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New Patient Consultation

Date: May 9, 2017

Location: Skierniewice, Poland

Patient Name: Jadwiga Kocieba

Date of Birth: Jan. 6, 2016

Email Address: joanna.kocieba@gmail.com

Jadwiga is a 1-year-old girl born with left congenital femoral deficiency type IA and fibular hemimelia type II. Her only treatment to date is a solid AFO with shoe lift. She is otherwise healthy and and has 1 older sibling who is unaffected.

On examination: She has full range of motion of left hip knee and ankle. The left knee has some AP instability. The left hip can abduct 15° compared to 45° on the right hip. She has 90° of external rotation and 0° of internal rotation of the left hip compared to 45° internal and 90° external on the right hip. The thigh foot axis is +5° bilaterally. The left ankle has valgus instability and can evert almost 45°. The lateral and medial malleolus are at the same level on the left side.

Radiographic examination: A long AP pulldown radiograph from September 28, 2016 shows a 6 cm leg length discrepancy of which 1.3 cm from the tibia and 4.7 cm are from the femur. At birth the leg length discrepancy measured radiographically with 3.5 cm. The predicted discrepancy at skeletal maturity based on the most recent radiograph using the multiplier method is 20 cm. The predicted discrepancy immaturity for the tibia is 4.3 cm and for the femur 15.5 cm. The left femur has a significant coxa vara with a neck shaft angle of 100°. There is mild delayed ossification of the left femoral neck compared to the right. I am still classifying this as Paley type IA because I believe that it will fully ossify by the time of treatment. The left ankle shows hypoplastic fibula with the distal fibular physis 8 mm proximal to the ankle joint.

Recommendations and plan: Jadwiga has both fibular hemimelia and congenital femoral deficiency and his predicted to have a significant leg length discrepancy in both femur and tibia totaling 20 cm at maturity. I have outlined a life plan of surgical treatment that will allow them to achieve equalization of limb length together with correction of hip knee and ankle deformities and deficiencies.

Surgery 1: This is preparatory surgery to any lengthening by operating on the left hip knee and ankle. At the left hip she requires the SUPERhip procedure including the femoral and pelvic components of the osteotomy surgery. At the left knee she needs ligamentous reconstruction by this SUPERknee procedure. At the left ankle she needs to undergo relative shortening of the tibia to the hypoplastic fibula using the SHORDT procedure. All of these her operations that I developed over the last 30 years of my career treating thousands of patients with congenital malformations of the lower limb. These procedures when performed in my hands have greater than a 99% success rate in returning the hip knee and ankle to normal. All 3 of these procedures need to be done prior to any lengthening surgery.

Surgery 2: Lengthening left femur with external fixation. This surgery is done 1 year after the SUPERhip procedure. Left femur can be lengthened up to 8 cm safely using the appropriate external fixator articulated across the knee to the tibia. This requires very intensive daily physical therapy. This is not any usual physical therapy, but rather a specialized limb lengthening therapy program. This type of program was pioneered and developed at the Paley Institute and all of our patients undergo this program on a daily basis during the distraction phase. It will take 4 months of distraction to lengthen 8 cm in the left femur. As long as she is able to maintain good knee motion we can continue to lengthen to a maximum of 8 cm. After the distraction phase the family could go home to Poland for the 4 months of consolidation. The external fixator is removed after full bony consolidation which is usually 4 months after the end of distraction. With lesser amounts of lengthening both the distraction and consolidation phase as her shorter. They would need to return to the Paley Institute once radiographic consolidation is seen. This would be determined from the monthly radiographs that the family would send me electronically during the consolidation phase. These radiographs can be taken in Poland. At the time of removal special care is taken to debride all the pin sites which minimizes any risk of bone infection. We also inserted a specialized rod into the bone (SLIM rod) to prevent fracture after removal of the apparatus. Within a few months Jadwiga would regain all of her knee motion and return to normal activities.

Surgery 3: Lengthening left femur with implantable lengthening nail. This surgery is done between age 8 and 10 years. The second lengthening of the femur is performed using an implantable lengthening nail. By the age of 8 the diameter and length of the femur can accommodate an implantable lengthening nail. I would recommend performing a 4 cm lengthening and then allowing the bone to consolidate. This would take 8 weeks from surgery after which the family could return home. This is a much quicker and simpler lengthening than the previous one. Bone consolidation would occur with the implantable nail in

place and due to the child's small weight she could fully weight-bear from the time the lengthening ends and return to normal activities much faster. During the time in Florida daily physical therapy would be carried out. A special brace to protect the knee is used during the lengthening.

Surgery 4: Lengthening left femur with implantable lengthening nail. The second lengthening with the implantable nail would be done between age 12-14 years. Since the nail is already in place it would only involve a minor outpatient surgery to re-break the femur around the nail and reactivate the nail. The postoperative protocol however is the same with 4 cm of lengthening over the course of 8 weeks and a knee brace to protect the knee. Consolidation and return to activities are the same as in the previous lengthening.

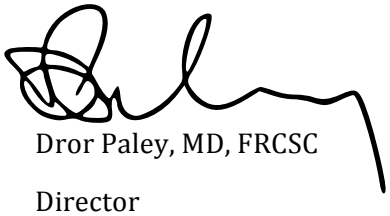
Surgery 5: Equalization of tibial length discrepancy. The tibia leg length discrepancy can be equalized either by epiphysiodesis or by lengthening. No decision on this needs to be made at this stage. Since Jadwiga appears to be tall for her age and since both of her parents are tall and loss of 5 cm of height by doing a contralateral proximal tibial epiphysiodesis which seem to be the simpler quicker option. The epiphysiodesis is not a difficult procedure technically. The critical part is the calculation of the correct timing. In most cases to achieve a 5 cm correction in the upper tibia epiphysiodesis would have to be performed between age 9 and 10. I mentioned this only as a guideline at this stage and not as a planned date for surgery. It is possible that it could be combined together with 1 of the other femoral surgeries. This is open for future planning and discussion. The alternative is to lengthen the tibia with an implantable lengthening nail. If that option is chosen, I would wait until skeletal maturity between age 14-15 to perform this final lengthening.

The above list of surgeries represents the surgery life plan that the parents should refer to in planning Jadwiga is limb length equalization. This like plan is based on successful experience that I had in treating many other children with similar conditions. In my hands, this would achieve a 99% chance of a successful outcome. I defined successful outcome as equalization of limb length by skeletal maturity, full range of motion of hip knee and ankle, stable joints at the hip knee and ankle, and normal gait with no limp. I realized that this is a very high bar and we have set for ourselves that it is based on actual results rather than the results we would like to achieve.

A cost estimate for the preparatory surgeries as well as the first lengthening will be prepared and sent to the parents. They are welcome to send me any

questions via email. We will be happy to help them plan for this treatment at the Paley Institute should they choose to pursue treatment with us.

Sincerely,



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Director