Addressing Congenital Hand Deformity

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For children with hand deformities, the functional, communication and psychological issues associated with the deformity are often the key drivers toward reconstructive surgery:

- **Functional.** For children born without an opposable thumb – one of the defining features of the human hand – grasping and manipulating objects with a single hand is nearly impossible.

- **Communication.** Over 90 percent of communication is accomplished through body language, voice inflection and tone, not words. As a result, if an individual with a hand deformity chooses to keep his or her hand in a pocket when interacting with others, communication is severely restricted.

- **Psychological.** Body image issues, coping with peer pressure, and dealing with parental guilt about their “difference” all contribute to the psychological issues associated with a hand deformity.

**Case Study:** A five-year-old girl with no family history of deformity presented with bilateral six-fingered hands with no thumbs. The decision was made to perform a pollicization of the radial second digit and amputate the radial-most ray in order to create a more normally functioning hand.

This operation was originally described in detail by Dr. Dieter Buck-Gramcko. It was modified to meet the requirements of this deformity. The proximal 80 percent of the second metacarpal was removed. A new trapezium was created by rotating the retained head of the metacarpal and placing it at the base of the hand in the usual position of the trapezium. The pollicized digit was rotated into 160 degrees of pronation. The thumb was now in a position of opposition. The proximal phalanx became the metacarpal of the new thumb, the middle phalanx became the proximal phalanx, and the distal phalanx remained the distal phalanx.

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What would you do if you had end-stage ankle arthritis? There are several excellent options, including an ankle-foot orthosis (AFO), ankle arthrodesis and total ankle arthroplasty. An AFO may diminish symptoms, but is unacceptable to many patients. A total ankle arthroplasty preserves motion and decreases stress on adjacent joints, but has a higher risk than an arthrodesis. Perioperative wound problems, a reported 20 percent reoperation rate and approximate 80 percent ten-year survival rate all contribute to this increased risk.

It is often quoted that ankle arthrodesis leads to subtalar and transverse tarsal arthritis because of the increased stress placed on these joints following an ankle fusion. There are no prospective or well-controlled studies, however, that confirm this conclusion. In all long-term retrospective studies, arthritic changes in adjacent joints were invariably present at the time of the ankle fusion. It is unlikely that a patient with a well-positioned ankle fusion will develop symptomatic arthritis in the subtalar or transverse tarsal joints. A patient preserves 25 percent of sagittal motion after an ankle fusion. In shoe wear, gait approaches normal and is equivalent to a patient with a total ankle arthroplasty.

At Cedars-Sinai, one of the clear indications for total ankle arthroplasty is concurrent arthritis or loss of motion in the ipsilateral subtalar joint. Once the subtalar joint is compromised, sagittal motion decreases along with the fluidity of gait. Preservation of ankle motion is essential in order to avoid the poor functional outcome of a pan-talar or tibial-to-calcaneal fusion. The patient in Figure 1 is 58 years old and had trauma to his lower extremity in a car accident when he was 32. He wears an AFO, but it no longer controls his symptoms. His ankle is painful and, although there is no subtalar arthritis, he has only 10 percent of inversion-eversion. An injection of lidocaine into his ankle joint relieved his symptoms for several hours, confirming the ankle as the source of pain. His postoperative X-ray (Figure 2) shows an excellent result with an Agility™ Total Ankle.
Pediatric Hip Fusion for Slipped Capital Femoral Epiphysis

Robert M. Bernstein, MD

Slipped capital femoral epiphysis (SCFE) is a well-known disorder of the hip in children that is characterized by displacement of the capital femoral epiphysis from the metaphysis through the physis. In the vast majority of cases, SCFE generally manifests between the ages of 10 and 16. Its etiology is unknown, but is frequently associated with childhood obesity.

A 14-year-old male presented in August 2002 with very limited motion and severe pain in his left hip. As a result of the pain, he was unable to place his foot on the ground and had been walking on crutches for months.

In 2001, the patient had been diagnosed with displaced SCFE by another physician. Based on the extent of his hip pain at that time, an open reduction and internal fixation with screws was performed. In February 2002, the patient’s hip pain returned. Femoral head collapse was detected and a second operation was performed to remove the screws previously inserted.

In August 2002, new X-rays showed further collapse of the femoral head. As a result, a left hip fusion, including a subtrochanteric osteotomy and the application of an external fixator, was performed in October 2002.

During a hip fusion, usually the hip joint is opened, any remaining cartilage is removed and screws are inserted across the hip joint to hold it together as it heals. Because the femur acts as a lever, which can result in nonunion, a subtrochanteric osteotomy is performed and the patient is immobilized in a body cast.

For this patient, a subtrochanteric osteotomy was performed and, in lieu of a body cast, an external fixator was used to hold the hip and leg in position. The osteotomy and fusion healed beautifully and the external fixator was removed four months later.

In 2008, at the age of 20, the patient is doing exceptionally well. Very active in sports, he is an excellent swimmer, plays basketball, runs and rollerblades. He wears a lift under his left foot to accommodate a 2 cm leg length discrepancy and has occasional slight back or knee pain, but otherwise leads a normal, active life.

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The extrinsic tendons of the pollicized finger were shortened and re-routed to provide proper flexion, extension and radial abduction. The intrinsic muscles were reconstructed in order to provide adduction, abduction and opposition. Skin flaps were transposed, advanced, trimmed, and reconstructed to create a new “first” web space. The radial-most ray was amputated.

After the surgery, the hand was immobilized for three weeks. At the end of three weeks, exercises were begun to restore range of motion, and subsequently strength. Functional training enabled the patient to use her hand well for all activities.

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Photographs taken after pollicization of the patient’s fifth digit and amputation of her sixth digit to create a normally functioning hand with an opposable thumb.
Questioning Assumptions Leads to Better Outcomes in Complex Cases

Earl W. Brien, MD

Experience and clinical expertise play an important role in the handling of complex orthopaedic cases. The following case studies illustrate two such situations in which prior experience helped me avoid assumptions and successfully treat patients with challenging orthopaedic conditions.

Case study #1
A female patient in her early 30s presented at a local hospital with what was believed to be a soft-tissue tumor of the foot. Upon biopsy, the tumor was found to be a soft-tissue sarcoma and the recommendation was made to amputate the foot. The patient then came to Cedars-Sinai for an evaluation, where the pathology slides and the imaging studies were reviewed. Although the slides were consistent with a soft-tissue sarcoma, I felt the clinical findings were not. In addition, my review of a current MRI did not support a diagnosis of sarcoma.

Rather than doing a definitive sarcoma procedure, I decided to re-biopsy the site, which confirmed my clinical suspicion: there was no tumor seen. Although it is not uncommon for a pathologist to see no tumor cells after a prior procedure, I felt additional information was needed before recommending observation only. I decided to evaluate the DNA of the original tumor specimen against the current specimen to test the possibility that the outside hospital had mixed up the slides. Not only was there no DNA match, but the DNA from the first specimen was from a male, not female.

The hospital that handled the original biopsy was immediately contacted and directed to track down the patient from whom the original specimen was taken. Such a case illustrates the importance of seeking specialized expertise in a field of rarities that may impact life and limb.

Case study #2
Advances in systemic chemotherapy and radiation therapy have led to a considerable improvement in the prognosis of patients with metastatic disease. As a result, orthopaedic surgeons are being asked with increasing frequency to evaluate and treat the manifestations of skeletal metastases.

A female patient in her mid-40s was diagnosed at a local medical center with breast cancer metastases to the femur. She was treated with plate fixation that failed in less than one year and was repeated. (Figure 1). This again failed shortly.

My prior experience of similar cases led me to recommend removal of the radiated bone that had failed to heal as the best approach for this difficult problem, and I performed a proximal femoral resection and hip replacement (Figure 2). While proximal femur replacement is a more aggressive approach than an internal fixation, patients tend to be happier. They avoid the multiple surgeries generally involved in an internal fixation, which can limit their lifestyle choices and activities.

Proximal femur replacement is a complex surgery that requires removing the femoral head and proximal femur, then reconstructing the hip abductor mechanism. Since proximal femur replacement puts the hip at risk for dislocation, a constrained total hip with multiple screws into the pelvis was also done. The patient was walking with the assistance of a physical therapist one day post-surgery and recovered beautifully. She was horseback riding within six to eight weeks of her surgery.

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Figure 1: X-ray of mid-40s woman who presented at Cedars-Sinai with a plate fixation that failed after less than one year.

Figure 2: X-ray of patient after proximal femoral resection and hip replacement.