DAVID S. FELDMAN, MD

ORTHOPEDIC SURGERY

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LIMB LENGTHENING AND DEFORMITY CORRECTION FOR ACHONDROPLASIA

The stature of achondroplastic individuals falls significantly below the growth charts for most children. The average adult height for achondroplastic men at skeletal maturity is 131 centimeters or 4 feet three inches and for women is 125 centimeters or 4 feet 1 inch. In comparison, the low end of height at skeletal maturity for men without achondroplasia is 160 centimeters or 5 feet two inches, and for women is 151 centimeters or 4 feet 11 inches. Tall parental height is associated with a relatively taller height in achondroplastic children. The short stature of patients with achondroplasia is due to an independent growth pattern and not due to a slow growth rate.

Any person less than 5 feet in height will quickly discover that society may not be suitable for them. Reaching light switches, using public facilities as well as general living is impacted. Spinal growth is not affected to the same extent as limb growth in individuals with achondroplasia, thus individuals have a relatively normal size trunk, which is why limb lengthening is possible as a treatment, as it restores body proportion. We believe this is a patient and family decision and that lengthening in dwarfism should be available to those who wish to undergo such a procedure. Any person interested in lengthening of this nature should ensure they elect to have it done at specialized centers with extensive experience.. Long bone lengthening and/or deformity correction of the upper and lower extremities at the Paley Orthopedic and Spine Institute can be done safely, reliably, and reproducibly.

Our goals for treatment are to increase overall height, correct secondary deformities which occur in achondroplasia such as bow leggedness, improve reach, restore body proportions and preserve patient function.

Our treatment plan involves staged lengthenings, typically consisting of two to three lengthenings of the lower extremities and one lengthening of the upper extremities. Surgical

treatment aims to correct the upper and lower limb deformities while simultaneously increasing stature. This typically results in limb lengthening between 30-40 cm (11-15 in) of total added length for the average patient with dwarfism. Each lengthening procedure acquires approximately 10-15 cm (4-6 in) of length. The length achieved when lengthening the arms ranges from about 10-12 cm (4-5 in), which is performed at a separate stage from the leg lengthenings.

Lengthening can be achieved with internal versus external fixation, depending upon the length and diameter of the bone and whether or not the bone can accommodate an internal device. Most often lower extremity lengthening is performed with internal growing rods, at which point deformity correction can also be obtained. Lengthening and deformity correction is obtained by performing osteotomies (surgical breaking of the long bones), to mobilize the long bones so they are able to be distracted with either an internal or external device.

Because achondroplasia and other skeletal dysplasias are associated with spinal problems. The spine is evaluated from the base of the brain, to the neck, thoracic spine and to the lower back. Specific questions are asked, radiographs and often an MRI of the spine are obtained. If this is cleared and ok then it is safe to proceed with lengthening.

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, Dr. Huser 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 and Dr. Huser 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 and Huser’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 our Executive Assistant, **Jennifer Enterkin** at **jenterkin@paleyinstitute.org** or **(561) 844-5255 ext. 310**. All scheduling for follow up appointments should be referred to **Andrea Mower** at **amower@paleyinstitute.org** or **561-844-5255 ext. 245**.

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

For a faster process, you may upload the images yourself to the **My Medical Images** website. Please have the imaging facility place the images on a disc for you so that you can upload them yourself to the My Medical Images website at **www.mymedicalimages.com**. Please notify **Africa Tyrell** at **atyrell@paleyinsitute.org** or at **(561) 844-5255 ext. 234** when you are ready to upload your images so she can send you an email with a one-time free upload link. She will need the patient's full name, date of birth, which email address to use to send you the link, and the type of imaging being uploaded (x-ray, MRI, CT, etc). Once you receive the email with the link, follow the prompts to upload your images. There is no need to create an account.

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**

Please keep in mind the following:

- The site is not supported by IE (internet explorer) MUST use Google Chrome or Safari.
- You will need a CD drive/reader to upload the images.
- There is NO need to create an account to use the one time free upload
- However if you choose to create an account, the cost is approximately $30/year. You may use referral code ORTHOMD for $10 off for the first year.
- If you have any trouble uploading the images please reach out to My Medical Images support line at **(855) 800-2851**.
- Once you have completed the process, please once again notify Africa Tyrell who will transfer all imaging from the MMI site to our Paley PACS System and notify Drs. Feldman and Huser's team that they are available for review. You can expect a call or email once these images have been reviewed after a few days.

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**
- **Aaron Huser, DO: ahuser@paleyinstitute.org**
- **Tiffany Brown, PA-C: feldmanpa@paleyinstitute.org**
- **Alyssa Clarke, PA-C: feldmanpa@paleyinstitute.org**
- **Katie Gauger, PA-C: feldmanpa@paleyinstitute.org**

Kristen DeAndrade is Director of Patient Advocacy for Dr. Feldman and Dr. Huser. As a former patient, she is familiar with the surgical and rehabilitative process, and the highs and lows that patients and their families face before, during and after treatment. She has undergone extended limb lengthening, deformity correction, and spinal fusion surgery. Kristen is a direct connection to Dr. Feldman, Dr. Huser and the team and is available to help make the journey as manageable for patients and their families as possible. From a patient standpoint, she can be a valuable resource, please do not hesitate to reach out to her.

- **Kristen DeAndrade**
  Patient Advocate for Dr. Feldman and Dr. Huser
  kdeandrade@davidsfeldmanmd.com

Mia Johnson is the Family Liaison for Dr. Feldman and Dr. Huser. Mia and her husband adopted 4 children internationally with orthopedic needs, who are patients of Dr. Feldman and Dr. Huser. She has thorough experience with limb length discrepancy, cerebral palsy and skeletal dysplasia as well as various orthopedic care procedures, pre-surgical planning and rehabilitation. Mia is available to provide patients and families with resources and advice in regards to lodging, clinic visits, hospitalization and support services.

- **Mia Johnson**
  Family Liaison for Dr. Feldman and Dr. Huser
  mjohnson@davidsfeldmanmd.com

******Find Mia and Kristen on Facebook, in the group 'Patients and Families of Dr. David Feldman at The Paley Institute (https://www.facebook.com/groups/patientsofdrdavidfeldman) where they can help answer questions and you can connect with other patients and families.

If you need assistance with lodging, our Patient Coordinator, **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, our certified Child Life Specialist, **Kaile Jo Scott**, is 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 her at **kscott@paleyinstitute.org or (561) 334-9135.**

If you are an inpatient, after surgery, you will be followed daily by our clinical staff which includes Dr. Feldman, Dr. Huser and other Paley Institute physicians, Tiffany, Alyssa, and Katie 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 management 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 During Lengthening

At the Paley Orthopedic & Spine Institute, we use a variety of orthopedic devices to lengthen bones. Based on each person's unique situation we will choose which device will work best. There are two phases to every limb lengthening procedure whether the device used is internal or external: lengthening and/or correction and consolidation. We typically begin with lower extremity lengthening between the ages of 8 to 10 years of age, with a total length gained of 4in or 10cm (5cm in the femur and 5cm in the tibia). The lengthening course for a 5 cm goal takes approximately 10-12 weeks of distraction, and an additional 10-12 weeks of consolidation (the time period in which the bones heal together). This equates to a total time being in the external fixators somewhere between 5 and 6 months for a 10cm lengthening.

We do request that you remain local during the lengthening/deformity correction phase to attend daily physical/occupational therapy and for frequent follow up visits and x-rays with Dr. Feldman, Dr. Huser and their team. **Jessie Smith** can assist with making lodging accommodations. Her email is **jsmith@paleyinstitute.org**, and she can be reached at **(561) 866-6866.**

Once the desired correction and/or length has been achieved, you may return home for the consolidation phase. Monthly x-rays can be sent to our team to assess your healing progress and determine when a timeframe for removal of the lengthening devices.

If you wish to move forward with additional lengthenings, then the second lengthening can be scheduled around the age of 13 and again is achieved with either internal or external fixation. Typically by this age and after the long bones have undergone one lengthening, they can accommodate internal fixation in all 4 segments (bilateral femur and tibia). If all 4 segments are being lengthened, an expected goal of 5-6 in or 12-15 cm (8 cm in the femur and 5-7 cm in the tibia) can be achieved, of course barring no complications and that you are able to maintain function and range of motion. Again, please anticipate staying local to the West Palm Beach area for 12-16 weeks while undergoing the second lengthening stage. You will require postoperative therapy and clinical follow-up appointments just as you did with the first lengthening procedure.

The third lengthening course involves lengthening of the upper extremities (humerus) and is achieved with external fixators. The goal of length in the upper extremities is dependent upon the lower extremity lengthening goal. If the patient anticipates a third lower extremity lengthening, to preserve proportion, we will lengthen the humerus to approximately 5 in or 12 cm. If the patient is not pursuing a third lengthening of the lower extremities, the recommended humerus lengthening would then be 4 in or 10 cm. You and your family may not have to remain local for the duration of this lengthening process. This can be determined by your progress and staying at least one month is advisable. Monthly x-rays from your primary care clinic can be sent

to our team for review and to determine when the external fixators can be removed from your arms.

The final lower extremity lengthening can occur around 16 years of age and a total of 6 in or 15 cm of length can expect to be achieved (8 cm in the femur and 7 cm in the tibia). The same protocol applies in terms of duration of treatment, needing to stay local and when one may return home.

What to Expect with Internal Fixation (Precise or Stryde)

Internal lengthening can be performed utilizing a device such as the PRECISE intramedullary nail or STRYDE. During surgery, a rod is placed in the long bone (femur and/or tibia) and Dr. Feldman and/or Huser creates an osteotomy (breaks the bone), allowing for distraction or new growth at that site. This rod can also correct any rotational malalignment simultaneously. If there is a rotational malalignment, you may be required to have a hip CT and/or MRI pre-operatively to further assess this.

The internal lengthening rod is magnetized which allows for lengthening to occur without turning screws on an external fixator. After surgery, patients are typically partially weight bearing with the use of an assistive device such as a walker. One week post-op, you will have a follow up appointment in the office and obtain your lengthening schedule. A typical schedule is 0.75 mm per day over the course of 3 sessions in a day. This may be sped up or slowed down depending on the healing process. You will also receive the magnetic device which will lengthen you the appropriate amount. The device will act as the external magnet and be applied to the surface of the limb to a specific area, typically marked using a Sharpie marker (some patients use temporary tattoos). You will hover the machine over the internal rod and its magnet. These two communicate and turn the mechanism allowing for elongation of the rod and thus lengthening of the bone. You will be given instructions for lengthening and placement of the external device at you one week post operative appointment as well. Lengthening sessions should be spaced out appropriately throughout the day, one in the morning, afternoon, and at night. The machine will automatically turn off once lengthening is complete during each session. It is imperative that you keep a regular lengthening schedule as the machine is set on a 24 hour clock and will only allow for the amount of lengthening programmed. Thus, if you are to lengthen 3 times a day, you will only be able to do so within that 24 hour period. If you lengthen at 1 am, this will go towards the next consecutive day and you will have missed a turn cycle. In the event that this happens, the missed lengthening can be made up in the office or at the end of your lengthening schedule.

You and your family must remain local to West Palm Beach during the lengthening treatment of the lower extremities to attend daily physical therapy as well as clinic visits with x-rays every 2 weeks to monitor your progress, ensure regenerate bone is being formed, and range of motion of the joints above and below are being maintained. With upper extremity lengthening, if the lengthening is being continued while you are home then x-rays are obtained every two weeks followed by telehealth appointments to determine if range of motion is adequate etc.

Once the desired amount of length is achieved, you will begin the consolidation phase and may return home for this portion of your treatment. During consolidation, the regenerate bone that formed at the distraction site will further strengthen and solidify. You will be required to send monthly x-rays to determine changes in weight bearing status and activity level. Once fully

healed and confirmed on x-ray, removal of the rod can be planned approximately six months to a year after you have completely consolidated. The rod removal is an outpatient, ambulatory procedure. After surgery, you will be weight bearing as tolerated and will go home from the hospital the same day.

What to Expect with External Fixation

Depending on the size of the long bones and/or degree of deformity, your lengthening and deformity correction may be done using external fixators using the Taylor Spatial frame ™, Drive rail ™, and/or Orthex frame ™. Lengthening with external fixation works on the same principle of bone regeneration as internal fixation.

During surgery, Dr. Feldman/Huser cuts the bone into two segments. Each of the two segments of the bone is then attached to a fixator with special screws and/or wires. For a circular fixator, each segment is attached to a separate ring. Circular fixators can have special rods, called struts, placed in between the rings. These struts allow the bone to be lengthened, rotated, and/or angulated in any direction. For a rail fixator, each bone segment is attached to an individual pin clamp. As the two bone segments are slowly pulled apart, new bone forms in the space between them. This new bone increases the overall length of the bone. Adjustments are made three times a day which equates to one millimeter a day.

Lengthening typically begins five to seven days after your surgery date. You will be scheduled to meet with one of our orthopedic technologists and they will show you how to lengthen and give you your turn schedule. Any other instructions will be provided to you at this time. As a general rule, patients are fully weight bearing with the external fixators in place.

After the correction has been achieved, the consolidation phase begins in which the regenerate bone slowly hardens. During this phase of treatment, the external fixator normally remains in place so that it can support the bone as it heals. The bone has consolidated when the new regenerate bone has completely hardened and calcified. The consolidation phase depends on how the individual patient heals. Typically we say one month of consolidation for every inch of new growth. So, if you lengthened a total of four inches (ten centimeters) you will consolidate in the frame for about four months.

Beginning the day after surgery, you will start with a physical therapist and learn how to walk and exercise with your fixator(s). You are able to put weight on the operated leg immediately after surgery since the external fixator will protect and support your leg.

Regardless if you are lengthening with internal or external fixators, once pin sites and/or all incisions are healed (about two weeks after your surgery date) you are allowed to swim. Pin site infections do occur with external devices in place, and are treated with oral antibiotics. We teach appropriate pin care instructions on how to clean the external device in an effort to minimize the risk of infections. All other instructions for distraction/correction will be provided to you post operatively.

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. Whether your procedure is inpatient or outpatient, prior to your discharge, you will be given all equipment and pain medication prescriptions. 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 re-cover the incision and steri-strips with sterile gauze and paper tape after each shower. Showers can begin two days after surgery and should be daily with normal soap and shampoo and they clean the skin around the pins. 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 3 weeks or 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 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 Rehabilitation 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.

If you need to schedule physical therapy appointments or have questions, please contact the **Rehabilitation Department** at **(561) 844-7878**.

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.

Returning 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**.

What to Expect after Consolidation

Normal childhood activities can be resumed in between lengthenings.

Hardware removals are considered an ambulatory or outpatient procedure, meaning the patient may go home the same day as surgery. Because the bone is theoretically weakened following removal of hardware, patients are sometimes placed in a cast or removable boot, or they may undergo prophylactic rodding of the long bones to protect the integrity of the bone and prevent fractures from occurring. A cast or boot is usually in place for about 4 weeks, depending on individual healing progress, and patients are fully weight bearing following removal of hardware, even if in a cast or boot. We do recommend that you remain local for a few days following hardware removals. Then you may either fly or drive home and the remaining postoperative care can be achieved from afar by sending x-rays and through telemedicine appointments.

Anti-inflammatory medications (Advil, Aleve, Ibuprofen, Motrin, etc.) are prohibited during the lengthening and/or deformity correction phase, however Tylenol is safe to take. Once your bones are healed and completely consolidated, such anti-inflammatory medications are safe to resume.

No outline can be complete or totally understandable. If you have any questions, please feel free to email us and hopefully we can help with all logistics and information.

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/Dr. Huser and their team to be involved in your care.