
Development of a framework to define the functional goals and outcomes of botulinum toxin A spasticity treatment relevant to the child and family living with cerebral palsy using the International Classification of Functioning, Disability and Health for Children and Youth.

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OBJECTIVE: To define a sub-set of functional goals and outcomes relevant to children and families living with cerebral palsy following treatment with botulinum toxin type A using the International Classification of Functioning, Disability and Health for Children and Youth. METHODS: We identified treatment goals and treatment outcomes from medical case records in 2102 assessments of 239 children with cerebral palsy treated with botulinum toxin between 1994 and 2009. Goals were set through assessment and discussion by experienced clinicians, therapists, parents and children. RESULTS: There were 61 separate goals, mapping to 40 categories, falling mostly within Body Functions: b710 Mobility of joint functions (414 times); b770 Gait pattern functions (374 times); b7351 Tone of muscles of one limb (117 times). A total of 93 separate treatment outcomes were identified, mapping to 51 categories. Two of the 3 most common outcomes correspond to the 2 most common goals (gait pattern and mobility of joint functions). CONCLUSION: The International Classification of Functioning, Disability and Health for Children and Youth provides a useful framework to categorize the reasons for using botulinum toxin in children and focuses the clinical consultation not only on impairments but also functional outcomes.

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Does gender influence the long-term outcome of single-event multilevel surgery in spastic cerebral palsy?

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This study compared the long-term outcome of single-event multilevel surgery in spastic diplegic cerebral palsy (CP) on the basis of sex. We hypothesized that boys would have a worse outcome than girls. Thirty-four children (19 boys and 15 girls) with diplegic spastic CP and a minimal follow-up of 10 years were included. The Gillette Gait Index was the main outcome measure. We found no differences in surgical treatment, and both groups initially benefited from the surgery. However, although girls maintained the enhanced level of walking, walking ability in boys deteriorated constantly. Such a finding suggests that sex might have an important influence on treatment outcomes in children with CP.

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The effects of ankle foot orthoses on energy recovery and work during gait in children with cerebral palsy.

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BACKGROUND: Studies suggest that 50% of children with cerebral palsy are prescribed ankle foot orthoses. One of the aims of ankle foot orthosis use is to aid in walking. This research examined the effects that ankle foot orthoses have on the energy recovery and the mechanical work performed by children with cerebral palsy during walking. METHODS: Twenty-one children with spastic diplegia walked with and without their prescribed bilateral ankle foot orthoses. Ten of the subjects wore articulated (hinged) orthoses and 11 subjects wore solid orthoses. Three dimensional kinematic data were collected and between and within group repeated measures ANOVAs were applied to the dependent measures. FINDINGS: The results were similar for both groups. There was an increase in stride length, energy recovery, and potential energy and the kinetic energy variation. There was no change in the mechanical work performed to walk or the normalized center of mass vertical excursion. Unfortunately, the increase in energy recovery did not alter the external work, as it was offset by increased variation in the potential and kinetic energies of the center of mass. There was a great deal of variability in the measured work, with both large increases and decreases in the work of individual subjects when wearing orthoses. INTERPRETATION: These results suggest that current ankle foot orthoses can reduce the work to walk, but do not do so for many children with cerebral palsy. This research suggests that ankle foot orthosis prescription could be aided by measuring the mechanical work during walking.

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Balance in Relation to Walking Deterioration in Adults With Spastic Bilateral Cerebral Palsy.

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Background: Balance function is central in walking, and impaired balance function may be related to walking deterioration in adults with spastic bilateral cerebral palsy (CP). Objectives: The purposes of this study were: (1) to compare balance confidence, fear of falling, and balance ability in adults with spastic bilateral CP, with and without self-reported walking deterioration; (2) to characterize balance confidence, fear of falling, and balance ability across all participants; (3) to examine the relationship between balance confidence and balance ability across all participants. Design: A case-control design was used. Methods: Sixteen adults from a 7-year follow-up study who had spastic bilateral CP and were under 40 years of age in the 2006 survey participated. Eight participants reported walking deterioration (cases), and 8 participants did not report walking deterioration (controls). Outcome variables were: the Activities-specific Balance Confidence (ABC) Scale, the Falls Efficacy Scale-International (FES-I), and the Balance Evaluation Systems Test (BESTest). Results: No differences in any of the outcome variables were found
between the cases and the controls. Across all participants, the ABC Scale and FES-I scores were 62% and 24 points, respectively. Reduced ABC Scale scores and increased FES-I scores were found when using escalators, walking in crowds, and walking on slippery surfaces. The BESTest subscale scores were 60% to 79% of the maximum score, but only 31% and 42% of the maximum score in postural responses and anticipatory adjustments, respectively. Balance confidence correlated positively with postural responses, sensory orientation, stability in gait, and BESTest total score. Limitations: The lack of reliability and validity tests for the outcome variables in this study population and the small number of participants were limitations of the study. CONCLUSIONS: Self-reported walking deterioration in this group could not be explained by differences in balance confidence, fear of falling, or balance ability. Across all participants, most balance problems seemed related to reduced postural responses and anticipatory adjustments.

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Comparison of two techniques in achieving planned correction angles in femoral subtrochanteric derotation osteotomy.

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Increased femoral anteversion in cerebral palsy alters biomechanics of gait. Femoral subtrochanteric derotational osteotomies are increasingly performed to improve gait in cerebral palsy. The amount of angular correction can be determined and planned preoperatively but, accuracy in achieving planned angular correction has not been tested experimentally before. The aim of this study was to evaluate the accuracy of the two techniques in achieving planned angular correction. Sixteen dry femora were used in this study. Specimens in both groups were derotated to achieve a desired amount of correction with two different techniques, consecutively. In technique one, the cross section of the femur was assumed to be circular and the desired amount of angular correction was calculated and expressed in terms of surface distance by a geometric formula (surface distance=2π×radius of femur). In both groups, derotations were made based on this surface distance calculation. Consecutively the same specimens were derotated by pins and guide technique. Femoral anteversion of specimens were measured before and after derotation by computerized tomography. There was a statistically significant difference in planned and achieved correction angles (P=0.038) in both subgroups derotated by the surface distance technique. When the two techniques were compared, there was significant difference (P=0.050) between high magnitude correction subgroups (subgroups 2 vs. 4). In conclusion, the results of this study highlighted the difficulty in achieving accurate derotation angles. Derotations based on guide-pins technique yielded more accurate results than derotations based on surface distance technique. In addition, surface diameter technique was not suitable when higher degrees of derotations are needed. In achieving a planned derotation angle two techniques are described for accuracy. Both the techniques have potential pitfalls resulting in malrotations. Surgeons must be aware of these obstacles and try to avoid them.

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Tuning Algorithms for Control Interfaces for Users with Upper-Limb Impairments.

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OBJECTIVE: Approximately 40% of Americans with disabilities cannot operate wheeled mobility devices and computers adequately because of diminished upper-limb motor control, sensory limitations, and cognitive impairments. We developed tuning software that can customize control interfaces for individuals with upper-limb impairments. This study compared the differences in each parameter among different diagnostic groups. DESIGN: The age of the subjects ranged from 18 to 80 yrs. The participants were classified into the following groups: athetoid cerebral palsy, spastic cerebral palsy, multiple sclerosis, upper-limb spasticity, and control. We used a validated tuning software protocol to customize an isometric joystick before a virtual tracing or driving task. Tuning parameters were then compared across groups. RESULTS: Seventy-five subjects were included. Gain, the parameter responsible for force-to-output ratios, in each directional axis (leftward gain: \( P = 0.018 \); rightward gain: \( P = 0.003 \); reverse gain: \( P = 0.007 \); forward gain: \( P = 0.014 \)) was significantly different across the diagnostic groups. Post hoc analyses showed that the control group required smaller leftward gain than spastic cerebral palsy, multiple sclerosis and upper-limb spasticity groups and smaller gain in all other directions compared with spastic cerebral palsy. CONCLUSIONS: Gain may be a useful parameter in tuning by clinicians, and efforts aimed at gain customization may aid the development of commercially available tuning software packages.

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Linking cerebral palsy upper limb measures to the International Classification of Functioning, Disability and Health.

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BACKGROUND: Intervention studies describe outcomes as measuring specific domains of the International Classification of Functioning, Disability and Health (ICF). However, the same measure may be described by different authors as assessing different domains, resulting in considerable confusion and inconsistent reporting of outcomes. OBJECTIVE: To systematically link the scored items from the Melbourne Assessment of Unilateral Upper Limb Function, Quality of Upper Extremity Skills Test and Assisting Hand Assessment to domain(s) of the ICF. METHODS: The meaningful concept for each scored item was defined. Using ICF linking rules, the concepts were assigned ICF codes to determine the outcome's overall domain of measurement. RESULTS: The Melbourne Assessment predominantly evaluates concepts in the body function domain. Coding of the Quality of Upper Extremity Skills Test indicated that dissociated movement, weight-bearing and protective extension predominantly measure concepts in the body function domain. Grasp was the only domain where concepts were coded in both the body function and activity domains. The Assisting Hand Assessment was the only measure where the majority of items assessed concepts in the activity domain. CONCLUSION: Measures of upper limb function can be categorized according to ICF domains. These findings should resolve confusion surrounding the classification of these measures and provide a reference for reporting the impact of intervention.

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Assessment of the hand in cerebral palsy.

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Cerebral palsy is the musculoskeletal manifestation of a nonprogressive central nervous system lesion that usually occurs due to a perinatal insult to the brain. Though the cerebral insult is static the musculoskeletal pathology is progressive. Some patients with cerebral palsy whose hands are affected can be made better by surgery. The surgical procedures as such are not very technically demanding but the assessment, decision-making, and selecting the procedures for the given patient make this field challenging. When done well, the results are
rewarding not only in terms of improvement in hand function but also in appearance and personal hygiene, which leads to better self-image and permits better acceptance in the society. This article focuses on the clinical examination, patient selection, and decision-making while managing these patients.

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Optimal sedative dose of propofol to start MRI in children with cerebral palsy.

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BACKGROUND: This study was designed to determine the optimal sedative dose of propofol to start brain magnetic resonance imaging (MRI) in children with cerebral palsy (CP). METHODS: Twenty children, aged 0.5-5 years, were administered propofol to achieve a University of Michigan Sedation Scale (UMSS) score ≥ 3 in the MRI room. The proper dose of propofol was determined using the up-and-down method. RESULTS: The ED50 and ED95 for successful sedation with a UMSS ≥ 3 were 2.07 mg/kg (95% CI 1.69-2.56) and 2.69 mg/kg (95% CI 2.35-5.59). Respiratory events occurred in 5 patients and were resolved with neck extension, chin lift, or transient respiratory assistance with successful sedation. CONCLUSIONS: Low dose propofol can safely facilitate the initiation of MRI in children with CP.


Acute Sialadenitis Secondary to Submandibular Calculi After Botulinum Neurotoxin Injection for Sialorrhea in a Child with Cerebral Palsy.

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Children with cerebral palsy and other neurologic diseases often present with sialorrhea. Intraglandular botulinum neurotoxin is being increasingly reported to be clinically effective for the treatment of sialorrhea. This treatment is becoming more popular in recent years because of being less invasive than surgical procedures. In addition, fewer adverse effects have been documented compared with oral or topical anticholinergic medication. We report the first case in a child with cerebral palsy who developed serious acute sialadenitis with submandibular sialolithiasis after intraglandular botulinum neurotoxin injection for sialorrhea.

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A Proposed Multidisciplinary Approach for Identifying Feeding Abnormalities in Children With Cerebral Palsy.


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Children with neurodevelopmental disabilities, such as cerebral palsy, frequently have associated oral motor dysfunction, which leads to feeding difficulties, risk of aspiration, prolonged feeding times, and malnutrition with its attendant physical compromise. The authors propose a comprehensive multidisciplinary assessment, including neurological and dysphagia examination and ear, nose, and throat examination, to evaluate clinical indicators and severity of feeding impairment in children affected by neurodevelopmental disorders. A representative sample of 40
children with cerebral palsy (spastic, dyskinetic, or mixed), intellectual disability, and feeding problems was included in the study. A specific multidisciplinary evaluation and standardized mealtime observation in patients with cerebral palsy appear feasible and appropriate to recognize proactive indicators of dysphagia and to establish personalized programs of gastric and rehabilitative interventions.

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Prevalence and predictive factors of sleep bruxism in children with and without cognitive impairment.

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Studies have found a higher prevalence of sleep bruxism (SB) in individuals with cognitive impairment. The aim of this study was to identify the prevalence and factors associated with the clinical manifestation of SB in children with and without cognitive impairment. The sample was made up of 180 individuals: Group 1 - without cognitive impairment; Group 2 - with Down syndrome; Group 3 - with cerebral palsy. Malocclusions were assessed based on the Dental Aesthetic Index (DAI); lip competence was assessed based on Ballard's description. The biopsychosocial characteristics were assessed via a questionnaire and clinical exam. Statistical analysis involved the chi-square test (p < 0.05) and multivariate logistic regression. The prevalence of bruxism was 23%. There were no significant differences between the groups (p = 0.970). Individuals with sucking habits (OR [95% CI] = 4.44 [1.5 to 13.0]), posterior crossbite (OR [95% CI] = 3.04 [1.2 to 7.5]) and tooth wear facets (OR [95% CI] = 3.32 [1.2 to 8.7]) had a greater chance of exhibiting SB. Sucking habits, posterior crossbite and tooth wear facets were identified as being directly associated with the clinical manifestations of bruxism.

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Self-injurious behavior (SIB) is causing deliberate harm to the body without suicidal intent; this problem occurs in a number of psychiatric, behavioral, and developmental disorders. This report describes the case of a girl, aged 19 months, whose lower lip injuries due to SIB were successfully treated through the use a combination of extraoral and intraoral prostheses. A custom-made lip guard with a custom-made adjustable head strap was effective in the management of the lip trauma. SIB was prevented and there was no recurrence even at the 17-month follow-up appointment.

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Failure to thrive in childhood.

Nützenadel W.


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Botulinum toxine treatment and the formation of antibodies to botulotoxin in patients with cerebral palsy. [Article in Russian]

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The aim of the study was to search for the association between efficacy of botulinum treatment (dysport) and the formation of antibodies to botulinum toxin in children with cerebral palsy (CP). Fourteen children with CP received dysport, 42 children (29 with CP and 13 patients with paresises of other etiologies) were not treated with this drug. No association between the presence of antibodies in blood serum and efficacy of treatment was found. However, further research of the role of antibody genesis as a factor influencing the formation of reaction to treatment is needed.

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Prevention and Cure


Prediction of outcome at 5 years from assessments at 2 years among extremely preterm children.


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Aim: Examine predictive value of early assessments on developmental outcome at 5 years in children born extremely preterm. Methods: Prospective observational study of all infants born in Norway in 1999-2000 with gestational age (GA) <28 weeks or birth weight <1000g. At 2 years of age, pediatricians assessed mental and motor development from milestones. At 5 years, parents completed questionnaires on development and professional support before cognitive function was assessed with WPPSI-R and motor function with the Movement Assessment Battery for children (ABC-test). Results: 26/372 (7%) had cerebral palsy at 2 and 29/306 (9%) at 5 years. Of children without major impairments, 51% (95% CI 35%-67%) of those with and 22% (95% CI 16% - 28%) without mental delay at 2 years had IQ <85 at 5 years, and 36% (95% CI 20%-53%) with and 16% (95% CI 11%-21%) without motor delay at 2 years had an ABC score >95(th) percentile (poor function). 56% of those without major impairments but IQ <85 or ABC score >95(th) percentile had received support or follow-up beyond routine primary care. Conclusion: Previous assessments had limited value in predicting cognitive and motor function at 5 years in these extremely preterm children without major impairments.


Long-term neurodevelopmental outcome after intrauterine transfusion for hemolytic disease of the fetus/newborn: the LOTUS study.


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OBJECTIVE: To determine the incidence and risk factors for neurodevelopmental impairment (NDI) in children with hemolytic disease of the fetus/newborn treated with intrauterine transfusion (IUT). STUDY DESIGN: Neurodevelopmental outcome in children at least 2 years of age was assessed using standardized tests, including the Bayley Scales of Infant Development, the Wechsler Preschool and Primary Scale of Intelligence, and the Wechsler Intelligence Scale for Children, according to the children's age. Primary outcome was the incidence of neurodevelopmental impairment defined as at least one of the following: cerebral palsy, severe developmental delay, bilateral deafness, and/or blindness. RESULTS: A total of 291 children were evaluated at a median age of 8.2 years (range, 2-17 years). Cerebral palsy was detected in 6 (2.1%) children, severe developmental delay in 9 (3.1%) children, and bilateral deafness in 3 (1.0%) children. The overall incidence of neurodevelopmental impairment was 4.8% (14/291). In a multivariate regression analysis including only preoperative risk factors, severe hydrops was independently associated with neurodevelopmental impairment (odds ratio, 11.2; 95% confidence interval, 1.7-92.7). CONCLUSION: Incidence of neurodevelopmental impairment in children treated with intrauterine transfusion for fetal alloimmune anemia is low (4.8%). Prevention of fetal hydrops, the strongest preoperative predictor for impaired neurodevelopment, by timely detection, referral and treatment may improve long-term outcome.

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Aberrant high-gamma oscillations in the somatosensory cortex of children with cerebral palsy: A meg study.


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Objective: Our study is to investigate somatosensory dysfunction in children with spastic cerebral palsy (CP) using magnetoencephalography (MEG) and synthetic aperture magnetometry (SAM). Methods: Six children with spastic CP and six age- and gender-matched typically developing children were studied using a 275-channel MEG system while their left and right index fingers were stimulated in random order. The latency and amplitude of somatosensory evoked magnetic fields were analyzed at sensor level. The patterns of high-gamma oscillations were investigated with SAM at sensor level. Results: In comparison to the children with typical development, the latency of the first response of somatosensory evoked magnetic fields (SEFs) in the children with spastic CP was significantly delayed (p<0.05). High-gamma oscillations were identified in the somatosensory cortex in both children with CP and typical developing children. Interestingly, children with spastic CP had significantly higher incidence of ipsilateral activation in the somatosensory cortex following right and left finger stimulation, compared to typically developing children (p=0.05). Conclusion: The results suggest that children with spastic CP have a measurable delay of SEFs and high-gamma oscillations. The high rates of ipsilateral cortical activation imply the impairments of functional lateralization in the developing brain. This is the first MEG study to demonstrate abnormal high-gamma oscillations in children with spastic CP.

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oscillations of somatosensory cortices representing the finger in children with spastic CP.

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Phenylketonuria in Hong Kong Chinese: a call for hyperphenylalaninemia newborn screening in the Special Administrative Region, China.

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Hyperphenylalaninemia is one of the commonest inborn errors of metabolism affecting approximately 1 in 15,000 livebirths. Among Chinese, BH4 deficiency leading to hyperphenylalaninemia is much commoner than in Caucasians. Exact diagnosis is important for the treatment and genetic counseling. In 2000, newborn screening for phenylketonuria is mandatory by law in China throughout the whole country. However, it is not yet included in the newborn screening program of the Hong Kong Special Administrative Region, China. Published data on hyperphenylalaninemia among Hong Kong Chinese are largely lacking. We report a 1-year-old Hong Kong Chinese girl with severe 6-pyruvoyl-tetrahydropterin synthase (PTPS) deficiency. The patient presented with infantile hypotonia and was misdiagnosed as cerebral palsy. She had very mild hyperphenylalaninemia (95 μmol/L), significantly high phenylalanine-to-tyrosine ratio (3.1), and elevated prolactin of 1109 mIU/L. Genetic analysis confirmed a homozygous known disease-causing mutation PTS NM_000317.1:c.259C>T; NP_000308.1: p.P87S in the proband. In our local experience, while the estimated prevalence of hyperphenylalaninemia due to PTPS deficiency was reported to be 1 in 29,542 live births, not a single case of phenylalanine hydroxylase deficiency has been reported. Furthermore, there is a general lack of awareness of inherited metabolic diseases in the community as well as among the medical professionals. Very often, a low index of clinical suspicion will lead to delay in diagnosis, multiple unnecessary and costly investigations, prolonged morbidity and anxiety to the family affected. We strongly recommend that expanded newborn screening for hyperphenylalaninemia should be implemented for every baby born in the Hong Kong Special Administrative Region, China.

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A new cerebral hemorrhage model in cynomolgus macaques created by injection of autologous anticoagulated blood into the brain.

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The aim of this study was to establish and validate a clinically relevant model of intracerebral hemorrhage (ICH) via injection of autologous blood into the brains of cynomolgus macaques (Macaca fascicularis). Eight male cynomolgus macaques received 1.5 mL of fresh anticoagulated autologous femoral artery blood into the inner side of the claustrum near the right basal ganglia under stereotactic guidance. Animals were evaluated with MRI and positron emission tomography (PET) scanning before and 24 hours after surgery and once per week thereafter. A neurological deficit scale was used to assess the animals on days 1, 2, 3, 7, 14, 21, and 28 after surgery. Animals showed focal neurological signs corresponding to the MRI-located hematoma. The behavioral impairment progressively ameliorated over time, but never fully resolved. The hematoma was absorbed over time but was still present 4 weeks after surgery, with persistent metabolic deficit detected using PET scanning. Histological examinations confirmed the in vivo findings. This ICH model in a non-human primate mimics human ICH in the basal ganglia and may be useful for assessing the safety and efficacy of neuroprotective agents.
Clinical Implications of MR Imaging Findings in the White Matter in Very Preterm Infants: A 2-year Follow-up Study.


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Purpose: To explore the association between diffuse excessive high signal intensity (DEHSI), punctate white matter (WM) lesions, and ventricular dilatation around term-equivalent age (TEA) and at clinical follow-up at 2 years in very preterm infants and the effect on neurodevelopment.

Materials and Methods: Ethical approval for this prospective study was given by the institutional review board, and informed parental consent was obtained. An unselected cohort of 110 preterm infants (gestational age, < 32 weeks) was imaged around or after TEA. Clinical follow-up was performed at a corrected age of 2 years and consisted of a neurologic examination and a mental and developmental assessment (Bayley Scales of Infant Development). Univariate analyses and logistic and linear regression were performed to examine the relationships between variables.

Results: DEHSI was found in 58 of 65 (89%) infants imaged around TEA. DEHSI was never detected in infants imaged after postmenstrual age of 50 weeks and showed no association with neurodevelopmental outcome. Punctate WM lesions and ventricular dilatation were significantly associated with mental (P = .02 for punctate WM lesions) and psychomotor developmental delay (P < .001 and P = .03, respectively), motor delay (P = .002 and P = .02, respectively), and cerebral palsy (P = .01 and P = .03, respectively). Conclusion: Because of its high incidence in preterm infants around TEA, its absence after a postmenstrual age of 50 weeks, and its association with normal neurologic outcome at a corrected age of 2 years, DEHSI should not be considered part of the spectrum of WM injury, but rather a prematurity-related developmental phenomenon. © RSNA, 2011.

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