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A unique presentation of recurrent cavus foot of an adolescent patient with Marfan syndrome: A case report

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The typical foot and ankle presentation of Marfan syndrome includes joint hypermobility, ligamentous laxity and a pes planus foot type. We present a unique case in which an adolescent patient had recurrent severe cavus foot, claw toe contractures and associated painful metatarsalgia following his original surgical correction eight years previously. This case report describes our surgical approach for definitive deformity correction and relief of the patient's symptoms upon re-evaluation. Osteotomies are the mainstay of treatment of cavus foot deformities in patients age five to skeletal maturity. Such joint sparing procedures may provide substantial relief for a juvenile patient without compromising future surgical options. Joint destructive procedures should be reserved until after skeletal maturity to achieve definitive deformity correction and pain relief. A 17-year-old Ethiopian male presented with severe cavus foot deformity with significant digital contractures secondary to Marfan syndrome. At age nine, the patient underwent numerous joint sparing osteotomies, which were successful in achieving pain relief for eight years. Upon closure of his growth plates, definitive arthrodesis procedures with extensive soft tissue releases were performed. Correction of the cavus foot type and hammered digits deformity was noted. The patient was non-weight bearing for three months. At last follow up, the patient reported improvement in pain scores and ambulatory tolerance. No recurrence in deformity or over-correction was noted. This case study details our treatment of severe recurrent cavus foot-type and digital contractures in a pediatric patient with Marfan syndrome. Utilizing joint sparing procedures prior to skeletal maturity was integral in providing temporary pain relief, while allowing for future surgical options for long term correction once skeletal maturity was reached.

Keywords: Marfan syndrome, cavus foot

Marfan syndrome, first described in 1896, is an inherited connective tissue disorder that affects multiple organ systems. The typical foot and ankle manifestations include joint hypermobility, ligamentous laxity and pes planus [1,2,3].

We present a unique case in which an adolescent patient had recurrent severe cavus foot, claw toe contractures and associated painful metatarsalgia following his original surgical correction eight years previously. This case report describes our surgical approach for definitive deformity correction and relief of the patient's symptoms upon reevaluation.

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Literature Review

Cavus foot type presents with an abnormal elevation of the medial arch with weightbearing. Typically, there is a component of forefoot pronation, which involves first metatarsal plantarflexion relative to the hindfoot. Cavovarus and calcaneocavus are most common [4,5,6].

When assessing these patients, a thorough neurologic evaluation is mandatory as the deformity is often associated with an underlying spinal cord or neuromuscular disorder responsible for intrinsic and extrinsic muscle imbalance [4,5,6]. The deformity can be progressive or caused by paralytic soft tissue disease, osteoarticular disease or trauma [5]. A systematic approach should be utilized during evaluation (Table 1). Anterior posterior and lateral standing foot views should also be obtained [4].

Is there a concomitant neurological disorder?
Is the deformity unilateral or bilateral?
What is the hindfoot position (i.e. cavovarus, calcaneocavus or cavovalgus)?
Are joints rigid or supple?
Where is the apex of deformity?
What is muscles strength in all lower extremity quadrants?
Are there neurologic changes?
Any digital deformities?

Table 1 Clinical Evaluation of Cavus Foot Deformity [7].

The goals of treatment should be to create a plantigrade, pain-free, stable, balanced foot. Conservative management includes physical therapy, casting in progressive abduction, insoles and customized shoes [5]. Should conservative measures fail, surgical options include plantar fascia release for flexible deformity, tendon transfers to restore muscle balance and osteotomies for fixed deformities [4,5,6]. Osteotomies are the mainstay of treatment of rigid cavus foot deformities in patients age five to skeletal maturity and should be performed at the apex of deformity. This is determined by the intersection of talar and first metatarsal lines on a lateral weight bearing radiograph [5,8]. Such joint sparing procedures may provide substantial relief for a juvenile patient without compromising future surgical options.

In rigid, severe deformities that have failed previous joint sparing procedures, tarsectomies, midfoot osteotomies and arthrodesis procedures can be performed [4,5]. The goal of arthrodesis is to achieve definitive deformity correction and pain relief with minimal foot architecture disruption. Arthrodesis procedures are contraindicated prior to growth plate closure and should be used with caution for patients with sensory loss [4].

Case Report

A 17-year-old Ethiopian male presented with severe bilateral cavus foot deformity, secondary to Marfan Syndrome (right foot worse than left) (Figure 1). Additional past medical history included myopia, lens dislocation, hypermobile joints, dilated aortic root and mitral valve prolapse with mild insufficiency. Past lower extremity surgical history significant for previous right foot reconstruction at age nine. Procedures included: gastrocnemius lengthening, Dwyer calcaneal osteotomy, cuboid closing osteotomy, medial cuneiform plantar based opening wedge osteotomy, V to Y skin-plasty and capsulotomy with pinning of the 4th and 5th metatarsophalangeal joints. The original surgery provided eight years of relief; however, due to the nature of his connective tissue disorder, the foot deformity recurred and progressed.

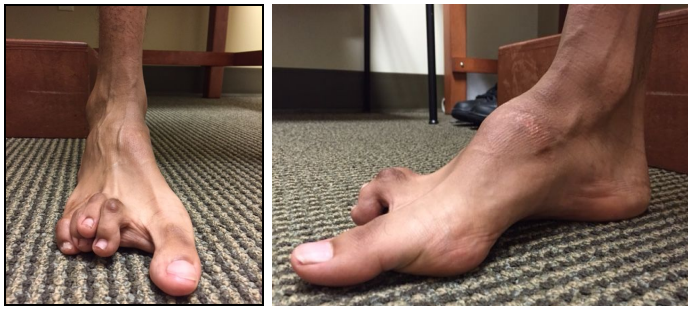

Figure 1 Preoperative weightbearing clinical appearance of right foot cavus deformity with significant digital contractures.

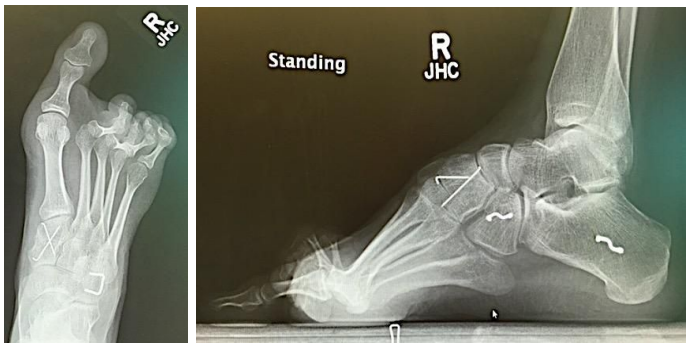

Figure 2 Preoperative anterior posterior and lateral weightbearing radiographs revealed global cavus deformity, increased calcaneal inclination angle, increased Meary's angle and dislocated second, third and fourth metatarsophalangeal joints. Retained deep hardware also noted from the index procedure.

At the time of presentation, the patient exhibited severe, rigid anterior cavus foot deformity with significant digital contractures bilaterally (right foot worse than left). Calluses along the metatarsal heads, metatarsalgia and an ataxic gait were also noted (Figure 1). Sensation was intact bilaterally. Radiographs revealed global cavus deformity, increased calcaneal inclination angle, increased Meary's angle and dislocated second, third and fourth metatarsophalangeal joints. Retained deep hardware also noted from the index procedure (Figure 2).

Due to the fact his growth plates were fused, the decision was made to perform definitive arthrodesis procedures with extensive soft tissue releases and capsulotendon rebalancing. The patient underwent a triple arthrodesis, first tarsometatarsal joint arthrodesis, peroneus longus to peroneus brevis transfer, lengthening of the extensor hallucis longus, 2nd and 3rd proximal interphalangeal joint arthrodesis, 4th proximal interphalangeal joint reduction with pin fixation and 1st, 2nd, 3rd, 4th and 5th metatarsophalangeal joint tenotomies and capsulotomies with pinning (Figure 3).

Results

Correction of the cavus foot type and hammered digits was noted immediately postoperatively (Figure 3). Pins were removed at 6 weeks. The patient was non-weight bearing for 13 weeks. He completed 8 weeks of physical therapy for strength and stability training. At 17 weeks postoperatively, regular shoe wear was resumed.

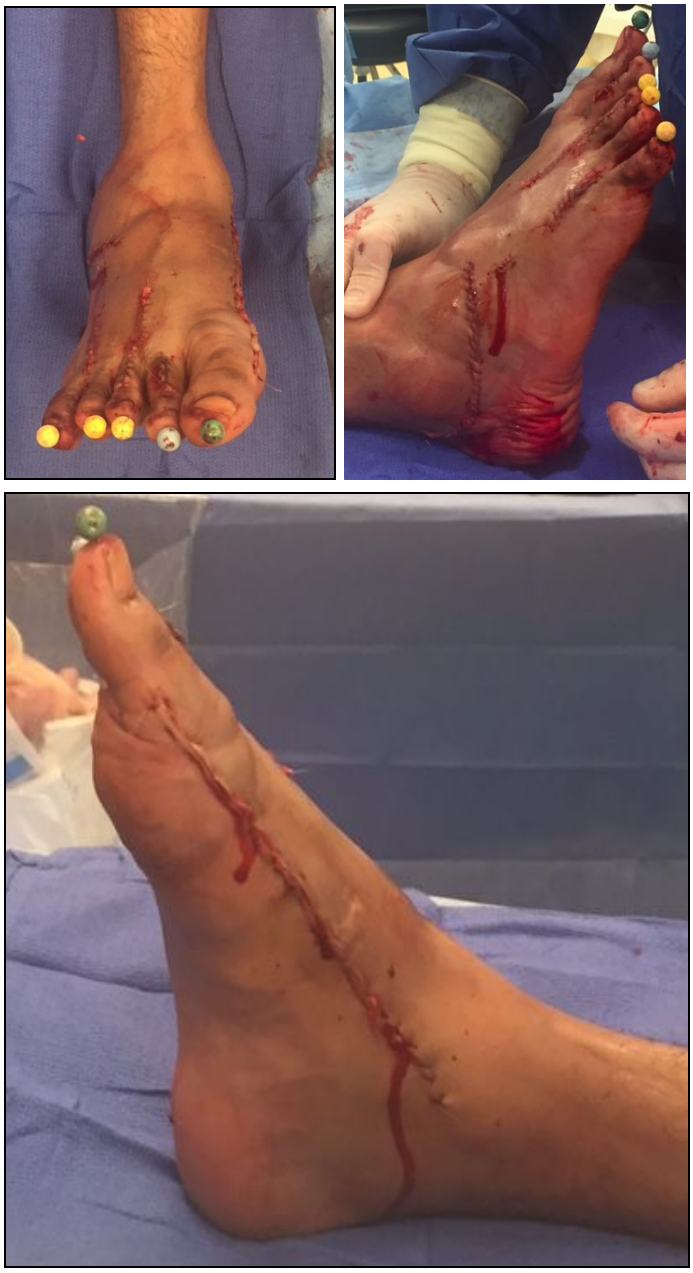

Figure 3 Correction of the cavus foot type and digital contractures was noted immediately postoperatively.

Plain films revealed a rectus foot, decreased Meary's angle and excellent bony consolidation along the arthrodesis sites (Figure 4).

At last follow-up, the patient reported improvement in pain scores and ambulatory tolerance. His severe plantar forefoot calluses had completely resolved (Figure 5). No recurrence in deformity or over-correction was noted.

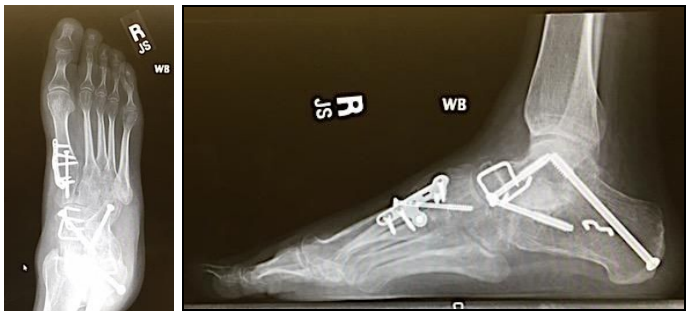

Figure 4 Correction of the cavus foot type and digital contractures was noted immediately postoperatively.

Discussion

To our knowledge this is the first case report of recurrent cavus foot deformity in an adolescent with Marfan syndrome. Cavus deformity is atypical for this connective tissue disorder, which is commonly associated with pes planus.

Prior to initial surgical intervention at age nine, the patient was seen by a neurologist to rule out a concurrent underlying neuromuscular disorder, due to his atypical presentation. The original foot reconstructive surgery included osteotomies and soft tissue rebalancing as to not violate the open growth plates.

At skeletal maturity, definitive procedures became feasible and arthrodeses were chosen. In the hindfoot, a triple arthrodesis was chosen over tarsectomy or midfoot osteotomy as the patient was ambulatory and bony architecture disruption was to be kept at a minimum. The use of the arthrodesis also ameliorates the risk of future hindfoot arthritis as seen with other salvage procedures. In the midfoot, a Lapidus procedure was chosen for definitive correction of the plantarflexed first ray as the apex of his cavus deformity was at the first tarsometatarsal junction. This procedure was supplemented by peroneus longus to peroneus brevis tendon transfer. Capsulotendon balancing of the forefoot with interphalangeal fusions of the 2nd and 3rd digits were performed to allow for toe purchase during ambulation. This decision was carefully considered due to the potential for neurovascular compromise and digital ischemia.

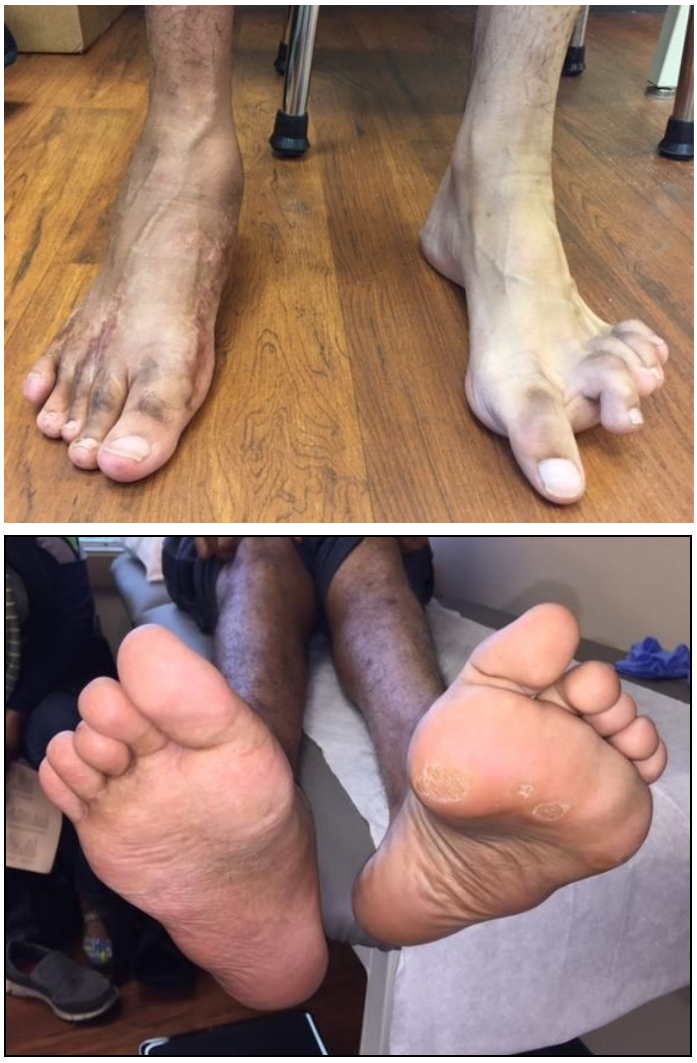

Figure 5 Seventeen weeks postoperative clinical appearance: Severe plantar forefoot callusing had completely resolved and no recurrence in deformity or over-correction was noted.

In conclusion, utilizing joint sparing procedures prior to skeletal maturity in this case study was integral in providing temporary pain relief, while allowing for future definitive surgical options for long term correction once skeletal maturity was reached.

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