

THE MHE COALITION
MHE and Me

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The MHE Coalition

is a 501(c)(3) non-profit organization dedicated to providing support and information to those whose lives are affected by Multiple Hereditary Exostoses, and to funding and participating in research to help find the causes, more comprehensive treatments and ultimately the cure for this rare bone disorder.

MHE and Me: A Support Group for Kids with MHE and Their Families

is a member organization of The MHE Coalition dedicated to providing resources, education and a supportive community to children and teens with MHE. Programs include "We Care", where gift packages are sent to children having surgery and their siblings, and The Bumpy Bone Club, which provides kids and teens with free membership kits and a magazine written by and for them.

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THE MHE COALITION

6783 York Road, #104, Parma Heights, OH 44130
1-440-842-8817, CheleZ1@yahoo.com
www.mhecoalition.org

**MHE and Me—A Support Group for
Kids with MHE and their Families**

PO Box 651, Pine Island, NY 10969
1-845-258-6058, mheandme@yahoo.com
www.mheandme.com

The ABC's of MHE

Orthopaedics

Diagnostic Tools and How MHE Can Effect Each Part of the Body

Harish Hosalkar, MD.; John Dormans, MD., Chief of
Orthopedic Surgery, Children's Hospital of
Philadelphia, Professor of Orthopaedic Surgery,
University of Pennsylvania School of Medicine;
Notes from The MHE Coalition

Multiple Hereditary Exostoses and the Lower Limb and Multiple Exostoses of the Forearm

Dror Paley, MD
Paley Avanced Limb Lengthening Institute
St. Mary's Medical Center,
West Palm Beach, Florida

Physical Therapy for Patients with Multiple Hereditary Exostoses

Elena McKeogh Spearing, MA, PT, DPT, PCS
Physical Therapy Manager
Children's Hospital of Philadelphia

**The MHE Coalition greatly appreciates the
time, effort and expertise that Drs. Hosalkar,
Dormans and Paley, and Elena McKeogh, MA,
PT, DPT, PCS shared with our organization and
all those affected by MHE/HME/MO**



DIAGNOSTIC TOOLS

Harish Hosalkar, MD.; John Dormans, MD., Chief of Orthopedic Surgery, Children's Hospital of Philadelphia, Professor of Orthopaedic Surgery, University of Pennsylvania School of Medicine;
Notes from The MHE Coalition

Important features of orthopedic exam: It is important to follow each step of the exam during every office assessment of MHE patients.

- Patient should be *comfortable with adequate exposure and well-lit surroundings* (lest some important physical finding is missed).
- It is important to assess how the patient moves about in the room before and during the examination as well as during various maneuvers. *Balance, posture and gait pattern* should also be checked.
- *General exam* findings should look for any lumps and bumps that can be felt on any anatomical sites that can be easily palpated. Chest wall, abdominal and deep pelvic exam should be performed in all cases. Sessile or pedunculated nature of the bumps should be ascertained if possible.
- *General body habitus*, including developmental milestones should be noted
- It is important to note any obvious *spinal asymmetry, deformities, trunk de-compensation, and evidence of para-spinal muscle spasm or elevation suggestive of spinal lesions*.
- The patient's height should be measured and monitored on subsequent visits. Individuals with MHE are frequently of short stature, with most having heights 0.5 to 1.0 SD below the mean. (The lesions tend to enlarge while the physes are open proportionate to the overall growth of the patient, and the growth of the osteochondromas usually ceases at skeletal maturity).
- It is essential to perform and document a *thorough neurologic exam*. Asymmetric abdominal reflex is a subtle sign of spinal pathology and may be associated with spinal cord compression. Motor, sensory and reflex testing should be performed and recorded.

Characterization of Lesions:

Location: Whether the tumor is located in the limb bones, chest wall or ribs, skull, or any other sites in the body.

Pain: Is the lesion associated with pain? If yes:

◆ **Intensity:** Can be assessed and documented in a lot of ways.

i. On a pain scale of 1 to 10.

ii. The MHE and Me Website now features the *Bumpy Bone Tracker, Pain Tracker and Pain Diary*, all of which can be used as a tool to discuss pain with a child during an examination

◆ **Quality:**

Do you experience pain on and off (intermittently) throughout the day, or all the time (constantly)?

Does the pain occur at specific times, or with specific activities (ex. Upon waking in the morning, when walking, etc.)

Is pain interfering with your general activities?

Do you need to use special accommodations at work or school?

Is pain affecting your mood?

Do you take medications for pain, or use other treatments (i.e. heat, ice, rest). If so, are these treatments effective?

Tumor pain is often unrelenting, progressive, and often present during the night.

Is there any shooting pain? (Suggestive of nerve compression)

- ◆ **Radiation:** Pain radiating to upper or lower extremities or complaints of numbness, tingling or weakness suggest neurologic compression and requires appropriate workup.

Onset:

How was the tumor noticed?

Was there any history of trauma?

When did the pain actually start?

Duration:

What has been the duration of this new lesion?

How long have the other lesions been around?

Progress: Whether the exostosis has remained the same, grown larger, or gotten smaller?

Associated symptomatology:

- ◆ **Gait and posture** disturbances (especially during follow-up of skull or spinal lesions and in cases of limb-length discrepancies and deformities).

- ◆ Any specific history of backpain. If yes, its complete characterization.

- ◆ Change in **bladder or bowel habits** (for evidence of spinal lesions causing cord compression)

- ◆ Gynecologic function alterations in girls with pelvic lesions.

- ◆ **Scoliosis** may be associated with spinal lesions and may need to be monitored.

- ◆ **Night pain** if present is a worrisome symptom and needs complete evaluation. Night pain is different than chronic pain in that the pain is not constant and characteristically wakes the patient from sound sleep. Chronic pain on the other hand is persistent and would interfere with the sleep pattern by making the patient restless. Chronic night pain is especially common in MHE cases where the location of the bump may cause pressure on the exostoses when lying down. Soft beds, air cushions, lateral positioning and frequent turning may prove to be helpful in these cases.

- ◆ **Neurologic symptoms** may be associated spinal or skull lesions. More commonly, local compression of peripheral nerves due to expanding lesions is encountered in arms and legs. In addition, several cases of Reflex Sympathetic Dystrophy (RSD) following MHE surgeries to knees and wrists have been noted. Many patients also experience other nerve-related symptoms following surgery, including long-lasting pain and sensitivity around surgical sites long after incisions have healed.

- ◆ Bursa formation and resulting **bursitis** may occur as a result of

the exostoses and should be recorded. A bursa is a fibrous sac lined with synovial membrane and filled with synovial fluid and is found. The function of a bursa is to decrease friction between two surfaces that move in different directions. Therefore, you tend to find bursae at points where muscles, ligaments, and tendons glide over bones. These bursae can be either anatomical (present normally) or may be developmental (when the situation demands). The bursae can be thought of as a zip lock bag with a small amount of oil and no air inside. In the normal state, this would provide a slippery surface that would have almost no friction. A problem arises when a bursa becomes inflamed. It loses its gliding capabilities, and becomes more and more irritated when it is moved. Bursitis can either result from a repetitive movement or due to prolonged or excessive pressure.

Note from The MHE Coalition: Children who have been experiencing MHE-related pain may be hesitant to share this information with their doctors, fearing that the doctor will recommend surgery. It is not unusual for children to try to minimize their symptoms. We advise families to use the pain tracker tools available to keep an ongoing record between check-ups. It is also important for both parents and physicians to let children know that pain does not necessarily mean surgery, but that it is important to let the doctor know about symptoms they have been experiencing.

DIAGNOSTIC WORK-UP

Physical examination.

A thorough physical examination of the patient is extremely important in the assessment of MHE patients.

Radiographs

High quality plain radiographs (X-rays) (anteroposterior and lateral views) should be ordered in cases presenting with exostoses. Standing postero-anterior and lateral views of the entire spine on a three-foot cassette should be ordered when spinal lesions are suspected. Special views like tangential views of the scapula may need to be ordered in some cases. Plain films help to localize the lesion and give a fairly good idea about its size and dimensions in 2 planes. Also **scanograms** help to assess the extent of limb-length discrepancy and its localization. Oblique views of the spine and special skull views may be ordered in suspected cases.

Advanced Imaging

Radionuclide bone scan is sensitive to pathologies causing increased bone activities within the skeleton. In combination with **SPECT** (single photon emission computed tomography), it gives excellent localization of the area of increased uptake. This is extremely useful in MHE to locate multiple lesions, especially those that are situated in deeper areas not amenable to clinical palpation. Further imaging if required, can then be focused. **Thallium and PET (positron emission Tomography) scans** are also modalities that can help define the tumor metastasis especially in those rare cases of malignant degeneration.

Computed Tomography (CT)

CT scans are useful in visualizing the bony architecture particularly as an adjunct to plain radiographs or bone scans. Thin slice CT cuts may be necessary in small lesions. Two and three-dimensional reconstructions are possible and add to the information. Rarely the CT may be combined with the myelogram to effectively delineate the size of the lesion especially for intraspinal lesions.

Magnetic Resonance Imaging (MRI)

This is an excellent modality for defining the spinal cord, nerve roots, soft tissue structures and cartilage. Cortical bone is not seen as well as compared with CT. Cartilage caps of the exostoses and their compression effects on soft-tissues, nerves and adjacent vessels can be very well delineated. It is a study of choice in suspected cases of malignant transformation. MRI studies must be reserved for those cases in which clinical signs and symptoms deem them appropriate. Clinicians must make a point to communicate clinical information and suspected differential diagnosis to the radiologists.

Other Diagnostic Tools

Ultrasound

May be necessary to diagnose compression of arteries. The principle for ultrasound, or ultrasonography, is the same as for underwater sonar or echo sounding. An apparatus sends an ultrasonic wave through the body at a speed of about 1,500 meters per second. At the [interface](#) between two types of tissue, the wave will be refracted or 'broken up', and part of the wave will be reflected back and detected by the apparatus. The rest of the ultrasonic wave continues deeper into the body, and is reflected as an echo from the surface of tissues lying further inside the body. How much is reflected depends on the densities of the

respective tissues, and thus the speed of the sound wave as it passes through them. The time taken for the reflected wave to return indicates how deep the tissue lies within the body. In this way, one obtains a picture of the relative locations of the tissues in the body, in the same way that one may visualize the contours of a school of fish with sonar. An ultrasound can help ascertain the status of the blood flow through the arteries as well and is therefore important for assessment of suspected compression.

EMG (Electromyography, myogram)

May be necessary in cases of suspected nerve damage

What is EMG

Electromyography is a test that measures muscle response to nervous stimulation (electrical activity within muscle fibers).

How the test is performed

A needle electrode is inserted through the skin into the muscle. The electrical activity detected by this electrode is displayed on a monitor (and may be heard audibly through a speaker). Several electrodes may need to be placed at various locations to obtain an accurate study. After placement of the electrode(s), you may be asked to contract the muscle (for example, by bending your arm). The presence, size, and shape of the wave form (the action potential) produced on the monitor provide information about the ability of the muscle to respond when the nerves are stimulated.

Each muscle fiber that contracts will produce an action potential, and the size of the muscle fiber affects the rate (frequency) and size (amplitude) of the action potentials.

A [nerve conduction velocity](#) test is often done at the same time as an EMG.

Why the test is performed

EMG is most often used when people have symptoms of [weakness](#) and examination shows impaired [muscle](#) strength. It can help to differentiate primary muscle conditions from [muscle weakness](#) caused by neurologic disorders.

EMG can be used to differentiate between true weakness and reduced use due to pain or lack of motivation.

Histology

Clinical examination and Imaging findings can help establishing the diagnosis in most cases. Biopsy should be performed when a malignant change is suspected.

Laboratory evaluation

Genetic linkages to MHE have been documented which form the basis of genetic testing.

Test methods:

Sequence analysis of the EXT1 and EXT2 genes are offered as separate tests. Using genomic DNA obtained from buccal (cheek) swabs or blood (5cc in EDTA), testing of EXT1 proceeds by bi-directional sequence analysis of all 11 coding exons. The EXT2 gene consists of 15 exons, and all coding exons (2-15) are sequenced in the analysis.

Test sensitivity:

In patients with MHE, mutations are found in approximately 80% of individuals. Of those in whom mutations are identified, 70% of the mutations are found in the EXT1 gene and the remaining 30% in the EXT2 gene. Thus, the method used to screen the EXT1 is expected to identify approximately 60% of mutations in MHE. In individuals who are found to be negative on analysis of the EXT1 gene, screening of the EXT2 gene will identify the molecular basis of the disease in a further 25% of affected individuals. To date, there are no known distinguishing features within the clinical diagnosis of MHE known to predict which gene is more likely to have a mutation. Multiple exostoses can be associated with contiguous deletion syndromes, which are not detected with these methods.

HOW MHE CAN AFFECT EACH PART OF THE BODY

MHE usually manifests during early childhood more commonly with several knobby, hard, subcutaneous protuberances near the joints.

The likelihood of involvement of various anatomical sites as observed in a large series is as follows:

Anatomical location	Percentage Involvement
Distal femur	70
Proximal tibia	70
Proximal fibula	30
Proximal Humerus	50
Scapula	40
Ribs	40
Distal radius and ulna	30
Proximal femur	30
Phalanges	30
Distal fibula	25
Distal tibia	20
Bones of the foot	10-25

THE SKULL

Lesions in the skull, although reported are extremely rare. Mandibular osteochondromas, typically of the condyle, skull wall lesions and even intracranial lesions have been reported.

Affects of MHE on Skull

Exostoses can cause problems if they compress or entrap cranial nerves or cause extrinsic compression on the brain. Effects can range from bumpy external lesions that cause cosmetic problems, compression of adjacent structures, cranial nerve involvement and even focal neurological deficits due to compression. Even seizures are likely due to intracranial lesions.

Diagnostic Procedures

The orthopedist will manually feel for exostoses along the outer table of the skull, check movements of the mandible and also of the upper cervical spine. The orthopedist will also

check cranial nerve function and perform a thorough neurological evaluation. X-rays or other imaging tests including CT and MRI may be ordered.

Possible Treatment Options

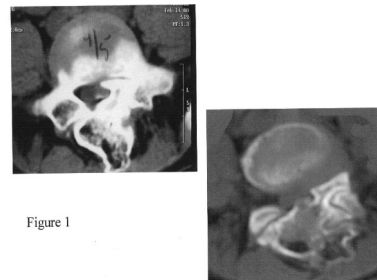
- ⇒ Minor lesions on the outer table of the skull that are flat can sometimes be closely observed.
- ⇒ Bigger lesions on the skull, mandibular lesions causing TM joint instability, and intracranial lesions causing pressure signs may need to be removed by neurosurgical intervention.
- ⇒ Upper cervical spinal tumors, especially of the atlanto-occipital region may be dealt with by orthopedists. Decompression and or stabilization may be performed as required.

What Parents Should Watch Out For

- ⇒ Pain. Is your child experiencing pain from exostoses?
- Visible lumps on the face or skull.
- ⇒ Any symptoms of tingling, numbness, weakness in the hands or legs suggestive of focal deficits.
- ⇒ Episodes of seizures or findings of cranial nerve involvement like altered smell, taste, ringing in ears etc.
- ⇒ Problems in chewing, restricted motion of the jawbone or instability of the mandible.
- ⇒ Parents can ask dentists and orthodontists to be on the lookout for signs suggestive of jawbone instability or joint involvement during their office visits especially in symptomatic cases.

SPINE

The spine extends from the base of the skull to the tailbone. Spinal exostoses are rare (Figure 1). Spinal cord impingement is also a rare, but documented, complication of MHE. Cervical, thoracic or lumbar region can be affected. Scoliosis secondary to spinal osteochondromas and instability has been reported.



Affects of MHE on the Spine:

This section of the body is not commonly involved with MHE. Involvement of isolated vertebrae has been noted. Affects can range from instability to neural root or cord compression that can manifest as tingling, numbness or weakness in the involved roots or even major neurological deficits like paraparesis or quadriparesis in untreated cases. Rarely compression effects in the form of dysphagia, intestinal obstruction or urinary symptoms may occur.

Diagnostic Procedures:

With any of the red flags mentioned earlier, the orthopedist will perform a thorough spinal and neurological evaluation. Plain x-rays of the spine and if required, advanced imaging may be performed. The presences and extent of the lesion are best delineated with CT,

while MRI of the spinal cord demonstrates the area of spinal cord impingement. In rare cases of peripheral nerve compression electromyography may be performed to check status of the nerve.

Possible Treatment Options:

- ⇒ Minor lesions not causing compressive symptoms or neurologic manifestations may be kept under close observation.
- ⇒ Progressive scoliosis and spinal instability may need to be treated with surgical stabilization involving spinal fusion.

What Parents Should Watch Out For:

- ⇒ Any red flags in terms of tingling, numbness, weakness, night pain or bladder and bowel changes and get them evaluated.
- ⇒ Any deformity in the spine or evidence of shoulder or pelvic imbalance.
- ⇒ Gait or posture disturbances. Remember that gait and posture disturbances can be caused by hip or leg exostoses as well (due to either limb-length discrepancy or deformity) and do not necessarily mean tumors in the spine. In any case evaluation by a clinician is important.

RIBS AND STERNUM

Affects of MHE on the ribs and sternum

The typically flat bones of the ribs are prone to effects of MHE, with approximately 40% of MHE patients having rib involvement. Prominent chest wall lesions are common although intrathoracic lesions including rare presentations like spontaneous hemothorax (build-up of blood and fluid in the chest cavity) as a result of rib exostoses have been described. Typically, these lesions create issues of cosmesis due to their obvious visibility. Other symptoms may include shortness of breath and other breathing difficulties, pain when taking a deep breath, when walking or exercising, or pain from exostoses "catching".

Diagnostic Procedures

The orthopedist will probably manually feel for exostoses along the chest wall and the ribcage. Size and extent of the lesions are noted. A thorough pulmonary evaluation is warranted in all cases when specific symptoms of cough, chest pain or breathing problems are encountered. X-rays or other imaging tests may be ordered.

Possible Treatment Options:

- ⇒ Minor bumps can sometimes be kept under observation. Cosmetic problems, rapid increase in size, large size, and signs of compression are some indications for early removal.

Consult may be required with specialists:

- ⇒ Pulmonary: when there are severe breathing difficulties with increasing chest pain.
- ⇒ *Thoracic surgeons: when intrathoracic (within the chest wall) exostoses may need to be excised.*

What Parents Should Watch Out For:

- ⇒ Breathing difficulties, shortness of breath
- ⇒ Pain when taking deep breath.

SHOULDER GIRDLE

The scapula is a fairly common site (40%) of involvement in MHE. The lesions may be located on the anterior or posterior aspect of the scapula. Anterior scapular lesions may lead to discomfort during scapulothoracic motion. Winging of the scapula due to exostoses has been described. Clavicle (collar bone) involvement has also been described (5% cases).

What is winging?

The scapula (also known as shoulder blade) is a triangular flat bone that is located in the upper back and takes part in forming the shoulder joint. The scapula usually lies flat on the chest wall without any prominence. Winging of the scapula is a phenomenon when a part of the scapula including the inferior angle becomes prominent either at rest or during movements. The two most common causes for this are

- ⇒ Exostosis on the inner (chest wall) aspect of the scapula.
- ⇒ Damage to the nerve (long thoracic) causing weakness or paralysis of muscles (serratus anterior) attached to the scapula.

Diagnostic Procedures

The orthopedist will probably manually feel for exostoses along the outer aspect of the shoulder blade. Some limited areas of the inner aspect are amenable to clinical examination. Range and feel of the scapulothoracic motion is helpful in clinical assessment. It is important to check individual groups of scapular muscles to rule out nerve compression leading to winging of scapula. X-rays (including special tangential views of the scapula) or other imaging tests may be ordered.

Possible Treatment Options

Both outer aspect lesions and inner ones may need excision in symptomatic cases. Smaller lesions on outer aspect amenable to clinical palpation may be observed with regular clinical follow-up.

What Parents Should Watch Out For:

- ⇒ Crunching or crackling sound when moving that area
- ⇒ Pain
- ⇒ Tingling, numbness

Note from The MHE Coalition: *Exostoses in the shoulder girdle (collarbone (clavicle) and shoulder blade can impact a child's ability to raise his/her arm in class, write on a blackboard, participate in certain sports, or wear a backpack. In addition, adolescent girls (and women) may be unable to wear a bra because of pressure not only on shoulder girdle exostoses but also on rib exostoses.*

ARMS

Upper Arm (Hummers)

Elbow

Forearm (Radius and Ulna)

Wrists

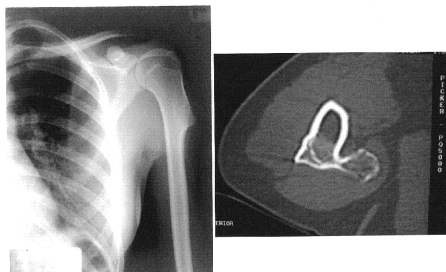


Figure 2

The arm bone is called the humerus while the forearm bones are the radius (curved bone) and the ulna (straighter bone of the two).

Osteochondromas of the arm are often readily felt but rarely cause neurologic dysfunction (Figure 2). Osteochondromas of the upper extremities frequently cause forearm deformities. The prevalence of such deformities has been reported to be as high as 40-60%. Disproportionate ulnar shortening with relative radial overgrowth has been frequently described and may result in radial bowing. Subluxation or dislocation of the radial head is well-described sequelae in the context of these deformities.

The length of forearm bones inversely correlates with the size of the exostoses. Thus, the larger the exostoses and the greater the number of exostoses, the shorter the involved bone. Moreover, lesions with sessile rather than pedunculated morphology have been associated with more significant shortening and deformity. Thus, the skeletal growth disturbance observed in MHE is a local effect of benign growth. Exostoses in the forearm are known to involve both the radius and the ulna. Since movements of the forearm (pronation and supination) are dependant on the radius moving in an arc of motion around the ulna, mobility may be restricted depending upon the severity of presentation. Also the lower end radius exostoses can lead to compression of the median nerve (in a closed space at the level of the wrist called the carpal tunnel) and present with weakness, tingling and numbness in the hand. Exostoses in the carpal bones can seriously hamper the wrist motion and cause pain.

Complete dislocation of the radial head is a serious progression of forearm deformity and can result in pain, instability, and decreased motion at the elbow. Surgical intervention should be considered to prevent this from occurring. When symptomatic, this can be treated in older patients with resection of the radial head.

Diagnostic Procedures:

The orthopedist will clinically feel for exostoses along the arm, elbow and forearm, and check range of motion ("ROM") by moving the arm in different directions. The orthopedist will also check measurements on each arm and forearm to see if there is a difference. X-rays or other imaging tests may be ordered.

Possible Treatment Options:

- ⇒ Indications for surgical treatment include painful lesions, an increasing radial articular angle, progressive ulnar shortening, excessive carpal slip, loss of pronation, and increased radial bowing with subluxation or dislocation of the radial head
- ⇒ Minor lesions can sometimes be observed with careful follow up.
- ⇒ Bowing and some length discrepancies and be treated with a surgical procedure called "stapling," where surgical staples are inserted into the growth plate of the bone growing faster than the other. This will hopefully give the slower growing bone the chance to "catch up" and the forearm will straighten over time.
- ⇒ Limb Lengthening with a fixator. (See Section on Fixators)
- ⇒ Resection of the radial head
- ⇒ Excision of exostoses
- ⇒ Osteotomy
- ⇒ Epiphysiodesis
- ⇒ Non-surgical measures for treatment of soft-tissue compression, irritation or inflammation (anti-inflammatories, heat, rest, etc.)
- ⇒ Adaptive devices to aid those with shortened forearms, such as grippers, long-handled sock aides, etc.

What Parents Should Watch Out For:

- ⇒ Any red flags in terms of sudden increase in size of swelling, pain, nerve compression, tingling, numbness, or weakness.
- ⇒ Possibility of exostoses irritating or catching on overlying tissue, such as muscles, tendons, ligaments, or compressing nerves.
- ⇒ Loss of range of motion
- ⇒ Pain
- ⇒ Difficulty and/or pain when raising arm(s), lifting, carrying

HANDS AND FINGERS

Hand involvement in MHE is common. Fogel *et al.* observed metacarpal involvement and phalangeal involvement in 69% and 68%, respectively, in their series of 51 patients. In their series of 63 patients, Cates and Burgess found that patients with MHE fall into two groups: those with no hand involvement and those with substantial hand involvement averaging 11.6 lesions per hand. They documented involvement of the ulnar metacarpals and proximal phalanges most commonly with the thumb and distal phalanges being affected less frequently. While exostoses of the hand resulted in shortening of the metacarpals and phalanges, brachydactyly was also observed in the absence of exostoses.

Diagnostic Procedures:

The orthopedist will manually feel for exostoses in the hands and check range of motion ("ROM") in different directions. X-rays or other imaging tests may be ordered.

Possible Treatment Options:

Isolated lesions growing rapidly, or interfering with the smooth motion of tendons or joint motion may need to be excised. Multiple surgeries for small, insignificant lesions is usually not advocated.

Occupational therapy, physical therapy

Use of pencil grips, laptop computers, and other adaptive devices

What Parents Should Watch Out For:

Complaints of pain when writing

Some children will not complain of pain, but will have poor penmanship, write slowly, avoid writing, etc. Parents should also observe how the child holds writing and eating utensils.

Difficulty in rotating hand(s),

PELVIC GIRDLE (HIPS AND PELVIS)

Osteochondromas of the proximal femur (Figure 3) may lead to progressive hip dysplasia. There have been reported cases of acetabular dysplasia with subluxation of the hip in patients with MHE. This results from exostoses located within or about the acetabulum that may interfere with normal articulation.

Pelvic lesions (Figures 4 and 5) may be found on both the inner as well as outer aspect of the pelvic blades. Large lesions may cause signs of compression, both vascular and neurological. There have also been reports of exostoses interfering with normal pregnancy and leading to a higher rate of Cesarean sections.



Figure 3

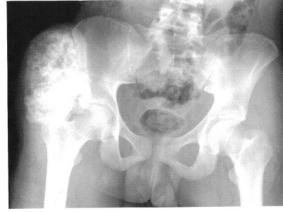


Figure 4 a

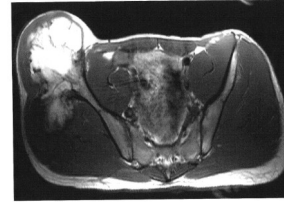


Figure 4 b



Figure 5 a



Figure 5 b

Diagnostic Procedures

Manual palpation is sometimes very difficult in these deep lesions. The orthopedist will check range of motion ("ROM") by manipulating (moving) the leg in different directions. The orthopedist will also check measurements on each leg to see if there is a difference in limb lengths. X-rays or other imaging tests may be ordered.

Possible Treatment Options

- ⇒ Minor length discrepancies can sometimes be effectively treated with the use of orthotics (specially made shoes or lifts that will equalize leg length).
- ⇒ Bowing and some limb length discrepancies can be treated with a surgical procedure called "stapling," where surgical staples are inserted into the growth plate of the leg bone growing faster than the other. This will hopefully give the slower growing bone the chance to "catch up" and the limb will straighten over time.
- ⇒ Limb Lengthening with a Fixator. (See Section on Fixators)
- ⇒ Pelvic lesions of concern may need to be surgically excised.
- ⇒ Osteotomies.
- ⇒ Hip replacement.

What Parents Should Watch Out For

- ⇒ Limping
- ⇒ Pain in hips, back, legs
- ⇒ Pain, discomfort, difficulty in sitting
- ⇒ Inability to sit "tailor" style
- ⇒ Stiffness in hips and/or legs after sitting
- ⇒ Pain and fatigue from walking

LEGS AND KNEES

Femur

Knees

Lower Leg (Tibia and Fibula)



Figure 6



Figure 7



Figure 10



Figure 9

The likelihood of involvement near the knee (figures 6 & 7) in at least one of the three locations is approximately 94%.

A clinically significant inequality of 2cm or greater has been reported with a prevalence ranging from 10-50%. Shortening can occur in the femur and/or the tibia; the femur is affected approximately twice as commonly as the tibia. Surgical treatment with appropriately timed epiphysiodesis has been satisfactorily employed in growing patients. In addition to limb-length discrepancies, a number of lower extremity deformities have been documented. Since the disorder involves the most rapidly growing ends of the long bones, the distal femur is among the most commonly involved sites and 70-98% of patients with MHE have lesions. Coxa valga has been reported in up to 25%; lesions of the proximal femur have been reported in 30-90% of patients with MHE. Femoral anteversion and valgus have been associated with exostoses located in proximity to the lesser trochanter.

Genu Valgum or *Knock-knee deformities* are found in 8-33% of patients with MHE. *Genu valgum* is defined as a mechanical malalignment of the lower limb when the knees knock against each other and the legs are pointed away from the body. Although distal femoral involvement is common, the majority of cases of angular limb deformities are due mostly to lesions of the tibia and fibula (Figures 8,9 & 10), which occur in 70-98% and 30-97% of cases, respectively. The fibula has been found by Nawata *et al.* to be shortened disproportionately as compared to the tibia, and this is likely responsible for the consistent valgus direction of the deformity. *Genu varum* or *Bowlegs* may also occur in some cases. This is defined as a mechanical malalignment of the lower limb when the knees drift away from the body and the legs are bowed and close together.

Diagnostic Procedures

The orthopedist will probably manually feel for exostoses along the leg, and check range of motion ("ROM") by manipulating (moving) the leg in different directions. The orthopedist will also check measurements on each leg to see if there is a difference. X-rays or other imaging tests may be ordered.

Possible Treatment Options

- ⇒ Minor length discrepancies can sometimes be effectively treated with the use of orthotics (specially made shoes or lifts that will equalize leg length).
- ⇒ Bowing and some limb length discrepancies can be treated with a surgical procedure called "stapling," where surgical staples are inserted into the growth plate of the leg bone growing faster than the other. This will hopefully give the slower growing bone the chance to "catch up" and the limb will straighten over time.

- ⇒ Limb Lengthening with a Fixator. (See Section on Fixators)
- ⇒ Excision of exostoses
- ⇒ Osteotomy

What Parents Should Watch Out For

- ⇒ Any red flags in terms of sudden increase in size of swelling, pain, nerve compression, tingling, numbness, or weakness.
- ⇒ Possibility of exostoses irritating or catching on overlying tissue, such as muscles, tendons, ligaments, or compressing nerves.
- ⇒ Leg cramps, bluish color, difference in skin temperature may indicate compression of an artery (most often the popliteal artery, located behind the knee).
- ⇒ Compression of the peroneal nerve, which runs along the outside of the leg, can cause a condition known as "drop foot", in which the foot cannot voluntarily be flexed up. Compression can be caused by exostoses growth, or as a complication of surgery.
- ⇒ Limping, pain when walking
- ⇒ Bowing of leg(s)
- ⇒ Exostoses on inside of legs bumping into each other
- ⇒ Exostoses interfering with normal movements, either by blocking movement or by causing pain (bending, sitting, walking up or down stairs)
- ⇒ Pain and fatigue when walking
- ⇒ Gait problems (awkwardness, limping, slow movements, etc.)

Notes from The MHE Coalition: *A good way to track possible bowing in your child's legs is to take a photo of your child dressed in shorts, standing straight with back to wall, legs together. Every few months take another photo of your child in the same position, and date and keep these photos to show your child's doctor. You can even have your child hold a sign with the date you take the picture (written dark enough to be picked up by the camera!).*

ANKLES

Valgus deformity of the ankle is also common in patients with MHE and is observed in 45-54% of patients in most series. This valgus deformity can be attributed to multiple factors including shortening of the fibula relative to the tibia. A resulting obliquity of the distal tibial epiphysis and medial subluxation of the talus can also be associated with this deformity, while developmental obliquity of the superior talar articular surface may provide partial compensation.

Diagnostic Procedures:

The orthopaedist will probably manually feel for exostoses along the leg, and check range of motion ("ROM") by manipulating (moving) the leg in different directions. The orthopaedist will also check measurements on each leg to see if there is a difference. X-rays or other imaging tests may be ordered.

Possible Treatment Options:

- ⇒ Minor length discrepancies can sometimes be effectively treated with the use of orthotics (specially made shoes or lifts that will equalize leg length).
- ⇒ Bowing and some limb length discrepancies can be treated with a surgical procedure called "stapling," where surgical staples are inserted into the growth plate of the leg bone growing faster than the other. This will hopefully give the slower growing bone the chance to "catch up" and the limb will straighten over time.

- ⇒ In more advanced cases, excision of exostoses with early medial hemiepiphyseal stapling of the tibia in conjunction with exostosis excision can correct a valgus deformity at the ankle of 15° or greater associated with limited shortening of the fibula.
- ⇒ Fibular lengthening has been used effectively for severe valgus deformity with more significant fibular shortening, (i.e. when the distal fibular physis is located proximal to the distal tibial physis).
- ⇒ Supramalleolar osteotomy of the tibia has also been used effectively to treat severe valgus ankle deformity.
- ⇒ Growth of exostoses can also result in tibiofibular diastasis that can be treated with early excision of the lesions.

What Parents Should Watch Out For:

- ⇒ Limping, pain when walking
- ⇒ Recurrent falls and instability while walking on uneven surfaces.

FEET AND TOES

Osteochondromas may occur in the tarsal and carpal bones, however they are often less apparent. Relative shortening of the metatarsals, metacarpals, and phalanges may be noted.

Diagnostic Procedures

Plain radiographs are probably more useful in defining the extent of involvement of the small bones of the feet in MHE. Other imaging studies may be ordered as and when required.

Possible Treatment Options

- ⇒ Large bumps can be surgically excised when symptomatic
- ⇒ Deformities of the foot (like hallux valgus) may be corrected by stapling of the growing epiphysis in younger children or by surgical osteotomy in older patients.

What Parents Should Watch Out For

Compression of the peroneal nerve, which runs along the outside of the leg, can cause a condition known as "drop foot", in which the foot cannot voluntarily be flexed up. Compression can be caused by exostoses growth, or as a complication of surgery.

Note from The MHE Coalition: Parents should know that finding shoes for children affected with ankle and/or foot exostoses can be challenging. Shoes must be found that do not cut into or press on exostoses. In some cases, they must be made specially for the child. In addition, some children will require lifts to help equalize a limb length discrepancy. Children who have exostoses on the bottom of their heel can sometimes benefit from gel cushions that are sold in drug/grocery stores. In addition, many children and teens will have difficulty tying shoes due to affected hands, shortened forearms, etc., and may need shoes with Velcro or shoes that slip on.

IN GENERAL

What Parents Should Watch Out For

- ⇒ If your child is limping, check to see if it is due to an injury, or is something that is occurring and continuing without obvious reason. Limping may signal a limb length discrepancy or other problem.
- ⇒ Bowing of one or both legs
- ⇒ Mobility problems. Is your child experiencing pain when walking or running?

- ⇒ Pain. Is your child experiencing pain from exostoses that bump each other? Is your child experiencing pain during certain activities, or pain at night. If so, keep a pain diary.
- ⇒ Any red flags in terms of sudden increase in size of swelling, pain, nerve compression, tingling, numbness, or weakness.

The MHE Coalition and its member organization, MHE and Me, offers patient support information, including:

“Tips for When Your Child Needs Surgery”

The Bumpy Bone Club Bumpy Bone Tracker and Pain Tracker and Pain Diary

The MHE and Me Handbook: A Guide for Families, Friends, Teachers and Classmates

School Needs Checklist for Kids with MHE

Common School Concerns for Kids with MHE

The Bump Bone Club (A free Membership kit includes membership certificate, Bumpy Bone Club Bone and Pain Tracker, a subscription to The Bumpy Bone Club Magazine, and special Bumpy Bone Club surprises.)

Information to help families work with schools to obtain necessary accommodations and resolve problems and challenges faced by children with MHE

The We Care Program provides gift packages for children undergoing surgery and their siblings, too. Please contact Susan Wynn at mheandme@yahoo.com about any of these programs or resources, or visit our websites (www.mhecoalition.org, www.mheandme.com)

What Adults Should Be Aware Of:

Sudden growth in an existing exostosis and pain can be symptomatic of a malignant transformation. It is smart to check out any changes with your orthopaedist. However, it is important to remember that chondrosarcoma is rare.

Years of wear and tear on joints can result in chronic pain. There is also the possibility of exostoses irritating or catching on overlying tissue, such as muscles, tendons, ligaments, or compressing nerves. Possible Treatment Options for these common problems, include pain medications, physical therapy (including stretching, strengthening and modalities), heat, rest, bracing (supportive orthosis acting as load sharing devices) etc.

GLOSSARY OF TERMS AND PROCEDURES:

Synonyms of multiple exostoses: *A number of synonyms have been used for this disorder including osteochondromatosis, multiple hereditary osteochondromata, multiple congenital osteochondromata, diaphyseal aclasis, chondral osteogenic dysplasia of direction, chondral osteoma, deforming chondrodysplasia, dyschondroplasia, exostosing disease, exostotic dysplasia, hereditary deforming chondrodysplasia, multiple osteomatoses, and osteogenic disease.*

Anterior: Situated in the front; forward part of an organ or limb

Ball and socket joints: Movable (synovial) joints, such as hips and shoulders, that allow a wide range of movement.

Bilateral: having two sides or pertaining to both sides.

Biopsy: Take a piece of the lesion to study the histological characteristics.

Cartilage: Form of connective tissue, more elastic than bone, which makes up parts of the skeleton and covers joint surfaces of bones.

Coxa-valga: When the thighbones are drawn farther apart from the midline due to an increase in the neck-shaft angle of the femur.

Coxa-vara: When the thighbones are drawn closer to the midline due to a decrease in the neck-shaft angle of the femur.

Dislocation: When the normal articulating joint surfaces have lost total contact.

Distal: Away from the midline or the beginning of a body structure (the distal end of the humerus forms part of the elbow).

Epiphysiodesis: To surgically stop the growth of the growing end of the bone either temporarily or permanently.

Excision: To surgically remove the lesion.

Genu-valgum: Knock-knees.

Genu-varum: Bowlegs.

Hinge joints: movable joints, such as knees and elbows, that allow movement in one direction.

Limb-lengthening: Process of increasing the length of bones using one of the various devices.

LLD: Limb-length discrepancy- difference in limb lengths.

Medial: Pertaining to the middle or toward the midline.

MHE: An autosomal-dominant disorder manifested by multiple osteochondromas and frequently associated with characteristic skeletal deformities.

Osteochondromas: Cartilage capped tumors found commonly at rapidly growing ends of bones.

Osteotomy: The surgical division or sectioning of a bone.

Pedunculated: Lesion with a stalk connecting it to the main bone.

Posterior: Situated in the back; back part of an organ or limb

Proximal: Near the midline or beginning of a body structure (the proximal end of the humerus forms part of the shoulder).

Sessile: Lesion without a stalk connecting it to the main bone.

Stapling: Process of insertion of a mechanical device (staple) following surgical intervention.

Subluxation: When the joint surfaces are still facing each other but not totally in contact.

Multiple Exostoses and the Lower Limb

Dror Paley, MD, FRCSC

**Paley Advanced Limb Lengthening Institute
Saint Mary's Medical Center, West Palm Beach, FL**

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 than 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), an 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 with external fixator lengthening is superficial pin infection. 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 two centers with a large experience with its use (Baltimore and Orlando). 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.

Multiple Exostoses of the Forearm

Dror Paley, MD, FRCS

**Paley Advanced Limb Lengthening Institute
Saint Mary's Medical Center, West Palm Beach, FL**

Introduction

The forearm consists of two bones (radius and ulna) and six joints (elbow: radio-capital and ulno-humeral; wrist: radio-carpal and ulno-triquetral; radio-ulnar: proximal and distal). Unlike the relationship between the tibia and fibula in the lower extremity the radius and ulna move functionally relative to each other to produce the movement of supination and pronation. Relative to the elbow they move together (flexion and extension). Although most wrist motion and stability comes from the articulation between the radius and the carpus, the ulna provides support for the ulnar side and prevents excessive ulnar deviation of the hand. The relationship between the radius and the ulna is therefore one of the most functional relationships between any two bones.

Exostosis formation of either bone can easily interfere in the function of the elbow, wrist or forearm rotation. Since osteochondromas form from the growth plates they are usually found at the ends of the bones but migrate towards the shaft of the bone with growth.

Ulnar Osteochondromas: osteochondromas most commonly form from the distal growth plate. Unlike those of the radius the ulnar exostoses are typically sessile (no stalk) while those of the radius are often pedunculated (on a stalk). The osteochondromas of the ulna often lead to delayed growth of the ulna relative to the radius. The radius gradually gets longer than the ulna. The slower growing ulna tethers the growing radius leading to increased tilt of the radius towards the ulna with increasing ulnar deviation of the wrist. Over time, the discrepant rate of growth leads to subluxation and then dislocation of the proximal end of the radius (radial head) from the elbow (radio-capitellar joint). Dislocation of the radial head from the joint causes the upper end of the radius to deform into valgus (bent position). Occasionally an osteochondroma can develop from the ulna side of the proximal radio-ulnar joint. This can also contribute to dislocation of the radial head by pushing the radial head laterally.

Radial Osteochondromas: osteochondromas from the radius can be divided into those that protrude towards the ulna and those that don't. The latter don't impede supination-pronation motion, while the former do. The radius and ulna may develop 'kissing exostoses' that meet in the interosseous space.

Distal radius deformity: the distal radius has a normal inclination towards the ulna of 23°. In MHE the slower growing ulna may tether the distal radius on the ulnar side leading to increased distal radial tilt. This increased tilt appears as ulnar deviation of the hand. With time the carpus will subluxate ulnarly and proximally.

Proximal radius deformity: the ulnar tether also exerts a dislocating force on the radio-capitellar joint. As the radial head subluxes it comes to rest against the lateral condyle of the humerus. To adapt to this chronic position the radial neck may grow into valgus. With time, the radial head may completely dislocate and protrude posteriorly.

Length discrepancy: The entire forearm is shorter than the other side. The shortening is predominantly in the ulna. Some shortening is also present in the radius.

Clinical signs and symptoms: Patients are limited in their forearm rotation range of motion. The wrist is usually ulnarly deviated. There may be a prominence or bump if the radial head is subluxed or dislocated. This may be tender to being bumped. Elbow flexion and extension is usually not affected. A flexion deformity of the elbow may be present.

Treatment considerations

Exostoses that are obviously impeding forearm rotation (e.g. kissing exostoses, are usually resected. It is important to do this via two separate incisions to avoid a cross union between the radius and ulna.

Lengthening and deformity correction can be performed as the first stage in the absence of exostoses that limit motion, or as the second stage if exostoses are resected first.

Lengthening Reconstruction Surgery (LRS): LRS refers to distraction surgery using external fixation to lengthen and correct deformities of the forearm. The problem in MHE ranges from simple to complex.

Simple cases: In simple cases, the primary deformity is relative shortening of the ulna. The radial tilt is minimal and does not need to be addressed. There is no subluxation/dislocation of the radial head. The problem is therefore just shortening of the ulna. If this is left untreated the secondary deformities of the radius will develop. The treatment is to perform an isolated lengthening of the ulna. I prefer to do this with a circular external fixator even though the lengthening is linear. A circular fixator allows simultaneous fixation of the radius to the ulna. Without fixation of the radius, lengthening of the ulna will transport the radial head distally. This occurs because of the tough interosseous membrane between the radius and the ulna. The osteotomy of the ulna is usually at its proximal end. This allows correction of any flexion deformity of the ulna (elbow) and leads to faster healing than if the osteotomy is made through the mid-diaphyseal (middle) section of the ulna.

Complex cases: In more complex cases the surgical plan includes correction of the distal radial deformity and or radial head dislocation. A circular external fixator is used. Proximally both the radius and ulna are fixed. The ulnar osteotomy is made proximally and the radial osteotomy is made distally. This type of frame simultaneously corrects shortening of the ulna and tilt of the distal radius. If the radial head is dislocated then the treatment is staged. The first step is to lengthen the ulna with a pin connecting the radius and ulna distally. This transports the radius distally and reduces the radial head. If the radial head does not reduce spontaneously then at a second stage surgery the radio-capitellar joint is opened and the radial head reduced at surgery and is held with an olive wire. If there is both distal radial tilt and dislocation of the radial head then the radial head is reduced first and then at a second stage the wire pulling the radius and ulna distally is removed and the distal radius osteotomized for deformity correction and lengthening.

With staged surgeries many of the deformities of MHE of the forearm can be corrected. Combined with removal of the obstructing exostoses improved range of motion of forearm rotation is obtained.

Does hemiepiphysiodesis stapling have a role in MHE? I have no experience with this in the upper extremity. Theoretically, it should work for the distal radius. We are considering correction of the distal radial tilt by stapling in combination with overlengthening of the ulna. Overlengthening of the ulna can help delay recurrence. Overlengthening of up to 2 cm is practical. Fixation of the hand is not required if the lengthening of the radius is less than 3 cm.

Physical Therapy for Patients with Multiple Hereditary Exostoses

Elena McKeogh Spearing, MA, PT, DPT, PCS

Physical Therapy Manager

Children's Hospital of Philadelphia

INTRODUCTION:

What is Physical Therapy?

Physical therapy is a profession that specializes in the diagnosis and management of movement dysfunction with the goal of restoring, enhancing, maintaining and promoting not only optimal physical function but optimal wellness, fitness and quality of life as it relates to movement and health. (1)

Physical Therapy can only be performed by a licensed Physical Therapist. Physical Therapists possess specialized training at the post-graduate level and have a license to practice Physical Therapy. Many people who have suffered an injury, disease or disability can benefit from Physical Therapy intervention. People who want to prevent illness and disability can benefit from Physical Therapy as well. (1)

What does a Physical Therapist do?

A Physical Therapist performs an examination of many systems in the body. These are the cardiovascular system, the neuromuscular system, the musculoskeletal and the integumentary system. The physical therapist looks specifically at how much a joint can move (range of motion) and how strong the muscles of the body are, including the heart. They look at what activities are hindered by pain or loss of motion or strength. Physical Therapists also examine balance, coordination and walking abilities.

After a complete assessment of the information and an interview with the patient, the Physical Therapist develops a plan of care to address any issues that are present. Based on a patient's personal goals, the Physical Therapist will develop a plan of care with specific interventions to help the patient to achieve those goals. Physical Therapists strive to provide patient and family centered care, which recognizes the importance of the patient and their family in the decision making process.

Physical Therapy interventions can include stretching, strengthening, postural and aerobic exercises, functional activities and activities of daily living training. Physical Therapists also educate patients on the importance of wellness and injury prevention.

Physical Therapists work as a team with other health care professionals including physicians, nurses, social workers, occupational therapists, speech therapists, recreational therapists, psychologists, and nutritionists.

PHYSICAL THERAPY FOR PATIENTS WITH MHE

How can Physical Therapy help patients with MHE?

For patients with Multiple Hereditary Exostoses, Physical Therapy is very important.

The physical therapist works together with the orthopedic surgeon to determine the best course of treatment for exostoses.

Pre-surgery:

As described throughout this book, exostoses can be present in any bone of the body. Depending on the location and amount of pain and disability, the orthopedic surgeon may or may not recommend surgery. Prior to surgery, the focus of Physical Therapy is to prevent or slow the loss of range of motion and function that can be caused by exostoses. Conservative treatment of exostoses may include physical modalities for pain relief. Although there is no evidence that exostoses growth can be prevented or slowed with Physical Therapy, the disability associated with the exostoses can sometimes be managed effectively with therapeutic interventions. Flexibility and strengthening exercises have been shown to decrease progressive disability in patients with other musculoskeletal disorders like fibromyalgia and rheumatoid arthritis (1, 2, 3).

Another focus of physical therapy, before and after surgery is required, may be to accommodate some of the deformities that occur as a result of the exostoses. These could include shoe lifts to make lengths of the legs equal, splints to protect joints and cushions to make certain positions more comfortable. Equipment can also be provided to make activities of daily living easier and less painful. These include long handled utensils, brushes, reachers and grippers. Problems with mobility can be addressed with walking aides like canes and crutches.

Additionally, it has been shown that cardiovascular exercise can decrease pain and improve overall well being in patients with musculoskeletal impairments (4, 5, 6, 7).

A supervised exercise program that includes aerobic exercise and strength training may also help to decrease the pain and stiffness associated with MHE. While there are many benefits to exercise, anything that causes increased pain in the area of exostoses should be discontinued and reported to the medical professional.

When the decision to have surgery is made, the patient's individual needs after surgery should be anticipated. Often, patients can be seen for a pre-operative Physical Therapy visit. During this visit, patients can learn how to use some of the equipment that they may need to use after the surgery. Practicing these new skills, like walking with crutches, or moving around with a cast or fixator, can make the patient less apprehensive about the rehabilitation process that will take place after the surgery.

This pre-surgery visit is also beneficial to problem solving obstacles to post-surgery rehabilitation. For example, many patients with painful exostoses under their arms, may not be able to use traditional axillary crutches to maintain decreased weight bearing on their legs after surgery. In this case, forearm crutches or a walker may be more appropriate for the patient. Additionally, a patient who does not demonstrate sufficient endurance may need a wheelchair to use for going outside of the house after surgery.

Similarly, the patient who may have decreased weight-bearing abilities after surgery will need to practice new approaches to everyday activities. These might include going up and down stairs, getting into and out of a car, using the toilet, bathing and going to school or work. The patient and the therapist can simulate these activities and problem solve together, before the surgery, so that they are prepared with successful strategies after the surgery is performed.

Post-surgery:

If it is determined that surgery is indicated to remove painful exostoses and increase a patient's function, physical therapy is important following surgery. Depending on the surgery, there may be a period of rehabilitation and the potential for a temporary decrease in function due to pain and muscle weakness. The focus of Physical Therapy after surgery is to minimize the pain and maximize the patient's movement potential around the area that the exostoses were removed.

Some patients with MHE may only require a brief hospital stay after removal of exostoses; others may require a rehab stay where more frequent and intense therapy is required. This depends on the location of the surgery, the extent of the surgery, the amount of function that is lost by the exostoses and the patient's prior level of functioning. During rehabilitation, therapy occurs daily and includes specifically stretching and strengthening of the muscles around the area of surgery. If pain and function were limited prior to the surgery, there may be some soft tissue limitations that are present after the surgery that will require special attention.

Based on the evaluation and orthopedic recommendations, weight bearing will be monitored and progressed as directed. In procedures, which include limb lengthening, physical therapy will also address the joints that surround the fixator to prevent further contractions.

Once surgery incisions are healed, a heated pool may be a good environment for therapy. The water's property of buoyancy can decrease the pain that may occur with weight bearing. Aquatic therapy can also provide an environment where muscles can be strengthened in a fun way with swimming.

Once the patient's goals are achieved and intense physical therapy is not required, transition planning will occur and recommendations for the home, work or school and community will be provided. The development of a home exercise program will maintain the gains that have been achieved through surgery and therapy and prevent secondary complications that are due to pain and immobility.

Is Physical Therapy painful?

Some activities that are performed in Physical Therapy can be uncomfortable because it is hard work. There are things that a therapist can do for their patient to make therapy and exercise more comfortable. Some examples of this are relaxation techniques such as deep breathing and imagery. There are also modalities like heat and ice, which can ease the discomfort caused by exostoses or surgery. Music therapy has been shown to be effective at decreasing pain in patients with other types of chronic pain (8).

Are there other diagnoses that are associated with MHE and/or MHE surgery?

There are some neurological disorders that can be associated with MHE. These include peripheral neuropathy and Complex Regional Pain Syndrome. (9,10,11) Secondary complications from these can also be addressed with Physical Therapy.

In **peripheral neuropathy**, the nerves that control the muscles are damaged due to being compressed by exostoses. This leads to a loss of nerve conduction to the muscle and resulting weakness in that muscle. It can affect the motor part of the nerve or the sensory part of the nerve. Impairment can range from slight to complete. Because the nerves are not central to the nervous system, they can regenerate once the compression is relieved surgically. This is, however, a slow process. Physical Therapy can address this with strengthening exercises and bracing, while the nerves to the muscles are healing. (9,10)

Complex Regional Pain Syndrome, type I, (also known as reflex neurovascular dystrophy "RND" or reflex sympathetic dystrophy "**RSD**") is a common condition characterized by extreme limb pain associated with autonomic dysfunction. This condition is associated with mild trauma to an extremity, as in the case of a painful exostoses or surgical removal of exostoses. In this situation, there can be temperature, hypersensitivity, and trophic changes to the effected extremity. Treatment of this disorder in adults ranges from medications, surgical sympathetic nervous system blocks and psychotherapy. In children, studies show the symptoms of this condition can be controlled with physical and occupational therapy. Treatment for this includes de-sensitization techniques where various textures are applied to the affected area

for prolonged periods of time. This usually begins with very light touching with cotton and progresses to different textures, such as cloth and brushes. Weight bearing activities are also essential to retraining the sensory system. Progressing weight bearing to the patient's tolerance is an important part of treatment. If weight bearing is restricted after surgery, deep pressure applied to the extremity in non-weight bearing positions can be substituted, until partial weight bearing is allowed. Exercise is also an integral part of treatment for complex regional pain syndrome and has been shown to be an effective treatment for this chronic pain disorder without the use of medications. (11)

Note from The MHE Coalition:

It can be very disconcerting for parents to see a child with RSD experience the episodes of burning pain associated with this disorder. It is important to remember how important treatment for this disorder is, and to work with your child's physical therapy team in designing a program that you can carry out at home. In addition to the normal range of exercises prescribed, there are beneficial "games" that the child can play, both at physical therapy and at home. If RSD is affecting the foot, fill a pan with play sand in which you have buried some marbles and coins. Have the child put his foot in the pan and try to pick up the items with his toes. This can help with desensitizing the foot, and promote movement, which the child might be avoiding because of pain. Children also enjoy kicking balloons and lightweight balls, when they are able. Another game is to have the child pick up marbles with his toes and place them into a bowl or cup, and try to beat the previous score each session the game is played. Appealing to the child's spirit of competition and fun can sometimes make the child forget that she is doing something therapeutic, and concentrating on the "score" can sometimes lessen the discomfort of the exercises. By working with your child, you will also discover the best way to handle episodes of pain. During some episodes, the child may not be able to stand touch, while at other times it may be necessary to apply firm pressure on the effected area.

Is it safe for patients with MHE to participate in sports and recreation?

Fitness and well-being is important for everyone, including the patient with MHE. Every opportunity for continuing exercise in a supervised manner should be encouraged. Sports and physical education can often be safe to participate in with a doctor's approval as long as the patient is being monitored by the orthopedic surgeon and physical therapist. Non-contact sports like swimming, cycling, dancing, tai chi, yoga and "Pilates" can be safe and fun forms of exercise for some patients with MHE. It is important to remember that MHE affects patients differently and certain sports and recreational activities may not be appropriate choices for people whose range of motion is limited by hip, pelvis and lower extremity exostoses. Additionally, if anyone with MHE experiences pain while participating in sports or recreational activities, he/she should stop doing the activity and speak with their doctor.

Some people with MHE tire very quickly when doing physical activity. Children with MHE need frequent rest breaks. Some children may be unable to participate fully in their school's physical education class. In these cases, there are federal laws under the Americans with Disabilities Act (ADA) and Individuals with Disabilities Education Act (IDEA), which entitle these patients to adaptations and accommodations and prohibit discrimination based on physical disability. Additionally, Physical Therapists can work with schools to adapt physical education classes or provide alternative activities in order to meet the curriculum's health and physical education requirement.(34 C.F.R. § 300.26) Parents may also request a copy of their school district's detailed physical education curriculum to review with their child's orthopedic surgeon to determine if there are any activities planned which are medically restricted for the child. Some school districts have allowed students to meet Physical Education curriculum requirements with specially designed instruction like written work on Physical Education topics. While this may be an acceptable physical education substitution for some children with MHE who are very limited in their physical mobility, it may be difficult for other children with MHE to do excessive written work due to exostoses in their hands and wrist.

Physical Fitness is an individualized concept. There are many types of activities that can be beneficial even if a person with MHE cannot participate in competitive or recreational sports due to fatigue or mobility impairments. Options for fitness include adapted wheelchair sports, seated aerobics and dance, and water aerobics. By encouraging as much independence as possible for a child in his/her school setting, he or she can get a significant amount of physical activity from walking to and from classes, sitting through classes, and keeping up with their studies. Physical Therapists can work with patients to determine their optimal activity level and options for fitness and well-being.

Note from the MHE Coalition

Many children with MHE enjoy playing sports and taking gym. In some school settings, families discuss the child's condition with the appropriate school and PE personnel, with the understanding that the child should not be forced to "just try" or do any activity that causes pain or that the child knows may injure an area affected by exostoses. Other children may have pain, fatigue and mobility issues that make any activity beyond that required to get to their classes and do their school work impossible. Parents may have to act as advocates on behalf of their children in order to receive the best possible accommodations or modifications for their child's Individual Education Plan. Your child's doctors and physical therapists should be advised of any mobility, pain and fatigue issues, not only as regards treatment, but also so that they can be included in your child's medical records. Medical and physical therapy reports are important documentation in support of requested modifications.

Additionally, chronic fatigue issues can also prohibit some children's ability to perform excessive writing activities. Some school districts have accepted a student's performance of the home exercise program that has been instructed by his/her Physical Therapist as specially designed instruction for meeting physical education curriculum requirements. An Individualized Education Plan (IEP) team will work with parents to ensure that the child is

CONCLUSION:
An overview of Physical Therapy for patients with MHE has been provided; however, each patient is individual and not all Physical Therapy interventions are indicated for all patients. All Physical Therapy should be patient and family centered in its approach. Exercise and fitness can be a family event and an activity that children, their siblings and parents can share to benefit each other.

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