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ARTHROGRYPOSIS

Arthrogryposis literally translates to “stiff joints”. For most patients, there is no known cause for this condition. Arthrogryposis Multiplex Congenita (AMC) is this most common type. In fact, patients with AMC or Amyoplasia should be discussed specifically and not confused with genetic forms of stiff joints. These other forms with a genetic basis which include Escobar Syndrome, Popliteal Pterygium Syndrome, Polymicrogyri, Distal Arthrogryposis, Larsen Syndrome, conditions caused by RYR1 Mutations as well as others.

AMC will present at birth or even in utero with lack of fetal movement. The child often appears the worst in the newborn period and it can be quite frightening for the parents and the physicians who are unfamiliar with this condition. The stiff child has been cramped in the mother’s uterus for nine months and as a result, it can take up to two years to know what function will return. Physical therapy helps a great deal in infancy as it is determined how much can resolve and which muscles are present and which are absent. This diagnosis may impact the arms, legs and trunk causing the child to be weak and stiff. It can also affect the spine. The severity of the condition varies from person to person but encompasses joint contractures that are present at birth.

Affecting approximately 1 in 10,000 people, arthrogryposis has no known cause and when tested genetically is normal. Whether it is a failure of migration of tissues during fetal development not allowing muscles to form or a viral illness that impacts the early embryo/fetus, there is no known cure or prevention.

Less commonly, individuals have a genetic form, such as distal arthrogryposis, that predominantly involves the hands and feet or Escobar Syndrome which is characterized by webbing at the joints, called pterygium.

Given that the condition is relatively rare, few surgeons and therapists are experienced enough to treat this effectively. AMC requires very complex treatment and should only be undertaken by physicians, surgeons, and allied health professionals who are not only familiar with the condition but also have a high level of expertise in treating these patients.

We believe that if an individual is to have surgery, there must be an expected outcome that can change their life. To achieve the best functional outcomes, we take into account the underlying muscle strength of the patient, outline realistic goals, assess the potential benefits of treatment, and partner with an exceptional team of experienced medical professionals to provide treatment. This ensures that patients do not undergo multiple painful surgeries that have minimal positive impact on their condition.

I have been working with AMC patients for more than twenty five years and over the past five years, I and the exceptional team at the Paley Orthopedic & Spine Institute have made great strides in treating the spine, arms, hips, knees and feet in patients with AMC.

Most often initial evaluations include an appointment with me, a physician assistant, physical therapist and occupational therapist. We create an extensive treatment plan in order to achieve hips and knees with increased range of motion and flexibility, without the use of external fixation and without causing increased stiffness, arms that can bend and even actively flex (with muscle transfers) and spines that can move but not progressively curve. Each patient is unique and for some where muscles are more present more can be achieved and others less, but almost every patient can function better than the natural history.

We avoid external fixation because, while they achieve straight limbs, as I performed until 2014, they cause even more stiffness. We know, having a leg that is straight but stiff allows the child to walk if before they could not, however in the long run this is a significant disability for activities of daily living. If the knees cannot bend, as the child grows it is difficult to sit in classrooms, cars, airplanes, theaters, work desks, etc. The goal is to increase the arc of motion, not lessen it.

Fixed, flexed knees are corrected to fully straighten and maintain a good deal of bending. Elbows can be loosened and muscles transferred to allow for active elbow flexion. Wrists can be brought up from the flexed position if this allows the person to be more functional. In addition, thumbs can be brought out of the palm and fingers separated. Patients with AMC may develop scoliosis, a twisting and curving of the spine, and kyphosis, bending forward of the spine. These must be treated very carefully in AMC patients so that the ability to sit or stand is not lost.

While each patient is individualized, as the severity of the disease varies greatly between patients, AMC can be managed and often patients can be helped to be much more independent than ever thought before.

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. After your appointment in the Kimmel building, you will then go up to the Paley Institute clinic to see one of the physician assistants and Dr. Feldman. They may take another set of x-rays. You will then go over your surgical plan. Please voice any questions or concerns to Dr. Feldman 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 and (561) 844-5255 ext. 310**

Prior to surgery, you may be sent for additional imaging over at St. Mary's Hospital main building. 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 have the study performed elsewhere. 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.

The Paley Institute
Attention: Africa Tyrell
901 45th Street

West Palm Beach, Florida 33407

Should you have further questions regarding imaging studies or 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**

You also have 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.

If you have any clinical questions or concerns for the team, please email them and they will respond within a few days.

- **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 at **(561) 866-6866**.

<u>What to Expect the Day of Surgery and During the Hospital Stay</u>

A day or two prior to surgery you will be given your arrival time. 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 be admitted and transferred to St. Mary's/Palm Beach Children's Hospital or Waters 3 (adults aged 18+). Surgery time depends on the procedure and is often estimated and discussed during your pre-surgical visit. The length of stay in hospital, outpatient versus inpatient is also discussed at that time.

For children, Jessie Smith, our 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**

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 get you mobilized 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 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**.

What to Expect with Flexion Deformity Surgery

For correction of flexion deformities of the knees, soft tissue releases are performed in conjunction with surgically cutting and shortening of the long bone to achieve full extension. You will likely be placed in a knee immobilizer postoperatively instead of a cast to prevent stiffness. This should be worn around the clock to maintain extension achieved in the operating room. It can be removed for physical therapy and stretching exercises done at home. The limb will also be wrapped in an ace wrap and webril which can be removed seventy two hours after surgery.

The surgical dressings should be left in place until you are seen at your first follow-up appointment two weeks after your surgery date. At this appointment, the bandages will be removed, your wounds will be checked, and x-rays will be obtained. At this point, you will likely be measured for bracing such as a HKAFO or KAFO which typically takes a week or so to complete.
At four weeks post-op, Dr. Feldman will assess the fit of the braces and another x-ray will be taken to confirm sufficient healing has taken place and you will likely progress to weight bearing as tolerated and can begin gait training in physical therapy.

What to Expect with Latissimus Transfer Surgery

Many arthrogrypotic patients lack the biceps brachii muscle which is necessary to perform so many daily activities, including eating. Dr. Feldman specializes in the transfer of the latissimus dorsi or pectoralis major muscle to this location which can give you increased independence. Prior to surgery, you will have a chest MRI taken to assess the availability of muscle to transfer.

The procedure involves the latissimus dorsi or pectoralis major being transferred and secured with an insertion site mimicking that of the typical biceps brachii. This will enable you to achieve more function of your upper extremity. Primarily, the goal is the ability to actively, independently bring your hand to your mouth without assistance. The muscle transfer is done carefully as to maintain innervation and circulation to the muscle.

Our occupational therapists will create a custom molded splint that will be worn around the clock postoperatively. It is imperative to the success of the surgery that the tendon is given time to heal at the insertion site. To do so, the arm cannot be extended past 90 degrees and no active motion using the muscle should occur prior to six weeks post-op. Our occupational therapists will work diligently on passive range of motion, training you to fire the muscle and, once cleared by Dr. Feldman, active flexion using the muscle. Active flexion is likely to happen around six weeks post-op.

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 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 postoperative appointment. If you have black stitches, these will be removed by a medical assistant in clinic two to three weeks after your surgery date. You will have steri-strips applied to the incision, which is then covered with a waterproof tegaderm dressing. The tegaderm will remain in place for one week. You may remove this tegaderm island dressing and shower on postoperative day seven. You should recover the incision and steri-strips with sterile gauze and paper tape after each shower. Do this dressing change daily until seen in your follow up appointment. 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 your incisions 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 usually vital to the success of your surgery, allowing motion that was achieved intra-operatively to be maintained post-operatively. 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 braces 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.

Your return to school or work will be discussed after your first post-operative 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.