

ARTHROGRYPOSIS

PATIENT CARE GUIDE

EVERYTHING YOU NEED TO KNOW

Arthrogryposis is a condition that may impact the arms, legs and trunk causing them to be weak and stiff. It can also effect the spine. The severity of the condition varies from person to person but encompasses joint contractures that are present at birth. It may be referred to as Amyoplasia or Arthrogryposis Multiplex Congenita.

Most forms of arthrogryposis have no known cause. Less commonly individuals have a genetic form, such as distal arthrogryposis, that predominantly involves the hands and feet. Another form of arthrogryposis, Escobar syndrome, has webbing at the joints, called pterygium.

AMC affects approximately 1 in 10,000 people. It is characterized as malformed and stiff joints causing limited mobility and function. The contractures are caused by short and abnormally tight soft tissues which prevent the extremities from moving properly.

The treatment of arthrogryposis varies based on the individual and their specific deficits. It remains a collaborative effort implementing physical and occupational therapy, casting, and surgical interventions to achieve a desired outcome. These modalities are integrated to improve baseline physical abilities, primarily the ability to ambulate. Treatment at a young age allows children to gain functionality and become stronger enabling them to have improved joint mobility and function. This provides the foundation for improved long term outcomes of independence.

There are certain underlying principles of treatment and indications for intervention. We recommend surgery if a significant deformity is present with involvement of the joint leading to a loss of range of motion and function, specifically in the lower extremities which are necessary for mobility. Each surgery is specific to the type of deformity and the location of the deformities, and will be discussed at your consultation/preoperative visits. The goal is not just to make a straight limb but one that moves and has an increased range of motion.



ORTHOPEDIC & SPINE INSTITUTE
at St. Mary's Medical Center

901 45th Street, Kimmel Building - West Palm Beach, FL 33407 • Phone 561.844.5255 • Fax 561.844.5245

NON SURGICAL TREATMENT

SERIAL CASTING & ACHILLES TENOTOMY. Paley Orthopedic and Spine Institute uses the Ponseti serial casting method with Ponseti trained physical therapists to treat clubfoot. The Ponseti method of clubfoot correction consists of manipulation of the foot by abducting the foot in supination while applying counter pressure over the lateral aspect of the head of the talus to prevent rotation of the talus at the ankle. All components of clubfoot deformity are corrected simultaneously, except for the ankle equinus. Manipulation is performed for approximately one minute followed by application of a well molded, long leg plaster cast. Casts are applied and worn for four to six days. The cast will be removed in the office and the feet will be manipulated further to achieve greater correction and then another cast will be applied. Most children undergo 3 weeks of serial casting with two casts per week. However, this may vary based on the initial severity and the correction achieved with each cast.

Approximately 80 percent of infants with clubfoot require a tenotomy of the Achilles tendon in order to achieve full correction of clubfoot. Depending on the child's age, this non-invasive procedure may be done by the surgeon in office or with sedation in the operating room.

If the tenotomy is required, the last cast remains in place for three weeks to allow time for the tendon to heal. Fully reduced clubfoot should demonstrate 70 degrees of abduction and 15 degrees of dorsi flexion. Once this is achieved, the patient is ready for the next phase of treatment.

To evaluate and schedule clubfoot casting please contact the **Rehabilitation Department** at **(561) 844-7878**.

OCCUPATIONAL THERAPY AND SPLINTING

Occupational therapy (OT) is a multidisciplinary treatment designed to increase functioning, enhance motor skills, and prevent disability in patients with musculoskeletal conditions and injuries. When it comes to OT, "occupation" isn't limited to a job; it refers to any type of meaningful activity.

The therapy the patient receives depends on the type and severity of the patient's condition and the goals you want to achieve. Our occupational therapists take a whole-person approach, working with the entire team at Paley Orthopedic & Spine Institute to ensure all of the patient's health care needs are incorporated into OT.

The occupational therapist may recommend splinting for the patient's hand, fingers, or toes to help capture and maintain range of motion. The splints are made in office during an occupational therapy appointment. The splinting regimen may include suggested hours to wear the splints, when to remove the splints for stretching, and multiple visits to receive new splints to help capture any range of motion gains.

To evaluate and schedule occupational therapy please contact the **Rehabilitation Department** at **(561) 844-7878**.

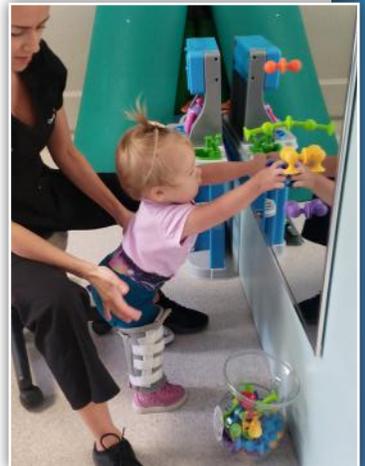
HOW TO PREPARE FOR SURGERY:

The patient will be scheduled to have a PAT (Pre-Admission Test) before surgery. At this appointment they will have blood work completed, if he/she has not already done so with their primary care provider. Specific instructions such as when to discontinue eating and drinking the night before your surgery will be discussed. As well, a scrub to be used while showering the night before and morning of surgery will be provided. You will also meet with anesthesia at this appointment to discuss postoperative pain control options.

The patient should be off all anti-inflammatory medications/NSAIDS (both prescription and over the counter including: advil / aleve / motrin / ibuprofen / Naprosyn / naproxen / Celebrex / celecoxib / voltaren / diclofenac / toradol / ketorolac / Mobic / meloxicam, etc) for two weeks prior to surgery and 3 months after.

The patient cannot be exposed to first hand or second hand smoke of any kind for one month prior to surgery and for at least 6 months following surgery.

The patient should discontinue all blood thinning medications such as aspirin, multi-vitamins, over the counter supplements, etc. 7 days before surgery. You will be instructed when to resume these upon discharge from the hospital.





You **MUST** notify us if the patient has a personal history or a family history of early cardiac disease, phlebitis, blood clot to the leg or lung (pulmonary embolus) or a history of a bleeding disorder.

If the patient cannot receive blood products due to a religious reason or they have an objection to receiving blood products for any other reason, you must notify us of this before surgery.

"All scheduling for surgery should be referred to **Jennifer Enterkin** - jenterkin@paleyinstitute.org or 561-844-5255 ext. 310

Psychosocial support in preparing your child for surgery or inquires about touring the hospital prior to surgery should be referred to the Certified Child Life Specialist, **Kaile Jo Scott** - kscott@paleyinstitute.org

Clinical Surgical questions for the team is best emailed and we will usually respond within a few days.

David Feldman, MD

dfeldman@paleyinstitute.org

Tiffany Brown, PA-C

tbrown@paleyinstitute.org

Nicole Odonnell, PA-C

nodonnell@paleyinstitute.org

Lauren Moir, PA-C

Imoir@paleyinstitute.org



The patient may be sent for additional imaging prior to surgery. If you need assistance scheduling such studies, please contact **Maribel Almonte**, her email is malmonte@paleyinstitute.org, and she can be reached at 561-844-5255 extension 309. It is preferred that these studies be done at St. Mary's Hospital, if this is not feasible, you may have the study performed elsewhere and the front office will send you the prescription to do so. If you are having 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, attention **Africa Tyrell**. She can be reached at atyrell@paleyinstitute.org or at 561-844-5255 ext 234.

The Paley Institute

ATTN: Africa Tyrell

901 45th Street

West Palm Beach, Florida 33407

WHAT TO EXPECT ON THE DAY OF SURGERY AND DURING YOUR HOSPITAL STAY:

The patient will arrive at the Kimmel Outpatient building at the designated time you were provided. Surgery will take place in the Kimmel building, and they will then be admitted and transferred to St. Mary's/Palm Beach Children's Hospital. The surgery typically takes between 3-4 hours and the expected inpatient hospital stay ranges from 3-4 days, depending upon the individual's progress. They will followed daily by our clinical staff which includes physicians, PA's and/or ARNP's.

Post-operative pain control varies depending on the individual patient. The patient will be receiving pain medication intravenously as well as orally, and will be discharged home with oral pain medication. As they progress in their 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 is Tylenol. Again you must refrain from all anti-inflammatories for approximately the first 3 months after surgery.

The patient will be mobilized the first day after surgery by physical therapy (PT) and will receive PT daily during your hospital admission. PT will review proper body mechanics and positioning with you as well. There are no restrictions with regards to position of sleep.

All durable medical equipment (DME) such as a walker, 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.

The patient may be in a knee immobilizer immediately following surgery, this is a removable brace that will protect the limb until the braces are made. Knee immobilizers should be worn intermittently throughout the day, removed for physical therapy, and slept in nightly. The patient will be molded for the braces at their first post-operative appointment (2 weeks post surgery).

If the patient has a urinary catheter placed during surgery, this will be removed on post-operative day #2. The patient may also have drains along your incisions which are typically removed just prior to discharge from the hospital.



There is the possibility the patient will require a blood transfusion based on the amount of blood loss during surgery and what the post-operative labs are, however this is unlikely that they will require a transfusion. If the patient opted to donate blood beforehand, these units of blood will be given to them in lieu of receiving blood from the hospital blood bank. It is best to have a trusted family member or friend donate on behalf of the patient, rather than depleting the patient's blood volume just prior to surgery. Our office can assist with how to donate blood on behalf of the patient; this is done through the One Blood donation centers.

WHAT TO EXPECT ONCE YOU LEAVE THE HOSPITAL

The first postoperative appointment will be between 10-14 days after surgery. If you are from out of town, it is typically advised that you remain local for 3 weeks. This will allow for close monitoring during the post-operative phase. If you are from out of town and need assistance with lodging, **Jessie Smith** can assist with making lodging accommodations. Her email is jsmith@paleyinstitute.org, and she can be reached at **561-866-6866**.

WOUND CARE: The suture is an absorbable suture which will dissolve in ~2-3 weeks. You may notice the clear suture coming from each end of the incision, these strings will be removed for you at your first post-operative appointment. You will have steri-strips applied to the incision, which is then covered with a waterproof island tegaderm dressing. This island dressing will remain in place for one week. The patient may shower on post-operative day #7, do not remove the tegaderm island dressing. After showering, pat dry, do not rub the tegaderm island dressing. At the first follow up appointment, the clinic staff will remove the dressing.

PHYSICAL THERAPY: The patient will likely be placed in a knee immobilizer post-operatively instead of casting to prevent stiffness. Upon discharge from the hospital, you may begin to follow the prescribed PT regimen. PT is vital to the success of the operation and allows the motion that was achieved intra-operatively to be maintained post-operatively. It is important that you attend and diligently partake in the prescribed exercises. It is important to 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.

AT HOME STRETCHING: Following surgery, physical therapy becomes of utmost importance to maintain the range of motion achieved in the operating room. Physical therapy sessions will begin while inpatient and continue once you are discharged from the hospital. By the end of week one, physical therapists will have taught you at home stretches to be done daily for the hip, knee, and ankle. These should be done daily in addition to formal PT. These basic stretches will emphasize focus on the motion that was most deficient prior to surgery. For example, if the knees were stuck in flexion prior to surgery, extension will be the focus post-operatively.

FIRST POST-OP APPOINTMENT: The first postoperative appointment is typically scheduled between 10-14 days after surgery. X-rays will be obtained at this time and the first wound check will be performed at this visit. The patient will be fitted for bracing at this appointment, and the turnaround of the bracing to be made is typically one week, thus staying in town locally for 3 weeks should allot enough time for the braces to be made and ready prior to your departure.

SECOND POST-OP APPOINTMENT: Your second postoperative appointment will be scheduled approximately 4-6 weeks after surgery if you are local. X-rays will be taken at this visit. If you are not local, we will provide you with a prescription to have x-rays taken which you will then send to us for review. Please send imaging to the below address:

Paley Orthopedic & Spine Institute
ATTN: Africa Tyrell
901 45th Street
West Palm Beach, Florida 33407

PAPERWORK

The patient can typically return to school/work after their first post-operative appointment so long as they are off all narcotic medications. We will be happy to provide any such documentation or forms required by your school or work, our medical assistants, **Keisha Bourne** (kbourne@paleyinstitute.org) and **Dalia Hanna** (dhanna@paleyinstitute.org) can assist with this and can also be reached at 561-844-5255, ext 240 and ext 243.

