

Introduction

It is a great privilege for me to provide the Bunnell Traveling Fellowship Report for 2002 travel period. My fellowship concentrated on the diagnosis and treatment of diseases that affect children, including congenital anomalies, brachial plexus birth palsies, and tetraplegia. I visited many Centers of Excellence throughout the world and the knowledge acquired has enhanced my practice. This report is designed to highlight the fellowship and to provide educational benefit to the reader. Each visit is divided into separate sections and a “tricks of the trade learned” section included for additional information.

Paris, France

Paris was the first stop of the fellowship and Alain Gilbert and Carolyn LeClerq served as hosts. Dr. Gilbert is a remarkable man, with an intense energy directed toward the care of children with brachial plexus birth palsy (Figure 1). His clinic and operating room schedule is replete with brachial plexus birth palsies. Microsurgical inclusion criteria primarily include no palpable elbow flexion three months after birth. Palpation of biceps contraction or active elbow flexion is a relative contraindication for microsurgery; although delayed reconstruction may be required after spontaneous recovery has occurred.

This approach to microsurgical reconstruction provides a large surgical experience and Dr. Gilbert is a surgical phenomenon. His technique for an upper plexus lesion includes a quick exposure, resection of the neuroma, and sural nerve grafting. Fibrin glue is used for nerve graft coaptations, instead of epineurial sutures. Fibrin glue is commercially available and has been incorporated into my practice to augment nerve repairs. During the surgical procedure, no intra-operative nerve studies are utilized as avulsions or ruptures are determined by direct inspection. The presence of viable axons is confirmed via direct visualization of nerve fascicles after neuroma excision. Avulsions are treated by nerve grafts that bypass the irreparable lesions or adjacent nerve transfers (e.g., spinal accessory nerve transfer).

Time was also spent with Carolyn LeClerq, a caring physician with an intense interest in the treatment of persons with tetraplegia. We traveled to the outskirts of Paris and spent time at the Centre de rééducation neurologique et fonctionnelle de Coubert, a large 240 bed rehabilitation center for persons with spinal cord injury, stroke, and traumatic brain injury. Patients are evaluated using a multi-disciplinary team approach consisting of physiatrist, neurologist, hand surgeon, therapist, and nurse. The upper extremity treatment algorithm for tetraplegia has been defined according to the International Classification of Surgery of the Hand in Tetraplegia (Table 1).¹ Despite this schema, the exact reconstruction is dependent upon a careful examination of the patient and an assessment of their needs. Dr. LeClerq excels in this aspect of tetraplegia management and is able to individualize the reconstruction based upon the patient's needs, goals, and expectations. Irrespective of their hand reconstruction, 70% of persons with tetraplegia require restoration of elbow extension.² The ability to extend the elbow results in functional gains including an increase in workspace, the ability to perform pressure relief maneuvers, better manual wheelchair propulsion, and independent transfer.³ Posterior deltoid-to-triceps and biceps-to-triceps transfers are available methods to restore active elbow extension. Dr. LeClerq prefers the posterior deltoid transfer and performs the procedure with the patient positioned prone to allow complete access to the posterior shoulder and elbow. The posterior deltoid is carefully isolated and mobilized in a proximal direction to the level of the axillary nerve. A second incision is made over the triceps insertion. A

Dacron tendon enhancer is used as interpositional material to bridge the gap between the posterior deltoid and triceps (Figure 2). This material augments both coaptation sites and allows for a secure connection. Post-operative management is carefully modulated with limitation of shoulder motion until healing occurs across the tendon repair sites.⁴ A torso brace with limb attachment is fabricated prior to surgery and carefully applied in the operating room following the tendon transfer. This attention to intra-operative and post-operative detail directly influenced the results obtained by Dr. LeClerq. Patients often achieve anti-gravity restoration of elbow extension.

The Narakas Brachial Plexus meeting met in Paris and enhanced the overall visit to Paris. Brachial plexus surgeons from around the world convened to discuss adult and pediatric brachial plexus issues. This two-day event covered the gamut of brachial plexus problems beginning with diagnosis, venturing into early microsurgery, and conversing about late reconstruction. The wealth of knowledge at the meeting was inspiring and included stalwarts such as Hans Millessi, Panupan Songcharoen, Kazuteru Doi, Simo Vilkki, Allen Bishop, Pepi Borero, and my long time mentor, Mike Wood.

Texas Scottish Rite Hospital

Texas Scottish Rite Hospital was a memorable visit of the fellowship. Marybeth Ezaki and Peter Carter served as hosts (Figure 3). Marybeth and Peter are a dynamic duo and spend time together in the operating room and clinic. Patients are seen interchangeably and difficult problems are evaluated by both. The pediatric hand service is replete with energy and camaraderie. Hours upon hours were spent discussing the spectrum of congenital deficiencies.

Madelung's deformity is a particular area of interest and their surgical approach includes a combined release of Vicker's ligament and biplanar dome osteotomy in the metaphysis of the radius via a palmar incision.⁵ This tactic results in substantial correction in both the sagittal and coronal planes and rotates the distal fragment to obtain better coverage beneath the lunate. This surgical procedure has been incorporated into my practice to manage Madelung's deformity. Currently, mild deformity is treated by isolated release of Vickers ligament and considerable deformity necessitates concomitant dome osteotomy.⁶

Arthrogryposis is one of the most difficult congenital anomalies to treat. The amyoplasia congenita form is especially frustrating as the child's arms are positioned with the shoulder in internal rotation, elbow in extension, and wrist in flexion. The hand has a thumb-in-palm deformity and digital stiffness. Active and/or passive elbow flexion and wrist extension is deficient, which prohibits many activities of daily living. Early restoration of passive elbow flexion and active wrist extension can facilitate function. Many surgical procedures have been described for restitution of wrist extension, including serial casting, proximal row carpectomy, dorsal wedge osteotomy of the radius, and distraction histiogenesis with multiplanar external fixation. Marybeth and Peter have worked on a technique that consists of a biplanar wedge osteotomy of the midcarpal joint plus extensor carpi ulnaris tendon transfer (Figure 4). This wedge resection of the midcarpal joint realigns the wrist and provides active wrist extension in a single setting. Early surgery repositions the limb and offers the child the opportunity to accomplish daily tasks with a wrist positioned for use.⁷

The management of the shoulder in infants with residual brachial plexus palsy was another frequent topic. The Scottish Rite Hospital group equates acquired dysplasia of the shoulder after brachial plexus palsy to developmental dysplasia of the hip.⁸ The clinical signs of decreased motion, apparent shortening of the affected limb, and asymmetric skin folds are synonymous. Lack of shoulder external rotation is analogous to lack of hip

abduction and implies underlying glenohumeral deformity. Their preferred diagnostic modality is ultrasound, which provides a dynamic view of the shoulder.⁹ The shoulder can be visualized during passive external and internal rotation to determine the relationship between the glenoid and humeral head. This modality requires some expertise and provides an indication of the “reducibility” of the glenohumeral joint. We still prefer magnetic resonance imaging using a cartilage sensitive axial images with a minimal interslice gap to visualize the specific glenoid morphology.¹⁰

Minneapolis

A trip to Minneapolis Shriners Hospital for Children and Gillette Children’s Hospital was hosted by Ann Van Heest and Bill Cooney. The fellows from Mayo Clinic, including Tim Leddy, Mark Rizzo and Steve Moran, accompanied Dr. Cooney for a day filled with surgery at the Minneapolis Shriners Hospital (Figure 5). Unfortunately, a water pipe broke in the operating room and all procedures were cancelled. This event necessitated a quick change in plans and patients were assembled for examination and discussion. The time evolved into an educational symposium with open deliberation, chalk talk, and didactic components.

Ann Van Heest and I also spent additional time at Gillette Children’s Hospital. The highlight was time spent in the motion lab with a dedicated team of individuals determined to analyze upper and lower extremity motion in impaired children. Part of the lab is dedicated to the evaluation of limb motion in children with residual brachial plexus palsy. The Vicon light system (Oxford Metrics, Lake Forest, CA) is used to digitize their movement, which allows a three-dimensional representation of motion to objectively evaluate changes in limb and trunk patterning before and after the surgery.

Chicago

Michael Bednar and Terry Light organized a visit to Chicago (Figure 6). Time was spent at Loyola University and Shriners Hospital for Children. Children remained the focus of the fellowship throughout the Chicago visit and Dr. Light’s wealth of experience was readily apparent. The treatment of radial deficiency about the thumb and forearm were discussed in practical terms and in the clinical setting. The technical details of pollicization were highlighted during surgery on a child with a pouce flottant (Figure 7). Dr. Light’s experience in pollicization, coupled with his previous time spent with Buck Gramko, was readily apparent. Time was spent discussing flap design, carpometacarpal joint alignment, and intrinsic muscle reconstruction. Placement of the index metacarpophalangeal joint into extension via a suture prior to repositioning was a particularly useful technical pearl. This step will prevent unwanted thumb hyperextension at the newly reconstructed carpometacarpal joint.

The trials and tribulations of treating recurrent radial deficiency were also discussed.¹¹ Children with recurrent deformity develop secondary changes within the soft tissues and bone that prevent immediate correction. Gradual correction and soft tissue stretching of the tight radial structures can be accomplished using a multiplanar external fixation device.¹² Drs. Light and Lubicky applied this concept to a nine-year old patient with recurrent deformity. An Ilizarov device was applied to the limb with offset hinges to allow simultaneous correction of both angulation and length. After gradual distraction histiogenesis, repeat centralization or formal arthrodesis can be performed. Although these type devices are being employed more frequently, the outcome after such treatment remains in question.

Mike Bednar and I spent additional time discussing brachial plexus birth palsies, a common theme throughout the fellowship. The difficulties in maintaining a concentrically reduced shoulder, along with the intricacies of tendon transfers about the shoulder were discussed. Glenohumeral dysplasia needs to be recognized early to prevent considerable deformity and permanent sequelae.¹³

Boston

The fellowship included a trip to Boston to see my friend and colleague, Peter Waters (Figure 8). Boston Children's Hospital is wealth of orthopaedic surgery and Peter is the leader of the upper extremity section. He has been instrumental in developing a global brachial plexus data network that has received grant support from both the Pediatric Orthopaedic Society of North America and the American Society for Surgery of the Hand. The goal of this center-randomized project is to answer many of the controversies that surround brachial plexus birth palsies. The accumulation of a large enough patient cohort to answer these difficult questions requires this multi-center approach. Continued support of this study is necessary to determine the precise indications for microsurgery, timing of surgical intervention, anticipated outcome of various techniques, and effect of tendon transfers. This information will be invaluable both to physicians that treat brachial plexus birth palsies and to families with affected children.

Korea

Part of the Bunnell Traveling Fellowship included a venture to the 4th International Congress of Asian Pacific Federation of Societies for Surgery of the Hand. Tom Wright, Longen Chen, Dean Sotereanos, John Taras, and Jai Ryu also traveled to the meeting (Figure 9). Dr. Tarek, Congress Chairman, and Dr. Kim, Chairman of the Scientific Committee put forth an exceptional meeting. The meeting encompassed the gamut of hand surgery, with the theme "Hand Surgery Update in the New Millennium". While in Korea, time was spent at Gunpo Medical Center, under the auspices of Sang Su Kim. Dr. Kim and his colleague, Dr. Choe, are experts in performing contralateral C-7 nerve transfer. Over 55 cases have been performed for avulsion injuries of the brachial plexus. The procedure is done in two stages to allow interim time for nerve regeneration. Dr. Kim is a facile surgeon with extraordinary technical ability and these attributes were witnessed during a contralateral C-7 nerve transfer (Figure 10).

Time was also spent at Seoul National University with Drs. Baek and Chung. Dr. Baek represents a rising star in the field of congenital hand surgery. Cases were shown of congenital or acquired radial head dislocation treated by distraction histiogenesis of the ulna with or without annular ligament reconstruction. This concept of a primary bony correction for radial head reduction is a tactic that can be applied to the forearm length inequalities encountered in multiple hereditary exostosis.¹⁴

China

Mainland China was visited after Korea. The cities of Shanghai, Beijing, and Wuxi were part of the itinerary. Numerous hand surgeons were seen throughout China. There are 15 hand surgeons at Shanghai University. Dr. Gu is chairman of the department and a pioneer in brachial plexus surgery having described the use of both contralateral and ipsilateral (i.e. from the same side) C-7 nerve transfer (Figure 11). Dr. Xu is currently in charge on the hand section and he insured our stay was full of camaraderie, discussion, education, culture, and surgery.

During a day in the operating room, an ipsilateral C-7 nerve transfer was performed on a 20 year-old patient nine months after injury (Figure 12). Drs. Chen and Hu visualized avulsions of C-5 and C-6 at surgery. A normal C-7 was cut and transferred directly to the upper trunk. A simultaneous spinal accessory to suprascapular nerve transfer was performed. Later that evening, examination of the patient revealed no weakness in triceps extension or finger extension. The group has performed approximately 20 ipsilateral C-7 nerve transfers without any considerable deficit.

A symposium on hand and microsurgery was also held during our stay in Shanghai. Surgeons from around China congregated to present material and discuss the intricacies of hand surgery. Dr. Xu presented a technique of endoscopic harvesting of the phrenic nerve to allow primary coaptation during brachial plexus reconstruction. Dr. Bingfang from the Sixth Peoples' Hospital discussed free tissue transfers for upper and lower limb reconstruction. Multiple free transfers to restore function to the injured hand are commonplace in China.

Wuxi is the home of the only hospital in China dedicated solely to microsurgery. The 200-bed hospital is staffed by 10 hand surgeons and 20 residents. Dr. Rei is in charge of the hand surgeons and acted as our host (Figure 13). A tremendous wealth of microsurgical experience was apparent. Free tissue transfers are a daily occurrence to reconstruct the injured hand. Their technical ability is admirable and the entire team is extremely skilled at microsurgical procedures.

Hosts everywhere throughout my fellowship were unbelievable, especially in China. Hospitality is genuine and was greatly appreciated by all of us. China is a country of 3 billion people that has tremendous resources that made this visit most memorable. Extraordinary sociable experiences included climbing the Great Wall of China at Juyongguan, walking through Tianamen Square, visiting the Shanghai Museum, and watching the Shanghai acrobatic show (Figure 14).

Camp Sunshine

A trip to Camp Sunshine, a retreat for critically ill children and their families located in Portland, Maine, was another highlight of the fellowship. Invited by the Fanconi Anemia Support Group, time was spent with children and their families afflicted with Fanconi's anemia, a rare, inheritable disease that leads to aplastic anemia. Fanconi children are born with hypoplastic thumbs, but that remains the least of their problems. This dreadful disease is often fatal without bone marrow transplant.

Fanconi children are typically born with hypoplastic thumbs, but do not have signs of bone marrow failure at birth, which prevents immediate hematologic diagnosis. The median age of aplastic anemia is approximately seven years and the majority of children experience signs between the ages of three and twelve. A chromosomal challenge test, however, is currently available that allows detection of the disease prior to the onset of bone marrow failure^{15,16}. This assay subjects a sample of the child's lymphocytes to diepoxybutane or mitomycin C, which causes chromosomes within Fanconi anemia cells to break and rearrange. In contrast, lymphocytes in unaffected children are stable to these agents. Since bone marrow transplant is the only cure for Fanconi's anemia, this prefatory diagnosis is crucial for the child and affected family. Early diagnosis provides ample time to search for a suitable bone marrow donor or to consider pre-implantation genetic diagnosis (PGD)^{17,18}. PGD is a sophisticated technique that involves in vitro fertilization, sampling of the blastocyte to ensure HLA similarity without Fanconi's disease, and reimplantation until birth (Figure 15). At delivery, cord blood is harvested from the newborn and used as a source of stem cell transplant to the affected sibling. Since the

PGD process takes time, early detection via a chromosomal challenge test is critical and may ultimately save the affected child.

The Camp Sunshine meeting consisted of a collection of scientists that have been instrumental in the development of techniques to allow early diagnosis and innovative treatment regimens, such as PGD. These assays and methods are changing the way we manage certain inheritable diseases. These techniques are not about designer babies, but rather about saving lives. Stem cells harvested from sibling core blood can be used as donor cells to rescue lives and help distressed families. All physicians that manage children with hypoplastic thumbs should consider the diagnosis of Fanconi's anemia. The diagnosis can be readily made by a chromosomal challenge test that would allow ample time for the family to consider PGD or to find a bone marrow match. Many of the children at the camp had undergone thumb surgery without testing for Fanconi's anemia and the parents remain bitter about the lack of early diagnosis.

Acknowledgments

I would like thank all the people that have helped me during my fellowship and career. My hosts were gracious with both their time and knowledge. Their effort and knowledge will be forever remembered (Table 2). A special thanks to Suzanne Sharif who was instrumental during my stay in Paris, Alain Gilbert for his hospitality, Mike Wood for his mentorship, Mike Bednar for his camaraderie, Ann Van Heest for her insight, Marybeth Ezaki and Peter Carter for their companionship, Bo Frederick for his kinship, Allen Bishop and Dick Berger for their friendship, my comrades in travel during the fellowship, and the hand surgeons at Shanghai University.

Lastly, none of this could have been done without the support of my family and children. My parents and children have born the brunt of the required travel and extensive work schedule necessary to accomplish and complete the Bunnell Traveling Fellowship. A special thank-you to my children, Bryan and Samantha, who represent the most important thing in life to me. Their continued sacrifices when "Daddy's away" do not go unnoticed.

Table 1: International Classification of Surgery of the Hand in Tetraplegia (ICSHT) Classification

Sensibility	Group	Muscle	Function
O or Cu	0	No muscles below elbow suitable for transfer	
	1	Brachioradialis	Flexion of elbow
	2	Extensor carpi radialis longus	Weak wrist extension with radial deviation
	3	Extensor carpi radialis brevis	Wrist extension
	4	Pronator teres	Forearm pronation
	5	Flexor carpi radialis	Wrist flexion
	6	Extensor digitorum communis	Finger metacarpophalangeal joint extension (extrinsic extension of the fingers)
	7	Extensor pollicis longus	Thumb interphalangeal joint extension (extrinsic extension of the thumb)
	8	Digital flexors	Extrinsic finger flexion
	9	All muscles except intrinsic	
	X	Exceptions	

Table 2:Tricks of the Trade Learned

Brachial plexus palsies- microsugery

- Microsurgical reconstruction can be accomplished via expeditious exposure, resection of the neuroma, and sural nerve grafting. Fibrin glue can be used for nerve graft coaptations to augment epineurial sutures.
- A global brachial plexus data network is required to solve the controversies surrounding precise indications for microsurgery, timing of surgical intervention, anticipated outcome of various techniques, and effect of tendon transfers.
- Contralateral C-7 nerve transfers can be safely performed for avulsions about the brachial plexus. The outcome after transfer remains a substantial question.
- Direct ipsilateral C-7 nerve transfer can be performed for avulsions of C-5 and C-6 with minimal effect on limb function. The exact indications are still evolving.

Brachial plexus birth palsies- shoulder dysplasia

- Developmental dysplasia of the hip and acquired dysplasia of the shoulder after brachial plexus palsy present with similar physical findings. The clinical signs of decreased motion, apparent shortening of the affected limb, and asymmetric skin folds are synonymous. Lack of shoulder external rotation is analogous to lack of hip abduction and implies underlying glenohumeral deformity.
- Ultrasound can be used to visualize the dysplastic glenohumeral joint and provides a dynamic view of the shoulder during external and internal rotation.
- Motion analysis techniques for the lower extremity motion can be applied to the upper extremity. Upper extremity motion after residual brachial plexus palsy requires three-dimensional representation to objectively evaluate changes in limb and trunk patterning before and after the surgery.

Tetraplegia

- Deltoid-to-triceps transfer can be performed in the prone position. A Dacron tendon enhancer can be used to bridge the gap between the posterior deltoid and triceps and augment both coaptation sites. Post-operative management must be carefully modulated with limitation of shoulder motion until healing occurs across the tendon repair sites.

Congenital

- Madelung's deformity can be corrected by release of Vicker's ligament and biplanar dome osteotomy in the metaphysis of the radius via a palmar incision.
- Arthrogryposis wrist flexion can be repositioned by biplanar wedge osteotomy of the midcarpal joint plus extensor carpi ulnaris tendon transfer in a single setting. Early surgery places the limb in a functional position for activities of daily living.
- Pollicization remains the premier procedure for the severely hypoplastic or absent thumb. Placing glabrous skin along the proximal phalanx of the index improves the contour and outcome. Placement of index metacarpophalangeal joint into extension via a

suture prior prior to repositioning prevents unwanted hyperextension at the newly reconstructed carpometacarpal joint.

- The Ilizarov device can be used to correct the neglected or recurrent radial deficient limb. The definitive procedure following angular correction remains controversial. Repeat centralization or formal arthrodesis are both options, although the outcome after such treatment remains in question.
- Children with Fanconi anemia are typically born with hypoplastic thumbs, but do not have signs of bone marrow failure at birth. A chromosomal challenge test is currently available that allows detection of the disease prior to the onset of bone marrow failure. Bone marrow transplant is the only cure for Fanconi's anemia and early diagnosis is crucial for the child and affected family to provide ample time to search for a suitable bone marrow donor or consider pre-implantation genetic diagnosis.
- Reduction of an acquired radial head dislocation requires correction of any deformity in the ulna. Distraction histiogenesis is a valuable technique that can be applied to forearm length inequalities (e.g., multiple hereditary exostosis).

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**Figure
Legends**



Figure 1: Alain Gilbert and myself at Clinique Jouvenet in Paris.

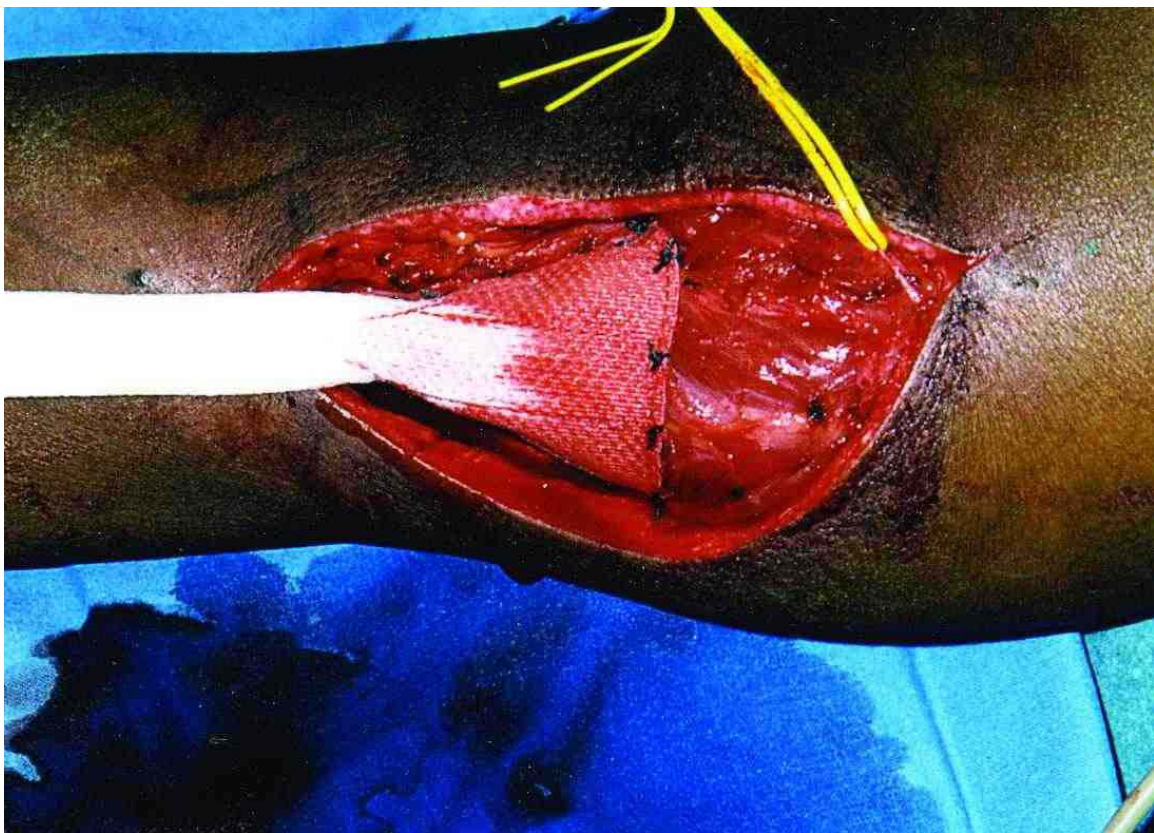


Figure 2: Dacron tendon enhancer used to bridge the gap between the posterior deltoid muscle and triceps tendon.



Figure 3: Marybeth Ezaki and Bob Bucholz were gracious hosts during my stay at their home.

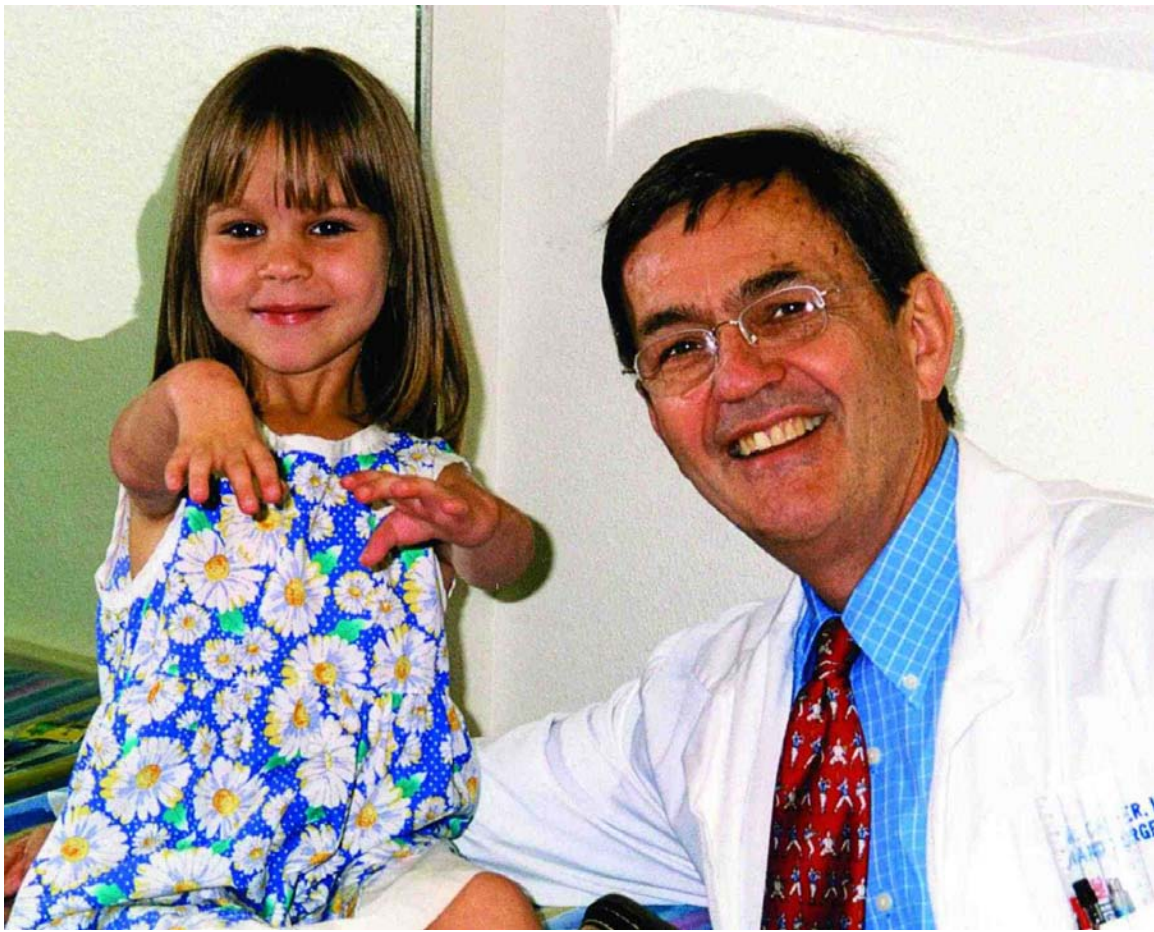


Figure 4: Peter Carter and a child with arthrogyriposis after right wrist biplanar osteotomy and tendon transfer.



Figure 5: Ann Van Heest, Bill Cooney and the fellows from Mayo Clinic at the Minneapolis Shriners Hospital.

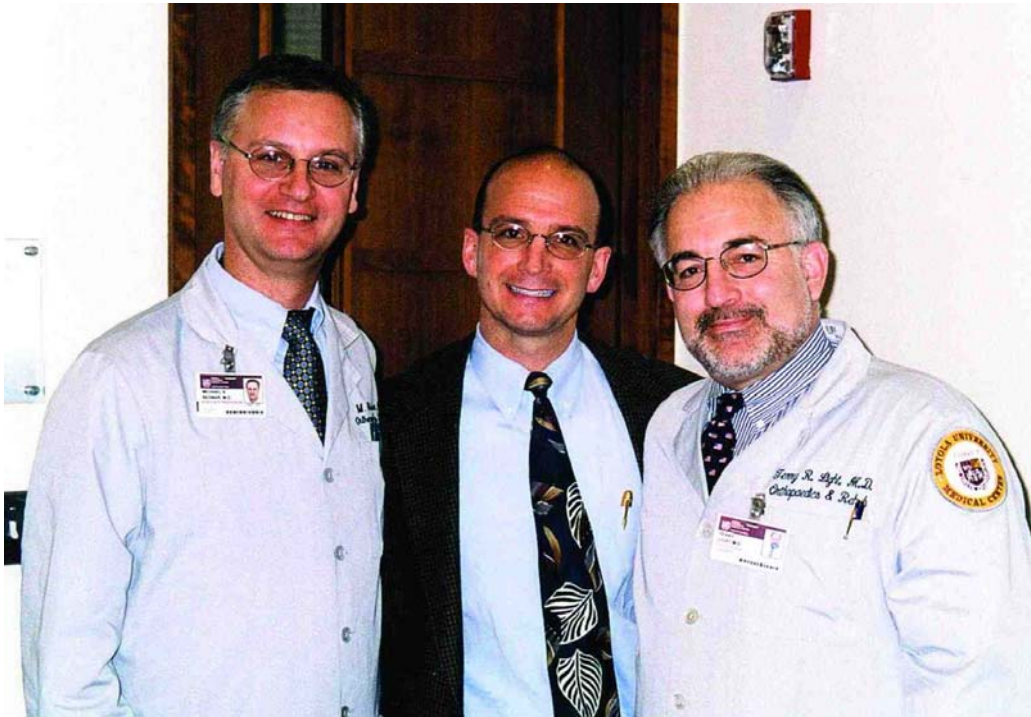


Figure 6: Michael Bednar, Terry Light, and myself during a visit to Loyola University and Shriners Hospital for Children.



Figure 7: Child with a pousse flottant prior to index finger pollicization.

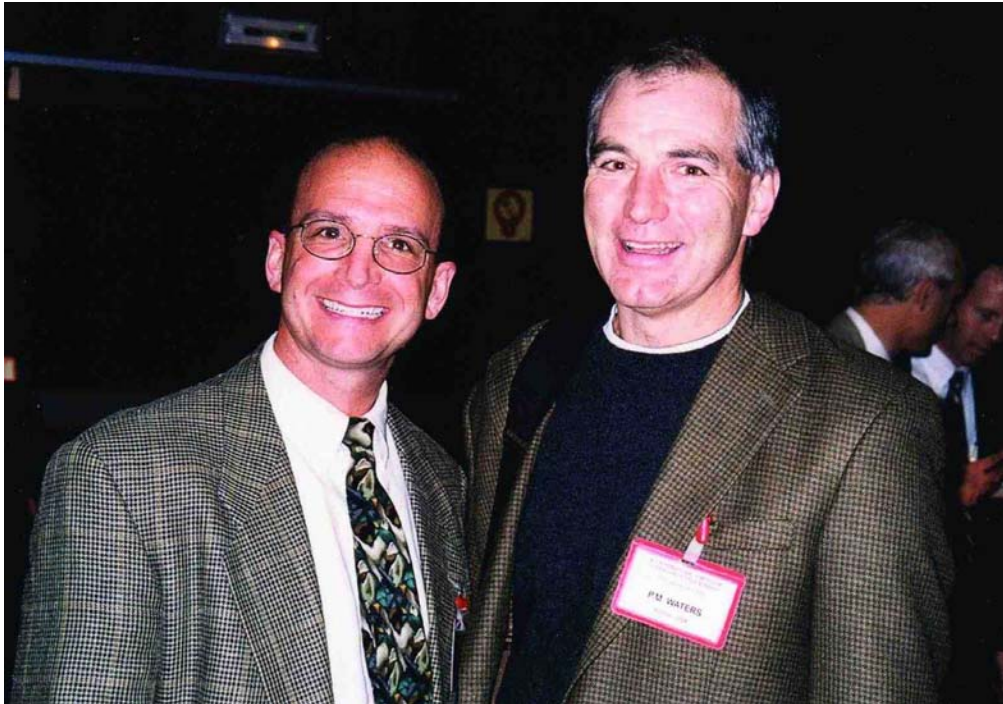


Figure 8: Peter Waters has been instrumental in developing a global brachial plexus data network.



Figure 9: Tom Wright, Longen Chen, Dean Sotereanos, me, John Taras, and Jai Ryu at the 4th International Congress of Asian Pacific Federation of Societies for Surgery of the Hand.



Figure 10: Dr. Sang Su Kim is a facile surgeon with vast experience performing contralateral C-7 nerve transfers.



Figure 11: Dean Sotereanos and Longen Chen seen with Professor Gu.



Figure 12: Dr. Xu and colleagues accompanied us during a day in the operating room. An ipsilateral C-7 nerve transfer was performed on a 20 year-old patient nine months after brachial plexus injury.

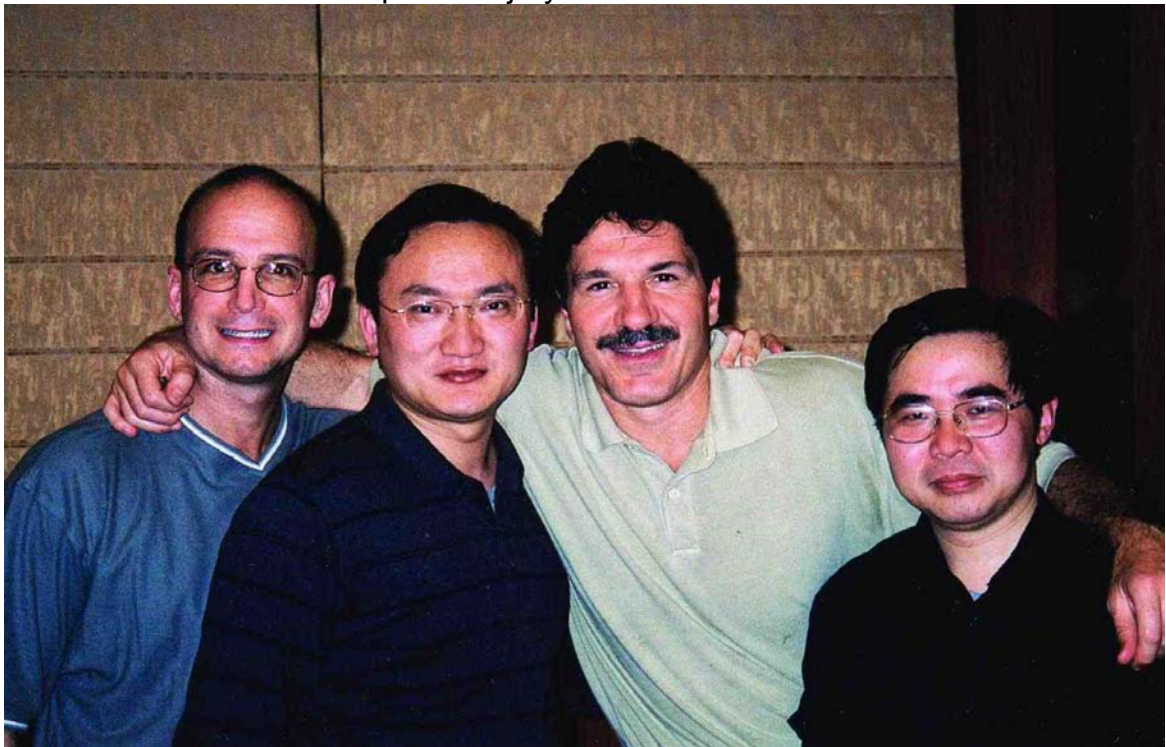


Figure 13: Dr. Xu from Shanghai and Dr. Rei from Wuxi are seen with Dean Sotereanos and myself during visit to Wuxi.



Figure 14: Great Wall of China at Juyongguan.



Figure 15: Mark Hughes is one of the scientists that were instrumental in the development of the technique pre-implantation genetic diagnosis.