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Interventions and Management

1. Arch Dis Child Educ Pract Ed. 2012 Aug;97(4):122-31.

Cerebral palsy: the whys and hows.

Fairhurst C.

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The descriptive term of cerebral palsy encompasses the largest group of childhood movement disorders. Severity and pattern of clinical involvement varies widely dependent on the area of the central nervous system compromised. A multidisciplinary team approach is vital for all the aspects of management to improve function and minimise disability. From a medical viewpoint, there are two pronged approaches. First a focus on developmental and clinical comorbidities such as communication, behaviour, epilepsy, feeding problems, gastro-oesophageal reflux and infections; and second on specifics of muscle tone, motor control and posture. With regards to the latter, there is an increasing number of available treatments including oral antispasticity and antidystonic medications, injectable botulinum toxin, multilevel orthopaedic and neurosurgical options and a variety of complementary and alternative therapies.

[PMID: 22868578](#) [PubMed - in process]

2. Assist Technol. 2012 Summer;24(2):78-86.

Wheelchair components and pulmonary function in children with cerebral palsy.

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OBJECTIVE: This study investigates the effects of four individual wheelchair components (upper extremity supports, lateral trunk supports, anterior pelvic belt, and 30 degree posterior seat tilt), on pulmonary function in prepubertal children with cerebral palsy (CP). **METHODS:** Participants who range in age from 5-10 years were evaluated using four wheelchair components in six configurations (conditions 1-6) using a planar seating simulator. The Respirationics Non Invasive Cardiac Output monitor (NICO) and MasterScreen Impulse Oscillometry System (IOS) measured pulmonary function parameters. Repeated measures ANOVA was used to analyze effect of

wheelchair conditions on total airway resistance (R(AW)). RESULTS: Eight participants completed the protocol. R (AW) and minute ventilation (MV) varied with wheelchair condition. Lowest R(AW) was seen with two upper extremity supports or two lateral trunk supports. Differences were not significant ($p = 0.253$). CONCLUSIONS: The NICO and IOS, independent of participant effort, measured R(AW) and MV, which varied by wheelchair seating condition. More research is needed with a larger sample to determine seating components' impact on pulmonary function. These methods objectively measured pulmonary function of young children with CP in wheelchairs and could facilitate further research into benefits of wheelchair postural support components.

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3. BMC Pediatr. 2012 Aug 9;12(1):120. [Epub ahead of print]

Evaluation of the effects of Botulinum toxin A injections when used to improve ease of care and comfort in children with cerebral palsy whom are non-ambulant: a double blind randomized controlled trial.

Thorley M, Donaghey S, Edwards P, Copeland L, Kentish M, McLennan K, Lindsley J, Gascoigne-Pees L, Sakzewski L, Boyd

BACKGROUND: Children with cerebral palsy (CP) whom are non-ambulant are at risk of reduced quality of life and poor health status. Severe spasticity leads to discomfort and pain. Carer burden for families is significant. This study aims to determine whether intramuscular injections of Botulinum Toxin A (BoNT-A) combined with a regime of standard therapy has a positive effect on care and comfort for children with CP whom are non-ambulant (GMFCS IV/V), compared with standard therapy alone (cycle I), and whether repeated injections with the same regime of adjunctive therapy results in greater benefits compared with a single injecting episode (cycle II). The regime of therapy will include serial casting, splinting and/or provision of orthoses, as indicated, combined with four sessions of goal directed occupational therapy or physiotherapy. **Method/design** This study is a double blind randomized controlled trial. Forty participants will be recruited. In cycle I, participants will be randomized to either a treatment group who will receive BoNT-A injections into selected upper and/or lower limb muscles, or a control group who will undergo sham injections. Both groups will receive occupational therapy and /or physiotherapy following injections. Groups will be assessed at baseline then compared at 4 and 16 weeks following injections or sham control. Parents, treating clinicians and assessors will be masked to group allocation. In cycle II, all participants will undergo intramuscular BoNT-A injections to selected upper and/or lower limb muscles, followed by therapy. The primary outcome measure will be change in parent ratings in identified areas of concern for their child's care and comfort, using the Canadian Occupational Performance Measure (COPM). Secondary measures will include the Care and Comfort Hypertonicity Scale (ease of care), the Cerebral Palsy Quality of Life Questionnaire (CP QoL-Child) (quality of life), the Caregiver Priorities and Child Health Index of Life with Disabilities Questionnaire (CPCHILD(c)) (health status) and the Paediatric Pain Profile (PPP) (pain). Adverse events will be carefully monitored by a clinician masked to group allocation. **DISCUSSION:** This paper outlines the theoretical basis, study hypotheses and outcome measures for a trial of BoNT-A injections and therapy for children with non-ambulant cerebral palsy. Trial registration Australia New Zealand Clinical Trials Registry:N12609000360213.

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4. Gait Posture. 2012 Aug 4. [Epub ahead of print]

Differences in implementation of gait analysis recommendations based on affiliation with a gait laboratory.

Wren TA, Elihu KJ, Mansour S, Rethlefsen SA, Ryan DD, Smith ML, Kay RM.

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This study examined the extent to which gait analysis recommendations are followed by orthopedic surgeons with varying degrees of affiliation with the gait laboratory. Surgical data were retrospectively examined for 95 patients with cerebral palsy who underwent lower extremity orthopedic surgery following gait analysis. Thirty-three patients were referred by two surgeons directly affiliated with the gait laboratory (direct affiliation), 44 were referred by five surgeons from the same institution but not directly affiliated with the gait laboratory (institutional affiliation), and 18 were referred by 10 surgeons from other institutions (no affiliation). Data on specific surgeries were collected from

the gait analysis referral, gait analysis report, and operative notes. Adherence to the gait analysis recommendations was calculated by dividing the number of procedures where the surgery followed the gait analysis recommendation (numerator) by the total number of procedures initially planned, recommended by gait analysis, or done (denominator). Adherence with the gait analysis recommendations was 97%, 94%, and 77% for the direct, institutional, and no affiliation groups, respectively. Procedures recommended for additions to the surgical plan were added 98%, 87%, and 77% of the time. Procedures recommended for elimination were dropped 100%, 89%, and 88% of the time. Of 81 patients who had specific surgical plans prior to gait analysis, changes were implemented in 84% (68/81) following gait analysis recommendations. Gait analysis influences the treatment decisions of surgeons regardless of affiliation with the gait laboratory, although the influence is stronger for surgeons who practice within the same institution as the gait laboratory.

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5. Hip Int. 2012 Aug 7:0. doi: 10.5301/HIP.2012.9453. [Epub ahead of print]

Hip flexion deformity improves without psoas-lengthening after surgical correction of fixed knee flexion deformity in spastic diplegia.

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Background: It is unclear if psoas lengthening surgery is required in the treatment of patients with cerebral palsy (CP) with hip flexion deformity and previous studies show equivocal results with regard to functional outcome. **Methods:** This study retrospectively assessed 12 patients with a diagnosis of spastic diplegia who underwent single event multilevel surgery in order to correct deformities in the sagittal plane distal to the hip. Both clinical and instrument gait analysis results were recorded preoperatively, at one year (short term) and at five years (mid term) postoperatively. **Results:** Clinically measured hip and knee movement improved at both short and mid term follow up. Correlations of clinically measured maximum hip and knee extension were significant at all three time points. Angles at terminal stance/toe off for hip and knee from kinematic data also showed significant correlations at all three time points. **Conclusions:** Our study demonstrates that the hip flexion deformities encountered in these patients will improve spontaneously when the distal fixed knee flexion deformity is surgically corrected. Therefore correction at the knee allows the ground reaction force to assume a more normal position resulting in correction at the hip over time. This then removes the need for surgery at the hip level. This fact is especially important when applied to psoas lengthening as this procedure can cause significant reduction in propulsion power.

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6. J Biomech. 2012 Aug 3. [Epub ahead of print]

Medial gastrocnemius muscle fascicle active torque-length and Achilles tendon properties in young adults with spastic cerebral palsy.

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Individuals with spastic cerebral palsy (CP) typically experience muscle weakness. The mechanisms responsible for muscle weakness in spastic CP are complex and may be influenced by the intrinsic mechanical properties of the muscle and tendon. The purpose of this study was to investigate the medial gastrocnemius (MG) muscle fascicle active torque-length and Achilles tendon properties in young adults with spastic CP. Nine relatively high functioning young adults with spastic CP (GMFCS I, 17±2 years) and 10 typically developing individuals (18±2 years) participated in the study. Active MG torque-length and Achilles tendon properties were assessed under controlled conditions on a dynamometer. EMG was recorded from leg muscles and ultrasound was used to measure MG

fascicle length and Achilles tendon length during maximal isometric contractions at five ankle angles throughout the available range of motion and during passive rotations imposed by the dynamometer. Compared to the typically developing group, the spastic CP group had 33% lower active ankle plantarflexion torque across the available range of ankle joint motion, partially explained by 37% smaller MG muscle and 4% greater antagonistic co-contraction. The Achilles tendon slack length was also 10% longer in the spastic CP group. This study confirms young adults with mild spastic CP have altered muscle-tendon mechanical properties. The adaptation of a longer Achilles tendon may facilitate a greater storage and recovery of elastic energy and partially compensate for decreased force and work production by the small muscles of the triceps surae during activities such as locomotion.

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7. J Neurol. 2012 Aug 10. [Epub ahead of print]

Botulinum toxin therapy: its use for neurological disorders of the autonomic nervous system.

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Botulinum toxin (BoNT) has gained widespread use for the treatment of overactive muscles, overactive exocrine glands and, most recently, non-muscular pain conditions. Autonomic conditions treated with BoNT include achalasia, gastroparesis, sphincter of Oddi spasms, and unspecific esophageal spasms in gastroenterology and prostate disorders in urology. BoNT's use for autonomic conditions related to neurology includes various forms of bladder dysfunction (detrusor sphincter dyssynergia, idiopathic detrusor overactivity, neurogenic detrusor overactivity, urinary retention and bladder pain syndrome), pelvic floor disorders (pelvic floor spasms and anal fissures), hyperhidrosis (axillary, palmar, and plantar hyperhidrosis, diffuse sweating, Frey's syndrome) and hypersalivation (hypersalivation in Parkinsonian syndromes, motor neuron disease, neuroleptic use, and cerebral palsy). Hyperhidrosis, hypersalivation, some forms of bladder dysfunction and pelvic floor disorders can easily be treated by neurologists. Most bladder dysfunctions require cooperation with urology departments.

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8. Spine (Phila Pa 1976). 2012 Aug 3. [Epub ahead of print]

Mortality and Morbidity in Early Onset Scoliosis Surgery.

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STRUCTURED: Study Design. Retrospective chart review. Objective. To accurately determine complication rates, particularly mortality rates, in surgically treated Early Onset Scoliosis. Summary of Background Data. The advent of modern segmental instrumentation for spinal fusion surgery in adolescent scoliosis has allowed for application of similar non-segmental un-fused techniques aimed at controlling scoliosis in the very young child. The dismal prognosis for these children without repeated spinal lengthening procedures is unquestioned though no controlled trials exist. Many if not most of these children need surgery, however the surgical complication rate is very high. Methods. During the study period all surgically treated children with EOS seen at our institution were identified. Inclusion criteria were: any patient who presented to our clinic with early onset scoliosis which was surgically managed. The total number of procedures, type of implants, number and type of complications, geographic origin of the cases and final outcomes were all assessed. Results. A total of 165 surgical procedures on 28 patients accrued

over the study time period, including index implantation of instrumentation, lengthening and definitive fusion as well as operations performed for complications such as wound debridement and revision of failed implants. Clinical diagnoses included congenital scoliosis, syndromic and chromosomal abnormalities, cerebral palsy and spinal muscular atrophy. There was a complication rate of 84% overall with a mortality rate of almost 18%. The only patients with no complications were those whose entire surgical course had been at our institution only. The mortality rate was equal in patients whose treatment was performed elsewhere versus exclusively in our center. Conclusion. This study underlines the grave severity of these scolioses particularly in syndromic children. The high mortality rate is alarming, suggesting that further study is needed in this area.

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9. Clin Linguist Phon. 2012 Sep;26(9):806-22.

Relationship between kinematics, F2 slope and speech intelligibility in dysarthria due to cerebral palsy.

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A multimodal approach combining acoustics, intelligibility ratings, articulography and surface electromyography was used to examine the characteristics of dysarthria due to cerebral palsy (CP). CV syllables were studied by obtaining the slope of F2 transition during the diphthong, tongue-jaw kinematics during the release of the onset consonant, and the related submental muscle activities and relating these measures to speech intelligibility. The results show that larger reductions of F2 slope are correlated with lower intelligibility in CP-related dysarthria. Among the three speakers with CP, the speaker with the lowest F2 slope and intelligibility showed smallest tongue release movement and largest jaw opening movement. The other two speakers with CP were comparable in the amplitude and velocity of tongue movements, but one speaker had abnormally prolonged jaw movement. The tongue-jaw coordination pattern found in the speakers with CP could be either compensatory or subject to an incompletely developed oromotor control system.

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10. Oral Surg Oral Med Oral Pathol Oral Radiol. 2012 Sep;114(3):e10-4. Epub 2012 Feb 28.

Oral myiasis: does an indication for surgical treatment still exist? Two case reports.

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OBJECTIVE: Oral myiasis is a rare infection for which treatment protocol has not yet been established. This article presents 2 cases treated with a combination of topical application of sulfuric ether and surgery. The reasons for the use of surgical therapy, as well as the possible advantages and disadvantages of drug-based treatments, are discussed. **CASE REPORT:** Two cases of oral myiasis are described, the first being observed in a 9-year-old child with hypotonic cerebral palsy, and the second in a 52-year-old adult, alcohol-dependant, both showing infection in the gingival sulcus. Both cases were successfully treated in a process that involved topical application of sulfuric ether, mechanical removal of larvae, and surgical debridement. **CONCLUSIONS:** Oral myiasis can be treated effectively with surgery after topical application of sulfuric ether. The use of drugs may suggest a therapeutic alternative, but still requires further study and experience to be implemented, especially in individuals with neurological disorders.

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11. Int J Rehabil Res. 2012 Aug 3. [Epub ahead of print]**Comparison between utility of the Thai Pediatric Quality of Life Inventory 4.0 Generic Core Scales and 3.0 Cerebral Palsy Module.**

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Health-related quality of life (HRQOL) is increasingly being considered in the management of patients with various conditions. HRQOL instruments can be broadly classified as generic or disease-specific measures. Several generic HRQOL instruments in different languages have been developed for paediatric populations including the Pediatric Quality of Life Inventory 4.0 (PedsQL 4.0) Generic Core Scale. This tool and a condition-specific tool, PedsQL 3.0 Cerebral Palsy (CP) Module, are widely used in children with CP. No psychometric properties have been reported for Thai PedsQL 4.0. Therefore, this study aimed to explore the psychometric properties of the Thai version of the PedsQL 4.0 Generic Core Scales and compare these with the values for the Thai PedsQL 3.0 CP Module reported previously. Thai PedsQL 4.0 Generic Core Scales and the PedsQL 3.0 CP Module were completed, respectively, by children with CP and their parents or caregivers twice within 2-4 weeks. Respondents were 97 parents or caregivers and 54 children. Minimal missing data were found in most scales. Acceptable internal consistency was supported, except for Emotional, Social, and School Functioning. Intraclass correlation coefficients for parent-proxy report and self-report were good to excellent (0.625-0.849). The feasibility and reliability of the Thai PedsQL 4.0 Generic Core Scales were supported. The Thai PedsQL 3.0 CP Module showed higher values for the psychometric properties. Low-to-good correlations were found among the scales between the PedsQL 4.0 Generic Core Scales and the 3.0 CP Module. Both instruments could be used to measure HRQOL for children with CP, and may provide different information.

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12. Soc Work Health Care. 2012;51(4):279-95.**Social workers as transition brokers: facilitating the transition from pediatric to adult medical care.**

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Transition from pediatric to adult medical care and the significant psychosocial considerations impacting this developmental process are a primary focus in health care today. Social workers are often the informal brokers of this complex and nuanced process and are uniquely trained to complete biopsychosocial assessments to understand the needs of patients and families and address psychosocial factors. Their extensive knowledge of resources and systems, along with their sophisticated understanding of the relationship issues, family dynamics, cultural implications, and basic person-in-context approach allow for unique collaboration with the health care team, family, and community supports to develop successful transition plans and programs.

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Prevention and Cure

13. Genet Couns. 2012;23(2):215-21.

Pure distal 9p deletion in a female infant with cerebral palsy.

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We report cytogenetic and molecular characterization of a 15.63-Mb pure distal deletion of chromosome 9p (9p22.3 -->pter) in a 1 1/2-year-old female infant with cerebral palsy and diffuse cerebral dysfunction. The deletion is of paternal origin and encompasses the genes of ANKRDS15, DOCK8, FOXD4 and VLDLR. We discuss the genotype -phenotype correlation in this case with neurological dysfunction and a distal 9p deletion of paternal origin.

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