

# Avascular Necrosis of the Tarsal Navicular Bone

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As an orthopedist, it is not unusual to encounter young toddlers generally four- or five-year-old to nine-year-old complaining of foot pain, impending on their ability to ambulate, with a characteristic pain worsening when running or playing at school. The child is often seen refusing to participate in gym activity. On physical examination, the medial aspect of the foot is painful to palpation and when asked to bear weight or ambulate, the child presents an antalgic gait with the flattening of the medial arch or even a complete collapse. Radiographic studies will often present a radio dense navicular bone. For most orthopedists, it is not difficult at this stage to diagnose a rare bone disease called “Kohler disease”, which represent an avascular necrosis of a small tarsal bone called navicular of the foot.

First described in 1908, this rare bone disease is a self-limiting disease, generally unilateral affecting especially boys but rarely seen before the age of two, unless the patient is a girl. Rarely, the presentation can be bilateral. We do not know the reason, but some experts advance many theories which may be related to the child's early onset of ossification. Others will find a better explanation in the theory of multiple traumas as a mechanical cause. The Navicular is, the last of the tarsal bone to ossify while the cuneiform and the calcaneus ossify earlier.



### Kohlars Disease

An “adult onset” form of this osteonecrosis of the navicular bone do exist as well and is called “Syndrome of Mueller-Weiss” because of its spontaneous appearance causing similar symptoms of pain to the mid and hindfoot with deformity. The blood flow appears to be cut off from the navicular bone, resulting in degeneration, deformity and fragmentation. In this adult form, the condition persists with pain over the medial arch lasting for months and progressing to severe and even disabling pain. This adult form is more often seen in the middle-aged women with a suspicion of trauma or following an acute injury in sport. The death of the bone can also be seen... but the true cause remains unknown.

The prognosis is considered excellent in Koehler’ disease because of the persistence of the blood supply and often a normal trabecular pattern may be observed. Radiographically, an appearance of uniformity and an increase in bone density is noted despite of a minimal fragmentation. We like to approach such a stage using a longitudinal arch support with medial wedge but occasionally the application of a short leg cast for six weeks may be needed. If the

pain persists, other causes like tarsal coalition may be looked for, complicating the picture. At 18-months of age, after the onset of such symptoms, most patients should recover from their symptoms.

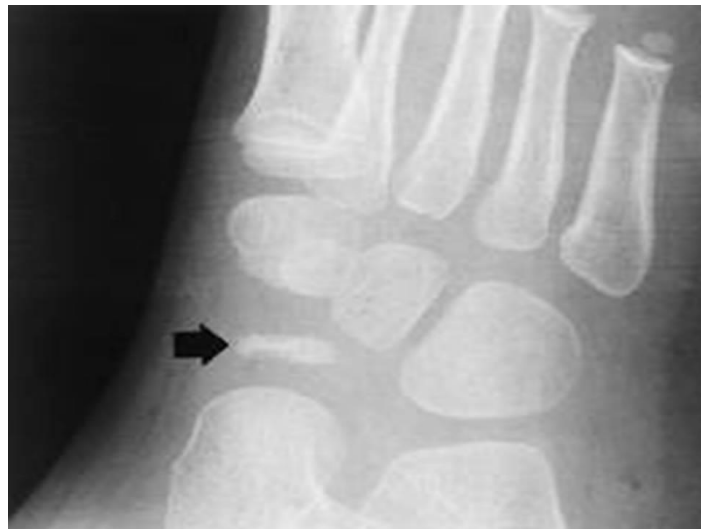
I chose to approach this topic because one of my favorite Tennis professional players is suffering from the complications of such a rare disease: Rafael Nadal. 35-year-old tennis champion. He is still considered on the top of the ranking in the world of Tennis (presently # 5) and has won recently the Australian Open on a surface that he masters so well. He has won the greater number of Grand Slam men's titles in history. Despite of these victories, he has been struggling with his left foot condition for years. He pulled out from the last year Wimbledon tournament and I hear him in an interview recently, saying that he believes that his career may be over but he is still contemplating a comeback. He has reached a point where we can guess that the day may be close for us to see him back on the courts, playing again.

In presence of Kohler 's disease, a conservative treatment remains the most popular approach to manage the symptoms but if persistence of pain is noted six months after, an arthrodesis may become the surgical treatment of choice. Often it can be performed arthroscopically or via an open procedure in a triple fusion as a talonavicular-cuneiform fusion to alleviate the symptoms of pain.

I know well that Nadal underwent a previous surgical treatment in September 2021 which facilitated his return to the court. This foot will certainly never return to normal nor that such operation will allow RAFAEL Nadal to emerge as the dominant player he was once. The Mueller-Weiss syndrome may not go away but the degeneration and the deformity of such peculiar bone will continue to be a cause for pain and discomfort, enabling him to remain on the top of his game. We will be witnesses of recurrent flare-ups. We will be unable

to tell the world that his best years are yet to come. I remain perplex and find myself unable to comprehend how such a champion has been able to manage a return with such a painful disease.

The navicular bone loses its blood supply for a while and as a result, part of the bone dies and results in a collapse but whenever treatment is offered, there are no long-term problems at recovery unless the onset was related to the adult age (Mueller-Weiss) form. Pain with a usual limp is expected often in this adult form and the patient may shift their weight on the lateral side of the foot to accommodate the gait. Bilateral radiological examination may reveal a unilateral or bilateral disease showing a sclerotic and flattened navicular bone. The affected foot needs to be treated by any means of support like arches for six months, pain relievers and protected weight bearing with crutches, short leg casts for six to eight weeks although others believe that various boots can offer the same support and protection. Moderate exercises can be beneficial as well as physical therapy.



## Surgical Treatment

Mueller-Weiss disease is certainly a rarely diagnosed disease in which a deformed tarsal navicular bone undergoes spontaneous osteonecrosis. Let us explore the surgical options for the treatment of such disease because until now, there has not been any widely accepted ways in treating this entity. Specialists so far are comparing their clinical and radiographical outcomes following procedures like a "Core decompression," as the least traumatic to an internal fixation as to treat the disease like it was considered as a fracture.

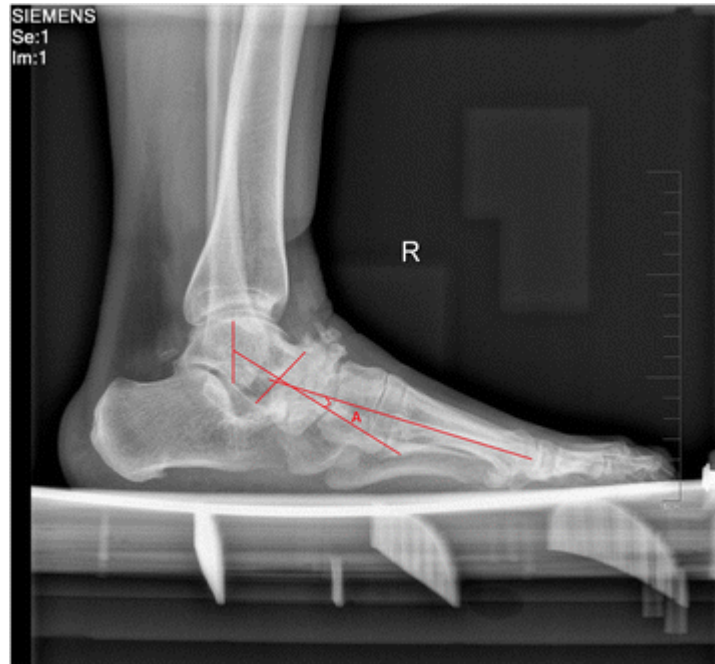
An arthroscopic arthrodesis of two tarsal bones (Talus and Navicular) or finally an open internal fixation of the same two bone to three bones (Talus, Navicular, and cuneiform). A triple arthrodesis versus (TNC: Talo Navicular Cuneiform) remains the operation for an advanced disease of Mueller-Weiss. Many have approached the disease in different stages. A salvage operation for the reconstruction of the medial column of the foot with removal of part of the navicular bone followed by a bony reconstruction with bone graft, this is what the "TNC" brings to the array of options.

This allows the orthopedic surgeon to realize really that Mueller-Weiss disease (MWD) is a completely different disease from Koehler osteochondritis. Mueller-Weiss disease (MWD) is a complicated idiopathic foot condition presenting with chronic midfoot pain and deformity related to a disease tarsal navicular bone. The prevalence and the incidence are still not well known but MWD is more often bilateral and more commonly seen in

women between 40 and 60 of age. The disease is more commonly encountered in Europe than the United States suggesting an environmental or even a nutritional link. Other causes related to osteonecrosis, with traumatic and biomechanical factors have explained the degenerative changes seen around the navicular bone. Scientists have stipulated the possibility of an abnormal evolution of the Kohler disease itself to explain the adult-onset findings with delays in ossification.

In Kohler, the disease is generally unilateral and rare in women, but once diagnosed, it is generally in a unilateral presentation for a male unless it is a female under the age of two. Some experts believe that the hindfoot present a varus deformity and most patient suffering with the disease, accuse chronic mid-foot pain and swelling with flatfoot deformity. In the advanced stages, a Pes Planus deformity can be seen, becoming the hallmark of MWD. Weightbearing radiographic studies are sufficient to assess the diagnosis of flat feet deformity with an increase in density and a medial and dorsal protrusion of the navicular bone often with fragmentation.

Maceira et al described 5 radiological stages of MWD in relation to the sagittal plane of deformation of the navicular bone and based on the orientation of the intersection line between the talar axis and the first metatarsal axis. This was called the "Meary-



Tomeno's angle".

seen in a foot with Mueller-Weiss Disease

A lateral weightbearing view of a foot with MWD showing severe sclerosis and fragmentation of the navicular bone, representing the stage 4 (Maceira classification) showing the Meary-Tomenos's angle.

It is also suggested to treat MWD, in its by early forms with a non-surgical approach using insoles, arch support and orthosis. Reinforce a decreased in sport activities. Others will go to a non-weight bearing status using even a cast for immobilization supplemented with anti-inflammatory medications. If no relief is obtained, an operative mean remains the last option, like te one described above. In advanced cases double or triple fusions

remains to date the best option for a speedy recovery: (TN, TNC). It remains uncertain of treatment is the best choice.

Once diagnosed, the conservative treatment described above is applied. If unsatisfactory, a minimally invasive “Decompression of the Navicular bone” with drilling techniques may take care of the problem but if despite all, the pain persists, and extensive bone destruction is noted, a fusion may be recommended.



This is a radiograph of a Talo-Navicular-Cuneiform (TNC) fusion of the left foot in its Post-operative stage and staples still in place, allowing to stabilize the medial column of the foot. A little what I would expect Rafael Nadal to have as a treatment for his left problematic foot. One can guess how a return to the high professional level of such an athlete can be difficult, allowing him to compete against athletes in top shape and playing matches lasting 4 to 5 hours



under the eyes of millions of telespectators. He must receive injections to ease his moves. We can't imagine what our champion has to go through to keep his standards. At 12-year-old, Nadal had to choose between Football and Tennis and this is when he started experiencing pain and discomfort.



This is now a Triple fusion of the right foot of an other athlete who suffer from advanced MWD.

Multiple fusions to stabilize a foot with Mueller-Weiss Disease...

Pre-operative Radiographic studies will generally reveal a medial extrusion of the navicular in a wedge shaped with loss of height and width of the bone. Remember that the disease can be bilateral as well while both feet can present with deformity and swelling. An arthrodesis involving the talonavicular and the navicular-cuneiform joint or a Triple Fusion are often the last options a surgeon may have to offer such patient, a foot free of

pain. Mueller-Weiss disease, although rare remains a spontaneous avascular necrosis seen during the adulthood still challenging the orthopedist. I wish a lot of luck to my favorite tennis player Rafael Nadal and a prompt recovery and return to the game.

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