PEDIATRIC ORTHOPAEDIC SOCIETY OF NORTH AMERICA's

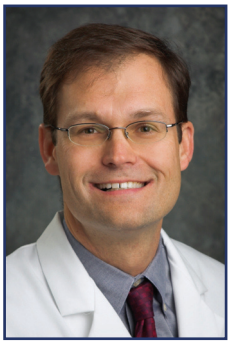
Resident REVIEW

CUTTING EDGE ORTHOPAEDIC INFORMATION ENHANCING RESIDENT EDUCATION

October 2012

From the Editor,

Steven L. Frick, MD



Welcome to the latest edition of the POSNA Resident Review-As summer ends, the cycle of residency education shifts from getting new interns and residents acclimated to new environments and responsibilities, and fall brings an opportunity to return to academic pursuits (maybe get back to that research project you have been planning to finish?). Another key fall activity for most residents is to begin to study in earnest for the OITE. The OITE is meant to provide a measuring stick to allow residents to see where they stand with regard to other residents across North America. The differences in how residency programs are organized, and the lack of a standardized curriculum, however, make it difficult to compare residents, especially in the first few years of training when everyone has not had exposure to all of the subspecialty areas of orthopaedics.

A major part of the Resident Review newsletter has been OITE-like questions on pediatric topics to help residents prepare for this examination. This edition focuses on neuromuscular disorders, and has 16 questions. In addition, the previous editions of the Resident Review are available on the POSNA website, and cover trauma, limb deformity, hip and foot disorders, spinal deformities, musculoskeletal infections and upper extremity problems. Hopefully these questions will help you prepare for the OITE, especially if you have not yet had the opportunity to go through your pediatric rotations.

In addition, this edition has other content to inform you about pediatric orthopaedic activities relevant to residents such as career opportunities in the Shriners Hospital system, the IPOS meeting and pediatric orthopaedic fellowship match.

Good luck preparing for the OITE (and getting some work done on that research project) this fall.

Congenital Clubfoot Update 2012: What we know, what we don't know and where we should go

Steven L. Frick, MD

What We Know

1. Congenital clubfoot is common (1 in 1000 births) and has been written about since the time of Hippocrates. It is bilateral in half of patients, and all of musculoskeletal structures beneath the knee seem to be affected (hypoplastic calf muscles, smaller foot). It is usually an isolated deformity, but can be associated with syndromes, developmental disorders and neurological abnormalities.

2. The components of clubfoot are cavus, adductus, varus and equinus, and can be remembered by the acronym CAVE.

3. Isolated clubfoot has a genetic basis in approximately 30% of cases (many being familial). The etiology in general is heterogeneous and an exact cause in isolated cases is rarely identifiable.

4. Even a well corrected clubfoot will have smaller, misshapen bones and limited range of motion of the ankle and foot compared to normal.

5. Current understanding of the principles of effective manipulation and casting, often combined with tendo Achilles tenotomy, can result in correction of deformity in the vast majority of patients. The most effective and least resource intense method of manipulative correction of clubfoot deformity is the Ponseti Method. Dr. Ponseti published his first paper on the results of his method in 1963, reported on those patients again in 1980, and there was a 30 year follow-up paper published in 1996. All of the papers described results that were at a minimum

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equivalent to the best results of other methods, and in most cases much better. Despite these positive publications, his method did not gain widespread acceptance until after he published a book in 1996, and importantly parental support groups began promoting the method in the late 1990s via the Internet.

6. Other manipulative methods can successfully correct clubfoot deformity, such as the French method popularized by Drs. Bensahel and Dimeglio. Comparative studies from TSRH in Dallas show slightly better results with the Ponseti method, and the Ponseti method utilized fewer resources.

7. After manipulative correction, clubfoot deformity tends to recur, mainly in the first few years of life when the foot is growing rapidly. Foot abduction orthoses worn at night for years lower the incidence of relapse.

8. Relapsed feet can be treated with repeat casting, tendon transfers to "balance" the foot to address supination "overpull" by the tibialis anterior, and repeat tendo Achilles tenotomy for recurrent equinus.

9. Clubfoot patients treated with surgical realignments that involve dividing ligaments, opening joints and pinning often end up with stiff, painful feet that function poorly.

10. A substantial number of childhood clubfoot patients undergoing joint-invasive surgery will have recurrent deformity and need repeat foot surgery in childhood. Some operatively treated clubfeet have significant, functional and life-altering complications.

What We Don't Know

1. What causes congenital clubfoot. An earlier study, in which pediatric orthopaedic surgeon Fred Dietz contributed, suggested that clubfoot was likely due to a single major gene defect. Another pediatric orthopaedic surgeon, Matt Dobbs, is currently leading research to look into the genetics of clubfoot, and has identified some genes that are involved in some families. We remain in search of "Ockham's razor", perhaps in vain.

2. Is the Ponseti Method the best we can do? The results of the Ponseti Method are obvious, nearly immediate, and are impressive to parents and practitioners. Some have suggested these criteria make the success of an intervention so self-evident that better evidence (EBM) is not needed. Our best long-term outcome study of Ponseti method patients involved only 45 patients, and 22% had a fair or poor outcome.

3. How much ankle dorsiflexion is enough? Most expert clubfoot surgeons using the Ponseti method perform Achilles tenotomy 90-95% of the time. The indications for tenotomy differ, with some using less than 10 degrees, while others use less than 15 or 20. How is this measured? And once the child is walking, how much ankle dorsiflexion is enough – 0 or plantigrade, 5, 10 or more degrees? We do not have validated information about this, and opinions seem to vary.

4. Does operative treatment for persistent or recurrent equinus lead to substantial improvement? One comparative study of Ponseti method verus Ponseti method plus posterior release found no difference in ankle dorsiflexion capability, and weaker active plantarflexion in the posterior release group. It is disappointing to divide all the tendons and ligaments that would limit passive ankle dorsiflexion and only get 5 or 10 degrees improvement- does this happen because of the abnormal shape of the talus?

5. While initial Achilles tenotomy in clubfoot seems to have few complications or functional implications, little is known about the indications for and long term results of repeat Achilles tenotomy or Achilles lengthenings after prior tenotomy.

6. How much can we expect the abnormal tarsal bone shape and size to change with successful Ponseti method treatment? Is the smaller, misshapen talus -one of the most consistent abnormalities described in morphological studies of clubfoot patients- able to be modified with any form of treatment, or is it a genetically programmed inevitability?

7. What is the ideal type and duration of abduction bracing? Many recent publications have noted non-compliance with bracing as the single most important factor related to relapse, yet many practitioners have patients that never werar their braces, yet do not relapse. Does everyone need to wear braces to prevent relapse, and for how long?

8. What is the best way to manage patients who do not respond as expected to Ponseti method manipulations and casting? This is uncommon, as most experienced Ponseti method practitioners report initial correction rates of 95-100%. But what should be the approach for the rare patient who does not respond? Do we apply the principles of "al a carte"

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Congenital Clubfoot Update 2012: What we know, what we don't know and where we should go

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surgery and address the residual deformities specific to that foot? Most frequently in my experience this will mean hindfoot surgery to address persistent equinus and varus- what are the long term results for these patients?

9. How many patients with clubfoot who relapse have neurological abnormalities that contribute to propensity to relapse? Some authors recommend neurology referral, electrodiagnostic testing and/or neuro-imaging for relapsing clubfeet. What are the indications for these types of tests, and what information do the tests yield?

10. What is the best approach for patients managed initially with the Ponseti method who relapse, are treated with repeat casting and then tibialis anterior transfer, but still have residual deformity leading to gait and functional abnormalities? What are the causes of persistent intoeing in clubfoot patients?

Where Should We Go?

1. Prior to his death in October 2009 at age 95, Dr. Ponseti saw his method gain worldwide acceptance, and in North America the application of his principles is credited with decreasing the number of annual clubfoot operative procedures from over 2000 cases to around 200 cases. Through education of residents in his home program at the University of Iowa, and then in courses beginning in 2000 and onward, Dr. Ponseti and others familiar with his methods have instructed many individuals in his specific principles and methods. This should continue to grow. The best way to learn the Ponseti method is self-study of his written materials, attending a hands-on course, and having some mentorship/feedback about management once you begin casting patients.

2. Research efforts to gain a better understanding of the etiology of congenital clubfoot should continue, utilizing the improved understanding and technology afforded by the human genome project. The theoretical idea of "fixing" a gene defect with gene therapy is attractive if a single gene cause can be identified, although currently it appears there are many candidate genes and a multifactorial etiology is suspected.

3. More centers with large numbers of clubfoot patients managed by the Ponseti method should collaborate to report results, especially regarding the best methods of management of the few patients per hundred who do not completely correct with initial treatment.

4. More prospective studies of brace duration and compliance are needed, with detailed data collection that will allow identification of low-risk and high-risk patients for relapse.

5. Define the amount of ankle dorsiflexion and how it is measured that is desired at the end of the acute correction phase (prior to final cast application) to guide indications for Achilles tenotomy, and define the amount of ankle dorsiflexion desired after walking begins to guide indications for repeat treatment of equinus.

6. Those surgeons who believe that the pendulum should swing back some to include some joint-invasive procedures in selected clubfoot patients (especially Ponseti method plus "limited" posterior release) should define and record their indications for surgical intervention, and report on mid- and long-term functional outcomes.

7. Experienced practitoners should combine series of patients with persistent gait and functional problems after relapse treatment with tibialis anterior transfer, and study/report on the results of differnet treatments for these patients. This would allow construction of a framework for clinical decision-making for these patients.

8. Other methods to prevent relapse after initial correction should be explored, such as peroneal muscle strengthening or stimulation, and the effects of defined passive range of motion exercises.

9. The indications for and expected outcomes of more intensive evaluations of neurological anatomy and function in children who have relapses should be studied.

10. More studies using sophisticated, non-ionizing radiation imaging to better understand the abnormal tarsal and mid-tarsal morphology and kinematics, and how these are affected by successful treatment.

"The author thanks Drs. Dobbs, Herzenberg, Raney, Talwalkar, Pirani, von Bosse, Mosca and Noonan for answering a survey about our current knowledge of clubfoot."

WEBINAR: Tips for the Orthopaedic Fellowship Match
September 24, 2012, 8:15 pm Eastern

To: All PGY-3 and PGY-4 Orthopaedic Residents

Tips for the Orthopaedic Fellowship Match
Choosing a fellowship is an important step in one's career. The AAOS Board of Specialty Societies (BOS) Match Oversight Committee has an exciting educational webinar planned for September 24th (8:15pm ET; 7:15pm CT; 6:15pm MT; 5:15pm PT) regarding the Fellowship Match process.

The **"Tips for the Orthopaedic Fellowship Match"** is an interactive webinar open to all residents wanting to learn more about the match. The webinar is hosted by Lisa Cannada, MD, Chair, BOS match Oversight Committee and the webinar faculty includes Fellowship Directors, a representative from San Francisco Matching Program and recent participants in the match process! Webinar participants will learn the history of the match, helpful match statistics for each subspecialty match for the past 3 years, tips from program directors and information on what to look for in choosing a fellowship. The participants will learn questions to ask when choosing a program and also have a chance to email questions to the faculty on the webinar.

Sign up now for this informative webinar designed to help you navigate the fellowship match process.

An Interview with Peter Armstrong

Peter Armstrong is Chief of Staff-Emeritus for the Shriners Hospitals for Children. He served as the Chief of Staff for the Shriners Hospitals for Children system from 2000-2011. Prior to that appointment, he was Chief of Staff at the Shriners Hospital for Children in Salt Lake City. In the interview below, Dr. Armstrong shares his thoughts about his career within the Shriners Hospitals for Children system and what the future holds for it.

1. Can you tell us how you ended up in pediatric orthopaedics?
I did my orthopaedic residency at the University of Toronto. When I did my 6 month rotation at The Hospital for Sick Children (Sick Kids), I knew for sure that I wanted to be a pediatric orthopaedic surgeon. I loved working with children and being able to do something that made a real difference in their lives. I also enjoyed interacting with the families. I still occasionally hear from my patients and families, both from my time in Toronto and my time in Salt Lake City. When I finished my residency, Dr. Robert Salter asked me to be his clinical fellow. He then played a key role in getting me a position on staff at The Hospital for Sick Children.

2. What was it like being a Canadian and practicing in the American system?
I can't say that I have ever really experienced the "American" system. I went from a "single payer" system in Toronto to a fully salaried position as Chief of Staff of the Shriners Hospital in Salt Lake City. As you can see, I have lived a life sheltered from the need to interact with insurance companies, CMS, etc. The one very strong perception that I had in moving from Canada to the US was the incredibly strong sense of competition that exists down here. For example, when the Ilizarov approach first came on the scene, we formed a group with participants right across Canada who were very willing to share their data for "the common good". The willingness to cooperate and collaborate freely seemed much more common in Canada.

3. How do you balance the demands of your job and the demands of your family?
This is a question that is better asked of my wife and children. To answer this question truthfully, I have to tell you that there were more times than I would like to admit when I failed miserably at this. It can be so easy to take those you love for granted. The pressures, particularly in academic medicine, are constant and insatiable. I realized that if I

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didn't proactively work hard on that balance, nobody else would do it for me. Cath prayed for me earnestly and encouraged me to spend more time in God's Word so that I would know, without a doubt, what God expected of me. It was very clear that my priorities must be God, Family and Work in that order. I believe I always knew that but there is a huge difference between knowing and doing. With His guidance, I began making the necessary changes to my priorities. That is what made the difference for me.

4. Can you tell us about why you chose to work within the Shriners System?
When I was on staff at Sick Kids, Colin Moseley announced that he had accepted a job as the Chief of Staff of the Shriners Hospital in Los Angeles. As we met at POSNA and other meetings, it was very clear how much Colin loved his work there. When I received the letter announcing the position of Chief of Staff of the Shriners Hospital in Salt Lake City, I discussed it with Cath and headed off to POSNA in Dallas in 2000. I had great respect for Sherm Coleman and thought of the opportunity to learn from him. Hugh Watts was the one who facilitated Newt McCollough and me getting together at that meeting. Newt told me about Shriners Hospitals in general and the hospital in Salt Lake City in particular. With Cath's support, I decided to interview for the position and was selected. I never regretted that decision although it was difficult to leave Sick Kids. The thing that I loved about the time there was that we could look at children, determine what they needed and do it without any hindrances such as copays, deductibles, concern about lengths of stay, etc. We truly did provide family centered care. There was a real sense of "Team" among all the hospital staff. I also greatly valued and enjoyed the camaraderie that existed among the medical staffs of the other 21 hospitals.

5. What has changed within the system from when you started until now? This can be applied to both the people in it and the system itself.
What changed was the economy. That brought Shriners Hospitals for Children face to face with the reality that, if we were going to survive in the long term, we absolutely had to begin operating much more efficiently and cost effectively. When the most recent economic downturn occurred, it became very clear to the "owners" (Shriners) and staff that we needed to embark upon the largest cultural and operational change that had ever occurred within the system. The Shriners made two critical decisions at their major meeting in July 2009. The first was that SHC would begin the process of implementing third party billing in an effort to help offset the cost of operating 22 hospitals. The second was that the Boards of Directors and Trustees at the national level were given the authority to change the care delivery model at any of the 22 hospitals. A Strategic Assessment Task Force was formed and is currently carefully looking at all 22 hospitals to determine what the optimal model for each facility is. Change is always difficult and not uncommonly, painful, especially for an organization that had functioned independently for so many years. Nevertheless, the change is necessary if we are to continue to exist well into the future.

"Nevertheless, the change is necessary if we are to continue to exist well into the future."

6. Who do you think is the ideal Shriners surgeon in 2013?
The ideal Shriners surgeon is one whose passion is for making a demonstrable difference in a child's life by utilizing the best medicine has to offer at the time (evidence based medicine). Quality and safety must be the highest attributes of the care provided. The surgeon must recognize that the only way this can be accomplished is through a multi-disciplinary team. He/she must recognize and respect the knowledge, experience and value of each member of the team. The surgeon must never be complacent in believing that what he/she is doing now is the best but rather constantly looking for ways to do it better. That is a commitment to research of varying degrees. Each surgeon must recognize that he/she has a responsibility to teach those who are coming along behind, as well as share their knowledge with others around the world through publication and presentation. He/she also needs to be very knowledgeable about the cost of

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An Interview with Peter Armstrong

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providing care and be proactively involved in making sure that the highest quality of care is provided at the lowest cost.

7. **What does the Shriners Hospital system look like in 2020?**
I believe it will still be true to its stated mission of excellence in patient care which is inextricably linked to research and education. There will be 22 locations but there will undoubtedly be variations in the care delivery models throughout the system. It is certainly possible that there may be additions/changes in the service lines offered in the facilities.

8. **Now that you are "Chief emeritus" what does the next phase of your life include?**
I am too young to retire completely! I do, however, hope to spend more time with Cath, our kids and grandkids. Cath and I hope to further develop our interest in photography. I may even get my sailboat out of the Public Storage garage in which it currently resides!

From the professional perspective, I plan on continuing, and perhaps increasing, my work as the Chairman of the Surgeon Advisory Board and member of the Board of Directors for OrthoPediatrics.

Focus: Neuromuscular Challenging Cases. What would you do?

CASE #1

A 14 year old boy presents to clinic with his parents for evaluation of pain under the lateral border of his feet with ambulation (Figure 1 a,b,c). He reports his feet started to look like this "years ago". The next step in the evaluation/treatment should be:

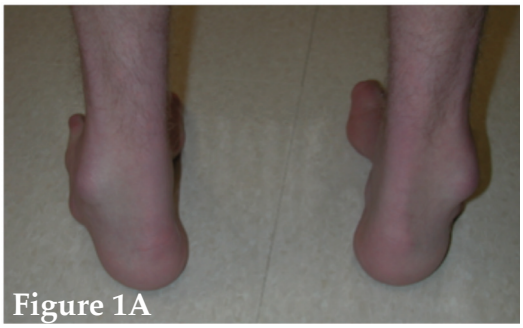
Figure 1A

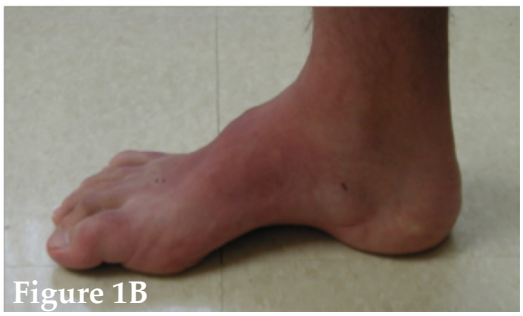
Figure 1B

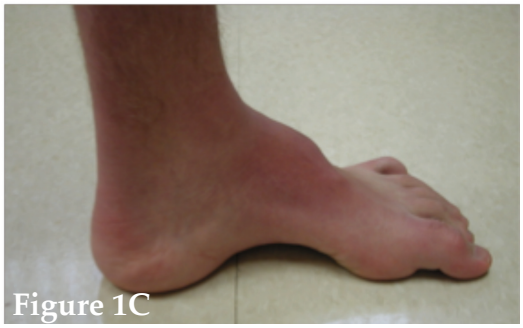
Figure 1C

A. Staged foot surgery consisting of tendon transfers and midfoot osteotomy with plantar fascia release.

B. MRI Scan of his spinal cord and posterior fossa.

C. Plain radiographs of his feet.

D. Examination of his parents feet and referral to neurology

E. Obtain a blood sample for CPK and aldolase and schedule a muscle biopsy.

Correct Answer is D.

This boy has bilateral cavovarus feet. Children with unilateral cavovarus feet may have this condition as a result of spinal dysraphism such as tethered cord and an MRI of the spinal cord is needed as a first step in these cases. Bilateral foot deformity can be due to hereditary motor sensory neuropathies (HSMN 1-7) (ex: Charcot Marie Tooth disease), Friedreich's Ataxia, Ataxia Telangiectasia and a host of other genetic disorders with phenotypic changes in the neuro-axis. Many of the HSMN's are autosomal dominant inheritance and therefore examination of the parent's feet is a good start in the evaluation. Further evaluation with a neurologist may include EMG-NCV testing or genetic testing for known defects such as PMP-22. Serum analysis for muscle enzymes will be of low yield as this is not likely a result of primary myopathy. Surgical planning with the aid of radiographs and execution of the plan with transfers and osteotomy should only be considered after a thorough evaluation.

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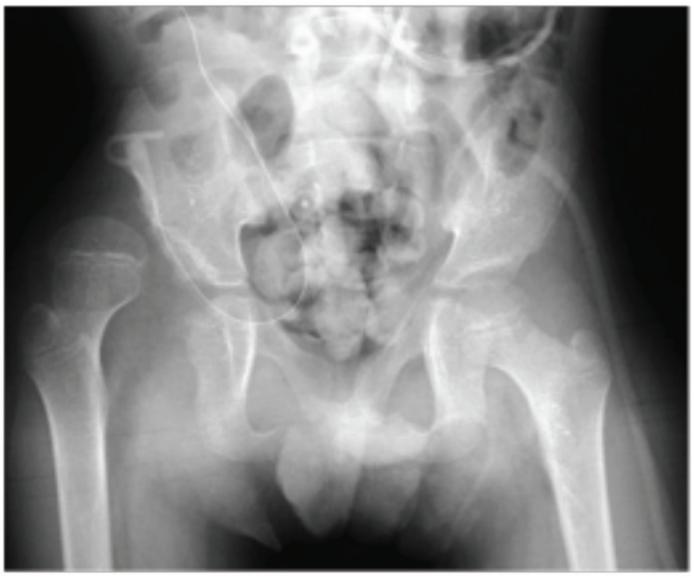
CASE #2

An 8-year-old boy with L2-3 level spina bifida has bilateral knee flexion contractures of 20 degrees and has had clubfeet reconstructions with 10-degree residual equinus deformity. He can stand and can ambulate with a swing-through gait with Lofstrand crutches and anterior floor reaction AFO's with a 2-centimeter shoe lift. A radiograph of his pelvis is included. The most likely factor that will prevent him from becoming a good community ambulator as an adult will be:



A. Progressive crouch at the knees with anterior knee pain

B. Residual equinus deformity and potential for foot sores

C. Limb length discrepancy and need for shoe lift

D. Muscle weakness

E. Osteoarthritis of the hip.

Correct Answer is D.

Children with myelomeningocele can develop a host of orthopaedic problems which include contracture and dislocation. The severity and incidence of these deformities is directly dependent on the motor level of the patient. The most important factor that predicts ambulatory potential is motor level. Adults with motor level functioning at L4-5 may have some ability for community ambulation. Adults with L2-3 level function usually get around in the community with a wheelchair.

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CASE #3

You are called to the operating room by a neurosurgical colleague who is performing a closure of a spina bifida defect in a 2 week old. There is a "mass" that he can't seem to cover with skin. His initial management was to apply a skin graft substitute over the area and attempt rotation flaps. Clinical photo and radiograph below. The appropriate treatment is:

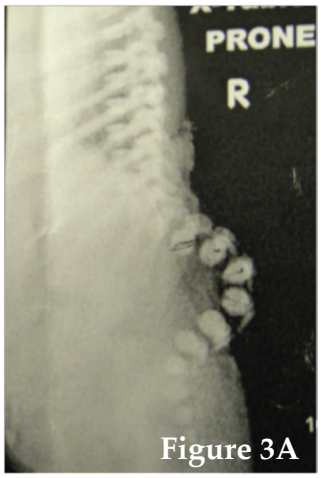

Figure 3A

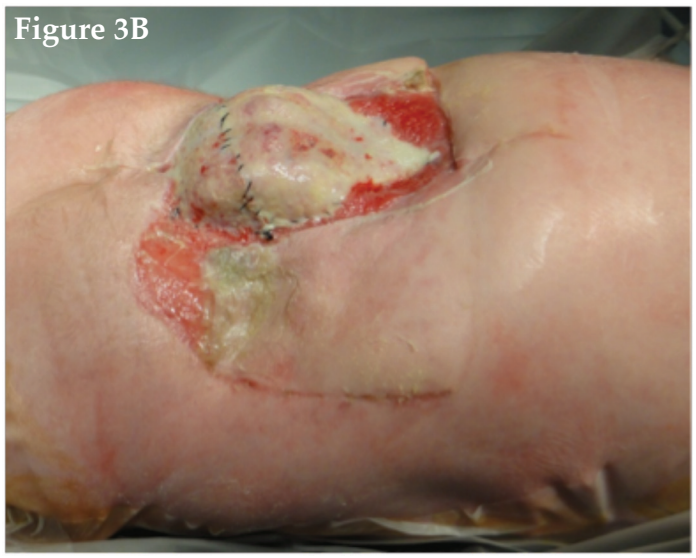

Figure 3B

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a. "shaving" the prominent bone

b. Instrumented limited fusion

c. Growing instrumentation (i.e. growth rod or VEPTR)

d. Kyphectomy

e. Free flap coverage by plastic surgical colleague

Correct Answer is D.

Defect closure in spina bifida is necessary to avoid infection and early death in children with myelomeningocele. This is usually done by addressing the vestigial neural elements and undermining and closing the skin. In some children, however, significant congenital deformity exists that precludes this, and in these cases neonatal kyphectomy is indicated. Fixation is usually with sutures supplemented with casting, as small size precludes instrumentation, growing or otherwise. Simply shaving the bone or removing the pedicle masses, or plastic surgical coverage, will not solve the problem. It is important to be aware of this potential issue in centers where in-utero repair is considered, as this may not be possible with a severe deformity. For this reason, prenatal MRI is being performed with increasing frequency in these cases.

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CASE #4

A 7 year-old presents to you because of gait abnormalities. He walks with significant lordosis of his spine and a wide based, waddling gait. He has very muscular appearing calves. When getting up from the floor, he "climbs" up his legs with his hands. The initial diagnostic step should be:

a. MRI of the lumbar spine

b. Muscle biopsy

c. Nerve conduction testing

d. Blood Creatinine Phosphokinase levels

e. Genetic testing

Correct Answer is D.

The patient has the classic phenotype of Duchenne's Muscular Dystrophy. As the proximal muscle weakness worsens, these patients go into lordosis to tension the anterior hip ligaments to compensate for weak hip extensors. Abductor weakness leads to a waddling gait. While their calves appear muscular, the enlargement is actually due to infiltration with fat and fibrous tissue- this is known as pseudohypertrophy. The Gower's sign is elicited by asking a child to stand from a seated position on the floor. Lower extremity weakness leads to the need to use the hands to push off of the legs. While muscle biopsy may lead to the diagnosis, it is not necessary in most cases. MRI and nerve testing are unlikely to reveal abnormalities. While genetic testing is usually done to further define the illness. CPK levels are simple and show massive elevation in children with DMD, often into the tens of thousands. Therefore the best initial step is D.

References

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CASE #5

A 4 year male presents to clinic with a chief complaint of new onset toe-walking. Patient met initial milestones but has had difficulty with running and jumping. Physical exam reveals a waddling gait, increased calf size, as well as a positive Gower sign. What is the most common inheritance pattern for this child's condition?

a. Autosomal dominant with anticipation.

b. Autosomal recessive.

c. X-linked recessive.

d. Maternal (mitochondrial) inheritance.

Correct Answer is C.

Discussion

Duchenne's Muscular Dystrophy is the most common of the muscular dystrophies, with an incidence of 1 per 3500-6000 males. The gene responsible for DMD is located on Xp21 region of the X chromosome which codes for dystrophin, a protein present in smooth, skeletal and cardiac muscle. The inheritance pattern for DMD is

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X-linked recessive with 1/3 of cases being spontaneous mutations. Diagnosis is typically made between 3 and 6 years of age with lower extremity weakness preceding upper extremity weakness. Ankle equinus leading to contracture and toe walking is often the first presenting sign. Progressive proximal muscle weakness coupled with contractures lead to further changes to the normal gait pattern. The Gower's sign may be present as early as 15 months of age in the patient with DMD: when a patient is asked to stand up from a seated position on the floor, the patient will walk his hands up his body to assist in elevating his trunk due to the proximal muscle weakness. The diagnosis is suppoted by initial tests of blood muscle enzyme levels, as the lack of dystrophin causes muscle cell membrane fragility leading to extremely high levels of muscle enzymes in blood. The diagnosis can now be confirmed almost all of the time with genetic testing for Dystrophin deletion/duplication.

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CASE #6

An 8 year old child with Cerebral Palsy is able to ambulate independently with the use of forearm crutches. The child would have a Gross Motor Function Classification System Level of:

a. I.

b. II.

c. III.

d. IV.

e. V.

Correct Answer is C.

Discussion

The GMFCS level was introduced in 1997 out of McMaster University. It is based on self-initiated movement by the patient with the emphasis being on the abilities of the patient in meaningful activities of daily life rather than on limitations. There are broad categories for the GMFCS that reflect developmental milestones: before the 2nd birthday, between 2 and 4, between 4 and 6, between 6 and 12 and between 12 and 18.

GMFCS I: walks without limitation

GMFCS II: walks with limitation (eg: holds onto railing while going up stairs)

GMFCS III: walks using a hand-held device (canes, crutches, walker, etc…)

GMFCS IV: self-mobility with limitation may use powered mobility.

GMFCS V: Transported in manual wheelchair.

References

GMFCS-E&R®Robert Palisano, Peter Rosenbaum, Doreen Bartlett, Michael Livingston, 2007; *CanChild* Centre for Childhood Disability Research, McMaster University

GMFCS®Robert Palisano, Peter Rosenbaum, Stephen Walter, Dianne Russell, Ellen Wood, Barbara Galuppi, 1997; *CanChild* Centre for Childhood Disability Research, McMaster University
(Dev Med Child Neuol 1997; 39:214-223)

CASE #7

An infant with myelomeningoceole undergoes repair of his neural tube defect at 2 days of life. He begins walking at 21 months of age with the aid of AFOs. At age 5, he demonstrates 5/5 strength in bilateral quadriceps, but he is insensate with no active motor function below the knee. His feet are flexible with mild hind foot varus and he shows no evidence of hip dysplasia. The child's parents ask for a comment on the anticipated function of their child when they graduate high school. Your best response should be "Several factors can influence your child's level of function including body mass, your child's motivation, and potential for change in the neurologic defect but I believe

1. Your child will be able to ambulate with a reciprocating gait orthosis (RGO) for exercise using a wheelchair at other times.

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2. Your child's function will improve as he grows and he may no longer require braces

3. Your child will likely walk with SMO (supramalleolar) orthotics

4. Your child will likely be able to walk with AFOs but may require other ambulatory aids.

5. I cannot predict your child's future function.

Preferred Response 4. This child has an L4 neurologic level. Strong quadriceps function is predictive of community ambulation but the lack of motor power below the knee demands an AFO to control the ankle and reduce energy demand. A reciprocating gait orthosis is used to allow exercise only ambulation for high lumbar or thoracic level patients. Supramalleolar orthotics may be adequate in the sacral level spina bifida patient with only foot deformity and some gastrocsoleus function.

CASE #8

A 9-year-old boy with an L-4 level myelomeningocele is scheduled to undergo surgery for equinovarus feet. In addition to shunt clearance, what other precautions should be taken in the perioperative period?

1. Latex-free environment

2. Avoidance of fiberglass casting material

3. Avoidance of intravenous narcotics

4. Hyperthermia prophylaxis

5. Preoperative echocardiogram

Preferred Response: 1. Patients with myelomeningocele are at risk for development of serious latex allergy. Risk factors include history of prior allergic reactions and multiple previous surgeries particularly urologic and orthopaedic procedures. Cardiovascular collapse during major thoracic or abdominal surgery is the most serious manifestation. Current practice is to perform surgery and all invasive procedures in a latex-free environment for all patients with myelomeningocele.

Equinus contractures are common in patients with myelomeningocele. Patients can initially be treated with passive manipulation. If the deformity persists when the child is ready to stand, lengthening of the heel cord may be indicated.

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Latex anaphylaxis during surgery in children with myelomeningocele. Dev Med Child Neurol 1993; 35: 543.

CASE #9

In the case of a mother (who is a carrier for Duchenne's muscular dystrophy gene) and a father (who does not have the Duchenne's muscular dystrophy gene), what is the chance that the son will be affected by Duchenne's muscular dystrophy?

1. No chance

2. 50%

3. 75%

4. 100%

5. Unable to determine since the father may be a carrier.

Preferred Response: 2. Duchenne's muscular dystrophy (DMD) is the most common form of muscular dystrophy, occurring in 1 in 3500 boys. It is transmitted in an x-linked recessive fashion whereby all affected persons are male; females are carriers of the gene. Molecular genetic testing has eliminated the need for muscle biopsy to establish the diagnosis. On very rare occasions, females with Turners syndrome may exhibit the disease because of their XO genotype.

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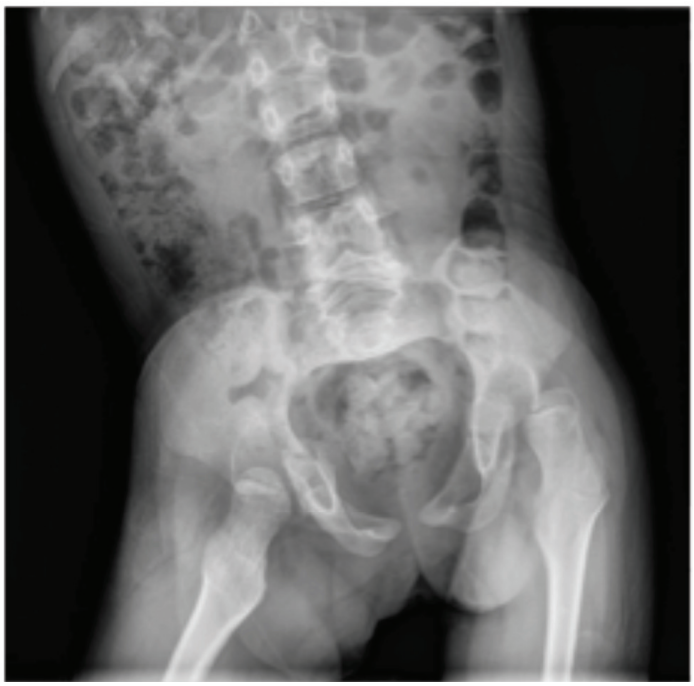
CASE #10

This nonambulatory 7-year-old girl with spastic quadriplegia has hip abduction of 30° on the right and 10° on the left. An AP pelvis radiograph is shown. Treatment should consist of



1. Hip abduction bracing.

2. Observation, with repeat AP pelvis radiographs in 1 year.

3. Botulimun toxin to the hip adductors bilaterally, and physical therapy.

4. Bilateral hip adductor releases without osteotomy, followed by abduction bracing.

5. Bilateral varus derotation femoral osteotomies and bilateral hip adductor releases.

Preferred Response: 5. Hip subluxation and dislocation is a common problem in children with cerebral palsy. Patients with spastic diplegia are at increased risk, and nonambulatory patients are at much higher risk than those who can walk. Hip subluxation develops in response to muscle imbalance; spasticity and contracture of the adductors and flexors overpower the hip extensors and abductors. The typical remodeling of femoral anteversion seen in a neurologically normal child does not occur and femoral anteversion persists. Coxa valga develops and the lesser trochanter elongates with pull of the iliopsoas. Bony surgery in the absence of soft tissue release is ineffective in correcting the subluxation and similarly if bony changes are seen, soft tissue surgery alone is likely to fail. Consideration should be given to correction of acetabular dysplasia at the same setting and many surgeons would add a pelvic osteotomy to the surgical prescription for the right side.

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Huh K, Rethlefsen SA, Wren TA, Kay RM. Surgical management of hip subluxation and dislocation in children with cerebral palsy: isolated VDRO or combined surgery? J Pediatr Orthop. 2011 Dec; 31(8):858-63.

Spiegel DA, Flynn JM: Evaluation and treatment of hip dysplasia in cerebral palsy. Orthop Clin North Am 2006; 37:185-196.

CASE #11

A posterior leaf spring ankle-foot orthosis would be appropriate for which foot and ankle misalignment pattern in a child with spastic-type cerebral palsy?

1. Absent heel strike, excessive plantar flexion in the swing phase, and 5 degrees of passive ankle dorsiflexion

2. Excessive ankle dorsiflexion in midstance caused by incompetence of the ankle plantar flexors

3. Crouch gait pattern with excessive ankle dorsiflexion, increased knee flexion, and increased hip flexion in midstance

4. Excessive supination of the hindfoot during stance, which is passively correctable

5. Significant knee instability and weakness with stance in a child who is minimally ambulatory

Preferred response: 1. Orthoses can be helpful in improving function in ambulatory patients with cerebral palsy. Ankle-foot orthoses (AFO) are prescribed to assist the child in positioning the ankle and foot during gait. Posterior leaf spring orthoses (PLSO) are made from more pliable polypropylene and provide a little push off at terminal stance. The factors that make the posterior leaf spring ankle-foot most appropriate are the clinical presence of absent heel strike and minimal (but some) dorsiflexion. The PLSO is designed to control excessive ankle plantar flexion (control equinus) in the swing phase and allow ankle dorsiflexion in midstance.

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A solid AFO is both a stance and swing-phase control orthosis, which can help when there is minimal dorsiflexion of the ankle. Hinged AFO's allow some dorsiflexion during stance but block plantar flexion thereby eliminating equinus. A ground reaction AFO has an anterior shell at the proximal tibia, to provide push back on the knee during stance and help the knee extend. A crouch gait pattern may attempt to be treated with a floor-reaction AFO. Supramalleolar orthoses are used to control flexible coronal plane deformities, such as excessive supination or pronation of the hindfoot.

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CASE #12

A 4-year-old girl with an L4 myelomeningocele presents for routine follow-up. Pelvic radiographs reveal a complete dislocation of the left hip with well-formed acetabulum, and a normal right hip. Her gait is symmetric with use of a walker and brace. Which of the treatment options should be offered to the patient at this time?

1. Right sided femoral shortening osteotomy

2. Continued observation and routine follow-up

3. Left greater trochanteric advancement

4. Left sided pelvic osteotomy

5. Open reduction of the left hip, correction of any bony abnormality and muscle transfers

Preferred response: 5. The most difficult hip deformity in patients with myelomeningocele is paralytic subluxation and dislocation. Paralytic dislocation of the hip occurs because of paralysis of the hip abductors and extensors w/ unopposed pull of the hip flexors and adductors. Surgical reduction of hips in patients with spina bifida is associated with a high failure rate and therefore treatment indications are controversial. If reconstruction of the dislocated hip is considered it must include (1) concentric reduction, usually by open reduction, (2) correction of bony abnormalities on the (femoral osteotomy to correct anteversion, Dega osteotomy on the pelvic side) and (3) balance the flexor-adductor, extensor-abductor imbalance. Transfer or release of muscles to balance the hip result in diminished muscle strength, even when balance is achieved. Some believe that unilateral dislocation in good ambulators strengthens the indications for hip stabilization to prevent limb length inequality, pelvic obliquity and scoliosis.

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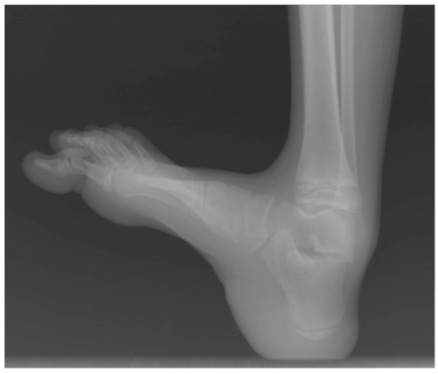
CASE #13

A 7-year-old boy presents to clinic with a diagnosis of spina bifida and progressive foot deformity. He can walk independently and on exam has motor function intact for his EHL, tibialis anterior, and common toe extensors but no gastrocsoleus function. His foot radiographs are shown in Figure 1 (attached). What level myelomeningocele is this patient, and what surgical procedure would help with his foot deformity?



1. S1 level, Symes amputation

2. S1 Level, Gastroc Lengthening

3. L5 Level, Tibialis Anterior Tenotomy

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4. L5 Level, Split Anterior Tib Transfer Posteriorly

5. L4 Level, Triple arthrodesis

Preferred Answer: 4. For the first part of the question, you have to determine what level myelomeningocele the patient is. This patient demonstrates intact motor function to his tibialis anterior, EHL, and common toe extensors, which represents a distal L5 level. He has no gastrocsoleus function, which would be S1 function. His radiographs demonstrate a calcaneus deformity with dorsiflexion of the calcaneus and the remainder of the midfoot relative the tibia. This represents overpull of the Tib Ant with no function of the gastrocsoleus to counteract. A tibialis anterior tenotomy would render the foot unable to dorsiflex or plantarflex, which would make ambulation more difficult, so the correct answer is to perform a split anterior tibialis transfer posteriorly. This would help with achieving some plantarflexion motion and a more plantigrade foot, which was demonstrated in the paper by Park et al.

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CASE #14

An 11 year old boy has a diagnosis of Duchenne Muscular Dystrophy. He has recently become wheelchair bound and pulmonary function tests reveal his FVC is 70% of normal. Upon routine screening radiographs in the orthopedic clinic, he is found to have a 23 degree thoracolumbar curve. What should the appropriate treatment be for his scoliosis?

1. No treatment is recommended because of the long-term poor prognosis of the disease process.

2. Bracing should be instituted because it will help delay progression of his curve until maturity

3. Posterior spinal fusion should be performed if his FVC drops below 60%

4. The patient should be monitored and posterior spinal fusion performed if and when the curve is greater than 50 degrees

5. The patient should undergo posterior spinal fusion based on the current radiographs

Preferred Answer 5. In patient with Duchenne Muscular Dystrophy (DMD), although there is a poor long-term prognosis, advancements in medical care have helped these patients live longer with better functional outcomes, so the spinal deformity is usually treated. Bracing has been shown to be ineffective for helping with the spinal deformity of DMD, so that should not be an option. With adolescent idiopathic scoliosis, current recommendations are to consider surgical treatment when the curve reaches 50 degrees, management of scoliosis in DMD should be more aggressive because of the pulmonary complications with increased curves shown in this patient population. Complications increase as the curve progresses and pulmonary function drops below 40%, so attempts should be made to perform surgical correction before pulmonary function deteriorates below that number. The role of surgery in improving pulmonary function and prolonging life in these patients is controversial, but patients who develop curves of 20 degrees or greater magnitude typically have substantial progression, so correction of the spinal deformity is recommended when the curve reaches 15-20 degrees to lessen the chances of pulmonary complications.

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Sussman M. Duchenne muscular dystrophy. JAAOS 2002; 10: 138-151

CASE #15

A 13 year-old non-ambulatory male with Duchene Muscular Dystrophy presents for evaluation of a mild spinal deformity. Radiographs of the spine are shown with the Cobb angle measuring 36 degrees. What is the most appropriate management for this patient's scoliosis.

1. Custom TLSO to prevent further curve progression

2. Observation and later spinal fusion if the curve progresses beyond 50 degrees

3. Selective Fusion of the lumbar portion of the curve only to maintain seating balance

4. Posterior Spinal Fusion from T2 to the Pelvis

5. Anterior and Posterior Spinal Fusion from T2 to the Pelvis

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Correct Answer is 4.

A considerable number of boys with Duchenne muscular dystrophy (DMD)develop progressive scoliosis following the loss of ambulation. Because pulmonary function in this patient population declines steadily with increasing age, surgery that is performed at a younger age (and lower curve magnitude) is felt to decrease the likelihood intra-operative and post-operative pulmonary complications. Accordingly, most authors recommend spinal fusion when the scoliosis reaches 20 to 30 degrees.

There is no role for bracing in boys with DMD as curves progress despite spinal orthotic use. Furthermore, using a brace to delay progression would merely result in surgery being required at an older age when cardiopulmonary function has declined and anesthetic risks have increased. For similar reasons, observation until curves increase in magnitude is not warranted in patients with DMD.

Selective fusions in patients with DMD are contraindicated as fusions that do not extend to the proximal thoracic spine may result in proximal or junctional kyphosis with a subsequent loss of head control. Though controversy exists as to the distal extent of spinal fusion, most authors agree that the fusion should extend to L5 or the pelvis. There is no role for anterior spinal fusions in patients with DMD as anterior approaches would further impair pulmonary function in an already compromised patient population.

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Smith AD, Koreska J, Moseley CF. Progression of scoliosis in Duchenne muscular dystrophy. J Bone Joint Surg Am. 1989; 71: 1066-74.

CASE #16

A mother brings her 10-year-old daughter in because of progressively high-arched cavus feet. The father also has cavus feet. The mother is concerned because the girl's teacher has noticed that the girl's gait is becoming more awkward. Physical exam reveals 1+ reflexes at the knees and ankles, calluses under the metatarsal heads, slightly decreased vibratory sensation on the bottom of the feet, and slight weakness of the peroneal and anterior tibialis muscles bilaterally. Xrays are obtained of the girl's feet and hips and are shown below. Not surprising, the teacher has also noticed:

A. A deterioration in the girl's reading comprehension

B. A deterioration in the girl's handwriting

C. The girl has been requesting more frequent trips to the rest room

D. The girl has been complaining of frequent headaches

E. The girl has been sitting out of gym complaining of back pain

This girl most likely has a form of Charcot-Marie-Tooth Disease, or hereditary motor sensory neuropathy (HMSN). Although more common in males, it tends to affect females more severely. Dysfunction of myelinization and degenerative changes in the motor nerve roots affect the peripheral nervous system. CMT1 (demyelinating) , CMT2 (axonal), and CMTX (X-linked dominant) are some of the subtypes that have been linked to over 40 genetic mutations [including peripheral myelin protein 22 (PMP22), myelin protein zero (MPZ), connexin 32 (Cx32), and numerous other genes.]. The feet are most frequently involved. Weakening of the intrinsic muscles, peroneus brevis, and tibialis anterior results in cavovarus deformity with claw toes. Similarly, peripheral neuropathy may cause intrinsic weakness in the hands, thus causing hand fatigue or deterioration in handwriting. Headaches, cognitive function, and central nervous system issues are not likely to be associated with a peripheral neurologic disorder.

Preferred Response: B.

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POSNA Tech Corner – Fall 2012

Orrin Franko

Orthopaedic textbook and journal publishers are beginning to offer mobile access to content via iPad apps. This month's column will highlight some of the most useful apps for accessing orthopaedic literature from your mobile tablet. For a complete list of reviews and apps for iPads, iPhones, and Android devices, visit www.TopOrthoApps.com.

AAOS eBooks:
AAOS has updated their new eBookstore to include an iPad app, called AAOS eBooks, that provides mobile access to titles purchased from their web-based store. As of writing, the store includes 8 popular titles, but is expected to increase to over 30 titles within the next few months. After logging in and registering purchased books, users can download them selectively, ranging in size from about 10-80 Mb. The app includes typical viewing options, including the ability to change the font, change text size, adjust brightness, highlight text, make notes, and place bookmarks. The annotation functions are primitive, at best, and even the instructions for highlighting require 4 separate steps each time. The books themselves are of high quality, text is crisp, and images are clear, however the user cannot increase the image size, which often prevents fully appreciating many of the detailed illustrations. Despite its limitations, I commend the Academy for recognizing a shift in the format of educational materials.

Bone & Joint
The British Editorial Society of Bone and Joint Surgery has also leapt into the mobile world with Bone and Joint, an iPad and iPhone app for viewing their collection of publications: *The Journal of Bone & Joint Surgery (British), Bone & Joint Research, and Bone & Joint*[360]. The app is available for free, however the full text version requires either a personal subscription or institutional access. When selecting articles the user can either view "abstract" or "full text." In addition, all references can be directly tapped to link directly to the abstract. I found the share feature to be surprisingly functional: the app generates an email that attaches the full-text PDF article. I thought this was a very useful feature.

Journal of Orthopaedic Trauma and Spine
Wolters Kluwer is beginning to offer digital app version of their publications with a current selection that includes the Journal of Orthopaedic Trauma, Spine, and Plastics and Reconstructive Surgery. Each app is the same, and organized into the "library" and "viewer." The journal is presented in entirety, including the table of contents and advertisements. The key feature, however, is the integration of links among the table of contents. Currently, many of the app and journals are offered for free, however a subscription will likely be required in the future.

Acta Orthopaedica
Acta Orthopaedica was the first orthopaedic journal to have developed a fully-functional mobile app. The app does everything one might expect from a journal app, and is even more impressive because the journal is a non-profit, Open Access journal. Thus, all content is completely free. The app allows for access to archives as early as 1930 and allows the user to download and read the full PDF article within seconds. The only missing function that would be useful is the ability to search articles. Of course, this could be done through Google Scholar or Pubmed, but integration in the app would be an obvious enhancement. Among its other useful features includes direct links to Google Scholar to see how many citations an article has, as well as the ability to view "Early Online" articles that may not yet be PubMed indexed.

JBJS Image Quiz
The Journal of Bone and Joint Surgery (American) recently released "JBJS Image Quiz" for both iPad and iPhone platforms. Although this app only provides access to the JBJS bi-monthly "Image Quizzes" that appear in the printed journal, the app is well-designed and easy to navigate. All cases are presented in thumbnail format, and as of writing, the current version includes 54 cases. The question section includes a detailed case history, often 2-3 paragraphs, as well as a number of relevant images which load quickly and clearly on the iPad. Once an answer is submitted, the user is automatically advanced to the "answer" section if correct, and is presented with a brief explanation. Selecting "learn more in the discussion" provides a more thorough explanation of the disease process and relevant clinical pearls. At a price of $0.99, the app is somewhat of an enigma. To those who already subscribe to JBJS, it seems odd that the journal would charge for access to these cases. For those who do not currently have a subscription, $0.99 for unlimited clinical cases and explanations is a phenomenal value. Overall, the convenience of having these cases available in a mobile format is a great benefit and well-worth the small price.

Pediatric Orthopaedic Fellowship Match

Scott J. Luhmann, M.D.

Over the last five years there has been a substantial change in the methodology in the placement of fellowship candidates into pediatric orthopedic fellowships. Five years ago there was no organized match process, multiple fellowship applications, varying interview times and exploding fellowship offers. In response to the applicant frustrations with the process, the Pediatric Orthopaedic Society of North America (POSNA) engaged the San Francisco Match Program (SFMP) to organize and administrate a match process. The match process the upcoming year will be the third year in which the SFMP has run the pediatric orthopaedic fellowship match. In general, the current SFMP match process has been a resounding success, when compared to the pre-SFMP era.

The current fellowship match will begin when the SFMP website goes "live" and began accepting applications on September 1, 2012. This will be for fellowships starting in 2014. Applications will continue to be accepted on the website up to March 31st, 2013. However, the earlier the application is submitted the better, as fellowships will begin evaluating their applicant pool in October and November and then extending interviews to applicants. The interview season will begin November 15th and will extend to March 31st. Interviews are permitted at the International Pediatric Orthopaedic Symposia (IPOS) and the annual meeting of the American Academy of Orthopaedic Surgeons (AAOS). POSNA encourages interviews at these meetings to be preliminary in nature and for applicants to have formal on-site interviews at each fellowship location. However, there may be financial and/or logistical constraints which may prevent an applicant to attend on-site interviews. Hence it is up to the discretion of the fellowship director and the applicant as to the number and location of interview(s). Unlike other orthopaedic subspecialty fellowships, communication between the applicant and fellowship, after the formal interview, is permitted. However **the applicant** must initiate the communication and aim to gain additional information about the fellowship. Fellowship programs are not permitted to contact the applicant after the formal interview. The reason post-interview communication is limited is to protect the applicant from any pressure or coercion from the fellowship.

Applicants must submit their match list onto the SFMP website by Thursday April 11th. The matching process is subsequently carried out and applicants will be notified of their matched fellowship position on Thursday April 18th. On April 22nd the SFMP will post a list on their website any fellowship positions which were not filled. At this time the unmatched applicant can contact one of these fellowship programs to inquire about a position.

Information about pediatric orthopaedic fellowships can be located on the POSNA website www.posna.org and the SFMP website www.sfmatch.org.

IPOS 2012 is just around the corner - November 28 - December 1, 2012, at the Loews Royal Pacific Resort at Universal Studios, Orlando.

Once again, expect an all star faculty with numerous concurrent hands on breakout sessions and main sessions including topics such as:

- Does this hip need an operation?
- Limb deformity and reconstruction.
- Idiopathic Scoliosis: State of the Art.
- On-call challenges: injuries and infections.
- Office orthopedics and practice management.
- Managing upper extremity injuries.
- Surgical solutions to improve function in children with CP.
- Hot topics in sports medicine with a spotlight on OCD.
- Complex spine with a spotlight on early onset and neuromuscular scoliosis.
- A special combined session with IOFAS on foot and ankle disorders and treatment.

We just launched a new IPOS website http://www.posna.org/ipos/ipos.asp and are planning on offering a range of new electronic functionality during the course including use of e-moderators, blog platforms and surgical skills simulation sessions.

We hope to see you all there!

Jack Flynn and Michael Vitale