Congenital talipes equinovarus

Abstract

Congenital talipes equinovarus, more commonly known as clubfoot, is an idiopathic congenital birth defect that affects in 1 in 1000 births.¹ The most widely used and well known treatment is the Ponseti method commonly followed by an Achilles tenotomy. This method is effective on roughly 90% of all cases. The severity of this idiopathic birth defect can be scored using a few different methods. The most common is the Pinar scoring system which is a simple system that is easily learned and implemented. The Pinar scoring system can also be supplemented by the use of ultrasound and Magnetic Resonance Imaging (MRI) to visualize the non-ossified bones and the surrounding soft tissue. Ultrasound can also show the stages of healing the Achilles tendon after the Achilles tenotomy. MRI has also been used to show the effect of congenital talipes equinovarus along all stages of development, from fetus to adulthood.

Introduction

Clubfoot is a defect that arises during fetal development. The origins of this birth defect at this point in time are unknown.² “A normally developing foot turns into a clubfoot during the second trimester of pregnancy… Therefore, like developmental hip dysplasia and idiopathic scoliosis, clubfoot is a developmental deformation”² (p.4) There are two common methods to correct the birth defect: the Ponseti method and surgically using a comprehensive clubfoot release. The Ponseti method is the preferred method for correcting clubfoot with up to a 90% success rate.³ It is a noninvasive method that utilizes manipulation of the foot through casting to abduct the foot to a normal position.⁴ The deciding factor for which treatment is utilized depends on the severity of the birth defect. Weight and age may also affect the advantages of the Ponseti method.⁴ There are several systems to decide the severity of a clubfoot; the most common scoring system used to determine the severity of the deformity is the Pirani score.⁴-⁶ This scoring system is widely used because it is reliable, easy to memorize, and quick. Imaging can also be used to see the relations of the bones. The modality that can offer a better view is ultrasound due to its ability to see non-ossified bone without the need for sedation. “The validity of [the Pirani scoring system] could be reinforced if supplemented by sonographic assessment, before and after
treatment, to confirm return of normal anatomic relationships between the different bones.\textsuperscript{6}(p. 2496) MRI has also been used to see the long term effect of talipes equinovarus.

**Ponseti method**

The Ponseti method is a noninvasive method to correct clubfoot. This treatment was first introduced in the early 1960s by Ignacio V. Ponseti and Eugene N. Smoley.\textsuperscript{7} Until this treatment was introduced, the only other option was invasive surgery. According to Ponseti “The early months of life offer a golden opportunity for the correction of club feet since the skeleton, which is to a great extent cartilaginous, is little deformed, and the joint capsules, ligaments, tendons, and muscles can be stretched without damage.” \textsuperscript{7}(p. 1141) The Ponseti method utilizes manipulation of the foot through casting to abduct the foot to a normal position. The severity of the defect is most commonly scored using the Pirani score. The Pirani score is the most common scale to measure the severity of congenital talipes equinovarus. This scoring system uses “…six clinical signs to quantify severity of each component of the deformity. Each component is scored as 0 (normal), 0.5 (mildly abnormal) or 1 (severely abnormal).” \textsuperscript{2}(p. 27) This scoring system uses the different views to help visualize the issues in the underling soft tissue and bony anatomy (see Figure 1). The casting, if needed, is followed by a heel cord release commonly known as Achilles tenotomy. This treatment has a success rate to avoid surgery by 90%. This method possibly can achieve higher numbers if the patient strictly adheres to the prescribed schedule for wearing the braces followed by casting. The treatment is recommended to start as early as the first week after birth.\textsuperscript{4} A study done by Awang et al\textsuperscript{4} compared the effectiveness of the treatment with the variables of the child’s age, weight, and Pirani score. This study was done to try to find the best time to start the treatment and the effectiveness of the treatment with the above factors.

The study conducted by Awang et al\textsuperscript{4} looked at infants four months or younger that had not received any treatment. Data was collected on 18 females and 20 males, with a total of 58 feet affected by this birth defect. Their age ranged from 7 to 120 days old, and the weight range was between 2.4 kg and 7 kg. When first introduced to the study, the children’s age, weight, sex and clubfoot severity rated by Pirani scoring test were recorded. It was also recorded if the children had bilateral or unilateral clubfoot. The evaluation and casting were done by orthopedic surgeons. “The initial stretching was aimed at correction of cavus by supinating the foot to bring the first metatarsal in line with the other metatarsals. The supinated foot was later gradually
abducted while applying counter pressure on the head of the talus until abduction of 60°." (p. 40) After casting achieved the desired abduction, the foot was dorsal flexed to assess range of motion. If the ankle could not flex beyond 15°, a heel cord release was performed to lengthen the Achilles tendon.4

The number of casts required to abduct the foot to the proper position ranged from 2-10 with an average of about 5. Linear regression analysis showed that age, weight, and sex had no association to the number of castings needed to correct the foot. Only the Pirani score had a correlation in the number of casts needed for correction. According to Awang et al4 “…castings required can be predicted based on the Pirani 10-point score at presentation as every one unit increase in Pirani score results in an increase of 0.544 units of the total number of required castings.” 4 (p. 42) Success of the treatment also relies on adherence to the brace requirements. Research also shows that, due to the mobility of older children, it has been difficult to keep them in the braces.4

Clubfoot relapses

After casting has achieved the desired foot position the child is required to start wearing braces to hold the feet in proper position. The child is also required to be seen by their clinician on the following schedule:

- After final cast removal braces are worn full-time.
- 3 months after final cast, braces need to be worn during nights-and-naps schedule until age 3.
- Patient should be checked every 4 months to monitor brace compliance and to check for relapses.
- Age 3 to 4 years check every 6 months.
- From 4 years until maturity check every 1 to 2 years.2

This schedule should be adhered to so the child can be monitored for brace compliance. It also helps to detect early signs of relapses. Some sings of relapse are a foot abduction and/or of dorsiflexion. Toddlers can also relapse. Some of the signs of relapse are described in the list below.

- As the child walks toward the examiner, supination of the forefoot.
- As the child walks away from the examiner there may be varus.
A seated child can show loss of ankle range of motion dorsiflexion.²

“The most common cause of relapse is noncompliance of the bracing program. Morcuende found that relapses occur in only 6% of compliant families and in more than 80% of noncompliant families. If relapse occurs in infants who are braced, the cause is an underlying muscle imbalance of the foot that can lead to stiffness and relapse.”²(p. 20)

**Ponseti method vs surgery**

There are two common methods to correct the birth defect known as clubfoot: the Ponseti approach and surgically using a comprehensive clubfoot release. According to Smith et al⁵ there have been many studies that have reported pain and decreased function in people who receive either treatments, but there was not information on the comparison of pain levels and function between the two groups. Smith et al⁵ compared two groups of adults who were treated with the Ponseti method and surgery. The two treatment groups underwent surgery or the Ponseti method between 1983 to 1987. The study compared the physical function, foot biomechanics, and quality of life of these two groups to those of a control group. The main purpose of this study was to verify which treatment was more advantageous for the patient to receive.

Physical function measured range of motion and gait analyses. Range of motion was diminished in both the surgery group and Ponseti group compared to the control group.⁵ The surgery group, however, displayed a reduced ability to planter flex the effected foot compared to the Ponseti group. Both the Ponseti and surgical group had decreased strength, with the surgical group being the weakest. In gait analyses, the Ponseti group was not significantly different from the control group. However, the surgery group had a notably lower score in almost all the categories. “[The] study showed that the biomechanical function and long-term outcomes of young adults treated with the Ponseti method more closely compare with function of individuals without clubfoot than to those treated using a comprehensive surgical method.”⁵ (p. 1289)

Osteoarthritis in the feet was assessed through the use of radiography. It was found that the control group was free of osteoarthritis. Both of the groups showed signs of moderate to severe osteoarthritis: “In the radiographic assessment, moderate to severe osteoarthritis was seen in 11 of 259 (4%) joints examined in the surgical group and six of 231 (3%) joints examined in the Ponseti group.”⁵(p.1284) The most common occurrence of osteoarthritis was found in the tibiotalar joint of the surgical group. The study also measured quality of life and the long term
impact of the treatments for clubfoot. Both treatment groups had worse hindfoot and midfoot scores when compared with the control group. The surgical group scored much lower than the Ponseti group and also experienced more pain than the Ponseti group.

The study concluded that young adults who were treated with a total surgical release were better off than most other treatments but still experienced side effects that were outside normal ranges, including a greater presents of arthritis. The Ponseti group more closely replicated the normal young adult. This was seen through range of motion, pain levels, and strength in the effected foot. The study supports the use of the Ponseti method in the use of clubfoot correction because of the results.

**Ultrasound review of non-ossified bones**

Until recently the only way to see the birth defect clubfoot was through x-ray or surgery. The problem with x-ray is that it does not show non-ossified bone. Through advances in technology, ultrasound is able to visualize the non-ossified bones found in the ankle joints of neonates and infants.

El-Adwar et al\textsuperscript{6} looked at three different views: medial, lateral, and dorsal. The medial view, which mimics an AP x-ray, was used to measure the distance from the medial malleolus to the navicular. This view showed a significant narrowing between the associations of these two bones. The medial view was also used to measure the medial soft tissue thickness.\textsuperscript{1,6} The measurement was taken from the medial border of the talus to the surface of the skin (see Figure 2). In the affected clubfoot of the soft tissue measurement was double when compared to the control group. The lateral view was used to see the displacement of the calcaneus to the cuboid. A line was made along the lateral border of the calcaneus, and a measurement was taken from that tangent down to the cuboid to show cuboidal displacement (see Figure 3). The dorsal view was used to see the navicular plantar displacement (see Figure 4). All three of these views were used to help confirm the severity of the clubfoot. The ultrasound views also can help to show if the Ponseti method is successful.\textsuperscript{6}

The Ponseti method was successful on all the affected feet studied by El-Adwar et al\textsuperscript{6}. The navicular had reduced subluxation to a more normal position (see Figure 5). The treatment also resulted in a reduction of the medial soft tissue thickness. The post treatment ultrasound in the lateral view showed a corrected alignment of the calcaneus and cuboidal (see Figure 6,7).
According to the post treatment ultrasound, the variables were useful in showing the true alignment of the non-ossified bones not visible on radiographs. This also shows that the Pirani scoring method is a good scoring system in evaluating clubfoot. “…[Ultrasound] can play an important role in assessing neonates and infants with clubfoot before and after treatment. Additional studies are needed to confirm if residual post-treatment [ultrasound] abnormalities are associated with future recurrences.” 6 (p. 2505) MRI can also be used to give superior images to evaluate the clubfoot birth defect. The advantage to ultrasound is that it does not require the infant to be sedated. According to Aurell et al1 a bilateral foot ultrasound could be performed on an infant without sedation in 10 minutes.

**Ultrasound review of healing of Achilles tenotomy**

Ultrasound can also be used to review the stages of healing after the Achilles tenotomy. A study by Mangat et al3 reviewed the phases of healing in children under 24 months using ultrasound. All of the children that participated in this study were treated with the Ponseti method. These patients also had a severe enough defect and were unable to dorsal flex the foot to a desired level so an Achilles tenotomy was performed. After the Achilles tenotomy the foot is casted in a dorsal flexed positon to help lengthen the Achilles tendon. There is also a palpable gap zone in the Achilles tendon.

The first phase according to Mangat et al3 was demonstrated on the scan that was preformed three weeks after initial tenotomy. This phase could be characterized by the appearance of distinct lines seen on ultrasound where the Achilles has been cut. The space between the cut lines is referred to as the gap zone. In the patients studied, the gap zone measured somewhere between 8 and 20mm. Ultrasound also showed in the gap zone and around the cut ends a bulbous formation with slight differences in echogenicity form a normal tendon. This difference in echogenicity was most likely a hematoma around the area. In this first phase patients showed some signs of irregular fibers spanning the gap. These irregular fibers were not common or abundant at this stage. At this point in healing, when the foot was dorsally flexed, the tendon appeared to move as one structure. This indicated that the tendon was continuous and healing. This view also can be used to confirm that the treatment did increase the length of the Achilles tendon.

6
The second phase of healing was scanned at six weeks. This phase showed a decrease in the hematoma formed around the gap zone. Phase two also showed an increase in fibers spanning the gap zone. These fibers also were more apparent and easier to find on the ultrasound scan when compared to phase one. In phase one, the fibers that did span the gap zone and seem to have a random pattern. In phase two, these fibers were much more linear. Although there were a greater number and more well organized fibers when compared to phase one, the tendon did not fully resemble a normal tendon. When compared to a normal tendon, these fibers were also not as well organized but show improvement from stage one. The gap zone was still easily seen in phase two.

Phase three of healing was demonstrated at 12 weeks. At this point in healing Mangat et al\textsuperscript{3} observed that the tendon was homogenous in appearance within the gap zone. This phase of near complete healing was demonstrated in 63\% of the patients studied. The remainder of the patients showed the same progression of healing, but the cut lines were still somewhat visible after twelve weeks of healing. Some of the patients in this study were scanned after the twelve week period. These scans showed that the tendon appeared the same as a normal tendon with no appearance of cut lines. Mangat et al\textsuperscript{3} also found that tendon healing was slower in children older than two years. This information can also be used in the treatment of relapse to help estimate a length of time for the tendon to heal.

**MRI analysis of muscle atrophy in unilateral congenital clubfoot**

One common attribute to unilateral congenital clubfoot is that the lower leg muscles on the effected side are smaller than the non-effected side.\textsuperscript{8-11} There has been debate the atrophy is due to the nature of the treatment, or it is a fundamental effect of the congenital defect. When it comes to treatment in some severe cases, a leg can be casted for up to two years.\textsuperscript{8} In a study done by Ippelito et al\textsuperscript{8} they compared the muscle volume, adipose tissue, and bone size across several age groups. There were several age groups in the study. The first group included two fetuses that were acquired after spontaneous abortion. The groups consisted of eight babies (age range 10 days to 2 weeks), eight children (age range 2–4 years), and eight adults (age range 19–23 years).

Ippelito et al\textsuperscript{8} found that in the fetuses the affected side had less muscle mass, more adipose tissue, and more bone mass. In the babies that were studied, it was found that the affected leg had less muscle tissue, more adipose tissue, and less bone mass. In the children
studied, the affected side had less muscle mass, more adipose tissue, and more bone mass. Finally in the adults studied, the affected side had less muscle mass, more adipose tissue, and more bone mass. The only variance between the four groups was that the subjects that were babies had less bone mass when compared to the unaffected leg. In a study done by Duce et al these finding are confirmed. Duce et al fond that the tibia in the affected leg was 8.5% smaller that the unaffected leg. Duce et al also stated that this difference was not due to surgery but more likely due to asymmetric leg development. The differences in adipose tissue and muscle was consistent across all groups. What can be derived from this information is that the muscle atrophy is not due to the treatment but most likely due to the defect. This can be seen since both new born babies and fetuses have less muscle mass that the non-affected side.

A study done by Moon et al studied the different tissue volumes when comparing treatment-resistant verses treatment-responsive clubfoot. Their findings were consistent with Ippelito et al. Moon et al found that the affected clubfoot had a greater amount of adipose tissue and less total muscle tissue. Moon et al also found that “…quantitative measurements of inter-leg differences in patients with unilateral clubfoot revealed that treatment-resistant clubfoot was associated with a greater difference in muscle and intracompartment fat areas compared with treatment-responsive clubfoot.” Unfortunately, the pathophysiological source for the increase in epimysial fat not known.

Conclusion

Congenital talipes equinovarus, also known as clubfoot, has many variables that affect people differently. The severity can range from mild to severe. This defect has been shown to have a decrease in total leg size with a greater chance of having chronic foot and leg pain. In the past, the most common treatment was surgery. Today, the most common treatment plan used is the Ponseti method. The Ponseti method should be utilized to help achieve the best possible results with the fewest side effects. This treatment has been shown to be the most successful treatment for congenital clubfoot. With continued use and research, this treatment has improved since its formation in the early 1960s. Through the utilization of ultrasound and MRI, the defect is better understood. These imaging modalities have given a real view into what is actually happing with the anatomy without the use of surgery. Ultrasound is a great tool due to the fact
that it can be performed without sedation. MRI has also shown that there are other factors to this birth defect that effect more than just the ankle and the foot.
References


Figure 1 demonstrates the six different variables involved in the use of the Pirani score.


Figure 2A–B “(A) A pretreatment line drawing and (B) the corresponding medial projection of the right foot of a 25-day-old neonate show Grade III medial displacement of the navicular over the talar head, an MMN distance (yellow line) of 3.1 mm, and a 12.8-mm MSTT (blue line). The red line delineates the cartilaginous border of the different bones. MM = medial malleolus; LM = lateral malleolus; TA = talus; OC = ossification center; and N = navicular.” Image from El-Adwar K, Taha Kotb H. The role of ultrasound in clubfoot treatment: correlation with the Pirani score and assessment of the Ponseti method. Clinical Orthopaedics and Related Research Sept. 2010;468(9):2495-2506
Fig. 3A–B “(A) A pretreatment line drawing and (B) the corresponding lateral projection of the same patient as in Figure 2 show medial displacement of the cuboid of 2.1 mm relative to the calcaneus. The red line delineates the cartilaginous border of the different bones. The yellow line represents the tangent along the lateral border of the calcaneus, whereas the blue line represents the measured CC distance. CAL=calcaneus; OC=ossification center; CU=cuboid.”

Image from El-Adwar K, Taha Kotb H. The role of ultrasound in clubfoot treatment: correlation with the Pirani score and assessment of the Ponseti method. Clinical Orthopaedics and Related Research Sept. 2010;468(9):2495-2506
Figure 4 A–B “(A) A pretreatment line drawing and (B) the corresponding dorsal projection of the same patient as in Figures 3 and 4 show a normally aligned navicular relative to the talus with no plantar displacement. The red line delineates the cartilaginous border of the different bones, and the yellow line represents a 13.8-mm talar length. TA = talus; OC = ossification center; N = navicular.” Image from El-Adwar K, Taha Kotb H. The role of ultrasound in clubfoot treatment: correlation with the Pirani score and assessment of the Ponseti method. Clinical Orthopaedics and Related Research Sept. 2010;468(9):2495-2506
Figure 5A–B “(A) A posttreatment line drawing and (B) the corresponding medial projection of the same patient as in Figures 2 through 4, obtained when the patient was 18 months old show an almost normally aligned navicular relative to the talar head, a MMN distance (yellow line) of 15.4 mm, and a 12-mm MSTT (blue line). The red line delineates the cartilaginous border of the different bones. MM = medial malleolus; LM = lateral malleolus; TA = talus; N = navicular.”

Image from El-Adwar K, Taha Kotb H. The role of ultrasound in clubfoot treatment: correlation with the Pirani score and assessment of the Ponseti method. Clinical Orthopaedics and Related Research Sept. 2010;468(9):2495-2506
Figure 6 A–B “(A) A posttreatment line drawing and (B) the corresponding dorsal projection of the same patient as in Figures 2 through 5 show a normally aligned navicular relative to the talus with no plantar displacement. TA = talus; N = navicular. The red line delineates the cartilaginous border of the different bones. The yellow line represents a talar length of 22.2 mm.” Image from El-Adwar K, Taha Kotb H. The role of ultrasound in clubfoot treatment: correlation with the Pirani score and assessment of the Ponseti method. Clinical Orthopaedics and Related Research Sept. 2010;468(9):2495-2506
Figure 7 A–B “(A) A posttreatment line drawing and (B) the corresponding lateral projection of the same patient as in Figures 2 through 6 show that the cuboid is normally aligned with the calcaneus. CAL = calcaneus; CU = cuboid. The red line delineates the cartilaginous border of the different bones. The yellow line represents the tangent along the lateral border of the calcaneus, whereas the blue line represents a CC distance of 0.3 mm.” Image from El-Adwar K, Taha Kotb H. The role of ultrasound in clubfoot treatment: correlation with the Pirani score and assessment of the Ponseti method. Clinical Orthopaedics and Related Research Sept. 2010;468(9):2495-2506