She was evaluated and shown to have full elevation overhead in the scapular plane with pain in the left shoulder, and while abducted, the arm could rotate 30° internally and 45° externally.

To fix the poor quality of the patient's anterior capsule of the left shoulder, it was restabilized using the Achilles tendon allograft. It was noted that the patient's subscapularis tendon was scarred and the capsule was lax[17, 18]. The procedures performed were a revision Bankart repair[15] and a standard inferior capsular shift[16]. For the Bankart repair, suture anchors were placed along the margin of the anterior glenoid to reattach the damaged labrum to the glenoid. The inferior capsular shift repaired the capsule to the humeral neck. A portion of the capsule is cut like a flap and then reattached so that it was tighter than before. To avoid too much tightening of the capsule, the arm was in 30° of flexion, abduction, and external rotation. This was done with the goal of balancing stability and adequate movement. If tightened too much, the joint would have a limited range of motion. The suture anchors from the Bankart repair are then used to attach the Achilles tendon allograft to the glenoid rim[19, 20]. The goal was to achieve maximum surface area of the allograft, so it was fixed to the glenoid rim vertically, on the humeral side, and the lateral side giving it a broad attachment area. Having a large surface area allows for the superior, middle, and inferior glenohumeral ligaments to be reconstructed. Following the repair of the subscapularis tendon and allograft, the arm was externally rotated at 30° and the anterior tissues showed adequate tension. After the procedure, the arm stayed in a sling for 8 weeks with slight motion at the four week mark. By 12 weeks, strength exercises were started to build up the muscular function in the shoulder. The patient felt like the procedure effectively resolved instability and pain in her left shoulder.

Though this procedure seemed to resolve the problems, 5 years later the patient had posterior subluxation even after strengthening the anterior part of the shoulder capsule. A similar approach was taken to the anterior repair with the Achilles tendon allograft, adding an infraspinatus tendon split to expose the capsule and a glenoid-based capsular shift. The Achilles tendon allograft was fixed on the glenoid's right side as well as the humerus medially and laterally. The rehabilitation process included no adduction across the body and internal rotation for 6 weeks. The next 6 weeks included a slight increase in motion of the shoulder and then more strengthening after total week 12. After this, the patient had stability in the anterior and posterior regions and no pain.

Seven years later, the patient stated she had pain and popping in her right shoulder for three years. She had tried physical therapy to fix this, but it did not work. She was diagnosed with multidirectional instability of her right shoulder. The treatment for this shoulder was simultaneous anterior and posterior capsular reconstructions with Achilles tendon allografts. The anterior was done first to reconstruct the anterior capsule. The allograft was fixed to the anterior glenoid rim and the humeral neck. The suture anchors placed in the humeral neck from the allograft were used also for a repair of the subscapularis tendon. The posterior was then fixed as well. The Achilles tendon allograft was fastened to the posterior glenoid margin and on the humeral side, while carefully adjusting the amount of tension to not be too loose or too tight. The postoperative management was similar to what was advised after the left shoulder operations. Cross-body adduction was to be avoided so that the posterior side of the shoulder would not be stretched too far. External rotation over 30° should be avoided because of the anterior repair. Motion could be restored after 6 weeks. After 3 years, there had not been any instability.

There are varying results for this combination of procedures. For the first procedure, the anterior capsule repair, the reconstruction of the superior, middle, and inferior glenohumeral ligaments were successful. Following the anterior repair, inspection done arthroscopically revealed the tension of the Achilles tendon allograft with the capsule was still adequate five years later[21]. At the time of follow up for the left posterior repair, the patient was able to fully raise her arm above her head, while maintaining shoulder stability anteriorly and posteriorly. The instability was completely resolved, and the patient was satisfied. Following the right shoulder stabilization, the patient exhibited no more instability of the joint after a three year follow up. For patients with recurrent failures of operative treatments for stabilization, it is reasonable to believe this is because of the abnormal collagen. This underlying irregularity from Ehlers-Danlos syndrome can cause deformation and stretching of the soft tissues. There is not any particular ideal surgical stabilization procedure for Ehlers Danlos syndrome patients, since many have positive short-term results, but the longevity of these positive results is not great. There is a high rate of failure long-term with complications. This is because Ehlers Danlos syndrome causes capsule stretching over time, even if fixed surgically. Even though reports from previous studies done with different types of allografts have had varied results, using the Achilles tendon as an allograft is a plausible effective way to restore glenohumeral stability after having previous failed stabilization attempts (Chaudhury et al).

This final study that will be discussed is of a larger group of pediatric subjects, each of whom exhibit a connective tissue disease, including Ehlers-Danlos syndrome. Each patient selected for this study had undergone a previous open inferior capsular shift for atraumatic shoulder instability. All patients also had failed attempts at treatment by physical therapy. The procedure performed for each of the 15 adolescent patients was an open inferior capsular shift. Of the patients, 4 were male and 11 were female, 18 shoulders were operated on in total, with one shoulder done on 12 of the patients and 3 of them had both sides operated on. The age range of the patients at the time of the tested procedure was 14 to 20 years old, with 17 being the mean age. Each of the patients met the clinical criteria of hyperlaxity, measured with a Beighton Score of over 6 points. Also genetic testing for Ehlers-Danlos syndrome was positive in 5 patients, so 36% of the subjects. This does not necessarily mean that the other patients do not also have Ehlers-Danlos syndrome since hypermobile Ehlers-Danlos is not diagnosed genetically. Nine of the patients had previous surgeries on Ehlers-Danlos syndrome related diagnoses, in other joints.

The purpose of this study was to measure the outcome of the open inferior capsular shift technique for adolescent patients exhibiting multidirectional shoulder instability and generalized lax ligaments or Ehlers-Danlos syndrome. The estimate of the pediatric population that presents with hyperlaxity and resultant joint hypermobility is in the range of 4-13%. Though not in association with a definitive connective tissue disease, this still is a large percentage of people with this possibly problematic issue. For young athletes like gymnasts, dancers, swimmers, racquet sports, and throwers this is more prevalent. This is because of the need for flexibility and larger range of motion as compared to other athletes. Problems arise in athletes with hypermobility because of the risk of shoulder dislocation. The result of this is instability in one direction from the structural damage caused by dislocation. More commonly is recurrent subluxations that later cause multidirectional instability of the shoulder. This happens by the lengthening of the typically static parts of the shoulder that are intended to keep a healthy range of motion. A treatment method for these individuals with complicated diagnoses for surgical procedures is needed that proves to be effective for a larger group. Nonoperative methods of physical therapy and modification of physical activity should always be tried first. In young patients, it is important to note that as they age, the hyperlaxity usually decreases. If the physical therapy has not been successful and the patient is still having instability symptoms, then surgical procedures may be implemented. Clinically, the outcomes of the open inferior capsular shift show that there is a reduction of symptoms in pain and instability, which is why it was performed for this study.

The description of an open inferior capsular shift is as follows. The approach was anterior and deltopectoral through a vertical incision in the anterior axillary crease. The subscapularis is removed from the capsule and is cut in the vertical direction. A laterally based T-capsulotomy is then performed. Each patient's humeral head, glenoid bone, and labrum were inspected, and for all the cases, there were no labral tears. Therefore, there was no need to repair the labrum. The inferior capsular flap was moved inferiorly, then the arm is placed in 30^{o} of abduction and external rotation to move the inferior flap superiorly. Then it is attached to the superior portion of the lateral capsule. The superior capsular flap is moved distally and attached to the inferior part of the lateral capsular remnant. Finally, the entire subscapularis was repaired. Following the procedure, each patient used a sling and pendulums for four weeks. After that, the sling was taken off and exercises to work on active range of motion began at four weeks. At six weeks, strengthening exercises were implemented. After four months, the patients were allowed to return to sports.

Upon follow up, the criteria measured for each patient was subjective clinical outcome, objective clinical outcome, and functional outcome scores. The follow up time after the procedure was a mean of 7.5 years, with a minimum follow up of 32 months. The subjective clinical outcome criteria includes pain, shoulder stability, patient satisfaction, and ability to return to activities like sports. Objective clinical outcome is based on the recurrence of dislocations or subluxations and if there were any complications, as well as subscapularis function. Functional outcome scores are based on the 11-item version of the Disabilities of Arm, Shoulder, and Hand (QuickDASH) and the American Shoulder and Elbow Surgeons score (ASES). The QuickDASH is a test measuring upper limb function and the ASES measures the functional shoulder outcome with a minimal clinically significant value of 6.4 points. The subjective outcomes reported by the patients following surgery state that 6 patients felt the pain was much better, 7 felt the pain was better, 1 patient felt no change in pain, and one felt like the pain deteriorated after the minimum follow-up of 32 months. Eleven patients felt their shoulders were much more stable, 2 felt that stability was improved, and 2 felt the shoulder was no more or less stable than before the surgery. Another criteria is the ability to return to sports activity. Of the 15 patients, 9 were able to return to sports. About a third returned with no change in level or ability, one third of them came in at a lower level following surgery. The other third was unable to return to sports. The objective clinical outcomes for recurrence of instability showed that 7 patients had no more instability or joint dislocation, 7 had recurrent subluxations, and 1 actually felt like the shoulder instability was increased[22]. Five patients had MRI scans after the procedure, and all of them had reports of intact subscapularis tendons, showing that the procedure had held up over time. It is stressed that there is a large importance of postoperative rehabilitation. This includes exercises to strengthen the glenohumeral joint, which will help to lower the risk of future instability. The recovery of the subscapularis is very important after shoulder stabilization procedures. If it does not heal properly, the procedural outcome will be poor. Overall, the data from this study shows that the open inferior capsular shift reduced symptoms of pain and instability, reduced recurrences of instability, satisfied the patients, and exhibited good functional outcome scores. The conclusion was that this surgical technique will provide predictable improvement in both subjective and objective functioning of the shoulder, as well as shoulder stability in adolescent patients with these disorders (Vavken).

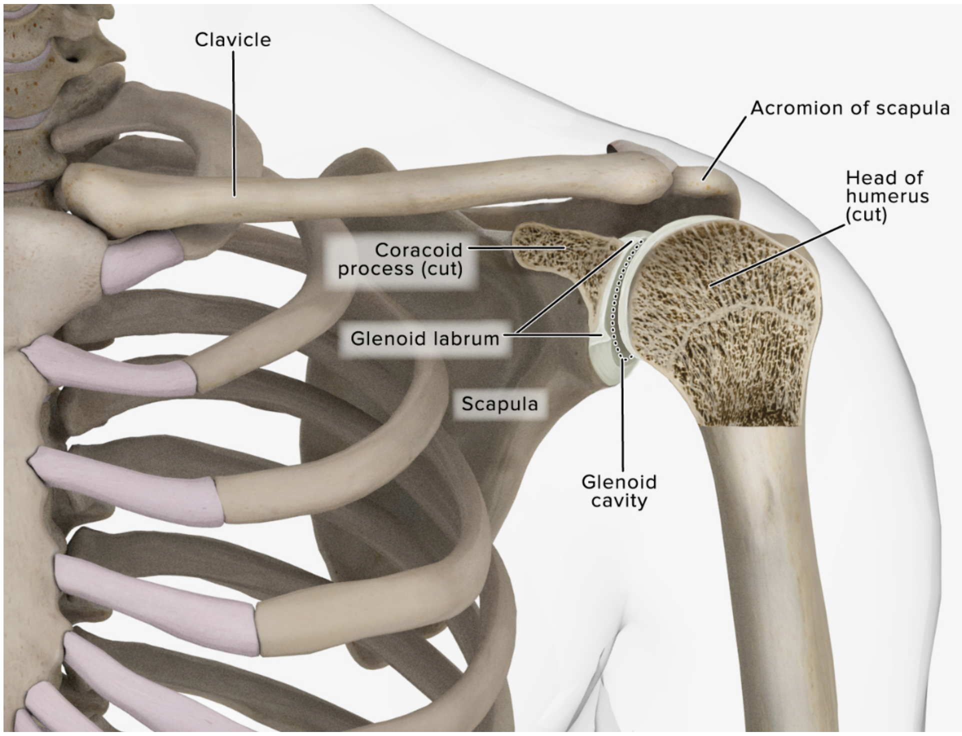
After examining these operative and non-operative studies, it is apparent that more research must be done in order to define a specific treatment with the greatest outcome, especially for long-term results. Though there is no clear standard treatment, it is supported that physical therapy exercises should be sufficient for more mild cases, whereas surgery may be required to treat more severe cases. Surgery is controversial since the long-term results of the procedures in Ehlers-Danlos syndrome patients is poor due to the ability of the abnormal connective tissues to stretch over time. Also, symptoms and severity vary widely from patient to patient, so there may not ever be an answer to what the most ideal treatment would be. Because of the lack of knowledge of the disorder as a whole, the Ehlers-Danlos Society has a free, global patient registry devoted to researching more about it. The registry collects health information for individuals living with Ehlers-Danlos syndrome and facilitates medical research with the recorded data. The Ehlers-Danlos Society is working to find links between Ehlers-Danlos syndrome and Hypermobility Spectrum Disorder with chronic pain, anxiety, neurological, gastrointestinal, mast cell, and autonomic disorders. It also is exploring genetic variants to see which commonalities exist in hypermobile Ehlers-Danlos syndrome patients, since this is currently the only subtype without a genetic diagnostic test. Ehlers-Danlos syndrome is gaining more awareness today, which hopefully will drive more research to be done worldwide in the near future for a better understanding of the rare disorder.

Images

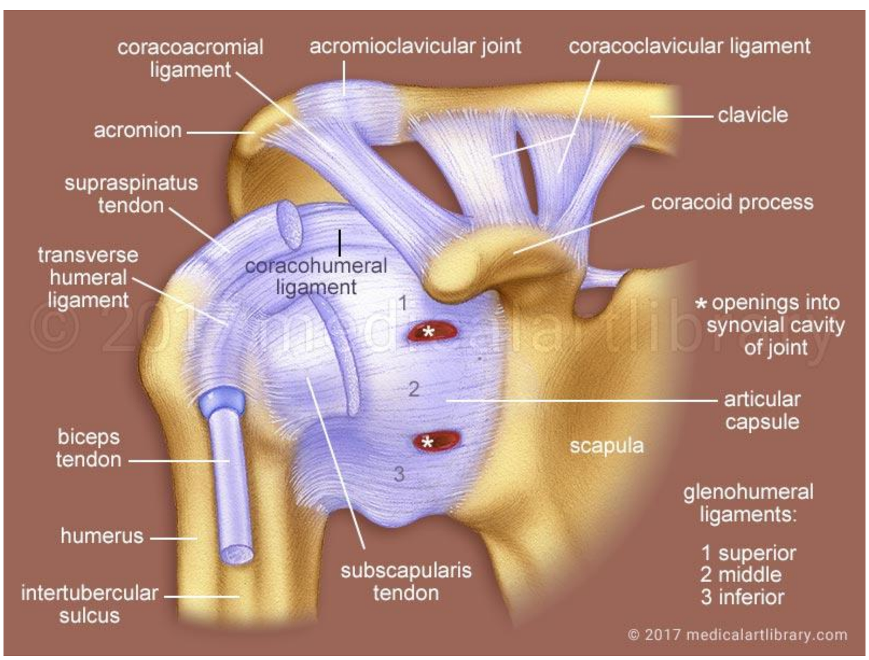
1. Ehlers-Danlos syndrome subtypes (“The Types of EDS”)

Name of EDS Subtype	***IP****	***Genetic Basis***	***Protein Involved***
Classical EDS (cEDS)	AD	Major: *COL5A1, COL5A2*	Type V collagen
		Rare: *COL1A1* c.934C>T, p.(Arg312Cys)	Type I collagen
Classical-like EDS (clEDS)	AR	*TNXB*	Tenascin XB
Cardiac-valvular EDS (cvEDS)	AR	*COL1A2* (biallelic mutations that lead to *COL1A2* NMD and absence of pro α2(I) collagen chains)	Type I collagen
Vascular EDS (vEDS)	AD	Major: *COL3A1*	Type III collagen
		Rare: *COL1A1* c.934C>T, p.(Arg312Cys) c.1720C>T, p.(Arg574Cys) c.3227C>T, p.(Arg1093Cys)	Type I collagen
Hypermobile EDS (hEDS)	AD	Unknown	Unknown
Arthrochalasia EDS (aEDS)	AD	*COL1A1, COL1A2*	Type I collagen
Dermatosparaxis EDS (dEDS)	AR	*ADAMTS2*	ADAMTS-2
Kyphoscoliotic EDS (kEDS)	AR	*PLOD1*	LH1
		FKBP14	FKBP22
Brittle cornea syndrome (BCS)	AR	*ZNF469*	ZNF469
		PRDM5	PRDM5
Spondylodysplastic EDS (spEDS)	AR	*B4GALT7*	ß4GalT7
		B3GALT6	ß3GalT6
		SLC39A13	ZIP13
Musculocontractural EDS (mcEDS)	AR	*CHST14*	D4ST1
		DSE	DSE
Myopathic EDS (mEDS)	AD or AR	*COL12A1*	Type XII collagen
Periodontal EDS (pEDS)	AD	*C1R*	C1r

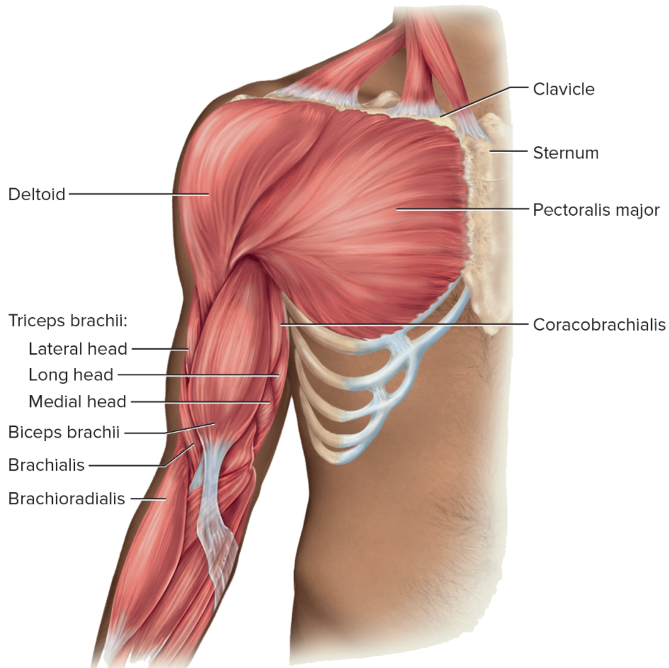
2. Glenohumeral joint - anterior (Saladin)



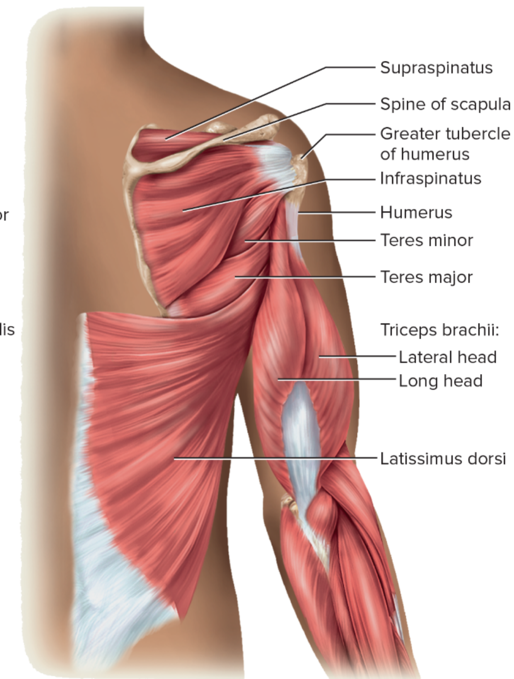
3. Ligaments and tendons of the glenohumeral joint ("Shoulder Joint Ligaments")



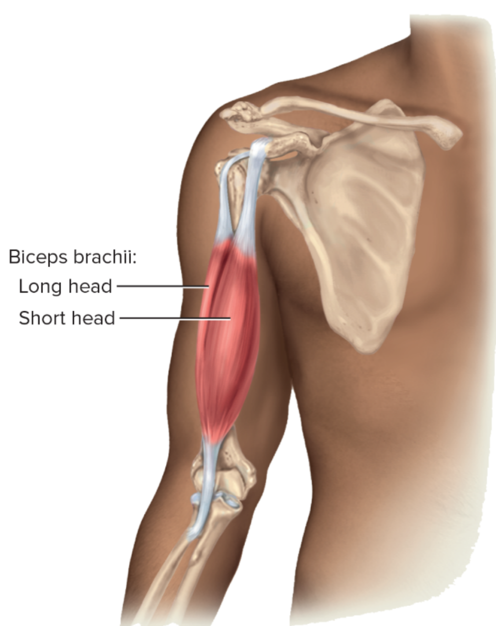
4. Muscular anatomy of the shoulder (Saladin)



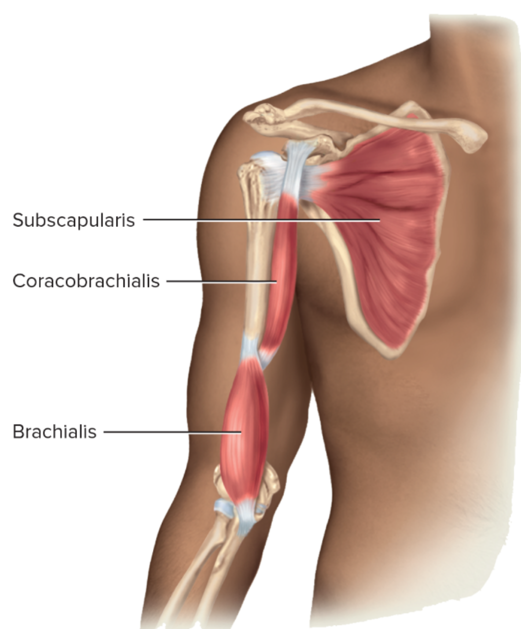
(a) Anterior view



(b) Posterior view

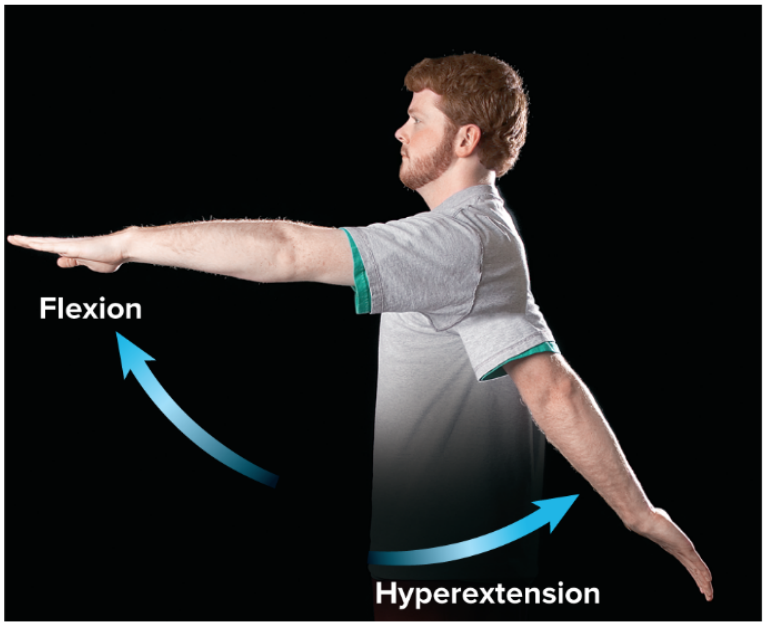


(c) Anterior view

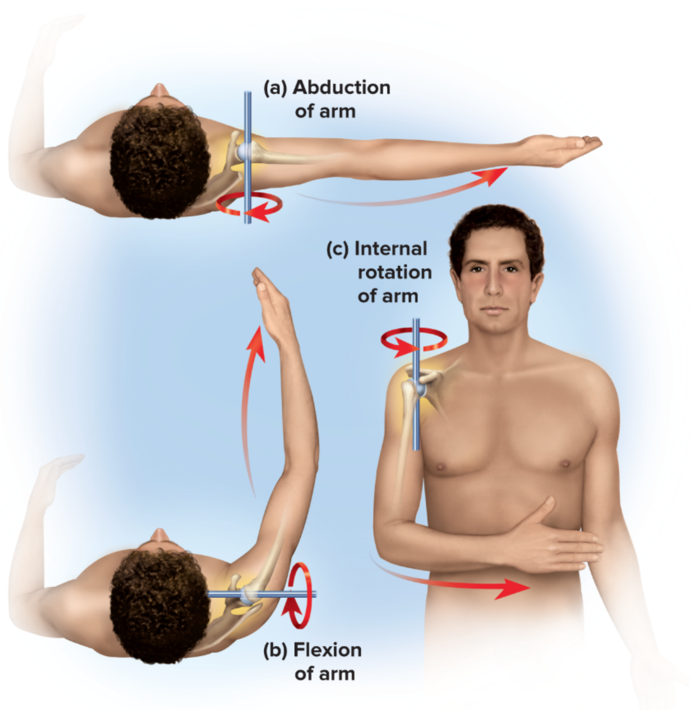


(d) Anterior view

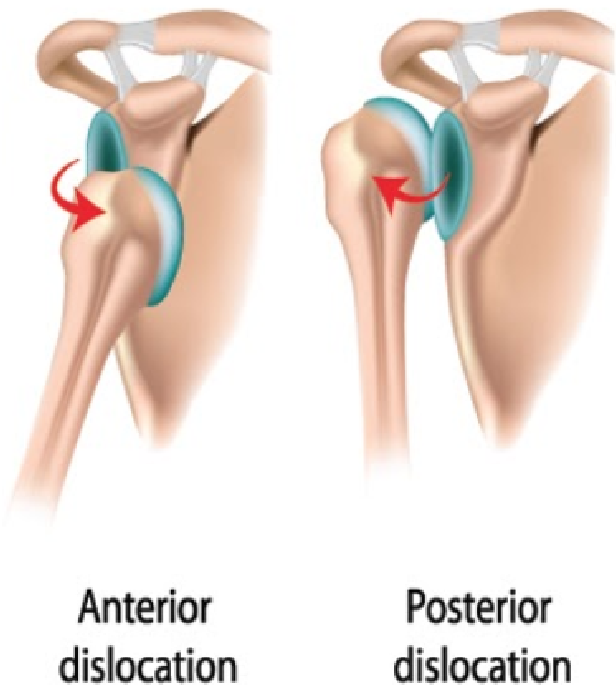
5. Flexion and extension (Saladin)



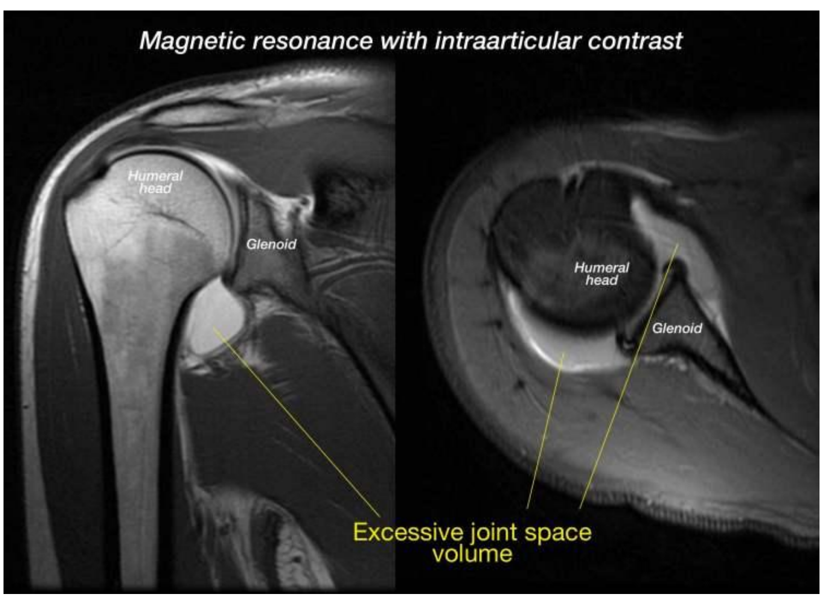
6. Abduction, flexion, internal rotation; adduction is opposite of abduction, extension is opposite of flexion, external rotation is opposite of internal rotation (Saladin)



7. Anterior and posterior shoulder dislocations



8. MRI showing excessive space in the joint with multidirectional instability (*Magnetic resonance with intraarticular contrast*)



9. Sulcus sign is pointed to in patient's shoulder (*Sulcus Sign)*

10. (Kitagawa et al.)

Table 1: Range of motion of shoulder joint on each time point.

	Baseline	After intervention A (3-month follow-up exam)	After intervention B (6-month follow-up exam)	Final evaluation (12-month follow-up exam)
Active flexion	30/35	45/45	155/150	160/160
Abduction	45/45	45/45	115/110	125/120
Passive flexion	25/30	35/30	35/30	35/35
Abduction	30/35	40/40	45/45	40/45

Table 2: The Rowe scores on each time point.

	Baseline	After intervention A (3-month follow-up exam)	After intervention B (6-month follow-up exam)	Final evaluation (12-month follow-up exam)
Stability score	0	0	30	10
Motion score	0	0	15	15
Function score	0	10	25	25
Total score	0	10	70	50

11. Beighton score (Beighton and Horan)

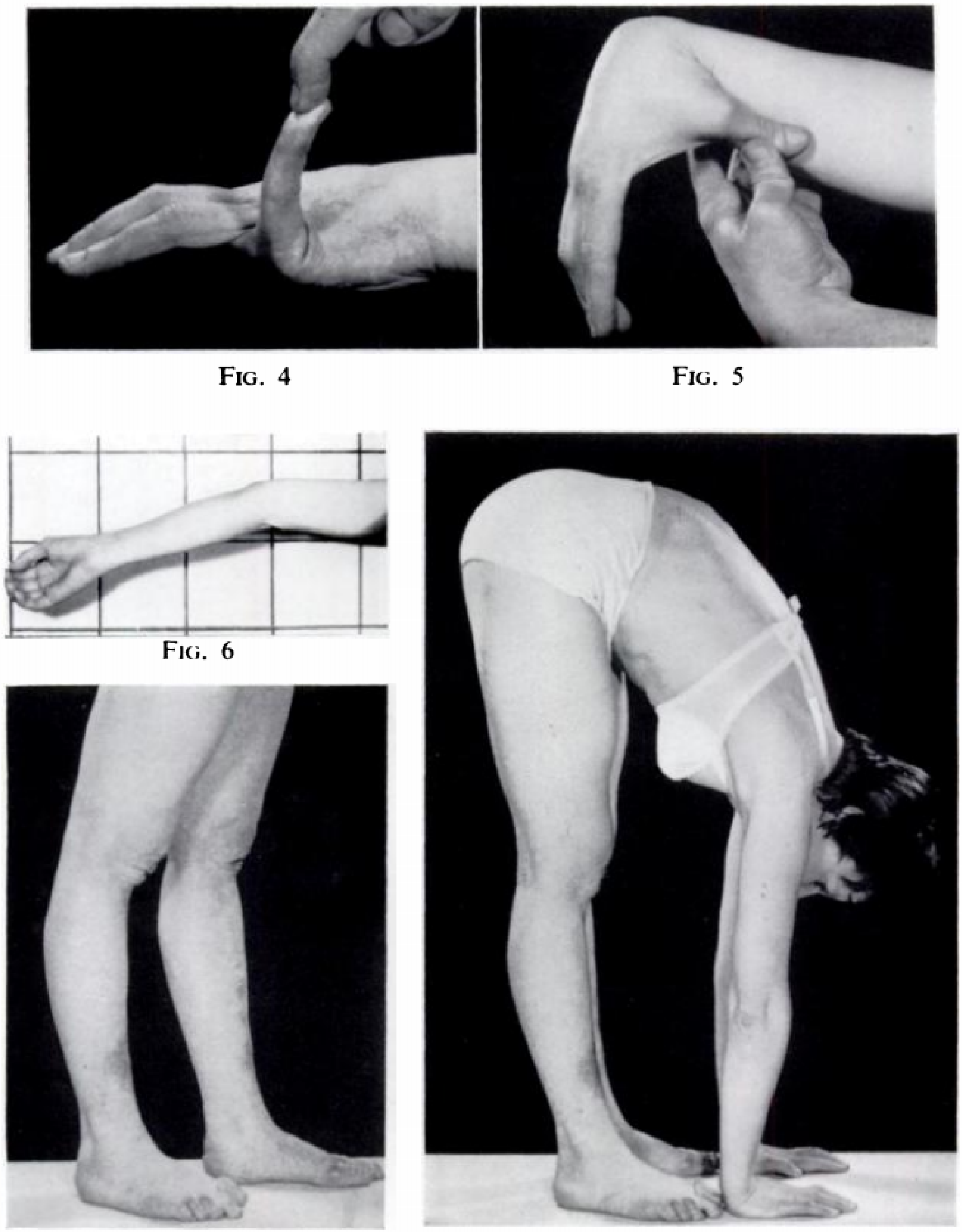


FIG. 4

FIG. 5

FIG. 6

FIG. 7

FIG. 8

Criteria for assessing hypermobility. Figure 4—Hyperextension of the little finger beyond 90 degrees. Figure 5—Passive apposition of the thumb to the flexor aspect of the forearm. Figure 6—Hyperextension of the elbow beyond 10 degrees. Figure 7—Hyperextension of the knee beyond 10 degrees. Figure 8—Forward flexion of the trunk so that the palms of the hands rest easily upon the floor.

12. (Spanhove et al.)

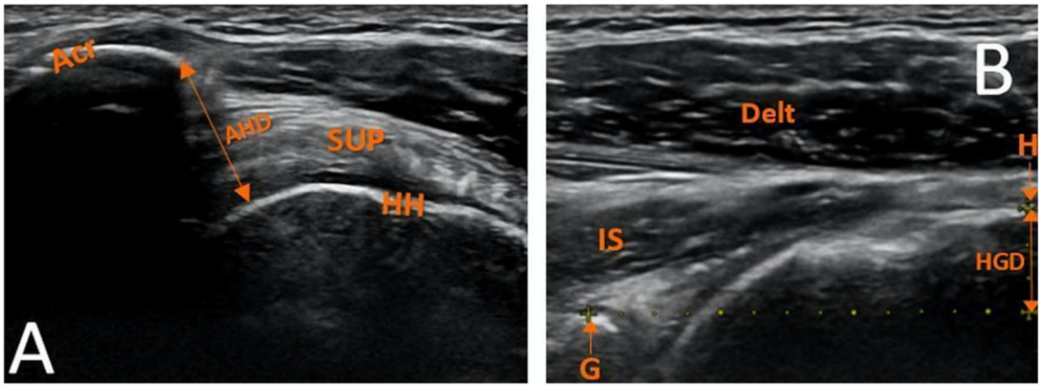


Fig. 1. Ultrasound measurements (A) of the AHD (B) of the HGD. AHD, Acromiohumeral distance; HGD, Humeral Glenoid Distance; SUP, Supraspinatus; IS, Infraspinatus; Delt, Deltoid; HH, Humeral Head; Acr, Acromion surface; G, posterior aspect of glenoid; H, most posterior edge of the humeral head.

13. Results: Table 2 and Table 3 (Spanhove et al.)

Table 2
Mean AHD and HGD values in patients and healthy controls. Rest and isometric values are expressed as Mean (SD). Δ values are expressed as mean difference ± (standard error): positive Δ values represent an increase in AHD (inferior translation)/HGD (posterior translation), negative Δ values represent a decrease in AHD (superior translation)/HGD (anterior translation).

	AHD (mm)		HGD (mm)	
	Patients	Controls	Patients	Controls
Shoulder ER				
• Rest	10.6 (1.6)	10.7 (1.5)	12.1 (4.4)	11.7 (3.4)
• Isometric	11.2 (1.7)	11.3 (1.8)	12.5 (4.9)	11.9 (3.5)
• Δisometric-rest	0.6 (0.2)*	0.6 (0.3)*	0.3 (0.3)	0.2 (0.3)
Shoulder Ext				
• Rest	11.3 (1.7)	10.9 (1.7)	10.5 (3.8)	10.7 (3.0)
• Isometric	10.6 (1.6)	11.0 (2.1)	10.5 (4.0)	10.5 (3.5)
• Δisometric-rest	−0.7 (0.2)**	0.0 (0.2)	−0.3 (0.3)	−0.3 (0.3)
Shoulder Flex				
• Rest	10.7 (1.7)	11.1 (1.7)	10.8 (3.6)	10.4 (2.4)
• Isometric	9.9 (1.5)	10.5 (1.7)	10.4 (3.8)	10.2 (2.6)
• Δisometric-rest	−0.8 (0.1)*	−0.6 (0.2)*	−0.6 (0.2)*	−0.3 (0.2)
Elbow Ext				
• Rest	10.9 (1.6)	10.9 (1.6)	10.2 (3.5)	10.8 (3.2)
• Isometric	9.3 (1.5)	10.5 (1.6)	10.5 (3.9)	11.2 (3.3)
• Δisometric-rest	−1.5 (0.2)**	−0.4 (0.3)	0.4 (0.2)	0.4 (0.3)

* Significant difference ($P < 0.05$) between isometric contraction and rest within the same group.
** Significant difference ($P < 0.05$) compared to the control group.

Table 3
AHD values in patients and healthy controls before and during dumbbell loading.

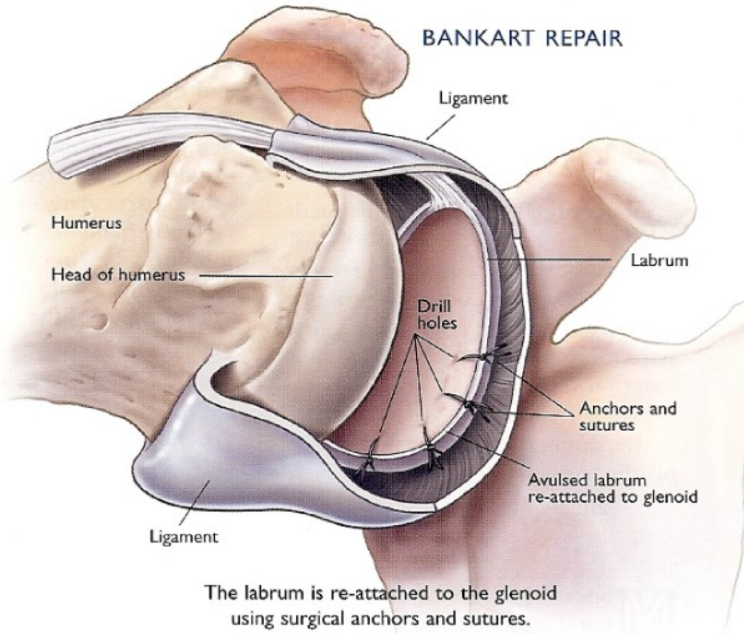
	AHD (mm)	
	Patients	Controls
Without dumbbell [a]	11.5 (1.7)	11.2 (1.7)
With dumbbell [a]	13.2 (2.8)	11.4 (1.7)
Δ with-without [b]	1.7 (0.3)**	0.3 (0.4)

[a] Values are expressed as Mean (SD).
[b] Δ values are expressed as mean difference (standard error): positive Δ values represent an increase in AHD.
** Significant difference compared to controls.

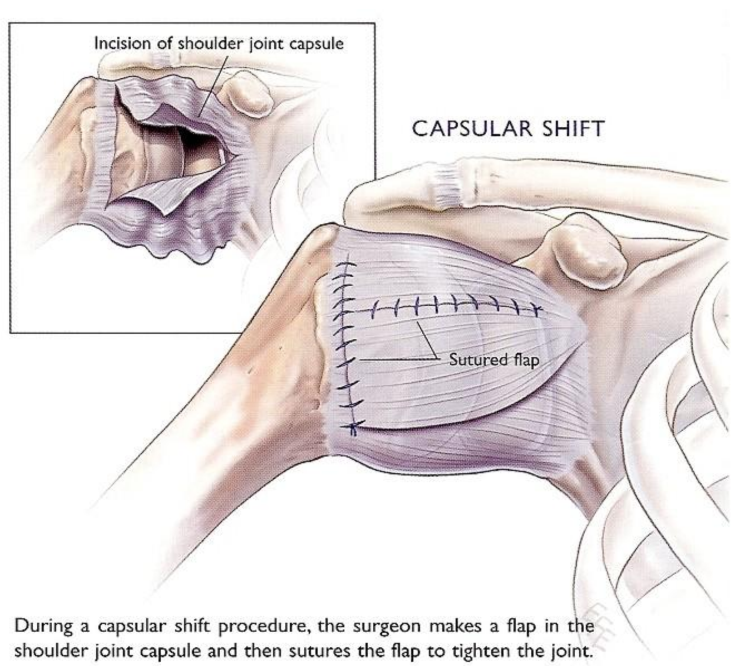
14. (Aldridge et al.)

FIGURE 1. MRI with the patient lying supine shows posterior glenohumeral subluxation.

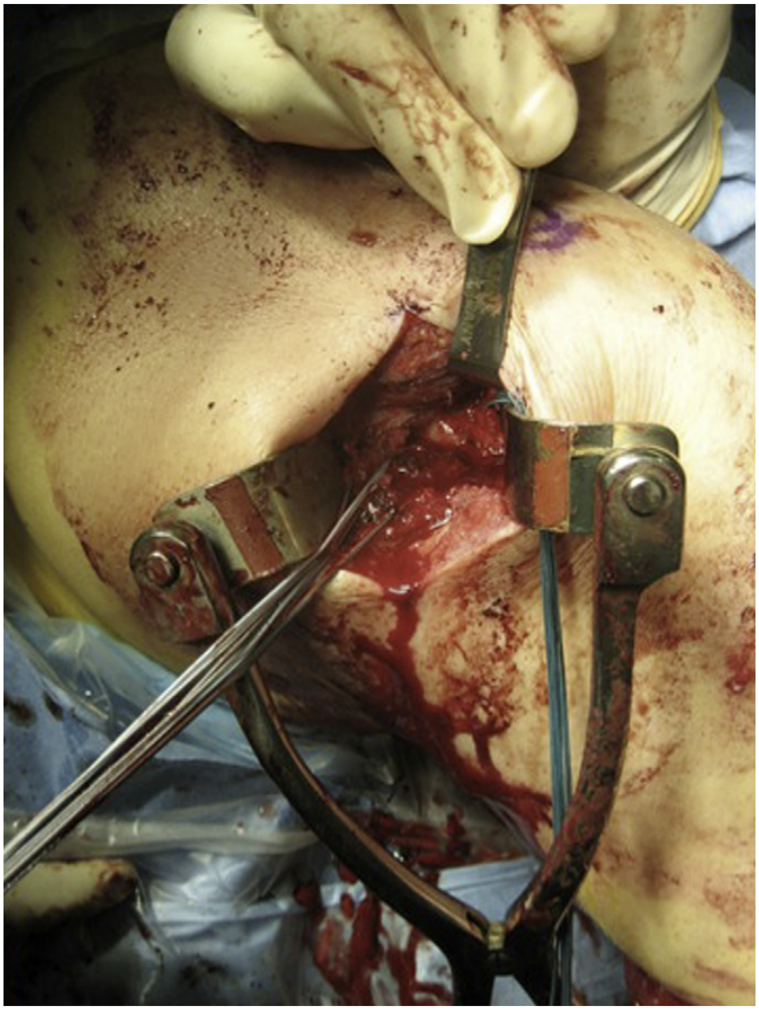
15. Bankart Repair (*Bankart Repair*)



16. Inferior Capsular Shift (Smith)

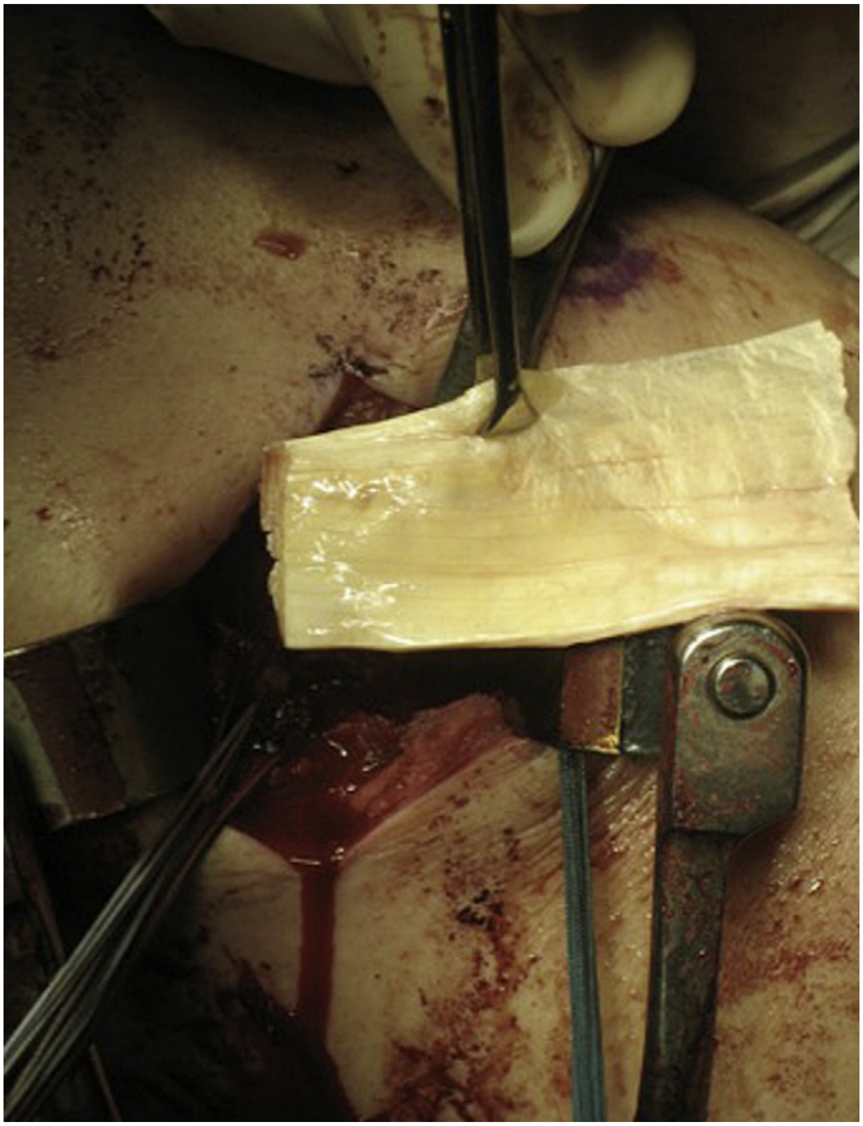


17. Standard deltopectoral approach, left shoulder subscapularis being divided (Chaudhury et al.)

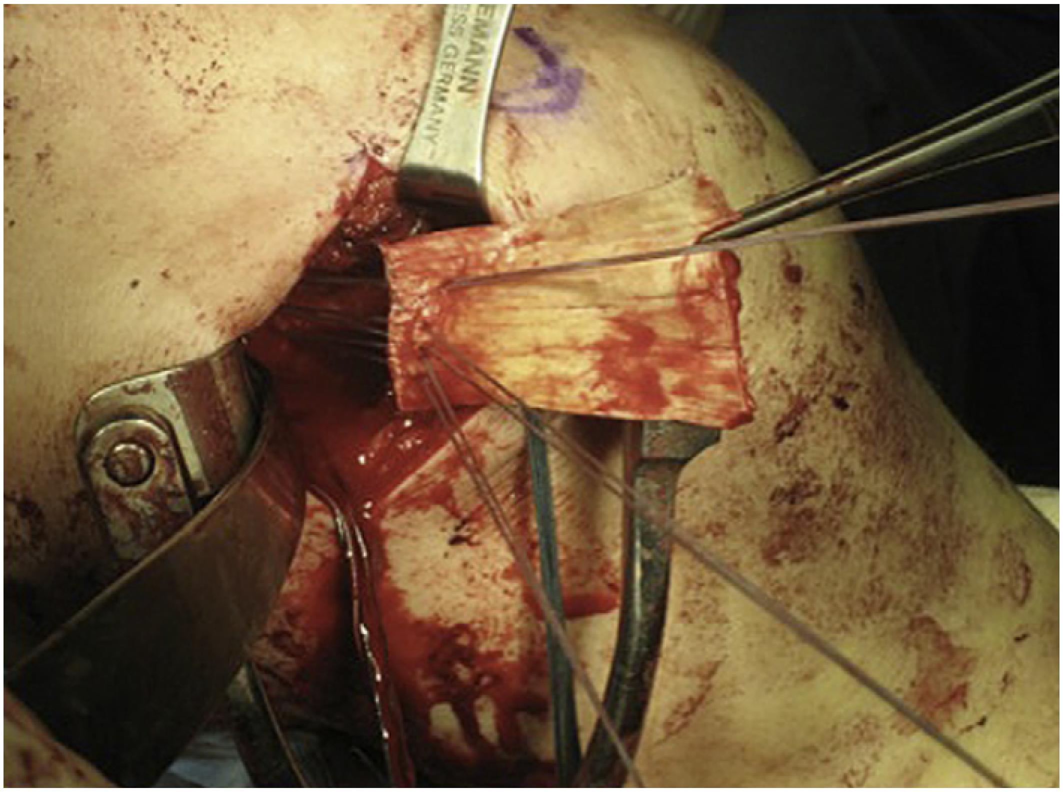


18. Capsule is divided to enter the joint (Chaudhury et al.)

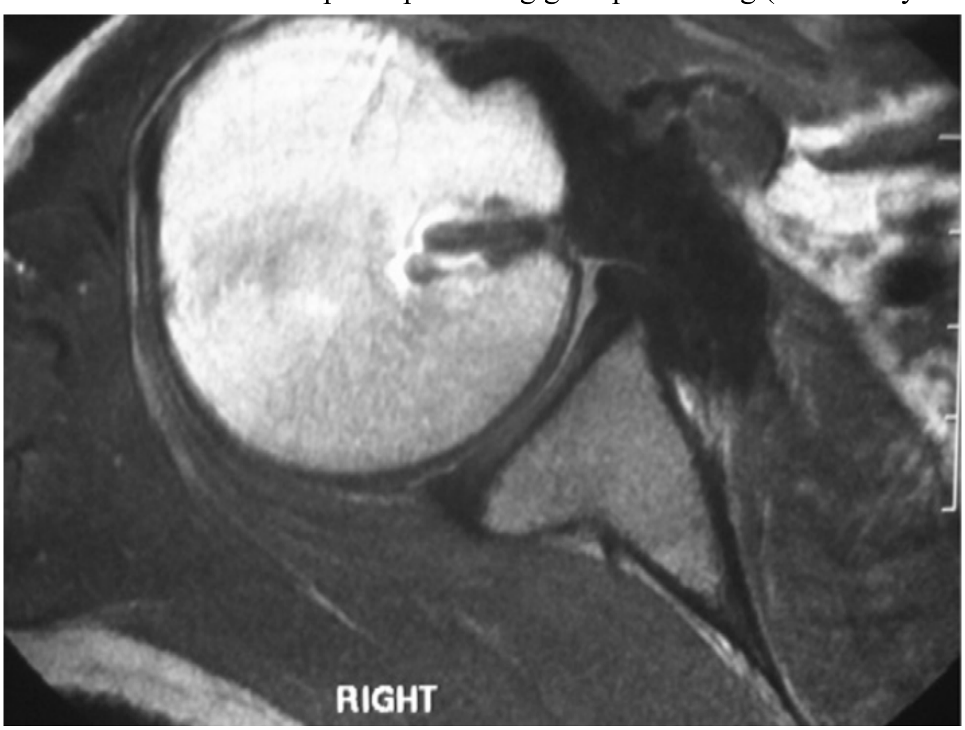
19. Achilles tendon allograft sized appropriately to imitate the anterior shoulder capsule (Chaudhury et al.)



20. Anterior capsule reconstruction of left shoulder, Achilles graft is placed with suture anchor guides from Bankart repair (Chaudhury et al.)



21. MRI taken at time of post-op showing graft positioning (Chaudhury et al.)



22. Statistical results (Vavken et al.)

Table II *P* values from the regression analysis of covariates

Covariates	Recalled number of dislocations before surgery	Genetic diagnosis of Ehlers-Danlos syndrome	Laterality of surgery (dominant, nondominant, bilateral)	Rotator interval closure performed
	P value	*P* value	*P* value	*P* value
Pain after surgery	.076	.278	.833	.278
Stability after surgery	.149	.205	.586	.205
Satisfaction with surgery	.200	.635	.698	.635
Return to sports	.483	.154	.092	.154
Recurrent dislocations	.682	.344	.902	.344
ASES score	.606	.027*	.174	.027*
QuickDASH score	.875	.044*	.133	.044*

ASES, American Shoulder and Elbow Surgeons; *QuickDASH*, 11-item version of the Disabilities of Arm, Shoulder and Hand.
* Statistically significant.

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