Join us for our

14th Annual
Arthrogryposis Multiplex Congenita
Support, Inc. Conference
Norfolk, Virginia
July 3-6, 2019

We hope that you can join us for the Annual AMCSI Conference which provides an opportunity for people affected by arthrogryposis multiplex congenita to gather and exchange ideas and support. During the three-day event, families and medical professionals participate in a variety of sessions and children’s activities providing different kinds of information to meet the needs of our ever-growing AMC family. Our planning committee is reaching out to medical professionals and speakers from around the country. Would you be interested in being a speaker?

Arthrogryposis Multiplex Congenita Support, Inc., started as an online support forum; therefore, we believe that these support sessions are just as important as those that offer medical information. Watch for our notices on Facebook and our website in early spring.

For more information visit us at amcsupport.org

Hilton Philadelphia at Penn’s Landing
Philadelphia, Pennsylvania, USA
September 24–26, 2018

3rd international symposium
on arthrogryposis

Jointly Provided by: The International Study Group on Arthrogryposis and Lewis Katz School of Medicine at Temple University

ABSTRACTS

ENDORSED BY
About the Symposium

The 3rd International Arthrogryposis Symposium is presented by The International Study Group on Arthrogryposis (ISGA) in joint providership with the Lewis Katz School of Medicine at Temple University, and in association with the Arthrogryposis Multiplex Congenita Support Inc (AMCSI), and is endorsed by the Pediatric Orthopaedic Society of North America (POSNA) and the European Pediatric Orthopaedic Society. The purpose of our meeting will be both scientific, and for networking. From the scientific standpoint, the meeting will be primarily podium presentations of submitted abstracts. There was a general call for abstracts, which were reviewed by panels of international experts, and selections were made on the basis of relevancy and advancement of knowledge on arthrogryposis multiplex congenita (AMC). As for networking, we hope to encourage both informal as well as formal interactions, to foster research collaborations. A number of groups interested in developing multicenter or multicountry studies will present their proposals to the audience, in hopes of generating interest as well as research partners.

Continuing medical educational credit will be provided by Temple University, and will be underwritten by a generous grant from the Shriners Hospitals for Children.

TARGET AUDIENCE

The intended audience of this international conference is all practitioners who work with patients with arthrogryposis multiplex congenita including physicians (geneticists, orthopaedists/plastic surgeons/hand surgeons, neurologists), physical and occupational therapists, and social workers, as well as researchers in the field of congenital contractures and arthrogryposis.

EDUCATIONAL OBJECTIVES

At the end of this meeting, participants should be able to:
1) Advance the treatment of children and adults with AMC
2) Explain the advances of newer research, procedures and treatments that benefit people with AMC
3) Advocate for the appropriate treatment of children born with AMC, by educating physicians and care givers of the potential of these children
4) Describe the value of interdisciplinary care for these patients
5) Foster an international community of physicians, care givers, researchers to further those goals
ACCREDITATION STATEMENT

Lewis Katz School of Medicine at Temple University is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide Continuing Medical Education for physicians.

CERTIFICATION STATEMENT

Lewis Katz School of Medicine at Temple University designates this live activity for a maximum of 19.5 AMA PRA Category 1 Credit(s)™. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

DISCLOSURE POLICY

It is the policy of the Lewis Katz School of Medicine at Temple University, The Albert J. Finestone, M.D, Office of Continuing Medical Education that the speaker and provider disclose real or apparent conflicts of interest relating to the topics of this educational activity, and also disclose discussions of unlabeled/unapproved uses of drugs or devices during their presentation(s). The Accreditation Council for Continuing Medical Education (ACCME) considers financial relationships to create actual conflicts of interest in CME when individuals have both a financial relationship with a commercial interest and the opportunity to affect the content of CME about the products or services of that commercial interest. The ACCME defines a commercial interest as “any entity producing, marketing, re-selling, or distributing health care goods or services consumed by, or used on, patients.” The ACCME does not consider providers of clinical service directly to patients to be commercial interests. The Lewis Katz School of Medicine at Temple University has established policies in place that will identify and resolve all conflicts of interest prior to this educational activity. Detailed disclosure will be made prior to the activity.
Sponsors

The International Study Group on Arthrogryposis thanks the following entities for their generous support.

Individual and Charitable Organization Support

**Platinum Level**
- AMCSI
- Roger’s Run 4 AMC
- Run for Jack
- Shriners Hospital for Children – Montreal and
- Shriners Hospital for Children – Philadelphia

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ALL faculty, planning committee members, and staff of the Office for Continuing Medical Education involved in the 3rd International Symposium on Arthrogryposis being held September 24–26, 2018 have provided disclosure information.

All faculty, planning committee members, and staff of the Office for Continuing Medical Education have provided disclosure information and indicated they do not have any financial relationships to disclose. All conflicts of interest identified have been resolved utilizing the Lewis Katz School of Medicine at Temple University conflict of interest policies and procedures in adherence with ACCME (Accreditation Council for Continuing Medical Education) guidelines.
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1 – Grant/Research Support from commercial interest (with name of company),
2 – Consultant, paid or unpaid, for a company or supplier,
3 – Speakers Bureau/paid presentation for a company or supplier,
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5 – other financial interests in a company or supplier.
Abstract # 1
Results and Functional Assessment of Long Head Triceps Transfer in Four Children with Arthrogryposis

Bart Kowalczyk, Alicja Fąfara
Arthrogryposis Treatment Centre University Children’s Hospital of Krakow, Krakow, Małopolskie, Poland

Background: Elbow extension contractures and lack of active elbow flexion in children with arthrogryposis significantly restrain their activities. Complex treatment of the upper extremities focus on functional aspects, mainly ability to self-care. In selected children with maintained passive elbow flexion without active flexor activity, long head triceps transfer may provide functional benefits.

Methods: Four children with amyoplasia underwent long head of triceps transfer procedure at our institution using the Ezaki et al. technique. The mean age at surgery was 11 years (range: 7–14). The mean follow up period was 36 months (range: 24–40 months). At follow up orthopedic examination including range of motion measurement with goniometer, assessment of elbow positioning and residual contractures. Functional pre- and post-operative status was calculated and compared using Pediatric Outcome Data Collection (PODCI) and WeeFIM.

Results: All four children presented with satisfactory elbow flexion of at least 90° (range: 90°–130°), all gained hand to mouth activity. In two elbows flexion contractures of 20° and 40° occurred. Three children actively incorporate transfer, with occasional compensatory maneuvers to flex the elbow, one child still uses adaptive mechanisms in activities of daily living. All four children noted improvements in their functional status when compared to the preoperative assessments. The main improvement were noticed in the upper extremity and physical core domains (average improvement – 3 points) and in transfer and basic mobility core scales (mean improvement – 1.5 points).

Conclusions: Despite the series is short we consider long head triceps transfer as beneficial for upper extremity function in selected patients with arthrogryposis. Complications in form of elbow flexion contractures may occur in follow up period without obvious negative impact on the overall function.
Abstract # 2
Free Functional Muscle Transfer for Reanimation of Elbow Flexion in Children with Arthrogryposis

Vinay N. Itte, Robert D. Bains, Grainne Bourke, Simon P.J. Kay
Leeds Teaching Hospital NHS Trust, Leeds, West Yorkshire, UK

Introduction: Elbow flexion is essential for feeding and self care. These activities impart independence and confidence both to the child and their parents. The use of free functional muscle transfer (FFMT) is a safe and reliable method for reanimation of elbow flexion. In the present study, we discuss the surgical technique, post-operative follow-up and surgical outcomes following free functional gracilis muscle transfer used for restoration of elbow flexion in arthrogryposis.

Materials and Methods: We retrospectively reviewed medical notes of ten children (mean age 3.7 years, age range 1 year–10 years). Six bilateral and four unilateral neurotised free gracilis muscle transfers. The elbow function was assessed in terms of activities of daily living and elbow flexor muscle power pre- and post-operatively.

Results: The functional outcomes were assessed regarding the individual ability to manage activities of daily living — self-feeding, brushing teeth and manipulating objects. The outcomes are demonstrated with pre-operative and post-operative videos and photographs. The results also show improvement in active range of motion in elbow flexion and muscle strength.

Conclusion: In comparison to other techniques, free functioning gracilis muscle transfer is a reliable option to restore elbow flexion in children with arthrogryposis provided the gracilis is present. The gracilis is a suitable muscle with reliable pedicle length and nerve segment for neuro synthesis with the recipient’s nerves. This technique imports new muscle to the upper limb and is a valuable option in these children. We would propose it is undertaken in centres with microsurgical and rehabilitation expertise in children.
Abstract # 3
Our Approach for Reconstruction of Active Elbow Flexion in Children with Amyoplasia

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Introduction: The choice of donor area for children with amyoplasia for reconstruction of elbow active flexion is extremely limited. Latissimus dorsi, pectoralis major and triceps brachii are the most frequently used muscles for this purpose. The goal of this report is to determine the optimal donor area for reconstruction of elbow flexion in children with amyoplasia.

Materials: From 2008 to 2018 we performed reconstruction of elbow flexion in 115 children with amyoplasia by transposition of 157 muscles in different variants: pectoralis major (PM) to biceps brachii (61 cases), latissimus dorsi (LD) to biceps brachii (40), pectoralis major et minor to biceps brachii (11), long head of triceps (LHTB) to biceps brachii (31), long head of triceps (LHTB) et pectoralis major to biceps brachii (2 cases), triceps (TB) to biceps brachii (12). The age of patients was from 10 months to 17 years.

Results: (From 6 months to 7 years after operation) We compared the outcome results of transposition of different muscles to biceps brachii and found out that the best results had the patients with latissimus dorsi muscle transposition, the worst results were after triceps transfer (severe elbow flexion contracture). The patients with C6, C6–C7 spinal cord disorders had the best structure of muscles about shoulder (LD, PM, LHTB, TB). The worst structure of muscles had the children with the level of damage of the spinal cord C5–T1. When we choose donor muscle for reconstruction of elbow flexion in patients with amyoplasia, we use the following principles:
1) We estimate the level of spinal cord injury and choose the optimal muscle (6- LD, 6–7- LD or PM, C5–C7 - LD or PM, C5–Th1 - PM or LHTB).
2) A CT scan is done of the thorax, and shoulder. This method allows to exam soft tissue and bone deformities in one procedure (in different regimes). The more so it chipper and more easy to perform in children than MTI. We can indentified muscles in zone of interest, there structure and size.
3) Intra-operative testing (muscle stimulation). If we see good muscle tissue and a good muscle contraction, this is an ideal prognostic sign.
4) If donor muscle is only a grade 2–3, we usually use two muscles.

Conclusion: Preoperative examination of patients with amyoplasia, identification of the level of the spinal cord disorders helps to choose the optimal variant of treatment and restore daily activity of children with this pathology.
Abstract # 4
Functional Improvement After Posterior Elbow Release in Arthrogrypotic Children

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Background: Upper limb deformities and contractures in patients with arthrogryposis restrain their activities of daily living i.e. loss of elbow passive flexion makes impossible self-feeding and self-care. Posterior elbow release allows the patient new functional activities to become more independent in daily living. The purpose of the study was to analyze improvements in self-care independance after posterior elbow releases in eleven children with arthrogryposis.

Methods: 11 patients aged 4.1 years on average at the time of surgery (range 1–12 years) underwent posterior elbow releases in form of posterior capsulotomy and triceps V-Y lengthening due to extension contractures. All underwent uniform post-op physical therapy and orthotic treatments at our institution. Follow up period was 3.5 years on average (minimal 12 months). The final examination included assessment of the elbow passive range of motion, presence of adaptive maneuvers, functional outcomes and patient/parents satisfaction using Pediatric Outcome Data Collection (PODCI) and International Classification of Functioning (ICF) in carrying, moving and handling objects; hand and arm use and self-care.

Results: 9 children achieved passive elbow flexion of at least 90°. One child required repeated release to increase functional flexion. The remaining child achieved 70° of flexion which was graded as unsatisfactory. In three children 10°–40° of flexion contracture occurred. All children were able to reach the mouth using passive assistance. Functional assessment revealed improvements in upper limbs PODCI and ICF domains in 9 children, especially in using fork and knife and possibility to put on a coat.

Conclusion: Posterior elbow capsular release with triceps lengthening improved the passive arc of motion allowing the hand to reach the mouth for independent feeding and self-care. Improvements were noted in upper limb PODCI and ICF domains.
Abstract # 5
Elbow Release and Tricepsplasty in Arthrogrypotic Patients—A Long Term Follow Up Study

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Background: Obtaining elbow flexion to improve hand to mouth reach capability is an essential component of achieving functional independence in pediatric patients with arthrogryposis. We analyzed the long-term outcomes of elbow release and tricepsplasty in a series of arthrogrypotic children at our institution.

Methods: In an IRB approved retrospective study, medical records of patients with arthrogryposis who underwent elbow release and tricepsplasty at our institute from 1993 to 2015, with at least 2 years of follow-up, were reviewed. Collected measures included pre-operative (pre-op) elbow passive range of motion (ROM), post-operative (post-op) elbow passive and active ROM, shoulder passive and active ROM and pediatric outcomes data collection instrument (PODCI) scores. Paired analysis was used to compare pre and post-op final follow-up of elbow passive ROM and student's t-test was used to compare patients' PODCI scores against age adjusted norm.

Results: Forty-eight patients with arthrogryposis undergoing elbow release and tricepsplasty were identified. Seventeen patients meeting inclusion criteria were successfully reached with average age at final follow-up 11 years of age (4–20 years) and average duration of follow-up at 8.4 years (2–18.2 years). Statistical significance was found in passive elbow flexion (pre-op 42.1° (10°-85°) vs. post-op 100.6° (65°-135°)), passive arc of motion (pre-op 39.2° (10° to 80°) vs. post-op 62.6° (25° to 112°)) and average flexion contracture (pre-op 2.9° (-10° to 30°) vs. post-op 39.3° (0° to 80°)). As would be expected, most parent and self-reported PODCI scores in our patients were significantly less than the age adjusted normal population, however, in domains of Comfort/Pain and Happiness, our arthrogryposis patient scores were not significantly different.

Conclusion: Long-term follow-up of elbow release and tricepsplasty in patients with arthrogryposis reveals both increase and sustainability of elbow flexion and arc of motion, indicating improved ability of hand to mouth reach and maximum independence promotion. Although PODCI scores were significantly lower when compared to the age adjusted norm, Arthrogrypotic patients were just as happy and had no more discomfort than unaffected norms.
Abstract # 6
Outcome of Thumb in Palm Surgery in Children with Arthrogryposis

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Introduction: Thumb in palm or abducted thumb is a common problem in patients with Arthrogryposis. Due to the involvement of all structures on the palmar and dorsal side surgical correction is difficult and not rarely can result in some remaining deformity that needs correction. Primary surgery can include skin and soft tissue release within the thumb web transferring a flap from the radial aspect of the index finger with or without additional full thickness skin grafts. This may involve detachment of adductor pollicis distally as well as division of the fascia over 1st dorsal interosseus down to the CMC joint, ‘Z’ lengthening of FPL at the wrist, EPL plication and volar plate release and/or arthrodesis or chondrodesis of the 1st MCP joint.

Method: A retrospective review collected data of all patients who have undergone thumb surgery for Arthrogryposis from 2000 to 2018. Data collected includes type of surgery, number of surgery and outcome.

Results: We will report on the Birmingham experience on correcting thumb in palm deformity in children with Arthrogryposis, the amount of residual deformity and secondary surgery.

Discussion: Although the type of surgery including release and replacement of palmar skin, flexor lengthening and extensor tightening is well described, our data shows that resulting deformities will not rarely require consecutive surgery.
Abstract # 7
Does Open Reduction of Arthrogryptic Hips Cause Stiffness?

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Purpose: Congenital hip dislocation in arthrogryposis multiplex congenita occurs in 15–30% of patients. Despite good reported outcomes after open reduction of arthrogrypotic hips, many still feel that treating these hips risks iatrogenic hip stiffness. We believe that arthrogrypotic hips have pre-existing limitations of motion which are only mildly affected by relocation. This retrospective study compares pre and postoperative hip ranges and total arcs of motions, and evaluates ambulatory abilities of patients having undergone open reductions of arthrogrypotic hips.

Methods: Since 2008, 40 consecutive patients with arthrogryposis underwent 58 open reductions of congenitally dislocated hips (22 bilateral), with 2 years’ minimum followup. Average age at surgery was 22 months, followup averaged 47 months. Open reductions were performed through a medial approach, with femoral shortening osteotomies as needed for reduction stability. The ligamentum teres was used to tether the femoral head into the acetabulum. Hip motions were recorded pre-operatively, at hardware removal, and at latest followup, as was ambulatory ability.

Results: Flexion contractures were >20° in 25 hips preoperatively (average 33°), improving an average of 24° at followup; 51 hips had <45° frogleg abduction (average 26°), improved an average 14°; 35 hips had <30° abduction preoperatively (average 20°) which improved an average 10°, all p-values <0.001. Hips that had <90° of flexion showed no significant change from pre-operative to followup. Flexion-extension total arc of motion (TAM) for all hips decreased only 4° from pre-operative to followup (p = 0.09), and the internal-external rotational TAM decreased 22° (p = 0.004). Comparing the 22 unilateral dislocated hips to their opposite side, the various TAMs were not statistically distinguishable at followup. Of the 40 patients, 26 were independently ambulatory at followup, most with braces, and 8 were progressing but still walker dependent. Six were non ambulatory at followup.

Conclusion: Open reduction of congenitally dislocated hips in children with arthrogryposis does not lead to stiffness. The pre-existing hip motion limitations were slightly worsened, whereas the lower limb positioning was improved and more appropriate for ambulation. Specifically, hip extension and abduction were improved, flexion and adduction mildly decreased. The majority become independent ambulators.
Abstract # 8
Reorientational Proximal Femoral Osteotomies for Arthrogrypotic Hip Contractures

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**Purpose:** Hip involvement in arthrogryposis multiplex congenita occurs in 50–85% of patients. Severe hip contractures are multi planar, typically flexion/abduction/external rotational, precluding or complicating ambulation. The reorientational osteotomy at the intertrochanteric level aligns the femoral shaft with the body axis, leaving the hip joint itself in its natural position. Pre-operative hip motion is preserved but placed it in a more functional range for sitting and ambulation. This retrospective study compares pre and postoperative hip ranges and total arcs of motions, and evaluates ambulatory abilities of patients having undergone the reorientational osteotomy.

**Methods:** Since 2008, 65 patients with arthrogryposis had 117 reorientational proximal femoral osteotomies (52 bilateral), with a minimum 2 years’ followup. Age at surgery ranged from 13 months to 12 years (average 48 months). An intertrochanteric wedge osteotomy was performed to align the lower extremity appropriately with the body axis. Hip motions were recorded pre operatively, at hardware removal, and at latest followup, as was ambulatory ability.

**Results:** Flexion contractures >20° in 81 hips preoperatively (average 52°) improved an average 35°; 84 hips had <15° adduction (average -20°), improved an average 42°; 101 hips had <30° internal rotation (average -16°) which improved an average 35°, all p-values <0.0001. Flexion-extension total arc of motion (TAM) for the 117 hips improved 14°, and the abduction-adduction TAM improved 9° (p <0.0001 for both), all other TAM axes were unchanged. Of the 65 patients, 36 were independently ambulatory at followup, most with braces, and 20 were walker dependent. Of the 9 non ambulatory patients, 5 had the procedure done specifically to improve seating.

**Conclusion:** Hip contractures are the main lower limb deformity that prevents efficient ambulation in children with arthrogryposis. The reorientational osteotomy described allows the lower limb to be positioned appropriately for ambulation, altering the range of motion but not the TAM. Most of the walker dependent children had less than 3 years followup; from our experience many of them are expected to become independent over time as well, especially once their knee flexion contractures are corrected.
Abstract # 9  
**A Long-term Outcome After Surgeries for Lower Limbs in Children with AMC**

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**Purpose:** To evaluate the long-term outcome after surgeries for lower limbs in children with AMC who were followed-up until the skeletal maturity.

**Methods:** 12 amyoplasia patients (6 males and 6 females) with a mean age of 23.7 years (15–41) at the final follow-up were investigated on a walking ability based on the Hoffer’s classification, usage of lower limb orthosis, the number of surgeries, a range of motion (ROM) of the hip, knee and ankle joints, and an occupation.

**Results:** 11 patients were community ambulators and one was a household ambulator. The total numbers of surgeries for lower limbs were 93, which meant an average 7.75 each patient. Thirteen surgeries for 11 hips in 8 patients, twenty surgeries for 15 knees in 12 patients, and fifty surgeries in 24 feet in 12 patients were performed. Hip dislocation was treated by an open reduction with femoral and/or pelvic osteotomies. Recurrent hip subluxation was treated by femoral and pelvic osteotomies for 2 hips. Quadricepsplasty was performed for 7 knees in 5 patients with knee extension contracture, and posterior release was performed for 7 knees in 5 patients with knee flexion contracture. Open reduction was performed for one knee with patella dislocation. Recurrent knee deformities were treated by soft tissue release in 4 knees, and femoral and tibia osteotomy in one knee. Foot deformities were corrected by soft tissue release for 12 feet in 10 patients and takedown for 4 feet in 2 patients. Secondary surgery consisted of calcaneocuboid fusion in 6 feet, mid-tarsal osteotomy in 6 feet, takedown in 5 feet, and soft tissue release in 2 feet, ankle osteotomy in one foot, and calcaneus osteotomy in one foot, respectively. Femoral osteotomy was performed for lower limb realignment in 10 limbs. The mean ROM were 67.5° flexion, -2.9° extension in the hip joint; 73.9° flexion, -12.1° extension in the knee joint; -7.7° dorsal flexion, 12.1° planter flexion in the ankle joint. Four patients did not use orthoses; eight needed lower limb orthoses. Five patients worked in offices, four were students and three were unemployed.

**Conclusion:** AMC patients maintained a walking ability at the final follow-up after several surgeries for lower limbs. The ankle joints were treated more frequently and finally demonstrated a more severe ROM limitation compared to the knee or hip joints. We should decrease the number of surgeries for foot deformities.
Abstract #10
Pirani Scores Better Demonstrate Clinical Change in Idiopathic and Non-idiopathic Clubfoot

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Purpose: We have compiled an eight year retrospective database of all clinical measurements in the treatment of clubfoot with a high proportion of the patients having an arthrogryposis diagnosis. As a pilot inquiry we compared the results of Ponseti-like management between non-idiopathic and idiopathic cohorts.

Methods: Dimeglio and Pirani scores were obtained and recorded at multiple visits for patients presenting with clubfoot at a single institution. These scores were recorded along with demographic data and other clinical information including procedures performed such as achilles tenotomy and number of casts applied. Comparisons were made between total Dimeglio and Pirani scores, age, number of casts, and follow up time.

Results: Data from 63 patients were analyzed. 14 idiopathic and 49 non-idiopathic with 223 idiopathic and 892 non-idiopathic measurements made separately for left and right sides during clinic visits. The groups were similar in follow up time, number of visits and number of casts applied. There was a significant difference in age, with idiopathic somewhat younger at initial visit (32 ± 53 vs 44 ± 39 months, p=0.05). Dimeglio scores were found to be lower in the idiopathic (mean 6.2 R, 6.4L) versus non-idiopathic (8.6R, 8.7L) and were affected by number of casts applied and gender but not age. Pirani scores were significantly lower in idiopathic (mean 2.0R, 1.8L) compared to non-idiopathic (mean 2.7R, 2.8L) based on 239 idiopathic and 978 non-idiopathic measurements. Pirani score scatter showed improvement over time in idiopathic patients with only a slight trend towards improvement in non-idiopathic patients over time up to 8 years.

Conclusion: Preliminary data illustrates higher severity as measured by total Dimeglio and Pirani scores for non-idiopathic versus idiopathic clubfoot with long term follow-up. Total Pirani scores appear to be more strongly associated with clinically improvement over time compared to Dimeglio scores.
Abstract # 11
Orthopaedic Scientific Literature Has Poor Uniformity in Defining Clubfoot Relapse

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**Purpose:** Congenital clubfoot (talipes equinovarus) treatment is largely successful by Ponseti technique. Recurrence is unfortunately a problem, commonly due to non-compliance with bracing. It remains a struggle to determine when and how to intervene for optimal correction and clinical improvement. Lack of uniformity in describing recurrence may make interpretation of scientific evidence about efficacy of intervention for relapsed clubfoot difficult.

**Methods:** Pubmed inquiry found 334 published articles from 1960–2017 including the term “clubfoot relapse” in English language. Exclusion criteria included articles that did not primarily include data on idiopathic clubfoot relapse, were case studies or reviews, or inaccessible online. Articles were identified and analyzed by two contributors for language to define relapse, clinical outcome measures, and results.

**Results:** 60 original research articles were identified. Eleven were published by Journal of Pediatric Orthopaedics(JPO), 7 by Clinical Orthopaedics and Related Research, 7 by Journal of Bone and Joint Surgery(British), and 4 by JPO B. Twenty-one (35%) contained no definition, no specific definition for relapse in the inclusion criteria or elsewhere. The most common definition for relapse were described as “requiring further treatment” (8 articles constituting 13% of the examined articles), any cavus, adductus, varus, or equinus (CAVE) deformity component recurrence (8;13%), a particular CAVE component recurrence (6;10%), a previously published clinical scale (as in Dimeglio, Pirani) or other (5;8%). Less than 50% used a reproducible clinical measurement to define relapse. 63% of studies analyzed results of a surgical intervention.

**Conclusion:** Scientific literature on relapses of clubfoot lacks uniformity in defining relapse. In the past 57 years 35% of articles on this subject contain no definition at all. Significance: It has not established what degree of relapse is clinically significant or which intervention is most appropriate and effective. A consensus agreement or prognostic classification system may help guide practicing orthopaedic surgeons.
Abstract # 12
Effectiveness of the Ponseti Method in Treating Foot Deformities in Children with Arthrogryposis

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Introduction: Arthrogryposis is characterized by joint contractures with foot deformities, of which clubfoot is the most common. Studies that investigate the effectiveness of the Ponseti method to treat children with arthrogryposis are few. This study evaluates the Ponseti method in children with arthrogryposis using passive range of motion (PROM), parent reported outcomes, and dynamic foot pressure. Significance: Clubfeet in children with arthrogryposis are typically stiffer than idiopathic clubfeet, but improved outcomes with conservative treatment can offer clinical benefit to this population.

Methods: Children treated with the Ponseti method were evaluated retrospectively pre, post/short term (ST; ≥ 6 months), and post/longer term (LT; 6 months – 1 year) serial casting. Outcome measures include PROM, pedobarograph, and the Pediatric Outcomes Data Collection Instrument (PODCI; age 2 years+). Children were grouped into younger (< 1 year) and older (> 1 year). With the Ponseti method, the equinovarus foot is serially casted; a percutaneous tenotomy of the Achilles tendon is done before the final cast is applied to the plantigrade foot.

Results: 31 children were included (n=78 feet; avg. age 4.8±4.0 years) and stratified into two groups: younger (n=19 feet; avg. age 7.9±3.7 months) and older (n=59 feet; avg. age 6.5±3.5 years). PROM was collected 1.5±3.4 months prior to casting (n=78 feet), 1.1±2.5 months (ST; n=78), and 9.0±2.9 months after casting (LT; n=22). Foot pressure and PODCI results were collected before and (4.6±3.9 months) after casting (n=12 children). Serial casting was repeated weekly for 2 - 9 weeks depending on progress. PROM improved in ankle dorsiflexion (DF) and forefoot abduction (FFABD) (p< 0.0001) in ST, but only ankle DF improved in the LT (p = 0.0002). Categorization by age revealed ankle DF (p < 0.05) and FFABD (p< 0.01) improved at ST in both younger and older age groups. Foot pressure had reduced abnormal lateral forefoot (p=0.02) impulse post casting. PODCI data improved in sports/physical functioning (p=0.07) and happiness scores post serial casting (p=0.04).

Conclusions: Serial casting in children with arthrogryposis is effective in improving PROM, dynamic foot pressure pattern and parent reported outcome in the short term.
Abstract # 13
Patient Reported Outcomes Measurement Information System (PROMIS) Scores for Children with AMC

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Introduction: PROMIS tools are standardized, validated patient-reported health measures for people with chronic conditions. We sought to determine whether they would be useful for clinicians caring for children with arthrogryposis multiplex congenita (AMC).

Materials and Methods: PROMIS Mobility, Upper Extremity (UE) Function, Pain, and Peer Relationships measures were administered to children with AMC who presented to Hand Clinic, as part of an initiative to gauge the health-related well-being of children with chronic musculoskeletal diseases. Responses were converted to a T-score which rescales the raw score to a mean of 50 and a standard deviation of 10. For Mobility, UE Function, and Peer Relationships, a T-score ≥ 50 is within normal limits; 40–49 indicates mild impairment; 30–39 indicates moderate impairment, and 0–29 indicates severe impairment. For Pain Interference, a T-score ≤ 49 is within normal limits; 50–59 indicates mild impairment; 60–69 indicates moderate impairment; and 70–78 indicates severe impairment.

Results: 21 children (age 5–17 years) with AMC were studied (April–December 2017). 19 children completed the Mobility measure (2 used wheelchairs for mobility). Their mean score was 39 (range 23–59, S.D.11). 37% (7 children) reported normal function or mild impairment, and 63% (12) reported moderate or severe impairment. All 21 children completed the remaining measures. The mean UE Function score was 30 (range 13–57, S.D. 12). 20% (4) reported normal function or mild impairment, and 80% (17 children) reported moderate or severe impairment. The mean Pain Interference score was 50 (range 34–68, S.D. 12). 61% (13) reported no or mild pain interference with activities; 39% (8) reported that pain interfered with their activities to a moderate or severe extent. The mean Peer Relationship score was 58 (range 43–62, S.D. 7); 95% (20) reported normal function and 5% (1) reported mild impairment.

Conclusion: PROMIS discerns impairment in children with AMC presenting to Hand Clinic. The majority had moderate or severe impairment of mobility and upper extremity function, mild pain interference with activities, and normal peer relationships. These findings indicate that PROMIS scores can be used over time and pre- and post-interventions, to better understand and treat the challenges that children with AMC face regarding mobility, upper extremity function, and pain.
Abstract # 14  
Patient Reported Outcomes in Arthrogryposis

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Introduction:  Patient reported outcomes help to understand patient status and optimize care, but little is known about common measures in the arthrogryposis population. Utilizing Patient-Reported Outcome Measurement Information System (PROMIS) and Pediatric Outcomes Data Collection Instrument (PODCI) questionnaires, we investigated functional and psychosocial measures in arthrogryposis.

Methods:  98 patients with arthrogryposis were identified from a prospective cohort (CoULD) from 2014 to 2018. Demographics and patient reported outcome measures were collected, including the PROMIS (upper extremity (UE) function, pain, depression, anxiety, and peer relations) and PODCI questionnaires (UE function, pain, happiness, and global function).

Results:  29 patients had complete PROMIS and PODCI data. This cohort was divided into amyoplasia and distal arthrogryposis, with 15 and 14 patients in each group respectively. There were 7 females in the amyoplasia group with an average age of 11 years; 8 females in the distal group, average age of 12 years. For both cohorts, the UE function PROMIS scores were well below population norms, at 22 for amyoplasia and 32 for distal arthrogryposis. PODCI UE function was lower for amyoplasia (42 with SD 24) than for the distal cohort (75 with SD 17). PROMIS pain, depression, anxiety, and peer relations were in the normal range for both amyoplasia and distal arthrogryposis. PODCI pain and happiness ranged from 77–84. Comparing PROMIS and PODCI in this population, there was almost perfect correlation between UE function scores for both groups, amyoplasia r=0.8 and distal r=0.9. Pain scores were moderately correlated between the questionnaires and the depression score for PROMIS was substantially, inversely, correlated with happiness for PODCI.

Conclusion:  Arthrogryposis patients have lower UE functional scores than the population norm, but they have emotional states that are consistent with normal values. Additionally, the PROMIS scores were found to correlate well with the PODCI questionnaire for UE functional assessment and moderate or better for mental health states.
Abstract # 15
Participation Among Children and Youth with Arthrogryposis Multiplex Congenita: A Scoping Review

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Background and Rationale: Individuals with AMC display variable clinical features, which may include contractures of varying severity to the upper and lower extremities, the spine and the jaw and may impact and limit independence in mobility, self-care, activities of daily living, and participation. Results from a qualitative study on 27 youth with AMC, caregivers and clinicians highlighted the importance of participation, especially from the youth’s perspective. Studies have shown that participation is reduced in terms of frequency, community engagement, and meaningfulness for children with congenital conditions. The objective of this scoping review was to identify what is known surrounding participation among children with AMC using the WHO’s International Classification of Functioning, Disability and Health (ICF) as a framework and inform treatment intervention. Specifically, the following questions were addressed: 1) What participation areas are children with AMC engaged in? 2) What are the outcome measures used to evaluate participation? 3) What are the interventions used to promote participation? 4) What are the facilitators and barriers to participation in this population using the ICF as a framework?

Methods: A scoping review using the Arksey and O'Malley framework was used to systematically select and summarize existing literature. Searches were conducted using a combination of keywords in the databases MEDLINE, CINAHL, EMBASE, PsycINFO, and OTSeeker as well as the grey literature to search for theses, dissertations and publicly available information using ProQuest, OpenSigle, and YouTube videos for English or French reports of participation in children with AMC. Two team members reviewed the abstracts, full texts and online videos, relevant information was extracted into a data extraction sheet. Methodological quality was reported using the McMaster Appraisal Checklist for quantitative and qualitative studies. Online videos were appraised using the CRAAP Evaluation Test.

Results: The number and details of included studies will be presented. This is a proposal for a participation rating tool development.

Conclusion: Expected contributions on the importance of participation among children and youth with AMC, measurement and intervention will be discussed.
Abstract # 16

Using the Oswestry Disability Index for Pain and Disability in Arthrogryposis Multiplex Congenita

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Introduction: Chronic pain and disability is often experienced by adults with arthrogryposis multiplex congenita (AMC), a rare condition involving multiple joint contractures present from birth. However, there are currently no pain or disability questionnaires available specifically for individuals with AMC and no impact analysis on quality of life has been performed. Therefore, the purpose of this study was to validate an existing disability tool, the Oswestry Disability Index (ODI), as a pain and disability outcome measure for the AMC population. The ODI was originally designed to measure disability from low back pain and assess limitations in lower extremity activities of daily living.

Methods: This study used a mixed methods approach to investigate pain and disability of individuals with AMC and to determine the content and construct validity of the ODI as an outcome measure for this population. Fifty adults with AMC recruited from an international arthrogryposis study via email participated in this pain study. Participants completed five pain and disability questionnaires and a 30-minute open-ended semi-structured interview. Content validity of the ODI was assessed by the percentage of participants who responded to open-ended questioning with domains already included in the ODI. Construct validity was assessed by correlating the results of the ODI against the results of the other three pain and disability measures using Pearson’s correlation coefficients (r-values) and R2 values.

Results: The majority (67%) of activities of daily living, such as walking, standing, personal care, sitting, lifting and sleeping, that were already included in the ODI were independently identified by participants. All three measures produced R2 values greater than 0.25, demonstrating the strength of construct validity of the ODI in individuals with AMC.

Conclusion: This study is the first to validate an existing disability tool, the ODI, as a pain and disability assessment tool for the AMC population. We conclude that the ODI has yielded sufficient content and construct validity to assess low back and lower extremity related disability in the AMC population. However, there were several activities identified in the interview that were not addressed in the ODI, most involving the use of upper extremities – which is a limitation of the ODI in adequately assessing all pain experienced by individuals with AMC.
Abstract # 17
Long-term Follow Up on Quality of Life and Function of Adults with Arthrogryposis

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Introduction: Arthrogryposis refers to a group of 300+ different disorders characterized by multiple congenital joint contractures and stiffness affecting the upper and/or lower extremities and trunk. Although there is an abundance of literature describing clinical characteristics of children with arthrogryposis, there are few long-term studies evaluating function and quality of life (QOL) among adults with arthrogryposis. The purpose of the current study was to describe long-term functional and QOL characteristics of individuals with arthrogryposis following transition into adulthood using standardized measures.

Materials and Methods: 23 adults with a primary diagnosis of arthrogryposis followed at a single pediatric orthopedic hospital as a child/adolescent (average age 23y6m +/- 4y; 9M, 14F) were included. Participants completed questionnaires related to demographics, mobility, activities of daily living, and QOL. The Patient Reported Outcomes Measure Information System (PROMIS®-57 Profile v. 2.0 and the Global Health Scale) and the Satisfaction with Life Scale outcome tools were completed to evaluate QOL and life satisfaction.

Results: General health was reported as good to excellent for 83% of participants. 30% of participants lived independently, 69% were community ambulators, and 57% were employed. Participants reported lower physical function than the general US population. Scores representing anxiety, depression, fatigue, participation, pain interference and sleep disturbance were consistent with the general US population. The average pain intensity was 2.6 on a scale of 0 to 10, with pain most frequently reported in the legs and feet. 56% were satisfied to extremely satisfied with their life. Five individuals who were dissatisfied with their current life also reported lower physical function, scores indicating anxiety, depression and fatigue, as well as pain in multiple joints.

Conclusions: Although most young adults with arthrogryposis presented with pain and limitations in physical function; overall, they reported good QOL. Dissatisfaction with life was associated with poor physical function, psychological well-being and pain. Findings from the current study will help clinicians anticipate the needs of individuals with arthrogryposis as they transition from pediatric to adult care.
Abstract # 18
Compensatory Strategies for Performing Activities of Daily Living in Adults with Amyoplasia

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Introduction: Knowledge about how adults with amyoplasia adapt in managing Activities of Daily Living (ADL) is sparse. Increased knowledge is necessary to better understand the challenges persons with this diagnosis have to face, and to support function-enhancement. A cross-sectional study on adults with amyoplasia was conducted in order to gain knowledge about physical function and coping strategies in daily activities. Other findings from the study were presented at the 2nd International Symposium on Arthrogryposis and were published in 2017. The aim of this part of the study was to explore and describe compensatory strategies used by the participants when having reduced active movements of the joints.

Materials and methods: 22 adults, aged 20 – 91, participated in the study. Ranges of motion and muscle strength were measured. The ability to perform ADL was mapped by Functional Independent Measure (FIM), which is a commonly used measure and estimates the level of assistance needed to complete basic daily activities. Compensatory strategies and adaptations used for ADL were described and documented by photographs.

Results: Independence in ADL was related to antigravity muscle strength and/or a high degree of range of motion. With muscle strength greatly reduced, passively movements of the joints showed to be important for managing daily activities independently, by using compensatory strategies. Use of compensatory strategies was common, especially in eating, grooming and dressing. The strategies were often based on supporting the arm to the surroundings, as a table or a doorframe, and then move the body in order to bend the joint passively. Only simple assistive devices and small adjustments in the surroundings were used to manage the activities, but the electric wheelchairs were usually individually adapted. During the presentation, the compensatory strategies will be documented by photos.

Conclusions: The physical limitations imposed by amyoplasia have influence on how these patients manage in daily life. When the ability to move actively was limited, a high degree of passive joint motion was crucial for independence in daily activities, by using creative compensatory strategies. Knowledge about these strategies is of importance, for both the patients and the professionals, in order to promote functional enhancement.
Abstract # 19
Muscle MRI in Amyoplasia: Diagnostic Biomarker and Further Evidence for a Motoneuron Disease

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Objectives: Amyoplasia is the most common form of Arthrogryposis multiplex congenita (AMC). However, information about the pattern of muscle involvement in amyoplasia is limited. The aim of this study was to determine if there are specific patterns of muscle involvement in amyoplasia and to find out if there is a fibro-adipose infiltration of the muscles with age.

Methods: Between 2008 and 2017, we examined 65 patients affected by amyoplasia (33 children and 32 adults), at the Centre Hospitalo-Universitaire Grenoble Alpes. 14 patients had atypical clinical findings. MRI was performed with T1-weighted turbo spin echo (T1W-TSE) sequences. We analyzed the fibro-adipose infiltration and “grelot” sign of the muscles of the neck, the shoulder and pelvic girdle, trunk, upper and lower limbs.

Results: Compared with atypical patients, the absence of the biceps brachii, brachialis, tibialis anterior, gracilis, and sartorius was significantly more frequent in typical patients. Mercuri scores were higher in adult patients in tensor fasciae latae, semimembranosus and vastus lateralis muscles. The adductor longus and brevis were selectively preserved.

Conclusions: Our data provide evidence that muscle MRI can identify a specific pattern of muscle involvement in amyoplasia patients and constitutes a diagnostic biomarker with a high positive predictive value. Increasing fibro-adipose infiltration with age was observed in some muscles in a subset of patients. However further evidence is needed to determine if this reflects disease progression. The “grelot” sign gives further arguments for the congenital neurogenic origin of amyoplasia.
Abstract # 20
Arthrogryposis - Genetic Update

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Arthrogryposis (multiple congenital contractures) is a sign with many etiologies. Well over 300 specific disorders have been reported. Various approaches to the differential diagnosis have been developed. The advent of whole genome sequencing has uncovered over 400 specific genes which, when mutated, are associated with decreased fetal movement and the development of multiple congenital contractures. Decreased fetal movement is the common feature. Why does decreased joint movement lead to joint contractures at all ages? There are many, many causes of decreased movement, including limited space, muscle disease, endplate deficiency, neurologic problems both central and peripheral, myelin defects, maternal illness, vascular compromise, etc. Each of these causes have underlying pathways made up of multiple proteins produced by genes, which when intact, enables movement.

Over 400 genes have been identified to have mutations which are associated with forms of arthrogryposis. Gene ontology programs allow the dividing of these genes into 22 functional groups, and subsequently the recognition of metabolic/biochemical pathways that are necessary for normal fetal movement. The recognition of these pathways has already led to new therapeutic approaches.

The functional groups identified by gene ontology programs fall into biologic processes, molecular function, and cellular components. Some genes have multiple functions. For instance, under Biologic Processes, there are genes involved in 18 metabolic processes, 10 in developmental processes, 7 in cellular processes, 6 cell communication, 5 transport, 5 systems, 4 cell activators, 3 cellular component organization, 3 cell cycle, 2 immune system, 2 apoptosis, 2 reproduction, and 1 gene involved in response to stimuli.

Under Protein Classification, there are 5 nucleic binding, 5 hydrolase, 4 phosphatase, 4 transcription factors, 3 extracellular matrix proteins, 3 cytoskeleton proteins, 3 transferases, 3 cell adhesion molecules, 2 ligases, 2 enzyme modulators, 2 defense/immunity proteins, 2 isomerases, 2 receptors, 1 transprotein, 1 calcium-binding protein, 1 transfer/carrier protein, 1 membrane traffic protein, 1 structural protein, 1 kinase, and 1 storage protein.

The value of information technologies to classify genes into pathways is obvious. The challenge is to find ways to convert this knowledge into prevention and therapy.
Abstract # 21
Natural Progression of Orthopedic Deformities in Bruck Syndrome

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Introduction: In 1897, Alfred Bruck described a syndrome characterized by joint contractures and bone fragility, considered as a combination of Arthrogryposis Multiplex Congenita AMC and Osteogenesis Imperfecta. Two types of BS (FKBP10 and PLOD2) are phenotypically indistinguishable. The clinical identification of BS by a combination of joint contractures and bone fragility seems to be problematic. Breslau-Siderious suggested that BS is not a subtype of OI or AMC but rather is a distinct disorder. The aim of this study is to describe the orthopedic characteristics of four patients with BS. We hypothesized that there is a pattern of progression of orthopedic deformities during growth.

Material and Methods: An observational study was conducted of four cases with BS treated in our facility from 2002 to 2017. Data were collected from medical records.

Results: Infancy: joint contractures resembled AMC with more distal involvement. Long bones were normal in shape with mild osteopenia. The feet had severe equinus, adductus (metatarsus) and varus (tarsal and metatarsal bones). The upper extremities presented ulnar deviation of the wrist, hyperextension of metacarpal joints and flexion of phalangeal joints. Early childhood: elongation of bones in the metaphyseal cutback zone, resulting in very thin diaphysis (gracilis shaped bones). There were fractures in the diaphysis (occasionally metaphysis) of long bones of lower extremities with good callus formation. There was mild thoracic kyphosis. Childhood: fractures of the lower and upper extremities. In spine: cervical kyphosis, progression of thoracic kyphosis, beginning of high thoracic scoliosis, and significant lower lumbar lordosis which may progress to include spondylolysis. Late childhood: multiple diaphyseal fractures of long bones of the lower and upper extremities with deformities that required orthopedic instrumentation, including pelvis deformity and possible nonunion. In the spine: progression of cervical kyphosis, thoracic kyphoscoliosis, and lumbar lordosis.

Conclusion: Bruck Syndrome has a pattern of progression of orthopedic deformities during growth: joint contractures, more severe distally. Long bones present initially as “normal” in shape and then become thin, gracilis and osteopenic during growth, predisposing to fractures and deformity. The spine develops progressive cervical kyphosis, thoracic kyphoscoliosis and lumbar lordosis.

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Abstract # 22
Defining AMC: Towards a Consensus on Terminology

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Background and Rationale: Arthrogryposis multiplex congenita (AMC) has been described and defined in thousands of articles, but terminology has been inconsistent. Some have described it as a diagnosis or syndrome, others as a term or clinical finding. Some use the terms arthrogryposis, AMC, and amyoplasia interchangeably, whereas amyoplasia has been defined as a “sub-type” of AMC in one of the existing AMC classifications. This has led to confusion in clinical and research communities as no common language has been set. Objective: This project aims to establish a definition of AMC using international expert opinion to reach a common language and understanding.

Methods: A literature review was conducted and general AMC definitions were extracted from articles. The most commonly used words in the definitions were identified. A group of 8 experts in AMC (researchers, clinicians, and individuals with AMC) identified elements considered to be critically important to the definition. Based on the responses and the literature review, the research team drafted a definition. Final consensus was achieved using a modified Delphi method. A first electronic survey was sent to 46 individuals and completed by 25. The results were extracted and are being analyzed. The revised draft definition will be modified accordingly and a second survey will be sent. We expect to reach consensus on the definition after two rounds of surveys. The final definition will be shared with all participants.

Analysis: Definitions from the literature were combined with the elements identified by the group of experts. Results from the electronic survey were extracted and analyzed qualitatively and quantitatively. The Delphi method will be repeated until 80% agreement on each element of the definition is achieved.

Conclusion: A consensus definition of AMC will help harmonize research and clinical endeavors and should facilitate exchanges among the AMC community (youth, adults, caregivers, support groups), clinicians and researchers. To share and integrate the new definition within the scientific community and the AMC population, a variety of knowledge translation initiatives will be undertaken.
Abstract # 23
An Initial Report on a Grading System for Global Assessment of the Child with Amyoplasia

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Background: Amyoplasia often presents with differences between particular patients, concerning i.e. severity of their contractures, which possibly may impact their prognosis and function. We propose a numeric scale based on clinical examination of spine, upper and lower extremities passive and active range of motion (ROM) for uniform and simple grading system allowing for comparisons between patients, prognosis, and documentation of changes with treatment course.

Methods: 10 passive parameters (passive ROM) were defined and graded 0-1-2-3 depending on the severity. 7 active parameters were based on presence (0 points) or absence (1 point) of selected active movements. All points were added for each patient to assess the severity of contractures at upper and lower limbs and summed up for global assessment. Subdivision into four categories of general severity was proposed as follows: mild 0–10, moderate 11–17, severe 18–24, extreme 25–34. 23 children with amyoplasia were evaluated independently by three investigators (orthopedic surgeons, physiotherapists). Kendall’s coefficient of concordance W was used to assess agreement among the raters. Friedman’s test was performed to detect differences between raters in sums of points for each patient. P-value of 0.05 or less was considered significant.

Results: 1 child with lower extremities involvement was graded as mild amyoplasia, 6 with upper limbs involvement were graded as mild in 2, moderate in 2, severe or extreme in another 2 cases. The remaining 16 with 4-limb involvement were graded as mild in 6, moderate in 6, severe in 4 cases. In 87.5% ratings the differences between the raters were less than 2 points, in 12.5% less than 3 points. W Kendall coefficient for all patients equaled 0.980 and was statistically significant (p<0.001) pointing that the overall concordance between raters was high and statistically significant. Although small differences in ratings were common, they were not statistically significant using Friedmann test (p=0.169).

Conclusions: We conclude that the proposed method of rating is simple and repetitive allowing for global assessment and grading of arthrogrypotic contractures. Further studies will be undertaken to assess the impact of the grading to the prognosis of treatment in amyoplasia.
Abstract # 24
Relationship of Ambulatory Function Based on Infantile Lower Extremity Posture Types in Amyoplasia

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Background: Arthrogryposis multiplex congenita (AMC) is a non-progressive syndrome with multiple rigid joints, fibrotic periarticular tissue, and muscular fibrosis. The most common subgroup is amyoplasia. Ambulation is one of the most significant functions of the lower extremities. There is no predicative scale to determine ambulation at maturity for the infant with amyoplasia. Lower extremity resting position of infants with amyoplasia potentially correlates with ambulation at maturity. The purpose of this study was to classify the infantile position of lower extremities and muscle strength to predict ambulation potential at maturity.

Methods: Children with amyoplasia were classified into groups based on infantile position of hip-knee alignment and limb muscle function. Sitting, standing, and walking skills from infancy into adulthood were evaluated. The ambulation function was correlated with the infantile position of the lower extremities.

Results: Amyoplasia cases (n=66) were sorted into five types and correlated with ambulatory potential. Type I: (n=13) mild ambulatory impairment with infantile position of flexed knees and hips but full range of motion. At maturity, all were community ambulators. Type II: (n=14) moderate ambulatory impairment having infantile position of hip flexion, hip external rotation, and knee flexion contractures. Hip abductors and external rotators had antigravity strength. All stood and walked during the first decade of life with knee ankle foot orthoses. Type III: (n=12) severe ambulatory impairment having infantile position of hip flexion, abduction, external rotation, and knee flexion contractures but lacked hip muscle recruitment. All used wheelchairs at maturity. Type IV: (n=12) mild ambulatory impairment with infantile position of extended knees and flexed dislocated hips. At maturity, 90% were community ambulators. Type V: (n=15) variable ambulatory impairment having asymmetric hip and knee alignment with unilateral hip dysplasia with extended knee and opposite limb flexed. Ambulation skill varied at maturity with 27% full-time wheelchair users.

Conclusions: Amyoplasia can be sorted by infantile position of lower extremities and muscle strength into five types to predict ambulatory function.
Abstract # 25
Disability in Adults with Arthrogryposis is Severe, Partly Invisible, and Varies by Genotype

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Objective: There is a recent interest to understand the handicap of adults with Arthrogryposis Multiplex Congenita (AMC), a rare disease spectrum characterized by at least two joint contractures at birth. This study is the first to describe disability patterns of a cohort of adults with AMC, in relation with their genotypes.

Methods: Retrospective analysis of the data of 43 unselected persons with AMC referred to the French Center for adults with AMC from 2010 to 2016. All patients underwent a pluri-professional systematic and comprehensive investigation of the deficits, the activity limitation, and the participation restriction (ICF), and genetic analysis when indicated. Mean age was 33.2 (13.4) years and sex ratio 27F/16M; 28 were affected by amyoplasia and 15 by other types.

Results: Beyond joint stiffness and deformities and muscle weakness, the well-known core symptoms quantified in the study, and for which the first line treatment was represented by technical aids, this study revealed that other less visible disorders contribute to severe participation restriction. These were particularly pain and psychological suffering including anxiety, fatigue, difficulty in sexual life, altered self-esteem, and feeling of solitude. Severe respiratory disorders are not frequent and linked to a specific genotype, as well as swallowing and speech disorders. Respiratory impairment did not explain gait disorders. Functional independence was worse in patients with amyoplasia than in patients with other types of AMC.

Conclusions: This study reveals that the handicap of adults with AMC is influenced by genotypes, and that the invisible part of this handicap is important.
Abstract #26
Functional Outcomes Among Children with AMC: Retrospective Review on 114 Charts

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Background and rationale: Arthrogryposis multiplex congenita (AMC) is a term used to define joint contractures in two or more different areas of the body and has an overall prevalence of 1 in 3,000 live births. Contractures can lead to decreased range of motion and strength, and may affect ambulation and autonomy. AMC can be categorized into 3 types: 1) amyoplasia is a sporadic condition with no specific gene, typical clinical presentation includes internally rotated shoulders, extended elbows, ulnar flexed wrists, hips and knees in either flexion or extension and clubfeet 2) distal arthrogryposis (DA) has a genetic etiology and typically involves the distal upper and lower joints 3) other/CNS Involvement is a form with the typical contractures as well as central nervous system dysfunction. There is a current need to better understand the clinical and surgical management of children with AMC to offer evidence-based care and develop future research priorities. The aim of this retrospective chart review was to describe the physical characteristics and functional status of children with AMC who were followed at a pediatric orthopedic setting.

Methods: A chart review of children with AMC followed at SHC - Canada between January 1979 and July 2016 was conducted. Of these, 6 charts were excluded due to misdiagnosis and/or insufficient chart information, and 114 were retained. Patient demographic information, type of AMC, comorbidities, surgical treatments, level of ambulation autonomy in self-care and transfers were recorded.

Results: At last clinic follow-up, there were 54 males and 60 females with a mean age of 10 years 3 months. Amyoplasia and DA were equally represented, 42% and 43% respectively. The most common area of contractures in the lower extremities were the ankles/feet and in the upper extremities, wrist/hands. The majority of children in all three groups were ambulators. A large proportion of children with amyoplasia (40%) were dependent in self-care as compared to DA (5.4%). Most children with AMC regardless of type were independent in transfers.

Conclusion: As AMC is a rare condition, this large sample size gave a clearer understanding of the physical and functional challenges associated with AMC. These findings demonstrate the importance of genetic testing to provide accurate diagnosis and classification which can then guide treatment as diagnosis and classification based on retrospective clinical information is less reliable.
Abstract # 27
Health-related Quality of Life and Orthosis Use in a Swedish Population with Arthrogryposis

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Introduction: Gait efficiency and functional exercise capacity have been reported as lower in children with AMC than in healthy controls (HC). To enable or facilitate walking orthoses are often used. The aim of this study was to describe health-related quality of life (HRQoL) and satisfaction with orthoses in a group of ambulatory children with AMC.

Methods: Thirty-three children with AMC with a mean age 10.5 (SD 4.2) years participated in the study. Questionnaires measured HRQoL (CHQ-PF and EQ-5D-Y), mobility and self-care (PEDI), and satisfaction with orthoses (QUEST 2.0). Eighteen children used orthoses either to enable or improve walking, of which nine children were dependent on orthoses for walking (Ort-D). Nine children used orthoses but were able to walk short distances indoors without orthoses (Ort-ND). Fifteen children did not use orthoses (Non-Ort). CHQ-PF (Parent Form) was compared between AMC and a Swedish reference group of 60 HC, mean age 12.9 (SD 1.5). Statistics were performed with SPSS version 23.0.

Results: Children with AMC had significantly lower CHQ scores in nine of 12 subscales compared to HC. When comparing groups with AMC, Ort-D had lower CHQ physical functioning than Ort-ND (p=0.011) and Non-Ort (p=0.002). The children’s reported perception of health with EQ-5D-Y did not show any difference between the groups. PEDI showed less mobility in Ort-D than in Non-Ort (p=0.012), whereas there was no difference in self-care. In total, both Ort-D and Ort-ND were “quite satisfied” with their orthoses, however Ort-D was less satisfied with orthosis weight than Ort-ND (p=0.014). As the three most important factors concerning the orthoses, Ort-D preferred “comfort”, “safety”, and “easy to use” and Ort-ND preferred “easy to use”, “comfort”, and “effectiveness”.

Conclusion: Children with AMC had lower physical HRQoL than HC. Of the groups with AMC, lowest physical functioning was found in Ort-D. As confirmed with PEDI, mobility was lowest in Ort-D. When reported by the children, EQ-5D-Y did not discriminate in perception of health between the groups. As reported by QUEST, children in both groups were “quite satisfied” with their orthoses. This study contributes to knowledge of HRQoL in ambulatory children with AMC and how they perceive their orthoses, emphasizing the importance of each child’s opinion when prescribing orthoses.
Abstract # 28
Preliminary Report on a Method of Assessing Functional Hip Motion

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Pediatric outcomes measures for hip procedures have been difficult to establish, and need to be fundamentally different from those devised for adult hip procedures. Adult hip scales are primarily evaluate outcomes for total hip replacements and focus largely on the patient’s symptomology, and not a functional range of motion. Pediatric hip pathologies in arthrogryposis are congenital, either dislocations or contractures, and are rarely painful during childhood. Evaluating activity limitations in pediatric hip conditions, before and after surgical treatment is also a less reliable outcome measure compared to adults. Many conditions are treated in infancy, before full acquisition of milestones, making functional comparisons infeasible. Also, children usually are less impeded by physical impairments than adults, attempting to adapt to their circumstances in order to participate. The existing pediatric outcomes scales, such as the PODCI, are very useful but do not evaluate the global improvement in hip motion after hip reconstruction in the child.

We will present preliminary work on a method to assess the total or global range of motion of the hip, then use it to assess motion before and after a treatment, thereby judging the effectiveness of a treatment. We created a model to graph hip range of motion in three dimensions, with the axes of flexion-extension, abduction-adduction, and internal-external rotation in the X-Y plane. Connecting the data points on the 3 axes creates a “radar” or “spider web” plot. Normative data was used to create an ideal or standard hip plot. This standard was made three dimensional, with the Z-axis, projecting perpendicularly, emanating up from the point corresponding to 30° of flexion, all other axes neutral (the most functional position for a fused or immobile hip). The resulting 3 dimensional graph is tent-like. A subject’s hip motion is plotted on the same radar plot, and projected straight up, intersecting the “roof of the tent”. The volume of standard hip graph captured by the outline of the subject’s plotted hip data is expressed as a percentage of the entire volume underneath that standard hip graph, i.e. a percentage of “functional hip motion”. Comparisons of the percentage of “functional hip motion” before and after a treatment would be meaningful in understanding how much a given surgical intervention improves a patient’s global hip motion range, which can then be compared a patient’s functional abilities over time.
Abstract # 29
Outcomes of Adults with Arthrogryposis Multiplex Congenita

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We have conducted the largest international study for identifying the long-term outcomes of adults with Arthrogryposis Multiplex Congenita (AMC). We recruited 177 participants from over 15 countries. They provided demographic information, such as living situation and mobility, and completed two standardized outcome measures: quality of life and physical activity. The information was collected using an online survey. The data were compiled, and descriptive analyses were performed. The study group consisted of 72% females and a mean age of 39 years. More than 90% of participants had upper and lower limb involvement, 35% had scoliosis or lordosis while 16% had jaw problems. Participants had an average of nine surgeries. The majority (75%) of respondents lived independently of family members (on their own or with a partner). Participants reported lower physical function scores than the general US population. They also reported similar or higher scores for the other quality of life domains of the SF-36. Half of participants experienced chronic back pain and 60% reported joint pain. Nearly half of the participants took regular pain medications. Based on this initial study, we pursued the validation of existing standardized surveys such as the Oswestry Low Back Disability Questionnaire for AMC. Additionally, as part of a follow-up we collected data regarding age and types of surgeries, long-term outcomes and complications to evaluate efficacy of the operations.
Abstract # 30
Arthrogryposis Multiplex Congenita: A Multi-site Pilot Registry in Pediatrics

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Introduction: Arthrogryposis multiplex congenita (AMC) describes a heterogeneous group of conditions with multiple congenital contractures. Etiology and natural history of AMC have yet to be described as studies are often single-site and are composed of small sample sizes. This limits comparability and pooling of findings across individual studies. Similarly, medical and rehabilitation management of AMC have also been poorly described to date. Preliminary hypotheses regarding certain etiologies and effectiveness of certain treatment have been proposed however rigorous research methods to prove their effectiveness are lacking. To support high quality studies to drive this field forward at both the epidemiologic and clinical levels, a multicenter pilot registry for children with AMC has been developed.

Methods: A data set was developed for the pilot phase of this registry using expert consensus. Information regarding the child’s first month of life, pregnancy, delivery and parental lifestyle habits were included. To standardize data collection methods, an operations manual was also elaborated. Forty families of children from birth to 21 years of age with AMC will be invited to complete the registry for the pilot phase.

Analysis: Descriptive statistics and regression analyses will be used to summarize relevant data and to explore associations to generate hypotheses regarding factors contributing to AMC. Qualitative analysis will also be used to better describe the phenotype of AMC and to determine ways to expand the registry and improve on current questionnaire. Recruitment methods as well as attrition rates will also be investigated.

Conclusion: The pilot registry for children with AMC will provide the platform for a comprehensive AMC registry that will generate multiple research avenues to enhance current care and establish new therapies. Following this pilot study, the participant selection criteria will be refined; data sets will be expanded to include comorbidities, rehabilitation and surgical outcomes, and genetic sequencing.
Abstract # 31
The creation of an Adult Arthrogryposis Research Registry

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Background: Arthrogryposis (AMC) research is challenging due to its relatively rare occurrence and variability in presentation. Many questions arise in parents about what will happen to their children. These are still difficult to answer due to the lack of longitudinal studies on adults. The second challenge is the lack of standardized and validated outcomes for ongoing research. Recently, a large cross-sectional study of adults with AMC involving 177 participants worldwide used two self-reported standardized outcome measure tools for quality of life (SF36) and physical activity (PASIPD) utilizing an online survey format. This study precipitated the work of validating a self-reported standardized outcome measure, the Oswestry Disability Index (ODI), in the AMC population. This is the beginning of creating the first international adult AMC registry. This paper describes the process by which the registry was created. The purpose of this study was to develop an online registry for collection of longitudinal data on adults with AMC which is meaningful to people with AMC and useful for clinicians and researchers.

Methods/result: A systematic review was done to identify validated and reliable outcome measures suitable for adults living with AMC, however aside from the now validated ODI there were no other measures. Based on focus groups of adult AMCers at the International Arthrogryposis Symposium in Oklahoma (2016) and the WHO’s International Classification of Function, Disability and Health (ICF), a set of standardized, validated and reliable outcome measures, applicable to AMC were selected to include in the registry. Experts, including patients, clinicians and researchers, prioritized the outcome measures to be included in the registry using two rounds of survey using a Delphi methodology (not sure my correction here is better). The final selection of questionnaires was done by consensus at a face-to-face meeting in May 2018.

Conclusion: The main anticipated outcome of this project, by people and for people with AMC, is to create a meaningful and clinically relevant online registry for adults living with AMC who want to contribute to a longitudinal study that explores the various changes in function and well-being over their adult lifetime. The systematic collection of data longitudinally will provide a platform for future research avenues providing information on natural history, outcomes and long term prognosis.
Conception of Achilles Tenotomy in Children with Arthrogryposis

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Introduction: Achilles tenotomy is the last step in clubfoot correction by Ponseti method, but severe soft tissue rigidity in patients with arthrogryposis make often us to reconsider the conception of achillotomy. Aim of our study was to analyze effectiveness of achillotomy in children of the first year of life with arthrogryposis.

Material and Methods: 54 patients (108 feet) with arthrogryposis multiplex congenita (AMC) and 18 children (32 feet) with distal type (DA), treated by Ponseti method were selected for the study. Mean number of casts in AMC group was 7.8±0.21 (from 6 to 12) and in DA group was 4.7±0.6 (from 3 to 7). During conservative treatment achillotomy was performed in 56 feet of AMC group and 32 feet in DA. Other children with AMC had residual deformity and required surgery.

Results: Analyze of effectiveness of the procedure, performed for equinus to be eliminated revealed that mean angle of calcaneus position change in AMC group was 23±2°. Mean angle of equines before treatment in this children was 40±1.4° (from 30 to 70°) and after achillotomy 15±1.4° (from -5 to 25°). The patients with DA demonstrated more significant correction. Mean angle of calcaneus position change in this children was 34±1.17° (p <0.05). Mean angle of equines before treatment in this children with DA was 27±2° (from 15 to 45°) and after achillotomy -7°±1.3°, i.e. 7° of dorsiflexion (from -15 to 0°). Residual equinus after achillotomy, not excideing 20° eliminated in all patients with further weekly casting and correction of hill position during 3–4 weeks. Thus mean angle of correction was 40° (23° after achillotomy and 20° after further casting).

Conclusion: We come to conclusion that effectiveness of achilolotomy in order equinus to be eliminated depends on type of the pathology (it is the most effective in children with distal type). If equinus deformity exceeds 40° achillotomy is not helpfull. Such patients should be considered for surgery. Significance: Consideration of angle of equines before treatment help to choose the highly effective management of clubfoot in children of the first year of life with arthrogryposis.
Abstract # 33
Treatment of Arthrogrypotic Clubfeet Using Ponseti Casts and Posterior Ankle Release

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Background: The initial treatment for arthrogrypotic clubfeet (ACF) using the Ponseti method is considered a standard, however relapses are common and satisfactory correction of equinus after Achilles tendon tenotomy remains controversial. The aims of our study were: evaluation of the efficacy of Achilles tenotomy and assessment of the early results of posterior ankle release in those ACF where the correction of equinus was unsatisfactory following the Achilles tenotomy.

Methods: This is a retrospective study performed between 2014–2017. Included into the study were children with arthrogryposis, without obvious central nervous system involvement, treated by the authors and aged 0-12 months during the beginning of the treatment. Excluded were children who didn't meet the above inclusion criteria or were lost to follow up. The final cohort consisted of 14 children (27 feet, 7 boys, 7 girls), aged 4.4 months (range: 1.6 – 9.4 mo.) during surgical intervention. 7 children had amyoplasia and 7 had other forms of arthrogryposis without CNS involvement. The mean follow up period was 21.2 months (range: 6 – 47.7 mo.). The treatment consisted of the Ponseti casts followed by surgical treatment in the form of initial Achilles tenotomy – if dorsiflexion or neutral position of the ankle was obtained the procedure was completed by cast application. However, if any equinus persisted immediate open posterior release was performed until desired correction was obtained. Final follow up included assessment of the need for repeated casting or surgery and functional grading of the deformity according to Carlson.

Results: In all but one foot, the correction after Achilles tenotomy was unsatisfactory, and those 26 feet required a posterior ankle release. 7 children (13 feet, 48%) required at least 1 repeat casting session after a mean time interval of 9.3 months (range: 3.3; 14.6 mo). 5 children (10 feet, 37%) required revision surgery after a mean time interval of 20.7 months (range: 8.4; 30.6 mo.). At final follow up all children had a good or satisfactory outcome according to Carlson.

Conclusions: Unlike in idiopathic clubfeet, Achilles tenotomy does not provide satisfactory equinus correction in ACF. Performing an immediate posterior ankle release allows achievement of ankle dorsiflexion and satisfactory early-term results of the treatment.
Abstract # 34
Longitudinal Management of Recurrent Arthrogrypotic Clubfeet with Ponseti Management

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Purpose: The management of clubfoot in arthrogryposis is notoriously challenging with a high recurrence rate compared to idiopathic clubfoot. Our practice treatment philosophy manages recurrences with minimal intervention using a Ponseti style technique and repeat Achilles tenotomies. The goal is to minimize deformity to achieve a plantigrade, bracable foot, while preserving comfort with avoidance of large procedures. Our observation is that over time there is a sinusoidal rate of recurrence which tapers as growth slows and is hastened with Ponseti management. We therefore describe the longitudinal appearance of recurrence using Pirani scores for a cohort of 25 patients.

Methods: 25 patients were sequentially identified from our clubfoot database (2008–2017) with the diagnosis of arthrogryposis and talipes equinocavovarus. Inclusion criteria required two years of follow-up data with complete Pirani scores treatment and recorded minimum of two cycles of Ponseti management consisting of a series of casts followed by a tenotomy. Patients were excluded for incomplete measurements. Ordinal composite values for Pirani scores were calculated for initial deformity and final result after completion of a series for each series. Analysis consisted of comparison of averages.

Results: Average initial Pirani score (n=45) at the start of the first recorded casting is 4.27 and at completion is 1.99 representing a change of 2.28 points or 53%. The second casting begins with an average initial Pirani score of 3.74 (12% lower than first casting) and achieves a final average Pirani score of 2.52 (27% higher than prior) representing a more narrow change of 1.22 points or 33%. A third series was seen less frequently in the observed time range (n=9) which demonstrates an average initial Pirani score of 4 which decreases by an average 0.75 points to 3.25.

Conclusion: Clubfoot in arthrogryposis will recur. Control of recurrence of arthrogrypotic clubfoot can be repeatedly maintained with Ponseti technique, but may be most powerful earlier in treatment with effective, yet diminished, ability to elicit change over time. Value in accepting some residual deformity may be elucidated in comparison with cohort of patients who receive dramatic interventions using patient satisfaction outcome measures.
Abstract # 35
Identifying Equinocavus Variant of Arthrogrypotic Clubfoot

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Purpose: Equinocavus (EC) is a term used descriptively to explain foot deformity consisting mainly of equinus and cavus and is a variety of foot deformity in the child with arthrogryposis. We have found that this deformity responds differently to traditional Ponseti management and is easily overcorrected into extreme valgus when confused with traditional equinocavovarus (ECV). We aim to illustrate the difference in initial presentation with a matched cohort using Pirani exam scores.

Methods: Eight known patients with suspected EC arthrogryposis clubfoot variant were identified. One was excluded due to late presentation at 7 years of age. The remaining were all assessed at less than 14 months of age and had prior minor procedures such as serial casting or Achilles tenotomy in the past. A cohort of seven random patients was selected from our clubfoot database meeting the criteria of age < 14 months, primary diagnosis of amyoplasia or distal arthrogryposis, and only minor prior procedures performed. All patients were bilaterally effected and each foot was treated as an instance. T-test comparisons were made between initial recorded equinus and initial Pirani score along with Pirani score subsets for hindfoot and midfoot.

Results: Equinus comparison was not statistically different with average initial dorsiflexion (EC=-33.4°; ECV=-38.9°; p=0.53) and initial Pirani hindfoot contracture score (EC=2.5; ECV=2.9; p=0.25). The Pirani midfoot contracture score (EC=1.1; ECV=2.6) and total initial Pirani scores (EC=3.6; ECV=5.6) were found to be significantly different (P<0.0001) correlating with the decreased acuity of varus and adduction in EC.

Conclusion: EC is a variant of arthrogrypotic foot deformity which clinically displays main deformity in equinus and cavus with relatively minor contributions from varus and adduction as compared to traditional ECV. The total Pirani and subsets of midfoot contracture are statistically lower in EC than in ECV. This may help to identify EC variant at initial assessment. We have found that EC variant is best treated with a different manipulation technique (4-finger technique) versus the traditional Ponseti method which may result in overcorrection into extreme valgus. This preferred technique for EC initially corrects the cavus by mainly extending the forefoot and midfoot to the level of the hindfoot with only gentle midfoot correction as needed. This is followed by correcting equinus of the hindfoot by casting and/or tenotomy.
Abstract # 36
Treatment Severe Deformities of Plano-valgus Feet in Children with Arthrogryposis

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Plano-valgus deformity of the foot occurs mainly in children with distal forms of arthrogryposis. The purpose was to determine the age indications for different types of surgical intervention. Materials and methods: 48 children with distal forms of arthrogryposis were examined and treated. The age of patients was 1 months–16 years. They had vertical talus. Conservative treatment with plastering staging was performed in 10 patients (20 feet) at the age of 6 months. In children older than 1 year conservative treatment was aimed at stretching the tissues along the dorsally-external surface of the feet before surgical treatment. 40 patients (69 feet) were exposed to an open correction of the talus with extended release and stretching tendon. Extended release of the feet with skin plastics was added by Ilizarov device in 5 patients (7 feet) with severe degree of plano-valgus deformities older than 5 years old. 5 patients (5 feet) had repeated interventions in the form of extraarticular subtalar arthrodesis or triarticular arthrodesis. Results: The result of treatment was assessed the AOFAS scale. Stage plastering in children up to 6 months allowed surgery and in children up to 2 years reduce the amount of intervention. After open correction of the talus, good results were observed in 70.4%, satisfactory in 17.6%, unsatisfactory in 11% of cases, which required further intervention on the bones or repeated releases on the foot with Ilizarov device.

Conclusion: The best results were observed in the group of children whose treatment was started before the age of 1 year. The tissues of children with arthrogryposis are so rigid that even the use of a minimally invasive method with a talon-navicular release and subcutaneous achillotomy did not allow proper interrelation in the joints of the foot. The expansion of the scope of surgical intervention was required.
Abstract # 37
Reducing Recurrence of Equinus in Tibial Dorsiflexion Osteotomies by Tendon Transfer and Wolff’s Law

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Introduction: Tibial dorsiflexion osteotomy for recalcitrant equinus in clubfeet is reported to have a high recurrence rate with growth, when done in children who grow for several years after surgery. Our hypothesis is that dorsiflexion osteotomies weaken the dorsiflexors by making them lax and worsen the imbalance between the dorsiflexors and plantarflexors, which favors recurrence due to the unopposed pull of the plantarflexors. We believe that correcting this imbalance will reduce the recurrence by Wolff’s law. We used this method as a salvage procedure in patients who had been referred with equinus following multiple surgeries and some of the patients had a flat top talus resisting dorsiflexion through the ankle.

Materials and Methods: We present the results of closing wedge distal tibial osteotomies and tendon transfers in 15 feet and 11 patients, who presented with recalcitrant equinus. The mean age at presentation was 7 years (range 3.98 to 12.38). Three patients had a diagnosis of arthrogryposis and three others had non functioning tibialis anterior muscles. One patient had subtalar coalition and a ball and socket ankle. The initial surgeries were done at other hospitals and included achilles lengthening and posteromedial releases. All had small scarred calf muscles. A closing wedge distal tibial osteotomy (DTO) was done in all patients as a first stage procedure. Fixation was done with Kirschner wires. A tendon transfer to the lateral cuneiform produce dorsiflexion and eversion was done in all cases after the patients resumed unaided normal gait, except in one patient where it was done at the time of the DTO. Tibialis anterior lateral transfer was used in 9 feet, the tibialis posterior in 2 feet, peroneal tendon in 2 feet and the flexor digitorum longus in one foot. Patients with arthrogryposis continued to use an ankle foot orthosis.

Results: At a mean follow up of 4.16 years, 11 of the 15 feet had neutral or more dorsiflexion and 4 had a recurrence of equinus. One patient had a pin site infection which required a wound washout and resulted in a delay of the second stage tendon transfer. Mean correction of dorsiflexion was 24.5° and mean correction of anterior distal tibial angle was 10.9°.

Conclusion: The loss of correction of tibial dorsal tilt through remodeling appears to be slowed down by the tight check rein effect of the transferred tendon, which promotes dorsiflexion and influences bone growth by Wolff’s law.
Abstract # 38
An AMC Multidisciplinary Clinic: Using the COPM to Ensure a Family-centered Model of Care

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Introduction: Children with arthrogryposis multiplex congenita (AMC) have multiple needs requiring the involvement of multiple professionals. Historically, these children were seen by various healthcare professionals (HCP) at different visits, leading to fragmented care. A multidisciplinary clinic combining orthopedics, genetics, nursing, rehabilitation and social services was created to provide comprehensive care. Considering the importance of involving families in the management of their care, the Canadian Occupational Performance Measure (COPM) has been integrated into the clinic process to guide interventions and promote seamless communication among HCPs and families.

Methods: Families were contacted prior to the clinic to set goals using the COPM. A tracking sheet was used to document the COPM goals, and a personalized copy was given to each family when they arrived for their clinic visit. During the clinic, the tracking sheet was used to promote communication between HCPs and to ensure recommendations and intervention plans were linked with the goals established. Family-centeredness of the clinic was evaluated using the Measure of Process of Care questionnaires (MPOC) completed by family members and HCPs.

Results: Twenty-four families established goals using the COPM; 3 families have had the chance to complete the COPM twice due to return visits in clinic. Goals were divided into 4 domains, namely self-care (n=15), physical (n=37), equipment (n=2) and health concerns (n=26). HCPs treatment plans will be correlated to these goals to determine extent to which interventions were influenced by these initial goals. The setting (hospital vs. local center) in which the treatment plan was carried out will be determined. Thirty-two families completed the MPOC-20, these results will be analyzed and presented.

Conclusion: The use of the COPM to guide intervention planning in an acute care multidisciplinary clinic, and increase cohesiveness between HCPs and with families is promising. This model can be used throughout many organizations facing similar challenges of engaging patients and families and providing comprehensive coordinated services.
Abstract # 39
AMC Patient Management at the University Hospital Grenoble/France: a Single Center Experience

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Objective: Describe multidisciplinary patient care in a nationwide reference center for AMC.

Patients and methods: As a result of the governmental incentives in France to create Reference Centers for rare diseases, we established multidisciplinary consultations for children with AMC in 2007 as center for developmental anomalies. We established adult consultations from 2009 on. Patients are either addressed via the national patient organization “Alliance Arthrogrypose,” or via our medical clinical network (geneticists, neuro pediatricians, others), whereas some patients contact us directly. Patient evaluations are performed on a 2-day and 4-day basis respectively for children and adults. Evaluations for children include rehabilitative, orthopedic, genetic and psychologic consultations, as well as systematic muscle MRI, and x-ray imaging and molecular analyses when appropriate. Evaluations for adults include rehabilitative, rheumatologic, pneumology, and genetic consultations, as well as systematic muscle MRI, x-ray imaging, osteodensitometry, spirometry and when appropriate molecular analyses.

Results: We have seen more than 150 patients since 2007. Patients with Amyoplasia represent 60% of all patients, followed by subtypes of Distal Arthrogryposes. Unlike initial management plans from local institutions that vary but are satisfactory in a majority of cases, a precise diagnosis has rarely been made. Our goal as expert center is to provide information on the diagnosis, the rehabilitative and orthopedic management, recurrence risk and prognosis for each patient, and identify best suited local centers for patient care with which to collaborate. Due to an increasing demand, we have started follow-up evaluations since 2017 for patients that have already been seen in our institution.

Perspectives: In order to harmonize nationwide patient care, we are working on management guidelines for patients with AMC. Furthermore, we are planning to perform a patient education program focusing especially on pain, a major concern especially in adults.
Abstract # 40
The Development of an MDT Approach to Arthrogryposis at Birmingham Children’s Hospital

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Introduction: Children with Arthrogryposis have complex needs. Their families have multiple concerns, attend many hospital visits and depend on specialist advice. To enable families to receive holistic care, an MDT clinic has been established at Birmingham Children’s Hospital over the last 15 years.

Setup: We run 12 clinics/year with a team of 2 orthopaedic, 2 plastic, a Genetic Consultant, an occupational and a physiotherapist and at times a paediatric neurologist. A member of TAG is invited to the clinic to meet with the families. A dedicated administrative staff liaises with the families and provides support. Patients are reviewed and an individualised diagnostic and management plan is discussed with the family. Surgical procedures are planned in combination with the plastic and orthopaedic consultants, so that the procedures can be combined. Therapy is crucial for these children and establishing a supportive relationship between the families and the therapists is vital.

Results: Birmingham Children’s Hospital is a tertiary referral centre, with a catchment area of 6 million. Through regional paediatricians, we hope to invite all babies with a possible diagnosis of arthrogryposis to attend within a few weeks of birth. In the early 2000’s, a joint clinic between an orthopaedic and plastic surgeon was established to manage several patients with arthrogryposis. This clinic developed into a full MDT setting. In 2016–2017, a total of 106 patients were seen. Currently, approximately 25 new patients are seen each year. Investigations may include MRI, NCS/EMG studies, muscle biopsy and blood tests for evidence of a muscle, nerve or metabolic conditions guide genetic testing. A neuromuscular consultation is often included. The treatment plan is always individualized, and may involve a combination of physio- and occupational therapy, serial casting, orthoses and surgery. The team provides verbal and written supportive information.

Conclusion: A patient centered MDT clinic has improved the holistic care of children with Arthrogryposis, and has lead to a better understanding of the condition. It reduces the number of hospital visits for the children and families, streamlines the investigation and decision making process. Hence patient satisfaction is improved as is quality of care. The interactive learning of all team members and the ability to keep up to date with new developments regarding this rare condition cannot be emphasized enough.
Abstract # 41

Foot Deformities and Gait Deviations in Children with Arthrogryposis

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Introduction: Arthrogryposis is a congenital condition characterized by joint contractures with foot deformities and gait deviations. The most common foot deformity in arthrogryposis is clubfoot. The aim of this study was to quantify gait deviations and foot deformities in children with arthrogryposis using three dimensional, instrumented gait analysis. SIGNIFICANCE: This study investigates dynamic data from gait and foot pressure analyses as well as physical exam to quantify foot deformities and gait deviations in children with arthrogryposis.

Methods: Children with arthrogryposis were evaluated retrospectively in the gait laboratory and compared to data for typically developing children. Data included multi-segment and single-segment foot kinematics, kinetics, foot pressure, and physical examination. Children were also grouped by age, orthotic use, and surgical history.

Results: 42 children with arthrogryposis age 10 ± 5 years old were evaluated. Foot deformity included 77 clubfeet of which 29 underwent Ponseti treatment and 48 who underwent surgical release. 7 feet had a congenital vertical talus with 6 undergoing surgical intervention. At the time of gait analysis, 11% of children wore KAFO’s, 44% of children wore AFO’s, and 45% of children wore no orthotic or an in-shoe orthosis. Physical exam and kinematic data showed that children with arthrogryposis walked with a crouched gait, exhibited stiffness in the hips, knees, and ankles and showed limitations in their gross motor functioning (p<0.006). Power generation was low at the ankle and was high at the hip (p<0.01). Multi-segment foot kinematics revealed stiffness in hindfoot plantarflexion and residual forefoot adduction (p<0.03). Foot pressure showed reduced heel impulse, excessive midfoot contact, and overall varus foot position (p<0.002). Categorization by age revealed greater stiffness at the hips and knees in older children (p<0.01). Children with KAFO’s showed the most stiffness (p<0.05). No significant differences were seen between the foot posture of children with clubfoot treated operatively or by Ponseti except for greater internal rotation at the ankle (p=0.0158) in Ponseti treated feet.

Conclusions: Motion analysis demonstrated significant gait deviations in patients with arthrogryposis. The foot posture of children with clubfeet were similar whether treated with the Ponseti method or with open surgical release.
Abstract # 42
AFO with Dynamic Carbon Fibers Improves Selected Gait Parameters in Children with Arthrogryposis

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Background: Carbon fibers used in Ankle-Foot-Orthosis (AFO) have an ability to capture mechanical energy, which is stored during dorsiflexion, unloaded in terminal stance and returned during push off. We suspected that AFO with carbon fibers improves gait parameters in patients with amyoplasia. Aims: To compare gait parameters between similar patients with amyoplasia walking bare-foot or using custom made AFO with carbon insert (Dynamic Ankle Foot Orthosis - DAFO) or Rigid AFO (RAFO) for walking.

Methods: 14 patients with amyoplasia of similar walking abilities, presenting with similar deformities and course of treatment were selected for the study. Gait analysis was performed and recorded by Vicon and TMSi system with EMG assessment. This system allowed to trace Ground Reaction Forces (GRF) in real time with observation of Shank to Vertical Angle (SVA). Every patient was recorded three times while walking bare-foot, with DAFO and RAFO.

Results: Bare-foot walking in all of patients resulted in decreased velocity of gait and shortened length of step accounting for 60% of the norm for age. The contribution of the double support phase in the walking cycle was increased and the phase of the single support was reduced. Contact with the ground in both limbs started with the forefoot. The use of both types orthosis (DAFO and RAFO) resulted in better alignment of ground reaction forces in all patients during stance phase. Walking in RAFO and DAFO ensured appropriate clearance of the foot during swing phase. In all patients the length of the step was 10% longer while using DAFO than RAFO. In 12 patients DAFO allowed for better positioning of the knee in the sagittal plane without soleus overstretch (10-12° inclined of Shank to Vertical Angle) in comparison to the RAFO measurements. The SVA value in DAFO was greater that in RAFO indicating that GRF pushed knee more to the front and provided better balance between stance and swing phase.

Conclusions: Walking bare-foot was possible but less functional than using DAFO or RAFO. DAFO allowed for most functional walking with longer steps and velocity.
Abstract # 43
Less Invasive Treatment of the Lower Limb and Carbon Fiber Spring Orthoses May Improve Gait in AMC

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Children with AMC not only lack muscle, but are also prone to deformed joints, such as hip flexion contractures, knee flexion contractures, knee luxations, clubfeet and vertical talus. Trunk muscles are typically spared, and may be used for ambulation if the lower limb joints are stabilized in orthotics. However, malalignment of the joints hinders beneficial biomechanics, and reduces the chance for independent walking. The new concepts of very early redressions, modified Ponseti casting and guided growth has reduced the number of major surgeries and has led to greater success in lower limb alignment while sparing as much muscle as possible, and reducing the need for e.g. triple arthrodesis. In order to stabilize joints and improve propulsion, knee-ankle-foot orthosis (KAFO) or ankle-foot orthosis (AFO) can be used.

We present two subjects with amyoplasia, both with four limb involvement and similar in their extensive lack of musculature in the lower limbs; A) a now 11 year-old child whose hips, knees and feet have been treated with redressions, casting, Achilles tendon tenotomy, club foot surgery left, vertical talus surgery right, knee and hip surgery and guided growth of knees; B) a now 5 year-old child whose club feet have been treated with repeated Ponseti casting, including two Achilles tenotomies and one open Achilles lengthening with dorsal capsulotomy. They have been fitted with articulated KAFOs for walking ever since the Ponseti redressions. When they had achieved enough motor control of the trunk and pelvis, they were fitted with orthoses that have carbon spring ankle joints. The spring is loaded during stance, and the power is then used for propulsion. The carbon spring thus acts as a calf muscle, and makes the gait more efficient.

They are compared with a now 28 year-old adult with distal arthrogryposis who has had a number of foot surgeries, including an Ilizarov frame, but still has deformed and stiff feet. He is fitted with carbon spring AFOs.

Clinical examinations, x-ray, surgery, use of orthoses and clinical gait analysis will be presented and discussed.
Abstract # 44
Growth Modulation for Recurrent Flexion Deformity in Arthrogryposis

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Background: Dorsal Carpal Wedge Osteotomy has been shown to be an effective treatment method for the wrist flexion deformity in Arthrogryposis patients. However, at times a recurrent flexion deformity occurs and repeat wedge osteotomy is not an option. In this subset of patients we have found that growth modulation offers a good alternative for treatment.

Methods: Medical records from the past three decades were retrospectively collected at a tertiary institution for patients demonstrating a carpal wedge osteotomy with an additional growth modulation procedure. Clinical and radiographic data specifically related to the procedures were assessed.

Results: A cohort of 5 patients (4 males and 1 female) underwent an initial carpal wedge osteotomy (CWO) with an additional growth modulation procedure for recurrent wrist flexion deformity. Average age at initial CWO was 4.8 years of age (range 2.5–7.5 years) with an increment average of 6.6 years (range 2.1–10.5 years) to the respective upper extremity growth modulation procedure. The average age at time of growth modulation occurred at 10.8 years of age (range 5–12 years) with an average follow up of 21 months (range 6–27 months). Initial resting position following carpal wedge osteotomy ranged from 0 to 10° extension, with recurrent flexion deformity averaging 35° prior to modulation procedure. All patients had significant outcome improvement of resting wrist position and maintained range of motion following growth modulation surgery.

Conclusion: Growth modulation offers an excellent option for recurrent wrist flexion deformity in patients with arthrogryposis when repeat osteotomy is not feasible. Although short-term results have shown improvement in resting wrist position, long-term follow-up is required to confirm the sustainability of this procedure.
Abstract # 45
Comparison for Techniques of Treatment of Wrist Flexion Deformity in Arthrogryposis

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Purpose: We are comparing dorsal wedge carpectomy (DWC) and volar wrist release (VR) for treatment of wrist flexion deformity in arthrogryposis.

Methods: Retrospective analysis of data sheets of 22 surgically treated wrists in 12 amyoplasia patients. The average age was 3 years. Average wrist flexion deformity was 23°. The patients were divided into 2 groups;

1. Osteotomy group: nine wrists treated with DWC
2. Release group: thirteen wrists treated with VR and flexor carpi ulnaris (FCU) transfer.

We reported the results regarding parents’ satisfaction, postoperative range of motion (ROM), wrist extension range and Percentage Of Improvement (POI) =

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\text{POI} = \frac{\text{Preoperative extension} - \text{postoperative extension}}{\text{preoperative extension}} \times 100 
\]

(Degree of extension is the complementary to a line perpendicular to the axis of the forearm). We analyzed the results in relation to ambulatory status, sex and age of intervention. Average follow up was 10 months.

Results: There was significant improvement of wrist extension in all wrists \((p<0.01)\). POI showed better improvement in the osteotomy group (75.5%) than the release group of (56%) with significant difference \((p=0.011)\). The postoperative total ROM increased in the release group (average 7°) and decreased in the osteotomy group (average -2°). Although the total ROM increased in Release group and decreased in Osteotomy group, there was more improvement of extension in Osteotomy group, but without significant difference \((p=0.44)\) between the two groups. There was more wrist extension in patients using their hands for ambulation. The outcome was not related to the age, or sex. Parents’ satisfaction was excellent and good in 16 hands, fair in 5 hands and poor in one hand.

Discussion: We preferred using the POI for comparing the results of treatment. Using the POI showed significant improvement in DWC in comparison to VR group. It reflects the real improvement even in cases with similar extension ROM. This study is in favor of DWC over VR for treatment of wrist flexion deformity in arthrogryposis. But these results are preliminary, too short follow up to report recurrence.
Abstract # 46
Wrist Contractures in Children with AMC: Long-term Follow Up of Surgical Treatment

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Introduction: Wrists contractures in patients with AMC are varied and damage wrist function and make activities of daily living difficult or even impossible. Multiple surgical procedures have been proposed to correct wrist contractures, but results were controversial because of further limit of motion and recurrence of deformity.

Materials and Methods: 90 patients (162 upper extremities) with the wrist contractures in AMC were examined and treated at the age from 6 months to 17 years. Clinical, X-ray study and neurophysiologic examination were performed. Flexion contracture, flexion contracture with ulnar deviation and isolated ulnar deviation were observed, which were combined with fingers contractures and thumb-in-palm deformities. Based on segmental muscles innervations patients were divided into 3 groups: with C6–C7, C5–C7 and C5–Th1 level of spinal cord lesion. 162 operations were performed on the wrist using following technique - tendon transfers, carpal wedge osteotomy and shortening osteotomies of forearm bones and combined with 155 additional operations for fingers contractures.

Results: 90 patients (162 wrists) were evaluated from 1 to 10 years after surgical treatment. With the increasing of amount of damaged segments passive correction, active movements, functional capacity for grasps decreased and increased the frequency of fingers contractures. The following criteria were taken into consideration for results assessment: wrist resting posture, active extension, functional capacity for grasps and cosmetic appearance. All results were divided into - good, satisfactory, unsatisfactory. Children with C6–C7, C5–C7 level of spinal cord lesion had 12%, 16% of satisfactory and 88%, 84% of good results, restoration of active wrist extension and improvement of functional capacity for grasps and cosmetic appearance were achieved. In C5–Th1 group patients had 11% of good, 79% of satisfactory and 10% of unsatisfactory results of treatment. Minimal increasing functional capacity for grasps and improvement of wrist position were observed. Recurrence of wrist deformity were revealed in 13% (21 wrists), in most of cases in patients with C5–Th1 level of spinal cord lesion (13 wrists).

Conclusion: Varied surgical approach to wrists and fingers contractures treatment can improve wrist function as much as possible. Determination of the level of spinal cord lesion can provide predictable results in children with AMC.
Abstract # 47
Guided Growth for Knee Flexion in Arthrogripsyosis: Neurovascular Encroaching with Bone Funnelization

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Introduction: Knee flexion deformity in arthrogrypsis multiplex congenita is reportedly treated by serial casting into extension, distal femoral osteotomies, distal femoral guided growth, hemiepiphysiodesis, external fixation, capsulotomy, and soft-tissues releases. We are aware of four cases treated by distal anterior femoral guided growth with tension band plates in which an unreported complication occurred: the screws of the tension band plates penetrated the posterior cortex of the femur during remodeling with metaphyseal funnelization risking the neurovascular bundle.

Materials and Methods: Inclusion criteria were cases with arthrogrypsis multiplex congenita and knee flexion deformity, treated at our institution by distal anterior femoral guided growth with tension band plates, and radiographic evidence of posterior cortex screw penetration during remodeling from growth.

Results: Six knees (four cases) met the inclusion criteria. The average of age at the distal anterior femoral guided growth with tension band plates was 5.8 years. The average of knee extension angle in the presurgical assessment was -52°, and the average of presurgical posterior distal femoral-physis angle was 97°. After an average of 5.6 years of follow up, radiographs showed that the screws of the tension band plates, which at surgery were intra-metaphyseal, had penetrated the posterior cortex of the femur. The average of knee extension angle in the postsurgical assessment was -38°, and the average post postsurgical posterior distal femoral-physis angle was 120°. Four knees (two cases) had diffuse pain around the knee to lower leg area, and instrumentation removal alleviated the symptoms.

Conclusions: During distal anterior femoral guided growth with tension band plates for knee flexion deformity from arthrogrypsis multiplex congenita, we found that the screws of the tension band plates, which were initially located inside the metaphysis, may protrude through the posterior bone cortex during metaphyseal funnelization with growth, and may encroach upon the neurovascular tissues. Significance: Report of potential neurovascular bundle injury during guided growth with tension band plates in knee flexion deformity in arthrogrypsis multiplex congenital.
Abstract # 48
Guided Growth with Hinge Tension Band Plate for Lack Of Extension of the Knee in Arthrogryposis

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Introduction: Lack of extension of the knee (LEK) and Fixed flexion of the knee (FFK) may occur in patients with Arthrogryposis Multiplex Congenita (AMC). Recommended treatments have included bracing, physical therapy and in severe cases: popliteal posterior release, distal femoral osteotomy or progressive distraction with external fixation. These treatments can fail due to complications, being one of the most relevant the rebound of the deformity. Kramer and Stevens reported correction of these deformities using staples for hemiepiphysiodesis of the distal anterior femur. Klatt and Stevens reported in a further experience “some limitations of stapling include relatively slow correction and occasional hardware migration”, in the same report they present their results using a pair of anterior tension band plates for hemiepiphysiodeses with less complications. The aim of this study is to evaluate how effective the Hinge Tension Band Plate is to treat the LEK in this population.

Material and Methods: We present our experience with guided growth using an alternative way to do anterior hemi-epiphysiodeses with Hinge Tension Band Plate in patients with LEK and FFK with AMC. We studied 5 patients with AMC (10 knees). The mean age was 6 years (3–8) and the mean LEK was 40º (20º–60º). The mean follow up was 50 months (from 25 months to 64 months). Clinical assessment included measurement of knee range motion, radiographic evaluation, gait analysis, and screening for concomitant deformities.

Results: In base of our findings, we describe two patterns: Type 1: FFK with limitation of flexion and extension: 6 knees; Type 2: LEK allow to flex: 4 knees. One patient had previous treatment with staples that failed due to forward migration and 2 patients had been previously treated with custom plates (without hinges). These 3 patients were treated by replacing staples or the plates with Hinge Tension Band Plate. 6 knees (60 %) had a full correction in a mean time of 16 months (10 to 24 months); the others had a partial correction and need a distal femoral osteotomy with bone shortening to achieve the extension. We did not find screws loosening or migration. There were no undesired deformities. In patients that reach full correction, the plates were removed to avoid recurvatum of the knee.

Conclusions: We observed that guided growth is effective with Hinge Tension Band Plate for correction of LEK and FFK in AMC.
Abstract # 49
Correction of Mild to Moderate Arthrogrypotic Knee Flexion Contractures with Guided Growth

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Background: Knee flexion contractures in children with arthrogryposis range from severe to relatively mild. The mild and moderate require treatment as they do cause gait inefficiencies. This study assesses the effectiveness of anterior distal femur hemiepiphysiodesis, with or without a concomitant posterior knee release, for knee flexion contractures of <45°.

Methods: Medical records and radiographs of 37 consecutive pediatric patients (60 knees) with arthrogryposis and knee flexion contractures treated with anterior distal femoral hemiepiphysiodesis were reviewed. Mean age at surgery was 6.9 years, and the mean length of follow-up was 3.2 years (11 months–7.8 years). A posterior knee release was done at the time of surgery in 48 knees, including hamstrings lengthenings, proximal gastrocnemius release, and release of posterior capsule. Patients were treated with a nighttime knee-ankle-foot orthosis (KAFO) in full extension, which continued after plate removal.

Results: Mean knee flexion contracture was 35° (10°–55°) pre-operatively and averaged 6° at hemiepiphysiodesis plate removal. The plates were removed 19 months after implantation (6–41 months), for an average correction rate of 6°/100 days. Contractures ≤ 25° that did not have a posterior release corrected well. Knees with 30° - 40° contractures corrected well with the addition of a posterior release, but >40° saw only 50% of knees correcting to ≤10°. The mean total arc of motion was 70° preoperatively and 65° at followup. At latest follow-up, the mean contracture was 16°, and all knees were ligamentously stable, although 5 patients (9 knees) underwent repeat hemiepiphysiodesis for contracture recurrence at 38 months after plate removal. Seven patients who were previously nonambulatory became ambulatory, and 10 that ambulated with a walker became independent.

Conclusions: Knee flexion contractures in growing children with arthrogryposis can be treated with anterior distal femoral hemiepiphysiodesis alone for contractures ≤25°, and relatively reliably for contractures ≤40° if a posterior knee releases is included. Greater than 40° contractures may respond in certain cases. Recurrence of the contracture is common due to growth. A KAFO is essential to help stretch soft tissues during the correction as well as after the plates have been removed, to help prevent or slow recurrence. At follow-up, the gradual loss of correction that occurred did not impact on the ambulatory gains made.
Abstract # 50
Improving Knee Range of Motion in Arthrogrypotic Knee

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Introduction: Hip and knee contractures in arthrogrypotic patients can be difficult regarding decision making and treatment. The purpose of this study was to describe the treatment for the arthrogrypotic knee and concomitantly the hip. We reviewed pre- and post-operative knee and hip motion and described a classification to guide treatment.

Methods: We reviewed the charts of arthrogrypotic patients with knee contractures with or without hip contractures, consecutively treated from 2015 to 2018. We classified the patients into three main categories based on the type of knee contracture with a hip modifier: type 1 - flexion contractures, type 2 were extension contractures and type 3 were combined flexion and extension contractures (Table1). Treatment for type 1 included posterior knee release and femoral shortening, for type 2 we performed quadsplasty and anterior release and for type 3 we performed both. Hip flexion and/or extension contracture was treated at the same time. We used the paired T-test to determine statistical significance (p = .05).

Results: A total of thirteen patients (age: 5 yrs. 9 mo. ± 4 yrs. 2 mo.) were analyzed for each affected leg (n = 22). There were thirteen affected legs of type 1, six affected legs of type 2, and two affected legs of type 3. In the type 1 group (flexion contractures) knee ROM increased from 48 ± 28° to 81 ± 13° (t = 3.5, p = .003). In the type 2 group (extension contractures) knee ROM increased from 29 ± 31° to 78 ± 8° (t = 4.0, p = .010). Finally, in the type 3 group (combine contractures) knee ROM increased from 35° to 90° but statistics were not calculated due to small sample size. The hip ROM in all types went from a mean of 67° ± 30° to a mean of 88° ± 7° which was significant (t = 3.6, p = .001). Mean follow up is 12 months (range 6–27 months).

Conclusion: Using this new classification, we stratified each knee and hip to the appropriate treatment. Our results indicate that knee arc and range of motion can be increased through surgical intervention with the largest increases seen in the flexion group. We found that hip flexion and extension contractures were successfully treated with soft tissue releases at the same time. The abduction contracture most often resolved in the postoperative period with physical therapy. Further study will include long term follow up and quality of life measures.
Abstract # 51
Treatment of Severe Knee Flexion Contractures in Patients With Arthrogryposis

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Background: Knee flexion contractures in children with arthrogryposis are difficult to treat. The purpose of this study was to assess the effectiveness of posterior knee releases and gradual contracture distraction with a circular external fixator in correcting and maintaining correction of severe knee flexion contractures.

Methods: The medical records and radiographs were reviewed of 44 consecutive pediatric patients (72 knees) with arthrogryposis and who had their severe knee flexion contractures corrected with a posterior knee release and gradual distraction with a knee spanning circular external fixation frame. The mean age at surgery was 7.8 years, and the mean length of follow-up was 3.6 years. Posterior knee releases included hamstrings tenotomies, proximal gastrocnemius release, and release of posterior capsule.

Results: The mean total fixator time was 104 days, with a mean correction rate of 0.69° per day. The mean knee flexion contracture was 68° (range 30°–105°) preoperatively, and all knees were corrected to full extension, ±5°. At latest follow-up, the mean contracture was 23°, and all knees were ligamentously stable. The mean total arc of motion was 56° preoperatively and 33° at followup. Twenty-three patients who were previously nonambulatory became ambulatory. Complications included 5 lower leg neuropraxias that resolved; 20 fractures (6 while in frame treated with frame adjustment, 14 at frame removal treated within the post-operative cast); 3 draining pins sites in post-operative casts causing severe skin maceration, resolved with wound care; a superficial femoral artery pseudo aneurysm; a proximal tibial physeal arrest; and a single return to OR for frame adjustment. Thirteen knees in 9 patients underwent anterior distal femur hemiepiphyseodesis at an average of 55 months after frame removal for 35° contracture.

Conclusions: Posterior knee releases and flexion contracture distraction by circular external fixator was effective in improving the mobility of pediatric patients with arthrogryposis. The procedure is well tolerated by patients, but can have complications. Patient selection is very important. At follow-up, the gradual loss of correction that occurred did not impact on the ambulatory gains made in most cases, but a substantial number of patients will require further treatment for contracture recurrence.
Abstract # 52
Delayed Surgery in Amyoplasia-Does it Work?

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Introduction: Although there is consensus on early treatment of contractures in children with Amyoplasia some parents are neglecting treatment. They want their children to develop after their own needs. How do we as professionals cope with the parents and children? What treatment can be recommended and what would be the right time for treating the children?

Material and Methods: A nine year old girl born with contractures in all 4 extremities. Clinical diagnosis was Amyoplasia. Genetical analysis was not done. She had bilateral clubfeet treated after Ponseti with serial casting and achillotenotomy. Knees were in flexion. The right knee was treated with hamstringstenotomy at two months of age together with achillotenotomy. The parents refused further treatment including serial casting. At seven years of age she was referred to our clinic. We recommended surgery for the knee contracture and foot surgery to get her in a standing position. The parents agreed now and at 8 years of age she was operated with bilateral anterior hemiepiphysiodesis of the distal femur and bilateral talectomy and posterior release of the ankles.

Results: After 15 months of follow-up she is now standing every day 30 minutes in a standing wheel chair. Flexion contractures of the knees improved from 90° to 130°. Preoperatively the feet had 60 ° equinus, a midfootbreak in plantarflexion and contractures in the toes. Both feet are now plantigrade and well maintained in an ankle foot brace. She is now able to use normal shoes which were her biggest desire and motivation for surgery.

Conclusion: The follow-up time is too short to draw a final conclusion. This is only a single case report. According to literature the results for talectomy shows a lot of variation. Satisfaction rate for talectomies in bigger series reports is around 45% and 50%. There is a risk for clubfoot relapses and a risk for calcaneotibial arthrodesis (Bosse et al, J Pediatr Orthop 2017). The results so far are promising. Even in an eight year old girl with 90° of flexion contractures in the knees and 60° of equinus position of the feet it was possible after surgery to get her in a standing position.
Abstract # 53  
Pioneering Wrist Arthroscopy in a Patient with Arthrogryposis

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Patients with arthrogryposis characteristically present with flexion and ulnar contracture of their wrist that is functionally disabling. This is a multifactorial problem involving all structures around the wrist joint. Surgical treatment for these patients can comprise of a combination of midcarpal wedge osteotomy, tendon transfer, volar fascia release and fusions. Good results are obtained in just over half of the cases with the more extensive lesions resulting in, expectantly, worse overall results. (Oishi SN, 2017)

We present a case of a 16 year old patient with arthrogryposis spectrum who had significant and sustained improvement of her wrist pain and range of motion following arthroscopic arthrofibrolysis of the radiocarpal joint. She had presented to our institution with severe left wrist pain thought to be arising from ulnar abutment syndrome or triangular fibro cartilaginous complex pathology rather than her arthrogryposis per se. Clinical examination revealed a wrist held in flexion (0-0-70° AROM). Imaging revealed an ulnar plus variant, a wide radio-ulnar joint and radiocarpal joint space narrowing. In the course of treatment of the ulnar sided wrist pain, the patient underwent a wrist arthroscopy. Intra-operatively, arthrofibrotic plica bands in the radio-carpal joint and thickening were removed. The TFCC appeared intact but stretched over the ulnar head. Following the operation, the patient had significant pain relief and was able to extend her wrist actively to 15° beyond neutral (15-0-70° AROM) which was sustained on one year follow up.

This is a rare case, where arthrolysis of fibrous bands could improve the active wrist range of movement. In very selected cases arthroscopic arthrolysis may add to the treatment of these patients.
Abstract #54
Effects of the Playskin LiftTM Exoskeletal Garment on Arm Function for Children with Arthrogryposis

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Introduction: Arthrogryposis is a rare condition. Only few centres look after this very selected group of patients therefore details and outcome of various upper limb interventions and outcome data is scarce. Surgery to skin, soft tissue, joints, tendons and muscle has some value, but each patient needs to be individually assessed and discussed as no existing function must be diminished but enhanced. The timing of surgery is controversial. Decisions need to be made after a thorough multidisciplinary assessment of existing and potential function. The basic functions of the upper limb, which enable independent living, require the child to be able to get one hand to the mouth for feeding and one hand to reach the back side for toileting. The ability to use a keyboard is a further vital skill for independent living. The aim of this study is to look at the types of upper limb surgery performed and question the patient and or parents whether in their opinion the goal of functional improvement has been achieved or not.

Method: A retrospective review collected data of all patients who have undergone upper limb surgery from 2000 to 2018. We subdivided the interventions into 6 subcategories, thumb, finger, wrist, forearm, elbow and combined surgeries. We also looked at the number at upper limb interventions for each patient in combination or on its own. Patients will then be sent a validated questionnaire, Reach out, to determine their satisfaction with the intervention.

Results: Results will be presented.

Discussion: Any upper limb surgery in patients with Arthrogryposis is aimed at improving activities of daily leaving, participation and quality of life. Patient centred outcome assessment is a vital part of determining the success of the intervention. Obviously as surgeons we aim to not only improve “objective” factors e.g. range of movement or certain abilities but especially patient and parent satisfaction with the results of the intervention. The Reach Out questionnaire has been validated for pediatric upper limb assessment as it includes various domains of the ICF including satisfaction.
Abstract # 55
Sensory Modulation and Emotional Regulation in Posterior Capsular Elbow Release Patients

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Introduction: This study seeks to examine the effect of music and occupational therapy co-treatments in the reduction of anxiety behaviors in post-operative AMC patients. The study takes place at Shriners Hospital for Children (SHC) in Philadelphia, PA. Patients in this study participate in three days of post-operative rehabilitation for posterior capsular elbow release surgery with triceps lengthening at SHC. Elbow flexion and extension stretches are indicated as part of the clinical guidelines and home exercise program for the surgery and are crucial for elbow flexion maintenance. Music interventions involve musical entrainment around 60bpm as well as patient-preferred music and instrument play. The study explores music as a sensory modulation and emotional regulation tool during elbow flexion and extension stretches.

Methods: A series of case studies will be performed. Data will be collected via caregiver questionnaires, caregiver interviews, and observational data. Pre-surgical and post-surgical elbow flexion and extension range of motion will be measured.
Abstract # 56
Patient and Parent Satisfaction After Upper Limb Surgery

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Introduction: Arthrogryposis is a rare condition. Only few centres look after this very selected group of patients therefore details and outcome of various upper limb interventions and outcome data is scarce. Surgery to skin, soft tissue, joints, tendons and muscle has some value, but each patient needs to be individually assessed and discussed as no existing function must be diminished but enhanced. The timing of surgery is controversial. Decisions need to be made after a thorough multidisciplinary assessment of existing and potential function. The basic functions of the upper limb, which enable independent living, require the child to be able to get one hand to the mouth for feeding and one hand to reach the back side for toileting. The ability to use a keyboard is a further vital skill for independent living. The aim of this study is to look at the types of upper limb surgery performed and question the patient and or parents whether in their opinion the goal of functional improvement has been achieved or not.

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Abstract # 57
An Upper Extremity Evaluation Tool for Children with Arthrogryposis Multiplex Congenita

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Arthrogryposis multiplex congenita (AMC) is a term used to describe multiple contractures present at birth in at least two body parts and has an overall prevalence of 1 in 3000. Individuals with AMC display variable clinical features involving the upper and lower extremities, spine and the jaw which may impact and limit independence in self-care and activities of daily living (ADLs). For this reason, early rehabilitation is warranted to provide splinting, range of motion exercises, strengthening programs as well surgical interventions to correct deformities and thus promote daily function.

Rehabilitation is essential for children with AMC to minimize limitations of the UE and promote function through ADLs. However, adequate evaluations tools to guide best practice in this population are lacking as current tools do not: 1) apply to children with a hand deformity, 2) guide clinicians as to which specific joint or muscle limit functional performance, and 3) allow the use of normative data when adaptations are made to accommodate the limb deformities present in children with AMC. The overall aim is to develop an outcome measure targeting the UE for children with AMC in order to: describe the functional status of children with AMC, evaluate patient changes over time, guide surgical and non-surgical treatment planning, and determine treatment effectiveness. Specifically, this project aims to develop a prototype of an UE AMC evaluation tool; establish content validity with a group of clinicians by creating a questionnaire that will address the administration and overall test utility; and determine the psychometric properties of the tool, including construct validity, inter-rater and intra-rater reliability.

The deliverables of this project will contribute to the advancement of OT and promote the delivery of evidence-based care to children with AMC. The development of the UE AMC evaluation tool will be established by working in partnership with clinicians throughout the province as well as with an international expert, in order to address their clinical needs relative to evaluation and decision making in both rural and urban settings.
Abstract # 58
Rehabilitation Needs in Arthrogryposis Multiplex Congenita: Multiple Stakeholder Perspectives

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Introduction: Arthrogryposis multiplex congenita (AMC) is a term used to describe multiple contractures present at birth, which can involve upper and lower limbs, the spine and jaw. Although early intensive rehabilitation is warranted for children with AMC, very few studies have documented the pediatric rehabilitation process and services for this population. Objective: To identify the barriers surrounding rehabilitation as experienced by youth with AMC, their caregivers and clinicians.

Methods: This project draws on an integrated knowledge translation methodology, involving the following stakeholder groups: youth with AMC, family members, clinicians, and researchers. Six youth (range: 16–20 years, 2 females), eleven caregivers, and ten clinicians (5 occupational and 5 physical therapists) completed a semi-structured interview. A series of three focus groups: one with clinicians, one with youth and caregivers, and one joint group took place to discuss ideal treatment scenarios. Interviews and focus groups were transcribed verbatim. A template analysis was executed using the International Classification of Functioning, Disability and Health (ICF).

Results: All three stakeholder groups reported challenges at the body function and structure and environment domains of the ICF. Each stakeholder group yielded three themes. Youth: AMC and me, navigating my resources, and life with AMC; Caregivers: caring, supporting, managing care; Clinicians: communicator and collaborator, institutional factors, enabler. It was demonstrated that early active rehabilitation began at birth and that frequency decreased as school years approached. A need for family centered care throughout the pediatric year in rehabilitation was also identified. Clinicians reported the lack of a condition-specific outcome measure, thus complicating evaluation.

Conclusion: Moving forward, family-centered rehabilitation guidelines for youth with AMC will be created. These guidelines will benefit all stakeholder groups as they would standardize evaluation and best practice. Knowledge between specialized centers and local therapists should be shared and the development of an AMC-specific tool would be beneficial.
Abstract # 59
Towards the Development of Best Practice Guidelines for Rehabilitation

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Background: Early intensive rehabilitation is essential for children with AMC to promote independence in mobility and daily activities, improve upper extremity function, execute transfers, and participate in physical activity. However, there are currently no rehabilitation guidelines to inform best practice for AMC. Since AMC is rare, rehabilitation providers in community settings often feel unprepared and uninformed when providing care.

Objective: To develop practice guidelines for rehabilitation in collaboration with youth with AMC, caregivers, and occupational and physical therapists (OT, PT).

Methods: In the first phase, interviews with six youth with AMC, ten caregivers and ten OTs/PTs identified rehabilitation needs (manuscript submitted). The following areas were identified as priorities: i) promoting independence in daily activities, ii) accessing rehabilitations services, iii) addressing psychosocial needs, iv) managing pain, v) addressing physical needs. The next phase is to co-develop the practice guidelines for these five priorities. First, a scoping review of the literature will be conducted for each of these five priorities. The findings from the scoping review will be summarized and communicated with a panel of experts. Second, a survey to identify the treatment methods used and associated outcome measures will be sent to OTs/PTs working with children living with AMC. Third, information gathered from the scoping review and the survey will be gathered and practice guidelines for each priority will be put forward. These recommendations will be shared with an advisory board comprised of youth and adults with AMC and caregivers to validate the practice guidelines using an iterative process. Practice guidelines will then be translated into a comprehensive and clear format, easy for clinicians to consult. Future steps include the evaluation of implementation of the practice guidelines in various clinical settings to tailor the guidelines, refine the dissemination plan and evaluate the impact of these guidelines on clinical practice, client-centeredness and satisfaction.

Expected contribution: The co-development of family-centered rehabilitation guidelines will promote evidence-based, coordinated services and communication among clinicians and families, and improve the care provided to children with AMC.
Abstract # 60
Measuring Efficacy of Physical Rehabilitation in Children and Adults with Arthrogryposis

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Introduction: When looking at the lifespan of individuals who have Arthrogryposis Multiplex Congenita (AMC), the ability to track functional change over time cannot be underestimated. This population can be genetically and phenotypically very different making it a challenge to have a cookbook approach to identifying outcome measures. When an individual who has AMC comes to physical or occupational therapy, the hope is to make a functional change for the individual and the challenge is to make that change and be able to track it. The reason to measure is based on need to qualify for services, baseline and end point during an episode of care, as well as periodically to track abilities along a lifespan. If in a multidisciplinary clinic environment, it is helpful to do data collection at least on an annual basis. Outcome measures help to qualify one for service, track change over time and adapt programing based on changing needs. It allows comparing the individual to oneself, to others who have AMC, to others with a similar type of AMC, as well as the general population.

Methods: Review some of the testing tools presently in use including FMS, PROMIS, GMFM, Observational Gait Score, TUG, timed tasks, sit and reach with positives and negatives related to them.

Results: No tool grows with the individual from childhood through adulthood. PROMIS has been helpful to tease out areas of need. Scoring ambulation is helpful for tracking ability and change. Many standardized test do not tease out functional skills for this population. Timed tasks help to track present skill and change over time.

Conclusions: Gaining consensus on outcomes measures allows for future multicenter research across the lifespan.
Abstract # 61
Serial Derotational Spinal Casting in Early Onset Scoliosis Secondary to Arthrogryposis Multiplex

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Introduction: Early-onset scoliosis has been reported in 20-70% of individuals diagnosed with arthrogryposis multiplex congenita (AMC). As such, a range of growth-sparing treatments may be employed in order to delay or avert the necessity of definitive spinal arthrodesis. This study aims to evaluate the safety and efficacy of serial derotational spinal casting (SDSC) to cure or control curvature in patients with scoliosis secondary to AMC.

Methods: Data were retrospectively collected on a single-center cohort of 14 consecutive patients receiving SDSC for treatment of early-onset scoliosis associated with AMC between December 2002 and March 2018. Treatment was initiated once patients reached sitting-age and had a curve of ≥25°, and all patients had a minimum 1 year of follow-up from initial treatment. Statistical analysis was performed with Pearson’s correlations and student-t tests.

Results: Eight of the 14 patients in the series were male, average age of first spine casting 28 months (range 10–57), with an average curve size of 51° (range 33°–77°). After an average of 9 casts (range 4–20) and 35 months since first cast (range 13–75), the average coronal Cobb angle was 51° (p=0.96) (range 6°–97°). Kyphosis changed from 46° pretreatment to 43° at follow-up. No complications were documented in the series. A significant improvement in coronal Cobb angles was observed subsequent to application of first cast (p=0.004). Of note, the average T1-S1 height increased significantly between pretreatment (210mm) and last follow-up (263mm) (p<0.0001), average rate of 24mm/year (range -1 – 45 mm). No other significant differences were observed between any intervals with respect to clinical or radiographic variables. At follow-up, only 1 patient’s curve was cured, 2 patients were still undergoing casting, one patient died from unrelated causes, and one was lost to followup. The remaining 9 were managed in braces (average 55° curve and 79 months of age), with 1 undergoing a spinal fusion, and 5 receiving an expandable spine implant (average 78° curve and 72 months of age), 2 of which were unilateral constructs for severe C-shaped curves.

Conclusions: Scoliosis associated with AMC in infants and young children can rarely be cured by SDSC, but is an important treatment modality for controlling or arresting progression of spinal deformity, allowing for several years of growth prior to advancing to invasive treatment measures.
Abstract # 62
Magnetically Controlled Growing Rods in Scoliosis Secondary to Arthrogryposis Multiplex Congenita

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Introduction: Early-onset spinal deformity has been reported in 20–70% of children diagnosed with arthrogryposis multiplex congenita (AMC). Non-fusion spinal instrumentation (NFSI) allows for curve control during growth, and magnetically controlled growing rods (MCGRs) allow for non-invasively performed lengthenings. This study evaluates the efficacy of MCGRs in patients with scoliosis secondary to AMC.

Methods: Radiographic and clinical data were retrospectively reviewed on 7 consecutive patients receiving MCGRs for treatment of AMC associated scoliosis with 6 months minimum followup. All patients were ≥4 years of age at time of surgery. Indications were a curve of ≥50° in a cast or brace, and/or respiratory compromise related to brace/cast treatment. Four patient had bilateral rod constructs, and 3 had severe spine and chest deformities, allowing for only a unilateral rod on the curve concavity. Rods were lengthened every 2 months for unilaterals and every 3 for unilaterals.

Results: Average age at surgery was 88 months (range 52–139). Average curve sizes pre- and post-operatively were 75° (62°–97°) and 48° (27°–64°) respectively (p=0.004), but stayed stable at 42° (34°–61°)(p=0.22) during 23 months of follow-up. Bilateral rod constructs underwent an average of 9 lengthening for a total of 21mm (1–55mm), unilateral rods gained 30mm over 7 lengthenings. T1-S1 height change pre- to post-operative (260mm to 275mm) was not significant (p=0.067), but when compared to followup (289mm) both changed significantly (p=0.039 and p=0.036 respectively). Kyphosis changed little (from 48° preoperatively to 56° at follow up, p=0.102), except one unilateral rod patient whose proximal kyphosis increased from 81° to 109°. A 14 year old girl had her MCGRs substituted with solid rods without formal fusion after 3 years and 20mm total distraction; her curve improved from 62° initially to 34° post-treatment. The complications were one patient who developed a latent hardware infection secondary to an infected renal calculus, treated with antibiotics, another who died of a viral infection 2 weeks post-operatively.

Conclusions: For growing children with AMC and associated scoliosis, MCGRs are an effective treatment modality for delaying or avoiding the need for spinal fusion. They are more effective treating coronal deformities than kyphosis. Many of these children have severely compromised health, necessitating a multidisciplinary care team before and after surgery.
Arthrogryposis Acumens

Arthrogryposis Acumen #1
External Tibial Torsion Accompanying Arthrogrypotic Clubfoot Deformities

Harold J. P. van Bosse
Shriners Hospital for Children – Philadelphia, Philadelphia, PA, USA

Many of the children with AMC and clubfoot deformities have marked external tibial torsion, often seen in the severe or frequently relapsing clubfoot. Derotational osteotomies may be indicated in certain cases to help prevent relapses.

Arthrogryposis Acumen #2
The Power of the Wedge

Harold J.P. van Bosse
Shriners Hospital for Children – Philadelphia, Philadelphia, PA, USA

Children with arthrogryposis often have hip and knee contractures, as well as foot deformities, making foot-flat standing impossible. Coupled with a very delicate ability to balance, many are unable to stand or walk independently. But a simple heel wedge can help them align their hips over their heels, allowing for functional balance, and independent standing or ambulation.

Arthrogryposis Acumen #3
Standing balance: the Art of the Shoe Wedge

Maureen Donohoe
Nemours/Alfred I duPont Hospital for Children, Wilmington, DE, USA

Those with AMC using braces to support upright standing and walking alignment may have foot flat and level pelvis challenges due to contractures. If the brace cannot balance in the shoe, the person wearing the brace needs more energy to stand upright. Balance and biomechanical advantage for hip extension can be obtained with shoe wedges. Balance leg length and then predict heel wedge. Contractures predict wedge height. Knee flexion + plantar flexion + hip extension = $\times \degree / 10 = \text{cm heel height}$. 
Arthrogryposis Acumen #4
Prediction of Ambulation

Maureen Donohoe
Nemours/Alfred I duPont Hospital for Children, Wilmington, DE, USA

Based on strength of, gluteals, quadriceps and upper body, as well as trunk alignment, level of bracing and potential for ambulation into adulthood is predictable. Conceptual framework for decision making will be provided.

Arthrogryposis Acumen #5
Lower Limb Amyoplasia Presenting with Severe Scoliosis and Unusual Skin Dimpling

Ted Matthew Padilla Evangelista
University of Santo Tomas Hospital, Manilla, Philippines

Amyoplasia, the classical form of arthrogryposis. There are only 85 cases of lower limb-only Amyoplasia identified in literature, and there are no published cases from the Philippines. We discuss a rare case of lower limb-only Amyoplasia presenting with unique features of severe scoliosis (managed with spine corrective surgery) and multiple symmetrical bilateral gluteal dimpling in a Filipino child.

Arthrogryposis Acumen #6
Maintaining a Delicate Balance

Michelle A. James
Shriners Hospital for Children - Northern California, Sacramento, CA, USA

Children with arthrogryposis use alternative strategies to accomplish tasks and achieve mobility. For example, they may cross arms at the wrists and use shoulder adduction and elbow extension to grasp, or bear weight on the dorsum of their hand to scoot. When planning surgical reconstruction that changes the position of the elbow or wrist, the surgeon and therapist must carefully observe the child’s strategies, and strive to maintain their delicate balance of position and function.
**Arthrogryposis Acumen #7**

**Tendon Transfer in the Arthrogrypotic Thumb**

Giorgio Pajardi  
Ospedale San Giuseppe - Università di Milano – MultiMedica, Milan, Italy

First ray involvement in the hand implies thumb in palm, MP joint flexion and 1° web contracture. Extensor tendons are present but usually extension is poor at MP level. We propose a double reinforce of extensor apparatus by rerouting EPL in a more radial position on the dorsal hood of the MP. Moreover we planned a tendon transfer of a slips of APL on the EPB, usually present but rudimental, to improve MP extension.

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**Arthrogryposis Acumen #8**

**The Thumb in AMC**

Dan A. Zlotolow  
Shriners Hospital for Children – Philadelphia, Philadelphia, PA, USA

Children with Distal Arthrogryposis have primarily MP joint flexion contractures and CMC extension contractures of the thumb. Amyoplasia typically results in a flexed CMC joint with hypo-pronation of the thumb. The MP joint is usually straight or slightly flexed. Tips for improving thumb function in Amyoplasia will be discussed.

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**Arthrogryposis Acumen #9**

**Does Age Matter?**

Dan A. Zlotolow  
Shriners Hospital for Children – Philadelphia, Philadelphia, PA, USA

The age at which surgery is considered varies greatly among clinicians, with some operating on multiple joints as early as 1 year of age and others adopting a more guarded approach. We will consider the evidence for the timing of surgery and prioritize surgical interventions.
Patients with Arthrogryposis receive treatments that can cause an increase in fear, anxiety, and long-term implications of negative medical experiences. Certified Child Life Specialists are trained medical professionals who work as part of the multidisciplinary team and focus on the psychosocial needs of patients and families. This presentation aims to equip attendees with the awareness of pediatric patient vulnerabilities and interventions to support patients, siblings, caregivers, and staff.
NOTES
Join us for our

14th Annual
Arthrogryposis Multiplex Congenita
Support, Inc. Conference
Norfolk, Virginia
July 3-6, 2019

We hope that you can join us for the Annual AMCSI Conference which provides an opportunity for people affected by arthrogryposis multiplex congenita to gather and exchange ideas and support. During the three-day event, families and medical professionals participate in a variety of sessions and children’s activities providing different kinds of information to meet the needs of our ever-growing AMC family. Our planning committee is reaching out to medical professionals and speakers from around the country. Would you be interested in being a speaker?

Arthrogryposis Multiplex Congenita Support, Inc., started as an online support forum; therefore, we believe that these support sessions are just as important as those that offer medical information. Watch for our notices on Facebook and our website in early spring.

For more information visit us at amcsupport.org

Hilton Philadelphia at Penn’s Landing
Philadelphia, Pennsylvania, USA
September 24–26, 2018

3rd international symposium
on arthrogryposis

Jointly Provided by: The International Study Group on Arthrogryposis and Lewis Katz School of Medicine at Temple University

ABSTRACTS

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