Common Anomalies Associated To Congenital Vertical Talus: A Single Center Experience

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Abstract

Background: Congenital vertical talus is defined as a foot deformity in which the calcaneus is in equinus, the talus is plantarflexed, and there is a rigid and irreducible dislocation of the talonavicular joint complex, with the navicular articulating on the dorsolateral aspect of the talar neck. It is often associated with systemic involvement.

Aims: To identify the most common anomalies accompanying CVT (Congenital Vertical Talus). No literature investigating similar clinical data was found in the literature review.

Study design: CVT has a systemic effect and is accompanied by many anomalies. At the same time as this study, anomalies were frequently found accompanying CVT. The aim of this study is to evaluate the prenatal, natal and postnatal clinical findings associated with Congenital Vertical Talus.

Methods: Between 2012 and 2018, a total of 20 CVT patients (12 males, 8 females) were included in the study. Children between the ages of 0 and 18 who applied to the dysmorphology clinic for the study were included.

Results: Five parameters associated with CVT were detected in this study. These include intrauterine growth retardation, hypoxic birth history, hypotonia, microcephaly and cryptorchidism.

Conclusion: A detailed and multidisciplinary approach to CVT is required.

Keywords

Microcephaly, Vertical talus, Intrauterine growth retardation

Introduction

Congenital vertical talus is rare. The first clinical, anatomical, pathological and roentgenographic study of this condition was published by Henken, in 1914. A congenital vertical talus, also known as congenital convex pes valgus, is a rare foot deformity characterized by a fixed dorsal dislocation in talar head and neck navicular. The incidence is estimated to be one in 10,000. Approximately half of all cases (idiopathic) are associated with deformity and 2-5 neuromuscular and genetic disorders in the remaining cases. There is evidence that some isolated deformities are transmitted as an autosomal dominant feature with incomplete penetrance [1-4]. The deformity is also known by congenital “rocker-bottom” flatfoot because of its rigid deformity with the forefoot dorsiflexed and the hindfoot plantarflexed. The term “congenital convex pes valgus” is also frequently used. To make a definite diagnosis, it is important to demonstrate that the navicular is dislocated dorsally on the neck of the talus when the foot is maintained in extreme plantar flexion [5,6]. Congenital vertical talus is characterized by hindfoot equinus, hindfoot valgus, forefoot abduction, and forefoot dorsiflexion at the midtarsal joint. This is usually recognized in the newborn period by the rigidity of the deformities, but it must be differentiated from the more common calcaneovalgus foot, posterior medial bowing of the tibia, and flexible flatfoot. In congenital vertical talus, the plantar surface of the foot is convex creating a rocker-bottom appearance [7].

Dysmorphology allows identification of occurred before or after birth non-normative human forms that enable the identification and classification of various congenital malformations. This term provides an comparable description that all body characteristics (stature, feet, hands, neck) and face (like shape of head, nose length, position of ears, thickness of vermillion, etc.) of individuals who the same age group and same ethnicity. A genetic etiology should be suspected if a child has dysmorphic vision with one of these features: (a) Congenital anomalies; (b) Growth retardation; (c) De-
Cryptorchidism is the absence of at least one testis in the scrotum (Figure 1). The incidence of cryptorchidism is high and almost 3% of boys in the western countries are operated on for this condition. Abnormalities in the hypothalamo-pituitary-testicular axis may result in cryptorchidism, and cryptorchidism is nearly always present in individuals with a testis and abnormal sexual differentiation [15].

The presence of a hypoxic birth story in the postnatal period was investigated. For this purpose, the birth APGAR score of the patients was investigated. At the same time, these patients were followed for a long time in medical genetic clinic. Especially whether hypotonia was present or not. The times of motor development, language development and social development stages were investigated.

The APGAR score used for fetal well-being is another parameter we questioned. This score includes color, heart rate, reflexes, muscle tone and respiration. There are 3 categories according to the results of the APGAR score; low (0-3); intermediate (4-6) and normal (7-10). Low Apgar scores at birth are consistently associated with increased risk of neurological disease, such as cerebral palsy, epilepsy and cognitive impairment [16,17].

Materials and Methods

Between 2012 and 2018, a total of 20 CVT patients (12 males, 8 females) were included in the study. Hospital records of the patients were reviewed in detail and the anomalies associated with CVT were identified. The records of the patients from the Prenatal period were scanned. Here, fetal characteristics were investigated in prenatal period. Fetal movements, oligohydramnios, polyhydramnios, intrauterine growth retardation. In addition, maternal complications such as hypertension, diabetes, preeclampsia were investigated.

Three parameters related to Natal turnover of the patients were investigated. Birth style, birth weight and birth time. If the cesarean birth type was preferred, the reason for this was investigated.

Collected data were analyzed by Statistical Package for Social Sciences version 18.0 (SPSS Inc., SPSS IBM, Armonk, NY, USA). Continuous data were expressed as mean ± standard deviation (range: minimum-maximum) whereas categorical data were denoted as numbers or percentages where appropriate. Chi-square test was used for the statistical comparisons. Two-tailed p values less than 0.05 were accepted to be statistically significant.

Results

In the study, the most common comorbidities were found in CVT patients.
Anomalies in CVT patients can be divided into 2 major categories. These are: central nervous system diseases and neuromuscular diseases (the frequency of both was found equal).

There are very few publications in the literature reporting relation cryptorchidism with CVT. Vertical Talus and cryptorchidism are common in multiple pterygium syndrome cases [19,20].

In a study by Merrill and colleagues, 36% of patients with CVT had neuromuscular disease (Such as Cerebral palsy, Myelomeningocel, Caudal regression syndrome, Hydrocephalus) [21]. Fetal brain abnormalities are the most common finding associated with congenital vertical talus, occurring in 63% of our cases. Abnormal conditions were usually severe and included microcephaly, migration anomalies, cerebrum, cerebellum and cerebellum underdevelopment in addition to myelomeningocele-associated Chiari II malformation. While useful in assessing USG at head growth, MRI has found an important help in identifying brain abnormalities, particularly migration and sulcation anomalies [22].

46% of patients at this study have spinal abnormalities such as scoliosis, myelomeningocele, and tethered cord. Although spinal column malformations can be easily seen by the USG, cord and conus position are better evaluated with MRI [22].

It was associated with CVT and trisomy 13, 15 and 18. 25% of the patients in the study of Rubio, et al. had chromosomal abnormalities that is trisomy 18 (most common chromosomal anomaly in this study.). This is in keeping with prior studies describing an association of rocker bottom foot and aneuploidy of trisomy 13, 15 and 18 [7,22]. In our study, the chromosomal anomalies were the trisomy 18 that we detected at the most common. We found trisomy 18 in 6 of 20 patients in total (30%).

Discussion
We haven’t been found any article in the literature investigating similar clinical data. Therefore, it is difficult to compare the data of our patients with the literature.

No information was found in the literature about intrauterine growth retardation in CVT cases. In the study conducted by Romo, et al. in 2009, the incidence of IUGR in the general population was found between 3% and 7% [18]. Compared to the general population and patients with CVT, the IUGR rate was much higher (80%) at have CVT patients.

According to our results in this study, neurological anomalies in CVT patients can be divided into 2 major categories. These are: central nervous system diseases and neuromuscular diseases (the frequency of both was found equal).

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Conclusion
In this study, 5 parameters were found to be fre-
quently associated with CVT cases. These parameters; intrauterine growth retardation, hypoxic birth history, hypotonia, microcephaly and cryptorchidism. Vertical talus is a multi-systemic disease and multidisciplinary approach is needed. Vertical talus cases should be investigated in detail for prenatal, natal and postnatal period findings.

References


