Assessment and Treatment of Children with Cerebral Palsy.

Chan G1, Miller F2.

Children with cerebral palsy are prone to development of musculoskeletal deformities. The underlying neurologic insult may result in a loss of selective motor control, an increase in underlying muscle tone, and muscle imbalance, which can lead to abnormal deforming forces acting on the immature skeleton. The severely involved child is one who is at increased risk for developing progressive musculoskeletal deformities. Close surveillance and evaluation are key to addressing the underlying deformity and improving and maintaining overall function.

Intramuscular connective tissue differences in spastic and control muscle: a mechanical and histological study.

de Bruin M1, Smeulders MJ1, Kreulen M2, Huizing PA3, Jaspers RT3.

Cerebral palsy (CP) of the spastic type is a neurological disorder characterized by a velocity-dependent increase in tonic stretch reflexes with exaggerated tendon jerks. Secondary to the spasticity, muscle adaptation is presumed to contribute to limitations in the passive range of joint motion. However, the mechanisms underlying these limitations are unknown. Using biopsies, we compared mechanical as well as histological properties of flexor carpi ulnaris muscle (FCU) from CP patients (n=29) and healthy controls (n=10). The sarcomere slack length (mean 2.5 µm, SEM 0.05) and slope of the normalized sarcomere length-tension characteristics of spastic fascicle segments and single myofibre segments were not different from those of control muscle. Fibre type distribution also showed no significant differences. Fibre size was significantly smaller (1933 µm², SEM 190) in spastic muscle than in controls (2572 µm², SEM 322). However, our statistical analyses indicate that the latter difference is likely to be explained by age, rather than by the affliction. Quantities of endomysial and perimysial networks within biopsies of control and spastic muscle were unchanged with one exception: a significant thickening of the tertiary perimysium (3-fold), i.e. the connective tissue reinforcement of neurovascular tissues penetrating the muscle. Note
that this thickening in tertiary perimysium was shown in the majority of CP patients, however a small number of patients (n=4 out of 23) did not have this feature. These results are taken as indications that enhanced myofascial loads on FCU is one among several factors contributing in a major way to the aetiology of limitation of movement at the wrist in CP and the characteristic wrist position of such patients.

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**Effects of Task-oriented Approach on Affected Arm Function in Children with Spastic Hemiplegia Due to Cerebral Palsy.**

Song CS.

Purpose: The purpose of the present study was to examine the effects of task-oriented approach on motor function of the affected arm in children with spastic hemiplegia due to cerebral palsy. Subjects: Twelve children were recruited by convenience sampling from 2 local rehabilitation centers. The present study utilized a one-group pretest-posttest design. All of children received task-oriented training for 6 weeks (40 min/day, 5 days/week) and also underwent regular occupational therapy. Three clinical tests, Box and Block Test (BBT), Manual Ability Measure (MAM-16), and Wee Functional Independence Measure (WeeFIM) were performed 1 day before and after training to evaluate the effects of the training. Results: Compared with the pretest scores, there was a significant increase in the BBT, MAM-16, and WeeFIM scores of the children after the 6-week practice period. Conclusion: The results of this study suggest that a task-oriented approach to treatment of the affected arm improves functional activities, such as manual dexterity and fine motor performance, as well as basic daily activities of patients with spastic hemiplegia due to cerebral palsy.

**PMID: 25013269** [PubMed]


**Efficacy of Constraint-Induced Movement Therapy and Bimanual Training in Children with Hemiplegic Cerebral Palsy in an Educational Setting.**


We examined the efficacy of modified constraint-induced movement therapy (CIMT) and hand-arm bimanual intensive therapy (HABIT) in a special education preschool-kindergarten in Israel. Twelve children (1.5-7 years) with congenital hemiplegic cerebral palsy were randomized to receive modified CIMT (n = 6) or HABIT (n = 6). Occupational and physical therapists administered usual and customary care for 8 weeks; children then crossed over to receive CIMT or HABIT 2 hr/day, 6 days/week for 8 weeks from their occupational therapist. The Assisting Hand Assessment and Quality of Upper Extremity Skills Test were administered 2 months prior to the intervention, immediately before, immediately after intervention, and 6 months after the first baseline assessment. Both groups demonstrated no change during baseline and comparable improvement following CIMT and HABIT (p < .001), which was maintained at 6-month follow-up. Results suggest that modified CIMT and HABIT provided in school-based settings can lead to improvements in quality of bimanual skill and movement patterns.

**PMID: 24983295** [PubMed - as supplied by publisher]


**Effects of Neuromuscular Electrical Stimulation on the Wrist and Finger Flexor Spasticity and Hand Functions in Cerebral Palsy.**

Yildizgören MT1, Nakipoglu Yüzer GF1, Ekiz T2, Ozgirgin N1.

PURPOSE: To evaluate the effects of neuromuscular electrical stimulation on wrist range of motion, wrist and finger...
flexor spasticity, and hand functions in patients with unilateral cerebral palsy. METHOD: Twenty-four children with unilateral spastic cerebral palsy (14 boys and 10 girls) between the ages of 5 and 14 years were randomized into neuromuscular electrical stimulation and control groups. Conventional exercises were applied, and static volar wrist-hand orthosis was administered to all patients 5 days a week for 6 weeks. Additionally, 30-minute neuromuscular electrical stimulation sessions were applied to the wrist extensor muscles in the neuromuscular electrical stimulation group. Patients were evaluated by Zancolli Classification System, Manual Ability Classification System, and Abilhand-Kids Test. RESULTS: Compared with baseline, a significant increase was evident in active wrist extension angle at the fourth and sixth weeks in both groups (all P < 0.001), more prominent in the neuromuscular electrical stimulation group at the fourth and sixth weeks (P = 0.015 and P = 0.006, respectively). A decrease was observed in the spasticity values in the neuromuscular electrical stimulation group at the fourth and sixth weeks (P = 0.002 and P = 0.001, respectively) and in the control group only at the sixth week (P = 0.008). Abilhand-Kids values improved only in the neuromuscular electrical stimulation group (P < 0.001). CONCLUSION: Neuromuscular electrical stimulation application in addition to conventional treatments is effective in improving active wrist range of motion, spasticity, and hand functions in cerebral palsy.

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Transfer of motor learning from virtual to natural environments in individuals with cerebral palsy.

de Mello Monteiro CB1, Massetti T2, da Silva TD2, van der Kamp J3, de Abreu LC4, Leone C4, Savelsbergh GJ5.

With the growing accessibility of computer-assisted technology, rehabilitation programs for individuals with cerebral palsy (CP) increasingly use virtual reality environments to enhance motor practice. Thus, it is important to examine whether performance improvements in the virtual environment generalize to the natural environment. To examine this issue, we had 64 individuals, 32 of which were individuals with CP and 32 typically developing individuals, practice two coincidence-timing tasks. In the more tangible button-press task, the individuals were required to 'intercept' a falling virtual object at the moment it reached the interception point by pressing a key. In the more, less tangible task, they were instructed to 'intercept' the virtual object by making a hand movement in a virtual environment. The results showed that individuals with CP timed less accurate than typically developing individuals, especially for the more task in the virtual environment. The individuals with CP did-as did their typically developing peers-improve coincidence timing with practice on both tasks. Importantly, however, these improvements were specific to the practice environment; there was no transfer of learning. It is concluded that the implementation of virtual environments for motor rehabilitation in individuals with CP should not be taken for granted but needs to be considered carefully.

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A randomized trial of upper limb botulimun toxin versus placebo injection, combined with physiotherapy, in children with hemiplegia.

Ferrari A1, Maoret AR2, Muzzini S3, Alboresi S4, Lombardi F5, Sgandurra G6, Paolicelli PB7, Sicola E8, Cioni G9.

The main goal of this study was to investigate the efficacy of Botulinum Toxin A (BoNT-A), combined with an individualized intensive physiotherapy/ortheses treatment, in improving upper limb activity and competence in daily activity in children with hemiplegia, and to compare its effectiveness with that of non-pharmacological instruments. It was a Randomized Clinical Trial of 27 children with spastic hemiplegic cerebral palsy, outpatients of two high speciality Centres for child rehabilitation. Each child was assigned by simple randomization to experimental group (BoNT-A) or control group (placebo). Assisting Hand Assessment (AHA) was chosen as primary outcome measure; other measures were selected according to ICF dimensions. Participants were assessed at baseline (T0), at T1, T2, T3 (1-3-6 months after injection, respectively). Every patient was given a specific physiotherapeutic treatment,
consisting of individualized goal directed exercises, task oriented activities, daily stretching manoeuvres, functional and/or static orthoses. BoNT-A group showed a significant increase of AHA raw scores at T2, compared to control group (T2-T0: p=.025) and functional goals achievement (GAS) was also slightly better in the same group (p=.033). Other measures indicated some improvement in both groups, without significant intergroup differences. Children with intermediate severity of hand function at House scale for upper limb impairment seem to have a better benefit from BoNT-A protocol. BoNT-A was effective in improving manipulation in the activity domain, in association with individualized goal-directed physiotherapy and orthoses; the combined treatment is recommended. The study brings more evidence for the efficacy of a combined treatment botulinum toxin injection-physiotherapy-orthoses, and it gives some suggestions for candidate selection and individualized treatment.

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Bone strength is related to muscle volume in ambulant individuals with bilateral spastic cerebral palsy.

Noble JJ1, Fry N2, Lewis AP3, Charles-Edwards GD4, Keevil SF4, Gough M2, Shortland AP2.

OBJECTIVE: To investigate how bone strength in the distal femur and proximal tibia are related to local muscle volume in ambulant individuals with bilateral spastic cerebral palsy (CP). METHODS: Twenty-seven participants with CP (mean age: 14.6±2.9 years; Gross Motor Function Classification System (GMFCS) levels I-III) and twenty-two typically developing (TD) peers (mean age: 16.7±3.3 years) took part in this study. Periosteal and medullary diameter in the distal femur and cortical bone cross-sectional area (CSA) and thickness (CT) in the distal femur and proximal tibia were measured along with nine lower limb muscle volumes using MRI. Additionally, the polar section modulus (Zp) and buckling ratio (BR) were calculated to estimate bone bending strength and compressional bone stability respectively in the distal femur. The relationships of all measured parameters with muscle volume, height, age, body mass, gender, and subject group were investigated using a generalized linear model (GZLM). RESULTS: In the distal femur, Zp was significantly positively related to thigh muscle volume (p=0.007), and height (p=0.026) but not significantly related to subject group (p=0.076) or body mass (p=0.098). BR was not significantly different between groups and was not related to any of the variables tested. Cortical bone CSA was significantly lower in the CP group at both the distal femur (p=0.002) and proximal tibia (p=0.009). It was also positively associated with thigh muscle volume (p<0.001) at the distal femur, and with subject height (p=0.005) at the proximal tibia. CONCLUSIONS: Bending and compressional strength of the femur, estimated from Zp and cortical bone CSA respectively, is associated with reduced thigh muscle volume. Increasing muscle volume by strength training may have a positive effect on bone mechanics in individuals with CP.

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Long-term Ambulatory Change After Lower Extremity Orthopaedic Surgery in Children With Cerebral Palsy: A Retrospective Review.

Yu S1, Rethlefsen SA, Wren TA, Kay RM.

BACKGROUND: Long-term studies of lower extremity orthopaedic surgery in children with cerebral palsy (CP) tend to focus on gait kinematics and kinetics, with little to no emphasis on gross motor ambulatory function. The current study was undertaken to examine the long-term impact of surgery on ambulatory function in patients with CP enrolled in a government-funded, outpatient therapy program. METHODS: Retrospective medical record review was conducted of 127 children with CP, Gross Motor Function Classification System (GMFCS) levels I to IV, followed up to 14 years after lower extremity orthopaedic surgery. Data were extracted from medical/operative records and routine physical therapy evaluations performed over the course of follow-up. Functional Mobility Scale (FMS) scores were assigned based on gross motor function information contained in each 6- to 12-month physical therapy evaluation. Data were compared statistically among GMFCS levels. RESULTS: Average length of follow up
was 11.8±4 years. Subjects underwent 0.61±0.43 surgical procedures per person-year in 0.16±0.09 operative sessions per person-year with no differences between GMFCS levels. Subjects at GMFCS level I improved significantly in community (P=0.02) but not household ambulation, reflecting the ceiling effect of the FMS. Subjects at GMFCS levels II to IV showed statistically significant improvements at all distances. Subjects at level III gained more in household than long-distance ambulation (P=0.002). Subjects functioning at GMFCS level II improved by 1 FMS level for household and school distances, and 2 FMS levels for community distances (P<0.02). Subjects at level IV exhibited small ambulatory gains at all distances (P<0.04). CONCLUSIONS: Significant long-term improvement in functional ambulation is seen after surgery for children at all GMFCS levels. Children with more independence tend to make gains in long-distance ambulation, whereas those who use assistive devices tend to improve more in short-distance ambulation. This information may be useful to clinicians when counseling patients and their families regarding potential for ambulatory improvement after lower extremity orthopaedic surgery.

LEVEL OF EVIDENCE: Level IV: case series.

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The effect of femoral derotation osteotomy on transverse plane hip and pelvic kinematics in children with cerebral palsy: A systematic review and meta-analysis.


The purpose of this study was to systematically review the current literature to determine the effect of a femoral derotation osteotomy (FDRO) on hip and pelvic rotation kinematics during gait compared to no intervention in children with spastic cerebral palsy (CP). We performed a systematic search for prospective and retrospective cohort studies of children with CP, who were treated with a FDRO, and were assessed with pre and post surgery three-dimensional gait analysis. Medline, CINAHL, EMBASE, the Cochrane Library and Web of Science were searched up to December 2013. Data sources were prospective and retrospective studies. Mean differences were calculated on pooled data for both pelvic and hip rotation kinematics. Thirteen of 196 articles met the inclusion criteria (5 prospective, 8 retrospective). All included studies were of sufficient quality for meta-analysis as assessed using a customised version of the STROBE checklist. Meta-analysis showed that FDRO significantly reduced pelvic retraction by 9.0 degrees and hip internal rotation by 17.6 degrees in participants with unilateral CP involvement and hip internal rotation by 14.3 degrees in participants with bilateral CP involvement. Pelvic symmetry in children with unilateral spastic CP is significantly improved by FDRO. Patients with bilateral involvement do not improve their transverse plane pelvic rotation profiles during gait as a result to FDRO, although this result should be interpreted with caution due to the heterogeneous nature of these participants and of the methods used in the studies assessed.

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Intraoperative experiments show relevance of inter-antagonistic mechanical interaction for spastic muscle’s contribution to joint movement disorder.

Ates F1, Temelli Y2, Yucesoy CA3.

BACKGROUND: Recent intra-operative knee angle-muscle force data showed no abnormal muscular mechanics (i.e., a narrow joint range of muscle force exertion and peak muscle force availability at flexed joint positions), if the spastic gracilis muscle was stimulated alone. This can limit inter-muscular mechanical interactions, which have been shown to affect muscular mechanics substantially. We aimed at testing the hypothesis that the knee angle-muscle force curves of the spastic gracilis muscle activated simultaneously with a knee extensor are representative of joint movement disorder. METHODS:
Experiments were performed during remedial surgery of spastic cerebral palsy patients (n=6, 10 limbs tested). Condition-I: muscle forces were measured in flexed knee positions (120° and 90°) after activating the gracilis exclusively. Condition-II: knee angle-muscle force data were measured from 120° to full extension after activating the vastus medialis, simultaneously. FINDINGS: Condition-II vs. I: Inter-antagonistic interaction did not consistently cause a gracilis force increase. Condition-II: Peak muscle force=mean 47.92N (SD 22.08N). Seven limbs showed availability of high muscle force in flexed knee positions (with minimally 84.8% of peak force at 120°). Knee angle-muscle force curves of four of them showed a local minimum followed by an increasing force (explained by an increasing passive force, indicating muscle lengths unfavorable for active force exertion). High active gracilis forces measured at flexed knee positions and narrow operational joint range of force exertion do indicate abnormality. The remainder of the limbs showed no such abnormality. INTERPRETATION: Our hypothesis is confirmed for most, but not all limbs tested. Therefore, tested inter-antagonistic mechanical interaction can certainly, but not exclusively be a factor for abnormal mechanics of the spastic muscle.

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Reid SL1, Pitcher CA, Williams SA, Licari MK, Valentine JP, Shipman PJ, Elliott CM.

Purpose: To investigate the muscle size-strength relationship of the knee flexors and extensors in children with spastic cerebral palsy (CP) in relation to typically developing children (TD). Methods: Eighteen children with spastic Diplegia, Gross Motor Function Classification System I-III (mean 7y 5mo SD 1y 7o) and 19 TD children (mean 7y 6mo SD 1y 9mo) participated. Muscle volume (MV) and anatomical cross-sectional area (aCSA) were assessed using MRI. Measures of peak torque (PT) and work of the knee flexors and extensors were assessed isometrically and isokinetically using a Biodex dynamometer, and normalised to bodymass (Bm). Results: Children with CP were weaker than their TD peers across all torque variables (p<0.05). MV and aCSA of the knee flexors (MV: p=0.002; aCSA: p=0.000) and extensors (MV: p=0.003; aCSA: p=0.0001) were smaller in children with CP. The relationship between muscle size and strength in children with CP was weaker than the TD children. The strongest relationship was between MV and isometric PT/Bm for TD children (r=0.77-0.84), and between MV and isokinetic work (r=0.70-0.72) for children with CP. Conclusions: Children with CP have smaller, weaker muscles than their TD peers. However, muscle size may only partially explain their decreased torque capacity. MV appears to be a better predictor of muscle work in children with CP than aCSA. This is an important area of research particularly in regard to treatment(s) that target muscle and strength in children with CP. Implications for Rehabilitation This research adds to the evidence that children with CP have smaller, weaker knee flexor and extensor muscles than their TD peers. However, unlike their TD peers, muscle size does not necessarily relate to muscle strength. The weak correlation between MRI-derived muscle volume and isometric peak torque suggests children with CP are underpowered relative to their muscle size. For children with CP, muscle volume appears to be the best predictor of isokinetic muscle torque output. Therefore, when assessing the capacity of a muscle, it appears preferable to measure total muscle volume and torque development through a range of motion (isokinetic strength).

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Changes in spastic muscle stiffness after botulinum toxin A injections as part of rehabilitation therapy in patients with spastic cerebral palsy.
Boyaci A1, Tutoğlu A1, Boyaci N2, Kocac I, Calik M3, Sakalar A1, Kılicaslan N2.

OBJECTIVE: To investigate changes in stiffness in the gastrocnemius (GCM) muscle following rehabilitation therapy and botulinum toxin type A (BTX-A) injections in patients with spastic cerebral palsy (CP). METHODS: The study included 16 children with spastic CP (Group 1) and 17 healthy children (Group 2). The children with CP received BTX-A injections and underwent rehabilitation therapy. The GCM and soleus muscles of the CP group
were evaluated according to the modified Ashworth scale (MAS). The thicknesses of the muscles were measured, and the elasticity score (ELX 2/1) index was calculated. RESULTS: The ELX 2/1 indices of the gastromedialis (GM) and gastrolateralis (GL) were significantly higher in Group 1 than in Group 2 (p < 0.05). The ELX 2/1 indices in the GM and GL muscles in the CP group were found to have decreased 4 weeks after the procedure (p < 0.05). Furthermore, the mean MAS score of the ankle decreased, from 3.4 to 2.6 (p < 0.05). GM ELX 2/1 in Group 1 (post-treatment) was significantly different from Group 2 (p < 0.05). CONCLUSIONS: The combined use of clinical scales and sonoelastography can provide valuable information for determining structural changes in the GCM muscle following rehabilitation therapy and BTX-A injections.

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Variability explained by strength, body composition and gait impairment in activity and participation measures for children with cerebral palsy: a multicentre study.

Oeffinger D1, Gorton G2, Hassani S3, Sison-Williamson M4, Johnson B5, Whitmer M6, Romness M7, Kryscio D8, Tylkowski C6, Bagley A4.

OBJECTIVE: To determine the amount of variability in scores on activity and participation measures used to assess ambulatory individuals with cerebral palsy explained by strength, body composition, gait impairment and participant characteristics. DESIGN: Multicentre prospective cross-sectional study. SETTING: Seven paediatric-orthopaedic specialty hospitals. PARTICIPANTS: Three hundred and seventy-seven ambulatory individuals (241 males, 136 females) with cerebral palsy, Gross Motor Function Classification System (GMFCS) levels I-III (I = 148, II = 153, III = 76), ages 8-18 years (mean 12 years 9 months, SD 2 years 8 months). METHODS: Participants completed assessments of GMFCS level, patient history, lower extremity muscle strength, Gross Motor Function Measure (GMFM-66), Pediatric Outcomes Data Collection Instrument (PODCI), instrumented gait analysis, 1 minute walk test, Timed Up-and-Go and body composition. Multiple linear regression and bootstrap analyses were performed for each outcome measure, stratified by GMFCS level. RESULTS: The amount of variability in outcome measures explained by participant characteristics, strength, and gait impairment ranged from 11% to 50%. Gait impairment was the most common predictor variable and frequently explained the greatest variance across all outcome measures and GMFCS levels. As gait impairment increased, scores on outcome measures decreased. Strength findings were inconsistent and not a primary factor. Body composition contributed minimally (<4%) in explaining variability. Participant characteristics (cerebral palsy type, gestational age and age at walking onset), were significant predictor variables in several models. CONCLUSIONS: Variability in outcome measure scores is multifaceted and only partially explained by strength and gait impairment illustrating the challenges of attempting to explain variation within this heterogeneous population. Clinicians treating individuals with cerebral palsy should consider this when developing treatment paradigms.

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Gait Changes Following Myofascial Structural Integration (Rolfing) Observed in 2 Children With Cerebral Palsy.

Hansen AB1, Price KS2, Loi EC3, Buysse CA3, Jaramillo TM4, Pico EL5, Feldman HM6.

Children with spastic cerebral palsy experience difficulty with ambulation. Structural changes in muscle and fascia may play a role in abnormal gait. Myofascial structural integration (Rolfing) is a manual therapy that manipulates muscle and soft tissues to loosen fascia layers, reposition muscles, and facilitate alignment. This study aimed to document (1) gait characteristics of 2 children with cerebral palsy and (2) effects of myofascial structural integration on their gait. Children received 3 months of weekly therapy sessions by an experienced practitioner. Gait parameters were recorded at baseline and after treatment using an electronic walkway. Children with cerebral palsy demonstrated abnormal velocity and cadence, decreased step length and single support times, and increased double support time. After treatment, both children demonstrated improvement for 3 months in cadence and double
support time. The objective gait analyses demonstrated temporary improvements after myofascial structural integration in children with spastic cerebral palsy.

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Pes planovalgus deformity in children with cerebral palsy: review article.

Kadhim M1, Miller F.

Pes planovalgus deformity is common in children with cerebral palsy. In planovalgus foot, the talus subluxates medially, the calcaneus is in valgus, and the forefoot is in supination and external rotation related to the midfoot. In young children, mild planovalgus feet can be managed with orthotics. Surgery is indicated if the deformity is not reducible or if the patient cannot tolerate orthotics during functional activities. The surgery can be calcaneal lengthening or subtalar fusion to restore a stable plantigrade foot and achieve hindfoot correction. Medial column fusion is important to reconstruct the medial arch in severe planovalgus feet.

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Defining the Mechanical Properties of a Spring-Hinged Ankle Foot Orthosis to Assess its Potential Use in Children With Spastic Cerebral Palsy.

Kerkum YL1, Brehm MA, Buizer AI, van den Noort JC, Becher JG, Harlaar J.

A rigid ventral shelf ankle foot orthosis (AFO) may improve gait in children with spastic cerebral palsy (SCP) whose gait is characterized by excessive knee flexion in stance. However, these AFOs can also impede ankle range of motion (ROM) and thereby inhibit push-off power. A more spring-like AFO can enhance push-off and may potentially reduce walking energy cost. The recent development of an adjustable spring-hinged AFO now allows adjustment of AFO stiffness, enabling tuning towards optimal gait performance. This study aims to quantify the mechanical properties of this spring-hinged AFO for each of its springs and settings. Using an AFO stiffness tester, two AFO hinges and their accompanying springs were measured. The springs showed a stiffness range of 0.01 to 1.82 Nm·deg⁻¹. The moment-threshold increased with increasing stiffness (1.13 to 12.1 Nm), while ROM decreased (4.91 to 16.5 degrees). Energy was returned by all springs (11.5 to 116.3 J). These results suggest that the two stiffest available springs should improve joint kinematics and enhance push-off in children with SCP walking with excessive knee flexion.

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Therapeutic effects of different doses of botulinum toxin A injection on tiptoe deformation in children with cerebral palsy [Article in Chinese]

Niu GH1, Zhang XL, Zhu DN, Cai ZJ, Li SS, Zhang W.

OBJECTIVE: To study the therapeutic effects of different doses of botulinum toxin A (BTX-A) injection on tiptoe deformation in children with cerebral palsy. METHODS: A total of 256 children with tiptoe deformation due to spastic cerebral palsy were classified into group A (muscle tension levels I-II, n=147) and group B (muscle tension levels III-IV, n=109). Group A was randomly divided into group A1 (injected with high-dose BTX-A, n=73) and group A2 (injected with low-dose BTX-A, n=74). Group B was randomly divided into group B1 (injected with high-dose BTX-A, n=55) and group B2 (injected with low-dose BTX-A, n=54). The dose of BTX-A was 6 U/kg for groups A1 and B1 and was 3 U/kg for groups A2 and B2. Before the injection and at 1,2,6, and 12 months after injection, the
muscle tension of limbs was evaluated with the modified Ashworth Scale, and the recovery of motor function of lower limbs was assessed with the Gross Motor Function Measure (GMFM). RESULTS: Before and after treatment, there were no significant differences in Ashworth and GMFM scores between groups A1 and A2 (P>0.05). After treatment, group B1 had a significantly reduced Ashworth score and a significantly increased GMFM score, and group B1 had a significantly lower Ashworth score and a significantly higher GMFM score compared with group B2 (P<0.05). For groups A and B, Ashworth score gradually declined post-treatment, reached the lowest point at 3 months after treatment, and returned to the level before treatment at 12 months after treatment; GMFM score gradually increased post-treatment and reached the peak level at 12 months after treatment (P<0.05).

CONCLUSIONS: The level of muscle tension should be considered when BTX-A injection is used for treating tiptoe deformation in children with cerebral palsy. It makes no difference to use high- or low-dose BTX-A when the muscle tension level is within I-II, but high-dose BTX-A has a better performance in reducing muscle tension and improving motor function when the muscle tension level is within III-IV.

PMID: 25008880 [PubMed - in process]


Incidence and risk factors for post-operative complications after scoliosis surgery in patients with Duchenne muscular dystrophy : a comparison with other neuromuscular conditions.

Duckworth AD, Mitchell MJ, Tsirikos AI.

We report the incidence of and risk factors for complications after scoliosis surgery in patients with Duchenne muscular dystrophy (DMD) and compare them with those of other neuromuscular conditions. We identified 110 (64 males, 46 females) consecutive patients with a neuromuscular disorder who underwent correction of the scoliosis at a mean age of 14 years (7 to 19) and had a minimum two-year follow-up. We recorded demographic and peri-operative data, including complications and re-operations. There were 60 patients with cerebral palsy (54.5%) and 26 with DMD (23.6%). The overall complication rate was 22% (24 patients), the most common of which were deep wound infection (9, 8.1%), gastrointestinal complications (5, 4.5%) and hepatotoxicity (4, 3.6%). The complication rate was higher in patients with DMD (10/26, 38.5%) than in those with other neuromuscular conditions (14/84, 16.7%) (p = 0.019). All hepatotoxicity occurred in patients with DMD (p = 0.003), who also had an increased rate of deep wound infection (19% vs 5%) (p = 0.033). In the DMD group, no peri-operative factors were significantly associated with the rate of overall complications or deep wound infection. Increased intra-operative blood loss was associated with hepatotoxicity (p = 0.036). In our series, correction of a neuromuscular scoliosis had an acceptable rate of complications: patients with DMD had an increased overall rate compared with those with other neuromuscular conditions. These included deep wound infection and hepatotoxicity. Hepatotoxicity was unique to DMD patients, and we recommend peri-operative vigilance after correction of a scoliosis in this group. Cite this article: Bone Joint J 2014; 96-B:943-9.


PMID: 24986949 [PubMed - in process]


Use of a proximal humeral plate for a paediatric peri-prosthetic femoral fracture.

Shaw CR1, Badhesha J2, Ayana G2, Abu-Rajab R2.

In this case an 18-year-old female with cerebral palsy sustained a peri-prosthetic femoral fracture adjacent to a blade plate previously inserted for a femoral varus osteotomy. The injury was treated using a long proximal humeral locking plate. The existing blade plate was removed. The fracture was reduced and held, and a 10-hole PHILOS™ plate applied with near anatomical reduction. There were no post-operative complications. Radiographic union was confirmed at 11 months. To our knowledge, this is the first reported use of a PHILOS™ plate in the management of a femoral peri-prosthetic fracture and successfully demonstrated a straightforward method for revision fixation.

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Early Pelvic Fixation Failure in Neuromuscular Scoliosis.
Myung KS1, Lee C, Skaggs DL.

BACKGROUND: To report on early failures of pelvic fixation in posterior spinal fusions for neuromuscular scoliosis.

METHODS: A retrospective review of posterior-only spinal instrumentation and fusion to the pelvis with iliac screws was performed. Forty-one patients with a mean age of 14 years and mean 16 levels fused met the inclusion criteria. Diagnoses include cerebral palsy (22), Duchenne muscular dystrophy (7), other neuromuscular (10), and spina bifida (2). Cox proportional hazards regression modeling was used to compare rates of failure. RESULTS: The mean preoperative primary Cobb angle was 82 degrees (range, 21 to 144 degrees). The pelvic obliquity correction was 76%. The fixation in the pelvis failed in 12/41 patients (29%). Failures include: screw head disengaged from screw shaft (5), iliac screw disengaged from rod (2), iliac connector disengaged from rod (2), iliac connector disengaged from iliac screw (4), and iliac screw loosened from bone (3). No failures occurred if there were at least 6 screws in L5, S1, and pelvis (0/7 patients). The failure rate with <6 screws in L5, S1, and pelvis was higher at 35% (12/34 patients) (P=0.16). When using traditional iliac screws with connectors to rods, all constructs had <6 screws in L5, S1, and pelvis. Only 1 failure occurred when S2-iliac screws were used, but was without clinical consequence. The mean time from surgery to failure was 18 months (range, 1 to 49 mo). CONCLUSION: Not placing bilateral pedicle screws at L5 and S1, in addition to 2 iliac screws, was associated with a 35% early failure rate of pelvic fixation.

LEVEL OF EVIDENCE: Level IV.

PMID: 24992353 [PubMed - as supplied by publisher]


Commentary on "measuring postural stability in young children with cerebral palsy: a comparison of 2 instruments".
Premysler CR1, Smith BA.

PMID: 24979090 [PubMed - in process]


Elizabeth Randall K1, Bartlett DJ, McCoy SW.

PURPOSE: To compare construct validity, interrater and test-retest reliabilities of the Pediatric Reach Test and the Early Clinical Assessment of Balance (ECAB), and their relationships with the Gross Motor Function Measure, 66-item version, Basal and Ceiling approach (GMFM-66-B&C) to appraise clinical utility of postural stability measures for children with cerebral palsy (CP). METHODS: A total of 28 children with CP, 2 to 7 years old, across all functional ability levels participated in 2 assessments over 2 weeks. Two assessors scored the measures during the first assessment. RESULTS: Both measures demonstrated construct validity, rs of 0.88 (P < .001). Both measures correlated with GMFM-66-B&C, rs > 0.95. Interrater and test-retest reliabilities were stronger for the ECAB than for the Pediatric Reach Test (intraclass correlation coefficients > 0.98 vs 0.87-0.94). The ECAB demonstrated lower measurement error and proportionately smaller minimal detectable change values. CONCLUSION: The ECAB is considered the better measure of postural stability among children with CP.

PMID: 24979089 [PubMed - in process]
Facilitation handlings induce increase in electromyographic activity of muscles involved in head control of Cerebral Palsy children.

Simon AD1, Pinho AS2, Grazziotin Dos Santos C3, Pagnussat AD4.

This study aimed to investigate the electromyographic (EMG) activation of the main cervical muscles involved in the head control during two postures widely used for the facilitation of head control in children with Cerebral Palsy (CP). A crossover trial involving 31 children with clinical diagnosis of CP and spastic quadriplegia was conducted. Electromyography was used to measure muscular activity in randomized postures. Three positions were at rest: (a) lateral decubitus, (b) ventral decubitus on the floor and (c) ventral decubitus on the wedge. Handlings for facilitating the head control were performed using the hip joint as key point of control in two postures: (a) lateral decubitus and (b) ventral decubitus on wedge. All children underwent standardized handlings, performed by the same researcher with experience in the neurodevelopmental treatment. EMG signal was recorded from muscles involved in the head control (paraspinal and sternocleidomastoid muscles) in sagittal, frontal and transverse planes, at the fourth cervical vertebra (C4), tenth thoracic vertebra (T10) and sternocleidomastoid muscle (SCM) levels. The results showed a significant increase in muscle activation when handling was performed in the lateral decubitus at C4 (P<0.001), T10 (P<0.001) and SCM (P=0.02) levels. A significant higher muscle activation was observed when handling was performed in lateral decubitus when compared to ventral decubitus at C4 level (P<0.001). Handling in ventral decubitus also induced an increase in EMG activation at T10 (P=0.018) and SCM (P=0.004) levels but not at C4 level (P=0.38). In conclusion, handlings performed in both positions may induce the facilitation of head control, as evaluated by the activity of cervical and upper trunk muscles. Handling performed in lateral decubitus may induce a slightly better facilitation of head control. These findings contribute to evidence-based physiotherapy practice for the rehabilitation of severely spastic quadriplegic CP children.

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PMID: 25010566 [PubMed - as supplied by publisher]

Correlation between Pediatric Balance Scale and Functional Test in Children with Cerebral Palsy.

Duarte Nde A1, Grecco LA2, Franco RC2, Zanon N3, Oliveira CS4.

Purpose: To investigate the correlation of functional balance with the functional performance of children with cerebral palsy. Subjects and Methods: This was a cross-sectional study of children with cerebral palsy with mild to moderate impairment. The children were divided into 3 groups based on motor impairment. The evaluation consisted of the administration of the Pediatric Balance Scale (PBS) and the Pediatric Evaluation Disability Inventory. Correlations between the instruments were determined by calculating Pearson's correlation coefficients. [Results] In Group 1, a strong positive correlation was found between the PBS and the mobility dimension of the Pediatric Evaluation Disability Inventory (r=0.82), and a moderate correlation was found between the PBS and self-care dimension of the Pediatric Evaluation Disability Inventory (r=0.51). In Group 2, moderate correlations were found between the PBS and both the self-care dimension (r=0.57) and mobility dimension (r=0.41) of the Pediatric Evaluation Disability Inventory. In Group 3, the PBS was weakly correlated with the self-care dimension (r=0.11) and moderately correlated with the mobility dimension (r=0.55). [Conclusion] The PBS proved to be a good auxiliary tool for the evaluation of functional performance with regard to mobility, but cannot be considered a predictor of function in children with cerebral palsy.

PMID: 25013281 [PubMed]

Commentary on "effects of short-term daily community walk aide use on children with unilateral spastic cerebral palsy".

Dholakia K1, Migliore S, Prosser L.

Effects of short-term daily community walk aide use on children with unilateral spastic cerebral palsy.

Pool D1, Blackmore AM, Bear N, Valentine J.

PURPOSE: To determine the effects of functional electrical stimulation (FES) on the main impairments affecting gait in children with unilateral spastic cerebral palsy. METHODS: A 20-week, multiple single-subject A-B-A design included a 6-week pre-FES phase, an 8-week FES phase, and a 6-week post-FES phase. Twelve children, aged 5 to 16 years, wore an FES device (the Walk Aide) daily for 8 weeks. Weekly measures included ankle range of motion, selective motor control, dorsiflexion and plantar flexion strength, gastrocnemius spasticity, single-limb balance, Observational Gait Scale (OGS) score, and self-reported toe drag and falls in the community. RESULTS: Compared with the pre-FES phase, the FES phase showed significant improvements in ankle range of motion, selective motor control and strength, and reductions in spasticity, toe drag, and falls, but no change in OGS score. These improvements were maintained during the post-FES phase. CONCLUSIONS: Intermittent, short-term use of FES is potentially effective for reducing impairments affecting gait in children with unilateral spastic cerebral palsy.

PMID: 24979084 [PubMed - in process]


Heart rate variability in children with cerebral palsy: Review of the literature and meta-analysis.

Amichai T, Katz-Leurer M.

OBJECTIVE: A systematic review which aims to assess the evidence regarding the function of the autonomic heart rate regulation system among children with cerebral palsy (CP). METHODS: The target population included children with CP of diverse severity, aged 1.5 to 18 years. Databases searched for English language studies from 1960 to 2013: PubMed, EMBASE, CINAHL, Cochrane Library, The Physiotherapy Evidence Database (PEDro), and ClinicalTrials.gov site. Search terms included 'cerebral palsy' or 'spastic diplegia' or 'hemiplegia' or 'quadriplegia' and 'autonomic nervous system' or 'heart rate variability' or sympathetic or 'parasympathetic'. Twenty five articles were identified and included if (1) participants were less than 18 years of age, (2) diagnosis of CP was made after the age of 18 months (3) more than 80% of cases had a diagnosis of CP and (4) autonomic cardiac heart rate regulation system state or response to a stimuli was described for all the participants. Six articles met the criteria for inclusion. RESULTS: Evidence suggests that reduced Heart Rate Variability (HRV) time domain parameters close to birth are associated with a CP diagnosis at the age of three years. In addition, HRV parameters’ mean values, are significantly lower among children with CP compared to typically developed (TD) control. While performing head up, tilt or standing position, HRV was significantly reduced only among TD control, but no effect was seen in those variables among children with CP. CONCLUSION: Further studies are needed to assess the potential to predict CP by assessing HRV parameters among newborn children. In addition, assessing HRV among children with CP may improve our understanding of the heart rate autonomic system and its response to different stimuli such as muscle contraction, paced breathing and aerobic training.

PMID: 24990007 [PubMed - as supplied by publisher]


Criterion-related validity of the Test of Children’s Speech sentence intelligibility measure for children with cerebral palsy and dysarthria.

Hodge M1, Gotzke CL.

Purpose: To evaluate the criterion-related validity of the TOCS+ sentence measure (TOCS+, Hodge, Daniels & Gotzke, 2009 ) for children with dysarthria and CP by comparing intelligibility and rate scores obtained concurrently
from the TOCS+ and from a conversational sample. Method: Twenty children (3 to 10 years old) diagnosed with spastic cerebral palsy (CP) participated. Nineteen children also had a confirmed diagnosis of dysarthria. Children's intelligibility and speaking rate scores obtained from the TOCS+, which uses imitation of sets of randomly selected items ranging from 2-7 words (80 words in total) and from a contiguous 100-word conversational speech were compared. Results: Mean intelligibility scores were 46.5% (SD = 26.4%) and 50.9% (SD = 19.1%) and mean rates in words per minute (WPM) were 90.2 (SD = 22.3) and 94.1 (SD = 25.6), respectively, for the TOCS+ and conversational samples. No significant differences were found between the two conditions for intelligibility or rate scores. Strong correlations were found between the TOCS+ and conversational samples for intelligibility (r = 0.86; p < 0.001) and WPM (r = 0.77; p < 0.001), supporting the criterion validity of the TOCS+ sentence task as a time efficient procedure for measuring intelligibility and rate in children with CP, with and without confirmed dysarthria. Conclusion: The results support the criterion validity of the TOCS+ sentence task as a time efficient procedure for measuring intelligibility and rate in children with CP, with and without confirmed dysarthria. Children varied in their relative performance on the two speaking tasks, reflecting the complexity of factors that influence intelligibility and rate scores.

PMID: 25011401 [PubMed - in process]


Association between objective measurement of the speech intelligibility of young people with dysarthria and listener ratings of ease of understanding.

Landa S1, Pennington L, Miller N, Robson S, Thompson V, Steen N.

Purpose: This study aimed to investigate the association between listeners’ ratings of how much effort it took to understand the speech of young people with cerebral palsy and the percentage of words listeners actually understood. Method: Thirty-one young people with dysarthria and cerebral palsy (16 males, 15 females; mean age = 11 years, SD = 3) were audio recorded repeating single words and producing speech. Objective measures of intelligibility were calculated for multiple familiar and unfamiliar listeners using a forced choice paradigm for single words and verbatim orthographic transcriptions for connected speech. Listeners rated how much effort it took to understand speech in each condition using a 5-point ordinal ease of listening (EOL) scale. Results: Agreement on EOL within rater groups was high (ICC > 0.71). An effect of listener was observed for familiar listeners, but not for unfamiliar listeners. EOL agreement between familiar and unfamiliar listeners was weak-moderate (ICC = 0.46). EOL predicted the percentage of speech actually understood by familiar and unfamiliar listeners (r > 0.56, p < 0.001 for all predictions). Strongest associations between EOL and intelligibility were observed for speakers with mild and profound impairments. Conclusions: The findings of this study demonstrate that listeners can judge how well they have understood dysarthric speech. EOL is associated with listener familiarity, speech task and speech impairment severity. EOL is appropriate for use in clinical practice as a measure of communication activity.

PMID: 25011400 [PubMed - in process]


Individual and environmental contributions to treatment outcomes following a neuroplasticity-principled speech treatment (LSVT LOUD) in children with dysarthria secondary to cerebral palsy: A case study review.

Boliek CA1, Fox CM.

This study describes the use of a neuroplasticity-principled speech treatment approach (LSVT(®)LOUD) with children who have dysarthria secondary to cerebral palsy. To date, the authors have treated 25 children with mild-to-severe dysarthria, a continuum of gross and fine motor functions, and variable cognitive abilities. From this data set, two case studies are presented that represent as weak or strong responders to LSVT LOUD. These case studies demonstrate how individual and environmental features may impact immediate and lasting responses to treatment. Principles that drive activity-dependent neuroplasticity are embedded in LSVT LOUD and may contribute to positive therapeutic and acoustic outcomes. However, examination of the response patterns indicated that intensity (within and across treatment sessions) is necessary but not sufficient for change. Weak responders may require a longer treatment phase, better timing (e.g., developmentally, socially), and a more prominent desire to
communicate successfully during daily activities. Strong responders appear to benefit from the intensity and saliency of treatment as well as from intrinsic and extrinsic rewards for using the trained skills for everyday communication. Finally, possibilities are presented for technological solutions designed to promote accessibility to the intensive task repetition and maintenance required to drive lasting changes.

PMID: 25011399 [PubMed - in process]


Nutritional problems in children with neuromotor disabilities: an Italian case series.


Background and aims: Several neuromotor disorders share exclusive, although often overlooked, nutritional problems. The objective of this study is therefore to delineate the frequency of malnutrition, evaluate the effectiveness of nutritional care, and identify issues needing to be possibly strengthened when caring for these patients into a general pediatrics department. Patients and methods: The study included 30 patients, 21 males and 9 females, aged between 2 and 15 years, affected by cerebral palsy, epileptic encephalopathy, and severe psychomotor developmental delay. Nutritional status was assessed by a dietary questionnaire administered to parents to investigate feeding difficulties; 3 days food diary to quantify daily calorie intake; anthropometrical (weight, height / length, body mass index percentiles, pliometry, specific body segments measurement) and blood (blood count, serum iron, albumin, transferrin, calcium, phosphorus) parameters. RESULTS: More than 44% individuals of the study population was at risk of malnutrition, according to feeding difficulties, progressive depletion of weight, reduced daily calorie intake, reduced albumin and transferrin levels. This occurred despite a massive caregivers commitment, as documented by almost universal parental constant assistance during their long-duration meals. CONCLUSIONS: Our results individuate the nutritional aspect being still a problem in the care of children with severe neuromotor disability.

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Measuring intellectual ability in children with cerebral palsy: Can we do better?

Sherwell S1, Reid SM2, Reddhihough DS3, Wrennall J4, Ong B5, Stargatt R1.

Standard intelligence tests such as the WPPSI-III have limitations when testing children with motor impairment. This study aimed to determine the proportion of children with cerebral palsy with sufficient verbal and motor skills to complete the WPPSI-III, to determine their comparative ability to complete tasks with and without a significant motor component, and to investigate short forms of the WPPSI-III as alternatives. Participants were 78 of 235 eligible 4-5 year old children with cerebral palsy resident in the Australian state of Victoria. Verbal IQ (VIQ), Performance IQ (PIQ), and Full-scale IQ (FSIQ) were determined using the WPPSI-III. Initial screening for pointing and verbal abilities determined which tests were attempted. The impact of speed was investigated by comparing scores on the Block Design subtest with and without an imposed time limit. FSIQ scores were calculated from two short forms of the WPPSI-III and compared to the full form. On screening, 16 children had inadequate pointing (14) and verbal abilities determined which tests were attempted. The impact of speed was investigated by comparing scores on the Block Design subtest with and without an imposed time limit. FSIQ scores were calculated from two short forms of the WPPSI-III and compared to the full form. On screening, 16 children had inadequate pointing (14) and verbal abilities (2). FSIQ was obtained in 62 (82%) children. Strong associations were seen between completion of the entire test battery and topographical pattern, level of manual ability and level of gross motor function. Scores on subtests requiring manual ability were depressed relative to other scores. Children performed better using short forms of the WPPSI-III and, for a minority, when time limits were disregarded. In summary, children with cerebral palsy often lack the fine and gross motor skills necessary to complete the WPPSI-III, scoring relatively poorly on tasks requiring a fine motor response. Using short-form estimations of FSIQ comprised of subtests without a significant fine motor component has the potential to increase a child's FSIQ by approximately 5 points. These findings have important clinical implications when assessing a child with both motor and cognitive limitations.

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Do children with cerebral palsy benefit from computerized working memory training? Study protocol for a randomized controlled trial.

Løhaugen GC, Beneventi H, Andersen GL, Sundberg C, Ostgård HF, Bakkan E, Walther G, Vik T, Skranes J.

BACKGROUND: Cerebral palsy (CP) is the most common motor disability in childhood (2 to 3 per 1000 live births), and is frequently accompanied by cognitive impairments and behavioural problems. Children with CP are at increased risk of attention deficit disorder with or without hyperactivity (Attention Deficit Disorder (ADD)/Attention Deficit Hyperactivity Disorder (ADHD)) including working memory deficits. The primary aim of this study is to evaluate if cognitive training may improve working memory in children with CP.

Methods/Designs: The study is an investigator-blinded, randomized controlled trial with a stepped-wedge design that will include 115 schoolchildren with CP. Eligible for participation are children with CP, aged 7 to 15 years, who are able to follow instructions and handle a computer mouse. Exclusion criteria are the presence of photosensitive epilepsy, Gross Motor Function Classification System (GMFCS) level V (most severe CP) (Phys Ther 80: 974-985, 2000) and severe visual or hearing impairments. Following assessment of eligibility and baseline cognitive assessment the participants will be randomized to either cognitive working memory training or treatment-as-usual (‘control group’). The intervention is a computer-based working memory training program consisting of 25 daily sessions to be performed over a 5 to 6-week period at home. A neuropsychological assessment will be performed before and 4 to 6 weeks after completed training. When the latter assessment has been completed in the intervention group, the ‘control group’ will start on the same training program. Both groups will meet for a final neuropsychological assessment six months after completed training by an examiner unaware of group adherence.

DISCUSSION: There is limited evidence for the effect of most interventions in children with CP, and evidence is completely lacking for interventions aiming to improve deficits in cognition, learning and behaviour. The proposed multicenter study, will bring forth comprehensive information about cognitive, neuropsychological, and daily-life functioning in children with CP aged between 7 and 15 years. In addition, the study will be the first to evaluate the effects of an intervention method to improve working memory in children with CP. If successful, computer-based working memory training may represent an efficient and cost-effective intervention for this group of children.

Trial registration: ClinicalTrials.gov Identifier: NCT02119364.

PMID: 24998242 [PubMed - as supplied by publisher] Free full text


Brazilian version of the instrument of environmental assessment Craig Hospital Inventory of Environmental Factors (CHIEF): translation, cross-cultural adaptation and reliability [Article in English, Portuguese]

Furtado SR1, Sampaio RF1, Vaz DV1, Pinho BA2, Nascimento IO2, Mancini MC3.

Background: Environmental factors are essential for the characterization of human functioning and disability; however, the shortage of standardized instruments to assess environmental factors has limited the design of scientific investigations directed at identifying barriers to and facilitators of social participation of people with disabilities. Objectives: To translate to Brazilian Portuguese, cross-culturally adapt, and verify the reliability of an environmental assessment questionnaire, entitled Craig Hospital Inventory of Environmental Factors (CHIEF). Method: The questionnaire was translated to Portuguese, analyzed, translated back to English, and compared with the original version. The final version (CHIEF-BR) was submitted to 47 caregivers of children and adolescents with cerebral palsy (CP). The intra-rater reliability was tested using quadratic kappa and intraclass correlation coefficients (ICC), through interviews of 23 caregivers drawn from the total sample, on two occasions 10 days apart. Results: During submission of the questionnaires, it was observed that examples were needed in order to facilitate the understanding of the questions related to the politics sub-scale. Quadratic kappa showed that test-retest reliability of each question varied from 0.28 to 1.0 for the frequency score and from 0.30 to 0.98 for the magnitude score. Intraclass correlation coefficients for total scores showed high consistency indices (ICC=0.92) for test-retest. Conclusion: The Brazilian version of the CHIEF was reproducible and applicable to the study sample. It may serve as an instrument to characterize the environmental barriers as well as a way to document the effects of
interventions aimed at minimizing the impact of such barriers on the participation of children and adolescents with CP.

**PMID: 25003279** [PubMed - in process] Free full text

## Prevention and Cure


**Joubert syndrome labeled as hypotonic cerebral palsy.**

Dekair LH1, Kamel H, El-Bashir HO.

Joubert syndrome (JS) is a rare autosomal recessive disorder with cerebellar vermis hypoplasia and complex brainstem malformation. The diagnosis of cases can be difficult as the presentation can be similar to cases of cerebral palsy. We present a case of JS in an 18-month-old girl who presented to pediatric rehabilitation with a diagnosis of hypotonic cerebral palsy and abnormal eye movements. The brain MRI confirmed the typical brain malformations.

**PMID: 24983287** [PubMed - in process]

### 37. Early Hum Dev. 2014 Jul 3;90(9):455-458. doi: 10.1016/j.earlhumdev.2014.06.002. [Epub ahead of print]

**Acute and massive bleeding from placenta previa and infants’ brain damage.**

Furuta K1, Tokunaga S1, Furukawa S1, Sameshima H2.

**BACKGROUND:** Among the causes of third trimester bleeding, the impact of placenta previa on cerebral palsy is not well known. **AIMS:** To clarify the effect of maternal bleeding from placenta previa on cerebral palsy, and in particular when and how it occurs. **STUDY DESIGN:** A descriptive study. **SUBJECTS:** Sixty infants born to mothers with placenta previa in our regional population-based study of 160,000 deliveries from 1998 to 2012. Premature deliveries occurring at<26weeks of gestation and placenta accreta were excluded. **OUTCOME MEASURES:** Prevalence of cystic periventricular leukomalacia (PVL) and cerebral palsy CP). **RESULTS:** Five infants had PVL and 4 of these infants developed CP (1/40,000 deliveries). Acute and massive bleeding (>500g within 8h) occurred at around 30-31weeks of gestation, and was severe enough to deliver the fetus. None of the 5 infants with PVL underwent antenatal corticosteroid treatment, and 1 infant had mild neonatal hypocapnia with a PaCO2 <25mmHg. However, none of the 5 PVL infants showed umbilical arterial acidemia with pH<7.2, an abnormal fetal heart rate monitoring pattern, or neonatal hypotension. **CONCLUSIONS:** Our descriptive study showed that acute and massive bleeding from placenta previa at around 30weeks of gestation may be a risk factor for CP, and requires careful neonatal follow-up. The underlying process connecting massive placental bleeding and PVL requires further investigation.

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**PMID: 24998493** [PubMed - as supplied by publisher]


**Effects of hypothermia for perinatal asphyxia on childhood outcomes.**


**BACKGROUND:** In the Total Body Hypothermia for Neonatal Encephalopathy Trial (TOBY), newborns with
asphyxial encephalopathy who received hypothermic therapy had improved neurologic outcomes at 18 months of age, but it is uncertain whether such therapy results in longer-term neurocognitive benefits. METHODS: We randomly assigned 325 newborns with asphyxial encephalopathy who were born at a gestational age of 36 weeks or more to receive standard care alone (control) or standard care with hypothermia to a rectal temperature of 33 to 34°C for 72 hours within 6 hours after birth. We evaluated the neurocognitive function of these children at 6 to 7 years of age. The primary outcome of this analysis was the frequency of survival with an IQ score of 85 or higher. RESULTS: A total of 75 of 145 children (52%) in the hypothermia group versus 52 of 132 (39%) in the control group survived with an IQ score of 85 or more (relative risk, 1.31; P=0.04). The proportions of children who died were similar in the hypothermia group and the control group (29% and 30%, respectively). More children in the hypothermia group than in the control group survived without neurologic abnormalities (65 of 145 [45%] vs. 37 of 132 [28%]; relative risk, 1.60; 95% confidence interval, 1.15 to 2.22). Among survivors, children in the hypothermia group, as compared with those in the control group, had significant reductions in the risk of cerebral palsy (21% vs. 36%, P=0.03) and the risk of moderate or severe disability (22% vs. 37%, P=0.03); they also had significantly better motor-function scores. There was no significant between-group difference in parental assessments of children's health status and in results on 10 of 11 psychometric tests. CONCLUSIONS: Moderate hypothermia after perinatal asphyxia resulted in improved neurocognitive outcomes in middle childhood. (Funded by the United Kingdom Medical Research Council and others; TOBY ClinicalTrials.gov number, NCT01092637.).

PMID: 25006720 [PubMed - in process]


Hypoxic/Ischemic and Infectious Events Have Cumulative Effects on the Risk of Cerebral Palsy in Very-Low-Birth-Weight Preterm Infants.

Wang LW1, Lin YC, Wang ST, Yeh TF, Huang CC.

Background: Hypoxia/ischemia and inflammation are two major mechanisms for cerebral palsy (CP) in preterm infants. Objective: To investigate whether hypoxia/ischemia- and infection-related events in the perinatal and neonatal periods had cumulative effects on CP risk in very-low-birth-weight (VLBW) premature infants. Methods: From 1995 to 2005, 5,807 VLBW preterm infants admitted to Taiwan hospitals were enrolled. The cumulative effects of hypoxic/ischemic and infectious events during the perinatal and neonatal periods on CP risk at corrected age 24 months were analyzed. Results: Of the 4,355 infants with 24-month follow-up, 457 (10.5%) had CP. The CP group had significantly higher incidences of hypoxia/ischemia-related events in the perinatal and neonatal periods, and sepsis in the neonatal period than the normal group. Three hypoxic/ischemic events, including birth cardiopulmonary resuscitation (OR 2.25; 95% CI 1.81 - 2.82), patent ductus arteriosus (PDA) ligation (2.94; 1.35 - 5.75) and chronic lung disease (3.14; 2.61 - 3.85) had the most significant contribution to CP. Relative to CP risk for infants with neither the three hypoxic/ischemic events nor sepsis, the CP odds increased 1.98-, 2.26- and 2.15-fold for infants with birth cardiopulmonary resuscitation, PDA ligation and chronic lung disease, respectively; while the combination with sepsis further increased the odds to 3.18-, 3.83- and 3.25-fold, respectively. Using the three hypoxic/ischemic events plus sepsis, CP rates were 10.0, 16.7, 26.7, 40.0 and 54.7% for infants with none, one, two, three and four events, respectively. Conclusions: Hypoxic/ischemic and infectious events across the perinatal and neonatal periods exerted cumulative effects on CP risk in VLBW premature infants. © 2014 S. Karger AG, Basel.

PMID: 25012626 [PubMed - as supplied by publisher]


Assessment of the structural brain network reveals altered connectivity in children with unilateral cerebral palsy due to periventricular white matter lesions.

Pannek K1, Boyd RN2, Fiori S3, Guzzetta A4, Rose SE5.

BACKGROUND: Cerebral palsy (CP) is a term to describe the spectrum of disorders of impaired motor and sensory function caused by a brain lesion occurring early during development. Diffusion MRI and tractography have been shown to be useful in the study of white matter (WM) microstructure in tracts likely to be impacted by the static brain lesion. AIM: The purpose of this study was to identify WM pathways with altered connectivity in children with
unilateral CP caused by periventricular white matter lesions using a whole-brain connectivity approach. METHODS: Data of 50 children with unilateral CP caused by periventricular white matter lesions (5-17 years; manual ability classification system [MACS] I = 25/II = 25) and 17 children with typical development (CTD; 7-16 years) were analysed. Structural and High Angular Resolution Diffusion weighted Images (HARDI; 64 directions, b = 3000 s/mm (2)) were acquired at 3 T. Connectomes were calculated using whole-brain probabilistic tractography in combination with structural parcellation of the cortex and subcortical structures. Connections with altered fractional anisotropy (FA) in children with unilateral CP compared to CTD were identified using network-based statistics (NBS). The relationship between FA and performance of the impaired hand in bimanual tasks (Assisting Hand Assessment - AHA) was assessed in connections that showed significant differences in FA compared to CTD. RESULTS: FA was reduced in children with unilateral CP compared to CTD. Seven pathways, including the corticospinal, thalamocortical, and fronto-parietal association pathways were identified simultaneously in children with left and right unilateral CP. There was a positive relationship between performance of the impaired hand in bimanual tasks and FA within the cortico-splinal and thalamo-cortical pathways (r(2) = 0.16-0.44; p < 0.05). CONCLUSION: This study shows that network-based analysis of structural connectivity can identify alterations in FA in unilateral CP, and that these alterations in FA are related to clinical function. Application of this connectome-based analysis to investigate alterations in connectivity following treatment may elucidate the neurological correlates of improved functioning due to intervention.

PMID: 25003031 [PubMed] PMCID: PMC4081979 Free PMC Article


Mendez-Figueroa H1, Dahlke JD, Viteri OA, Chauhan SP, Rouse DJ, Sibai BM, Blackwell SC.

OBJECTIVE: To describe the perinatal and infant and early childhood morbidity associated with preterm premature rupture of membranes (PROM) in a cohort of twin pregnancies evaluated prospectively with neonatal follow-up to 2 years of age. METHODS: This was a secondary analysis of a randomized controlled trial of magnesium sulfate for prevention of cerebral palsy. Inclusion criteria were twin gestation with preterm PROM diagnosed between 24 0/7 and 31 6/7 weeks of gestation and planned expectant management. Latency (time from membrane rupture to delivery) and perinatal outcomes were evaluated by gestational age at membrane rupture. Long-term neonatal outcomes were also analyzed. RESULTS: Among 151 women who met inclusion criteria, the median gestational age at preterm PROM was 28.1 weeks (range 24.1-31.6 weeks). Approximately one-third of women achieved a latency of at least 1 week. Gestational age at preterm PROM (odds ratio [OR] 0.75, 95% confidence interval [CI] 0.63-0.90 for each week after 24 weeks of gestation) and cervical dilation at admission (OR 0.66, 95% CI 0.49-0.90 for each centimeter of dilation) were inversely associated with a latency period of at least 1 week. There were no stillbirths (95% CI 0-1%), but the rate of neonatal mortality was 90 per 1,000 newborns (95% CI 57-112) with a 7.3% cerebral palsy rate among survivors (95% CI 4.4-10.3%). CONCLUSION: In twin pregnancies, preterm PROM from 24 to 31 weeks of gestation is associated with a neonatal mortality rate of 9.0% and an overall cerebral palsy rate of 7.3%. A longer latency period is associated with less advanced cervical dilation and later gestational age at PROM.

LEVEL OF EVIDENCE: II.

PMID: 25004345 [PubMed - as supplied by publisher]

42. Paediatr Drugs. 2014 Jul 11. [Epub ahead of print]

Clinical Features and Pharmacotherapy of Childhood Monoamine Neurotransmitter Disorders.

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Childhood neurotransmitter disorders are increasingly recognised as an expanding group of inherited neurometabolic syndromes. They are caused by disturbance in synthesis, metabolism, and homeostasis of the monoamine neurotransmitters, including the catecholamines (dopamine, norepinephrine, and epinephrine) and serotonin. Disturbances in monoamine neurotransmission will lead to neurological symptoms that often overlap with
clinical features of other childhood neurological disorders (such as hypoxic ischaemic encephalopathy, cerebral palsy, other movement disorders, and paroxysmal conditions); consequently, neurotransmitter disorders are frequently misdiagnosed. The diagnosis of neurotransmitter disorders is made through detailed clinical assessment, analysis of cerebrospinal fluid neurotransmitters, and further supportive diagnostic investigations. Early and accurate diagnosis of neurotransmitter disorders is important, as many are amenable to therapeutic intervention. The principles of treatment for monoamine neurotransmitter disorders are mainly directly derived from understanding these metabolic pathways. In disorders characterized by enzyme deficiency, we aim to increase monoamine substrate availability, boost enzyme co-factor levels, reduce monoamine breakdown, and replace depleted levels of monoamines with pharmacological analogs as clinically indicated. Most monoamine neurotransmitter disorders lead to reduced levels of central dopamine and/or serotonin. Complete amelioration of motor symptoms is achievable in some disorders, such as Segawa's syndrome, and, in other conditions, significant improvement in quality of life can be attained with pharmacotherapy. In this review, we provide an overview of the clinical features and current treatment strategies for childhood monoamine neurotransmitter disorders.

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