

DAVID S. FELDMAN, MD

ORTHOPEDIC SURGERY

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PSEUDOACHONDROPLASIA

Pseudoachondroplasia is an inherited bone growth disorder characterized by short stature, a large range of joint mobility issues, joint pain, deformity of the lower legs and is often accompanied with curvature of the spine. Pseudoachondroplasia is the result of a genetic mutation in the COMP gene which is a gene involved with the formation of cartilage.

Pseudoachondroplasia is inherited as an autosomal dominant trait, which means one copy of the altered gene in each cell is sufficient to cause the disorder. Most cases result from new mutations in the gene and occur in people with no family history of dwarfism. Children of an individual with pseudoachondroplasia have a fifty percent chance of inheriting the gene. This form of dwarfism is estimated to affect 1 in 30,000 individuals.

Individuals with pseudoachondroplasia are not unusually short at birth but by the age of two, their growth rate begins to fall below the standard growth curve. The average height of an adult male with this condition is 120 centimeters or 3 feet, 11 inches, and the average height of an adult female is 116 centimeters or 3 feet, 9 inches. Pseudoachondroplasia does have many of the observable features of achondroplasia and really should not have a similar name. It's major differences have to do with the lack of characteristic facial features in achondroplasia and its effect on the individual's joint cartilage. Therefore, a pseudoachondroplastic patient typically has more significant malalignment deformities of the lower limbs stemming from the hips, knees and ankles. This can also lead to an early onset of osteoarthritis. Some individuals have an unusually large range of joint movement (hyperextensibility) in the hands, knees, and ankles and a limited range of motion at the elbows and hips. In some cases, the legs may turn outward or inward, this is known as a valgus or varus deformity.

Spinal stenosis is almost unheard of in those with pseudoachondroplasia, while C1-C2 instability (neck instability) and scoliosis are quite common.

While pseudoachondroplasia is an uncommon condition, I have treated many children and adults with this form of skeletal dysplasia. Our approach to this type of skeletal dysplasia, here at the Paley Orthopedic and Spine Institute, involves aggressively maintaining the alignment of the limbs. This is done with physical therapy to strengthen muscles, judicious use of growth modulation and osteotomies (cutting of the bone) to maintain normal alignment and/or deepen the hip socket.

Spinal intervention is often necessary. This can range from physical therapy to neck and back spinal deformity correction. While we do not necessarily encourage lengthening of the limbs, I do perform this surgery when patients decide it is what they want. It can be done at the same time any deformity correction is performed.

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

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 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** or 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 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. Please mail in the disc, addressed as below:

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 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 the day 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 be admitted and transferred to St. Mary's/Palm Beach Children's Hospital. Length of surgery depends on the procedure and is estimated and discussed during your preoperative consultation.

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.

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

Your first postoperative appointment is typically scheduled ten to fourteen days after surgery. At your appointment, X-rays may be obtained and you will have your first wound check. Bracing may be ordered at that visit (turnaround time for braces is usually 7 days).

Your sutures will usually be absorbable (clear) or more rarely ones that need to be removed (black) in the clinic/office at the Paley Orthopedic & Spine 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 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.

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.