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Physical & Occupational  
Therapy

**CONSULTATION**

Name: **Krizanova, Natalia**

DOB: 11/21/2017

DOS: 11/22/2019

Physician: Dror Paley, MD, FRCSC

**History of Present Illness:**

Natalia is a 2-year-old girl from Slovakia who presents today regarding left congenital femoral deficiency, Paley type 3b. Natalia has been treated with prosthesis with an ischial bearing proximal section and a plantar flexion position non-articulated lower portion for the foot and ankle. She manages very well in this prosthesis. She is very active and otherwise developing normally. This is an isolated problem to the left lower limb.

On examination, Natalia has a well-formed left foot and ankle which are located at the level of the opposite normal right knee. The right lower limb is fully normally developed while the left lower limb is noticed extremely short. She has flexion deformity of the hip and palpable femoral segment which feels stiff relative to the knee joint and may not have any motion there. Her ankle on the other hand has excellent motion with 70° of plantar flexion and 30° of dorsiflexion for total arc of motion of 100°. She has excellent mobility of this left lower limb, both at the hip and at the ankle.

Radiographs confirm a short proximal femoral segment which does include metaphysis, physis, and epiphysis in a flexed position at the knee. The ankle is confirmed to be proximally at the level of the opposite knee. She has a little bit of hypoplasia of the fibula accounting for some increased valgus instability that was noted clinically.

**Problem List:**

1. Congenital femoral deficiency, Paley type 3b.
2. Severe congenital leg length discrepancy, left shorter than right.

**Recommendations and Plan:**

I have been communicating with this family prior to the visit today via email. I have offered the opinion that the best treatment in this case is the Paley-Brown rotationplasty procedure. Today, I am evaluating her in person including looking at her x-rays and I concur with my original recommendation. There is no femoral head and therefore the Paley-Brown procedure is the ideal

procedure in this case. The stiff knee joint can be treated by posterior capsulotomy at the time of the rotationplasty to regain the knee motion. The ankle and foot are ideal for the rotationplasty since it is a five-ray foot with hypermobility in plantar flexion and dorsiflexion of the ankle joint. It seems normally rotated and therefore may not even need a rotational osteotomy.

Nevertheless, after performing the fusion of the femoral segment to the pelvis we can decide whether any derotation of the distal tibia is also required. If so, it would be done all in one surgery. I have explained to them in detail the process of the procedure including a brief hospital stay of usually three to four nights which can include one night in the pediatric ICU. I have also explained to them that the child would be under anesthesia for a lengthy period of time because this operation takes between six to eight hours for just the rotationplasty part and then another two hours for the skin closure. Skin closure is performed by our plastic surgeon, Dr. Mark Pinsky, who comes in and repairs the complex wound closure and then also follows them up postoperatively. Most common and almost the only complication we have had is some wound breakdown or seroma after the surgery. This is related to the 180° rotation that we are performing with complete cut of the skin circumferentially. I showed them another patient from Poland who recently underwent this procedure six weeks ago. They also understand that after surgery they would need to stay here for rehabilitation. Typically, we start with passive range of motion of the new hip joint and active and passive range of motion of the ankle which is the new knee joint. This would be followed by active and passive range of motion of the hip and knee six weeks later. Between 6 and 12 weeks, there is no weightbearing allowed but active and passive range of motion are allowed of the new hip and knee. This is because the muscles have now healed into their transfer sites. Between 12 weeks onwards, we allow full weightbearing and we measure them for a prosthetic so they can start walking. The goal after 12 weeks is gait training. We train the child how to walk with a rotationplasty prosthesis. At a minimum, I recommend they stay for the first 12 weeks and an additional six weeks after that to do rehabilitation. They are very fortunate in that they have a close family relative living nearby (Jupiter). This is very helpful so that they have family here and they have a place to stay.

I explained to them that only four days ago I operated on another Polish patient who is here for rotationplasty and the third one is coming in February. All three of these patients were sponsored by the Polish National Health Fund. (NFZ) Poland has long ago recognized that there is no one in Poland or even in Europe who has expertise in treatment these Paley type 3a or b deficiencies. Therefore, the best outcomes for the Polish citizens have been to approve out of country funding for them to have this treatment performed here. I have highly urged

them to petition the Slovakian National Health Fund to approve the cost of treatment for this surgery. CFD is a rare condition and therefore very few surgeons have much experience in treating it.

Furthermore, rotationplasty is an extremely complex procedure usually reserved for tumor patients. Unfortunately, the type of rotationplasty done for tumor patients: the Van Ness or the Winkelmann rotationplasty are not appropriate for patients with congenital femoral deficiency. For congenital femoral deficiency, the best type of rotationplasty is either the Paley-Brown rotationplasty or when there is a femoral head the Paley type. In this case, we are recommending a Paley-Brown rotationplasty since there is no femoral head. I have performed approximately 50 of these procedures successfully over the last 22 years. To my knowledge, there is no other single surgeon in all of Europe who has performed the Paley-Brown type rotationplasty. Any other rotationplasty for this runs a risk of rotating back out of the correct position and having hip instability and having a less successful functional result. The goal here is to get as normal as possible walking, running, jumping including sports activity. I showed the family videos of other children doing these type of activities including one of my patients videos after 20 years followup at age 24 years old. A cost estimate for this treatment will be provided for them to submit to their health fund. The ideal age for treatment is between age 3 and 4 years old. I will be happy to provide this treatment for them which will include not only the price of the operation and hospitalization but also cost of the plastic surgeon who performs the closure, the cost of hyperbaric oxygen treatment for a week after surgery, the cost of rehabilitation, and the cost of the prosthetic device. This is certainly the biggest and longest and most expensive operation I perform. In one operation it solves the problem this child has and avoids the need for repeated expensive procedures and interruption of childhood. The results in my hands have been excellent and I can honestly state the best in the world. I look forward to helping this girl achieve the best possible functional outcome.

Sincerely,

A handwritten signature in black ink, appearing to read 'Dror Paley', with a stylized, cursive script.

Dror Paley, MD, FRCSC

Director, Paley Institute