Interventions


Parent proxy-reported quality of life for children with cerebral palsy: is it related to parental psychosocial distress?

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Background: Parent-proxy reports of quality of life (QOL) are often used to guide decisions about children with cerebral palsy (CP), although little is known about the factors that influence parent-proxy reports. The aim of this study was to examine (i) the relationship between parental psychosocial distress and parent proxy-reported QOL; and (ii) whether parental psychosocial distress mediates the relationship between child impairment and proxy-reported QOL. Methods: A sample of 201 primary caregivers of children aged 4-12 years with CP completed the Cerebral Palsy Quality of Life Questionnaire for Children, a condition-specific QOL instrument, and a measure of psychosocial distress, the Kessler 10. The children, evenly distributed by gender (56% male) were sampled across Gross Motor Function Classification System levels (Level I = 18%, II = 28%, III = 14%, IV = 11%, V = 27%). Results: Consistent with the hypotheses, parental distress was negatively correlated with all domains of parent proxy-reported QOL (r=-0.18 to r=-0.55). The relationship between impairment and proxy-reported QOL was mediated by parental distress for five of the seven domains of QOL (social well-being and acceptance, feelings about functioning, participation and physical health, emotional well-being and self-esteem, and pain and impact of disability). Child impairment did not predict access to services or family health. Conclusion: This is the first study that assesses the relationship between parental distress and proxy-reported QOL for children with CP. Although the cross-sectional nature of the available data precludes any statements of causality, the results suggest that, when using parent proxy, the parents' psychological state should also be measured. This is particularly important when, as is often the case for child disability research, proxy-reported QOL are the only available data.

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Objective: To systematically review the impact of different interventions on quality of life (QoL) for children with cerebral palsy. Methods: English articles were sought from five major English databases from inceptions until March 2010. Keywords used consisted of four components (and their variants): (i) clinical condition: cerebral palsy; (ii) outcome measures: quality of life, well-being; (iii) study design: clinical trials; and (iv) target population: people aged 0-18. Results: Eight studies satisfied the inclusion criteria, all of which are of good to excellent quality (a Jadad score of 4 or above). The Pediatric Evaluation of Disability Inventory, the Pediatric Quality of Life Inventory, the TNO-AZL Children's Health-Related Quality of Life and the Caregiver Priorities and Child Health Index of Life with Disabilities were used to measure QoL. Significant positive results were reported by two studies using medical interventions (diazepam and intrathecal baclofen therapy, effect sizes 5.9, 9.1 respectively) and two studies employing motor control approach training (strength training and exercise training, former effect size being 3.8). Conclusion: Current review suggests that positive effect was shown in medicinal and motor control interventions on QoL. However, no single interventional approach can demonstrate a consistent positive impact on QoL across different studies. Future studies are recommended to (i) provide a clear definition of QoL, and investigate the relationship between symptoms' severity and QoL; (ii) measure outcome at different time points to capture real effects of interventions; and (iii) make more use of valid outcome instruments, either self-report or parent/caregiver proxy reports.

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Functional decline in children undergoing selective dorsal rhizotomy after age 10.

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Aim: To compare function and gait in a group of children older than most children who received selective dorsal rhizotomy (SDR) with age- and function-matched peers who received either orthopedic surgery or no surgical intervention. Method: A retrospective study examined ambulatory children with diplegic cerebral palsy, aged between 10 years and 20 years and categorized in Gross Motor Function Classification System (GMFCS) levels I or II. Three groups were considered: (1) children who had selective dorsal rhizotomy (n=8; two females, six males; mean age 15y 4mo at SDR, 16y 8mo at follow-up); (2) children who had orthopedic surgery (n=9; three females, six males; mean age 14y 6mo at SDR, 15y 1mo at follow-up), and (3) children who had no surgical intervention (n=9; two females, seven males; mean age 15y 6mo at follow-up). Longitudinal measures of gait analysis (velocity, gait deviation index, and gait variable scores) and gross motor function (GMFCS level, Gross Motor Function Measure scores, and centiles) were examined. Results: No significant differences were found between changes in gait comparing rhizotomy with orthopedic surgery; however, the group who received orthopedic surgery demonstrated improved gait compared with the group without surgical intervention. Longitudinal comparisons of gross motor function demonstrated a decrease in the group who received SDR. Between-group analysis of outcomes also demonstrated worse outcomes of the SDR group compared with the orthopedic surgery group and with the no surgical intervention group. Interpretation: Rhizotomy in older children was associated with functional declines compared with similar children who had no surgery and with those who underwent orthopedic surgery. This suggests that age greater than 10 years might be a contraindication for SDR if the goal is to improve motor skills.


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‘Causal relation between spasticity, strength, gross motor function, and functional outcome in children with cerebral palsy: a path analysis’

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Publication Types - LETTER


Effects of a supported speed treadmill training exercise program on impairment and function for children with cerebral palsy.

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Aim: To compare the effects of a supported speed treadmill training exercise program (SSTTEP) with exercise on spasticity, strength, motor control, gait spatiotemporal parameters, gross motor skills, and physical function.

Method: Twenty-six children (14 males, 12 females; mean age 9y 6mo, SD 2y 2mo) with spastic cerebral palsy (CP; diplegia, n=12; triplegia, n=2; quadriplegia n=12; Gross Motor Function Classification System levels II-IV) were randomly assigned to the SSTTEP or exercise (strengthening) group. After a twice daily, 2-week induction, children continued the intervention at home 5 days a week for 10 weeks. Data collected at baseline, after 12-weeks’ intervention, and 4 weeks after the intervention stopped included spasticity, motor control, and strength; gait spatiotemporal parameters; Gross Motor Function Measure (GMFM); and Pediatric Outcomes Data Collection Instrument (PODCI).

Results: Gait speed, cadence, and PODCI global scores improved, with no difference between groups. No significant changes were seen in spasticity, strength, motor control, GMFM scores, or PODCI transfers and mobility. Post-hoc testing showed that gains in gait speed and PODCI global scores were maintained in the SSTTEP group after withdrawal of the intervention. Interpretation: Although our hypothesis that the SSTTEP group would have better outcomes was not supported, results are encouraging as children in both groups showed changes in function and gait. Only the SSTTEP group maintained gains after withdrawal of intervention.


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Botulinum toxin a (dysport®): in dystonias and focal spasticity.

Keam SJ, Muir VJ, Deeks ED.


Dysport®, a formulation of botulinum toxin A, blocks acetylcholine release at neuromuscular junctions causing denervation and temporary muscle paralysis. It is used to treat several medical conditions, including dystonias and focal spasticity. Subcutaneous Dysport® was effective in improving functional disability in adults with blepharospasm in a placebo-controlled trial with 16 weeks’ follow-up, and in adults with hemifacial spasm in case series. Similarly, intramuscular Dysport® was effective in improving symptoms of cervical dystonia in adults, focal spasticity in adults with post-stroke upper limb spasticity and dynamic equinus spasticity in paediatric patients with cerebral palsy in
placebo-controlled trials with up to 20 weeks' follow-up. However, in two 12-week, placebo-controlled trials in adults with focal lower limb spasticity (spastic equinovarus deformity after stroke and hip adductor spasticity associated with multiple sclerosis) intramuscular Dysport® had limited efficacy. Available longer-term data indicated that Dysport® treatment was effective over several treatment cycles in patients with cervical dystonia or upper limb spasticity. Dysport® was generally well tolerated in patients with dystonias or focal spasticity. Most adverse events were mild to moderate and transient.

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Quantitative evaluations of ankle spasticity and stiffness in neurological disorders using manual spasticity evaluator.

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Spasticity and contracture are major sources of disability in people with neurological impairments that have been evaluated using various instruments: the Modified Ashworth Scale, tendon reflex scale, pendulum test, mechanical perturbations, and passive joint range of motion (ROM). These measures generally are either convenient to use in clinics but not quantitative or they are quantitative but difficult to use conveniently in clinics. We have developed a manual spasticity evaluator (MSE) to evaluate spasticity/contracture quantitatively and conveniently, with ankle ROM and stiffness measured at a controlled low velocity and joint resistance and Tardieu catch angle measured at several higher velocities. We found that the Tardieu catch angle was linearly related to the velocity, indicating that increased resistance at higher velocities was felt at further stiffer positions and, thus, that the velocity dependence of spasticity may also be position-dependent. This finding indicates the need to control velocity in spasticity evaluation, which is achieved with the MSE. Quantitative measurements of spasticity, stiffness, and ROM can lead to more accurate characterizations of pathological conditions and outcome evaluations of interventions, potentially contributing to better healthcare services for patients with neurological disorders such as cerebral palsy, spinal cord injury, traumatic brain injury, and stroke.

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The presence of physiological stress shielding in the degenerative cycle of musculoskeletal disorders.

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The health of physiological tissue is governed by the continuous conversion of mechanical stimulus (stress) to biochemical response, a concept known as mechanical homeostasis. If this regulatory imperative becomes flawed, it may be detrimental, and consequently invoke or encourage the progression of various musculoskeletal disorders. This notion is corroborated by the quantification of altered function and irregular mechanical properties found within the articulations of such phenotypes as cerebral palsy. Although the divergence from healthy to irregular tissue behavior is likely multifactorial, the presence of imbalanced mechanical properties may promote the concept of physiological stress shielding. Extrapolating upon the stress shielding phenomenon may allow inferences to be drawn with respect to the pathomechanisms of progressive disorders. Further, recognition of this association may also provide a new platform from which to interpret the impact of conventional treatments aimed at such syndromes and, in turn, perhaps support new therapeutic avenues.

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Results and complications following spinal fusion for neuromuscular scoliosis in cerebral palsy and static encephalopathy using luque galveston instrumentation: experience in 93 patients.

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Study Design: Retrospective Cohort Analysis. Objective: To evaluate the results of spine fusion for neuromuscular scoliosis in cerebral palsy and static encephalopathy, using Luque Galveston instrumentation, with emphasis on the early and late complications especially those increasing the hospital stay or requiring additional surgery. Summary of Background Data: There are numerous studies in the literature on the treatment of neuromuscular scoliosis using Luque Galveston instrumentation analyzing the results and complications. Most series are small and evaluate some of the complications, none evaluating all the early and late complications and none assessing the impact of the complication on length of hospitalization or the need for additional surgical intervention. Methods: This was a retrospective review of a consecutive series of patients operated on for neuromuscular scoliosis in cerebral palsy and static encephalopathy with Luque-Galveston instrumentation at one institution from 1/1/1997 to 12/31/2003. Ninety-three patients were identified. Results: The average age at surgery was 14.3 years, with the average age at follow-up of 18.2 years, and an average follow-up of 3.8 years. The mean pre-operative scoliosis was 72° with correction post-operatively to a mean of 33°, maintained at 36° at final follow-up. There were 83 early complications in 54 patients for a complication rate of 58% of patients for the entire study. There were no peri-operative deaths or neurological complications. There was only one deep wound infection for an infection rate of 1.1%. Patients with one complication had a longer length of stay, nine vs. seven days, the difference being statistically significant (Mann-Whitney test, p<0.001). Two patients required re-operation during the initial hospitalization (1.1%), one for infection and one for proximal hook cutout and proximal junction kyphosis. There were a total of 81 late complications in 44 patients for a late complication rate of 47% of patients for the entire study. The majority of the complications were minor not requiring additional care or surgery. Seven patients had a pseudarthrosis (7.5%), presenting at an average of thirty months post-operatively. Eight patients underwent nine procedures for late complications, five for repair of a pseudarthrosis, three for removal of a prominent iliac screw, and one for superior junctional kyphosis. All the pseudarthrosis repairs were solid at follow-up. Conclusion: Spinal fusion in neuromuscular scoliosis with Luque-Galveston technique is a safe and effective procedure. Any early complication increased the length of stay, with a low rate of re-operation during the hospitalization. The majority of late complications were minor, not requiring additional care. Pseudarthroses were detected late, and were the main reason for additional surgery.

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Sleep in Children With Cerebral Palsy: A Review.

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Children with neurodevelopmental disabilities, such as cerebral palsy, are considered to be a population at risk for the occurrence of sleep problems. Moreover, recent studies on children with cerebral palsy seem to indicate that this population is at higher risk for sleep disorders. The importance of the recognition and treatment of sleep problems in children with cerebral palsy cannot be overstressed. It is well known that the consequences of sleep disorders in children are broad and affect both the child and family. This review article explores the types and possible risk factors associated with the development of sleep problems in children with cerebral palsy and the impact of this disorder on the child and family. In addition, a brief summary of current diagnostic and treatment modalities is provided. Finally, the characteristics, diagnostic techniques, and management of sleep-related breathing disorders in children with cerebral palsy are discussed.

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Visual Attention Study in Youth With Spastic Cerebral Palsy Using the Event-Related Potential Method.

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Youth with mild spastic cerebral palsy (n = 14) and a peer control group were compared on an oddball paradigm. Here, visual stimuli were presented with low and high probability and participants were instructed to count in silence the number of rare stimuli. The infrequent stimulus typically elicits an enhanced frontal central N2 and a centroparietal P300 event-related brain potential, reflecting orientation and evaluation of stimulus novelty. No differences in latency and amplitude of the N2-P300 complex were found between the 2 groups, indicating that some fundamental attention processes are intact in youth with mild spastic cerebral palsy.

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Obesity and overweight prevalence among adolescents with disabilities.

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INTRODUCTION: We examined overweight and obesity prevalence among adolescents with disabilities by disability type (physical vs cognitive) and demographic factors (sex, age, race/ethnicity). METHODS: Parents (N = 662) of adolescents aged 12 to 18 years with disabilities from 49 states responded to an online survey from September 2008 through March 2009. RESULTS: Prevalence of obesity among adolescents with physical and cognitive disabilities (17.5%) was significantly higher compared with that among adolescents without disabilities (13.0%). Obesity prevalence was higher among males, 18-year-olds, and youths with cognitive disabilities than among females, younger adolescents, and youths with physical disabilities. CONCLUSION: The higher prevalence of obesity among youths with disabilities compared with nondisabled youths, particularly in certain subgroups, requires further examination in future surveillance research.

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Cerebral palsy is the most common neurodevelopmental motor disability in children. The condition requires medical, educational, social, and rehabilitative resources throughout the life span. Several countries have developed population-based registries that serve the purpose of prospective longitudinal collection of etiologic, demographic, and functional severity. The United States has not created a comprehensive program to develop such a registry. Barriers have been large population size, poor interinstitution collaboration, and decentralized medical and social systems. The Cerebral Palsy Research Registry was created to fill the gap between population and clinical-based cerebral palsy registries and promote research in the field. This is accomplished by connecting persons with cere-
bral palsy, as well as their families, to a network of regional researchers. This article describes the development of an expandable cerebral palsy research registry, its current status, and the potential it has to affect families and persons with cerebral palsy in the United States and abroad.

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Prevalence and risk indicators of temporomandibular disorder signs and symptoms in a pediatric population with spastic cerebral palsy.

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OBJECTIVE: To determine risk indicators for signs and symptoms of temporomandibular disorders (TMD) in children with cerebral palsy (n = 60) and control subjects (n = 60). STUDY DESIGN: The subjects were assessed by means of questionnaire and clinical exam: 1) signs and symptoms of TMD; 2) malocclusions [Dental Aesthetic Index (DAI)]; 3) harmful habits; and 4) bio-psychosocial characteristics. Statistical analysis involved the chi-square, Fisher's exact tests (p < or = 0.05) and multivariate logistic regression (forward stepwise procedure). Variables that achieved a p-value < or = 0.20 were used as potential predictors of signs and symptoms of TMD and applied as co-variables in the multivariate analysis. RESULTS: The prevalence of at least one sign and/or symptom of TMD in the present sample was 1.7% (n = 1) among the individuals in the control group and 13.3% (n = 8) among the individuals with cerebral palsy. The presence of cerebral palsy (Odds Ratio: 9.08; p = 0.041), male gender (OR: 6.21; p = 0.027), severity of the malocclusion (OR: 4.75; p = 0.031), mouth breathing (OR: 5.40; p = 0.022) and mixed dentition (OR: 4.73; p = 0.035) were identified as risk indicators for signs and symptoms of TMD. CONCLUSIONS: It was concluded that children with cerebral palsy had a significantly greater chance of developing signs and symptoms of TMD.

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Developmental aspects of environmental neurotoxicology: Lessons from lead and polychlorinated biphenyls.

Winneke G.

The particular vulnerability of the developing nervous system for low-level exposure to chemicals is well established. It has been argued that some degree of developmental neurotoxicity was found for a large number of industrial chemicals. However, for only few of these, namely inorganic lead, arsenic, organic mercury and polychlorinated biphenyls (PCBs), human evidence is available to suggest that these may cause neurodevelopmental adversity and may, thus, be involved in contributing to neurodevelopmental disorders like autism, attention-deficit disorder, mental retardation or cerebral palsy. The focus of this overview is on PCBs and inorganic lead as developmental neurotoxicants at environmental levels of exposure. The adverse effects of inorganic lead on the developing brain have long been studied, and much emphasis has been on subtle degrees of mental retardation in terms of intelligence (IQ). The evidence is consistent, but the effect sizes are typically small. Research interest has also been devoted to studying aspects of "attention-deficit hyperactivity disorder" (ADHD) in children in relation to environmental exposure to lead in both cross-sectional and case-control studies. More recently, we have also studied core elements of ADHD according to ICD-10 and DSM-IV in relation to environmental exposure to lead, mercury and aluminum in asymptomatic school children in Romania. Both, performance measures (several attention tasks) and questionnaire-based behavior ratings from parents and teachers showed that lead, but not Hg or Al, was consistently and adversely associated with core elements of ADHD. These findings in asymptomatic children nicely fit into the overall pattern of observations and suggest that, apart from genetic influences, low-level exposure to lead contributes to this neurodevelopmental disorder. Polychlorinated biphenyls (PCBs) are persistent organic pollutants with lipophilic properties. Due to their persistence, they are still present in environmental media at potentially harmful concentrations, although production and use of PCBs was already banned in the early 1980s. Several prospective cohort studies-including our Düsseldorf study-have demonstrated that pre- and early postnatal exposure to PCBs is asso-
associated with deficit or retardation of mental and/or motor development, even after adjusting for maternal intelligence and developmental effects of the quality of the home environment. The pathophysiology is still unclear, although interference with thyroid metabolism during brain development is being discussed. Based on these reviews, three aspects, namely pre- vs. postnatal impact, effect scaling for comparative purposes, and integration of neurobehavioral findings into clinical and neuroscience contexts, are outlined as lessons learned from neurodevelopmental observations in children environmentally exposed to lead or PCBs.

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Cerebral palsy in preterm survivors.
Jobe AH.
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Predicting motor outcome and death in term hypoxic-ischemic encephalopathy.
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OBJECTIVES: Central gray matter damage, the hallmark of term acute perinatal hypoxia-ischemia, frequently leads to severe cerebral palsy and sometimes death. The precision with which these outcomes can be determined from neonatal imaging has not been fully explored. We evaluated the accuracy of early brain MRI for predicting death, the presence and severity of motor impairment, and ability to walk at 2 years in term infants with hypoxic-ischemic encephalopathy (HIE) and basal ganglia-thalamic (BGT) lesions. METHODS: From 1993 to 2007, 175 term infants with evidence of perinatal asphyxia, HIE, and BGT injury seen on early MRI scans were studied. BGT, white matter, posterior limb of the internal capsule (PLIC), and cortex and brainstem abnormality were classified by severity. Motor impairment was staged using the Gross Motor Function Classification System. RESULTS: The severity of BGT lesions was strongly associated with the severity of motor impairment (Spearman rank correlation 0.77; p < 0.001). The association between white matter, cortical, and brainstem injury and motor impairment was less strong and only BGT injury correlated significantly in a logistic regression model. The predictive accuracy of severe BGT lesions for severe motor impairment was 0.89 (95% confidence interval 0.83-0.96). Abnormal PLIC signal intensity predicted the inability to walk independently by 2 years (sensitivity 0.92, specificity 0.77, positive predictive value 0.88, negative predictive value 0.85). Brainstem injury was the only factor with an independent association with death. CONCLUSION: We have shown that in term newborns with HIE and BGT injury, early MRI can be used to predict death and specific motor outcomes.


Oligodendrocyte vulnerability following traumatic brain injury in rats.
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Experimental and clinical findings demonstrate that traumatic brain injury (TBI) results in injury to both gray and white matter structures. The purpose of this study was to document patterns of oligodendrocyte vulnerability to TBI. Sprague Dawley rats underwent sham operated procedures or moderate fluid percussion brain injury. Animals were
perfusion-fixed for quantitative immunohistochemical analysis at 3 (n=9) or 7 (n=9) days post-surgery. Within the
ipsilateral external capsule and corpus callosum, numbers of APC-CC1 immunoreactive oligodendrocytes were sig-
nificantly decreased at 3 or 7 days post-TBI compared to sham rats (p<0.03). At both posttraumatic survival peri-
ods, double-labeling studies indicated that oligodendrocytes showed increased Caspase 3 activation compared to
sham. These data demonstrate regional patterns of oligodendrocyte vulnerability after TBI and that oligodendrocyte
cell loss may be due to Caspase 3-mediated cell death mechanisms. Further studies are needed to test therapeutic
interventions that prevent trauma-induced oligodendrocyte cell death, subsequent demyelination and circuit dys-
function.

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