Clinical case
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Talar head replacement for treatment of Müller – Weiss syndrome: three clinical cases

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Abstract

Introduction Müller – Weiss syndrome is a rare condition that is characterized by avascular necrosis of the navicular bone leading to severe foot deformity, pain disturbing activities of daily living. There is no generally accepted treatment for avascular necrosis of the scaphoid, and the available treatments have disadvantages, which necessitates the need for new options.

The **objective** was to present short-term results of three patients with osteonecrosis of the scaphoid and local necrosis of the talar head treated with ceramic talar head prosthesis, plastic surgery, metal osteosynthesis of the scaphoid and related surgical techniques.

Material and methods Three patients diagnosed with Müller – Weiss syndrome were treated with ceramic talar head replacement, autologous bone grafting and metal osteosynthesis of the scaphoid bone with accompanying surgical techniques. Severity of pain and the condition of patients were assessed with the VAS and AOFAS AH scores.

Results Short-term results showed consolidation at the site of metal osteosynthesis in all patients with no signs of instability of the talus hemiprosthesis. VAS and AOFAS AH scores indicated decrease in the pain and improved condition of the patients.

Discussion Hemiarthroplasty of the talar head combined with plastic surgery and restoration of a congruent joint surface of the scaphoid, and associated surgical techniques may become an effective alternative to existing treatments for patients with Müller – Weiss syndrome, with further study.

Conclusion The short-term findings showed that hemiarthroplasty was practical for restoration of the talonavicular mobility maintaining stable fixation of the talus.

Keywords: talonavicular joint, avascular necrosis, scaphoid bone, Müller – Weiss syndrome, joint replacement, ceramic implant

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INTRODUCTION

The talonavicular joint is part of the Chopart joint complex and is one of the key joints to ensure the foot function. It plays a significant role in the walking cycle distributing load vectors and is involved in pronation and supination of the foot [1]. Severe degenerative changes in the joints of the mid- and hindfoot can reduce the patient's quality of life affecting the foot function [2]. Osteonecrosis of the articular surfaces of the talonavicular joint is characterized by functional impairment. Osteonecrosis can be caused by post-traumatic conditions, rheumatoid arthritis and Keller's disease. Müller – Weiss syndrome is a rare disease that is characterized by ischaemic necrosis of the tarsal scaphoid [3]. Müller – Weiss disease is managed initially with conservative treatment that are not always effective [4]. Surgical options are reserved for severely destructed articular surfaces of the talonavicular joint [5, 6]. Arthrodesis of functionally important joints can result in poor functionality at a long term, in particular. An original endoprosthetic model of the talar head and a method for its implantation were offered to maintain the talonavicular function of severely deformed and destructed joint [7].

The **objective** was to present short-term results of three patients with osteonecrosis of the scaphoid and local necrosis of the talar head treated with ceramic talar head prosthesis, plastic surgery, metal osteosynthesis of the scaphoid and related surgical techniques.

MATERIAL AND METHODS

The study was performed in accordance with ethical principles for medical research involving human subjects stated in the Declaration of Helsinki developed by the World Medical Association as revised in 2013. The study received a favourable opinion from the relevant research ethics committee. The patients gave informed consent for publication of the findings without identification. Three patients diagnosed with Müller – Weiss syndrome were treated with ceramic talar head replacement, autologous bone grafting and metal osteosynthesis of the scaphoid bone with accompanying surgical techniques. Hemiprosthesis of the talar head is an original design developed in Russia and made of yttrium-stabilized zirconium oxide ceramics. The implant contains an articular head with a smooth articular surface and a four-lobed stem. Press-fit fixation occurs with the hemiendoprosthesis stem being self-jammed in the prepared canal of the talus. Weight-bearing radiography of the feet and multislice computed tomography (MSCT) were performed pre- and postoperatively to assess the objective condition of patients. VAS score and AOFAS AH score Severity of pain and the condition of patients were assessed with the VAS (visual analog pain scale) and AOFAS AH (American Orthopedic Foot and Ankle Society — Ankle Hindfoot Scale) scores preoperatively and at 12 months.

RESULTS

Case 1

A 70-year-old patient reported pain in the mid- and hindfoot on the right side at rest, which intensified during walking and after physical activity. He had a history of an injury a year ago with his right foot being sprained. He did not seek medical assistance. Over time, he experienced severe pain with negative clinical dynamics. Physical examination showed moderate swelling of the soft tissues in the mid- and hindfoot on the right side. His medial longitudinal arch appeared to be flattened and calcaneus aligned in varus. Palpation revealed severe pain at the talonavicular joint of the right foot. He could not supinate or pronate with the right foot. Preoperative Vas scored 10 and AOFAS AH measured 22 points. Computed tomography demonstrated severe deformity and dislocation of the talonavicular articular surfaces, fragmentation of the scaphoid bone, linked wedging of the talar head and a scaphoid fragment (Fig. 1).

Fig. 1 Preoperative MSCT sections of the right foot (*a*) sagittal plane; (*b*) axial plane showing severe degenerative changes in the talonavicular joint with the talus and navicular bones impacted

The patient was diagnosed with stage 3 osteoarthritis of the talonavicular joint, osteonecrosis of the scaphoid and the talar head of the right foot, blockade of the talonavicular joint. Medical history and physical examination indicated a stress fracture of the scaphoid which caused destruction of the talar head, fragmentation of the scaphoid and development of Müller – Weiss syndrome. Surgical intervention included debridement, bone grafting of the scaphoid bone defect using the anterior articular process of the calcaneus, metal osteosynthesis (MOS) of the scaphoid bone with screws, hemiendoprosthetics of the talar head with a ceramic implant, lengthening resection arthrodesis of the calcaneocuboid joint with plastic surgery using a cancellous allograft and a staple, achilloplasty by Strayer. The patient could maintain full weight-bearing on the right lower limb and did not limp at 12 months. There was no swelling of the soft tissues at the hindfoot on the right side. Moderate flattening of the medial longitudinal arch of the right foot was observed with and the hindfoot alignment being satisfactory (Fig. 2). Postoperative scars heeled with no signs of inflammation. Palpation of the talonavicular joint was painless. The patient had full and painless range of motion in the right ankle. The range of painless supination-pronation of the right foot in the Chopart joint measured 10°–0°–10°. The AOFAS AH scored 72 and VAS was 2. Weight bearing radiographs of the right foot showed stable MOS of the scaphoid bone, calcaneocuboid ankylosis (Fig. 3), moderate flattening of the longitudinal arch with the foot function restored.

Fig. 2 Appearance of feet at 12 months showing moderate flattening of the medial longitudinal arch of the right foot, satisfactory alignment of the hindfoot

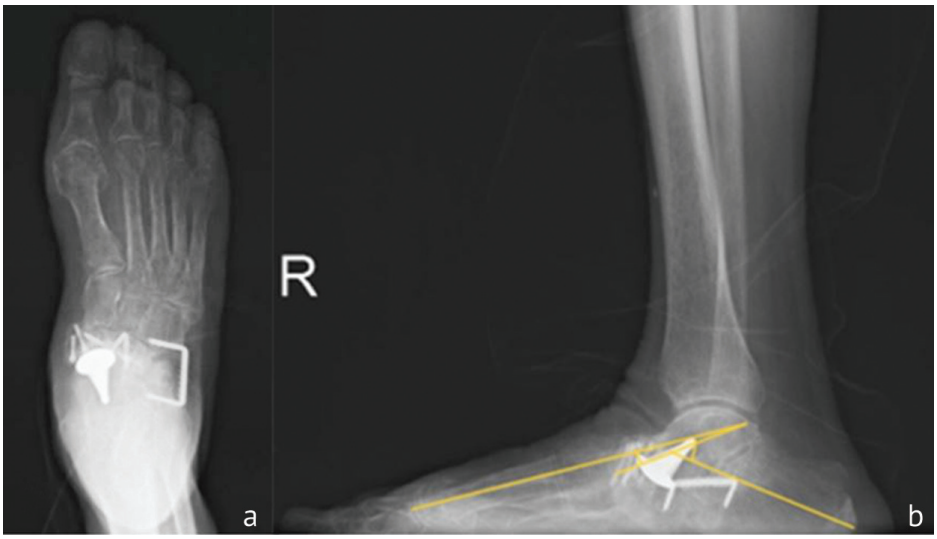

Fig. 3 Weight-bearing X-ray of the right foot at 12 months showing (*a*) AP view; (*b*) lateral view demonstrating Mary angle of 8° Kite and talocalcaneal angle of 45°

Case 2

A 43-year-old patient reported an acute pain in the mid- and hindfoot on the right side which developed for no apparent reason about one year ago. She underwent conservative treatment without positive dynamics. Clinical examination revealed pronounced swelling of the soft tissues of the mid- and hindfoot on the right side, flattening of the medial longitudinal arch and varus of the calcaneus of the right foot (Fig. 4).

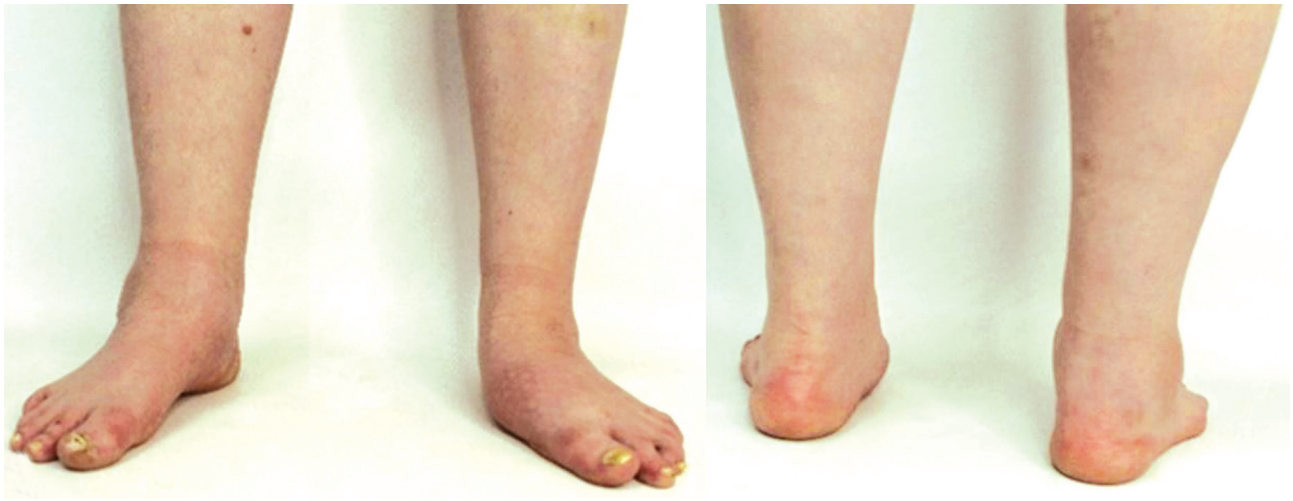

Fig. 4 Preoperative appearance of the feet showing varus of the hindfoot, pronounced flattening of the medial longitudinal arch of both feet, valgus of the hindfoot on the left side

The patient reported severe pain at the talonavicular joint on the right side on palpation. Supination and pronation of the right foot were not noted. No neurological deficit was identified. When testing the patient before surgery, Preoperative AOFAS AH scored 24 and the VAS scored 10. The left foot showed pronounced flattening of the medial longitudinal arch, valgus deformity of the hindfoot, and abduction of the forefoot. However, with the severe deformity, the foot was plantigrade and the patient reported no pain. Computed tomography of the right foot showed severe degenerative changes in the articular surfaces of the talonavicular joint, osteonecrosis, fragmentation of the scaphoid bone, and linked wedging between the talar head and a scaphoid fragment (Fig. 5).

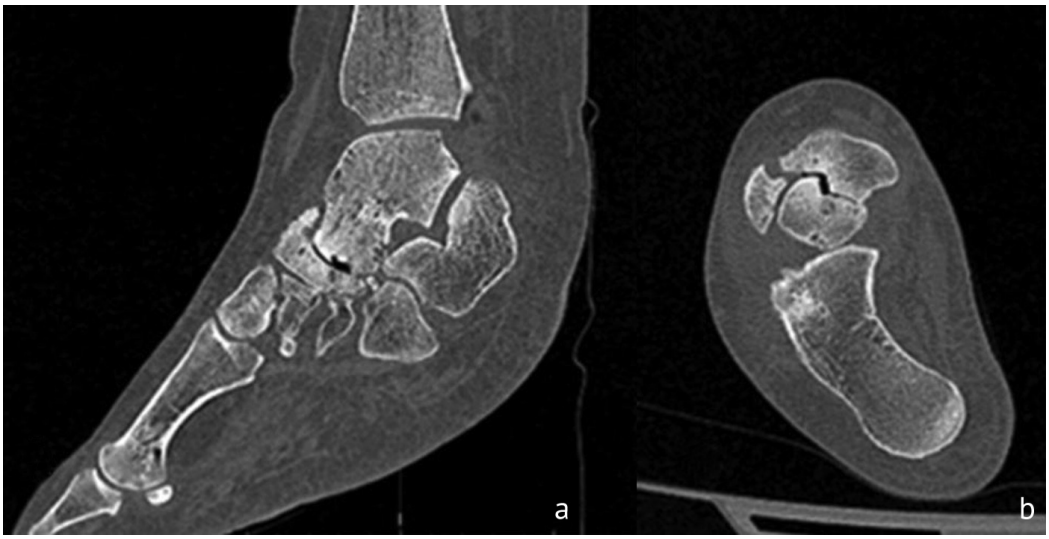

Fig. 5 Preoperative MSCT sections of the right foot in (*a*) sagittal plane; (*b*) axial plane showing severe osteoarthritis at the Chopart joint, fracture of the scaphoid, pathological angle of scaphoid inclination

The patient was diagnosed with Müller – Weiss syndrome, osteonecrosis of the navicular and the talar head of the right foot, blockade of the talonavicular joint. The first stage of surgical intervention included necrectomy of the scaphoid bone. MOS of the scaphoid bone was produced, pseudarthrosis of the scaphoid identified, and resected with an oscillatory saw down to the bleeding areas. The scaphoid bone fragments were reduced, fixed with a lag screw, and then with a mini-support plate with angular stability. The next stage included hemiendoprosthetics of the talar head with a ceramic implant, lengthening resection arthrodesis of the calcaneocuboid joint, plastic surgery with a bone cancellous allograft and fixation with a staple, and Vulpius achilloplasty. Physical examination showed slight swelling of the soft tissues of the hindfoot on the right side, the axis of the calcaneus aligned in a neutral position at 12 months (Fig. 6). Movements of the right foot in the Chopart joint, palpation of the talonavicular joint were painless, supination–pronation measured 10°–0°–5°. The AOFAS AH scored 95 and VAS scored 1. Weight-bearing radiographs of the right foot at 12 months showed a fracture of the staple, adequate osteosynthesis of the scaphoid and calcaneocuboid ankylosis identified (Fig. 7). The right foot was plantigrade and pain free.

Fig. 6 Appearance of the feet at 12 months showing slight swelling at the level of the right ankle joint, neutral position of the heel bone on the right

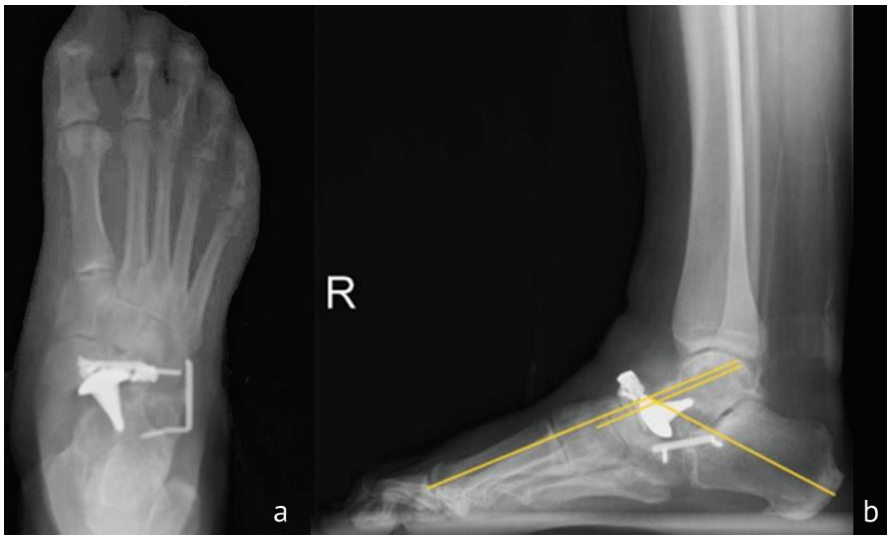

Fig. 7 Weight-bearing radiographs of the right foot at 12 months showing (*a*) AP view; (*b*) lateral view demonstrating Mary angle of 0°, Kite talocalcaneal angle of 50°. Scaphoid bone healed with MOS, satisfactory calcaneocuboid arthrodesis, broken connector bracket

Case 3

A 24-year-old female patient reported acute pain in the mid foot on her right side. She sustained an injury about 7 years ago, she sprained her right foot with no skeletal injury detected at the time. A physical examination revealed moderate swelling of the soft tissues in the mid- and hindfoot on the right side, flattening of the medial longitudinal arch of the right foot, and varus alignment of the calcaneus (Fig. 8).

Fig. 8 Preoperative appearance of the feet showing moderate swelling of the right foot, flattening of the medial longitudinal arch, varus of the calcaneus on the right

Severe pain in the right foot was detected upon palpation at the talonavicular joint. The supination-pronation movements of the right foot are rocking and sharply painful. When testing the patient before surgery, the AOFAS AH score was 39 points, and the VAS score was 9 points. Severe pain in the right foot is detected upon palpation in the projection of the talonavicular joint. She had swinging, sharply painful supination-pronation movements of the right foot. Preoperative AOFAS AH scored 39 and VAS scored 9. Computed tomography showed severe deformity of the articular surfaces of the talonavicular joint, osteonecrosis and fragmentation of the scaphoid bone and local avascular necrosis of the talar head (Fig. 9).

Fig. 9 Preoperative MSCT sections of the right foot in the sagittal plane (*a*) and axial plane (*b*) showing severe osteoarthritis of the talonavicular joint, cysts in the talar head, fragmentation of the scaphoid bone

The patient was diagnosed with Müller – Weiss syndrome, osteonecrosis of the scaphoid and the talar head of the right foot. Surgical intervention included necrectomy of the scaphoid bone, MOS with a mini-plate, hemiendoprosthetics of the talar head, Evans lengthening osteotomy of the calcaneus, plastic surgery with cancellous bone allograft and fixation with a connector staple. Physical examination at 12 months showed slight swelling of the soft tissues of the hindfoot on the right side and physiological position of the heel (Fig. 10). Painless supination and pronation of the right foot in the Chopart joint was 10°–0°–10° at 12 months. Palpation of the talonavicular joint was painless. AOFAS AH scored 87 and VAS scored 1. Weight-bearing radiographs of the right foot at 12 months demonstrated stable MOS of the scaphoid bone (Fig. 11). MSCT findings showed stable hemiendoprosthesis (Fig. 12).

Fig. 10 Appearance of the feet at 12 months showing clean postoperative scar, no signs of inflammation with the physiological position of the heel provided

Fig. 11 Weight-bearing lateral radiograph of the right foot at 12 months showing no signs of migration with neutral physiological position of the foot; Mary angle of 2°, Kite talocalcaneal angle of 50°

Fig. 12 MSCT sections of the right foot at 12 months in (a) sagittal plane and (b) axial plane showing consolidation of the scaphoid bone with MOS; adequate fixation of hemiprosthesis

DISCUSSION

Destruction of the articular surfaces of the talonavicular joint can be caused by primary osteonecrosis, previous trauma, osteochondritis, Keller disease, stress fracture of the scaphoid, rheumatoid arthritis or Müller – Weiss syndrome. Traumatic fractures of the scaphoid are a rare injury and diagnosis is difficult. About 30 % of all stress fractures of the foot involve injuries to the navicular bone [8]. The scaphoid is susceptible to osteonecrosis due to its complex anatomy and blood supply and an injury can lead to post-traumatic osteoarthritis of the surrounding joints. In childhood, avascular necrosis of the scaphoid is represented by Keller disease that normally develops at the age of 2–10 years [9]. In adults, the condition is termed Müller – Weiss syndrome and causes collapse and fragmentation of the scaphoid bone. Müller – Weiss syndrome was first described in 1927

by the German surgeon Walther Müller and the Austrian radiologist Konrad Weiss. However, some would suggest that this condition was first described by Schmidt in 1925 [10]. There is no generally accepted opinion about the cause of Müller – Weiss disease [11]. However, there are several theories to include osteonecrosis, osteochondritis, post-traumatic necrosis or a consequence of biomechanical disorders, osteoarthritis (OA) due to scaphoid dysplasia, congenital malformation, repeated stress fractures of the scaphoid and others. An etiopathogenetic theory has been recently accepted to involve dysplastic, mechanical, and socioeconomic environmental factors [12]. However, Doyle et al. did not find this relationship [13].

The blood supply to the navicular bone is provided by the medial plantar artery, the dorsalis pedis artery and the tarsal canal artery [14]. The central part of the scaphoid has a poorer blood supply, which also tends to decrease with age [15]. Tan et al. conducted the only pathological study to date and identified osteonecrosis in the material explored [16]. Other authors reported normal bone tissue [17].

In 2004, E. Maceira, R. Rochera reported dysplasia of the tarsal and scaphoid bones with delayed ossification and associated pathological load distribution [12]. The condition was associated with a significant increase in pressure on the lateral edge of the scaphoid. Varus deformity of the foot at th subtalar joint and a short first metatarsal bone could be predisposing factors. The prevalence of the disease in the population is unknown. The condition is asymptomatic and is often diagnosed with severe osteoarthritis of the talonavicular joint and an accurate diagnosis is difficult to make. This condition is most common in women aged 40–46 years, with an incidence rate of 6:1 or 2:1 compared to men. Bilateral involvement is observed in 50 % of cases [18]. Patients often experience pain along the dorsal-medial surface of the mid- or hindfoot, and there is a tendency for negative dynamics. The deformity is characterized by varus of the hindfoot, the so-called paradoxical flatfoot deformity (Pes planovarus, paradoxical flatfoot deformity) with flattening of the medial longitudinal arch and abductor deformity of the midfoot. In this case, manifestations of osteoarthritis are observed in the joints adjacent to the scaphoid bone. Progressive fragmentation of the scaphoid bone and collapse on the lateral surface, displacement of the talar head the lateral sphenoid bone are observed in the pathogenesis of the disease. Radiologic diagnostic criteria were offered by Maceira and Rochera in 2004 and summarized in Table 1 [12].

Table 1

Radiological classification of Müller – Weiss syndrome developed by Maceira and Rochera

Stage	Description
1	Standard radiographs, technetium scan, MSCT and MRI (intraosseous edema) Minor subtalar varus may be present
2	Lateral displacement of the talar head causes subtalar varus
3	Splitting or compression of the scaphoid bone leads to decreased height of the medial arch
4	Progressive compression of the navicular bone results in hindfoot equinus
5	Complete extrusion of the scaphoid and close contact of the talus and sphenoid

Radiological examination can show changes in the shape of the scaphoid bone: a concave shape (boat-shaped) is changed into a “comma” with decreased thickness along the lateral edge caused by progressive compression of the lateral aspect of the scaphoid bone. The cuboid bone can be displaced medially (cuboid sign). Radiographs show an open tarsal sinus, which is caused by varus alignment of the hindfoot. X-rays demonstrate a hypertrophied second metatarsal bone. The changes

are caused by the lateral shift and increased load on it. Increased plantar pressure in the midfoot and medial edge of the hindfoot can cause pathological changes in the foot [19]. MRI shows decreased signal intensity from the scaphoid on T1-weighted images (T1WI), hyperintense diffuse edema and hyperintense periarticular fluid on T2-weighted images (T2WI). Although conservative treatment can be applied for early stages of the disease (stages 1 and 2 as graded by Maceira and Rochera), and may fail to provide positive results, the treatment strategy is considered first-line therapy [3, 4].

Conservative treatment includes the use of orthopedic insoles, limited physical activity, immobilization, taking NSAIDs and analgesics and lasts at least 3 months. The use of orthopedic insoles with semi-rigid support of the medial longitudinal arch shows satisfactory results and the effectiveness of the treatment depends on the angle of inclination of the heel and abduction of the foot [20]. There is a variety of surgical treatments. Some authors recommend isolated arthrodeses of the talonavicular joint. The established methods of surgical treatment are various combinations of arthrodeses including double and triple arthrodeses [5, 6, 21]. Correction of rotational displacement of the talus followed by talonavicular or talonavicular-sphenoid arthrodesis can facilitate satisfactory treatment results [22]. Arthrodeses are used in combination with autologous bone grafting harvested from the iliac wing [23]. Scaphoid resection followed by medial column reconstruction using femoral head allograft for fusion using a plate is reported [16]. Blocking of the functionally significant joints leads to poor results, which become pronounced at a long term, therefore, arthrodesis of the talonavicular joint should be avoided [24, 25]. Joint-saving interventions in the treatment of Müller – Weiss disease include percutaneous decompression of the scaphoid in the early stages of the disease, debridement of the scaphoid and replacement with a bone cancellous graft using the ilium [26, 27], debridement of the scaphoid and bone grafting with a vascularized graft harvested from the medial femoral condyle [28]. It is difficult to draw conclusions about the effectiveness of the interventions due to the small number of observations. Realignment of the hindfoot is one of the important treatment goals that can be achieved with calcaneal osteotomy [20].

Lengthening osteotomy of the calcaneus unloads the lateral aspects of the navicular bone, corrects the deformity, relieves pain and improves foot function. Lengthening and stabilization of the lateral column can also occur due to lengthening calcaneocuboid arthrodesis with allograft reconstruction [29]. N. Wülker reports calcaneocuboid arthrodesis reducing the range of motion in the talonavicular joint by 19 % and after surgery measuring 32.95 ± 5.14° postoperatively [30]. In our opinion, this is not critical for the foot function of the and can stop progression of the deformity and reduce the load on the contact surfaces of the talonavicular joint. Maintaining the mobility of the talonavicular joint is important. The use of implants made by 3D printing can be promising in the treatment of osteonecrosis of the scaphoid and the talar head. The use of custom-made titanium scaphoid implants is reported in the literature. SB Adams and RM Danilkowicz reported a successful results for a patient with osteonecrosis of the scaphoid after implantation of a custom-made titanium scaphoid implant with a follow-up period of 4 years [24]. Zirconium ceramics cause less damage to the opposite cartilaginous surface than metal and is more preferable for hemiendoprosthetics [31. 32].

We have been using the technique of hemiprosthetics of the talar head since 2021; more than 20 surgical interventions aimed at replacing the articular surface of the talar head have been performed at the time of publication. We often used this surgical intervention for osteoarthritis of the talonavicular joint, accompanied by concomitant surgical techniques. These implants

are an original domestic development; patent No. 2788474 was received for the model of the endoprosthesis of the talar head and the method of its implantation [7]. The implant line is presented in 4 standard sizes, which allow you to select the necessary implant, regardless of the size of the patient's foot. We are conducting further work to evaluate the surgical technique in patients with lesions of the talonavicular joint of various origins.

CONCLUSION

The short-term findings showed that hemiarthroplasty was practical for restoration of the talonavicular mobility maintaining stable fixation of the talus.

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Ethical review *Not applicable.*

Informed Consent *The patients gave informed consent for publication of the findings without identification.*

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