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## Interventions and Management

### 1. Short-term Efficacy of Hand-Arm Bimanual Intensive Training on Upper Arm Function in Acute Stroke Patients: A Randomized Controlled Trial.

Meng G, Meng X, Tan Y, Yu J, Jin A, Zhao Y, Liu X.

Front Neurol. 2018 Jan 19;8:726. doi: 10.3389/fneur.2017.00726. eCollection 2017.

**BACKGROUND:** Rehabilitation training during the acute phase of stroke (<48 h) markedly improves impaired upper-limb movement. Hand-arm bimanual intensive training (HABIT) represents an intervention that promotes improvements in upper extremity function in children with cerebral palsy. This study repurposed HABIT in acute stroke patients and assessed recovery of upper extremity function when compared with a conventional rehabilitation program (CRP). **METHODS:** In a randomized trial, 128 patients with acute stroke were assigned to the HABIT or the CRP groups. The primary endpoint was clinical motor functional assessment that was guided by the Fugl-Meyer motor assessment (FMA) and outcomes of the action research arm test (ARAT). The secondary endpoint was an improved neurophysiological evaluation according to the motor-evoked potential amplitude (AMP), resting motion threshold (RMT), and central motor conduction time (CMCT) scores over the 2-week course of therapy. In both groups, scores were evaluated at baseline, 1 week from commencing therapy, and post-therapy. **RESULTS:** After 2 weeks, the HABIT group showed improved scores as compared the CRP group for FMA ( $51.7 \pm 6.44$  vs.  $43.5 \pm 5.6$ ,  $P < 0.001$ ), ARAT ( $34.5 \pm 6.2$  vs.  $33.3 \pm 6.3$ ,  $P = 0.022$ ), and AMP ( $1.1 \pm 0.1$  vs.  $1.0 \pm 0.1$ ,  $P < 0.001$ ). However, CMCT ( $8.6 \pm 1.0$  vs.  $9.1 \pm 0.6$ ,  $P = 0.054$ ) and RMT ( $55.3 \pm 4.2$  vs.  $57.5 \pm 4.1$ ,  $P = 0.088$ ) were similar when comparing between groups. **CONCLUSION:** HABIT significantly improved motor functional and neuro-physiological outcomes in patients with acute stroke, which suggested that HABIT might represent an improved therapeutic strategy as compared CRP.

[PMID: 29403422](#)

### 2. Evolution of self-care and functional mobility after single-event multilevel surgery in children and adolescents with spastic diplegic cerebral palsy.

Dequeker G, Van Campenhout A, Feys H, Molenaers G.

Dev Med Child Neurol. 2018 Feb 8. doi: 10.1111/dmcn.13683. [Epub ahead of print]

**AIM:** To explore the evolution of self-care and functional mobility after multilevel surgery in children and adolescents with spastic diplegic cerebral palsy and to identify factors affecting these outcomes. **METHOD:** Thirty-four participants (22 males, 12 females) were evaluated before surgery, and at 2 months, 6 months, 1 year, 18 months, and 2 years after surgery. Self-care was assessed with the Pediatric Evaluation of Disability Inventory Dutch edition. The Mobility Questionnaire<sup>47</sup> (MobQues<sup>47</sup>) and Functional Mobility Scale (FMS) were used to measure functional mobility. **RESULTS:** All outcomes revealed a significant decrease 2 months after single-event multilevel surgery (SEMLS) ( $p$ -value between  $<0.001$  and  $0.02$ ) followed by a significant increase at 6 months ( $p < 0.001$  and  $p = 0.045$ ). Between 6 months and 1 year, a significant increase was also revealed for Mobques<sup>47</sup> ( $p < 0.001$ ), FMS ( $p \leq 0.008$ ), and the Pediatric Evaluation of Disability Inventory Functional Skills Scale (PEDI-FSS) ( $p = 0.001$ ). Improvement continued until 18 months for the PEDI-FSS. Initial score, initial muscle strength, Gross Motor Function Classification System level, age, and number of surgical interventions significantly influenced

time trends for self-care and/or functional mobility. INTERPRETATION: Most preoperative scores are regained at 6 months after SEMLS. Further improvement is seen until 18 months. Participants with a higher functional level before surgery will temporarily lose more than participants with lower initial functional ability, but they also fast regain their function. WHAT THIS PAPER ADDS: Self-care and functional mobility decrease significantly in the first months after single-event multilevel surgery (SEMLS). Six months after SEMLS most preoperative scores are regained. Impact of SEMLS is more pronounced for functional mobility than for self-care. Muscle strength and functionality at baseline are important influencing factors on the evolution after SEMLS.

[PMID: 29417569](#)

### 3. Management of tone in children and young people with cerebral palsy. What is the evidence?

Carr LJ.

Dev Med Child Neurol. 2018 Feb 8. doi: 10.1111/dmcn.13695. [Epub ahead of print]

[This commentary is on the original article by Fehlings et al.]

[PMID: 29417558](#)

### 4. Minimally Invasive Surgery for Neuromuscular Scoliosis: Results and Complications in a Series of one hundred Patients.

Miladi L, Gaume M, Khouri N, Johnson M, Topouchian V, Glorion C.

Spine (Phila Pa 1976). 2018 Feb 6. doi: 10.1097/BRS.0000000000002588. [Epub ahead of print]

STUDY DESIGN: A retrospective review. OBJECTIVES: To report the results of an alternative technique using a minimally invasive fusionless surgery. The originality is based on the progressive correction of the deformities with proximal and distal fixation and on the reliability of the pelvic fixation using ilio-sacral screws on osteoporotic bones. SUMMARY OF BACKGROUND DATA: Spinal deformities are common in neuromuscular diseases. Conventional treatment involves bracing, followed by spinal instrumented fusion. Growing rod techniques are increasingly advocated but have a high rate of complications. METHODS: The technique relies on a bilateral double rod sliding construct anchored proximally by 4 hooks claws and distally to the pelvis by ilio-sacral screws through a minimally invasive approach. 100 patients with neuromuscular scoliosis underwent the same fusionless surgery extended from T1 to the pelvis. The average age at initial surgery was 11+6y. Diagnoses included cerebral palsy (61), spinal muscular atrophy (22), muscular dystrophy (10), and other neurological etiologies (7). Cobb angle and pelvic obliquity were measured before and after initial surgery, and at final follow-up. Complications were reviewed. RESULTS: At latest follow-up 3+9 y (range 2y - 6+3y), the mean Cobb angle improved from 89° to 35° which corresponds to 61% correction. Mean pelvic obliquity improved from 29° to 5° which corresponds to 83% correction. Mean T1-S1 length increased from 30.02cm to 37.28cm. Mean preoperative hyper kyphosis was reduced from 68.44° to 33.29°. Complications occurred in 22 patients including mechanical complications (12) and wound infections (16). No arthrodesis was required at last follow-up. CONCLUSION: This original fusionless technique is safe and effective, preserving spinal and thoracic growth. It provides a significant correction of spinal deformities and pelvic obliquity with a reduced complications rate. The strength and stability of this modular construct over time allow the avoidance of final arthrodesis.

[PMID: 29419720](#)

### 5. Reliability analysis of the Korean version of the trunk control measurement scale in cerebral palsy.

Heo JY, Shin HK.

J Phys Ther Sci. 2018 Jan;30(1):1-4. doi: 10.1589/jpts.30.1. Epub 2018 Jan 27.

[Purpose] The purpose of this study was to translate the Trunk Control Measurement Scale into a Korean version and to analyze the intra- and inter-rater reliability. [Subjects and Methods] Fifteen children with spastic cerebral palsy and four physical therapists with over 10 years of clinical experience participated in this study. A Korean-American physical therapist translated the trunk control measurement scale from English into a Korean version. Four physical therapists viewed the video data of 15 children and scored each child's trunk control measurement scale performance on seven separate days. Four testers analyzed the test-retest reliability and inter-rater reliability using the intra-class correlation coefficient. [Results] Intra-class correlation coefficients for test-retest and inter-rater reliability for the Korean trunk control measurement scale showed

significantly high reliability in all testers and sublevels. [Conclusion] The Korean version of the measurement scale is a reliable and suitable instrument for assessing trunk control in individuals with cerebral palsy in Korea.

[PMID: 29410555](#)

## **6. Preoperative hip rotation moments do not predict long-term development after femoral derotation osteotomy in children with cerebral palsy.**

Braatz F, Dreher T, Wolf SI, Niklasch M.

Gait Posture. 2018 Jan 26;61:215-219. doi: 10.1016/j.gaitpost.2018.01.017. [Epub ahead of print]

**BACKGROUND:** Femoral derotation osteotomy (FDO) is the standard treatment for internal rotation gait (IRG) in children with cerebral palsy (CP) although high rates of recurrence have been reported recently. Various factors associated with recurrence could be identified, but no predictor named. **RESEARCH QUESTIONS:** Does FDO lead to a change of internal transversal hip moments? Are preoperative internal transversal hip moments a predictor for recurrence of IRG? **METHODS:** 41 children with spastic bilateral CP and 72 limbs that received a FDO ( $10.4 \pm 2.7$  years at surgery) were included retrospectively. Kinematic data were analyzed pre- ( $2 \pm 3$  months), postoperatively ( $12 \pm 3$  months) and at long-term follow-up (at least five years postoperatively;  $84 \pm 13$  months), internal transversal hip moments were analyzed pre- and postoperatively. **RESULTS:** The maximum peaks of the internal hip rotation moment during loading response decreased significantly ( $p = 0.003$ ). The minimum during the second half of the stance phase increased significantly ( $p = 0.004$ ) and the initially internal externally rotating moment changed to an internal internally rotating moment. No correlation between changes in hip rotation from postoperatively to the long-term follow-up and the preoperative internal hip rotation moment could be identified. **SIGNIFICANCE:** FDO leads to changes in internal hip rotation moments. Preoperative internal hip rotation moments can't be used as predicting factor for recurrence of IRG. The data suggest, that recurrence of IRG depends less on patient specific motion patterns, but more on the time point of surgery and the therapy of all concomitant deformities during SEMLS.

[PMID: 29413787](#)

## **7. Selective dorsal rhizotomy in cerebral palsy: Selection criteria and postoperative physical therapy protocols.**

Nicolini-Panisson RD, Tedesco AP, Folle MR, Donadio MVF.

Rev Paul Pediatr. 2018 Jan 15:0. doi: 10.1590/1984-0462/;2018;36;1;00005. [Epub ahead of print]

**OBJECTIVE:** To identify selection criteria for selective dorsal rhizotomy (SDR) in cerebral palsy, to analyze the instruments used for evaluation, and to describe the characteristics of physical therapy in postoperative protocols. **DATA SOURCES:** Integrative review performed in the following databases: SciELO, PEDro, Cochrane Library, and PubMed. The terms in both Portuguese and English for "cerebral palsy", "selective dorsal rhizotomy", and "physical therapy" were used in the search. Studies whose samples enrolled individuals with cerebral palsy who had attended physical therapy sessions for selective dorsal rhizotomy according to protocols and describing such protocols' characteristics were included. Literature reviews were excluded and there was no restriction as to period of publication. **DATA SYNTHESIS:** Eighteen papers were selected, most of them being prospective cohort studies with eight-month to ten-year follow-ups. In most studies, the instruments of assessment encompassed the domains of functions, body structure, and activity. The percentage of posterior root sections was close to 50%. Primary indications for SDR included ambulatory spastic diplegia, presence of spasticity that interfered with mobility, good strength of lower limbs and trunk muscles, no musculoskeletal deformities, dystonia, ataxia or athetosis, and good cognitive function. Postoperative physical therapy is part of SDR treatment protocols and should be intensive and specific, being given special emphasis in the first year. **CONCLUSIONS:** The studies underline the importance of appropriate patient selection to obtain success in the SDR. Postoperative physical therapy should be intensive and long-term, and must necessarily include strategies to modify the patient's former motor pattern.

[PMID: 29412426](#)

## **8. Use of caudal epidural catheter in a child with cerebral palsy with prior posterior spine (T1-sacrum) fusion.**

Dixit D, Theroux MC, Dabney KW, Miller F.

Indian J Anaesth. 2018 Jan;62(1):80-82. doi: 10.4103/ija.IJA\_562\_17.

[No abstract available]

[PMID: 29416157](#)

## 9. Physical Activity in Adolescents and Young Adults with Cerebral Palsy.

Waltersson L, Rodby-Bousquet E.

Biomed Res Int. 2017;2017:8080473. doi: 10.1155/2017/8080473. Epub 2017 Dec 20.

The aim of this study was to examine the level of physical activity in adults with cerebral palsy (CP) and to analyse its relationship with physical activity as adolescents, pain, and gross motor function. A prospective cohort study was performed using data from the Swedish National CP Registry (CPUP) for all 129 individuals born in 1991-1993 living in Skåne and Blekinge who reported to CPUP at 14-16 years of age. Physical activity as adult was analysed relative to physical activity as adolescents, pain, and the Gross Motor Function Classification System (GMFCS). Seventy-one individuals at GMFCS I-V were followed up as adults and included in the analyses. Of these, 65% were physically active, but only 56% performed physical activity at least once a week. Their physical activity as adults differed relative to their physical activity as adolescents ( $p = 0.011$ ) but not to pain or GMFCS. Being physically active as an adolescent doubled the probability of being active as an adult (OR 2.1;  $p = 0.054$ ), indicating that physical activity in adults with CP is related to their physical activity as adolescents. Therefore, interventions to increase physical activity among adolescents with CP are likely also to improve physical activity in adulthood.

[PMID: 29423412](#)

## 10. Outcome of medial hamstring lengthening in children with spastic paresis: A biomechanical and morphological observational study.

Haberfehlner H, Jaspers RT, Rutz E, Harlaar J, van der Sluijs JA, Witbreuk MM, van Hutten K, Romkes J, Freslier M, Brunner R, Becher JG, Maas H, Buizer AI.

PLoS One. 2018 Feb 6;13(2):e0192573. doi: 10.1371/journal.pone.0192573. eCollection 2018.

To improve gait in children with spastic paresis due to cerebral palsy or hereditary spastic paresis, the semitendinosus muscle is frequently lengthened amongst other medial hamstring muscles by orthopaedic surgery. Side effects on gait due to weakening of the hamstring muscles and overcorrections have been reported. How these side effects relate to semitendinosus morphology is unknown. This study assessed the effects of bilateral medial hamstring lengthening as part of single-event multilevel surgery (SEMLS) on (1) knee joint mechanics (2) semitendinosus muscle morphology and (3) gait kinematics. All variables were assessed for the right side only. Six children with spastic paresis selected for surgery to counteract limited knee range of motion were measured before and about a year after surgery. After surgery, in most subjects popliteal angle decreased and knee moment-angle curves were shifted towards a more extended knee joint, semitendinosus muscle belly length was approximately 30% decreased, while at all assessed knee angles tendon length was increased by about 80%. In the majority of children muscle volume of the semitendinosus muscle decreased substantially suggesting a reduction of physiological cross-sectional area. Gait kinematics showed more knee extension during stance (mean change  $\pm$  standard deviation:  $34 \pm 13^\circ$ ), but also increased pelvic anterior tilt (mean change  $\pm$  standard deviation:  $23 \pm 5^\circ$ ). In most subjects, surgical lengthening of semitendinosus tendon contributed to more extended knee joint angle during static measurements as well as during gait, whereas extensibility of semitendinosus muscle belly was decreased. Post-surgical treatment to maintain muscle belly length and physiological cross-sectional area may improve treatment outcome of medial hamstring lengthening.

[PMID: 29408925](#)

## 11. A heel-strike real-time auditory feedback device to promote motor learning in children who have cerebral palsy: a pilot study to test device accuracy and feasibility to use a music and dance-based learning paradigm.

Pitale JT, Bolte JH 4th.

Pilot Feasibility Stud. 2018 Jan 29;4:42. doi: 10.1186/s40814-018-0229-0. eCollection 2018.

**BACKGROUND:** Cerebral palsy (CP) is a developmental disorder of movement and posture that occurs due to damage to the developing nervous system. As part of therapy, wearable sensors that trigger interactive feedback may provide multi-sensory guidance and motivation. A prototype of a heel-strike real-time feedback system has been developed which records the number of heel strikes during gait and indicates successful heel contact through real-time auditory feedback. The first aim of this feasibility study was to test the prototype accuracy. Since the end user for this device is a child, the device should be esthetically appealing and sufficiently motivating for children to perform repetitive challenging therapeutic movements. The second aim of this study was to collect feedback from the subjects with regard to the device usability and understand if the bell sound used as feedback used was motivating enough for children to continue using the prototype. This would help us in developing the next generation of the device. **METHODS:** The prototype was tested with typically developing children and children who have CP. The accuracy in detecting heel strikes was calculated. As part of the study, the subjects were also asked questions to test the device compliance and acceptability of the musical beats with the pediatric population. **RESULTS:** The device accuracy in

identifying heel strikes is 97.44% (95% CI 96.31, 98.88%). The subjects did not show any hesitation to put on the device and the sound feedback motivated them to move. Based on this pilot study, a minimum age limit of 5 years is appropriate and the intervention study should be conducted for no more than 30 min per week. **CONCLUSIONS:** The pilot study showed that a main study can be conducted to test auditory feedback as an intervention to promote motor learning in children who have cerebral palsy. No adverse event or safety issues were reported in the feasibility study.

[PMID: 29423260](#)

## 12. Quality of life and habitual physical activity in children with cerebral palsy aged 5 years: A cross-sectional study.

Keawutan P, Bell KL, Oftedal S, Davies PSW, Ware RS, Boyd RN.

Res Dev Disabil. 2018 Jan 29;74:139-145. doi: 10.1016/j.ridd.2018.01.008. [Epub ahead of print]

**OBJECTIVE:** To compare quality of life (QOL) according to ambulatory status and to investigate association with habitual physical activity (HPA) in children with cerebral palsy (CP) aged 5 years. **METHODS:** Fifty-eight participants were classified using Gross Motor Function Classification System (GMFCS) as level I = 33, II = 8, III = 6, IV = 3 and V = 8 and assessed for motor function using 66-item Gross Motor Function Measure (GMFM-66). Participants wore an ActiGraph® triaxial accelerometer for 3 days to measure HPA. Parents completed the parent proxy Cerebral Palsy Quality of Life questionnaire for Children (CP QOL-Child). Linear regression analyses were performed. **RESULTS:** Ambulant children with CP (GMFCS I-III) had better parent-reported QOL than non-ambulant children (GMFCS IV-V) in domains of feelings about functioning (mean difference (MD) = 20.0; 95% confidence interval (CI) = 11.7, 28.2), participation and physical health (MD = 14.5; 95%CI = 4.7, 24.4), and emotional well-being and self-esteem (MD = 12.5; 95%CI = 4.8, 20.1). HPA was not associated with QOL domains after controlling for motor function. GMFM scores accounted for 39% of variation for feelings about functioning domain (MD = 0.4; 95%CI = 0.2, 0.6). **CONCLUSIONS:** In children with CP aged 5 years, HPA was not associated with parent-reported QOL. Gross motor function contributed to QOL domains of feelings about functioning.

[PMID: 29413428](#)

## 13. Running in people with cerebral palsy: A systematic review.

Chappell A, Gibson N, Morris S, Williams G, Allison GT.

Physiother Theory Pract. 2018 Feb 6:1-16. doi: 10.1080/09593985.2018.1434846. [Epub ahead of print]

**BACKGROUND:** Running is a fundamental motor skill which is important for participation in recreational activities throughout the lifespan. **AIM:** To identify, appraise, and synthesize the evidence related to running in people with cerebral palsy (CP). **METHOD:** A search of electronic databases was conducted in April 2016. Articles were reviewed by two assessors and had to meet the following criteria: 1) population included people with CP; and 2) included information about running. Articles were assessed for quality using the Checklist for Measuring Study Quality by Downs and Black. **RESULTS:** Of 2607 articles initially identified, 56 underwent full-text review and 17 were included. Quantitative analysis was not possible. The quality of descriptive studies was fair-good, while intervention studies scored poor-good. Four of the 12 descriptive studies reported kinematic and kinetic data. Five intervention studies included running as an outcome measure, although running was not the focus of intervention. A few studies showed that sprint speed, agility, and running efficiency are impaired in people with CP, but mechanisms underlying these impairments have yet to be reported. **INTERPRETATION:** Research on running in people with CP is limited, methodology and findings are inconsistent and studies are generally fair quality. Further investigation is warranted.

[PMID: 29405796](#)

## 14. Validation of Manual Muscle Testing (MMT) in children and adolescents with cerebral palsy.

Manikowska F, Chen BP, Józwiak M, Lebedowska MK.

NeuroRehabilitation. 2018;42(1):1-7. doi: 10.3233/NRE-172179.

**BACKGROUND:** Weakness is a major impairment in many movement disorders, including cerebral palsy (CP), which presents as a decrease in muscle strength. Manual muscle testing (MMT) is very popular in clinical practice, however it has many limitations. **OBJECTIVE:** (1) Whether maximum voluntary contraction (MVC) measures differ across clinical MMT groups; (2) Whether an association exists between clinical MMT score groups and instrumental MVC measures. **METHODS:** Twenty-one participants with spastic CP were recruited (11 females and 10 males; age = 13.46 ± 3.62 years). To achieve the aims

of the study, we investigated the relationship between qualitative (MMT) and instrumental (MVC) measures of knee flexor muscles' strength in patients with CP. RESULTS: MVC values increased somewhat proportionally with increasing MMT score group ( $p=0.032$ ,  $MS=207.54$ ,  $F=3.75$ ). The differences in MVC values was only statistically significant between score groups 3 and 5. A weak correlation ( $R=0.4$ ,  $MVC=-2.54+4.50$  MMT,  $p<0.01$ ) was found between measured MVCs and the MMT score groups. CONCLUSIONS: In pediatric research studies, instrumental MVC should be preferred over MMT scoring. Also, MMT score groups higher than 3 should be modified in clinical testing of children and adolescents with cerebral palsy.

[PMID: 29400673](#)

### **15. A new clinical tool for measuring limited selective voluntary motor control in cerebral palsy.**

Rudisch J.

Dev Med Child Neurol. 2018 Feb 6. doi: 10.1111/dmcn.13692. [Epub ahead of print]

[No abstract available]

[PMID: 29405295](#)

### **16. Using the International Classification of Functioning, Disability, and Health Model to Gain Perspective of the Benefits of Yoga in Stroke, Multiple Sclerosis, and Children to Inform Practice for Children with Cerebral Palsy: A Meta-Analysis.**

Veneri D, Gannotti M, Bertuccio M, Fournier Hillman SE.

J Altern Complement Med. 2018 Feb 6. doi: 10.1089/acm.2017.0030. [Epub ahead of print]

OBJECTIVE: Research pertaining to yoga and children with cerebral palsy (CP) is negligible. The primary purpose of this study was to determine the domains of the International Classification of Functioning, Disability, and Health (ICF) model and levels of evidence for yoga and adults with stroke and multiple sclerosis (MS), and children. A secondary purpose was to decide whether any inferences could be made for children with CP. DESIGN: This study included a meta-analysis. INTERVENTIONS: A systematic review was performed of yoga and said populations. Outcome measures were categorized according to the ICF model domains of body structures and function, activity, and quality of life. Effect sizes (ESs) were calculated by using Cohen's  $d$ . Since there were few commonalities among outcome measures and reporting of outcomes within and among diagnostic groups, direct comparisons of ESs were difficult. Hence, we chose to evaluate the impact of yoga as compared with the control group or other physical exercise by using a General Linear Mixed Model. RESULTS: There were 5 yoga studies with stroke, 15 with MS, and 12 with children. Studies with children used outcomes related to body structure and function, whereas those with stroke and MS used outcomes across all three domains of the ICF. ESs varied from negligible to medium for stroke, from negligible to large for MS and children. CONCLUSIONS: The findings of this meta-analysis indicate that yoga is no better or worse than other exercise modalities as a treatment intervention for adults with stroke and MS, and children. Group yoga classes are typically social environments that can contribute to increased physical progress and feelings that contribute to quality of life, which may benefit individuals with CP. More research on yoga and particularly in children and adults with CP would yield valuable information for creating effective and safe yoga programs with a rich array of benefits.

[PMID: 29406768](#)

### **17. Optimization of fMRI methods to determine laterality of cortical activation during ankle movements of children with unilateral cerebral palsy.**

Hilderley AJ, Taylor MJ, Fehlings D, Chen JL, Wright FV.

Int J Dev Neurosci. 2018 Feb 1;66:54-62. doi: 10.1016/j.ijdevneu.2018.01.004. [Epub ahead of print]

Measurement of laterality of motor cortical activations may provide valuable information about lower limb control in children with unilateral cerebral palsy (UCP). Evidence from upper limb research suggests that increased contralateral activity may accompany functional gains. However, lower limb areas of activation and associated changes have been underexplored due to challenges with imaging motor cortical leg representations. In this study, methods for a task-based functional magnetic resonance imaging (fMRI) ankle dorsiflexion paradigm were refined with three pilot groups of participants: (i) adults ( $n = 5$ ); (ii) typically developing (TD) children ( $n = 5$ ) and; (iii) children with UCP ( $n = 4$ ). Parameters of experimental design, task resistance, reproducibility, and pre-scan procedures were tested/refined using a staged development approach with additions or changes introduced if image quality did not meet pre-defined standards. When image quality was acceptable for two

consecutive participants, the next participant group was recruited to test/refine the next parameter. The final paradigm involved an event-related design of a single dorsiflexion movement against individualized resistance, with two runs per leg. It included a pre-scan session to increase child comfort and determine task resistance. This paradigm produced valid data for laterality index (LI) calculations to determine the ratio of activity in each hemisphere. Ventricle and lesion masks were used in non-linear image registration, and individual thresholds were used for extent-based LI calculations. LI of dominant ankle movements were contralateral ( $LI \geq +0.2$ ) for TD children (mean  $LI = +0.89$ ,  $std = 0.27$ ) and children with UCP (mean  $LI = +0.86$ ,  $std = 0.26$ ). For the affected ankle of children with UCP, LI values indicated ipsilateral and/or contralateral activation (mean  $LI = +0.02$ ,  $std = 0.71$ , range  $-0.92$  to  $+1.00$ ). This fMRI paradigm will support investigations of cortical activation and mechanisms of skill improvement following lower limb interventions.

[PMID: 29413879](#)

### **18. Quantifying Effect of Onabotulinum Toxin A on Passive Muscle Stiffness in Children with Cerebral Palsy Using Ultrasound Shear Wave Elastography.**

Brandenburg JE, Eby SF, Song P, Bamlet W, Sieck GC, An KN.

Am J Phys Med Rehabil. 2018 Feb 5. doi: 10.1097/PHM.0000000000000907. [Epub ahead of print]

**OBJECTIVE:** A pilot study to longitudinally quantify effect of onabotulinum toxin A (BoNT-A) on passive muscle properties in children with cerebral palsy (CP) using ultrasound shear wave elastography (SWE). **DESIGN:** Prospective longitudinal cohort study **RESULTS:** Between 1 and 3 months post-BoNT-A, a significant improvement in the shear modulus of the lateral gastrocnemius was found at 10° plantar flexion (PF) ( $-7.57$  [ $-10.98$ ,  $-5.07$ ],  $p=0.02$ ) and 0° PF ( $-14.74$  [ $-18.21$ ,  $-9.38$ ],  $p=0.03$ ). There was a notable, but non-significant difference in shear modulus at 20° PF, 10° PF, 0° PF between pre-BoNT-A and 1 month post-BoNT-A. Pre-BoNT-A shear modulus was not significantly different than 3 months post-BoNT-A at all foot positions. No significant differences in ankle passive range of motion or spasticity were found. **CONCLUSION:** Despite no significant change in ankle range of motion or spasticity, SWE was able to detect a difference in lateral gastrocnemius passive muscle properties in children with CP following BoNT-A injections. The difference in passive muscle properties resolved by 3 months post-BoNT-A.

[PMID: 29406405](#)

### **19. Deep brain stimulation for dystonia due to cerebral palsy: A review.**

Elia AE, Bagella CF, Ferré F, Zorzi G, Calandrella D, Romito LM.

Eur J Paediatr Neurol. 2017 Dec 14. pii: S1090-3798(17)31936-0. doi: 10.1016/j.ejpn.2017.12.002. [Epub ahead of print]

Cerebral palsy (CP) is a heterogeneous group of syndromes that cause a non-progressive disorder of early onset, with abnormal control of movement and posture. Various aetiologies can cause the CP clinical spectrum, but all have a disruption of motor control in common. CP can be divided into four major types based on the motor disability: predominant spastic, dyskinetic, ataxic and mixed form. Dyskinetic CP (DCP) is the most common cause of acquired dystonia in children. The treatment of DCP is challenging because most individuals have mixed degrees of chorea, athetosis and dystonia. Pharmacological treatment is often unsatisfactory. Functional neurosurgery, in particular deep brain stimulation targeting the basal ganglia or the cerebellum, is emerging as a promising therapeutic approach in selected patients with DCP. We evaluated herein the effects of DBS on patients with DCP in a review of published patient data in the largest available studies.

[PMID: 29396170](#)

### **20. Pharmacological and neurosurgical interventions for managing dystonia in cerebral palsy: a systematic review.**

Fehlings D, Brown L, Harvey A, Himmelmann K, Lin JP, Macintosh A, Mink JW, Monbaliu E, Rice J, Silver J, Switzer L, Walters I.

Dev Med Child Neurol. 2018 Feb 6. doi: 10.1111/dmcn.13652. [Epub ahead of print]

**AIM:** To systematically review evidence for pharmacological/neurosurgical interventions for managing dystonia in individuals with cerebral palsy (CP) to inform a care pathway. **METHOD:** Searches included studies with a minimum of five participants with dystonia in CP receiving oral baclofen, benzodiazepines (clonazepam, diazepam, lorazepam), clonidine, gabapentin, levodopa, trihexyphenidyl, botulinum toxin, intrathecal baclofen (ITB), or deep brain stimulation (DBS). Evidence was classified according to American Academy of Neurology guidelines. **RESULTS:** Twenty-eight articles underwent data extraction: one levodopa, five trihexyphenidyl, three botulinum toxin, six ITB, and 13 DBS studies. No articles for oral

baclofen, benzodiazepines, clonidine, or gabapentin met the inclusion criteria. Evidence for reducing dystonia was level C (possibly effective) for ITB and DBS; level C (possibly ineffective) for trihexyphenidyl; and level U (inadequate data) for botulinum toxin. INTERPRETATION: For dystonia reduction, ITB and DBS are possibly effective, whereas trihexyphenidyl was possibly ineffective. There is insufficient evidence to support oral medications or botulinum toxin to reduce dystonia. There is insufficient evidence for pharmacological and neurosurgical interventions to improve motor function, decrease pain, and ease caregiving. The majority of the pharmacological and neurosurgical management of dystonia in CP is based on clinical expert opinion. WHAT THIS PAPER ADDS: Intrathecal baclofen and deep brain stimulation are possibly effective in reducing dystonia. Current evidence does not support effectiveness of oral medications or botulinum toxin to reduce dystonia. Evidence is inadequate for pharmacological/neurosurgical interventions impact on improving motor function, pain/comfort, and easing caregiving. The majority of the care pathway rests on expert opinion.

[PMID: 29405267](#)

## 21. Botulinum toxin treatment of pain syndromes -an evidence based review.

Safarpour Y, Jabbari B.

Toxicon. 2018 Jan 31. pii: S0041-0101(18)30031-X. doi: 10.1016/j.toxicon.2018.01.017. [Epub ahead of print]

This review evaluates the existing level of evidence for efficacy of BoNTs in different pain syndromes using the recommended efficacy criteria from the Assessment and Therapeutic Subcommittee of the American Academy of Neurology. There is a level A evidence (effective) for BoNT therapy in post-herpetic neuralgia, trigeminal neuralgia, and posttraumatic neuralgia. There is a level B evidence (probably effective) for diabetic neuropathy, plantar fasciitis, piriformis syndrome, pain associated with total knee arthroplasty, male pelvic pain syndrome, chronic low back pain, male pelvic pain, and neuropathic pain secondary to traumatic spinal cord injury. BoNTs are possibly effective (Level C -one class II study) for female pelvic pain, painful knee osteoarthritis, post-operative pain in children with cerebral palsy after adductor release surgery, anterior knee pain with vastus lateralis imbalance. There is a level B evidence (one class I study) that BoNT treatment is probably ineffective in carpal tunnel syndrome. For myofascial pain syndrome, the level of evidence is U (undetermined) due to contradicting results. More high quality (Class I) studies and studies with different types of BoNTs are needed for better understanding of the role of BoNTs in pain syndromes.

[PMID: 29409817](#)

## 22. Prevalence and factors associated with sleep disorders among children with cerebral palsy in Uganda; a cross-sectional study.

Munyumu K, Idro R, Abbo C, Kaddumukasa M, Katabira E, Mupere E, Kakooza-Mwesige A.

BMC Pediatr. 2018 Feb 5;18(1):26. doi: 10.1186/s12887-018-0996-z.

**BACKGROUND:** Sleep plays a prominent role in the growth and development of children. Children with cerebral palsy (CP) are more prone to sleep disorders (SDs) than their peers. Children with CP, have a higher prevalence of disorders involving; initiation and maintenance of sleep, sleep-wake transition, excessive sleepiness and arousal. These sleep disorders impact on the quality of life of these children. Despite, having a high prevalence of CP in Uganda, there is a paucity of data that focuses on sleep disorders in CP, including a lack of prevalence estimates of sleep breathing disorder (SBD) in CP. Understanding the prevalence and disorders of sleep within this population would help advise on the development of tailored interventions to address the needs of these children and improve their quality of life. This study determined the prevalence and associated factors of sleep disorders among children aged 2 - 12 years with cerebral palsy in Uganda. **METHODS:** This was a cross sectional study. All participants had a physical examination and screening with the Sleep Disturbances Scale for Children (SDSC) questionnaire to determine the prevalence of sleeps disorders. A total score (TS)  $\geq 51$  on the Sleep Disturbances Scale for Children was regarded as abnormal. **RESULTS:** A total of 135 participants were recruited. The prevalence of sleep disorders was 43/135 (32%) with 95% CI: (24.0-39.7). The most common type of sleep disorders was a disorder of initiating and maintaining sleep 37(27%). The factors associated with sleep disorders among children with cerebral palsy were bilateral spasticity ( $p = 0.004$ ); OR:(95%CI), 11.193: (2.1 - 59.0), lowest levels of gross motor function V ( $p < 0.001$ ); OR:(95%CI), 13.182: (3.7 - 47.0) or IV ( $p = 0.007$ ); OR:(95%CI), 12.921: (2.0 - 82.3), lowest level of manual ability V ( $p = 0.004$ ); OR:(95% CI), 11.162: (2.2 - 56.4) and presence of epilepsy ( $p = 0.011$ ); OR:(95%CI), 3.865: (1.4 - 10.9). **CONCLUSIONS:** The prevalence of sleep disorders among children with cerebral palsy in Uganda is high. Severe disability and presence of epilepsy were associated with sleep disorders among children with cerebral palsy.

[PMID: 29402250](#)

### 23. Nutritional and dental issues in patients with intellectual and developmental disabilities.

Ziegler J, Spivack E.

J Am Dent Assoc. 2018 Feb 1. pii: S0002-8177(17)31065-6. doi: 10.1016/j.adaj.2017.11.014. [Epub ahead of print]  
 BACKGROUND: People with intellectual and developmental disabilities are among the most disadvantaged and underserved groups of dental patients. Considerable health care disparities for this population have been identified, particularly oral and dental health as well as access to dental care services. People with Down syndrome and cerebral palsy have a variety of nutritional and dental considerations. CONCLUSIONS: These people have a higher prevalence of untreated caries and periodontal disease than the general population and may have higher rates of obesity, edentulism, and chronic oral and systemic diseases. Diet choices may affect the oral health and may play an important role in the systemic health of these people. Suggestions to improve and affect dietary intake are provided. CLINICAL IMPLICATIONS: Health issues within this population require a holistic approach to care. Concerns about oral health and diet must be addressed to support optimal health.

[PMID: 29397872](#)

### 24. The impact of breathing rate on the cardiac autonomic dynamics among children with cerebral palsy compared to typically developed controls.

Amichai T, Eylon S, Berger I, Katz-Leurer M.

Dev Neurorehabil. 2018 Feb 6:1-6. doi: 10.1080/17518423.2018.1434700. [Epub ahead of print]

PURPOSE: To describe the immediate effect of breathing rate on heart rate (HR) and heart rate variability (HRV) in children with cerebral palsy (CP) and a control group of typically developed (TD) age and gender-matched children. METHODS: Twenty children with CP at gross motor function classification system levels I-III and 20 TD children aged 6-11 participated in the study. HR was monitored at rest and during paced breathing with biofeedback. Respiratory measures were assessed by KoKo spirometry. RESULTS: Children with CP have lower spirometry and HRV values at rest compared to TD children. The mean reduction of breathing rate during paced breathing among children with CP was significantly smaller. Nonetheless, while practicing paced breathing, both groups reduced their breathing rate and increased their HRV. CONCLUSIONS: The results of the current work present the immediate effect of paced breathing on HRV parameters in CP and TD children. Further studies are needed to investigate the effect of long-term treatment focusing on paced breathing for children with CP.

[PMID: 29405815](#)

### 25. Subgrouping children with cerebral palsy from a broader perspective using two methods.

Jeevanantham D, Bartlett D.

Physiother Theory Pract. 2018 Feb 5:1-11. doi: 10.1080/09593985.2017.1422163. [Epub ahead of print]  
 The purpose was to develop two versions of a body function index in cerebral palsy (BFI-CP I and BFI-CP II) using two methods to explore the relationship and differences among them and the Gross Motor Function Classification System (GMFCS) and to explore the differences among subsets of the classifications that do not correspond to the ordinal levels of the GMFCS. Data on various measures from 405 children with CP between 18 months and 5 years of age were extracted from the Move & PLAY Study. The BFI-CP I was developed using a summing technique and the BFI-CP II was developed using cluster analysis. There was a strong correlation between the BFI-CP I and the GMFCS ( $r = 0.92$ ), between the BFI-CP II and the GMFCS ( $r = 0.93$ ), and between the BFI-CP I and II ( $r = 0.92$ ) all ( $p < 0.001$ ). There was also a significant difference between the BFI-CP I and the GMFCS ( $\chi^2 = 670.49$ ,  $df = 16$ ,  $p < 0.001$ ) and between the BFI-CP II and the GMFCS ( $\chi^2 = 685.57$ ,  $df = 16$ ,  $p < 0.001$ ). The findings of this study indicate that the two versions of the BFI-CP could be used as complementary methods in describing children with CP.

[PMID: 29400608](#)

### 26. Comparing Levels of Mastery Motivation in Children with Cerebral Palsy (CP) and Typically Developing Children.

Salavati M, Vameghi R, Hosseini SA, Saeedi A, Gharib M.

Med Arch. 2018 Feb;72(1):41-45. doi: 10.5455/medarh.2018.72.41-45.

INTRODUCTION: The present study aimed to compare motivation in school-age children with CP and typically developing children. MATERIAL AND METHODS: 229 parents of children with cerebral palsy and 212 parents of typically developing children participated in the present cross sectional study and completed demographic and DMQ18 forms. The rest of

information was measured by an occupational therapist. Average age was equal to 127.12±24.56 months for children with cerebral palsy (CP) and 128.08±15.90 for typically developing children. Independent t-test used to compare two groups; and Pearson correlation coefficient by SPSS software applied to study correlation with other factors. RESULTS: There were differences between DMQ subscales of CP and typically developing groups in terms of all subscales (P<0.05). The lowest motivation scores of subscales obtained in gross motor persistence (2.4870±.81047) and cognitive-oriented persistence (2.8529±.84223) in children with CP. Motivation was correlated with Gross Motor function Classification System (r= -0.831, P<0.001), Manual ability classification system (r=-0.782, P<0.001) and cognitive impairment (r=-0.161, P<0.05). CONCLUSION: Children with CP had lower mastery motivation than typically developing children. Rehabilitation efforts should take to enhance motivation, so that children felt empowered to do tasks or practices.

[PMID: 29416217](#)

## 27. Health-related quality of life of children and adolescents with cerebral palsy in low- and middle-income countries: a systematic review.

Power R, King C, Muhit M, Heanoy E, Galea C, Jones C, Badawi N, Khandaker G.

Dev Med Child Neurol. 2018 Feb 6. doi: 10.1111/dmcn.13681. [Epub ahead of print]

AIM: To systematically review literature on health-related quality of life (HRQoL) of children and adolescents (≤18yo) with cerebral palsy (CP) from low- and middle-income countries (LMICs) to identify trends in HRQoL and areas for future research. METHOD: We systematically reviewed six key bibliographic databases and two reviewers independently screened results. Peer-reviewed original articles examining HRQoL of children from LMICs were eligible. RESULTS: A total of 22 524 papers were identified, of which 16, from eight LMICs, were included. Four measures of HRQoL were used; Child Health Questionnaire Parent Form 50 (n=5); Paediatric Quality of Life Inventory 3.0 and 4.0 (n=4); CP Quality of Life Questionnaire for Children (n=4); Lifestyle Assessment Questionnaire CP (n=3). Children with CP from LMICs (n=1579; 2-18y) had significantly poorer HRQoL on all instrument dimensions when compared to age-matched controls (p<0.003) and on all except two dimensions when compared to peers in high-income countries (p<0.001). Physical well-being dimensions of HRQoL were poorest overall and associated with impaired motor function. INTERPRETATION: Research to improve HRQoL in LMICs is required and should address all aspects of HRQoL. Future research is recommended to incorporate multi-respondent assessment, utilize both general and CP-specific measures of HRQoL, and delineate adolescents as a unique cohort. What this paper adds Children with cerebral palsy from low- and middle-income countries are at high risk of poor health-related quality of life (HRQoL). Physical well-being was poorest dimension of HRQoL and associated with impaired motor function.

[PMID: 29405292](#)

## 28. Quality of life as assessed by adults with cerebral palsy.

Maestro-Gonzalez A, Bilbao-Leon MC, Zuazua-Rico D, Fernandez-Carreira JM, Baldonado-Cernuda RF, Mosteiro-Diaz MP.

PLoS One. 2018 Feb 5;13(2):e0191960. doi: 10.1371/journal.pone.0191960. eCollection 2018.

AIM: We explored the quality of life of adults with cerebral palsy without an intellectual disability and the predictors of quality of life. BACKGROUND: Because cerebral palsy is a disease that manifests in childhood, much of the research into quality of life for those dealing with it focuses on children; there are few studies that evaluate the quality of life of adults with cerebral palsy. Therefore, it is important to consider their perceptions in order to improve their general wellbeing and self-determination. DESIGN: This was a descriptive, cross-sectional study. METHOD: Quality of life was measured using the GENCAT Quality of Life Scale. Demographic and personal variables were also collected and examined. Participants comprised 75 adults (58.7 percent men, mean age = 40.84 years) with cerebral palsy who were members of the National Cerebral Palsy Association of Spain between 2014 and 2015. A linear multivariate model was examined as well. RESULTS: The overall mean score indicator of participants' quality of life was 103.29, which corresponds to the 56.6th percentile on the GENCAT scale. Examining the level of qualification, we found significant differences in the factors "personal development" and "self-determination," and those with a university education obtained higher scores than their less-educated counterparts. Having a partner was related to higher quality of life standard scores. After constructing a linear model, it was observed that maintaining sexual relationships was another factor that increased participants' quality of life. CONCLUSION: This study highlights the importance of social and romantic relationships to achieve a better quality of life in adults with cerebral palsy who do not have an intellectual disability. Social integration and sexuality education programs should be developed to improve their quality of life.

[PMID: 29401489](#)

### 29. Improved artefact removal from EEG using Canonical Correlation Analysis and spectral slope.

Janani AS, Grummett TS, Lewis TW, Fitzgibbon SP, Whitham EM, DelosAngeles D, Bakhshayesh H, Willoughby JO, Pope KJ.

J Neurosci Methods. 2018 Jan 31;298:1-15. doi: 10.1016/j.jneumeth.2018.01.004. [Epub ahead of print]

**BACKGROUND:** Contamination of scalp measurement by tonic muscle artefacts, even in resting positions, is an unavoidable issue in EEG recording. These artefacts add significant energy to the recorded signals, particularly at high frequencies. To enable reliable interpretation of subcortical brain activity, it is necessary to detect and discard this contamination. **NEW METHOD:** We introduce a new automatic muscle-removal approach based on the traditional Blind Source Separation-Canonical Correlation Analysis (BSS-CCA) method and the spectral slope of its components. We show that CCA-based muscle-removal methods can discriminate between signals with high correlation coefficients (brain, mains artefact) and signals with low correlation coefficients (white noise, muscle). We also show that typical BSS-CCA components are not purely from one source, but are mixtures from multiple sources, limiting the performance of BSS-CCA in artefact removal. We demonstrate, using our paralysis dataset, improved performance using BSS-CCA followed by spectral-slope rejection. **RESULT:** This muscle removal approach can reduce high-frequency muscle contamination of EEG, especially at peripheral channels, while preserving steady-state brain responses in cognitive tasks. **COMPARISON WITH EXISTING METHODS:** This approach is automatic and can be applied on any sample of data easily. The results show its performance is comparable with the ICA method in removing muscle contamination and has significantly lower computational complexity. **CONCLUSION:** We identify limitations of the traditional BSS-CCA approach to artefact removal in EEG, propose and test an extension based on spectral slope that makes it automatic and improves its performance, and results in performance comparable to competitors such as ICA-based artefact removal.

[PMID: 29408174](#)

### 30. Psychometric Evaluation of the 'Evidence Based Practice Competencies Questionnaire - Cerebral Palsy'.

Kerr C, Bowe SJ, Miyazaki K, Imms C.

Phys Occup Ther Pediatr. 2018 Feb 5:1-11. doi: 10.1080/01942638.2017.1420002. [Epub ahead of print]

**AIM:** To understand whether knowledge translation activities are effective, good measurement of practice is required. This study investigated the psychometric properties of a self-report measure of allied health practitioner (AHP) evidence-based behaviors when working with children with cerebral palsy. **METHODS:** Construct validity and reliability studies were undertaken for the 12-item Evidence Based Practice Competency Questionnaire - Cerebral Palsy (EBP-CQ-CP) using the Consensus-based Standards of Measurement Instruments methods. Factor analysis tested construct validity. Weighted Kappa tested chance-corrected agreement for each item and intra-class correlation coefficients (ICC) the reliability of factors derived in the validity study. **RESULTS:** In the validity study 259 AHP completed the EBP-CQ-CP on occasion 1, and 228 on occasion 2. In the reliability study 46 AHP completed the questionnaire twice. Exploratory factor analysis determined the EBP-CQ-CP contained two scales: 'communicating evidence based expectations' and 'evidence based assessment practices'. Confirmatory factor analysis using data from the second occasion of assessment supported the findings. Excellent consistency in ratings across factor scores were obtained from 46 pairs of raters: Factor 1, ICC = 0.93 (95% Confidence Interval 0.88-0.96); Factor 2, ICC = 0.94 (95% Confidence Interval 0.88-0.97). **CONCLUSIONS:** This study supports the interpretation of the EBP-CQ-CP in a clinically meaningful and psychometrically robust manner.

[PMID: 29400591](#)

## Prevention and Cure

### 31. Magnesium as a Neuroprotective Agent: A Review of Its Use in the Fetus, Term Infant with Neonatal Encephalopathy, and the Adult Stroke Patient.

Lingam I, Robertson NJ.

Dev Neurosci. 2018 Feb 7. doi: 10.1159/000484891. [Epub ahead of print]

Magnesium is an intracellular cation essential for many en-zymatic processes and cellular functions. Magnesium sulfate acts as an endogenous calcium channel antagonist at neuronal synapses, thought to prevent excessive activation of N-methyl-D-aspartate receptors by excitatory amino acids, such as glutamate, and by downregulation of proinflammatory pathways. Early intervention is essential in the prevention of the secondary phase of neuronal injury. The immature brain is particularly prone to excitotoxicity, and inflammation has been strongly implicated in the pathogenesis of cerebral palsy. This article explores the

current status of magnesium being used as an adjunct to hypothermia in term neonatal encephalopathy (NE) against a background of its use in other populations. There is some evidence for magnesium sulfate as a neuroprotective agent, however animal studies of NE at term equivalent age have been confounded by concomitant hypothermia induced by magnesium itself. Nevertheless, the combination of magnesium and cooling has been shown to be more effective than either treatment alone in adult rodents. In the preterm baby, magnesium sulfate given antenatally in threatened preterm labor has demonstrated a significant reduction in the risk of cerebral palsy at 2 years of age, though the benefit is not clear at school age. In adult clinical studies of ischemic and hemorrhagic stroke, there have been disappointing results for magnesium sulfate as a neuroprotective strategy. Importantly, clinical neurological scores may be affected by the increased hypotonia observed. We suggest that magnesium sulfate should be carefully re-evaluated as a neuroprotective agent given its favorable safety profile, relative low cost, and widespread availability.

[PMID: 29408814](#)

### **32. TSEN54 Gene-Related Pontocerebellar Hypoplasia Type 2 Could Mimic Dyskinetic Cerebral Palsy with Severe Psychomotor Retardation.**

Pacheva IH, Todorov T, Ivanov I, Tartova D, Gaberova K, Todorova A, Dimitrova D.

Front Pediatr. 2018 Jan 23;6:1. doi: 10.3389/fped.2018.00001. eCollection 2018.

Pontocerebellar hypoplasia (PCH) type 2 is a very rare autosomal recessive neurodegenerative disorder with prenatal onset that disrupts brain development. We present three patients (two siblings and one unrelated child) with PCH 2 linked to the most common mutation c.919G > T (p.Ala307Ser) in TSEN54 gene. The disease started soon after birth with feeding difficulties, extrapyramidal symptoms, psychomotor retardation, progressive microcephaly. Two of the patients were diagnosed with dyskinetic cerebral palsy (CP) at first. Despite the neurodegenerative character of PCH 2, the absence of regression and even some developmental progress in few patients, might erroneously lead to the incorrect diagnosis of dyskinetic CP. Megacisterna magna on brain ultrasound makes the diagnosis of PCH 2 highly probable and should prompt further imaging with MRI. MRI findings of PCH are pivotal for the diagnosis. Genetic testing for the most common mutation in TSEN54 gene should also be performed. Correct diagnosis of PCH 2 is essential not only for the prognosis of the patient, but also for prenatal diagnosis in future pregnancies. Knowledge of the clinical picture of PCH 2 will lead to correct and timely diagnosis. Advanced neuroimaging procedures and molecular genetic techniques provide valuable tools for prompt diagnosis of rare, but clinically important, neurogenetic imitators of CP.

[PMID: 29410950](#)

### **33. The general movement assessment in non-European low- and middle-income countries.**

Tomantschger I, Herrero D, Einspieler C, Hamamura C, Voos MC, Marschik PB.

Rev Saude Publica. 2018 Feb 5;52:6. doi: 10.11606/S1518-8787.2018052000332.

Abnormal general movements are among the most reliable markers for cerebral palsy. General movements are part of the spontaneous motor repertoire and are present from early fetal life until the end of the first half year after term. In addition to its high sensitivity (98%) and specificity (91%), the assessment of general movements is non-invasive and time- and cost-efficient. It is therefore ideal for assessing the integrity of the young nervous system, most notably in lowresource settings. Studies on the general movements assessment in low- and middle-income countries such as China, India, Iran, or South Africa are still rare but increasing. In Brazil, too, researchers have demonstrated that the evaluation of general movements adds to the functional assessment of the young nervous system. Applying general movements assessment in vulnerable populations in Brazil is therefore highly recommended.

[PMID: 29412374](#)

### **34. Outcome in early-onset fetal growth restriction is best combining computerized fetal heart rate analysis with ductus venosus Doppler: insights from the Trial of Umbilical and Fetal Flow in Europe.**

Frusca T, Todros T, Lees C, Bilardo CM; TRUFFLE Investigators.

Am J Obstet Gynecol. 2018 Feb;218(2S):S783-S789. doi: 10.1016/j.ajog.2017.12.226.

BACKGROUND: Early-onset fetal growth restriction represents a particular dilemma in clinical management balancing the risk of iatrogenic prematurity with waiting for the fetus to gain more maturity, while being exposed to the risk of intrauterine death or the sequelae of acidosis. OBJECTIVE: The Trial of Umbilical and Fetal Flow in Europe was a European, multicenter,

randomized trial aimed to determine according to which criteria delivery should be triggered in early fetal growth restriction. We present the key findings of the primary and secondary analyses. **STUDY DESIGN:** Women with fetal abdominal circumference <10th percentile and umbilical pulsatility index >95th percentile between 26-32 weeks were randomized to 1 of 3 monitoring and delivery protocols. These were: fetal heart rate variability based on computerized cardiotocography; and early or late ductus venosus Doppler changes. A safety net based on fetal heart rate abnormalities or umbilical Doppler changes mandated delivery irrespective of randomized group. The primary outcome was normal neurodevelopmental outcome at 2 years. **RESULTS:** Among 511 women randomized, 362/503 (72%) had associated hypertensive conditions. In all, 463/503 (92%) of fetuses survived and cerebral palsy occurred in 6/443 (1%) with known outcome. Among all women there was no difference in outcome based on randomized group; however, of survivors, significantly more fetuses randomized to the late ductus venosus group had a normal outcome (133/144; 95%) than those randomized to computerized cardiotocography alone (111/131; 85%). In 118/310 (38%) of babies delivered <32 weeks, the indication was safety-net criteria: 55/106 (52%) in late ductus venosus, 37/99 (37%) in early ductus venosus, and 26/105 (25%) in computerized cardiotocography groups. Higher middle cerebral artery impedance adjusted for gestation was associated with neonatal survival without severe morbidity (odds ratio, 1.24; 95% confidence interval, 1.02-1.52) and infant survival without neurodevelopmental impairment at 2 years (odds ratio, 1.33; 95% confidence interval, 1.03-1.72) although birthweight and gestational age were more important determinants. **CONCLUSION:** Perinatal and 2-year outcome was better than expected in all randomized groups. Among survivors, 2-year neurodevelopmental outcome was best in those randomized to delivery based on late ductus venosus changes. Given a high rate of delivery based on the safety-net criteria, deciding delivery based on late ductus venosus changes and abnormal computerized fetal heart rate variability seems prudent. There is no rationale for delivery based on cerebral Doppler changes alone. Of note, most women with early-onset fetal growth restriction develop hypertension.

[PMID: 29422211](#)

### **35. Association of Early-Onset Spasticity and Risk for Cognitive Impairment With Mutations at Amino Acid 499 in SPAST.**

Gillespie MK, Humphreys P, McMillan HJ, Boycott KM.

J Child Neurol. 2018 Jan 1:883073818756680. doi: 10.1177/0883073818756680. [Epub ahead of print]

Hereditary spastic paraplegia is a phenotypically and genetically heterogeneous group of neurodegenerative disorders characterized by lower extremity weakness and spasticity. Spastic paraplegia 4 (SPG4), caused by heterozygous mutations in the gene SPAST, typically causes a late-onset, uncomplicated form of hereditary spastic paraplegia in affected individuals. Additional clinical features in SPG4 have been reported on occasion, but no genotype-phenotype correlation has been established. Through targeted clinical testing, we identified 2 unrelated female patients with the same de novo p.Arg499His mutation in SPAST. Both patients presented with early-onset spasticity resulting in delayed motor milestones, which led to a diagnosis of cerebral palsy in one child and tethered cord in the other. Review of the literature identified several patients with mutations at amino acid 499 and early-onset symptoms associated with a risk of cognitive impairment. Early and accurate diagnosis of children with early-onset spasticity is important for informed prognosis and genetic counselling.

[PMID: 29421991](#)