

DAVID S. FELDMAN, MD

ORTHOPEDIC SURGERY

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ACHONDROPLASIA

Achondroplasia is an inheritable, short limbed, disproportionate form of dwarfism caused by the body's inability to convert cartilage into bone, especially in the long bones. Characteristics include but are not limited to a large head size, prominent forehead, small hands, feet and fingers, bowing of the lower legs, curvature of the spine (kyphosis) and limited range of motion in the elbows. The average height of an adult male with achondroplasia is 131 centimeters or 4 feet, 4 inches, and the average height for adult females is 124 centimeters or 4 feet, 1 inch.

The most common form of over two hundred different types of dwarfism, achondroplasia is inherited as a dominant trait. Eighty percent of cases, however, are due to a genetic mutation where the child is born to parents of average height. Achondroplasia can be diagnosed before birth with an ultrasound or amniocentesis. After birth, characteristic features can be confirmed by x-ray.

Achondroplasia affects approximately 1 in 40,000 individuals. It is cause for various health complications such as premature arthritis, sleep apnea, hydrocephalus, recurring ear infections, joint instability, lordosis or kyphosis (curvature of the spine), obesity and spinal stenosis (narrowing of the spinal column) among others.

My approach to achondroplasia at The Paley Institute takes into account the most common orthopedic problems and reasons for disability later in life. The impact of spinal stenosis (compression of the spinal cord and nerve roots) is taken into consideration beginning in infancy. It is important to avoid having an achondroplastic infant sit upright too early using devices like the Bumbo. Although this is controversial and some feel it does not play a role, I do believe that it may have an impact on the spine later in life. This can prevent kyphosis (round back).

Children with achondroplasia generally have delayed milestones and will begin to sit, stand and crawl once their neck and back muscles are fully developed. It is recommended that an infant diagnosed with achondroplasia be seen by a neurosurgeon for assessment of the foramen magnum (area at the base of the brain where the spinal cord exits from the skull). In infancy a sleep study should be done and if there is apnea this needs to be assessed fully. It is either caused by obstructive apnea such as the tonsils being enlarged and/or compression at the foramen magnum. Often a decompression at the foramen magnum is needed.

Early interventions will have an impact on the achondroplastic individual's life forever. Yearly x-rays and asking questions, beginning in childhood and continuing throughout the patient's life, are important in determining if treatment is needed.

Deformity correction is performed to avoid arthritis later in life and involves straightening of the lower legs (tibia and fibula) and/or straightening of the forearm (radius and ulna). In addition, if the patient is experiencing instability in the knee, this can also be corrected.

As mentioned above, spinal stenosis, or compression of the spinal cord or nerve roots, is the most common complaint and source of disability for patients with Achondroplasia. Intervention and treatment should not wait until the individual is weak, paralyzed or urinary incontinent. There are solutions that are quite effective and can be life changing.

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. Her email is **malmonte@paleyinstitute.org** and you can reach her by phone **(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: **(561) 866-6866**.

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

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 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**

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**.

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 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. 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 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. 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 4 weeks until the incision(s) and/or pin sites 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.

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.