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MULTIPLE HEREDITARY EXOSTOSES

Multiple Hereditary Exostoses is a benign condition characterized by multiple osteochondromas (bony growths) found on the surfaces of bones, often at or near the sites of tendon insertions. A genetic autosomal dominant disorder of most commonly the EXT1 and EXT 2 genes, the prevalence of MHE is 1 in 50,000. MHE is associated with characteristic progressive skeletal deformities of the extremities and shortening of one or both sides leading to limb length discrepancy and short stature. Two bone segments such as the lower leg or forearm are at greater risk of problems due to either osteochondromas 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. Osteochondromas can affect joint motion due to impingement of an osteochondroma with the opposite side of the joint or subluxation/dislocation related to deformity, impingement and incongruity. Osteochondromas can also cause nerve or vessel entrapment and/or compression including of 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. Other problems caused by osteochondromas include chronic pain and fatigue, issues with mobility and early onset arthritis. Patients with MHE are also at a mildly increased risk of developing malignant tumors.

Multiple Hereditary Exostoses (MHE) must be treated correctly to prevent problems from occurring and to allow a patient with MHE to live a full and painless life. This means, for example, that the elbow of MHE patients should not be permitted to dislocate. It also means if an osteochondroma is pressing on a nerve and causing severe pain, this should be treated. If there is a limb that is becoming crooked, it can be straightened. If there is a tumor injuring the spine, hip and/or ankle, it can be removed and the joints normalized. All these issues and more need to be addressed in the MHE patient. As well, we were involved in a drug trial with Palovarotene (PVO) in order to decrease the growth of tumors. This study has been on

temporary hold due to the use of the drug in a different condition (Ossificans fibrodysplasia progressiva).

The patient as a whole needs to be treated, listened to and managed. There is no reason for an individual with MHE to suffer from chronic nerve and/or joint pain. The principles of intervening on patients with MHE revolve around the following concepts. We would recommend surgery if a significant deformity is present, there is significant involvement of the joint leading to loss of range of motion, nerve entrapment leading to possible irritation or palsy, or a rapidly growing osteochondroma, which could represent a pre-malignancy. Once a patient reaches skeletal maturity, indications for surgical removal are pain at a known location of an osteochondroma or a rapidly growing tumor. Therefore, it is not uncommon for patients to undergo numerous surgical procedures throughout their lives to remove painful or deforming bony tumors, correct limb length discrepancies and improve joint range of motion.

MHE can affect various bones in the lower extremities including the pelvis, legs, ankles and feet. Osteochondromas of the hip are common and these may impinge on the hip movement. The hip also may start to dislocate. Both of these issues can be managed, often surgically, by removing the tumors and at times by redirecting the ball or socket of the hip to maintain the normal roundness of the hip and prevent arthritis. Osteochondromas of the knee (distal femur and proximal tibia) can be large, symptomatic and prominent. Patients may feel ‘catching’ within the knee joint during flexion and extension. In the ankle, a valgus deformity is very common, meaning that there is an outward (lateral) angle to the joint. We have designed operations to correct this problem. Another common issue with MHE is a limb length discrepancy due to growth inhibition.

Multiple areas of the upper extremities are also affected by MHE. From the shoulder all the way down to the fingers, the function of the forearm and wrist are often a main concern. The shoulder blade (scapula) is a common site for osteochondroma growth that can cause pain and nerve entrapment. The upper arm (humerus) is also prone to bony tumors. Depending on the degree of discomfort caused to the patient, osteochondromas in the humerus can be left in place or removed. Because the forearm is made up of two bones (radius and ulna) it can be particularly problematic. Osteochondromas that grow between the bones cause pain and limited movement. It is important that we pay close attention to these and remove them as soon as possible in order for the patient to maintain movement and range of motion and eliminate interference with musculature. Another common issue that may present in the forearm is differential growth of the ulna versus the radius which causes bowing. Treatment depends on the classification of the deformity and options will be discussed in length with the patient and their family.

We at the Paley Orthopedic & Spine Institute recommend that all patients with MHE undergo a spinal MRI by age eight. Spinal involvement may be present and studies show that a number of patients experience encroachment of the spinal canal. This is dangerous for numerous reasons and our goal is early detection.

When the decision to have surgery is made, the patient's individual needs before, during and after surgery should be anticipated. For children, Jessie Smith, our certified Child Life Specialist and Patient Coordinator and Kaile Jo Scott, our certified Child Life Specialist, are available prior to surgery and throughout the entire process to make things a little less scary, easier to understand and even fun. If you have questions or concerns regarding your child's experience with surgery please reach out to them.

- **Jessie Smith: jsmith@paleyinstitute.org or (561) 866-6866**
- **Kaile-Jo Scott: kscott@paleyinstitute.org**

Sarah Ziegler is our Rare Bone Disease Coordinator at the Paley Orthopedic & Spine Institute and head of the MHE Research Foundation. Please reach out to her with any questions or concerns at **sziegler@paleyinstitute.org** and **(561) 844-5255 ext. 237.**

How To Prepare For Surgery

In preparation for surgery, you will be scheduled to have Pre-Admission Testing (PAT) completed prior to surgery in the Kimmel building. At this appointment a nurse will perform a blood test and basic vital signs, unless you have already done so with your primary care provider within thirty days of the surgery date. Specific instructions such as when to discontinue eating and drinking the night before surgery will be discussed. A cleansing soap will be provided and must be used while showering the night before and morning of surgery. You will also meet with anesthesia at this appointment to discuss the process of 'going under' and postoperative pain control options. You will have a PAT appointment scheduled the same day as your preoperative consultation with Dr. Feldman and his physician assistants. Additional x-rays may be performed and you will then go over your surgical plan and post operative expectations. Please voice any questions or concerns to Dr. Feldman and his team at that time.

It is important that you discontinue all anti-inflammatory medications (NSAIDS), both prescription and over the counter, for two weeks prior to surgery and three months after (unless otherwise instructed). These include: Advil, Aleve, Motrin, Ibuprofen, Naprosyn or Naproxen, Celebrex, Celecoxib, Voltaren, Diclofenac, Toradol, Ketorolac, Mobic, Meloxicam, etc. In addition to NSAIDS, you should also discontinue all blood thinning medications such as aspirin (unless otherwise instructed by your medical doctor or cardiologist), multi-vitamins and any over the counter supplements at least seven days prior to your surgery date. You will be instructed when to resume these upon discharge from the hospital. If you have any questions regarding current medications and whether or not they can be taken prior to surgery, please reach out to Dr. Feldman's physician assistants.

You cannot be exposed to first hand or second hand smoke of any kind for one month prior to surgery and for at least six months following surgery. It is very important that you notify us if you have a personal history or a family history of early cardiac disease, phlebitis, blood clot

to the leg or lung (pulmonary embolism) or a history of a bleeding disorder. If you cannot receive blood products due to a religious reason or have an objection to receiving blood products for any other reason, you must also notify us of this before surgery.

In the event that you have any scheduling issues with your pre-operative appointment or surgery date, please contact Dr. Feldman's Executive Assistant, Jennifer Enterkin at **jenterkin@paleyinstitute.org or (561) 844-5255 ext. 310.**

Prior to surgery, you may be sent for additional imaging over at St. Mary's Hospital main building. Depending on the location of the osteochondroma that will be removed, Dr. Feldman may request such additional imaging such as an MRI in order to confirm the relationship of the osteochondroma to the nerves, arteries, and veins. This allows for advanced surgical planning. If you need assistance scheduling an MRI, CT Scan or additional x-rays, please contact Maribel Almonte at **malmonte@paleyinstitute.org and (561) 844-5255 ext. 309**

It is preferred that these additional studies be completed at St. Mary's Hospital. If this is not feasible, you may opt to have the study performed at an outside facility. In that case, the front office will send you the Rx to do so. If you have the study performed at an outside facility, you MUST bring the study with you on a CD-ROM to your pre-operative appointment or mail it in advance to our office with attention to Africa Tyrell. There is also a program that we can help you to download your CD to our system. The option of uploading the images yourself using My Medical Images. Please go to **mymedicalimages.com**, create an account and select to send the images to dfeldman@paleyinstitute.org. There is a one time annual storage fee required by MMI. Africa Tyrell, information below can help with this. If you prefer to mail the disc then address as below:

The Paley Institute
Attention: Africa Tyrell
901 45th Street
West Palm Beach, Florida 33407

Should you have further questions on how to mail in a copy of the disc, you can reach out to Africa Tyrell at **atyrell@paleyinstitute.org** or **(561) 844-5255 ext. 234**

If you have any clinical questions or concerns for the team, please email Dr. Feldman and/or his PA's who will gladly get back to you with their responses as promptly as possible.

- **David Feldman, MD: dfeldman@paleyinstitute.org**
- **Tiffany Brown, PA-C: tbrown@paleyinstitute.org**
- **Lauren Moir, PA-C: lmoir@paleyinstitute.org**
- **Alyssa Clarke, PA-C: aclarke@paleyinstitute.org**

If you need assistance with lodging, **Jessie Smith** can assist you with making lodging accommodations. Her email is: **jsmith@paleyinstitute.org** or she can be reached by phone: **(561) 866-6866**.

What to Expect the Day of Surgery and During the Hospital Stay

You will be provided with your arrival time a day or two prior to surgery. Please arrive at the Kimmel Outpatient building at the designated time you were provided. Surgery will take place, most often, in the Kimmel building and you will then if admitted be transferred via an underground walkway to Waters 3 (adult) within St. Mary's or Palm Beach Children's Hospital (children).. The length of your surgery will depend on the procedure (location, how many osteochondromas are excised, etc.) and will be estimated and discussed with Dr. Feldman during your preoperative consultation.

If you are an inpatient, after surgery, you will be followed daily by our clinical staff which includes Dr. Feldman and other Paley Institute physicians, Tiffany, Alyssa, and Lauren or other Paley Institute physician assistants as well as Marcia and Osiris, our nurse practitioners, who work on the floors of the hospital.

Post-operative pain control varies depending on the individual patient. You will receive pain medication intravenously or orally, and will be discharged home with oral pain medication. As you progress in your post-operative recovery, pain medication should be weaned in a tapering fashion. The ONLY over the counter medication that is acceptable to take for pain relief after surgery is Tylenol (unless otherwise instructed).

If you are an inpatient, on post-op day one, a physical therapist will come to your room to mobilize and you will continue to receive physical therapy daily during your hospital admission. The therapist will review proper body mechanics and positioning with you as well how to use any assistive devices such as crutches or a walker. There are most often no restrictions with regards to your sleeping position.

All durable medical equipment (DME) such as a walker, crutches, shower chair, wheelchair etc. will be provided to you before you are discharged from the hospital. **Emily Ward** can assist with any DME you will require, her email is **eward@paleyinstitute.org**.

Some surgeries are considered outpatient or ambulatory, meaning that you are discharged from the hospital the same day as your surgery. You will be given all medically necessary equipment and medication prescriptions prior to your discharge from the Kimmel recovery room. You will be discharged home with paperwork providing specific instructions for wound care, showering, and physical activity.

What to Expect Once You Leave the Hospital

Outpatient surgery, that is you return home, to a hotel, or to the Quantum House the same day as surgery, is for minor to moderate surgery where hospitalization is not needed. You will be given all equipment and pain medication prescriptions prior to your discharge from the Kimmel recovery room. If an elastic bandage feels tight, it can be loosened. Instructions about wound care and showering will be explained before discharge on a case by case basis. Typically, the bandage over your incisions should be left in place until seen at your first follow up appointment. Each operation is different and a protocol will be explained and detailed to you prior to surgery and before you leave the hospital.

Your sutures will usually be absorbable (clear) or more rarely ones that need to be removed (black) in the clinic/office at the Paley Institute. If you have absorbable ones, they will dissolve in two to three weeks. You may notice a long piece of clear suture coming from each end of your incision, these strings will be removed for you at your first post-operative appointment. If you have black stitches, these will be removed by a medical assistant in clinic two to three weeks after your surgery date. If you have dissolvable (clear) sutures, you will have steri-strips applied to the incision, which is then covered with a waterproof tegaderm dressing. The steri-strips will begin to fall off on their own. Please, do not actively remove them. Avoid submerging yourself in a bathtub or swimming pool for four weeks until the incision(s) are completely healed.

Upon discharge from the hospital, you may begin to follow the prescribed physical therapy regimen given to you by the St. Mary's inpatient physical therapy team. Physical therapy is vital to the success of your surgery, allowing motion that was achieved intra-operatively to be maintained post-operatively. Your rehabilitation will depend on the location of your surgery, the extent of the surgery, the amount of function that is lost by the exostoses and your level of functioning prior to surgery. During rehabilitation, therapy occurs daily and includes stretching and strengthening of the muscles around the surgical area. After your hospital discharge, you will receive a call from the Paley Institute Physical Therapy department to schedule appointments three to five days a week. It is important that you attend all scheduled physical therapy sessions as well as diligently do your prescribed exercises at home. Be mindful of the number of physical therapy sessions your insurance company will allow. Please contact your insurance company to determine what the coverage is for physical therapy to eliminate any confusion.

Your first postoperative appointment is typically scheduled ten to fourteen days after surgery. At your appointment, often x-rays will be obtained and you will have your first wound check. If you need any splints or braces, you will be fitted for them at this appointment. You will receive splints from occupational therapy the same day, the turnaround time for custom made machined orthotics is typically one week.

Pain medication, if it is narcotics, will legally need to be re-ordered with a visit every week.

Your second postoperative appointment will be scheduled at the time of the first.

Returning to school or work will be discussed after your first postoperative appointment but is ordinarily outlined before surgery. We will be happy to provide any documentation or forms required by your school or work. Please contact our medical assistants **Keisha Bourne** who can be reached at **kbourne@paleyinstitute.org** or **(561) 844-5255 ext. 240** and **Dalia Hanna** who can be reached at **dhanna@paleyinstitute.org** or **(561) 844-5255 ext. 243**.

If after surgery you need to reach us for a medical question, we can be reached at **(561) 844-5255**. There is someone on-call 24 hours 7 days a week. For medical emergencies, please call **911** and go to the nearest emergency room. If you are staying locally, St. Mary's Emergency Room is the most convenient for Dr. Feldman and his team to be involved in your care.

For more MHE patient resources, please contact **Sarah Ziegler** at **sziegler@paleyinstitute.org** and **(561) 844-5255 ext. 237.**