Minimally Invasive Surgical Technique for Ankle and Rearfoot Stabilization in Patients with Hypermobile Ehlers-Danlos Syndrome

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Statement of Purpose

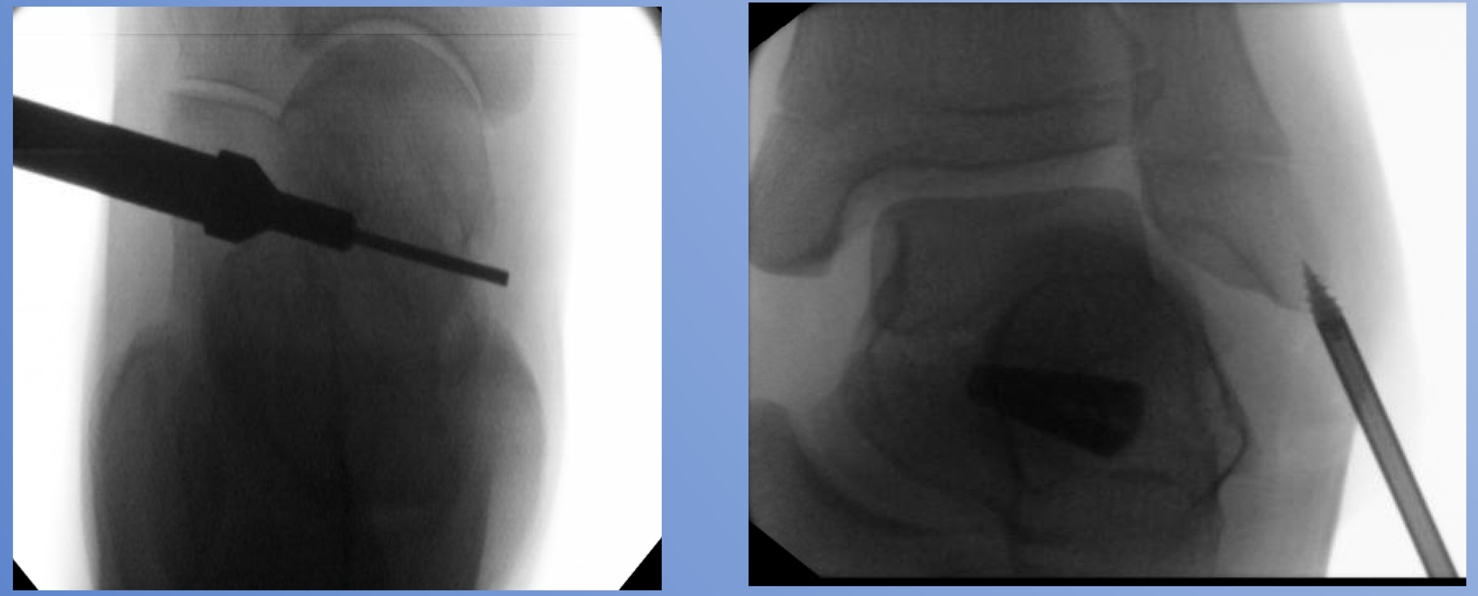
The purpose of this study is to present our observation that patients with hypermobile Ehlers-Danlos syndrome (hEDS) benefit from a minimally invasive procedure supplementing the lateral ankle ligaments while limiting hyperpronation.

Methods and Hypothesis

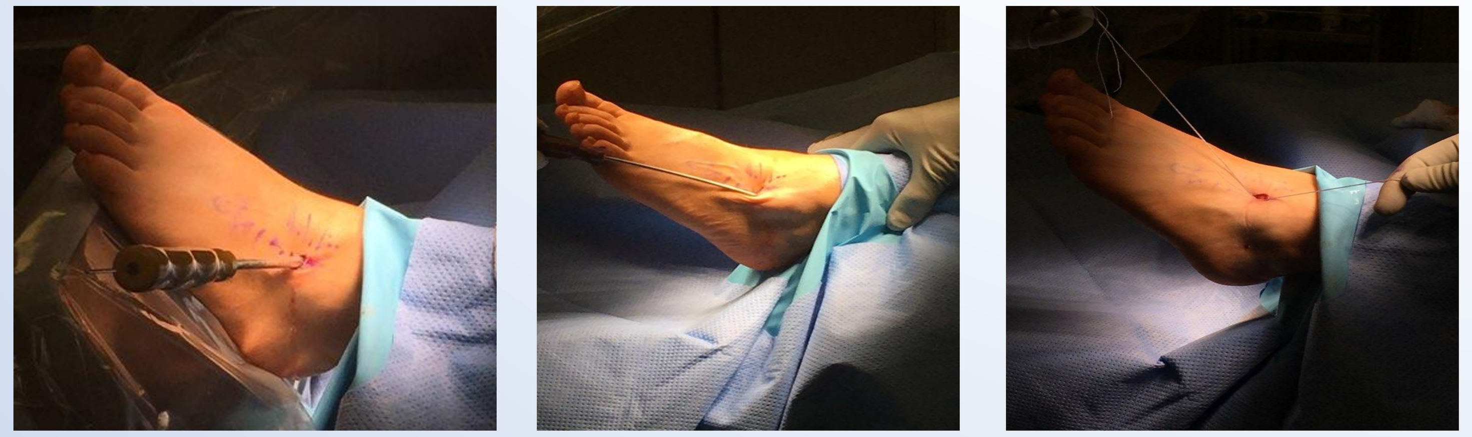
A retrospective review was conducted on 15 consecutive patients previously diagnosed with hEDS who presented with painful lateral ankle instability and pes planus. Patients were included after failing conservative treatment options such as bracing, orthotics, and physical therapy. All patients in this study underwent minimally invasive surgery, including subtalar joint arthroereisis and lateral ankle ligament supplementation. Surveys based on the American College of Foot and Ankle Surgeons (ACFAS) Universal Evaluation Scoring Scale were collected greater than six months after surgery to assess pain level, functional capabilities, and ankle sprain frequency. We hypothesized that patients would experience improvement in pain, less frequent ankle sprains, and less limitation in activity after surgery.

Procedure

The surgical technique consisted of a curvilinear skin incision less than one centimeter in length made over the lateral aspect of the sinus tarsi and a stab incision made over the lateral calcaneus near the insertion of the native calcaneal fibular ligament (CFL). Blunt dissection was carried down to the level of bone through each incision. A guide wire was placed within the sinus tarsi and the appropriately sized sinus tarsi implant was inserted over the wire (Figure 1a and 2a). Next, two titanium bone anchors were manually inserted through the incision over the sinus tarsi into the distal fibula (Figures 1b and 2b). Another titanium bone anchor was introduced through the same incision and inserted into the talus at the junction of the talar head and neck. A final titanium bone anchor was inserted through the stab incision over the CFL and into the lateral calcaneus. All bone anchors came with two strands of attached non-absorbable high-strength fiber suture. The suture from the anchor to the lateral calcaneus was passed superficial to the peroneal tendons to the incision over the sinus tarsi. Finally, the sutures from one anchor to the fibula and those from the anchor to the talus were securely tied together and the sutures from the other fibula anchor and those from the calcaneal anchor were tied together while holding the foot in slight eversion (Figure 2c). The incisions were closed using a combination of absorbable and non-absorbable sutures as needed.



Figures 1a and 1b. (a) Insertion of the sinus tarsi implant under fluoroscopy. (b) Insertion of fibular bone anchor under fluoroscopy.



Figures 2a , 2b, and 2c. (a) Insertion of the sinus tarsi implant through the small incision over the sinus tarsi. (b) Insertion of fibular bone anchor through the incision site over the sinus tarsi. (c) Suture from talar bone anchor and suture from fibula bone anchor being tied together.

Literature Review

Ehlers-Danlos syndrome (EDS) is a heterogeneous group of connective tissue disorders characterized by abnormal collagen synthesis resulting in joint hypermobility, skin hyperextensibility, and tissue fragility (1-6). The 2017 International Classification for the Ehlers-Danlos Syndromes recognizes thirteen subtypes of the disorder (5). The most common variant of EDS, hypermobile Ehlers-Danlos syndrome (hEDS), was previously known as EDS type III according to the Berlin nosology and EDS hypermobility type in the Villefranche classification (7). In the past, joint hypermobility syndrome (JHS) has been classified as a separate hereditary connective tissue disorder characterized by musculoskeletal pain and joint hypermobility. However, since JHS and hEDS have been deemed indistinguishable, the two disorders are now classified together under the subtype of hEDS (8). Unfortunately, hEDS has no known genetic etiology and therefore diagnosis of the disorder is made clinically using tools such as the Beighton scale to access hypermobility (7-10).

Hypermobile EDS is associated with the most debilitating musculoskeletal manifestations including pain, joint instability, and soft tissue overuse injury (11, 12). Recurrent dislocations are common and may occur spontaneously or with minimal trauma, often leading to early degenerative joint disease and functional impairment (6, 13, 14). A significant portion of musculoskeletal complaints involves the lower extremity, with 77.8% prevalence of foot and ankle pain reported (1). Foot appearance and functionality are severely altered by the pathology of hEDS, with severe pes planus revealed to be the most common foot deformity (1, 4, 15). Ankle sprains and dislocations are common in hEDS patients, leading to lateral ankle instability and frequent falls, with up to 95% of patients reporting falling within the past year (1, 16).

Treatment of the most common podiatric pathologies of pes planus and lateral ankle instability presents a challenge in patients with hEDS. Typical conservative treatment of these pathologies, including ankle braces and orthotics, are of limited use in hEDS due to the severe nature of the deformity and hypermobility (12, 14, 17). Physical therapy should be employed to improve instability and pain, however these efforts are limited as well (12). When conservative measures are unsuccessful, surgical treatment such as a modified Brostrum procedure for lateral ankle instability or tendon transfer and osteotomies for pes planus are usually employed in the general population (18, 19). Unfortunately, these procedures often depend on the stability of the native ligaments and tendons and are unreliable in patients with hEDS given the reduced mechanical properties of the tissue (3). Several studies have demonstrated that the presence of generalized joint hypermobility was significantly associated with clinical failure and recurrence of ankle instability after undergoing a modified Brostrum procedure (20-22). In addition, due to soft tissue fragility, common surgical complications in hEDS include bleeding, hematoma formation, abnormal scarring, delayed healing, and infection (15, 23). In fact, some experts have stated that surgery should be contraindicated in the EDS patient due to unpredictable healing implications (24). Given these findings, the need exists for a minimally invasive technique for correction of deformity and instability in patients with hEDS.

Results

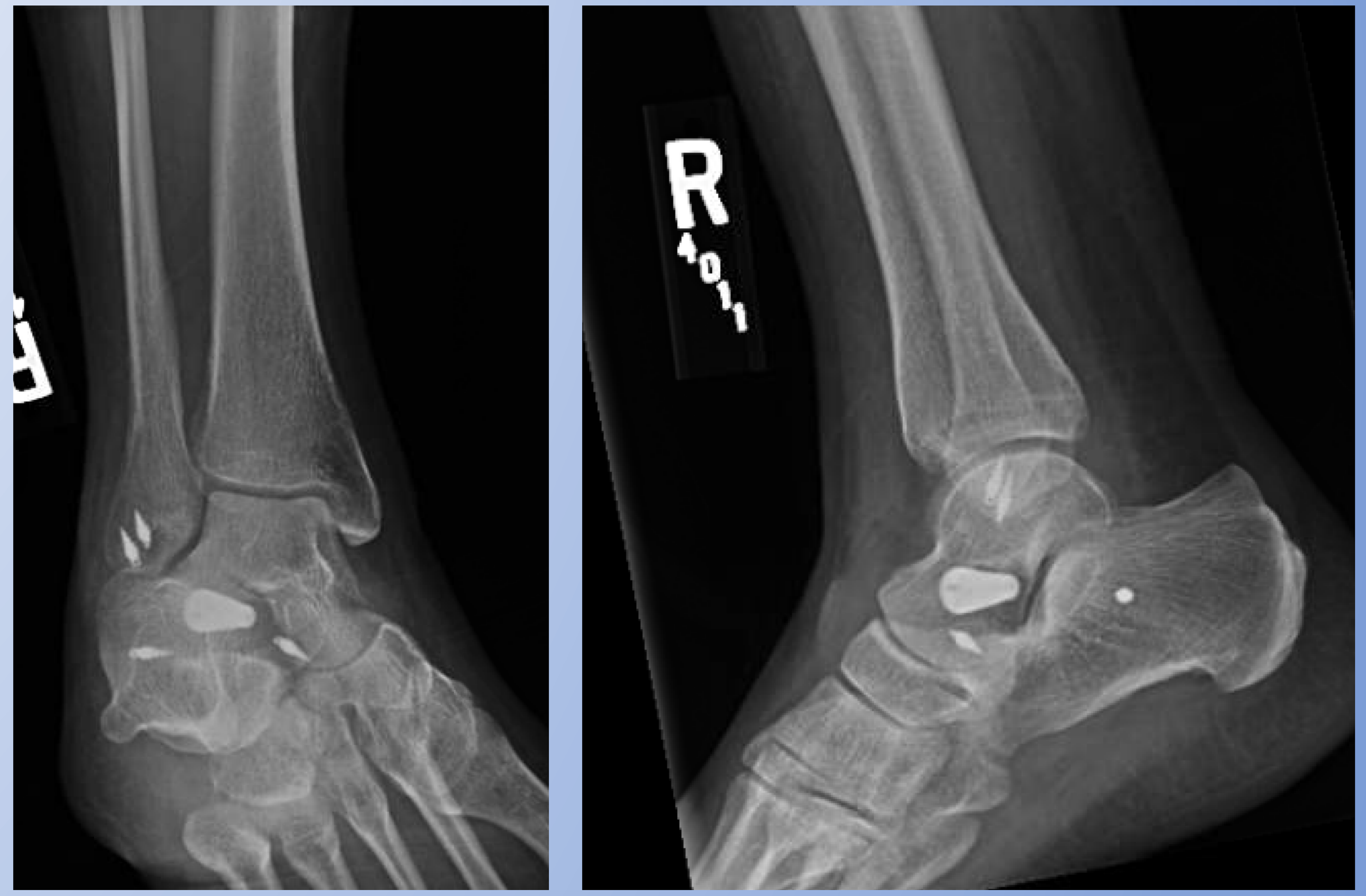
At greater than six months after surgery, 73% of patients with hEDS showed improvement in pain and function, with 53% reporting no limitation in activity. Specifically, 73% of patients reported less difficulty in ability to function on one flight of stairs, incline, and uneven terrain after completion of the surgery. Furthermore, 60% of patients were able to return to wearing normal shoes and only one patient still required custom-made shoes after surgery. Eighty percent of patients no longer used assistive devices after surgery. All 15 patients reported decreased frequency of ankle sprains since surgery with 80% reporting no sprains post-operatively.

Analysis and Discussion

Given the soft tissue fragility and wound healing complications found in patients with hEDS, a minimally invasive surgical technique addressing the hypermobility while not relying on the native ligaments seems most sensible (23). To our knowledge, a combination of STJ arthroereisis and lateral ankle ligament supplementation using a minimally invasive technique has not been previously used in the treatment of foot and ankle instability in hEDS. A single case study did report using STJ arthroereisis in an EDS patient to provide stability, however the lateral ankle ligaments were not addressed (24). In our study, 73% of patients showed improvement in pain and function which is higher than the satisfaction rates of 59-67% after orthopedic surgeries reported in the literature (23). In a study by Huang et al., 28% of patients with generalized joint hypermobility reported continued ankle sprains following a modified Brostrum procedure which is greater than the 20% of patients with hEDS reporting ankle sprains after our minimally invasive technique (25). Overall, satisfaction after undergoing orthopedic surgery in patients with EDS is much lower than that of the general population (12, 14).

Since direct repair of the ligaments is not recommended in the setting of generalized joint hypermobility, lateral ankle ligament reconstruction using allograft tendon has been suggested. However, compared to the modified Brostrum procedure, reconstruction using an allograft tendon is quite invasive requiring extensive soft tissue dissection which would almost certainly result in wound healing complications in hEDS patients (25). Also, a more extensive procedure would require a long duration of tourniquet use which can lead to vessel rupture in patients with hEDS. Experts recommend leaving sutures in place twice as long as normal in hEDS patients, however wound-healing complications are still common and therefore it is best to avoid extensive incisions (12).

In conclusion, hEDS is a debilitating disorder often involving the lower extremities, but our promising results indicate that a minimally invasive procedure addressing pes planus and lateral ankle instability improves pain and functionality while decreasing frequency of ankle sprains. A few limitations exist in our study, including a small patient population, the retrospective nature of the study, and that outcomes were subjectively reported by the patients. Also, we could not perform stress radiographs or anterior drawer testing due to the high probability of joint dislocation occurring during these tests in hEDS patients. The benefits of our proposed technique include minimizing wound healing complications with small incisions, lessening the risk of hematoma with minimal soft tissue dissection, reducing the chance of vessel rupture with no tourniquet use, and short duration of the procedure minimizing risk of anesthesia. Patients are also able to immediately bear weight after the surgery, minimizing the need for assistive devices such as crutches, which are often difficult for patients with hEDS to use. Future studies should involve a larger patient population and may consider using ankle joint arthroscopic debridement to address any ankle pathology that may be contributing to pain. However, given the positive results of this study, we recommend using our minimally invasive technique to address foot and ankle hypermobility and pain in patients with hEDS.



Figures 3a and 3b. Final post-operative images of minimally invasive lateral ankle stabilization and STJ arthroereisis procedures.

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