A Tale of Two Pathologies:
A New Case Study on Congenital Talipes Equinovarus (CTEV)

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CTEV is a worldwide problem. There is no nation or culture on earth that it has not been touched by this defect. Fortunately, children born with CTEV in North America are extremely fortunate as treatment for the condition is begun soon after birth. This is not so in the rest of the world, including much of Europe.

While not all cases of CTEV have one hundred percent success rates in the repair and management of malformed feet and ankles, it is rare that you see anyone under the age of fifty walking around in public with abnormal gait caused by CTEV. There are varying degrees of successful outcomes in managing this condition. The vast majority of less than perfect outcomes are hardly noticeable in society. Prior to the current mode of treatment, the fate of Albert, mentioned in part one of this series, was more the rule. Today, with advanced modes of management, the outcomes are much better, though complicated cases such as Tommy’s remain in very small numbers.

Skateboards have been around as a mode of pre-motor vehicle transportation for children and teens for roughly sixty years. Once the transit mode for beatniks and boardwalk beachcombers, it is used as a substitute for a wheelchair in many European countries. Just down the avenue from the historic Pantheon in Rome is the Basilica of Santa Maria Sopra Minerva. It is the church at the headquarters of the Dominican religious order and is built upon the site of the ancient Roman Temple to the goddess Minerva.

As you approach the entry door, a teenage girl opens it for you. She is sitting on a skateboard and is seeking donations to supplement her income. Bending down to make the donation, you notice something strange – misshapen feet and ankles. She has CTEV. She cannot walk. Her condition went untreated at birth.

Walking through the streets of the Eternal City, there are several skateboarding teens, all with CTEV. This is something that would never be seen in the United States. Even with nationalized healthcare for everyone in Italy, there are far too many that don’t get treatment for whatever reason.

CTEV occurs in anywhere from 1 to 2.29 cases per 1000 live births. The number is higher in less developed countries. Males outnumber females 2 to 1. In unilateral cases, the right foot is more affected. The incidence rates rise with family history. Siblings have a 1 in 35 chance with twins having a 1 in 3 chance. The etiology remains largely unknown, though there are several theories:
A TALE OF TWO PATHOLOGIES: A NEW CASE STUDY ON CONGENITAL TALIPES EQUINOVARUS (CTEV) (PART 2)

1. Mechanical factors in utero. Hippocrates believed that external uterine pressure caused the foot to be held in equinovarus during gestation. Parker (1824) and Browne (1939) held that a lack of amniotic fluid prevents fetal movement, making the fetus subject to external pressure.

2. Neuromuscular defect. The literature has approximately fifty percent on either side of the possibility.

3. Primary Germ Plasm Effect. During early stages of development at the base cell level, a marker is turned on to allow CTEV to occur. In a dissection of feet with CTEV, the talar neck is always short, anteriorly rotated medially and planterly.

4. Arrested fetal development. Intrauterine and environmental causes (e.g. rubella or use of thalidomide during pregnancy).

5. Heredity. The pathoanatomy shows gross changes in shape and position of the navicular, cuboid and calcaneus. Tendons, tendon sheaths, ligaments and fascia have adaptive changes that become fibrotic or contracted. The talocalcanealcuboid articulation is subluxated.

Pathogenesis: During embryonic development, the foot passes through three phases:

1. Initial position - foot is in straight line with the leg.
2. Embryonic phase - foot is in marked equinovarus adductus position.
3. Fetal phase - foot changes to a slight equinovarus adductus position (this phase is reached by 11 weeks gestational age).

Any interference with stages 1 to 3 can result in clubfoot. The severity of the clubfoot is dependent on the evolutionary stage at which the interference occurred. Many theories exist with these being the most plausible.

1. The theory of arrest of fetal development in the fibular phase is based on the phases of embryologic development. However deviation of the talus (as found on dissections) is thought to be the primary cause of clubfoot, because it is thought to result from a primary germ plasm defect.

2. Some investigators have found innervation changes in the muscles of clubfoot patients and suspect that these changes are the primary cause of this malformation.

3. Other investigators have found excessive amounts of fibrous tissue and thought that retracting fibrosis may be the primary cause of the clubfoot malformations.

4. Some authors have examined the contracted medial side and have suggested that the contracture of myofibroblast-like cells may have been enhanced by histamine release from the mast cells, which are also found in increased concentrations.

5. Anomalous tendinous insertions of the Achilles tendon, tibialis anterior, and/or the peroneal tendons have also been blamed as the primary cause.

Dimaggio (1991) described his classification system as having four categories based on joint motion and ability to reduce deformities.

1. Soft foot, aka postural, treatment with conservative methods and physical therapy.
2. Soft > Stiff. More than 50 percent reducible, initially responding to casting.
3. Surgery indicated if total correction not achieved in 7-8 months.
5. Stiff foot. Poorly reducible with severe equinus position of the calcaneus. Often bilateral and correction with surgery indicated.

Overall, 50 to 90 percent success rates are reported with conservative measures.

A 1983 study came to a similar conclusion offering three classifications: mild, moderate and severe with a 1985 study that described the condition as severe, resistant, mild and postural. This same study found that a calcaneus high in the heel pad of fat at 6 weeks was an early indicator for surgery.

Another indicator of the severity is the measurement of calf muscle mass and the presence of an accessory tendon. In conservatively managed cases, there was little difference with the normal side in unilateral cases, where in surgical groups, it was highly significant. By the age of four, unilateral feet were essentially the same. The operative group showed distinct differences in the range of motion in joint anatomy.

Results show a distinction between resistant and resolving CTEV. In the conservatively treated group, those without abnormal calf muscle measurements had normal anatomy and ankle range of motion. In the surgical group, the reduction of calf muscle mass at six weeks presents as an intrinsic structural problem. Even though the surgery generally corrected the deformity, the range of motion at the ankle was reduced and joint anatomy was reduced.

It was the opinion of this study team that resolving deformities, those generally corrected with conservative treatment alone, should be excluded from the definition of CTEV and left to those with intrinsic structural problems. The thought process is that resolving cases show favorable biases in skewing the success numbers as these cases resolve with conservative treatment alone.

The inability of researchers to establish a common standard as to what CTEV is makes for confusion in describing treatment modalities and success rates.

In a Swedish study published in 1992, 75 children’s feet were studied over a period of 6-11 years. 47 were treated conservatively beginning at 2 weeks of age. Three were treated with surgical intervention at 2-5 months. Physical therapy and bracing were
used for three years. 27 feet had multiple surgical procedures over the years. This study was concerned with cosmesis, functional, radiographic and surgical outcomes.

The protocol is rather aggressive, consisting of daily manipulation by physical therapy that is followed up by the parents later in the day resulting in two sessions daily. An AFO with a medial flange extending up and over the hallux. The flange extends superiorly to the medial malleolus, ending at the calcaneus.

The device is held in place in a straight lasted shoe similar to one used in a Denis-Browne splint. A surgical evaluation was performed in the 2-5 months of age range. Post op (soft tissue releases) was followed by placement in a cast for five weeks.

Once out of the cast, a dynamic splint was used. Physical therapy was performed twice daily for six months, then bi-weekly for one year, then at a 4-6 week interval. An anti-varus shoe was used when the child was walking. Active therapy ceased at three years of age.

Follow-up appointments were twice yearly until the child reached the age of seven. At the mean age of eight (actual ages 6-11) the cosmetic, functional and radiographic evaluations began.

Cosmetic result was considered successful with a plantargrade foot and absence of forefoot adduction. Acceptable results was a plantargrade foot with mild residual deformity. Poor result was significant residual deformities or overcorrection. Results: Good (62) Acceptable (12) and poor (1).

Functional assessment scale is based on activity level with no limitations, absence of pain and neutral heel position, 10 degrees or more ankle dorsiflexion, satisfactory subtalar joint motion and normal gait. Results: Excellent (51) Good (21) and Fair (3). None had functional failures.

Radiograph (X-Ray) studies measured various angles in the alignment of the bones and articulations.

Surgical: 67 of 75 feet studied received surgical intervention. Procedures include soft tissue releases, tendon transfers and osteotomy. No complications were reported.

Conclusions: Within the medical universe, there is no standard description of CTEV. This is one of those “you know it when you see it” pathologies. The real issue is causality.

There certainly are statistics that show genetics plays a role, as CTEV can run in families. It is doubtful that a single cause will be described any time soon. One critical difference in the literature is the use of the term CTEV.

It is clear that the vast majority of the cases are resolved with the standard conservative treatment: manipulation, serial casting, splinting. This type of presentation is described as “postural”. Some authors seek to reserve the term CTEV for the resistant type that require moderate to significant surgical intervention and follow-up care, including pedorthic modalities.

Scientifically, both postural and complicated versions qualify as the foot presents as talipes equinovarus, and is obviously congenital. Perhaps some resolution may come to the definition issue. How about Type 1 and Type 2? Once the medical and research community reach a conclusion about the description, the research on the cause(s) can move forward in earnest.

The next installment in the series will focus on the ‘Role of Genetics and Ancillary Conditions to CTEV’.

References: