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Grand Rounds from HSS

MANAGEMENT OF COMPLEX CASES

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FROM THE EDITOR

Hospital for Special Surgery was established in 1863 as the New York Society for the Relief of the Ruptured and Crippled, the first orthopaedic hospital in the United States. Celebrating 150 years of specialty care devoted to musculoskeletal medicine, from the outset, pediatric conditions have been perhaps the most important and challenging conditions facing orthopaedic specialists. This year the hospital opened the Lerner Children's Pavilion, a family-centric center that brings together pediatric orthopaedic and rheumatology specialists, nurses, anesthesiologists and rehabilitation specialists to provide special care for pediatric patients and their families.

In this volume of *Grand Rounds from Hospital for Special Surgery/Management of Complex Cases* we highlight three challenging cases demonstrating the state of the art in pediatric orthopaedics. The first case, Limb Lengthening for the Congenitally Short Femur, is presented by Roger Widmann, Chief of the Pediatric Service, demonstrating carefully staged procedures for limb lengthening in an effort to salvage the extremity and achieve a more functional gait. In the second case, John Blanco addresses the challenges in treating a chronic Monteggia injury. Finally, Dan Green describes a gratifying outcome achieved in the staged correction of genu valgum in a 9-year-old girl with hypophosphatemic rickets.

This publication is also available on **www.hss.edu/complexcases,** where you will find additional images and references as well as links to related articles. We hope you find these cases to be of interest and the principles presented informative. Comments are always welcome at **complexcases@hss.edu.**

— Edward C. Jones, MD, MA, Assistant Attending Orthopaedic Surgeon

in this issue

CASE 1

Limb Lengthening for the Congenital Short Femur

Case presented by Roger F. Widmann, MD and Benjamin T. Bjerke-Kroll, MD

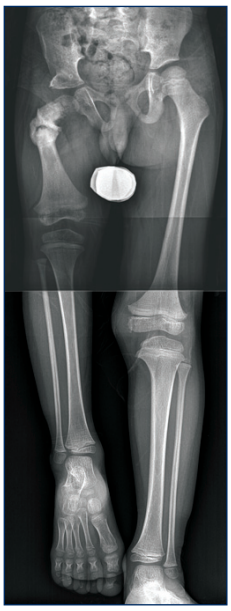

FIGURE 1

FIGURE 2

Figure 1: Radiograph of initial presentation of right Congenital Short Femur with 14cm of shortening. This shows a healed pelvic osteotomy, and intertrochanteric osteotomy with radiographic evidence of a nonunion and significant varus angulation.

Figure 2: Radiograph showing well healed corrective valgus osteotomy after plate removal.

FIGURE 3

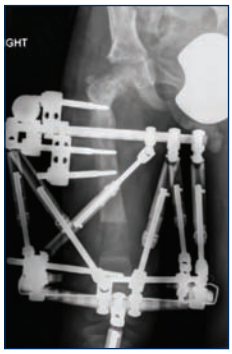

FIGURE 4

FIGURE 5

Figure 3: After a femoral diaphyseal osteotomy, a Taylor Spatial Frame (TSF) was applied to the femur and spanned across the knee joint.

Figure 4: After femoral diaphyseal osteotomy, TSF application and 5cm lengthening.

Figure 5: After removal of TSF and contralateral distal femoral epiphysiodesis with residual 8cm discrepancy.

CASE REPORT: A 4-year-old boy presented to Hospital for Special Surgery in 2008 with marked shortening of the right leg since birth. He was diagnosed *in utero* with Congenital Short Femur (CSF) (1), a rare non-inherited congenital anomaly affecting 1 in 50,000 births (2). The affected leg measured 14cm shorter, and the patient was able to walk only with a large lift under the right shoe. He had been treated previously with a Dega osteotomy of the pelvis and an intertrochanteric valgus-producing osteotomy. The pelvic osteotomy healed well, however the intertrochanteric osteotomy showed radiographic evidence of nonunion and was in significant varus angulation (Figure 1).

The patient's projected overall limb length discrepancy at skeletal maturity was 20-25cm. A staged procedure for limb lengthening was offered to the patient's family, in an effort to salvage the extremity and achieve a more functional gait. The hip was addressed first: an adductor tenotomy was performed, the nonunion resulting from prior valgus-producing osteotomy was taken down and a blade plate was placed after angular correction was achieved. This hardware was later removed, showing a radiographic union of the proximal femur and good correction of angular alignment (Figure 2). At age 6 years and 8 months, a femoral diaphyseal osteotomy was performed, and a Taylor Spatial Frame (TSF) applied to the femur and spanned across the knee joint (Figure 3). The femur was then gradually lengthened 5cm by distraction osteogenesis (Figure 4). After complete consolidation of the regenerate bone, the frame was removed. In order to slow growth of the contralateral leg, an epiphysiodesis of the left distal femur was performed at age 7 years and 7 months. The patient is now 9 years old, and has a total limb length discrepancy of 8 cm (Figure 5).

DISCUSSION: Significant limb length discrepancy in the growing child presents many challenges for the treating orthopedic surgeon. In a case where projected total discrepancy is greater than 20cm, amputation or Van Nes rotationplasty are generally considered (3). This creates a lifelong need for prostheses, and carries with it wound healing complications and stump breakdown. In addition, prosthesis use at any level will increase the energy expenditure needed for ambulation. Through the advances in the Ilizarov technique (4), what was once considered a "heroic" limb salvage procedure is a realistic alternative for the experienced surgeon. The TSF allows for distraction osteogenesis, angular correction in all three planes using computer-based software, and the ability to span adjacent joints to maintain range of motion and joint stability (5, 6).

In the case of significant length correction, soft tissue lengthening may determine the overall magnitude of correction. For this reason, the external fixation construct was spanned across the knee. The addition of tibial fixation allows for the knee to be locked in extension and to maintain range of motion. Congenital limb deficiency is frequently associated with absence or dysplasia of the cruciate ligaments and resultant knee instability. Posterior subluxation of the tibia and knee dislocation are known complications (7), and a knee-spanning construct facilitates stability about the joint. During the consolidation phase of lengthening, the patient sustained a proximal tibial Salter-Harris type I fracture and buckle fracture of the proximal fibula. This was noted to heal well in the frame with good alignment overall and no apparent growth disturbance at the affected physis. During any significant lengthening, fracture through the pin sites or regenerate bone is a known complication. The incidence increases when additional procedures or lengthenings are performed.

For this patient, an additional lengthening procedure is planned to further address the residual discrepancy. After a second lengthening, and likely revision epiphysiodesis of the contralateral distal femur, the final difference in limb length will be manageable with a small shoe lift if needed. This case demonstrates the importance of operative planning for the surgeon over multiple stages of lengthening. Pre-operative counseling and management of family expectations is

Continued on page 5

CASE 2

Chronic Monteggia Injury – Bado Type III

Case presented by John S. Blanco, MD and Christopher J. Dy, MD, MSPH

Figure 1A and 1B: Pre-operative AP and lateral of the elbow revealing lateral dislocation of the radial head with associated plastic deformation of the ulna.

Figure 2A and 2B: AP and lateral of the elbow and forearm following surgical correction and removal of intramedullary rod fixation. The radial head is located and radio-ulnar alignment is restored.

CASE REPORT: A 4-year-old boy presented with an injury to his dominant right upper extremity three months following a playground fall. He had been initially evaluated and treated elsewhere, at which time he was placed in an above elbow cast for several weeks for what was thought to be a non-displaced lateral condyle fracture. At the time of cast removal, his radiographs were concerning for a Monteggia injury. An attempt at closed reduction and casting was unsuccessfully performed and he was subsequently referred to the Pediatric Orthopaedic Service at Hospital for Special Surgery. On our exam he had an obvious deformity about his elbow with a prominent radial head laterally and restricted supination and pronation. His flexion/extension arc of motion was full and painless. He was neurovascularly intact. His radiographs revealed a lateral radial head dislocation along with a radially angulated proximal ulna fracture (Bado Type III Monteggia injury) (Figures 1A and 1B). Given his minimal symptoms, there was extensive discussion with the patient's family explaining the risks and benefits of attempted reconstruction.

At the time of surgery, an initial attempt at open reduction of the radial head was performed. The radial head had buttonholed through the anconeus musculature. Additionally, fibrofatty tissue prevented reduction. The annular ligament was intact. After addressing these soft tissue obstacles, the radial head was reducible but unstable. Through a separate incision, a proximal ulnar osteotomy at the site of maximal angulation was performed. The radial head was now stable. Intramedullary stabilization was performed and the radio-capitellar capsular tissues were repaired. A long arm cast was applied with the elbow hyperflexed and supinated for 6 weeks, at which time the intramedullary fixation was removed and physical therapy for range of motion initiated. The patient healed uneventfully (Figures 2A and 2B) and remains asymptomatic with full range of elbow motion and fully functional at 4 years post reconstruction.

DISCUSSION: Treatment of chronic Monteggia injuries in children is challenging. While Monteggia originally described a fracture of the proximal ulna in association with radial head dislocation in 1813, subsequent authors expanded the definition to include fractures in more distal portions of the ulna (1). Children with missed or neglected Monteggia injuries may function well initially, but concerns for long-term pain, instability, deformity, ulnar and radial nerve irritation, and secondary forearm/wrist pathology are often cited as the impetus for treatment. The delay in presentation makes treatment more difficult. While acute injuries can often be treated with closed reduction and immobilization, the delay allows formation of fibrous scar tissue in the proximal radioulnar joint space and healing of the ulna in a shortened and angulated position. Both of these factors impede attempts at closed treatment of chronic Monteggia injuries, likely necessitating open reduction and osteotomy of the ulna. Surgeons should counsel patients and families that the results of treatment are less predictable for chronic injuries compared to acute injuries, particularly if a radial head deformity has developed (1).

Following exposure of the radiocapitellar joint (through either a Kocher interval between the extensor carpi ulnaris and anconeus or through a muscle-splitting approach that spares the lateral ulnar collateral ligament), the surgeon must make an assessment of whether the radial head can be easily reduced. If the radial head cannot be easily reduced and maintained in stable position after open arthrolysis, a second incision and osteotomy of the ulna is recommended (1).

The treating surgeon may find it helpful to approach this injury as if it were an ulnar malunion. Many aspects of the ulnar osteotomy have been discussed, including location, orientation, and fixation (2). The variability in the literature suggests that there is no single correct way to accomplish the task of maintaining a stable radioulnar reduction. Horii et al have recommended that the ulnar osteotomy should be placed in the proximal metaphyseal portion of the

Continued on page 5

CASE 3

Staged Correction of Bilateral Genu Valgum Secondary to Hypophosphatemic Rickets

Case presented by Daniel W. Green, MD, Roger F. Widmann, MD, and Timothy Downey-Zayas

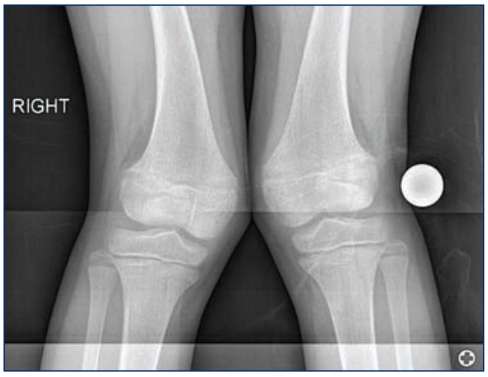

Figure 1: Preoperative AP radiographs of the knees showing genu valgum deformity

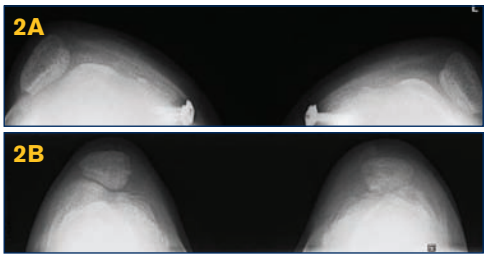

Figure 2A and 2B: Preoperative merchant views of the knees: showing patellar dislocation (2A) and showing postoperative correction (2B).

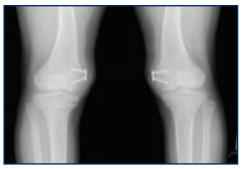

FIGURE 3

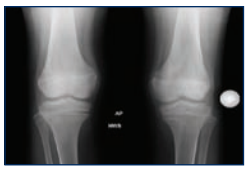

FIGURE 4

Figure 3: Postoperative AP radiographs of the knees showing correction of the genu valgum deformity after medial guided growth procedure using tension plate implantation.

Figure 4: Postoperative AP radiographs of the knees

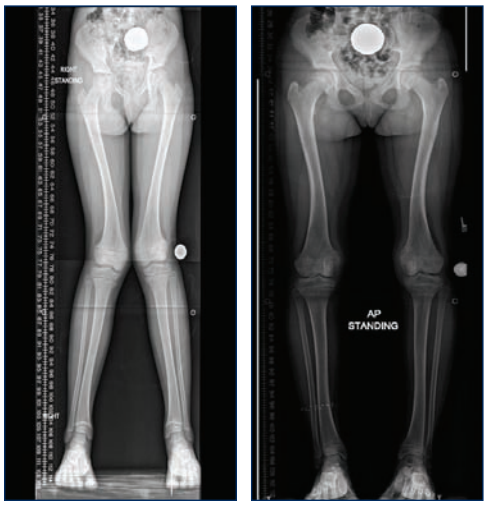

Figure 5: Standing Pre and Post operative radiographs showing valgus deformity and corrected alignment.

CASE REPORT: A 9-year-old girl with hypophosphatemic rickets presented to Hospital for Special Surgery with a chief complaint of patella instability and knock knee deformity. The patient was taking Calcitrol, K-Phosphate tabs and Advil for her hypophosphatemic rickets.

Physical examination revealed severe bilateral genu valgum approximately 20 degrees on each side. She ambulated and ran well in spite of this deformity. The patient had full active flexion and extension of both knees. Both patellae rested in the trochlear groove in extension but dislocated laterally upon flexion. The patella dislocation was non painful.

Initial imaging studies (Figure 1) demonstrated genu valgum deformity in both knees, with left knee femoral-tibial valgus of 12.4 degrees and 14.8 degrees in the right knee. The mechanical lateral distal femoral angles were measured as 72.7 degrees and 70.2 degrees in the left and right knee respectively, while the medial proximal tibial angles were measured as 93 degrees on the left and 92 degrees on the right. These angles are critical in making an informed decision for medical management of the patient's genu valgum (1). Widening of her lower extremity growth plates consistent with hypophosphatemic rickets can also be seen (Figure 1). The Merchant view radiographs demonstrated bilateral patellar dislocation (Figure 2A).

After thorough discussion with the patient and her family, the patient underwent a series of staged procedures. Initially, a bilateral distal femur medial guided growth procedure using tension plates was performed to address the genu valgum. The patient did well and had complete correction of the genu valgum deformity (Figure 3). Five months later, a Medial Patellar Femoral Ligament (MPFL) reconstruction with hamstring autograft, quadriceps lengthening, and lateral release was performed on the right knee. The eight plate that had previously been inserted was removed. Typically quadriceps lengthening and lateral release is not required in most cases of MPFL reconstruction, but in this case of obligatory patella dislocation the patella would not stay reduced in the trochlea without a lateral release and a quadriceps lengthening. Four months later, as with the right knee, a Medial Patellar Femoral Ligament (MPFL) reconstruction with hamstring autograft, quadriceps lengthening, and lateral release was performed on the left knee. The patient again tolerated the surgery well and upon follow-up six weeks later was doing well, with excellent quad strength and patella alignment (Figure 2B). Clinically, deformity correction was achieved and the patient had good strength in both knees. Postoperative imaging confirmed good anatomic alignment (Figure 4, Figure 5).

DISCUSSION: The management of angular deformities of the lower extremity in children with hypophosphatemic rickets is challenging. Severe angular deformities of lower extremities can cause an awkward gait and difficulty with running or sport activities. Early medical intervention is essential to minimize progression of deformities. After medical treatment is optimized, a standard treatment option is osteotomy (2). While corrective osteotomy is considered the gold standard for severe angular deformity, it has the downside of being a major surgical intervention that can lead to postoperative pain and prolonged healing time. While there is paucity of data in hemiepiphysiodesis in the hypophosphatemic rickets population, it is minimally invasive and shows good correction in restoring alignment, as well as preventing the need for more extensive surgery. (2, 3, 4) Additionally, in some cases the use of non locking plates have shown to correct deformities faster with fewer complications than osteotomy. (1) In this patient, having hypophosphatemic rickets and progressive genu valgum with bilateral patellar dislocation, we chose to perform a staged procedure using implant mediated guided growth followed later by bilateral patella reconstruction using MPFL hamstring autograft. Genu Valgum can significantly affect knee function, leading to patellar maltracking, overload of the lateral compartment of the knee and medial collateral ligament stress (2).

Continued on page 5

CASE 1 CONTINUED

paramount, as the lengthening procedure demands both effective counseling from the medical team as well as compliance from the patient and family. With modern operative techniques and surgical expertise, limb salvage and lengthening may be an option for patients with significant congenital limb abnormalities. ■

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CASE 2 CONTINUED

ulna to avoid disruption of the proximal attachment of the interosseous membrane on the ulnar shaft (2). The orientation of the opening ulnar osteotomy should be based on the deformity of the radial head (3). If the radial head is dislocated anteriorly, the osteotomy should be open posteriorly. If the radial head is dislocated posteriorly, the osteotomy should be open anteriorly. An oblique osteotomy is recommended to promote healing of the osteotomy, but a transverse cut is likely sufficient given the rarity of osteotomy nonunion.

The position of the osteotomy fragments should be dictated by the reduction of the proximal radioulnar joint. Intramedullary fixation provides the advantage of less prominent hardware and easier removal of hardware, while the plate/screw construct allows more rigid fixation and earlier motion, but necessitates removal of hardware in the formal surgical setting.

Annular ligament reconstruction, originally described with triceps tendon, is considered to supplement stability of the proximal radioulnar joint reduction (4). Initial enthusiasm for its added stability has waned due to concern for osteolytic change along the underlying radial neck, narrowing of the radial neck, limitations in forearm motion, and graft attenuation over time (2, 3). External fixation or radiocapitellar pinning can also be used to supplement stability of the reduction, but concerns for wire breakage, migration, and infection exist. Following wound closure, the forearm is stabilized in a position of maximum stability with a long-arm cast. Forearm and elbow motion is eventually allowed at the surgeon's discretion depending on the stability of the reduction and type of ulnar fixation used, with the earliest motion permitted at 2 weeks.

Outcomes are largely predicated on intra-operative stability of the reduction and secure fixation of the ulna. Horii et al. (2) reported their results before and after changing their surgical technique. There were 7 post-operative dislocations among 13 patients treated with transverse ulnar shaft osteotomies secured with intramedullary fixation. Following a change to oblique metaphyseal osteotomies fixed with plates and screws, there were no dislocations in 9 patients (2). Rodgers et al. (5) reported nerve palsies and forearm compartment syndromes in their series of 14 patients, suggesting that the treating surgeon should consider neurolysis and compartment releases in the appropriate clinical scenario (5). Clinical outcomes are generally good to excellent, but limitations in elbow and/or forearm motion may be noted, particularly with longer delays from acute injury to treatment (6, 7). ■

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CASE 3 CONTINUED

Hemiepiphysiodesis is a minimally invasive, outpatient procedure that has been shown to provide gradual deformity correction, while also allowing additional growth after implant removal. (5) ■

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