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Talonavicular-cuneiform arthrodesis in the management of Mueller-Weiss Syndrome: a retrospective case series

G. UZER[1], M. DEMIREL[2], D. KARA[1], B. TOKER[3], F. YILDIZ[1], V. UCAN[1]

[1]Department of Orthopedics and Traumatology, Bezmialem Foundation University, Istanbul, Turkey; [2]Department of Orthopedics and Traumatology, İstanbul University, İstanbul School of Medicine, İstanbul, Turkey; [3] Clinic of Orthopaedics and Traumatology, Acıbadem Fulya Hospital, Sports Medicine Center, İstanbul, Turkey.

Correspondence at: Vahdet Ucan, Department of Orthopedics and Traumatology, Bezmialem Vakif University, Fatih, Istanbul, Turkey 34093, Phone: +90 554 658 67 50, Fax: +90 212-4531700, Email: vahdetucan@hotmail.com

Mueller-Weiss Syndrome (MWS), characterized by spontaneous adult-onset tarsal navicular osteonecrosis, is an uncommon cause of chronic midfoot pain that can lead to functional impairment and progressive deformities. This study aimed to present clinical and radiological outcomes of talonavicular-cuneiform (TNC) arthrodesis in the treatment of patients with MWS. A retrospective study was performed on 8 consecutive patients (6 female, 2 male; mean age = 50 years; range = 33-64) who underwent TNC arthrodesis using plate fixation with autologous bone grafting for the treatment of MWS. To evaluate clinical status, the American Orthopaedic Foot and Ankle Society (AOFAS) Ankle-Midfoot Score was performed immediately preoperatively and at the final follow-up. In radiographic evaluation, talus-first metatarsal angle (Meary's angle) was measured preoperatively and at the final follow-up. Solid fusion was also examined on postoperative radiographs and computerised tomography. The mean follow-up was 35 months (range = 24-52). The mean AOFAS improved from 37 (range = 24-53) preoperatively to 85 (range = 80-93) at the final follow-up ($p < 0.001$). No major intra-operative complications were observed in any of the patients. According to the Maceira and Rochera radiological staging system, 5 feet was stage 3, and 3 feet was stage 4. The mean union time was 10 months (range = 5-15). Radiographic solid fusion was achieved in all but one foot that developed talonavicular non-union. TNC arthrodesis using plate fixation with autologous bone grafting seems to be an effective surgical method for reconstruction of MWS.

Keywords: Mueller-Weiss Syndrome, spontaneous osteonecrosis, tarsal navicular, talonavicular-cuneiform arthrodesis, navicular collapse.

INTRODUCTION

Koehler's disease, or childhood osteochondrosis of the tarsal navicular, is a well-known self-limiting disorder. Mueller- Weiss Syndrome (MWS) is a different clinical condition characterized by spontaneous adult-onset tarsal navicular osteonecrosis and should not be confounded by Koehler's disease[14]. MWS more often occurs in women than in men at a ratio of 6:4, and its true incidence remains unknown[5,18]. Previous studies have found patient age at the time of diagnosis was over 40 years[6,10,17].

MWS is an uncommon cause of chronic midfoot pain that can lead to functional impairment and progressive deformities secondary to the fragmentation and collapse of the navicular[10]. The exact etiology of the navicular collapse remains uncertain, but osteonecrosis is regarded as the most likely mechanism[15]. Paradoxic flatfeet involving hindfoot varus in lieu of traditional valgus represents the distinctive clinical feature of MWS, and the diagnosis may be readily delayed or missed because of the unusual clinical and radiological findings[12].

In the management of MWS, non-operative supportive measures such as shoe modification, bracing, and orthotics, are generally considered the first-line treatment, whereas surgery is mostly reserved for cases refractory to non-operative treatment. The main goal of operative treatment is to obtain pain-free, plantigrade foot with restoration of the medial column height[2, 12]. To achieve this goal, arthrodesis[2] or miscellaneous joint-sparing surgical methods (internal fixation of navicular bone[2], removal of the necrotic bone and replacement with autologous iliac cancellous bone graft[15], and percutaneous decompression[7] etc.) have been proposed in the literature. However, the latter techniques are based on single case reports, and to our knowledge, arthrodesis remains the mainstay of treatment for symptomatic relief and deformity correction in MWS.

A variety of techniques for arthrodesis have been proposed in the management of MWS, comprising talonavicular arthrodesis[3], talonavicular-cuneiform (TNC) arthrodesis[17], double arthrodesis[2], and triple arthrodesis[16,18]. Nonetheless, there is no consensus regarding the optimal surgical treatment for MWS, with a limited number of case series in the literature. This study aimed to introduce clinical and radiological outcomes of TNC arthrodesis in the treatment of patients with MWS in a consecutive case series.

MATERIALS AND METHODS

The medical records of 9 consecutive patients in whom MWS was diagnosed and treated by TNC arthrodesis from 2016 to 2019 at our institution were retrospectively identified. Inclusion criteria for the study were: (1) a diagnosis of MWS; (2) a minimum follow-up of 24 months; (3) complete medical records and radiographic images; (4) being willing to participate in the study. Exclusion criteria included: (1) diagnosis of other disorders that can involve the midfoot, such as Kohler disease, stress or traumatic fractures of the navicular bone, and Charcot arthropathy; (2) lost to follow-up; (3) inadequate medical and/or radiological records; and (4) being unwilling to participate in the study. One patient was lost to follow-up, thus 8 patients were included in the study and invited to a final follow-up appointment. The approval of institutional review board was obtained before data collection, and informed consent was obtained from all participants.

The diagnosis of MWS was established based on the history, clinical examination, and radiographical evaluation. Demographic and clinical data were collected from the hospital electronic database including age at the time of surgery, gender, involvement side, duration of preoperative symptoms, and follow-up duration. Intra-and post-operative complications were also recorded.

The feet were clinically examined preoperatively by the senior author for the presence of the following features: hindfoot alignment, subtalar motion, and medial longitudinal arch height. The American Orthopaedic Foot and Ankle Society (AOFAS) Ankle-Midfoot Score was performed immediately preoperatively and at the final follow-up[8].

All radiographic examinations were done on the weight-bearing anteroposterior (AP) and lateral radiographs of the foot by a single orthopedic surgeon with a special interest in foot and ankle surgery. Solid fusion was defined as an osseous bridging of at least three cortices across the arthrodesis site on the standard

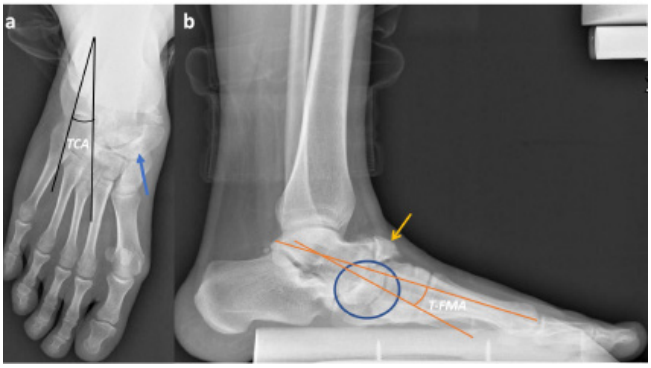

Figure 1. — Anteroposterior radiograph (a) shows the characteristic changes in the navicular bone secondary to Mueller-Weiss syndrome: compression and collapse of the lateral part of the navicular and subsequent development of comma shape (blue arrow). Note that TCA (talocalcaneal angle) refers to the angle between lines drawn from the lateral border of the calcaneus and axis of the talus. Lateral radiograph (b) shows condensation, fragmentation, and collapse of the navicular (yellow arrow). Note that T-FMA (talus-first metatarsal angle) indicates the angle between lines drawn from the longitudinal axis of the talus and first metatarsal.

postoperative radiographs. Computed tomography (CT) was used to confirm solid fusion in all patients.

On the preoperative AP radiographs, characteristic findings of MWS including "comma shape" sign, compression, and/or collapse of the navicular bone [13] were investigated, and the talocalcaneal angle (Kite's angle) indicating the position of calcaneus under the talus was measured (> 40° heel valgus and < 25° heel varus)[4] (Fig. 1).

On the preoperative lateral radiographs, other characteristic findings including flattening (increasement in the dorsoplantar length of the navicular) and condensation of the navicular bone[13] were examined. Talus-first metatarsal angle (Meary's angle) was measured on both preoperative and final follow-up lateral radiographs. Each case was graded preoperatively based on the Maceira and Rochera radiological staging system[9]. Osteoarthritis in the involved and adjacent joints was also investigated.

The main indication for TNC arthrodesis was chronic, intractable pain on the dorsum of the foot and perinavicular area unresponsive to nonsurgical treatment and functional impairment in daily life activities such as walking and climbing in all the patients. All operations were performed by a single experienced foot and ankle surgeon. The choice of surgical technique was based on the surgeon's preference and experience.

A standardized protocol of surgical technique was used under spinal anesthesia with a pneumatic thigh tourniquet in all patients. The patient was placed on

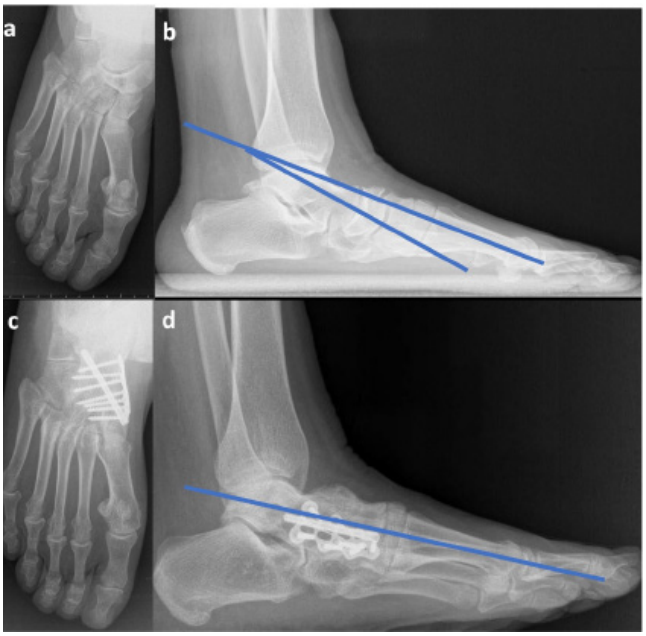

Figure 2. — Preoperative anteroposterior (a) and lateral (b) radiographs show a case of stage III Mueller-Weiss syndrome characterized by collapse of the navicular with an apex dorsal Meary's angle (talus-first metatarsal angle). Postoperative radiographs illustrate complete solid fusion along the medial column of the foot (a and b) and restoration of the medial longitudinal arch with an increase in Meary's angle (b).

a radiolucent table in a supine position. In addition to the affected lower limb, the ipsilateral iliac crest was draped to allow harvest autologous bone graft if needed. The talonavicular and naviculocuneiform joints were exposed via a longitudinal medial incision, originating from 1 cm proximal to the talar head and extending to the naviculocuneiform joint. In the preparation of articular surfaces, both joints were distracted by a lamina spreader. Then, all the cartilage, sclerotic or necrotic bone was removed from the talonavicular and naviculocuneiform joints using curettes, osteotomes, and rongeurs until viable sub-chondral bone was attained. Additionally, to enhance the fusion, multiple holes were drilled in the subchondral bone.

After preparation of articular surfaces, a tricortical autogenous bone graft was harvested in 4 patients from the iliac crest in a proper size of bony gap and inserted into the arthrodesis site. To obtain rigid fixation, a longitudinal screw was first inserted from the medial cuneiform to talar neck to ensure a more stable medial column, and then the arthrodesis site was affixed using a 3.5 mm LC-DCP plate with screws spanning talonavicular and naviculocuneiform joints. The lateral and medial cuneiforms was attempted to obtain with screws from the same plate inserted in each of the two cuneiforms after the preparation of articular surfaces.

Additionally, cancellous iliac crest bone graft was routinely inserted to the arthrodesis site in all patients. The construction and stability were assessed by direct observation and fluoroscopic imaging intraoperatively. After confirming the stability, wound closure was performed.

All statistical analyses were done using SPSS software (version 25, Armonk, NY: IBM Corp.). A P-value <0.05 was regarded significant. Normality tests were conducted using the Shapiro-Wilk test and histogram graphics. Data are presented as "minimum", "maximum", and "arithmetic mean". For differences from pre- to final follow-up, a paired sample T test was used.

RESULTS

8 feet of 8 patients were operated for symptomatic MWS. There were 6 females and 2 males with a mean age of 50 years (range = 33-64). All the patients were presented with unilateral, symptomatic MWS, but contralateral feet exhibited characteristic radiographic findings of the MWS in 5 patients albeit clinically asymptomatic. The mean follow-up was 35 months (range = 24-52). The mean duration of preoperative symptoms was 22 months (range = 9-38) (Table I).

In preoperative clinical examination, heel alignment was in neutral position in 7 feet and valgus in 1 foot. No restriction in subtalar motion was detected in any of the cases. Medial longitudinal arch was low in all the feet.

The mean AOFAS improved from 37 (range = 24-53) preoperatively to 85 (range = 80-93) at the final follow-up ($p < 0.001$) (Table I).

No major intra-operative complications were observed in any of the patients. One patient post-operatively developed superficial wound infection that resolved on treatment with antibiotic. One patient developed talonavicular nonunion and required bone grafting one year after initial surgery. In this patient, solid union was achieved three months after the revision surgery.

According to the Maceira and Rochera radiological staging system, 5 feet was stage 3, and 3 feet was stage 4. The mean union time was 10 months (range = 5-15). Radiographic solid fusion was achieved in all but one foot that developed talonavicular non-union. CT scan also confirmed solid fusion in all cases.

In the preoperative radiographic assessment of the navicular bone, the AP view revealed comma shape and compression signs in all feet in addition to collapse in 6 feet. The mean talocalcaneal angle was 18.5 °

Table I. — Patients' baseline characteristics and clinical outcomes

Number of patients	8
Gender (Female/Male)	6 / 2
Age (year), mean (min to max)	50 (33-64)
Number of the feet operated for symptomatic MWD	8
Contralateral radiographic involvement (clinically asymptomatic)	5
Follow-up (month), mean (min-max)	35 (24-52)
Duration of preoperative symptoms (month), mean (min-max)	22 months (range = 9-38)
Clinical findings	Heel alignment Neutral heel alignment → 7 feet Heel valgus → 1 foot Heel varus → 0 Subtalar motion No restriction in any of the cases. Medial longitudinal arch Low in all feet
AOFAS score, mean (min-max) Preoperative Final Follow-up	 37 (24-53) 85 (80-93)
Complications	Major complication → 1 patient • Talonavicular non-union Minor complication → 1 patient • Superficial wound infection
MWD = Mueller Weiss Disease; AOFAS = The American Orthopaedic Foot and Ankle Society.	

Table II. — Patients' radiographic assessment

Radiographic parameter	
AP X-ray examination	
Comma shape sign	All feet
Compression of the navicular bone	All feet
Collapse of the navicular bone	6 feet
Talocalcaneal angle (°), mean (min to max)	18.5 ° (15 °-30 °)
Flattening of the navicular bone	7 feet
Condensation of the navicular bone	All feet
Fragmentation	4 feet
Talus-first metatarsal angle (°), mean (min to max) *Preoperative* *Final follow-up*	 -8.5 (range = -5 - -11) -0.25 (range = -2-0)

(range = 15-30). Based on the talocalcaneal angle, 7 feet demonstrated heel varus while one foot showed neutral heel alignment.

The lateral view showed flattening of the navicular bone in 7 feet, fragmentation in 4 feet and condensation in all feet (Table II). The mean talus-first metatarsal angle was increased from -8.5 (range = 5-11) preoperatively to -0.25 (range= -2-0) ($p < 0.001$) at the final follow-up.

DISCUSSION

MWS is an uncommon cause of midfoot pain in adults that is characterized by the navicular collapse and further flatfoot deformity. The literature is controversial regarding the etiology, pathomechanism, and natural course of MWS. Conservative treatment has been suggested as first-line treatment by many authors for MWS and may contain patient education, non-steroidal anti-inflammatory medicines, weight loss, shoe modification, orthotics, bracing, and casting[2,10]. When conservative treatment fails, surgical treatment is indicated, and arthrodesis is considered as the treatment of choice. Nonetheless, joint-sparing calcaneal osteotomies has been recently reported with promising clinical and radiographic results[1].

Although various types of perinavicular arthrodesis have been performed by several authors[3,17,18] for the treatment of cases refractory to conservative treatment, the optimum fusion technique has not yet been determined in the literature[2]. TNC arthrodesis is one of the options that can be used to reconstruct the medial column collapse with arch height loss in patients with MWS, but there are, to our knowledge, a limited number of case series published due to rarity of MWS[3,17,18]. Accordingly, we primarily aimed to investigate the mid-term outcomes of TNC arthrodesis using plate fixation with autologous bone grafting in a consecutive case series of MWS. The results of this study have confirmed the feasibility and effectiveness of this technique.

In one of few retrospective studies on the TNC arthrodesis in the management of MWS, Zhang et al.[18] compared the results of TNC arthrodesis (5 patients; 6 feet) and triple fusion (5 patients; 5 feet) in patients with stage IV disease. The authors determined that both methods can provide significant improvement in AOFAS with a fusion rate of 100% for the management of such patients if the proper method is chosen according to a meticulous radiological examination. In contrast, Maceira et al.[9] supported the use of TNC arthrodesis with the rationale that triple fusion cannot address the naviculocuneiform degeneration. In another study, Cao et al.[3] compared the results of TNC arthrodesis (14 patients; 14 feet) and TN arthrodesis (16 patients; 16 feet) in patients with stage III and IV MWS. They found both TN and TNC arthrodesis are reasonable treatment option for stage III or IV MWS with a fusion rate of 100% and remarkable amelioration in AOFAS. In other study, Yu et al.[17] reported satisfactory clinical outcomes with a fusion rate of 100% in a case series of 7 patients (7 feet) by TNC arthrodesis using plate and screws. Although available data in the literature is limited, our results of TNC arthrodesis in the treatment of MWS are consistent with previous observations.

Stage III and IV MWS is often manifest with compression and fragmentation of the navicular, and thus the articular surfaces of the talonavicular and naviculocuneiform joints are typically disrupted[9,10]. Furthermore, collapse of the navicular results in medial column collapse and acquired flatfoot deformity[12]. Accordingly, rigid internal fixation spanning all affected joints may be a viable option for reconstruction of each component of the deformity. Fixation may be achieved by various instruments such as, staples, screws and/or plates[11].We preferred to use a 3.5 mm LC-DCP plate for TNC arthrodesis, and a longitudinal screw was routinely inserted from the medial cuneiform to talar neck to ensure a more stable medial column and eventually fusion. Since the navicular articulates with three cuneiforms, arthrodesis between the lateral and medial cuneiforms was attempted to obtain with screws from the same plate inserted in each of the two cuneiforms following the preparation of articular surfaces[10]. This approach depended on the case-by-case basis and senior author's preference. Alternatively, this can be achieved with bi-columnar fixation with multiple smaller plates (2.7 mm)[10]. However, we consider that to obtain sufficient axial stability of TNC arthrodesis and to prevent further implant failure, a single large plate should be applied to the medial side of the medial column. With this approach, solid fusion was achieved without implant failure in all but one patient who developed non-union of the talonavicular joint and required subsequent bone grafting one year after initial surgery.

With respect to bone grafting, different graft materials, such as allograft, autograft, and synthetic graft materials may be necessary to augment TNC arthrodesis due to poor bone quality and bone defect. Favorable results have been reported using both autograft and allograft in the management of MWS[11]. In our case series, autologous tricortical iliac crest graft was required in 4 patients, especially with stage IV MWS, because of the remaining large defect following the debridement of the unviable navicular. In the remaining patients, autologous cancellous bone grafting was sufficient to achieve stable fixation and to fill the navicular defect. That is, the need for grafting depends on the case-by-case basis, and which material to use is based on the surgeon's preference and experience. We support the use of autologous iliac graft to enhance bone fusion and achieve sufficient stability at the TNC arthrodesis site in the management of MWS.

Demographic data obtained from the current study are consistent with the former literature, as detailed in the methods section. Besides, the typical presentation of MWS patients is progressive midfoot pain as seen in our case series, and medial longitudinal arch height may be normal or low with hindfoot varus in physical examination based on the severity of the dorsolateral fragmentation and collapse of the navicular[2,11,12]. Paradoxic flatfoot characterized by hindfoot varus in lieu of traditional valgus represents the distinctive clinical feature of MWS. Heel varus is caused by lateral shift of the talar head secondary to medial extrusion of the navicular[12]. Thus, this phenomenon is related to the severity of the disease and can be hard to clinically detect[2]. In accordance with this, although all our patients exhibited flatfoot deformity, hindfoot varus was not clinically detected in any of our patients with stage III and IV MWS as per Maceira classification. However, AP radiograph revealed heel varus according to talocalcaneal angle in seven feet.

When interpreting the findings in this study, some limitations and strengths should be considered. The major limitations were its retrospective nature, limited sample size, and short-to mid-term follow. Despite these limitations, our study is one few studies[3,17,18] that present the results of TNC arthrodesis using plate fixation with autologous bone grafting in MWS.

CONCLUSION

There is no gold standard treatment for patients with MWS. The surgical procedure of choice should be individualized for each patient. TNC arthrodesis using plate fixation with autologous bone grafting seems to be an effective surgical method for reconstruction of MWS if applied properly, with significant improvement in clinical and radiographical results.

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