FIBULAR HEMIMELIA FOR PARENTS
Frequently Asked Questions

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Introduction: This information on fibular hemimelia (FH) is based on Dr. Paley's over 25 year experience in the treatment of this condition. Dr. Paley has performed over 15,000 limb lengthening and deformity correction surgeries of which at least 2000 were for FH.

What is fibular hemimelia?
Fibular hemimelia is a birth defect where part or all the fibular bone is missing and there are associated limb length discrepancy, foot deformities, and knee ligament problems.

How common is fibular hemimelia?
Fibular hemimelia occurs one in 40,000 births. Bilateral fibular hemimelia (fibular hemimelia affecting both legs) occurs much less commonly.

What are the genetics of fibular hemimelia?
Fibular hemimelia is usually not an inheritable condition. The vast majority of children born with this condition have no family history of other birth defects. It is important to obtain a family history of any birth defects. These can include different types of birth defects such as extra fingers or fingers fused together (syndactyly), etc.. If such a history is present, it can make one suspicious that there is a tendency towards birth defects in the family. Barring such a family history, it is extremely unlikely that fibular hemimelia is related to a genetic cause that is inheritable. This means that neither the parents of the child with fibular hemimelia nor the child themselves have any increased risk of producing additional children with this or other birth defects beyond the risk of one in 40,000 for fibular hemimelia. The exception to this is when fibular hemimelia is associated with birth defects in one or more the other limbs. For example, bilateral fibular hemimelia is often related to a gene disorder. In these cases, a thorough family history and possibly genetic testing are in order. When multiple limbs are affected by a limb deficiency, one can often assume that this was either an autosomal-dominant gene disorder (familial type of gene disorder) or related to a teratologic agent (birth defect causing agent such as a drug, radiation, virus, etc.).

What causes fibular hemimelia?
It is unknown why fibular hemimelia occurs. Some research in animals has demonstrated that if during early formation of the limb the genes guiding the formation of that limb are activated in an abnormal order, fibular hemimelia can occur. Other experiments have demonstrated that isolated mutations of genes in the forming limb bud can lead to missing parts of the limb. Since such gene mutations and abnormalities are occurring in the forming limb and not anywhere else in the child (especially not in their sperm or egg cells), these mutations are isolated to that child and cannot be transmitted to the next generation.

How does fibular hemimelia affect a child?

Children with fibular hemimelia have four main problems with their limb:

1. Limb length discrepancy.
2. Foot and ankle deformities.
3. Knee deformity.
4. Deficient cruciate ligaments of the knee.

1. Limb length discrepancy: Fibular hemimelia leads to a limb length discrepancy when it occurs on one side. This is because the main lower limb bone below the knee called tibia is growing at a slower rate than the tibia on the opposite side. In addition many children with fibular hemimelia have a slower growing femur (thigh bone) on the fibular hemimelia side than on the normal side. This combination of femur and tibia slower growth leads to a limb length discrepancy. The final component of the limb length discrepancy is the smaller foot. The foot tends to be shorter in length but also in height. The decreased height of the foot contributes to the limb length discrepancy. The limb length discrepancy with fibular hemimelia is proportional to the difference in growth between the two limbs and will be explained in the next section.

2. Foot and ankle deformities: The foot deformity is one of the biggest problems with fibular hemimelia. The foot deformity is related to the abnormal ankle joint as well as to missing parts of the foot. The ankle joint with fibular hemimelia may range from a normal ankle, which is stable, to a very abnormal shaped ankle which has limited mobility of the ankle joint and which is unstable. The fibula contributes to the stability of the ankle. The end of the fibular can be felt as a big bump on the lateral side of our ankles. Children with fibular hemimelia are missing part or all of the fibula and this bump may be completely missing. It is called the lateral malleolus. When the lateral malleolus is present, it buttresses the ankle bone (talus) and prevents it from coming out of joint. When it is missing, this stabilizing effect is absent. The ankle joint is primarily made up by the lower end of the tibia bone. This lower end of the bone is often severely deformed in fibular hemimelia. This deformity comes from a bend in this bone that can be located in the main shaft of the bone and form a large bend or knuckle like appearance of the tibia (often with a skin dimple over the knuckle). A more subtle deformity of the ankle is the one that is often not visible on the x-ray. This is the malorientation of the joint itself. The ankle joint frequently faces to the side (lateral) and to the back (posterior). This malorientation points the foot towards the outside and down creating what is called an equinovalgus deformity. This deformity was thought to be due to tight soft tissues such as the Achilles tendon and peroneal tendons as well as due to a fibrous remnant of the fibular bone. A fibrous remnant is often referred to as the ***anlage***, a word coming from German. In 1996, Dr. Paley was first to identify that the equinovalgus deformity in fibular hemimelia was not just due to tight muscles and to the fibular hemimelia anlage. He discovered that it was due to malorientation of the joint itself which was invisible because the joint is mostly made of cartilage at a young age and therefore cannot be seen on the x-ray. His findings have since been confirmed by MRI studies as well as by open surgical examination of this joint. This discovery led to the development of the only effective surgical treatment for reconstructing the foot and ankle for fibular hemimelia and now called ***superankle procedure***. This procedure will be described below:

In addition to the ankle deformities, the fibular hemimelia foot may also have a deformity between the ankle and heel bones (talus and calcaneus). Normally, these two bones are connected to through the subtalar joint. While the ankle joint moves the foot up and down. The subtalar joint allows the foot to move side-to-side and it is important for walking on uneven ground. The subtalar joint in fibular hemimelia is usually fused. This means that the joint is actually absent and the two bones are connected (fused) together. In a normal ankle fusion of the subtalar joint would prevent side to side motion of the ankle. In FH side-to-side motion is usually present due to a an abnormal shaping of the ankle joint called, ball and socket ankle joint. The ball and socket shape of the ankle bone (talus) and the lower end of the tibia allow side-to-side movement in addition to up and down movement. Therefore, the ankle joint functions for both the ankle and subtalar joints. This fusion of the subtalar joint is called a subtalar coalition. This means that the two bones are connected initially by cartilage and later by bone. If the subtalar coalition connects the talus and calcaneus in a normal position such that the heel is in line with the ankle bone then it does not contribute to additional deformity around the foot. If however this coalition is joint in an abnormal fashion, so that it is tilted outwards (valgus) or inwards (varus) then it leads to additional deformity around the foot and ankle.

Dr. Paley was one of the first who recognized the contribution of the coalition deformities to fibular hemimelia which helped form the basis of the superankle procedure to correct these deformities. In many patients, there is a combination of malorientation of the ankle and malposition of the subtalar coalition.

The third element of the foot deformity is the absence of some of the toes including the foot metatarsal bones. The metatarsals are the long bones in the foot that lead down to the toes. Normally, there are five metatarsals and five toes while in fibular hemimelia, there may be four, three, two, or one metatarsal bones and toes. In fact, in some patients, the numbers of toes do not correspond to the number of metatarsals. There can be two metatarsals and three toes etc. Some surgeons feel that the absence of metatarsals leading to excessive narrowing of the foot is an indication for amputation. I disagree with this and will discuss the role of number of toes or rays later in a later section. Some of the toes may also be joined together (syndactyly) and may be considered for separation at a later date. The big toe may also be pointing inwards away from the rest of the foot. This is due to a particular deformity called delta metatarsal and will be discussed later too.

3. Knee joint deformities: The knee joint frequently has a valgus (knocked knee) deformity. This knocked knee alignment can be related to the lower end of the femur bone (thigh bone), or the upper end of the tibia, or both. It is important to realign the knee as a part of the treatment of fibular hemimelia. This will be discussed in a later section.

4. Absent knee ligaments: Most patients with FH have partially or completely absent cruciate ligaments. In particular, the anterior cruciate ligament (ACL) is often hypoplastic (under developed) or absent. The absence of these ligaments may or may not cause any clinical problem to a patient affected by fibular hemimelia. No initial treatment for this is provided but ligament reconstruction should be considered if the children begin to develop problems of knee instability. Unlike athletes who tear there cruciate ligaments and require reconstruction, children with deficient or absent cruciate ligaments often do very well and do not require reconstruction. The absence of the ACL does not prevent them from participating actively in various sports that other children their age can participate in.

Classification of Fibular Hemimelia:

Fibular hemimelia is not one condition where all of the cases have the same amount of deformity or deficiency or limb length discrepancy. As such we classify fibular hemimelia into different groups so that we can recommend different treatments according to different degrees of severity. There are many classifications of fibular hemimelia that have been published. The problem with the majority of these is that they were developed at a time when surgical reconstruction for fibular hemimelia was unsuccessful. Therefore, the different groups of fibular hemimelia that have been described do not relate to different types of treatment since the majority of patients with this condition where recommended to undergo amputation and prosthetic fitting. The most common classification that was used for fibular hemimelia was called the Kalamchi classification which described the amount of fibula that was absent. We now know that the amount of leg length discrepancy and foot deformity which are the two biggest problems in fibular hemimelia do not relate to the amount of fibula that is missing. Furthermore, since many of the most severe cases are the once that have complete absence of the fibula. A classification is needed that can differentiate between different types that have complete absence of the fibula.

In 1995, Dr. Paley developed a classification for fibular hemimelia which is now known as the Paley classification. It divides fibular hemimelia into four groups; types 1, 2, 3, and 4. It then subdivides type 3 into four types; 3A, B, C, D. The Paley classification is related to treatment of fibular hemimelia. Each type and subtype has a different treatment.

Paley type 1 is a congenitally short tibia and fibula with a stable ankle joint. In many cases the ankle of type 1 cases appears completely normal and the fibula is only slightly shorter at its upper end compared to the opposite side. The predicted leg length discrepancy in type 1 cases is typical under 7.0 cm (3 inches).

Paley type 2 cases have a short tibia and fibula with a foot that stands flat to the ground but often goes into a valgus position. This means that the foot can come into a normal position but as the child gets older or more tired, it goes into valgus such that the heel is pointing outwards and the ankle is pointing inwards. Children with type 2 often have a tight Achilles tendon as well. Another common finding with type 2 is the ball and socket ankle joint.

Paley type 3 cases demonstrate a fixed foot deformity with the foot pointing down and to the side. This is known as fixed equinovalgus deformity. The foot has limited mobility and cannot brought into a 90-degree (plantigrade) position relative to the lower leg. While the more common birth deformity called club foot has the foot pointing down and in, this is the opposite deformity with the foot pointing down and out. This is the type of deformity that has malorientation of the ankle joint and/or malunited subtalar coalition. The type 3cases can be subdivided into four types. In type 3A, the fixed equinovalgus deformity is due to a malorientation of the ankle joint. In type 3B, the fixed equinovalgus deformity is due only to a malunited subtalar coalition. In type 3C, it is due to a combination of maloriented ankle joint and a malunited subtalar coalition. In type 3D, the subtalar joint is not fused and does not have the coalition but is maloriented. In fact in this condition, there is hypermobility of the subtalar joint. As will be described in a later section, the differences in anatomy between all of these different types of fibular hemimelia require different variations of the superankle procedure.

Paley type 4 also known as the club foot type of fibular hemimelia has a subtalar coalition maloriented into varus (foot turned inwards) and appears more like a club foot deformity instead of the equinovalgus typical fibular hemimelia. This equinovarus or club foot deformity can often be mistaken to be a from a true club foot. The treatment of a true club foot,is by manipulation and casting and Achilles tendon lengthing using the Ponseti technique. A fibular hemimelia treated mistakenly by this technique would fail to correct. Furthermore, such manipulation in the presence of a subtalar coalition can damage the ankle joint and growth plate of the lower tibia. I have even seen cases that due to this mistaken identity, underwent extensive club foot surgery which failed to correct the deformity. Therefore one must beware of fibular hemimelia masquerading as a club foot.

Reconstructive Treatment:

The surgical treatment for fibular hemimelia is designed to address all of the deformities and deficiencies and length discrepancies as described above. At the Paley Institute, we start with a **reconstructive life plan**. This involves evaluating all of the surgical deformities, deficiencies, and predicting the limb length discrepancy at maturity and then coming up with a surgical plan to correct these in the fewest number of surgeries spread apart as much as possible throughout the child's growing years, so that by skeletal maturity, the child has achieved equal leg length, a functional plantigrade foot (foot in a 90-degree position), excellent alignment of the hip, knee, and ankle, and when necessary stability of the knee joint.

Step 1-Predicting Leg Length Discrepancy and determining the number of lengthening surgeries:

First step is predicting the leg length discrepancy at skeletal maturity. This is carried out by obtaining x-rays of both lower limbs and measuring the lengths of both femora both tibiae and both feet heights. By comparing the sum of the femur tibia and feet height on the short leg with that on the long leg, we can determine the leg length discrepancy at the time of the x-ray. Using the multiplier method for limb length discrepancy prediction, we can then calculate the leg length discrepancy at skeletal maturity. The multiplier method is a method of leg length discrepancy prediction that was invented by Dr. Dror Paley in 1997 and published in The Journal of Bone & Joint Surgery in 2000. It is the simplest and most accurate method ever developed for limb length discrepancy prediction. Using a series of coefficients and simple mathematical formula, Dr. Paley demonstrated that one could predict the leg length discrepancy of children born with a congenital limb deficiency accurately with only one measurement of limb length discrepancy. Additional measurements at additional times improves the accuracy of prediction. The accuracy of prediction even with a single measurement arrives at a prediction that is accurate within 1.0 cm in over 85% of patients and within 2.0 cm in 95%. The Paley multiplier method is now the accepted worldwide standard for predicting limb length discrepancy. Its use has been simplified and facilitated by the Paley Growth app on the iPhone. Once the predicted leg length discrepancy at skeletal maturity has been calculated, a determination of the number of limb length equalization procedures can be made.

Under the age of four it is safe to lengthen up to 5.0 cm in the tibia (over 5cms, 2 inches, can lead to growth inhibition). Subsequent lengthenings can be performed preferably four years apart as needed to achieve limb length equalization at skeletal maturity. Lengthenings performed at an older age can achieve up to 8.0 cm of lengthening. Therefore, one lengthening at age four and one at age eight would achieve total of 5.0 cm plus 8.0 cm equals 13.0 cm in total (5.1 inches). One lengthening at age 4 plus one at age 8 and one at age 12 would achieve the total of 5.0 cm plus 8.0 cm plus 8.0 cm equals 21.0 cm (8 ¼ inches). If additional equalization is required, one can also consider leg growth slowing procedure called epiphysiodesis. Epiphysiodesis involves **slowing the growth of long leg** to allow the growth on the short leg to catch up. This method will be explained in a later section. Epiphysiodesis is typically performed at a specific age calculated with the Paley multiplier formulae. While limb lengthening can be performed at any age, epiphysiodesis has to be performed at a very specific age in order to achieve a specific amount of growth slowing. We rarely use epiphysiodesis to achieve more that 5.0 cm (2 inches) of limb length equalization. Therefore, if in addition to the lengthening strategy as outlined above we add an epiphysiodesis of 5.0 cm, we can increase the correction up to 25.0 cm in the example with three lengthenings plus an epiphysiodesis. This covers the majority of cases with limb length discrepancy due to fibular hemimelia. It is rarely ever necessary to perform more than three limb lengthening procedures and one epiphysiodesis procedure to equalize limb length discrepancy due to fibular hemimelia.

Cases that present with larger discrepancies than 25.0 cm, usually have some shortening in the femur and would be candidate for simultaneous lengthening of the femur and tibia. This will be discussed in a later section.

Step 2-Determining the Paley type of FH:
The next step is to determine what type of FH the child has. At the initial consultation, it is determined whether the patient has a fixed foot deformity that will require surgical treatment or even a dynamic foot deformity that will require surgical treatment. Fixed foot deformities are those where the foot cannot be brought to the midline into a plantigrade (foot at 90 degrees) position. A dynamic foot deformity is one in which the foot has a tendency to go into an abnormal position such as valgus but can easily come into a plantigrade position. Dynamic deformities include feet that reach plantigrade position but have tightness of certain tendons that will obstruct lengthening or likely create problems during lengthening and putting the foot into an abnormal position such as a toe down position (equinus). Most patients with type 1 do not require any foot surgery. Most patients with type 2 will require realignment of the ankle combined with lengthening of the gastrocsoleus muscles (the two muscles connected to the Achilles tendon). This should be planned and included as a part of the surgical procedure.

Type 3 and type 4 have fixed deformities that must be corrected early to allow the patient to walk with the foot in a plantigrade position and to be able to wear a shoe properly. Type 3 and type 4 are treated by the superankle procedure. The super ankle procedure is an operation that was developed by Dr. Dror Paley in 1996. It is the most successful method to correct the fixed equinovalgus of type 3 or fixed equinovarus of type 4 fibular hemimelia. The superankle is the first and only procedure that has reduced and almost eliminated recurrent foot and ankle deformity following treatment for fibular hemimelia. The superankle procedure is the most revolutionary advance in the treatment of fibular hemimelia. Prior to the advent of this procedure, the biggest cause of failure of surgical reconstruction was recurrent foot and ankle deformity. The previous procedures such as soft tissue releases or joint distraction had very high failure rates and lead to increased or permanent stiffness. The superankle procedure combines extra-articular soft tissue releases with intra-articular reshaping of the ankle joint with extra-articular osteotomy bony alignment of the ankle and subtalar joints. It is a very technically demanding surgery and will be discussed in more detail below. At the first consultation, a determination if the super ankle procedure is required is made. If this procedure is to be performed on its own, it can be done as early as 12 months of age. If it is to be performed in combination with lengthening, it is preferable to wait until 18 months of age. This is not to say that it cannot be performed at an older age. This is referring only to the youngest age at which we perform this surgery and the guidelines for how to choose that youngest age. My oldest patient to undergo the superankle was 16 years old.

In summary, at this first consultation an evaluation is made of the type of foot deformity and therefore the classification of the fibular hemimelia as well as to the predicted leg length discrepancy at skeletal maturity and the number of lengthenings and/or epiphysiodesis that are required and finally whether a valgus knee is present and should also be corrected. The parent is given a surgical plan as to when we

would perform the first surgery and then how many total surgeries and at what ages they would be required.

Example of reconstructive life plan: 12 month old patient with a Paley type 3A fibular hemimelia and predicted leg length discrepancy at skeletal maturity of 25.0 cm in which there is also a valgus knee deformity.

Surgery #1, at age 18 months, superankle procedure combined with lengthening of 5.0 cm combined with hemiepiphysiodesis of distal femur for valgus knee correction.

Surgery #2, at age eight years, lengthening 8.0 cm of tibia.

Surgery #3, at age 12 years, lengthening 8.0 cm of tibia.

Surgery #4, at age 13 epiphysiodesis of tibia on long leg for correction of 5.0 cm.

Total leg length equalization = 25cms (10 inches)

Thus the parents leave that first consultation with an approximate plan and a good understanding of what would be involved should they choose to reconstruct and lengthen their child's leg with fibular hemimelia.

What do I do until the first surgery?

One of the most common questions again asked is "how soon do I need to come to the Paley Institute to be evaluate for my child's fibular hemimelia?" Since no treatment is required prior to walking age, there is no urgency in the timing of the first consultation. I usually recommend parents to come in around age six months of age and to treat the child normally until that time with no specialized braces, shoes, or physical therapy. Physical therapy is not useful prior to surgical treatment. This is a common misconception. A lot of parents tell me that they can get free early intervention physical therapy and therefore because it is free they should probably do it. Physical therapy if carried inappropriately can actually be harmful to the foot. I highly recommend against any manipulation therapy of the foot and ankle includes casting or bracing prior to surgery. The first intervention is the use of a shoe lift at the time of walking. Once the child demonstrates that they are cruising the furniture, it is then appropriate to obtain a brace or shoe lift or both. By this time, they should have seen a consultant who can advise them and prescribe such treatment. In most cases, I recommend an ankle foot orthotic (AFO) which is custom molded to the foot. Then a shoe lift is added to the bottom of the shoe to equalize limb length discrepancy. We generally aim to prescribe a shoe lift that is 1.0 cm less than the leg length discrepancy. After that, the child should be booked for the first surgery. In most cases, this means a superankle procedure between age 12 and 18 months depending whether it is going to combine with lengthening and that decision will be discussed in the section of the supper ankle. No other treatment is required prior to this first surgery.

Are children with FH delayed in the age of walking: Children with fibular hemimelia learn to walk either at normal age or slightly delayed. All of them will walk and can walk with fibular hemimelia even in very extreme cases. There is no neurologic obstruction to learning to walk in children with fibular hemimelia.

Superankle Procedure:

As mentioned above the super ankle procedure was invented by Dr. Dror Paley in 1996. This procedure achieves a stable plantigrade foot and ankle in the first surgery. It can be combined with lengthening but it does not have to be. When there is a severe bend in the shaft of the tibia, if the superankle is combined with lengthening that bend (the knuckle) is corrected gradually with the lengthening using an external fixator. When no lengthening at the initial surgery, the bend is corrected by taking a wedge out of the bone and using internal pins and a cast. The lengthening is preformed at a later date. The decision between performing the superankle in combination with lengthening versus performing it without lengthening can be based on several criteria. If there is an extremely bad deformity, they cannot wait

until 18 months of age and needs to be corrected earlier I will perform a superankle without lengthening. This is more commonly done in bilateral cases where limb length discrepancy is not a consideration. The other situation is when there is a well-developed ankle joint with excellent motion. There is a theoretical advantage to separating the superankle procedure from the lengthening to better preserve the ankle moion. The rationale for this is that performing the lengthening right away leads to more stiffness of the ankle joint. Since the ankle joint has excellent range of motion before surgery, theoretically performing the superankle first to realign the foot and ankle and to stabilize the ankle joint followed by physical therapy will maximize and optimize the ankle range of motion. In theory, the lengthening could lead to loss of some range of motion since the external fixator is on for a longer period of time and since the muscles are tightened by the lengthening. This decision and discussion should be made with the surgeon and is really based on the surgeon's personal experience, which is why the more experience a surgeon has with this operation the better the decisions they can make on the best option to offer the patient. Ankles that have limited range of motion due to poor formation of the shape of the ankle joint (dysplastic ankle) cannot benefit from this two-stage superankle and lengthening approach. In these patients, it is preferable to perform all of the deformity correction of the superankle combined with the lengthening in order to decrease the total number of surgeries.

In the superankle procedure, the first step is to remove the fibrous fibular anlage (remnant). This remnant of the fibular bone tethers the lengthening and pulls the ankle and tibia into a valgus deformity (foot leaning to the outside). By removing this band it facilitates the lengthening and prevents recurrent valgus. The second step of the superankle is to lengthen the peroneal and Achilles tendons. The third step is to decompress the peroneal and posterior tibial nerves to prevent any damage to these structures from the deformity correction of the super ankle. The fourth step is carried out only in very dysplastic ankles and is a relatively newer modification of this procedure which many surgeons, who learnt this from Dr. Paley a long time ago, may not be familiar with. This involves reshaping the dome of the talus bone. The talus is the ankle bone and it normally has a cylindrical shape to its joint surface. The surface of talus resembles half of a cylinder. Dysplasia (malformation) of the talus leads to flattening of this cylindrical shape, so that the amount of rounding of the surface is limited. Furthermore, dysplasia of the talus leads to shortening of the front and back of the talus bone. This causes the front of the talus to bump into the front of the tibia when the foot is pulled up and the top of the heel bone to collide with the back of the tibia when the foot is pointed down. This leads to a very limited arc of motion within the ankle joint. In 2009, Dr. Paley started reshaping the talus and calcaneus bones to deepen the groove at the front and back of the ankle joint that permits the tibia to rock back and forth on the talus thus increasing the range of motion of the ankle joint. While this newer addition to the superankle procedure, it showing promise increasing the range of motion, it is still too early to confirm that the ankle range of motion can be significantly increase by this modification. This modification does not cause any harm and therefore there is no downside to adding these groove deepenings as part of the superankle procedure. The fifth step of the super ankle is to perform the osteotomies. In the case of type 3A, this involves a supramalleolar osteotomy of the tibia(bone cut above the ankle joint). In type 3B, this involves a subtalar coalition osteotomy (bone cut between the talus and calcaneus (ankle and heel bones) to realign the calcaneus to the talus. In type 3C, it involves the combination of supramalleolar and subtalar osteotomies. Type 3D involves a very specialized osteotomy of the talus bone to reposition the subtalar joint orientation. In some of these cases, they may also require a supramalleolar osteotomy.

Finally in type 4, it also involves a subtalar osteotomy often combined with a supramalleolar osteotomy but the subtalar osteotomy involves tilting of the heel bone the opposite way (out of varus) than in the more common subtalar osteotomy for type 3B.

In summary, knowing the Paley classification type of FH determines very specifically the type of osteotomy that should be performed.

The sixth step of the superankle involves pinning of the osteotomy with internal pins driven in through the heel pad and across the subtalar and ankle joints.

The seventh step is repair of the lengthened tendons of the Achilles and peroneal tendons.

The eighth step involves performing a procedure that allows for swelling of the muscles within the leg called a compartment release.

After these eight steps, the skin incisions (total of two) are closed. The final step is applying an external fixator and cutting the bone usually at the apex of the bend of the tibia. If lengthening is not going to be performed at the same time but there is a bend the tibia then before closing the skin, the tibia should be cut in either a wedge or more commonly a trapezoidal shape and a small segment of bone should be removed to allow complete straightening of the tibia. Once the tibia is straightened it can be fixed internally either with pins or metal plate and screws. The straightening of the shaft of the tibia is complicated because foot also has to be turned externally since it is internally twisted before that. With lengthening, this is carried out by a gradual correction of the bends and the portion of the bone using a computer controlled device called Taylor Spatial Frame external fixator. If the lengthening is performed, the patient is given an adjustment schedule to start lengthening usually five to seven days after the surgery.

Correction of Knee Valgus Deformity:
The valgus deformity (knocked knee deformity) of the knee has been referred to several times above. This deformity should be corrected with the first treatment. Valgus of the knee can negatively impact the correction on the foot. The valgus of the knee is most commonly from the lower femur. Even though, the child has fibular hemimelia and the primary deformities and deficiencies are below the knee, there is often an associated development change in the lower femur which is the top part of the knee joint. This deformity can very easily be corrected by tethering of the growth plate of the lower femur on the inside of the knee using a device called a hemiepiphysiodesis plate. You may hear this refer to as an 8-plate, peanut plate or hinge-plate device. Hemiepiphysiodesis temporarily prevents growth on one side of the growth plate while not impeding growth in the rest of the growth plate. This forces the growth plate to grow towards the tether. Therefore, if the knee is a knocked knee, the leg will grow to become straight and if the plate is not removed, it will actually become bow-legged. Therefore, it is critical to follow this correction every three months with an x-ray and then to remove this plate early enough to prevent overcorrection. A slight amount of overcorrection is acceptable because some rebound valgus can occur. An 8-plate hemiepiphysiodesis can be repeated more than once throughout the child's growth years if the valgus deformity recurs, which may occur in some cases. It is a very minor outpatient procedure when carried out on its own and can be combined with a lengthening and super ankle procedure as early as 12 to 18 months of age.

Toe and Metatarsal Surgery:
Some patients with fibular hemimelia have fusion of two metatarsals together or two toes together. This requires a specialized surgery to realign the toes and two metatarsals and particularly the first and second which are fused together. The big toe may be in a varus (pointing) position. It is some times joint to the second toe. This requires a specialized operation to release some of the muscles and tendon and decompress nerves in the foot in order to realign the toe and possibly to straighten the double metatarsal. It is a very complex surgery and may need to be performed prior to one year of age. Separation of the toes such as for syndactyly can also be combined with this surgery or when they occur an isolation typically are carried out prior to one year of age. This procedure may require a skin graft from the groin to be performed at the same time. This additional surgery was not discussed before but it is a part of the surgical plan and would be determined and decided at the first consultation. It is one of the surgeries that we perform in addition to all the other described above. In some cases, the fibular hemimelia have a "Y" shaped metatarsal where two separate toes come off from one metatarsal. At the end of the metatarsal, there are two separate heads in the shape of letter "Y". This also requires a special reconstruction. The parent should make sure to discuss these toe deformities with the surgeon. The one toe issue we cannot address is to add toes that are missing. Despite anxiety over this issue it does not produce any problem for the child except modification of shoe wear or the need for orthotics.

Knee Ligament Reconstruction:
Knee ligament reconstruction is rarely required for the patients with fibular hemimelia. The patients with combined femoral shortening and fibular hemimelia are more likely to require knee ligament reconstruction. Knee ligament reconstruction is performed using the Paley superknee procedure which was developed in 1994 by Dr. Dror Paley. The superknee procedure can be performed as young as 18 months of age and to and including skeletal maturity.

The Femoral Lengthening:
Femoral lengthening can be combined with the tibial lengthening. This is only carried out when there is a combined shortening of the femur and tibia and femoral shortening is of significant magnitude. In such cases, it is not unusual to perform the superankle procedure with application of the external fixator for lengthening and the femoral lengthening procedure by application of an external fixator on to the femur. In these cases, lengthening of the rectus femoris and fascia lata structures are also performed. If femoral lengthening is considered, it is factored into to the surgical life plan. Obviously, simultaneous femoral and tibial lengthening can yield much larger amounts of lengthening in one treatment that tibia lengthening alone. For example, simultaneous 5.0 cm femoral and 5.0 cm tibia lengthening together take a total of five months of external fixation. Isolated tibia lengthening of 5.0 cm takes a total of five months of external fixation. Therefore, in the first example combined femoral and tibia lengthening acheive 10.0 cm (4 inches) of leg length equalization compared to only 5.0 cm (2 inches) when only the tibia is lengthened. Of course, there is no indication to do femoral lengthening in the absence of femoral shortening.

How long will the external fixator be on child's leg?
Lengthening by external fixation involves two phases: Distraction phase and consolidation phase.

The distraction phase is the time period from the operation until the adjustments of the lengthening are concluded. The consolidation phase begins at the end of the distraction phase until the external fixator is removed from the leg. In most children, for a single level lengthening (one bone cut in the tibia) the total external fixation time is equal to one month per centimeter. This is an approximate average time and can vary between individuals. This means that some individuals take more than one month per centimeter and some less than one month per centimeter but that the majority are one month per centimeter. During the external fixation time, approximately half of it is the distraction phase and half is the consolidation phase. Therefore a 5-cm lengthening takes approximately two and a half months for lengthening and two and a half months for bone hardening. Similarly for an 8-cm lengthening, the total external fixation treatment time is expected to be eight months of which four months is the distraction phase and four months is a consolidation phase. Prior to beginning the distraction, there is a waiting period of typically five to seven days. During this time, the patient is usually discharged from hospital after approximately three to four days and begins physical therapy. The lengthening rate in fibular hemimelia is usually about 0.7 mm per day. This rate should be adjusted according to bone formation and range of motion of the joints. During the lengthening for fibular hemimelia, it is essential to undergo physical therapy. This will be discussed in a separate section. Please do not get confused between lengthening rate and the time in external fixation. The one month per cm is not the rate of lengthening. It is the sum of the time taken to lengthen and harden the new bone.

Physical Therapy:
Physical therapy is essential for almost all in lengthening procedures. Physical therapy for fibular hemimelia focuses on the knee range of motion and toe range of motion. It also focuses on gait training (walking). The tendency during the lengthening is for muscles to get tighter causing the toes to flex and the knee to bend. If these joints are no diligently stretched, they will become stuck (contracted) in the flexed position. The foot and ankle are immobilized by the external fixator. A common mistake is to perform lengthening in fibular hemimelia without fixation of the foot in a 90-degree position. With the superankle, the foot is fixed and pinned in a 90-degree position. This principle is the same for all subsequent lengthenings at an older age after a previous superankle. The foot must always be fixed with external fixation FH lengthening. Even though the superankle procedure leads to stability of the ankle joint, lengthening will destabilized that ankle joint and cause the ankle to start coming out joint (subluxation or dislocation) and the foot to go into an equinovalgus position once again. The only type of fibular hemimelia that can be considered for lengthening without foot fixation is type 1. All of the other types including types 2, 3, and 4 must have foot fixation during the lengthening.

In type 1 where the foot is not immobilized, a splint such as cast or special boot should be used to maintain the foot position at 90 degrees. Physical therapy in such cases should focus on stretching the ankle to stretch the Achilles tendon. In older children where implantable limb lengthening is considered and the foot and ankle are not immobilized by the implantable device, a special cast or brace must stabilize the ankle and foot at 90 degrees.

In some cases a specialized spring loaded splint such as the Dynasplint are required during the lengthening to prevent knee flexion contracture. For the toes, a special removable toe splint or forefoot splint is custom made before the lengthening begins and is used on a regular basis. If simultaneous femur and tibia lengthening are carried out then the knee extension bar needs to be applied every night to prevent knee contracture and knee range of motion is even more imperative for both flexion and extension.

Amputation Versus Reconstruction:

Until now all I have discussed is the reconstruction options for fibular hemimelia. Clearly, my **bias** is towards limb reconstruction and not amputation for this condition. This bias is based on over 25 years' experience of reconstructing more than 2000 fibular hemimelia limbs. (this is probably more than any surgeon in the world). Many of those patients had some or all of the procedures described above.

Amputation remains the most common option presented to parents with children who are born with fibular hemimelia. I am therefore asked by both parents and surgeons why I think reconstruction is a better option for almost all patients with FH. To explain that I have to start by explaining why amputation is offered as the main alternative. Performing an amputation at the level of the ankle joint (Syme's amputation) gives a nice round stump with the healed skin as a weightbearing surface. That combined with modern prosthetics leads to unrestricted excellent function. As we all saw demonstrated in the 2012 London Olympics, amputees and even bilateral below the knee amputees such as Oscar Pistorius fitted with advanced prosthetics can even complete at the highest level. There is no question that a patient with fibular hemimelia who undergoes a Syme's amputation and good prosthetic fitting and who has access to a technologically advanced prosthesis and prosthetic care on a regular basis (most children need a new prosthesis each year) will function normally for almost any activity. It is not uncommon to see video clips of children skateboarding, rock-climbing, and performing individual and team sports following a below knee amputation.

Nevertheless, if an amputation could be avoided and the foot and ankle and leg reconstructed to nearly normal function comparable to that afforded by a below the knee prosthetic, most parents and most individuals will choose to have the reconstruction. I do not think anybody wants to give up their foot or ankle unless there are no good alternatives.

When pediatric orthopedic surgeons are asked 'would you amputate the foot if all that was wrong with the leg was a foot or ankle deformity such as club foot or many other childhood foot deformities', the answer is universally "no". Despite this the results of some club foot treatments leave the child with chronic pain and a stiff deformed foot that might be better treated by amputation and prosthetic fitting. When orthopedic surgeons were asked will they amputate the leg of a child with no foot deformity and just a leg length discrepancy, the answer is almost universally "no". When orthopedic surgeons where asked will they amputate the leg of a child with combination of foot deformity and a leg length discrepancy, the answer frequently is "yes". The logic of this is escapes me since for a foot deformity our standard answer is to perform foot deformity correction surgery, and for a limb length discrepancy our answer is to perform a limb lengthening surgery. Therefore, it does not make sense that when there is a foot deformity combined with a leg length discrepancy, the answer is a foot deformity correction surgery combined with a leg lengthening. What I recommend is exactly that: foot deformity correction with the supernakle procedure combined with lengthening using an external fixator. We recently compared the results of the superankle procedure combined with lengthening to Syme's amputation.

Twenty-two patients that I personally treated with the superankle procedure where compared to an age matched group of patients who underwent Syme's amputation at the Dallas at Texas Scottish Rite Hospital. The results of the two groups demonstrated no difference in function between the two groups. Both groups where satisfied with their results and both groups were equally and functionally active and both groups had no pain. Both groups give comparable function to normal. The choice is therefore that of the parents as to which procedure they prefer for their child. With lengthening reconstruction surgery using the superankle and lengthening, the big advantage is that in addition to normal function, one retains a sensate foot that can feel the ground and that has balance and proprioception (balance and position) sense. No prosthesis provides sensibility or proprioception. Furthermore, the child and later the adult with the prosthesis must get an expensive high quality technically advanced prosthesis made every year throughout childhood and frequently every year throughout adult life. This is an important economic consideration. The total cost to health care of that many prosthetic changes is much greater than all of the medical costs related to the surgery of lengthening reconstruction surgery. This does not even factor in the frequent adjustments and modifications to prosthesis that are required, nor the intermittent skin irritation of the stump to the prosthetic that causes some pain and suffering and sometimes interrupts prosthetic use. It also does not factor in the children with fibular hemimelia with missing knee ligament and therefore adding a prosthesis with added stress to the knee and also does not factor in the valgus of the knee that still needs to be corrected in children who have amputation for fibular hemimelia. Our study also could not quantify the psychologic effects of having the prosthesis and going to the beach and having to take it off in order to get in the water or walking on the sand one legged having to remove the prosthesis or any stress to the individual created by having to wear shorts or a short skirt with the prosthesis. With lengthening reconstruction surgery, these are not considerations that one has to deal with.

I have plenty of examples and videos of my patients who have undergone the superankle procedure with lengthening as well as simpler cases who just underwent lengthening without the super ankle procedure. These videos demonstrate these patients are doing every manner of sport from baseball, football, basketball, tennis, soccer, gymnastics, rock-climbing, etc. Therefore, the decision is a personal one and **not one** that should be dictated by the surgeon. The results and success that we are able to achieve at the Paley Institute are now world-renowned and available for everyone to see. Patients travel from over 70 different countries and 50 states in order to obtain this treatment which unfortunately is not readily available elsewhere. This leads me to my last argument and that is that the option of amputation is too readily provided because of the lack of training and availability of the superankle procedure. While every pediatric orthopedic surgeon has been trained in amputation techniques and while amputation is not technically a difficult procedure, the superankle is a very technically challenging operation. It is also not well known by most orthopedic surgeons. In order to get good at the superankle procedure, one requires extensive experience and training. Since the fibular hemimelia is a rare diagnosis, the majority of the pediatric orthopedic surgeons do not see many cases annually with this condition. In order to good at this operation and all of its variations, for the different Paley types, one needs to perform this operation several times a year. Most pediatric orthopedic surgeons do not even see one such case every year. Therefore, it is impossible for them to get sufficient experience with this procedure even if they do obtain proper training. Therefore with the lack of training and experience, and the rarity of the condition, the superankle procedure will remain an underutilized and less well known operation than it should be and therefore out of the reach and recommendation to most patients. Purpose of this article is to educate patients and doctors as to its success and details, so that lengthening reconstruction surgery will be offered as an alternative and hopefully a preferable alterative to amputation surgery.