## Proteus Syndrome: Right Lower Extremity Deformity

#### **David Westberry, MD**



**Shriners Hospitals** for Children<sup>®</sup>— Greenville



- 16 year old male
  - Right LE deformity noted at birth
  - Progression of deformity through childhood
  - Resides in Middle East











# **Case History**

- 6 years of age
- Amputation recommended
  - Taken to operating room in Iraq, but dismissed after hours long wait without treatment
  - No other treatment attempted
- Increased deformity leading to functional and societal challenges



## **Clinical Presentation**

- External foot progression angle
- Right foot deformity
  - Enlarged right great toe and medial border
  - Severe pronation
- Anterior medial bowing of tibia
- Knee flexion contracture
- Knee ROM 20-40 degrees
- Patella enlargement
- Medial bowing of right femur
- External rotation contracture of right hip





# **Radiographic Findings**

- Right hip
  - Mild dysplasia
- Right femur
  - Medial bow
- Right tibia
  - Anterior medial bow
- Patella
  - Hyperplasia





### Treatment options?



# Recommendation for Treatment

- Right above knee amputation
- Patellectomy
- Femoral osteotomy with retrograde femoral nail







# **Postoperative Results**

- Patient happy with results
- Mild phantom pain
  - Medical management
- Returned home after several weeks in physical therapy with host family







