

Monday 1 November 2010

This free weekly bulletin lists the latest research on cerebral palsy (CP), as indexed in the NCBI, PubMed (Medline) and Entrez (GenBank) databases. These articles were identified by a search using the key term "cerebral palsy".

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## Interventions

### 1. *Am J Phys Med Rehabil.* 2010 Oct 21. [Epub ahead of print]

#### **Evaluating the Effect of Intensive Intervention in Children with Cerebral Palsy Using a Hypothetical Matched Control Group: A Preliminary Study.**

Yabunaka Y, Kondo I, Sonoda S, Saitoh E, Tsuruta Y, Konaka M, Konaka T, Kawarada S.

From the Department of Rehabilitation Science (YY), Osaka Health Science University, Osaka, Japan; Department of Rehabilitation (IK), Fujita Memorial Nanakuri Institute and Department of Rehabilitation Medicine (SS), Nanakuri Sanatorium, Fujita Health University, Mie, Japan; Department of Rehabilitation Medicine (ES), Fujita Health University, Aichi, Japan; Division of Rehabilitation (YT, MK, TK), Osaka Developmental Rehabilitation Center, Osaka, Japan; and Division of Rehabilitation (SK), Aomori Prefectural Asunaro Rehabilitation Center for Children, Aomori, Japan.

Yabunaka Y, Kondo I, Sonoda S, Saitoh E, Tsuruta Y, Konaka M, Konaka T, Kawarada S: Evaluating the effect of intensive intervention in children with cerebral palsy using a hypothetical matched control group: A preliminary study.

**OBJECTIVE:** To evaluate the effect of intensive intervention in children with cerebral palsy using a hypothetical matched control group based on motor growth curves. **DESIGN:** For pretest-posttest design using a hypothetical control group, a convenient sample of 39 children with cerebral palsy who received intensive intervention without surgical treatment was assigned to the experimental group. The hypothetical matched control group was created based on motor growth curves. Gains in Gross Motor Function Measure-66 score after intensive treatment in the experimental group were compared with those in the hypothetical matched control group using a mixed design for repeated-measures two-way analysis of variance. **RESULTS:** Gross motor function development in the experimental group was significantly accelerated compared with the hypothetical matched control group. **CONCLUSIONS:** In this preliminary study, using a hypothetical control group, the effectiveness of intensive intervention in children with cerebral palsy has been demonstrated. Although it is desirable to have a baseline phase to make sure whether gross motor function in the hypothetical control group changes in a similar way to that in the experimental group before an intervention phase, the hypothetical control group design is well worth considering as a research design option in the field of cerebral palsy research.

PMID: 20975523 [PubMed - as supplied by publisher]

## 2. Disabil Rehabil Assist Technol. 2010 Oct 27. [Epub ahead of print]

### The influence of the flexibility of the chair seat on pressure peak and distribution of the contact area in individuals with cerebral palsy during the execution of a task.

Braccialli LM, Sankako AN, Braccialli AC, Oliveira FT, Lucareli PR.

Unesp - Univ Estadual Paulista, Marilia, Brazil.

This study aimed to determine the influence of flexibility of the chair seat surface on the pressure peak and on the contact area during the execution of a task of handling an object on the seated position by individuals with spastic cerebral palsy. Ten individuals of both genders with diagnosis of spastic cerebral palsy, who had some control to voluntarily move the body and the upper limbs, participated in this study. Quantification of data was carried out in two experimental situations: (1) execution of a task of fitting with upper limbs, and with the individual placed on an adapted canvas seat; (2) execution of a task of fitting with the participant positioned on an adapted wooden seat. Data obtained were submitted to a non-parametric and descriptive statistical analysis using the Wilcoxon test. Results indicated that the use of canvas seat increased the contact area and decreased the pressure peak and the medio-lateral displacement of centre pressure on the seated posture.

PMID: 20977410 [PubMed - as supplied by publisher]

## 3. Res Dev Disabil. 2010 Oct 26. [Epub ahead of print]

### The determinants of daily function in children with cerebral palsy.

Tseng MH, Chen KL, Shieh JY, Lu L, Huang CY.

School of Occupational Therapy, College of Medicine, National Taiwan University, Taipei, Taiwan; Department of Physical Medicine and Rehabilitation, National Taiwan University Hospital, Taipei, Taiwan.

The aim of this study was to identify determinants of daily function in a population-based sample of children with cerebral palsy (CP). The study took into consideration factors from the entire scope of the International Classification of Functioning, Disability, and Health (ICF). Furthermore, the determinants of daily function were examined from the perspectives of capacity and performance respectively. A total of 216 children with CP (mean age 8.19 years, SD 3.39 years) and their caregivers participated in the study. The potential determinants of daily function from the dimensions of health condition, body functions and structures, environmental and personal factors of the ICF were collected. Stepwise multiple regression models showed that child's age, grade, preferred hand, educational placement, severity of gross and fine motor impairment, and prosocial behavior were important determinants, accounting for 88.29% of the variance of daily capacity. The aforementioned variables together with birth order were determinants of performance of daily function, and accounted for 89.53% of the variance in that performance. Knowledge of determinants of daily function helps clinicians and educators to plan intervention and educational programs targeted at these determinants to improve capacity and performance in daily function for children with CP.

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PMID: 21030207 [PubMed - as supplied by publisher]

## 4. Chang Gung Med J. 2010 Sep-Oct;33(5):524-31.

### Relationships between Developmental Profiles and Ambulatory Ability in A Follow-up Study of Preschool Children with Spastic Quadriplegic Cerebral Palsy.

Chen CL, Chen CY, Lin KC, Chen KH, Wu CY, Lin CH, Liu WY, Hsu HC.

Department of Physical Medicine and Rehabilitation, Chang Gung Memorial Hospital at Taipei; Graduate Institute of Early Intervention, Chang Gung University, Taoyuan, Taiwan.

Background: To investigate the follow-up course of developmental profiles in preschool children with spastic quadri-

plegic (SQ) cerebral palsy (CP) who had varying ambulatory abilities. Methods: Forty-eight children with SQ CP between 1 and 5 years old were classified into 2 groups, the ambulatory and non-ambulatory groups, based on Gross Motor Function Classification System (GMFCS) levels during the initial assessment. The developmental profiles, consisting of development quotients (DQs) of 8 domains, were evaluated during the initial assessment and the final assessment one year later. The DQ change index (%) was calculated as  $100\% \times (\text{final DQ} - \text{initial DQ}) / \text{initial DQ}$ . Results: The DQs of all developmental domains in the non-ambulatory group were lower than those in the ambulatory group on both initial and final assessments ( $p < 0.01$ ). As indicated by the DQ change indices, most DQs in the ambulatory group decreased slightly, whereas those in the non-ambulatory group decreased considerably ( $p < 0.05$ ). Furthermore, fine motor function increased proportionally with age in the ambulatory group, but not in the non-ambulatory group. Conclusion: The DQs of the developmental profiles varied in preschool CP children with different ambulatory abilities. The course of developmental profiles in preschool children with SQ CP evolves with age and relates to the degree of ambulatory function. Knowledge of these developmental profiles may be helpful in understanding, predicting, and managing the developmental problems of these children.

PMID: 20979703 [PubMed - in process]

## 5. Gait Posture. 2010 Oct 21. [Epub ahead of print]

### Arm posture score and arm movement during walking: A comprehensive assessment in spastic hemiplegic cerebral palsy.

Riad J, Coleman S, Lundh D, Broström E.

Department of Orthopedics, Skaraborgs Hospital, 541 42 Skövde, Sweden; Department of Woman and Child Health, Karolinska Institutet, 171 76 Stockholm, Sweden.

Patients with hemiplegic cerebral palsy often have noticeably deviant arm posture and decreased arm movement. Here we develop a comprehensive assessment method for the upper extremity during walking. Arm posture score (APS), deviation of shoulder flexion/extension, shoulder abduction/adduction, elbow flexion/extension and wrist flexion/extension were calculated from three-dimensional gait analysis. The APS is the root mean square deviation from normal, similar to Baker's Gait Profile Score (GPS) [1]. The total range of motion (ROM) was defined as the difference between the maximum and minimum position in the gait cycle for each variable. The arm symmetry, arm posture index (API) was calculated by dividing the APS on the hemiplegic side by that on the non-involved side, and the range of motion index (ROMI) by dividing the ROM on the hemiplegic side by that on the non-involved side. Using the APS, two groups were defined. Group 1 had minor deviations, with an APS under 9.0 and a mean of 6.0 (95% CI 5.0-7.0). Group 2 had more pronounced deviations, with an APS over 9.0 and a mean of 13.1 (CI 10.8-15.5) ( $p=0.000$ ). Total ROM was 60.6 in group 1 and 46.2 in group 2 ( $p=0.031$ ). API was 0.89 in group 1 and 1.70 in group 2 ( $p<0.001$ ). ROMI was 1.15 in group 1 and 0.69 in group 2 ( $p=0.003$ ). APS describes the amount of deviation, ROM provides additional information on movement pattern and the indices the symmetry. These comprehensive objective and dynamic measurements of upper extremity abnormality can be useful in following natural progression, evaluating treatment and making prognoses in several categories of patients.

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PMID: 20971011 [PubMed - as supplied by publisher]

## 6. Gait Posture. 2010 Oct 22. [Epub ahead of print]

### A regressed phase analysis for coupled joint systems.

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Department of Biomedical Engineering, Rutgers University, Piscataway, NJ 08854, USA; Clinical Brain Disorders Branch, National Institute of Mental Health, Bethesda, MD 20892, USA.

This study aims to address shortcomings of the relative phase analysis, a widely used method for assessment of coupling among joints of the lower limb. Goniometric data from 15 individuals with spastic diplegic cerebral palsy were recorded from the hip and knee joints during ambulation on a flat surface, and from a single healthy individual

with no known motor impairment, over at least 10 gait cycles. The minimum relative phase (MRP) revealed substantial disparity in the timing and severity of the instance of maximum coupling, depending on which reference frame was selected: MRP(knee-hip) differed from MRP(hip-knee) by  $16.1 \pm 14\%$  of gait cycle and  $50.6 \pm 77\%$  difference in scale. Additionally, several relative phase portraits contained discontinuities which may contribute to error in phase feature extraction. These vagaries can be attributed to the predication of relative phase analysis on a transformation into the velocity-position phase plane, and the extraction of phase angle by the discontinuous arctangent operator. Here, an alternative phase analysis is proposed, wherein kinematic data is transformed into a profile of joint coupling across the entire gait cycle. By comparing joint velocities directly via a standard linear regression in the velocity-velocity phase plane, this regressed phase analysis provides several key advantages over relative phase analysis including continuity, commutativity between reference frames, and generalizability to many-joint systems.

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PMID: 20971643 [PubMed - as supplied by publisher]

### 7. J Acoust Soc Am. 2010 Oct;128(4):2305.

#### Measuring tendon velocities using vector tissue Doppler imaging.

Eranki A, Sikdar S, Stanley C, Prosser L, Bellini L, Bland D, Alter K, Damiano D.

Dept. of Elec. & Comput. Eng., George Mason Univ., Fairfax, VA 22031, aeranki@gmu.edu George Mason Univ., Fairfax, VA 22031 NIH, Bethesda, MD 20892.

We have developed a vector tissue Doppler imaging (vTDI) method to quantify the magnitude and direction of tissue motion. The goal of this study was to quantify the repeatability of vTDI in measuring the contraction velocity of the tibialis anterior (TA) tendon in patients with cerebral palsy and foot drop (impaired dorsiflexion). vTDI was implemented on Ultrasonix Sonix Touch ultrasound system with a 5-14-MHz linear array transducer. The array was electronically split into two transmit and two receive apertures to estimate velocity vectors. Transmit and receive beams were steered by  $\pm 15$  deg. We conducted 42 trials on 7 subjects. Our preliminary results show that TA tendon velocities measured using vTDI have a strong linear correlation with the joint angular velocity estimated using a conventional 3-D motion capture system. We observed a peak velocity of  $5.20 \pm 1.58$  cm/s during dorsiflexion and  $8.45 \pm 2.06$  cm/s during the gravity-aided passive relaxation phase. The  $R^2$  values for all 42 trials were  $0.77 \pm 0.10$ . A second velocity measurement was made on three subjects after an interval of 4 weeks. We obtained repeatable velocity estimates with the standard deviation of the radius of action less than 0.13 cm. This demonstrates that vTDI is a feasible and reproducible method for measuring tendon velocities.

PMID: 20968591 [PubMed - in process]

### 8. J Nutr. 2010 Oct 27. [Epub ahead of print]

#### A DXA-Based Mathematical Model Predicts Midthigh Muscle Mass from Magnetic Resonance Imaging in Typically Developing Children but Not in Those with Quadriplegic Cerebral Palsy.

Modlesky CM, Cavaiola ML, Smith JJ, Rowe DA, Johnson DL, Miller F.

Department of Kinesiology and Applied Physiology, University of Delaware, Newark, DE 19716.

Valid methods for assessing regional muscle mass in children are needed. The aim of this study was to determine whether dual-energy X-ray absorptiometry (DXA) can accurately estimate midthigh muscle mass from MRI (muscle(MRI)) in typically developing children and children with quadriplegic cerebral palsy (CP). A mathematical model predicting muscle(MRI) from midthigh, fat-free soft tissue mass from DXA (FFST(DXA)) was developed using 48 typically developing children (6-13 y) and was validated using the leave-one-out method. The model was also tested in children with quadriplegic CP ( $n = 10$ ). The model produced valid estimates of midthigh muscle mass (muscle(DXA)) in typically developing children, as indicated by a very strong relationship between muscle(DXA) and muscle(MRI) ( $r^2 = 0.95$ ;  $SEE = 68$  g;  $P < 0.001$ ), no difference in muscle(DXA) and muscle(MRI) ( $P = 0.951$ ), and visual examination using a Bland-Altman plot. Muscle(DXA) was very strongly related to muscle(MRI) in chil-

dren with CP ( $r(2) = 0.96$ ;  $SEE = 54$  g;  $P < 0.001$ ); however, muscle(DXA) overestimated muscle(MRI) by 15% ( $P = 0.006$ ). The overestimation of muscle(MRI) by muscle(DXA) was strongly related to the lower ratio of (muscle(MRI)/FFST(DXA)) in children with CP ( $r(2) = 0.75$ ;  $P = 0.001$ ). The findings suggest that the DXA-based mathematical model developed in the current study can accurately estimate midhigh muscle mass in typically developing children. However, a population-specific model that takes into account the lower ratio of (muscle(MRI)/FFST(DXA)) is needed to estimate midhigh muscle mass in children with quadriplegic CP.

PMID: 20980659 [PubMed - as supplied by publisher]

### 9. Coll Antropol. 2010 Sep;34(3):981-8.

#### Intensive rehabilitation in children with cerebral palsy: our view on the neuronal group selection theory.

Polovina S, Polovina TS, Polovina A, Polovina-Proloscić T.

"Prof. Milena Stojcević-Polovina" Polyclinic for Physical Medicine and Rehabilitation, Zagreb, Croatia. poliklinika@zg.t-com.hr

Cerebral palsy (CP) is one of the major forms of developmental disorders. There are different approaches and controversies in rehabilitation treatment. The Neuronal Group Selection theory could provide theoretical explanation for Stojcević Polovina rehabilitation method. The aim of the study was to evaluate long-term impact of intensive and continuously performed rehabilitation on the motor autonomy level children with CR Motor autonomy levels, defined according to the Gross Motor Function Classification System (GMFCS) and Gross Motor Function Measure (GMFM), were analyzed in 24 children with CP at the beginning of the study and at the last visit. During rehabilitation, GMFM scores increased above the expected value of initial GMFCS level in the majority of patients. Intensive rehabilitation had significant influence on motor improvement in children with CP.

PMID: 20977092 [PubMed - in process]

### 10. J Neuroeng Rehabil. 2010 Oct 27;7(1):54. [Epub ahead of print]

#### Oromotor variability in children with mild spastic cerebral palsy: a kinematic study of speech motor control.

Chen CL, Chen HC, Hong WH, Yang FP, Yang LY, Wu CY.

**BACKGROUND:** Treating motor speech dysfunction in children with CP requires an understanding of the mechanism underlying speech motor control. However, there is a lack of literature in quantitative measures of motor control, which may potentially characterize the nature of the speech impairments in these children. This study investigated speech motor control in children with cerebral palsy (CP) using kinematic analysis. **METHODS:** We collected 10 children with mild spastic CP, aged 4.8 to 7.5 years, and 10 age-matched children with typical development (TD) from rehabilitation department at a tertiary hospital. All children underwent analysis of percentage of consonants correct (PCC) and kinematic analysis of speech tasks: poly-syllable (PS) and mono-syllable (MS) tasks using the Vicon Motion 370 system integrated with a digital camcorder. Kinematic parameters included spatiotemporal indexes (STIs), and average values and coefficients of variation (CVs) of utterance duration, peak oral opening displacement and velocity. An ANOVA was conducted to determine whether PCC and kinematic data significantly differed between groups. **RESULTS:** CP group had relatively lower PCCs (80.0-99.0%) than TD group ( $p = 0.039$ ). CP group had higher STIs in PS speech tasks, but not in MS tasks, than TD group did ( $p = 0.001$ ). The CVs of utterance duration for MS and PS tasks of children with CP were at least three times as large as those of TD children ( $p < 0.01$ ). However, average values of utterance duration, peak oral opening displacement and velocity and CVs of other kinematic data for both tasks did not significantly differ between two groups. **CONCLUSION:** High STI values and high variability on utterance durations in children with CP reflect deficits in relative spatial and/or especially temporal control for speech in the CP participants compared to the TD participants. Children with mild spastic CP may have more difficulty in processing increased articulatory demands and resulted in greater oromotor variability than normal children. The kinematic data such as STIs can be used as indices for detection of speech motor control impairments in children with mild CP and assessment of the effectiveness in the treatment.

PMID: 20979638 [PubMed - as supplied by publisher]



**11. Dev Disabil Res Rev. 2010 00;16(3):239-247.****Treatment of epilepsy in children with developmental disabilities.**

Depositario-Cabacar DF, Zelleke TG.

Neurology and Pediatrics, George Washington University Medical Center, Children's National Medical Center, Washington, DC.

Children with developmental disabilities are at increased risk for epilepsy with a prevalence rate higher than the general population. Some of the more common developmental disorders in childhood and the features of epilepsy in these conditions are discussed. Specifically, autism, cerebral palsy, mental retardation, and attention deficit and hyperactivity disorder are reviewed. Ideal treatment for developmentally-disabled children with epilepsy entails maximal seizure control without any significant adverse effects from the anti-epileptic drugs and good quality of life. Antiepileptic drugs' cognitive and behavioral adverse effects tend to occur more frequently in these children. Careful selection of the appropriate medication and close monitoring for drug adverse effects is important. The specific adverse effects of the older and newer antiepileptic drugs are also reviewed. © 2010 Wiley-Liss, Inc. Dev Disabil Res Rev 2010;16:239-247.

PMID: 20981762 [PubMed - as supplied by publisher]

**12. Dev Neurorehabil. 2010;13(4):248-57.****Use of microswitch technology and a keyboard emulator to support literacy performance of persons with extensive neuro-motor disabilities.**

Lancioni G, O'Reilly M, Singh N, Green V, Chiapparino C, De Pace C, Alberti G, Stasolla F.

Department of Psychology, University of Bari, Via Quintino Sella 268, Bari 70124, Italy. g.lancioni@psico.uniba.it

**OBJECTIVE:** To assess the effectiveness and acceptability of microswitch technology and a keyboard emulator to enable three participants with extensive neuro-motor disabilities to write words. **METHOD:** In Study I, two participants triggered an automatic scanning keyboard and selected/wrote letters via a small sliding movement of their hand(s) activating a touch/pressure panel (microswitch). In Study II, a third participant used the sliding movement and panel and a vocalization response with a voice-detecting microswitch. The sliding movement allowed her to light up the keyboard and select the letters and the vocalization to perform the scanning. **RESULTS:** Participants showed a better performance (shorter writing time) or an equally effective but less tiring performance with the new microswitch technology and response(s). They also preferred using this technology, and social validation ratings favoured such technology over previous solutions. **CONCLUSION:** The aforementioned technology may be useful to enable persons with extensive neuro-motor disabilities to write successfully.

PMID: 20629591 [PubMed - indexed for MEDLINE]

**13. Int J Pediatr. 2010;2010:717518. Epub 2010 Sep 29.****Infantile scurvy: two case reports.**

Ghedira Besbes L, Haddad S, Ben Meriem C, Golli M, Najjar MF, Guediche MN.

Pediatric Department, Fattouma Bourguiba Hospital, Monastir 5000, Tunisia.

**Background.** Ascorbic acid (vitamin C) is necessary for the formation of collagen, reducing free radicals, and aiding in iron absorption. **SCURVY**, a disease of dietary ascorbic acid deficiency, is uncommon today. It still exists in high risk groups including economically disadvantaged populations with poor nutrition. The incidence of **SCURVY** in the pediatric population is very low. **Cases Report.** Here we report two cases of **SCURVY** revealed by subperiosteal hematoma in children with cerebral palsy and developmental delay. **Conclusion.** **SCURVY** is extremely rare in children. Musculoskeletal manifestations are prominent in pediatric **SCURVY**. Multiple subperiosteal hematomas are an important indicator for diagnosis.

PMID: 20976262 [PubMed - in process]PMCID: PMC2952912Free PMC Article

**14. Spec Care Dentist. 2010 Jul-Aug;30(4):140-5.**

**Clinical and microbiological evaluation of the use of toothpaste containing 1% chlorhexidine and the influence of motivation on oral hygiene in patients with motor deficiency.**

de Andrade Meyer AC, de Mello Tera T, da Rocha JC, Jardini MA.

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Patients with motor deficiency have variable difficulties with mechanical plaque control, and as a consequence, the incidence of dental caries and periodontal disease can be higher in these patients. The objective of this study was to evaluate the clinical and microbiological efficacy of a toothpaste containing 1% chlorhexidine, which was used by patients with motor deficiency for 14 days. The reduction in plaque and gingival index and the impact on salivary microorganisms was evaluated. We conclude that the motivation of caregivers to carry out oral hygiene for patients with mental and motor deficiency is of great importance and is effective in reducing the formation of plaque as long as it is continuously reinforced. The use of chlorhexidine-containing toothpaste significantly reduced the plaque index and microorganism count between days 0 and 14. A reduction was also observed in the group that used a dentifrice without the chlorhexidine, but this difference was not significant.

PMID: 20618779 [PubMed - indexed for MEDLINE]

**15. AJNR Am J Neuroradiol. 2010 Oct 28. [Epub ahead of print]**

**Imaging Evaluation of Intrathecal Baclofen Pump-Catheter Systems.**

Miracle AC, Fox MA, Ayyangar RN, Vyas A, Mukherji SK, Quint DJ.

Neuroradiology Division, Department of Radiology, and Department of Physical Medicine and Rehabilitation, University of Michigan Medical Center, Ann Arbor, Michigan.

**SUMMARY:** ITB pumps are widely used in the treatment of intractable spasticity for many clinical indications, including cerebral palsy and spinal cord injury. High-dose intrathecal administration places the patient at significant risk for withdrawal in the event of device malfunction, necessitating rapid and complete evaluation of the pump-catheter system. This article reviews the approach to imaging evaluation of ITB pump-catheter systems, with specific emphasis on radiography, fluoroscopy, CT, and nuclear scintigraphy.

PMID: 21030478 [PubMed - as supplied by publisher]

## Epidemiology / Aetiology / Diagnosis & Early Treatment

*Please note: This is not yet a comprehensive outline of cerebral palsy prevention literature. It is expected that more research will be included when the search terms are expanded to include key terms other than "cerebral palsy". It is a work-in-progress and it will be expanded in coming months.*

**16. Ann Neurol. 2010 Nov;68(5):768-9.**

**Interleukin-6-174 CC polymorphism is associated with clinical chorioamnionitis and cerebral palsy.**

Resch B, D Müller W.

Research Unit for Neonatal Infectious Diseases and Epidemiology, Division of Neonatology, Department of Pediatrics, Medical University of Graz, Austria.

PMID: 21031589 [PubMed - in process]

**17. Dis Model Mech. 2010 Nov-Dec;3(11-12):678-88.**

**Towards improved animal models of neonatal white matter injury associated with cerebral palsy.**

Silbereis JC, Huang EJ, Back SA, Rowitch DH.

Departments of Pediatrics and Neurosurgery, Eli and Edythe Broad Institute for Stem Cell Research and Regeneration Medicine and Howard Hughes Medical Institute, University of California San Francisco, 513 Parnassus Avenue, San Francisco, CA 94143, USA.

Newborn neurological injuries are the leading cause of intellectual and motor disabilities that are associated with cerebral palsy. Cerebral white matter injury is a common feature in hypoxic-ischemic encephalopathy (HIE), which affects full-term infants, and in periventricular leukomalacia (PVL), which affects preterm infants. This article discusses recent efforts to model neonatal white matter injury using mammalian systems. We emphasize that a comprehensive understanding of oligodendrocyte development and physiology is crucial for obtaining new insights into the pathobiology of HIE and PVL as well as for the generation of more sophisticated and faithful animal models.

PMID: 21030421 [PubMed - in process]

**18. J Med Genet. 2010 Oct 23. [Epub ahead of print]**

**Adaptor protein complex-4 (AP-4) deficiency causes a novel autosomal recessive cerebral palsy syndrome with microcephaly and intellectual disability.**

Moreno-De-Luca A, Helmers SL, Mao H, Burns TG, Melton AM, Schmidt KR, Fernhoff PM, Ledbetter DH, Martin CL.

1 Department of Human Genetics, Emory University School of Medicine, Atlanta, Georgia, USA.

**Background:** Cerebral palsy is a heterogeneous group of neurodevelopmental brain disorders resulting in motor and posture impairments often associated with cognitive, sensorial, and behavioural disturbances. Hypoxic-ischaemic injury, long considered the most frequent causative factor, accounts for fewer than 10% of cases, whereas a growing body of evidence suggests that diverse genetic abnormalities likely play a major role. **Methods and results:** This report describes an autosomal recessive form of spastic tetraplegic cerebral palsy with profound intellectual disability, microcephaly, epilepsy and white matter loss in a consanguineous family resulting from a homozygous deletion involving AP4E1, one of the four subunits of the adaptor protein complex-4 (AP-4), identified by chromosomal microarray analysis. **Conclusion** These findings, along with previous reports of human and mouse mutations in other members of the complex, indicate that disruption of any one of the four subunits of AP-4 causes dysfunction of the entire complex, leading to a distinct 'AP-4 deficiency syndrome'.

PMID: 20972249 [PubMed - as supplied by publisher]