

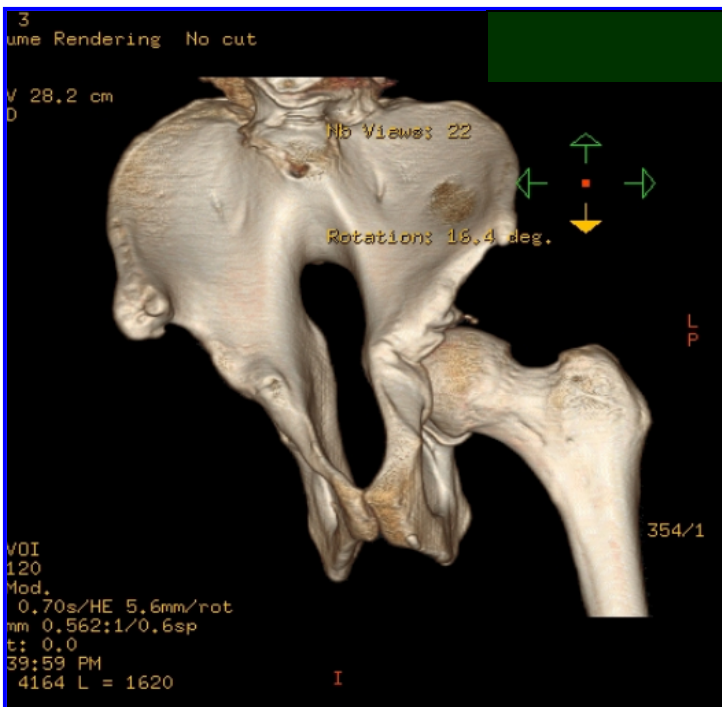
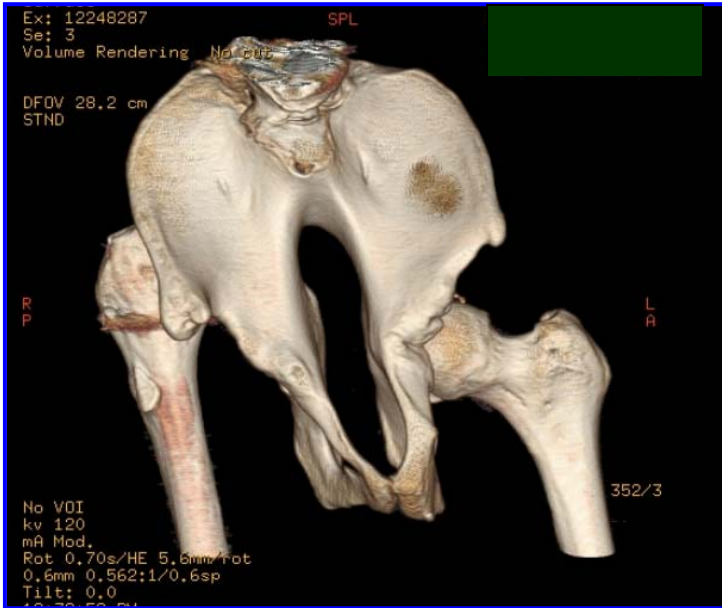
July 2010 Case Study: Sacral Agenesis s/p Complex Pelvic Reconstruction

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History of Present Illness: The patient is a 22-year-old female born with caudal Regression (sacral agenesis). She has had multiple surgeries including a colostomy, suprapubic urinary drainage, scoliotic deformity correction, femur fracture repair, leg lengthening and a dislocated right hip. Because of her caudal regression and multiple prior abdominal operations, the patient has an extremely narrow and abnormally formed pelvis. She complains of significant dyspareunia because of mechanical factors related to the extremely narrow pelvis. She presented to the orthopaedic office on referral to see if there was any way to increase the pelvic outlet. Her physical examination was notable for a right iliac wing, which was palpable, although the left one was difficult to appreciate. She had multiple abdominal scars. Her right greater trochanter was significantly higher than her left by nearly 2 inches. She has limited dorsiflexion and plantarflexion of both feet, and she had an ostomy and a urostomy in place. X-rays of her pelvis demonstrated a congenitally significantly narrow pelvis with a dislocated right hip. She had an intramedullary rod in place on the right, and she had spinal instrumentation in her spine. The pelvic outlet was approximately 0.5 to 1 cm in transverse diameter at its maximum. Because of her unique presentation, a CT scan and MRI scan was obtained pre-operatively. A 3-dimensional model of her pelvis constructed to better evaluate the unique anatomy. After consultation with my colleagues, we all felt that it would be possible to perform a number of osteotomies and internal fixation and possible soft tissue procedures to diminish the amount of dyspareunia, open up the pelvic outlet and reconstruct her pelvis to a more favorable configuration

Physical Examination: Height: 153 cm Weight: 121 lbs BP 124/64
HEENT: Normocephalic, atraumatic head with pupils round, equal. Neck: Full range of motion on flexion, extension, lateral bending, lateral rotation with no step off to palpation of the posterior elements and no pain. Heart: Regular rate and rhythm without detectable murmurs. Chest: Lungs are clear to auscultation throughout both lung fields. Abdomen: She has multiple scars and a urostomy. The urostomy is in the left lower quadrant. Pelvis: Her right iliac wing is higher than her left. Her lower extremities are cool to the touch and shiny. Distal pulses could not be appreciated to palpation. She appears to have normal quadriceps and hamstring function as well as for strength of the abductors and adductors of the hip bilaterally. She has limited dorsiflexion and plantar flexion of both feet. Her right foot demonstrates fixed varus of the hindfoot with the left demonstrating a calcaneovalgus alignment. Her right hip internally rotates 40° with only 10° of internal rotation on the left. Her right hip externally rotates 10° in the left hip externally rotates 45°. She has 45° of abduction on the right and 55° of abduction on the left.

Radiographs (CT scan with 3D Reconstruction):



CT examination of the pelvis showing severe dysplasia of the pelvis and right hip in the setting of sacral agenesis with superior, posterior and medial displacement of the right femur. There is severe narrowing of the pelvic outlet due to bony dysplasia

Procedure: Pelvic reconstruction with pelvic outlet expansion

Post-Op Radiograph:

