

Multiple Hereditary Exostoses

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Multiple hereditary exostoses (MHE), also known as multiple osteochondromas (MO), is an autosomal dominant skeletal disorder. Approximately 10–20% of individuals are a result of a spontaneous mutation while the rest are familial. The prevalence of MHE/MO is 1/50 000.

There are two known genes found to cause MHE/MO, *EXT1* located on chromosome 8q23-q24 and *EXT2* located on chromosome 11p11-p12. In 10–15% of the patients, no mutation can be located by current methods of genetic testing.^{1–7} These mutations are scattered across both genes. *EXT1/EXT2* is essential for the biosynthesis of heparan sulfate (HS). HS production in patients' cells is reduced by 50% or more.^{8–19} MHE/MO is associated with characteristic progressive skeletal deformities of the extremities and shortening of one or both the sides, leading to limb length discrepancy (LLD) and short stature.^{20–28} Two bone segments, such as the lower leg or forearm, are at greater risk of problems due to either osteochondromas (OCs) from one or both bones impinging on or deforming the other bone or a primary issue of altered growth causing one bone to grow at a faster or slower rate. OCs can also affect joint motion due to impingement of an OC with the opposite side of the joint or subluxation/dislocation related to deformity, impingement, and incongruity.^{20,24,26,29,30}

OCs can also cause nerve or vessel entrapment and/or compression, including the spinal cord and nerve roots. Tethering of tendons and muscles can lead to locking or restriction of range of motion of joints as well as pain. Finally, chronic pain, arthritis, and disability are associated conditions.³¹

Finally, the risk of malignant degeneration to chondrosarcoma has been reported in multiple publications to range between 1 and 5% of patients with MHE.^{7,8,17,20,26,32–38}

► MEDICAL TREATMENT

Since MHE is a single-gene disorder affecting the biosynthesis of a specific molecule, HS, medical treatment should be possible. HS impacts many signaling pathways in bone and cartilage, including FGF, Wnts, Ihh, BMP, TGF β , and SH2. Each of these pathways is a possible target for drug development. Since BMP signaling is critical for cartilage and bone formation, it may be the ideal therapeutic target.^{12,19,39–49}

Palovarotene (PVO) is a retinoic acid receptor γ selective agonist. Its safety profile was established in an emphysema trial. PVO was identified as an effective treatment for fibrodysplasia ossificans progressiva (FOP) by Pacifici and collaborators. In FOP, PVO acts as a suppressor of BMP signaling. Thus, PVO was predicted to be effective in the treatment of MHE/MO as well.^{45,50–54}

Animal studies demonstrated reduced OCs in a mouse model.⁵¹ In 2018, a Phase 2 clinical trial was begun on the use of this in humans. This is a multicenter, randomized, placebo-controlled company-sponsored study. The results are as yet unknown [*An Efficacy and Safety Study of Palovarotene for the Treatment of MO (MO-Ped) NCT03442985*].

► LOWER EXTREMITY

MHE can affect any of the bones in the lower extremity (pelvis, femur, tibia, foot bones). The indications for surgical treatment are numerous and are related to age, OC size and location, effects on joint range of motion, deformity and stability, alignment and length discrepancy, and nerve entrapment and pain.^{20–26,28,31,55,56}

Children with MHE will often require numerous OC removal. It is important to limit the number of surgeries so as to minimize the interruption of childhood.^{28,57–60} To this end, the indication for removal of OCs may be softened to include ones that are not yet symptomatic but are expected to become so. This way, the removal of numerous OCs is often combined with other reconstructive procedures, including osteotomies, hemiepiphysiodesis, lengthening, and nerve decompression. One should avoid repeated single OC surgery that interrupts childhood. When both upper and lower limb problems need to be addressed, one should consider doing all of the upper limbs at one time and all of the lower limbs at another time.

For the purpose of instruction, specific problems at each area will be discussed separately, even though many of these procedures may be combined into one more comprehensive surgery.^{28,57–63}

► HIP

MHE affects both sides of the hip joint, although the femur is more often primarily affected, which then leads to secondary dysplasia in the acetabulum^{20,24,64,65} (Figs. 1a,b, 2a-d).

Computerized tomography (CT) as well as magnetic resonance imaging (MRI) are useful modalities to evaluate the hip. A three-dimensional (3D) CT study is the most helpful in understanding the location of the offending OCs.^{66–70} These are most often located on the femoral neck but can also be present on the outside or inside of the acetabulum. OCs in this region lead to limitation of joint motion and pain, both due to impingement. They can protrude posteriorly and entrap or irritate the sciatic nerve. The femoral neck tends to grow into a valgus position. In some cases, the neck shaft angle can reach 180°. ⁶⁸ One should look for a break in Shenton's line as well as an enlargement of the medial joint space, both indicators of hip joint subluxation. Subluxation may be related to one or more of the following: coxa valga, medial femoral OCs, and intra-acetabular OC.⁷⁰ A 3D CT will help

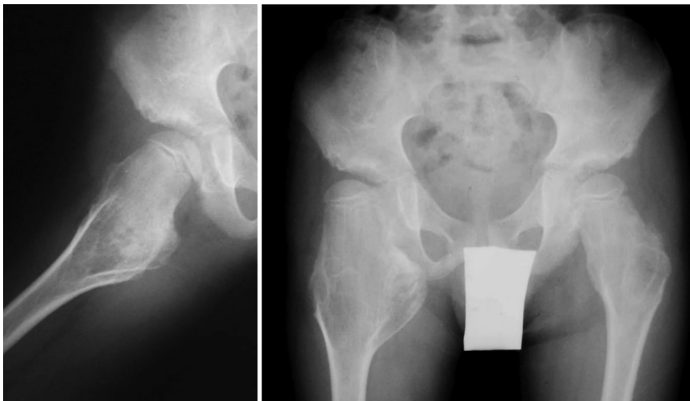


Figure 1a

AP (right) and frog lateral (left) radiographs of right femur showing large medial proximal femoral osteochondroma in an 8 year old boy with marked coxa valga.

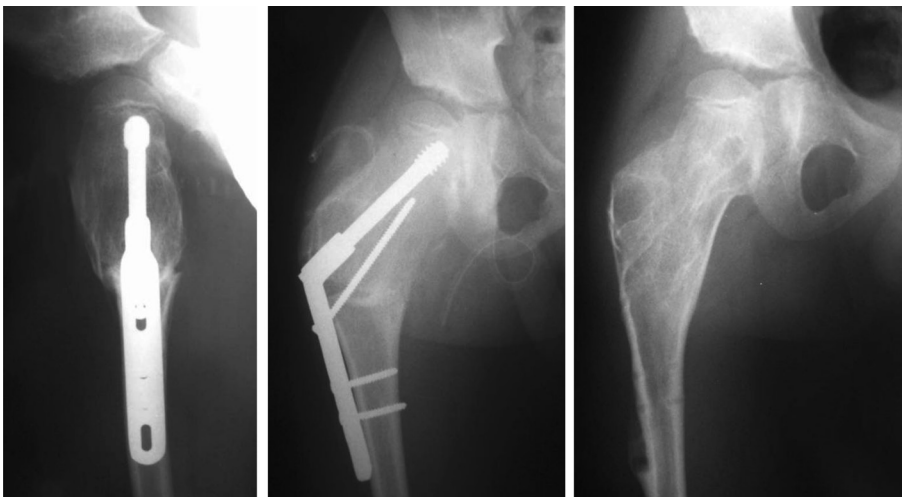


Figure 1b

Frog lateral (left) and AP (center) radiographs after osteochondroma excision through lateral approach without surgical dislocation combined with varus proximal femoral osteotomy and fixation with sliding hip screw device. The final AP radiograph of the hip after hardware removal (right) is shown.



Figure 2a

AP pelvis radiograph of both hips in a 30 year old man with MHE. He has large osteochondromas emanating from both proximal femurs limiting his hip range of motion.



Figure 2b

3D CT scan of pelvis and both hips showing details of the three dimensional location of these osteochondromas.

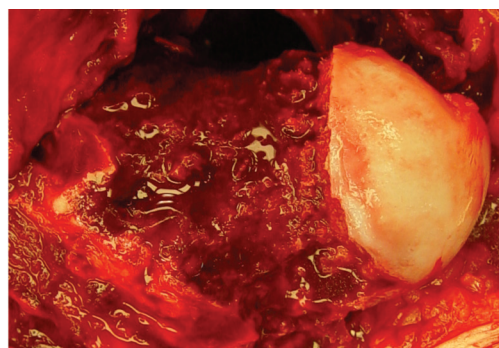
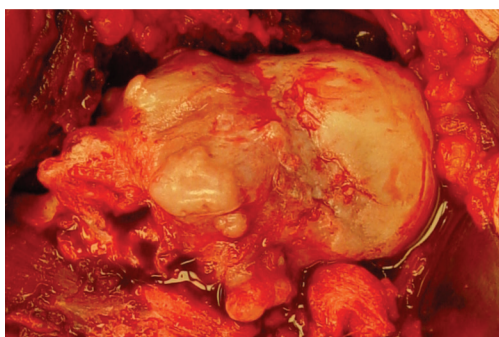


Figure 2c

Intraoperative photograph of the right hip after safe surgical dislocation (left). Note the cauliflower like exostoses emanating from the femoral neck and head. The osteochondromas were resected circumferentially around the femoral neck without damaging the retinaculum of the femoral neck and the blood supply of the femoral head (right).

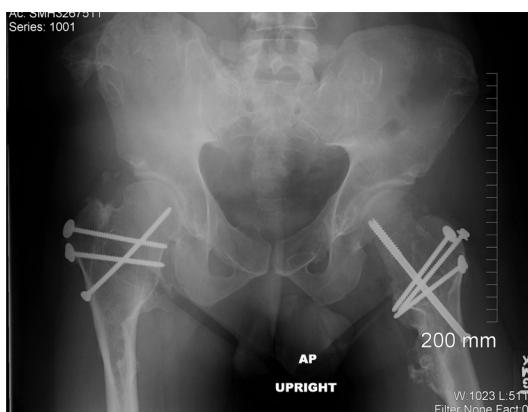


Figure 2d

AP pelvis radiograph showing complete resection of the osteochondromas from both femoral necks. The femoral necks were protected from stress fracture with a screw. The greater trochanters were fixed back in place with screws. The range of motion of the hips improved bilaterally. The two surgeries were performed three months apart.

identify which of these is the contributory cause. Associated genu valgus also exacerbates the coxa valga and lack of femoral head coverage during stance and gait.

Coxa valga without subluxation can be watched until it exceeds 150°. As the neck shaft angle increases beyond this amount, the tendency for the acetabulum to become dysplastic increases. It is preferable to treat the hip in MHE before the acetabulum becomes dysplastic to avoid the need for a pelvic osteotomy. Coxa valga should be treated by a varus intertrochanteric osteotomy at the level of the lesser trochanter to minimize the amount of medial translation required.⁶⁴ The center of rotation of angulation (CORA) of the coxa valga is at the level of the base of the lesser trochanter. Therefore, an osteotomy through or above the lesser trochanter is almost at the same level as the CORA, while an osteotomy below this level requires medial translation to maintain the colinearity of the piriformis fossa with the mid-diaphysis of the femur.⁷¹ If there are non-impinging OCs of the calcar region, these will move away from the pelvis with the varus osteotomy. If there are associated impinging OCs of the femoral neck or calcar, a safe surgical dislocation approach combined with the varus osteotomy is performed.

Limitation of hip range of motion, femoroacetabular impingement, and associated nerve irritation due to posterior OC of the proximal femur are treated by the Ganz safe surgical dislocation method. This involves a greater trochanteric osteotomy to flip the gluteus medius–minimus and vastus lateralis anteriorly while leaving the piriformis tendon behind to protect the ascending branch of the medial femoral circumflex vessels.^{68,72–75} A Z-shaped capsulotomy is performed and the femoral head is dislocated from the joint. The acetabular labrum is probed for tears and if present these are debrided and repaired. Any intra- or periarticular OCs of the acetabulum are removed. The OCs on the femoral head, neck, and calcar are removed with an osteotome. An osteochondroplasty is done to recreate the normal anterior superolateral offset of the femoral neck. In distinction from the more common cam lesions, the removal of the bone from the neck is much more extensive. One has to be careful not to damage the posterior retinaculum to avoid avascular necrosis. A prophylactic screw is inserted into the femoral neck to prevent fracture since the femoral neck is significantly weakened by this almost circumferential removal of the surrounding OCs. The redundant hip capsule is plicated after the femoral head is reduced in joint. The greater trochanter is replaced to its original location and fixed with screws. A varus femoral osteotomy is performed if the neck shaft angle is 150° or greater. If an intertrochanteric osteotomy is performed, it is fixed with a 130° locking plate with screws or with a 130° blade plate.

Postoperatively, an assessment and decision need to be made whether there is sufficient coverage of the femoral head. If the acetabulum is too dysplastic, then a periacetabular osteotomy should be performed. In children below the age of 8, it is preferable to avoid the hip dislocation so as not to disrupt the greater trochanteric apophysis. In most of these children, there is marked coxa valga but limited medial OCs. A surgical dislocation can be deferred to an older age.^{68,72–76} Isolated varus osteotomy is performed.

► KNEE

Frontal plane alignment is the most common problem at the knee (Figs. 3, 4). There is a tendency of the tibia to grow into valgus. This is often related to osteochondromatous involvement of the upper fibula, suggesting that there is lateral tethering of growth of the tibia. The simplest way to treat this problem is using a medial hemiepiphysiodesis plate.^{62,63,77} To confirm that the deformity is coming from the tibia and not from the femur, the Paley Malalignment test should be performed. This involves measuring the lateral distal femoral angle and the medial proximal tibial angle. Based on these, one can determine whether the valgus is coming from the femur or tibia or coming from both.⁷⁸ If there is significant OC involvement of the proximal fibula, the peroneal nerve should be decompressed at its two tunnels and the OCs of the proximal fibula removed. This includes an anterior and lateral compartment transverse fasciotomy.^{61,79–83}

Sagittal plane malalignment is also common but less clinically relevant. Hyperextension deformity of the distal femur is often present and rarely needs surgery. In contrast, flexion deformity of the distal femur leads to fixed flexion of the knee and requires osteotomy.

OCs of the distal femur and proximal tibia are often large, symptomatic, and prominent. Medial tibial OCs can lead to entrapment of the pes anserinus tendon and catching of the knee joint during flexion and extension. The femoral vessels are often in contact or displaced by medial distal femur OCs. Lateral distal femur OCs can entrap the iliotibial band.

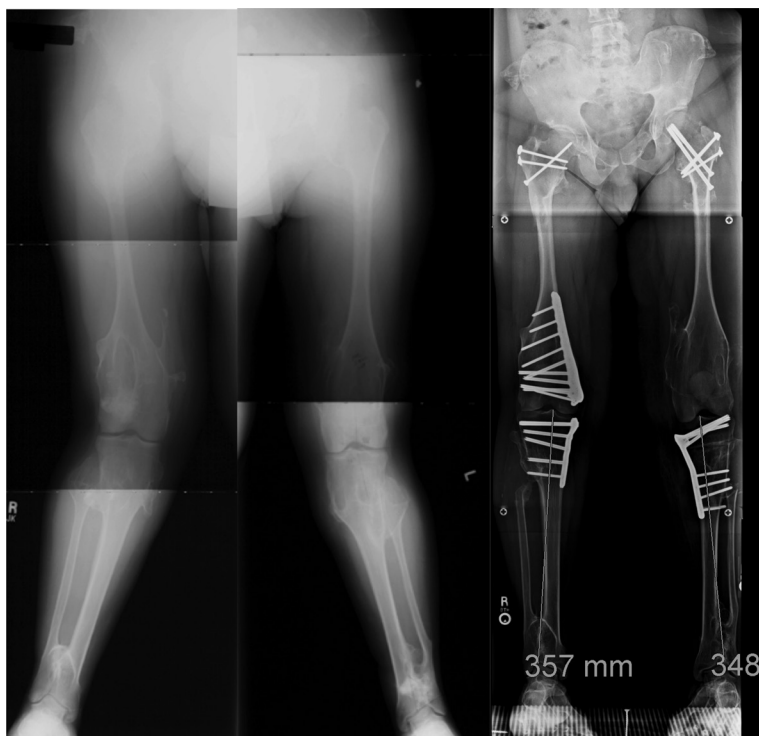


Figure 3

Standing long radiographs of both lower limbs in the previous patient showing bilateral genu valgum in addition to the hip osteochondromas (left). Standing radiographs after bilateral proximal tibial and right distal femoral osteotomy for realignment. The peroneal nerves were decompressed bilaterally together with resection of osteochondromas.

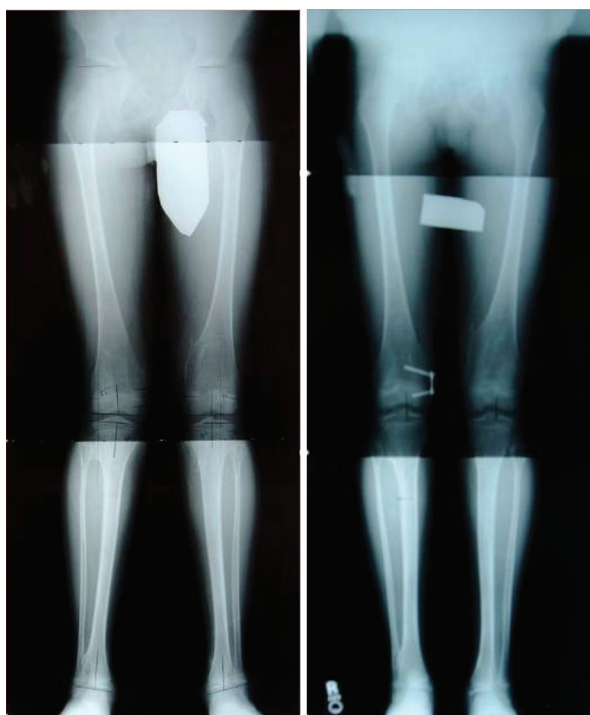


Figure 4

Preoperative standing AP radiographs of right genu valgum in an MHE skeletally immature patient (left). Postoperative standing radiograph a year and a half later after correction of genu valgum with hemiepiphysiodesis with a screw plate device (right). The right lower limb has achieved normal alignment.

Anterior distal femoral OCs can affect the patella and quadriceps mechanism. Posterior distal femur OCs can limit knee flexion range of motion due to impingement with the posterior tibia with knee flexion. This will resolve itself in most cases as the OC grows away from the joint. Posterior proximal tibial OC can displace the vessels or nerves depending on its location. When it is posterolateral, it can collide with the fibula. Kissing tibiofibular OCs are common both proximally and distally. Removal of femoral and tibial OCs requires careful, extensile anatomic exposure with special attention to the neurovascular structures at risk. It is often safer to remove an OC piecemeal rather than try and resect the entire exostosis as one piece. The cartilage cap should always be removed in its entirety since it contains the growth plate for enlargement of the OC. To prevent bleeding, bone wax can be pressed onto the base of the resected bone.

Neurovascular structures should be identified proximal and distal to the OC and then carefully teased off of the OC to avoid injury. After resection of the OC, there is a soft tissue dead space. This space is at risk of hematoma accumulation. A drain should be inserted into this dead space just before wound closure. The drain is left in until it is almost dry before it is pulled.

► ANKLE

Valgus deformity at the ankle is very common in MHE.^{20,84–90} This deformity may manifest in four stages (Figs. 5a-c, 6a-c, 7a-c)).

Paley–Feldman Classification of MHE Ankle Valgus (Fig. 8a)

Type 1: Valgus plafond (sometimes the distal epiphysis may be wedged), no talar shift, fibula at station.

Type 2: Valgus plafond, lateral talar shift, fibula migrated proximal.

Type 3: V-shaped distal tibial epiphysis, lateral talar shift, fibula migrated proximal.

Modifiers

Interosseous OCs distal tibia and fibula: present versus absent.

Subtalar motion: mobile versus fixed varus.

Ankle degenerative changes: absent versus present.

Treatment of Type 1 ankle: These cases can be easily treated by medial distal tibial hemiepiphyseodesis in the growing child or by supramalleolar osteotomy of the distal tibia and fibula in the skeletally mature individual.⁹¹

Treatment of Type 2 ankle: This requires a procedure called the shortening osteotomy realignment distal tibia (SHORDT).⁹² The distal fibula is proximally migrated in

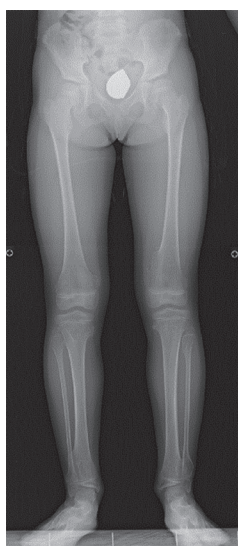


Figure 5a

Standing AP radiograph showing bilateral ankle valgus with fibula at station (distal fibular physis at level of ankle joint). This is classified as Paley-Feldman type 1.

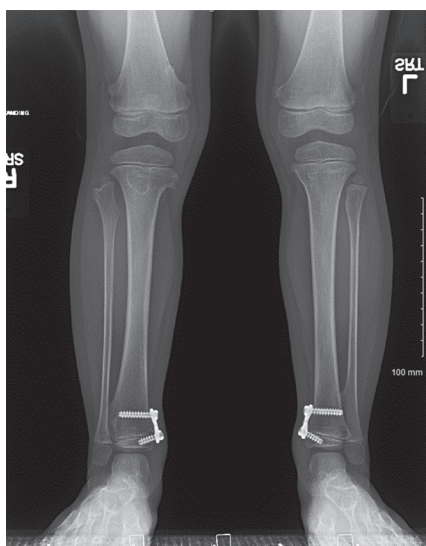


Figure 5b

AP bilateral tibial radiographs showing that the valgus of both plafond was corrected using hemiepiphyseodesis with screw plate devices. The fibulas remain at station.

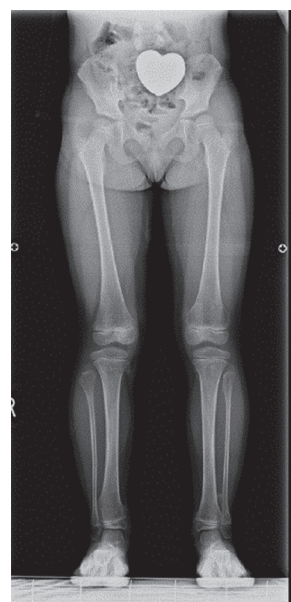


Figure 5c

Post correction and post removal AP standing radiograph of both ankles showing maintained correction a year later.

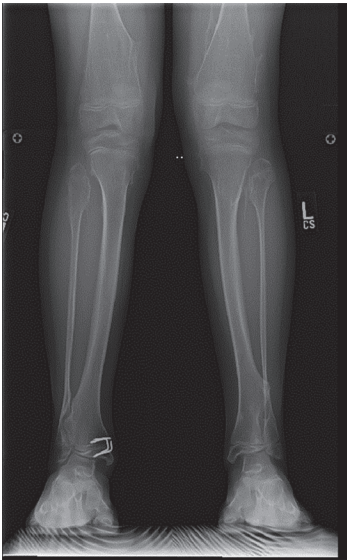


Figure 6a

Standing AP radiographs showing both ankles in significant valgus deformity. Note the lateral shift of the talus. There is a space created between the medial malleolus and the talus. The fibula is proximally migrated (not at station). Note that its physis is proximal to the ankle joint. The talus always follows the fibula. The right side had a failed attempt at hemiepiphysiodesis with staples. This may have contributed to the intra-articular V shaped plafond on the right side. There are interosseous osteochondromas present bilaterally between the tibia and fibula.

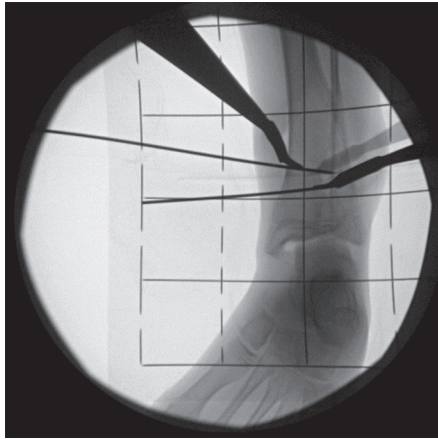


Figure 6b

Intraoperative fluoroscopic view of the left ankle showing the placement of reference wires used for guiding the resection of the trapezoidal segment of tibia. The distal tibiio-fibular ligaments and osteochondromas have already been released and resected respectively.



Figure 6c

Mortis view of the left ankle after varusization and shortening of the distal tibia and fixation with an anterior plate. The distal tibio-fbular syndesmosis was fixed with a ligament washer system. The fibula was brought down to station by shortening the tibia relative to the intact fibula.

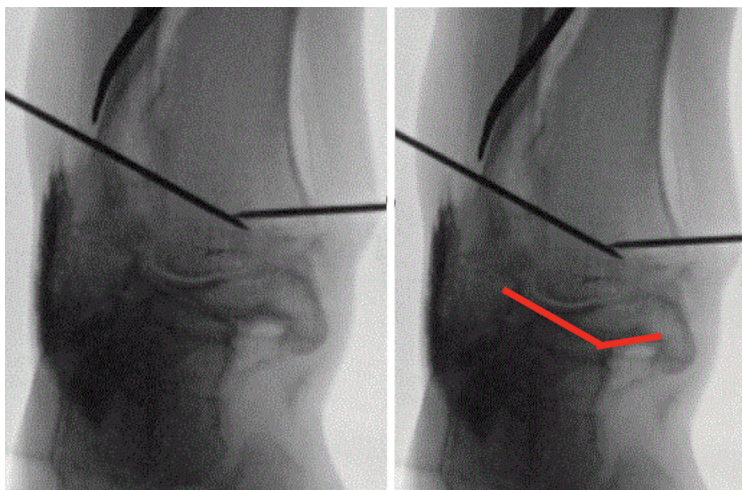


Figure 7a

Intraoperative fluroscopic view of the right ankle in the previous case, showing the pagoda shaped tibial plafond (red lines). Two K-wires were inserted one parallel to the medial and one to the lateral part of the plafond.

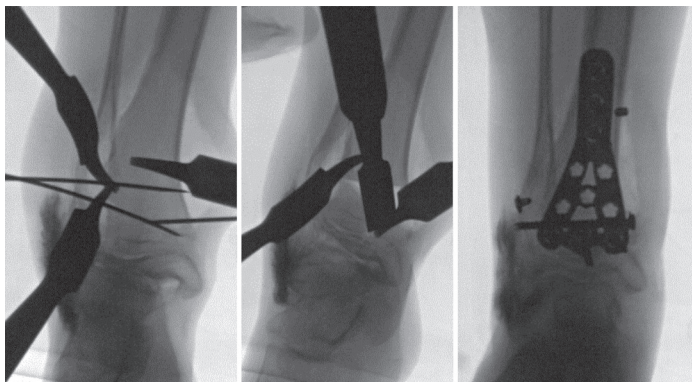


Figure 7b

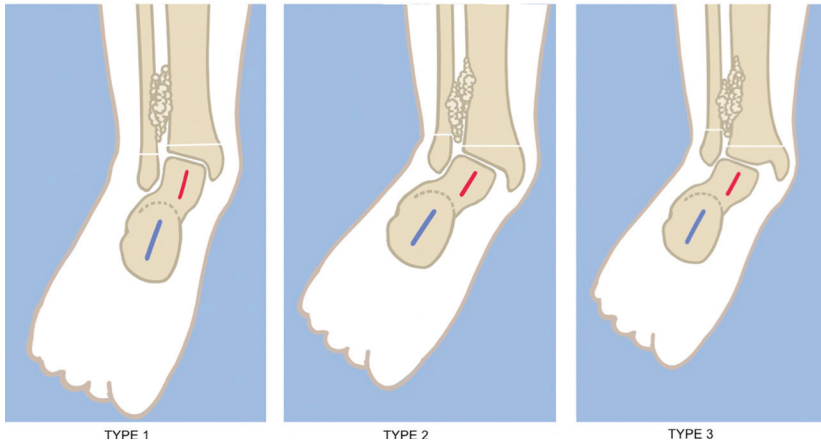
Intraoperative fluoroscopic views: A third wire inserted perpendicular to the proximal tibia shaft at a distance equal to the amount of desired shortening of the tibia to bring the fibula to station (left). An intra-articular osteotomy is made after first resecting the pentagon shaped bone segment (middle). The bone is shortened, the plafond is flattened by opening a wedge between the medial and lateral parts of the plafond. The intra-articular osteotomy does not penetrate the cartilage. The cartilage bends to accommodate the correction. An anterior plate is used for fixation (left).



Figure 7c

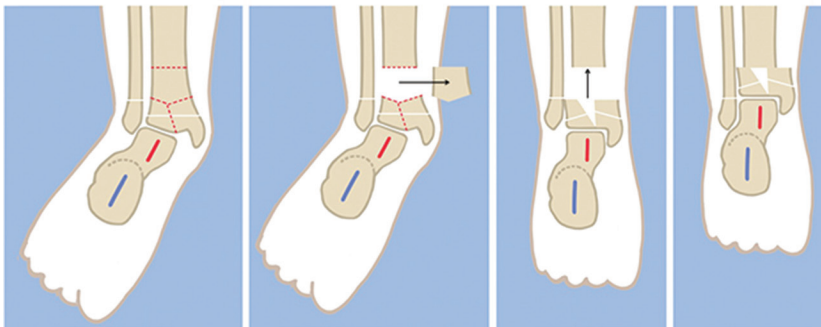
Postoperative standing AP radiographs showing complete realignment of both ankles with both fibulas at station and both tali reduced in the mortis.

PALEY-FELDMAN MHE CLASSIFICATION | ANKLE

**Figure 8a**

Paley-Feldman
MHE classification
of the ankle

PENTAGON OSTEOTOMY

**Figure 8b**

Paley's Intra-
articular Pentagon
Osteotomy

these cases. This position is referred to as the fibula not at station. To restore the fibula to station, the tibia is shortened relative to the fibula. This involves releasing the anterior and posterior distal tibiofibular ligaments combined with a varus supramalleolar osteotomy with shortening of the tibia by the amount of proximal migration of the fibula.

Treatment of Type 3 ankle: These cases require a procedure called the pentagon osteotomy (Fig. 8b). The distal fibula is proximally migrated with the talus following it and tilting into valgus. This leads to valgus deformation of the lateral distal tibial epiphysis. A valgus angle develops between the medial and lateral plafond. The talus articulates only with the lateral plafond. There is a space between the talus and the medial malleolus. The distal tibiofibular ligaments are released. The supramalleolar osteotomy is made in a V-shaped fashion with each limb of the V parallel to its section of the tibial plafond. A second osteotomy is made more proximally for shortening of the tibia to restore the fibula to station. Finally, an incomplete intra-articular osteotomy is added through the apex of the V to level the plafond. In growing children, this osteotomy is through the distal tibial physis. Bone wax is inserted in the opening wedge space to prevent a physeal bridge in the growing child.⁹³

Associated with the valgus ankle deformity types, there are often interosseous OCs between the distal tibia and fibula coming from one or both bones.^{20,24,94–96} The anatomy of this is best studied using a 3D CT scan. These OCs must be resected at the same time as the rest of the treatment. With Types 2 and 3, an anterior incision is made to allow such resection and osteotomy of the distal tibia. An anterior plate and screws are used for fixation.

In order to correct the valgus of the ankle, the subtalar joint must at least be able to move into its neutral position. An examination is done to determine whether the heel can be put into the same amount of valgus as the valgus of the tibial plafond. If the heel cannot evert enough, then there is a fixed subtalar varus contracture, which will limit how much varus correction can be done through the supramalleolar osteotomy. Otherwise, full correction will uncover the varus subtalar contracture, leaving the foot stuck in varus.

The presence of ankle degenerative changes is important. If the ankle degeneration is significant, then osteotomy surgery may not be indicated. An ankle fusion may need to be considered. To avoid degenerative changes, the proximal fibular migration with ankle valgus should be treated aggressively.

► LIMB LENGTH DISCREPANCY

LLD^{24,97} (Fig. 9) is a common problem with MHE. Since growth rate is less predictable in MHE, epiphysiodesis is a less reliable option. Furthermore, short stature is a common



Figure 9

Standing AP radiographs showing a 2cm leg length discrepancy (left). Radiograph of the right side showing lengthening with implantable nail (middle). Radiograph after full bone consolidation (right).

problem since growth inhibition affects both sides, making epiphysiodesis and its associated height loss less attractive. LLD is a result of more inhibition on one side than on the other. Discrepancy can be due to either or both femur and tibia. The preferred method of lengthening now is with an implantable lengthening device. Since the total LLD is usually 5 cm or less, waiting till closer to skeletal maturity is advisable. In cases where the discrepancy is predicted to be much larger and two lengthenings are anticipated, the first lengthening can be around age 8 and the second closer to maturity. Epiphysiodesis is also a consideration for discrepancies less than 5 cm.

► UPPER EXTREMITY

MHE often involves multiple areas of the upper extremities from the scapulae to the tip of the fingers. Specific areas are of concern, may be due to the location consistently causing pain or deformity, while others impinge upon nerves and blood vessels. Joint involvement of the wrist and elbow is quite common. The upper extremity has many unique features in MHE, in that the function of the forearm and wrist is often severely involved. The radiocapitellar joint is particularly prone to dislocation.^{24,29,98–100}

► SCAPULA

The scapula is a common site for OC growth. OC of the posterior (dorsal) aspect of the scapula is often not painful but may become quite apparent and can be large and disfiguring. They may be excised for relief of discomfort or cosmetic purposes. Dorsal scapular OCs are divided into supra- and infraspinous. The supraspinous tumors can entrap or be near the nerves to supra- and infraspinatus. Care must be taken to avoid injury to this nerve and to decompress the nerve at the scapular notch and from around the tumor if necessary. Infraspinous dorsal tumors can be approached directly.^{66,101–106}

Anterior scapula OCs (costal surface) (Fig. 10) are often painful as they contact the ribs when the patients raise and lower their arms. They also cause the appearance of winging of the scapula when they are large. The approach to the costal side of the scapula for excision is determined by location. More medial ones are approached along the vertebral border of the scapula, cutting through the trapezial fascia and then detaching the rhomboid major



Figure 10

3D CT scan views of large right scapular supero-medial osteochondroma

and minor and dissecting under the subscapularis muscle to locate and excise the OC. Care must be taken since the long thoracic nerve can lie on the surface of large protruding OC as you approach the axillary border of the scapula lying on the serratus anterior muscle. Through the vertebral border approach, one can reach almost to the axillary border, which is where the long thoracic and thoracodorsal nerves are at risk.

Lateral scapular OC can also be excised with a lateral incision. Care to protect the thoracodorsal nerve is paramount. The teres major and minor origins on the scapula may be detached with care not to injure the more laterally lying axillary and thoracodorsal nerves as well as the circumflex artery.

The scapula is one bone that is prone to malignant degeneration of tumors (Fig. 11). The vast majority of these are low-grade chondrosarcomas. These require marginal excision of the tumor, but in aggressive and locally recurrent cases, partial or complete scapulectomy may be required. One of the problems with scapular tumors is that the constant movement between the chest cavity and the scapula may break off cartilage rests that form free chondrosarcoma loose bodies in the soft tissues between the scapula and the ribs. These can be seen only on MRI.³³

► HUMERUS

The proximal humerus is a common location for OC growth. While many are asymptomatic and can be left in place, medial proximal humeral OCs (Fig. 12) may cause nerve and arterial and/or venous compression. Ulnar nerve compression is the most common, and in excising these tumors, the nerves and artery exiting the brachial plexus must be visualized and protected.^{20,24,95,107,108}

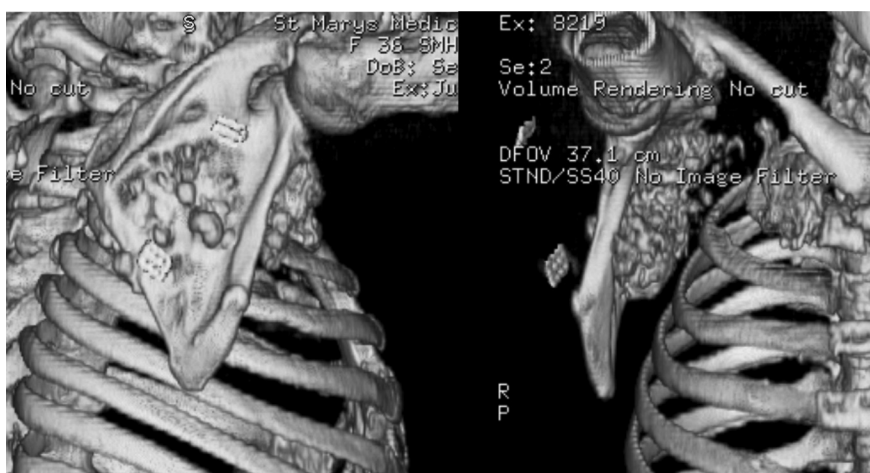
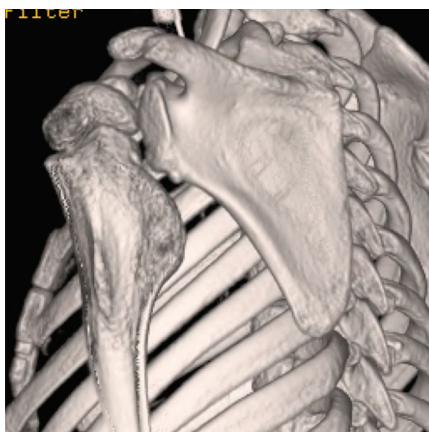


Figure 11

3D CT scan views of costal surface and dorsal surface scapular osteochondromas. The costal surface ones were diagnosed as Grade 2 chondrosarcoma. A scapulectomy preserving the glenoid was later performed.

**Figure 12**

3D CT scan of proximal humerus showing medial osteochondroma that was irritating the brachial plexus.

**Figure 13**

AP radiograph (left) and AP 3D CT (middle) of distal radius showing that the distal radius and ulna are approximately at the same length (Feldman-Paley type 1). There are kissing osteochondromas between the two bones. These were resected freeing up forearm rotation (right).

Excision of large humeral OC may weaken the shaft of the humerus. Prophylactic fixation of the humerus or excision and grafting may be needed (Fig. 14 a-c).

► FOREARM (FIGS. 13-16)

OCs are particularly problematic when they occur in the forearm. The two-bone structure of the forearm makes it prone to problems that do not occur in single-bone segments, such as the humerus.

OCs growing between the radius and the ulna cause disturbance and limitation of supination and pronation. This is mostly a mechanical problem. This can result from a single OC extending from the radius or ulna or kissing OCs extending from both bones toward each other. These OCs should be excised early to maintain or restore supination and pronation. Ulnar lesions are frequently sessile budding off of the distal ulna. This creates the appearance of what is referred to as the candlestick ulna. Pedunculated lesions are also common off of both bones. OCs may also interfere with muscle function and may be excised to allow for untethered muscle excursion.

MHE is associated with differential growth of one bone versus the other in the forearm. This is similar to the relatively short fibula compared to the tibia seen in the leg in these patients. Since these two bones normally grow evenly with each other, this differential growth between the two bones often leads to secondary deformities. Ulna bowing is one of these secondary deformities. The ulna may bow into the anatomic radial bow, narrowing the



Figure 14a

AP and lateral radiographs of right forearm showing shortening without angulation of the ulna. The radial head is well located. There are some distal ulnar and radial osteochondromas. This is classified as Feldman-Paley 2a



Figure 14b

AP radiograph of the forearm showing lengthening of the ulna with a monolateral external fixator.



Figure 14c

AP and lateral radiograph of the forearm after removal of the external fixator.



Figure 15a

AP radiograph of the forearm showing a large single ulnar osteochondroma. There is increased radial tilt with carpal slip greater than 50%. The ulnar is short and has angulation of its diaphysis narrowing the interosseous space. The radial head shows mild subluxation. This is classified as Feldman-Paley type 2b.

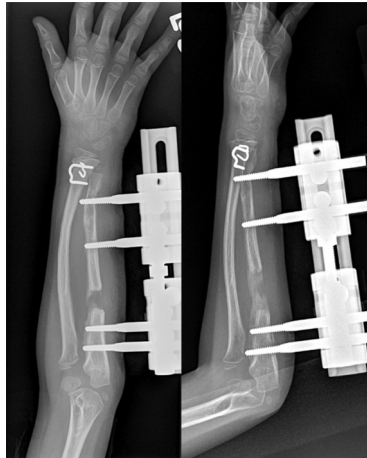


Figure 15b

AP and lateral radiographs showing lengthening of the ulna with a monolateral fixator. The distal radius had a closing wedge osteotomy fixed with two staples.



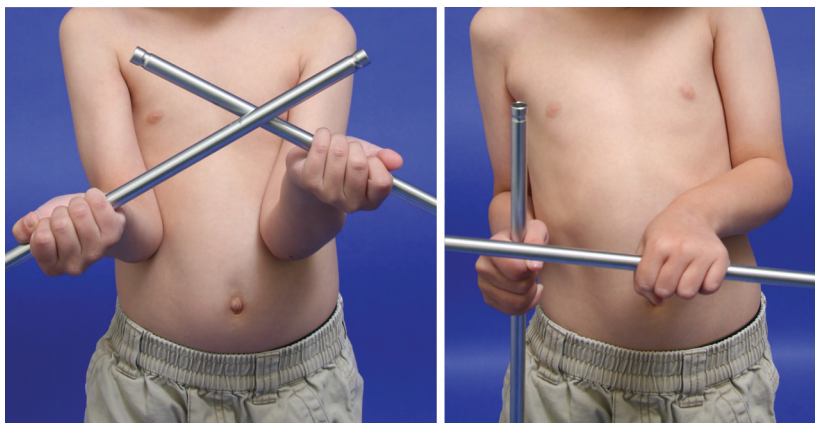
Figure 15c

AP and lateral radiographs of the forearm, 2 years after the procedure. The length of the two bones remains equal and the interosseous space is maintained. The distal radial tilt remains corrected. One of the staples had been removed. The radial head remains well reduced.

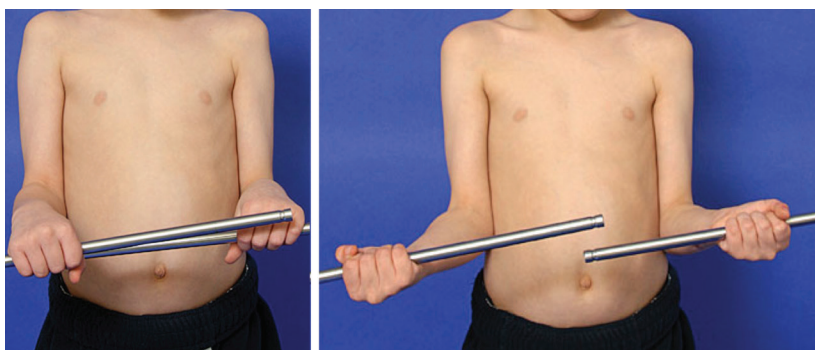


Figure 15d

Preoperative photographs showing full supination and 90° loss of pronation of the right forearm.

**Figure 15e**

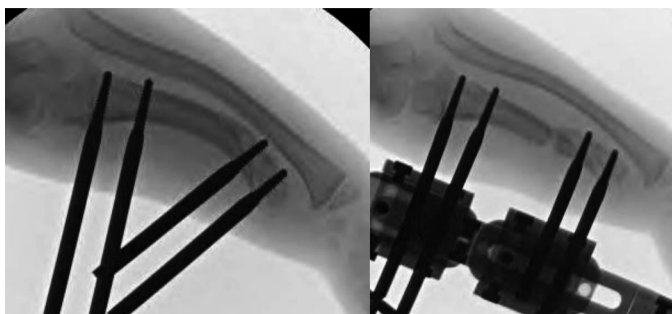
Postoperative photographs a year after surgery showing full supination and pronation.

**Figure 15f**

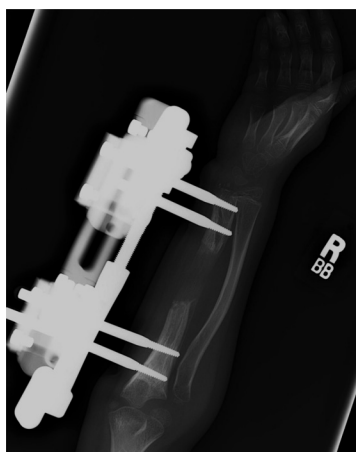
AP and lateral radiographs 5 years after surgery showing that the radius and ulna maintain normal anatomic relations.

**Figure 16a**

Photograph and radiograph of right forearm. This shows complete dislocation of the radial head and marked deformity and shortening of the ulna.

**Figure 16b**

Fluoroscopy views in surgery showing placement of the half pins perpendicular to each bone segment. The ulna was osteotomized at its apex and acutely straightened with a monolateral fixator.

**Figure 16c**

The ulna was then lengthened out to station and the radius transported distally.

**Figure 16d**

AP and lateral radiographs after removal of the external fixator. An open reduction and ligament reconstruction of the radio-capitellar joint was performed. The radial head is reduced.

interosseous space. This alters the normal bucket handle mechanics of the radius rotating around the ulna, contributing to loss of supination–pronation. This is often followed by gradual radial head subluxation and dislocation in a similar pattern to acute dislocations seen with Monteggia fracture–dislocations.¹⁰⁹ The radial head dislocates in the direction of the ulnar bow. The critical ulnar bowing threshold for subluxation/dislocation of the radial head is approximately 15° or greater. The shorter the ulna, the less bowing angulation is needed before dislocation of the radial head occurs. If ulnar shortening occurs without ulnar bowing, the radial head will not dislocate (unpublished data).^{24,26,29,98–100,110–125}

The ulna shortening also acts as a tether to the distal radial physis. The distal radial tilt in the frontal plane is on average 23°. In MHE, the radial tilt increases and is associated with ulnar shortening. Correspondingly, without ulnar head support, the increased radial tilt leads to ulnar carpal slip. This ulnar subluxation of the carpus weakens grip strength, and patients often complain of weakness and pain when gripping. It creates an ulnar deviation posture of the hand. It also leads to wrist joint degeneration over time. Carpal slip greater than 50% is therefore considered an indication for surgery.

The radial diaphyseal bow may also increase widening the interosseous membrane space. This does not usually interfere with forearm rotation unless the ulna also bows in the same direction. The increased radial bow does not impact the wrist or elbow and is usually not the primary concern.

Shortening of the radius relative to the ulna can also occur. This is less common than ulnar shortening. This causes ulnocarpal impingement and loss of forearm rotation. It does not cause dislocation of the radial head.

The commonly used Masada classification divides the types of MHE forearm based on the location of the OC and the presence or absence of a radial head dislocation.^{100,125} This classification does not guide treatment. It is also not predictive of radial head subluxation. We propose a new classification that is treatment based and takes into account that the ulna is the primary problem in radial head subluxation and dislocation.

Feldman–Paley Classification of MHE Forearm(Fig. 17)

Type 1: Radius and ulna same length.

Type 2: Ulna short relative to radius.

Type 2a: Ulnar short with no bowing, radial head located.

Type 2b: Ulna bowed with or without radial head subluxation.

Type 2c: Ulna bowed with radial head dislocation.

Type 3: Radius short relative to ulna.

Modifiers

1. Interosseous OC: absent or present.
2. Distal radial tilt:
 - (a) normal.
 - (b) Increased with no carpal slip.
 - (c) Increased with carpal slip.
3. Proximal radial valgus: absent or present.

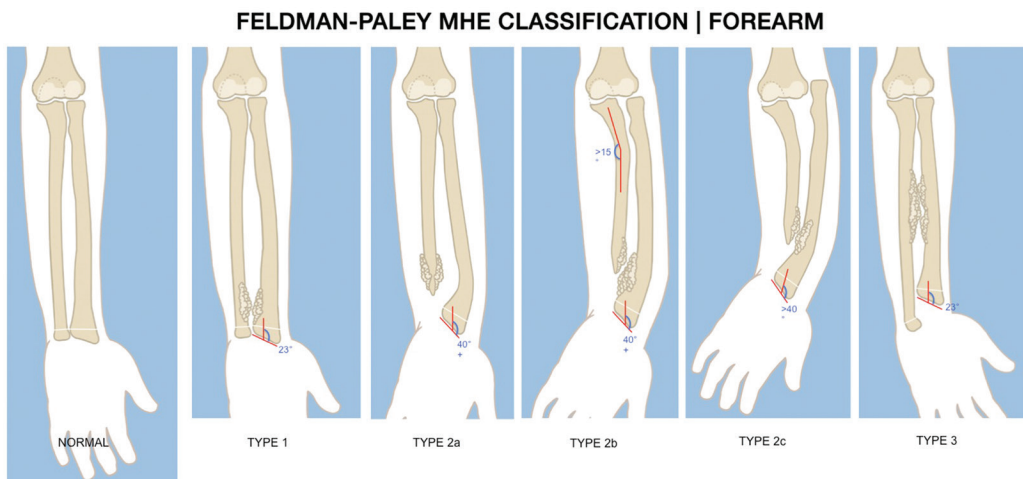


Figure 17a

Feldman-Paley MHE classification of the forearm.

Treatment for forearm MHE depends on the type. It can be divided into the following six surgical options, which are frequently combined:

Option 1: Resect OCs.

Option 2: Correct ulnar bowing to increase interosseous space and treat or prevent radial head subluxation.

Option 3: Correct distal radial tilt with hemiepiphysiodesis or closing wedge distal radial osteotomy.

Option 4: Correct length discrepancy between bones by lengthening or epiphysiodesis.

Option 5: Reduce dislocated radial head.

This classification aids in the treatment and prevention of sequela of the MHE forearm. Type 1A plus OC would simply have a resection of the OCs that are limiting motion and impinging on the other bone. While most such OCs are distal, proximal OC pushing out the radial head can also exist (Fig. 18).

Type 2b with normal radial tilt would have an ulnar osteotomy for straightening with or without ulna lengthening.

Type 2b with increased radial tilt with carpal slip would undergo resection of the OC, ulna straightening osteotomy, and a distal radial closing wedge osteotomy. If there was no carpal slip, the distal radius could be watched or hemiepiphysiodesis could be considered. Lengthening of the ulna may be beneficial as well to aid the buttress on the ulna side of the wrist. Getting the ulna out to length is known to improve grip strength.

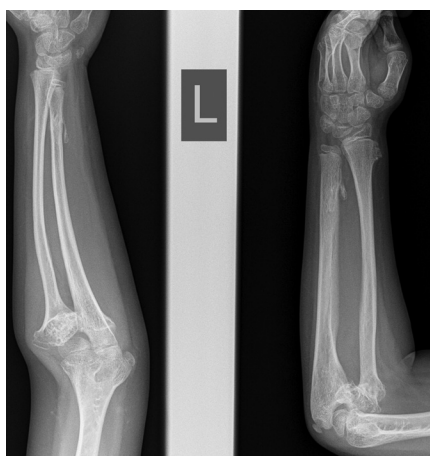


Figure 18a

AP and lateral radiographs of the forearm showing a proximal osteochondroma of the radial neck causing dislocation of the radiocapitellar joint.

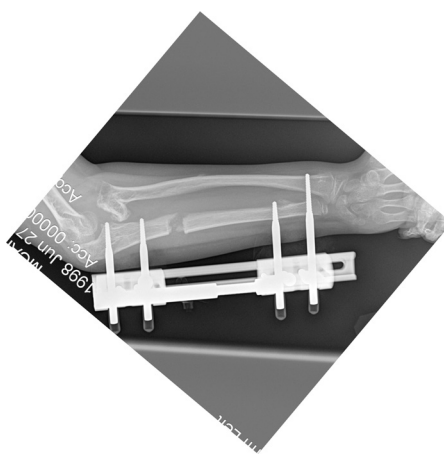
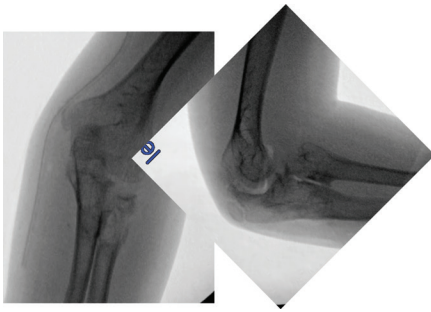


Figure 18b

The ulna was lengthened with a monolateral fixator. The radial neck osteochondroma is transported distally with the radius.

**Figure 18c**

Intra-operative fluoroscopy of showing reduction of the radio-capitellar joint and resection of the radial neck obstructing osteochondroma.

**Figure 18d**

AP and lateral radiographs two years later showing that the reduction is maintained. Some residual or recurrent exostosis is present but is not causing luxation.

Type 2c needs to undergo an ulnar lengthening while pulling down the radial head to station. This reconstruction is a salvage procedure and even with open reduction of the radiocapitellar joint and reconstruction of the annular ligament, the patient does not regain the loss of supination and pronation. The advantage of reducing the radial head is mostly to remove the bothersome prominence at the elbow, which is sometimes tender and is prone to be bumped. In some late cases, reduction of the radial head is also performed in conjunction with radial head prosthetic replacement.

The presence of proximal radial neck valgus may be significant when addressing radial head dislocation. In some cases, a varus osteotomy of the proximal radius is performed at the time of open reduction of the radiocapitellar joint.

The goal of treatment is not to allow the radial head to dislocate. This can be achieved by timely treatment of the Type 2b as the ulna bow approaches 15°.

This classification is treatment based and will aid in the decision-making tree in MHE forearm deformity.

► HAND AND FINGERS

OCs can grow anywhere within the hand and carpus (Fig. 19). Common problems include joint impingement by growth of an OC in the PIP or DIP joints. These often require excision with ligament reconstruction at times. Although resection of finger OCs and



Figure 19

Volar view of 3D CT of left hand (left) and AP radiograph left hand (right) showing multiple osteochondromas affecting volar distal radius, volar base of second metacarpal and multiple phalanges. The middle finger has an intra-articular osteochondroma affecting motion.

osteotomies of the phalanges are not difficult procedures, they are associated with a high risk of loss of motion of the PIP joint that is difficult to recover.

Clinodactyly with radial and ulna deviation of the digits is seen and may require straightening osteotomies if function is impeded.

Subungual exostoses are also seen and may cause pain as the nail bed is raised. These are treated by raising the nail and excision of the OC.^{126–128}

► SPINE

Spinal involvement in MHE has been reported as high as sixty eight percent of patients with twenty seven percent of patients having encroachment of the spinal canal.¹²⁹ Osteochondromas can grow in any location of the spine from the posterior lamina which are often palpable to transverse process and the deep lamina.

We have reviewed our series of 64 consecutive patients treated for MHE and found only 3 percent with spinal involvement and one percent with spinal encroachment. While OC can grow anywhere on the spine it is likely to be an uncommon clinical problem. We still recommend all patients with MHE undergo a spinal MRI by age 8 as undetected spinal encroachment can be clinically dangerous (Fig 20a,b).



Figure 20a

MRI T2 weighted sagittal image in a twelve year old boy demonstrating a thecal sac compression (large arrow) with a more distal resultant syringomyelia (small arrow).

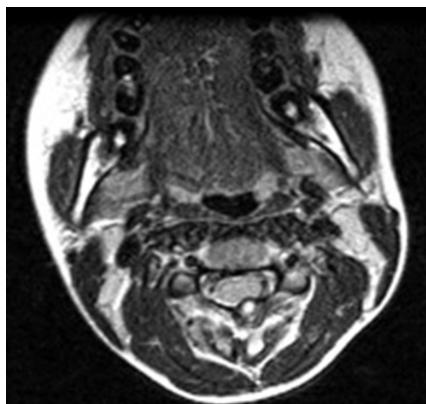


Figure 20b

MRI T2 weighted axial image demonstrating a ventral laminar osteochondroma compressing the thecal sac.

KEY LEARNING POINTS

- The Hip in MHE may be impacted by both OC and coxa valga. Both are correctable.
- Nerve compression by OC is common in MHE and should be treated by excision of the offending OC and nerve decompression.
- A short fibula in the ankle of MHE patients may cause disruption of the ankle mortise and can be treated by the SHORDT operation.
- Dislocation of the radial head can be prevented by straightening the ulna bow.
- Spinal involvement in MHE is possible and may cause spinal cord compression.

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