Consultant360 Multidisciplinary Medical Information Network

PHOTOCLINIC

PEER REVIEWED

Congenital Vertical Talus

Authors:
Tolulope F. Obafemi, BS; Sam N. Russo, MD; Elizabeth Becker, MD; and Lynnette J. Mazur, MD, MPH
Department of Pediatrics, McGovern Medical School at UT Health, Houston, Texas

Citation:
Obafemi TF, Russo SN, Becker E, Mazur LJ. Congenital vertical talus [published online August 12, 2019]. Consultant360.

A full-term boy was born by spontaneous vaginal delivery to a 37-year-old gravida 3, para 3 mother. His birth weight, length, and head circumference were 2885 g (16th percentile), 49 cm (32nd percentile), and 32 cm (third percentile), respectively.

At birth, physical examination findings were unremarkable except for an undescended left testicle and an everted, flat left foot (**Figures 1 and 2**). The right foot was normal in appearance, but crepitus was felt and heard in both feet. There was no evidence of flexion contractures, other joint abnormalities, or an underlying neuromuscular disease. The family history was negative for similarly affected individuals. The neonate passed screening tests for heart and hearing defects. Radiographs of the left ankle showed vertical orientation of the talus (**Figure 3**).

Figure 1. *Lateral view demonstrating distinct dorsiflexion of the left midfoot and forefoot with hindfoot equinovarus.*

Figure 2. *Anterior view demonstrating distinct dorsiflexion of the left midfoot and forefoot with hindfoot equinovarus.*

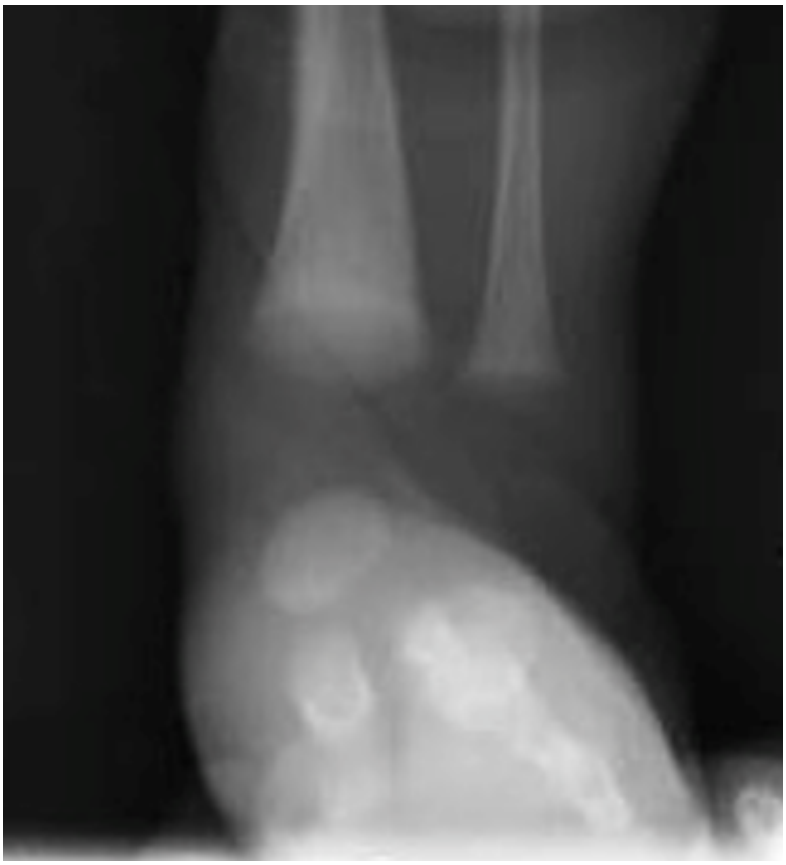

Figure 3. *Lateral plantar flexion radiograph demonstrating abnormal vertical orientation of the talus..*

Discussion. Congenital vertical talus (CVT), also known as rocker-bottom foot or congenital convex pes valgus, occurs in 1 in 10,000 infants, is bilateral in 50% of cases, and has a 1-to-1 male-to-female ratio.[1,2] It is characterized by a rigid irreducible dorsolateral dislocation of the talonavicular joint, in which the talus becomes vertically aligned.[1,2] In addition to the dislocation, soft tissue contractures commonly occur in the Achilles and extensor digitorum longus tendons.[1,2]

The differential diagnosis of CVT includes oblique talus, calcaneovalgus, pes planus (flat foot), and talipes equinovarus (clubfoot) (**Table 1**).[3,4] Differentiation can be achieved via thorough physical examination and imaging, the interpretation of which will determine whether surgical correction is required. Magnetic resonance imaging can be useful to rule out an underlying neurological disorder.[5]

Table 1. Differential Diagnoses of Congenital Vertical Talus

Congenital Deformity	Examination Findings	Radiography Findings
Congenital vertical talus	Rigid hindfoot equinovalgus and dorsiflexed and abducted forefoot —"rocker-bottom deformity"	Vertically positioned talus and irreducible dorsolateral dislocation of talonavicular joint; talus plantarflexion

Oblique talus	Comparable findings to congenital vertical talus	Oblique talus, talonavicular subluxation, talonavicular joint can be reduced with forced plantarflexion
Calcaneovalgus	Hindfoot in excessive dorsiflexion, dorsum often touches distal leg, flexible deformity, plantar flexion normal	Can appear normal, first metatarsal aligned with talus, nondislocated talonavicular joint
Talipes equinovarus	Shortened tibia, hindfoot in equinovarus, forefoot adduction, variable rigidity	Talocalcaneal angle <25°, metatarsal base convergence
Pes planus	Collapse of longitudinal foot arch, hindfoot valgus, forefoot abduction	Normal heel, parallel metatarsals, talonavicular joint sag, increased talocalcaneal angle

Approximately half of CVT cases have associated neuromuscular or chromosomal abnormalities; the other half are idiopathic or nonsyndromic (**Table 2**).[6]

Table 2. Clinical Associations of Congenital Vertical Talus

Central nervous system	Myelomeningocele, spina bifida, spinal muscular atrophy, caudal regression syndrome, hydrocephalus
Chromosomal abnormalities	Trisomy 13, trisomy 18, trisomy 21
Genetic syndromes	Prune-belly syndrome, Costello syndrome, De Barsy syndrome, Rasmussen syndrome
Muscular abnormalities	Distal arthrogryposis, multiple pterygium syndrome, neurofibromatosis
Single-gene defects	*HOXD10*, *GDF5* (*CDMP1*)

In isolated CVT, a positive family history occurs in 12% to 20% of patients.[7] In families with an autosomal dominant inheritance pattern, incomplete penetrance has been detected.[7] Although the etiology of the disease unknown, it is likely multifactorial. One study of a family with complete penetrance of CVT found a single missense mutation in *HOXD10* in each family member, while another study in patients with sporadic CVT did not show *HOX* mutations.[8,9] Another study linked a *GDF5* (formerly *CDMP1*) mutation to deformities in the hands and feet, including a child with isolated CVT.[10] Genetic testing for children with multiple anomalies is recommended by the American Academy of Pediatrics.[11] Testing with chromosomal microarray analysis has a higher diagnostic yield (15%-20%) than conventional G-banding karyotype

analysis (3%).[12,13] However, because our patient had an isolated deformity, no genetic testing was deemed necessary.

If left untreated, CVT may result in painful calluses, skin breakdown, foot pain that affects the child's gait, and difficulty in finding proper footwear.[14] Treatment options include casting and surgical correction.[7] Traditionally, children up to the age of 3 years are offered 1- or 2-stage open reduction of the talonavicular joint via release of tendons and ligaments of the foot.[7] When performed between 9 and 12 months of age, the long-term prognosis is improved. After the age of 3 years, a more permanent deformity persists; these children are usually treated with open reduction and arthrodesis.[7]

Complications after surgical procedures include degenerative arthritis and avascular necrosis of the talus in adulthood.[7] To mitigate surgical complications, a minimally invasive method—the Dobbs method—has been proposed, involving serial casting followed by a percutaneous Achilles tenotomy and limited open reduction of the talonavicular joint.[7,15] This method is similar to the Ponseti method for correction of clubfoot with serial casting.[7,15] For CVT, approximately 4 to 6 casts are needed. Casts are changed weekly to achieve adequate talonavicular reduction prior to percutaneous surgery.[7,15] Following the operation, a dynamic shoe-bar cast is worn 23 hours a day for the first 3 months of life and then 12 to 14 hours a day (naps and nighttime) for 2 more years.[7] Stretching exercises with every diaper change also improve outcomes.[7]

Our patient's parents opted for serial casting of the foot, and the boy was referred to orthopedics specialists.

References

1. Miller M, Dobbs MB. Congenital vertical talus: etiology and management. *J Am Acad Orthop Surg.* 2015;23(10):604-61
2. Hart ES, Grottkau BE, Rebello GN, Albright MB. The newborn foot: diagnosis and management of common conditions. *Orthop Nurs.* 2005;24(5):313-321.
3. Harris EJ. The oblique talus deformity: what is it, and what is its clinical significance in the scheme of pronatory deformities? *Clin Podiatr Med Surg.* 2000;17(3):419-442.
4. Becker-Andersen H, Reimann I. Congenital vertical talus: reevaluation of early manipulative treatment. *Acta Orthop Scand*. 1974;45(1):130-14
5. Thometz JG, Zhu H, Liu X-C, Tassone C, Gabriel SR. MRI pathoanatomy study of congenital vertical talus. *J Pediatr Orthop.* 2010;30(5):460-464.
6. Ogata K, Schoenecker PL, Sheridan JJ. Congenital vertical talus and its familial occurrence: an analysis of 36 patients. *Clin Orthop Relat Res.* 1979(139):128-132.
7. Alaee F, Boehm S, Dobbs MB. A new approach to the treatment of congenital vertical talus. *J Child Orthop.* 2007;1(3):165-174.
8. Shrimpton AE, Levinsohn EM, Yozawitz JM, et al. A *HOX* gene mutation in a family with isolated congenital vertical talus and Charcot-Marie-Tooth disease. *Am J Hum Genet.* 2004;75(1):92-96.
9. Gurnett CA, Keppel C, Bick J, Bowcock AM, Dobbs MB. Absence of HOXD10 mutations in

idiopathic clubfoot and sporadic vertical talus. *Clin Orthop Relat Res.* 2007;462:27-31.
10. Dobbs MB, Gurnett CA, Robarge J, Gordon JE, Morcuende JA, Bowcock AM. Variable hand and foot abnormalities in family with congenital vertical talus and CDMP-1 gene mutation. *J Orthop Res.* 2005;23(6):1490-1494.
11. Committee on Bioethics; Committee on Genetics; American College of Medical Genetics and Genomics Social, Ethical, and Legal Issues Committee. Ethical and policy issues in genetic testing and screening of children. *Pediatrics.* 2013;131(3):620-622.
12. Ellison JW, Ravnan JB, Rosenfeld JA, et al. Clinical utility of chromosomal microarray analysis. *Pediatrics.* 2012;130(5):e1085-e1095.
13. Miller DT, Adam MP, Aradhya S, et al. Consensus statement: chromosomal microarray is a first-tier clinical diagnostic test for individuals with developmental disabilities or congenital anomalies. *Am J Hum Genet.* 2010;86(5):749-764.
14. Lui TH. Correction of neglected vertical talus deformity in an adult. *BMJ Case Rep.* 2015;2015:bcr2014208187.
15. Dobbs MB, Purcell DB, Nunley R, Morcuende JA. Early results of a new method of treatment for idiopathic congenital vertical talus. *J Bone Joint Surg Am.* 2006;88(6):1192-1200.