Paley Advanced Limb Lengthening Institute

Dror Paley, M.D., F.R.C.S.C.
dpaley@lengthening.us

Pediatric Orthopedic Surgery ▪ Limb Reconstruction Surgery ▪ Joint Preservation Surgery

November 8, 2013

Re: Drabik, Julia
DOB: 02/10/2008

To Whom It May Concern:

Julia is a 5-year 9-month old girl from Poland. She presents today for evaluation of her right fibular hemimelia and foot deformity. Julia was born with VACTERL syndrome. Her specific congenital anomalies include imperforate anus and vagina, tracheoesophageal fistula, one functional kidney, mild congenital scoliosis, and right fibular hemimelia and congenital femoral deficiency. She is here for evaluation and treatment of the latter.

Julia still has difficulties related to her tracheoesophageal fistula and perhaps some mild aspiration and wheezing. She is receiving treatment for this. Otherwise, she is healthy.

On examination she has an obvious leg length discrepancy which today measured 6.1 cm, shorter on the right than the left. Most of this is in the right tibia while a slight amount is in the femur. She has an obvious foot deformity with a hypoplastic foot approximately half the length of the other foot with only two toes. These two toes are a duplication of the big toe which are syndactylized together. She has no lateral toes. This big toe is supinate and flexed and slightly floppy. She has a good soft tissue mass of the foot laterally but it is devoid of any bone or cartilage. Despite this she has excellent range of motion of the hind foot and mid foot which combine to give her a 20 degree dorsiflexion and 50 degree plantar flexion arch of motion. Her heel remains in a mutual alignment position and the ankle seems to be stable. Her knee is excellent and goes from 0 to 140 degrees with only mild anteroposterior instability. Her hip has full range of motion and functions normally. Her right calf is thinner than her left while her thighs are of similar girth.

Radiographic examination demonstrates a 6.1 cm leg length discrepancy of which 1.3 cm is in the femur, 2.6 cm in the tibia, and 2.2 cm is from the foot. In total this adds up to 6.1 cm. The predicted leg length discrepancy based on her age, sex and these measurements is 9.5 cm at skeletal maturity. Her radiographs also demonstrate a subtalar coalition from a single calcaneous talus bone followed by an ossified navicular followed by three bones in the mid foot in the region that would normally have the cuneiforms and cuboid. These are then followed by a single wide metatarsal bone which has both proximal and distal physis to it. In articulation with that is some strange bone that I can only assume is a type of delta proximal phalanx of the big toe connected to two additional phalanges of each of these other toes.

Recommendations:

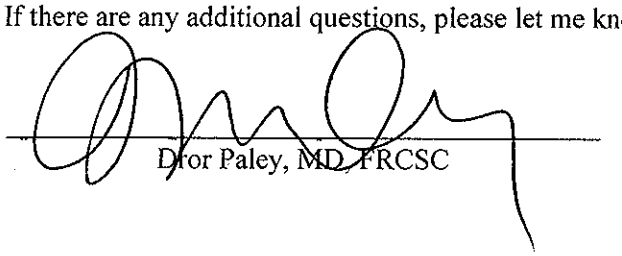
Julia would thus be treated with leg equalization procedures as well as foot reconstruction. The leg equalization strategy I would recommend is lengthening of the right tibia approximately 5 to 6 cm using the Taylor Spatial Frame computer dependent external fixator device. I will also extend this device down

to the foot in order to correct the foot deformity. Specifically, I performed a specialized oblique osteotomy of this first metatarsal avoiding injury to its growth plates. I would then lengthen it distally to gain some length in her foot but also to remove it laterally at the same time in order to widen the forefoot. Only a single metatarsal forefoot is very narrow. This would gain us probably 2 cm of length and 2 cm of width of the forefoot. This would be performed simultaneously with the lengthening of the tibia.

In the future, she would require another lengthening of 4 to 5 cm or an epiphysiodesis of the contralateral proximal tibia. The decision on which of these procedures to choose does not need to be taken now.

For Julia's treatment, she would need to plan to have the external fixator on between five to six months of which half that time would be lengthening and half of that time bone consolidation. She will have daily physical therapy in order to maintain range of motion and function during this time. At the end of this period the external fixator would be removed under general anesthesia and a cast applied. One plate of the cast could be removed and she could resume physical therapy. This is a very specialized type of treatment and the combination of lengthening of this atrophic fibular hemimelia leg with reconstruction of the complex foot deformity including lengthening and widening of the foot is something that is not available in Poland nor in Europe. For these reasons it would be in Julia's best interest to come to the Paley Institute in Florida to obtain this specialized treatment. A cost estimate for this treatment would be prepared for them so they can apply to the Polish National Health Fund. I have explained to them that our center is well known to the Polish National Health Fund and that we have treated numerous patients with fibular and tibial hemimelia over the past several years. Our success rate with this is over 99% which is another reason for her to obtain treatment here since it is much more guaranteed than if they try to do similar treatment in Poland or in Europe. As I believe this type of treatment is not available in Poland and since the only recommendation for treatment that they receive today is to amputate the leg, it seems appropriate to request such funding and to save her leg. There is no question that Julia's foot even with its current deformities is still a better foot than any prosthetic foot both due to it having sensation and mobility. Therefore, this should be taken in to consideration to give this child the benefit of doubt for obtaining this possible treatment which in her case is reconstruction and lengthening.

If there are any additional questions, please let me know.



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Name:	Drabik, Julia	DOS:	11/08/2013
DOB:	02/10/2008	Chart:	
Provider:	Dror Paley, MD, FRCSC	Ref. Phy:	

CONSULTATION

History of Present Illness:
Julia is a 5-year-9-month-old female with a history of VACTERL association. She has a history of tracheoesophageal fistula as well as anal fistula. She also has one functional kidney and does not have vagina as well as some mild congenital scoliosis. She does have issues with swallowing and gets pneumonias often and has had multiple surgeries on her trachea. She comes to us for evaluation for her right fibular hemimelia. They are from Poland and come in today to talk about all possible treatments. They have previously seen other doctors whose only recommendation has so far been for amputation. She has done previous surgeries on her foot which was to correct the position of her toes to make them more straightened, so they are deviating more medially. She walks with prosthesis. She has an AFO type prosthesis which is attached to a long approximately 5 cm lift. Otherwise, she has been healthy. Mother would like to know what reconstruction options they have here.

Past Medical History:
Significant for VACTERL association as above, tracheoesophageal fistula, anal fistula, no vagina, single functional kidney, frequent pneumonias, fibular hemimelia, and mild CFD.

Past Surgical History:
Previous surgeries on the patient's trachea as well as her anus as well as right toe surgery.

Developmental History:
The patient was born at 31 weeks via C-section for preeclampsia. Otherwise, she has been healthy. The patient sat at nine months and walked at about two and a half years.

Family History:
Negative for fibular hemimelia and any other congenital deformities.

Medications:
Albuterol or other similar type inhaler.

Review of Systems:
Negative except for HPI.

Physical Examination:
General: The patient is awake, alert, and healthy-appearing girl.
Extremities: She has obvious deformity to her right leg, which is shorter and the right calf is atrophic compared to the contralateral side. Thigh-foot angles are approximately +10 on both sides. Her hips have full range of motion with 110 degrees of flexion bilaterally and 45 degrees of abduction in extended position and 60 degrees of abduction in the flexed position. She has approximately 45 degrees of internal rotation and 10 degrees of external rotation on the right and about 60 degrees and 10 degrees on the left. Her knee

flexes and extends from 0 to 140 degrees. Ankle dorsiflexes about 5 to 10 degrees on the right and in combination with midfoot approximately 15 to 20 degrees and plantar flexes about 40 degrees on the left side and about 15 degrees of dorsiflexion and 45 degrees of plantar flexion on the left. She has no subtalar motion on the right and about 20 degrees of inversion and 10 degrees of eversion on the left.

Physical Examination:

Extremities: Examination of right foot reveals a **narrowed** foot with two toes, which are essentially duplicated and syndactyly of her greater toe which are sitting in neutral position, however, slightly supinated. Her foot also sits in a neutral position overall. Her knees are clinically straight. There is no significant genu valgum or varum. She has mild grade 2 AP instability, varus and valgus minimal instability compared to the contralateral site. She does seem to have active tibialis anterior and peroneal and Achilles tendons. I think I am also able to palpate a posterior tibial tendon as well.

Radiographic Findings:

X-rays: Standing EL reveals approximately 6.1 cm discrepancy. She is standing on a 5 cm block on the right and has approximately 1.1 cm discrepancy measuring from the top of her hips. Measuring her pelvis, there is approximate 3 cm discrepancy with the right smaller than the left. Femora measure approximately 13 mm discrepancy with the right shorter and the tibiae measure approximately 26 mm discrepancy with the right shorter and this gives an approximate 2.2 cm discrepancy between the foot. X-ray of AP and lateral and Saltzman view of her foot reveal she has essentially fused talus and calcaneus associated with subtalar joints. She has what appears to be a navicular and what appear to be three cuneiform type bones followed by a single metatarsal with two growth plates with a delta proximal phalanx and which seems to be attached to the middle and distal phalanx of her syndactyly. Her fibula is hypoplastic. It extends about two thirds of the way up to the length of the tibia. It is also a little bit high on the ankle and she has what appears to be the beginning of a ball and socket type ankle joints. Otherwise, mechanical axis is essentially neutral, may be 3 mm mechanical axis deviation on the right but essentially neutral on the left.

Assessment:

Right fibular hemimelia, VACTERL association, and complex foot deformity.

Plan:

Overall, Julia has a pretty functional foot. Her knee and ankles are actually quite stable and quite functional with good muscle motion as well as good sensation distally. We calculated her total discrepancy to be projected 9.5 cm at skeletal maturity. With this in mind, our recommendation will be to do a total of two lengthenings during her lifetime. We would also try to correct her complex foot deformity during the first lengthening with the frame. We will plan on doing a right tibia osteoplasty and lengthening for about 5 to 6 cm. At the same time, we will also do an oblique osteotomy of the metatarsal in order to lengthen as well as widen her right foot. We will do this in about one to two years when she is a little bit older. We will then plan on doing a second lengthening versus epiphysiodesis of the contralateral limb. We can potentially do an internal lengthening device at that time. We would likely undertake this several years later when she is an adolescent. A cost estimate will be sent to Polish embassy and Dr. Paley dictated a letter to them today. We will need to see Julia back in about a year or so until we decide on a final surgical plan and get approval for surgery. We will plan on doing her surgery while is about 7 or 8 years old.

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