

Multiple Exostoses / Multiple Osteochondroma of the Lower Limb Guide

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Located on the MHE Research Foundations website is a companion educational videos, Biology of Limb Length, Limb Lengthening Methods by Dr. Paley

Multiple Osteochondromas

There are a variety of problems related to the exostoses of Hereditary Multiple Osteochondromas. The majority of these problems relate to bothersome bony protrusions with their affect on surrounding joints, muscles, tendons, nerves, blood vessels and skin. Osteochondromas can also affect growth plates and lead to limb deformities and length discrepancies. The focus of this article will be on the limb deformities and discrepancies secondary to the multiple osteochondromas.

Lower Limb

Osteochondromas are believed to bud off the growth plates. The cartilaginous cap of the osteochondroma has the same structure as the growth plate. It grows in length and width in the same fashion as a growth plate leads to growth in length and width of the end of a bone. For reasons unknown some osteochondromas tether the growth of the growth plate when they bud off. This can lead to asymmetric growth (less growth on the osteochondroma side and more growth on the opposite side of the growth plate) and consequently limb deformity. This tethering effect can also decrease overall limb growth leading to a shorter final limb length than expected. If the opposite lower limb is not as affected then the result is a lower limb length discrepancy (LLD). Although both lower limbs often appear to be equally affected by osteochondromas, LLD is not uncommon indicating that one side is more tethered at the growth plate then the other.

The tethering effect of the osteochondroma on growth is directly related to the size of the growth plate it came from. The larger the growth plate the less effect the osteochondroma has on longitudinal growth because the force of growth in the remaining healthy part of the growth plate is so great. The smaller the growth plate the greater is the tethering effect since the percent of the growth plate involved is so great. Good examples of this are the fibula in the lower limb and the ulna in the upper limb. We shall discuss the ulna in a future article. In the lower leg where there are two adjacent bones (tibia and fibula), a osteochondroma tethering the growth of one bone and not the other will lead to a deformity since

the two bones are attached together. Therefore if the fibula is growing slower than the tibia the leg will grow towards the fibula. This leads to a valgus deformity (knock-kneed) of the upper tibia and a valgus deformity of the ankle (tilted outward). Osteochondromas between the tibia and fibula can also lead to deformity of the adjacent bone. For example an osteochondroma of the distal tibia can lead to deformity of the adjacent fibula near the ankle.

Osteochondromas of the distal femur (lower end of femur near the knee), do not typically lead to any deformity or length discrepancy on their own. They protrude into the surrounding soft tissues and can lead to symptoms related to soft tissue impingement due to their bulk. On occasion they do lead to deformity of the knee which is related to the tethering of soft tissues and not to bony deformity. For example osteochondromas around the knee can lead to locking and flexion deformity of the knee joint (The knee joint catches in a certain position and will not straighten out)

Osteochondromas of the upper femur sprout off the femoral neck. Depending on the direction they come from they lead to different problems. Commonly they lead to asymmetric growth of the neck of the femur resulting in a valgus femoral neck (more vertical than usual). This is usually not a problem. Valgus of the neck of the femur is usually symmetric and therefore does not lead to a leg length discrepancy.

When the osteochondroma is too near the hip joint or if it expands the capsule of the hip joint, this can result in a hip joint contracture or subluxation. The typical hip joint contracture is fixed flexion deformity of the hip from an anterior osteochondroma. Patients present walking leaning forward with hyperlordosis of the spine (sway back) as they try and compensate for the leaning forward effect of the hip by arching their back.

Subluxation of the hip occurs due to the effect of the osteochondroma pushing the hip out of joint combined with the effect of the valgus of the femoral neck.

Treatment of the Lower Limb Deformities.

Valgus Knee Deformity (Knock knee deformity)

This deformity is usually in the upper tibia. There is usually a large osteochondroma involving the upper end of the fibula. The fibular osteochondroma often tethers or envelops the peroneal nerve. This is a very important nerve that is responsible for controlling the muscles that pull the foot up and out. Injury to this nerve results in a drop foot (inability to pull the foot up). Correction of the valgus deformity of the upper tibia requires an osteotomy (bone cut) of the upper tibia. All osteotomies of the upper tibia to correct valgus stretch the peroneal nerve even in patients without HME. In patients with HME and a fibular exostosis

the nerve is very tethered and stretched even before surgery. The nerve can actually be inside the bone if the osteochondroma envelops it. Therefore to correct the deformity safely the nerve must first be found above the fibula and decompressed around the neck of the fibula. The osteochondroma of the fibula should be resected. If the upper fibular growth plate is considered to be damaged beyond recovery then a segment of the fibula should be removed so that the two ends of the fibula do not join together again to prevent re-tethering of the tibia.

Only after all of this is performed can an osteotomy of the tibia be carried out safely to correct the valgus deformity. The valgus deformity can either be corrected all at once or gradually. Correcting it all at once is usually performed by taking out a wedge shaped piece of bone and then closing the wedge to straighten the tibia. This can be fixed in place with a metal plate or with an external fixator. Gradual correction is carried out by minimal incision technique to cut the bone. The correction is achieved by use of an external fixator. This is a device that fixes to the bone by means of screws or wires that attach to an external bar or set of rings.

Adjustment of the external fixator slowly corrects the deformity. This opens a wedge instead of closes a wedge of bone. This has the advantage of adding length to the leg which if the leg is short already is advantageous. This type of external fixator is also used for limb lengthening. Therefore if there is a LLD the angular correction can be performed simultaneous with lengthening. Gradual correction is safer than acute (all at once) correction for correction of the valgus deformity. Another way to address the valgus knee deformity without addressing limb length discrepancy is hemi-epiphyseal stapling of the growth plate. This is perhaps the most minor procedure possible and involves insertion of one or two metal staples on the medial side (inside) of the growth plate of the upper tibia. The metal staple straddles the growth zone on the medial side preventing growth of the medial growth plate while permitting growth on the lateral side. This allows the tibia to slowly autocorrect its alignment. It is a very slow process and may require several years. Once the tibia is aligned the staple can be removed permitting resumption of growth from the medial side. There is a small risk of damaging the medial growth plate which could lead to a varus bowing deformity of the tibia. Stapling can also be used in the distal tibia to correct the ankle deformity.

Valgus deformity of the ankle

Patients complain of walking on the outer border of the foot. Viewed from behind this posture of the foot is very apparent. This deformity is often well tolerated. The lower end of the tibia tilts outwards towards the fibula. The lower end of the fibula is the lateral malleolus. It is important for stability of the ankle. Since the fibula grows less than the tibia the lateral malleolus is often underdeveloped

leading to lateral shift of the talus (ankle bone). This can eventually lead to arthritis of the ankle. Lateral tilt of the ankle joint is compensated by the subtalar joint (joint under the ankle) by inversion of the foot (turning of the foot in). Since this is a longstanding process the subtalar joint becomes fixed in this position of compensation for the ankle joint. Therefore if one tries to fix the ankle joint tilt completely the foot will end up tilted inwards and the patient will be standing on the outer border of the foot. Therefore one either has to accept the valgus ankle or correct it together with the subtalar joint fixed deformity. This is best done with a circular external fixator (Ilizarov device). This correction involves gradual correction of a minimally invasive osteotomy of the lower tibia and fibula together with distraction (pulling apart) of the subtalar joint contracture.

Flexion deformity of the knee

This deformity is usually related to tethering or locking of the soft tissues around the knee by distal femoral or proximal tibial osteochondromas. The treatment involves resection of the offending exostosis and lengthening of the hamstring tendons if needed.

Flexion deformity of the hip / subluxation of the hip / valgus upper femur.

This is treated by resecting the offending osteochondroma of the femoral neck. This hip capsule has to be opened to access these. At the same time to reduce the hip subluxation (hip coming out of joint) a varus osteotomy of the upper femur should be done (bending the femur inwards towards the joint). The bone can be fixed either by an internal metal plate or an external fixator.

Limb Length Discrepancy

Limb length discrepancy under 2cm is usually not noticeable and does not require treatment. LLD over 2cm is usually noticed by the individual affected leading to self compensation by walking on the ball of the foot (toe down) or by tilting the pelvis and curving the spine (scoliosis). Untreated LLD can lead to lower back pain, and long leg arthritis of the hip. These take many years to develop. Individuals who compensate for LLD by walking on the ball of the foot often develop a tight Achilles tendon. The easiest way to treat LLD is by using a shoe lift. I generally prescribe a shoe lift one cm less than the LLD. Shoe lifts of up to 1cm can be easily accommodated inside a shoe. Greater than 1cm should be added to the outside of the shoe. Wearing a shoe lift prevents problems of the back, hip and ankle from developing. LLD can also be equalized surgically. This can be done by either shortening the long leg or lengthening the short leg. In children shortening the long limb is achieved by surgically closing the growth plate of the lower femur or the upper tibia prematurely (epiphysiodesis).

This is a small minimally invasive procedure with few complications. The accuracy of this method depends on the ability of the surgeon to predict the LLD at maturity and the rate of growth of the long limb. The accuracy of LLD equalization with this method is $\pm 1\text{cm}$. After growth of the skeleton has ceased (skeletal maturity) epiphysiodesis is no longer an option. Shortening in adults is carried out by removing a segment of the bone and fixing the bone in place with a metal rod that is inserted into the marrow cavity (locked intramedullary nail). In the femur this procedure can be done through very small incisions, and shortening up to 5cm (2 inches) can be safely achieved. In the tibia this procedure requires bigger incisions and has greater risk and is usually limited to 3cm (1.25 inches).

Lower limb lengthening is the other way to correct LLD and can be carried out in both children and adults and at almost any age. To lengthen a limb the bone is cut through a very small incision (1cm) and then the two ends of the bone are pulled apart at a gradual rate of 1mm/day (1/25 inch/day). Since bone is a living substance it grows new bone to repair the break. By pulling the bone apart at a gradual rate, we prevent the bone ends from joining together. Instead new bone is formed in the growing gap between the bone ends. Once the desired lengthening is achieved the bone is held in place until it joins together. The new bone that was formed in the gap becomes stronger as calcium accumulates in it.

Eventually this new bone achieves the strength of normal bone. There are various devices that are used for limb lengthening. The majority of these are external fixators. An external fixator is an external frame or brace that attaches directly to the bone by means of thin (1.8mm- 1/16") tensioned wires or thicker (6mm- 1/4") screws (half-pins). The frame of the fixator is either shaped like a bar (monolateral fixator: e.g Orthofix, EBI, Wagner, monotube) or has rings and arches (circular fixator: Ilizarov, Taylor Spatial Frame, Sheffield). More recently these systems have become hybridized and have elements of both monolateral and circular fixators.

The circular fixators can be attached to the bone by means of wires that go from one side of the limb to the other passing through the skin on one side, then through the bone and then exiting the skin on the other side. Wires have much smaller diameters than half-pins and achieve their strength by being tensioned across the ring, like tensioning a guitar string. Half pins are of much larger diameter and only pass through the skin on one side. They fix to the bone by means of a screw-like thread. To lengthen the limb the fixator has a screw mechanism which allows for small adjustments that pull the bone apart. The bone is pulled apart because the fixator which is attached to the bone above and below the break in the bone, lengthens as the screw mechanism is turned. The typical lengthening rate is 1/4mm, 4 times a day, for a total of 1mm/day. There is even a motorized attachment which can be used for lengthening (Autogenesis). This

lengthens at the same rate of 1mm/day divided into hundreds of small lengthenings. This may reduce the pain of lengthening. It is also more gentle on the soft tissues (nerves, muscles) that must stretch and grow as the bone is pulled apart.

The most common complication and care with external fixator lengthening is superficial pin infection. (Refer to fixator care guide) This minor complication is to be expected. It is also easily treated by taking oral antibiotics at the first sign of infection (redness, tenderness, and drainage around a pin site). Deeper infection of the soft tissues and bone is quite rare, but if it occurs usually requires removal and possible replacement of the problem pin, IV antibiotics and sometimes surgery to debride (remove dead tissue) the soft tissue and bone. Other complications include tightness of muscles which can limit the range of motion of the adjacent joints or even pull the adjacent joints into a fixed position that interferes with function (e.g. equinus contracture of the ankle (fixed toe down position) is due to tightness of the Achilles tendon that develops during lengthening).

To prevent problems with joints and muscles it is essential to do daily range of motion and stretching exercises with physical therapy, and to maintain that stretch by using foot or knee splints. Sometimes it is necessary to either immobilize a joint by extending the external fixation across the joint to hold the joint in a functionally good position (e.g. foot fixation at 90° with tibial lengthening to prevent equinus). In some cases it may be necessary to surgically lengthen some of the tendons or fascia to prevent or treat contractures (e.g. Achilles tendon lengthening). Bone complications can also occur. These include too rapid or too slow bone formation. Too rapid formation (premature consolidation) can prevent further lengthening and requires rebreaking the bone to continue lengthening. To prevent this, the lengthening rate may have to be increased. Poor bone formation can also occur (delayed consolidation). This requires more time in the external fixator until the bone is fully healed. Complete or partial failure of bone formation leads to a bone defect and may require a bone graft to get the bone to heal.

There are two phases to the lengthening process. The first is the distraction phase when the bone is being pulled apart at one mm per day. The second is the consolidation phase when the bone is hardening while it is being held in place by the external fixator. The fixator cannot be removed until the bone is completely healed. If the fixator is removed before that time the bone will bend, shorten and/or break. The best way to tell if the bone is fully healed is by x-ray. Even with x-rays it is not uncommon to misjudge the strength of the bone and remove the fixator prematurely. In many cases we apply a cast for an additional month of protection to minimize the risk of refracture. It is better to leave the fixator on an extra month than to take it off a day too early. Patients are often impatient at this stage and push their doctors to take the frame off. An experienced limb

lengthening surgeon turns a deaf ear to these frustrations and refuses to remove the frame until the x-rays suggest that the bone is strong enough that it will not break or bend upon removal. Most of the complications of lengthening occur during the distraction phase or after removal. Few complications other than pin infection arise during the consolidation phase.

External fixator lengthening has been the standard for the past one hundred years of the history of limb lengthening. In the past decade internal lengthening devices have emerged. These permit gradual lengthening by means of a fully implantable telescopic intramedullary rod (a metal rod that fits inside the marrow cavity of the bone). While there are several of these devices in use worldwide, there is only one at present FDA approved in the USA. This is called the Intramedullary Skeletal Kinetic Distractor (ISKD). It is manufactured by Orthofix, Inc. At present it is on a limited release with only a small number of surgeons trained to use it and of those only a few centers with a large experience with its use. This device can only be used in patients who are skeletally mature and therefore is not applicable in growing children. It is also limited in its ability to correct deformities.

Nevertheless it eliminates all of the problems related to the pins of the external fixator, especially pin site infections, scars and pin site pain. It also reduces the muscle tethering from the pins and makes the physical therapy easier. The ISKD does present some new problems not experienced with external fixator lengthening. There is less control of the lengthening rate and rhythm which can lead to contractures, nerve problems and bone healing problems. In the femur there is a higher rate of premature consolidation while in tibia there is a higher rate of delayed consolidation. Some patients experience severe pain at the onset of lengthening and require an epidural for several days until this pain goes away. All in all however we consider this a major advance. We have performed over 50 such surgeries with good success. None have been for MHE. Deciding between lengthening and shortening is based on a few factors. Shortening is only applicable for discrepancies less than 5cm. Shortening is a much smaller procedure while lengthening is a bigger procedure and longer treatment. Lengthening has a higher complication rate. Shortening cannot correct deformity on the short leg. Lengthening can simultaneously correct deformity and length discrepancy.

Shortening will decrease the patients height by the amount of shortening (max 5cm : 2 inches). Lengthening does not decrease height. Therefore in someone with less than 5cm of LLD and no deformity who is not short or concerned about the height loss, epiphysiodesis or shortening are good alternatives for equalization or LLD. Most cases do have associated deformities and therefore our preference is to perform one operation to simultaneously correct the LLD and the deformity at the same time.