

Monday 6 December 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. Arch Phys Med Rehabil. 2010 Dec;91(12):1891-6.

Influence of age on step activity patterns in children with cerebral palsy and typically developing children.

Stevens SL, Holbrook EA, Fuller DK, Morgan DW.

Department of Health and Human Performance, Middle Tennessee State University, Murfreesboro, TN.

Stevens SL, Holbrook EA, Fuller DK, Morgan DW. Influence of age on step activity patterns in children with cerebral palsy and typically developing children.

OBJECTIVE: To document the influence of age on step activity patterns in children with cerebral palsy (CP) and typically developing (TD) children. **DESIGN:** Cross-sectional. **SETTING:** All step activity data were collected in free-living environments. **PARTICIPANTS:** Children with CP (n=27; age, 4-18y; 22 boys, 5 girls; Gross Motor Function Classification System levels I and II) and 27 age- and sex-matched TD children were recruited through public advertisements and contacts with local clinicians. CP and TD participants were stratified into younger (<10y; n=14) and older (10-18y; n=13) age groups. **INTERVENTION:** Daily step activity was monitored using a step activity monitor that was individually programmed to account for the gait characteristics of each participant. Step activity data were collected in 1-minute epochs during waking hours on 3 weekdays and 1 weekend day. Stored data were analyzed to yield average values of daily step activity, percentage of inactive time (0 steps) over the entire day, and percentage of total daily active time spent in low step activity (1-15 steps/min), medium step activity (16-40 steps/min), and high step activity (>40 steps/min). **MAIN OUTCOME MEASURES:** Daily step activity, percentage of inactive time, and percentage of active time spent in low-, moderate-, and high-intensity step activity. **RESULTS:** A significant ($P<.05$) interaction was observed between age (younger, older) and condition (CP, TD) for daily step activity, percentage of inactive time, and percentage of active time spent in low- and high-intensity step activity. The main effect of age was significant for each physical activity measure except for relative high-intensity step activity, and the main effect of condition was significant for all physical activity measures. Follow-up analyses ($P<.025$) revealed that older children with CP took fewer daily steps and displayed higher relative levels of inactivity and low-intensity activity and lower relative levels of high-intensity activity compared with older TD children. Older children with CP also exhibited lower daily step activity, demonstrated higher relative levels of inactivity and low-intensity activity, and displayed lower relative levels of moderate-intensity activity compared with younger children with CP. **CONCLUSIONS:** Compared with younger children with CP and age- and sex-matched TD youth, older youth with CP generally displayed step activity patterns typified by lower levels of physical activity and a greater degree of inactivity. These findings highlight the need to provide multiple opportunities for adolescents with CP to engage in a variety of physical activities that are appropriate to their needs, abilities, and preferences and that can aid in maintaining functional mobility, health, and quality of life.

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PMID: 21112431 [PubMed - in process]

2. Arch Phys Med Rehabil. 2010 Dec;91(12):1952-6.

Youth with cerebral palsy with differing upper limb abilities: how do they access computers?

Davies TC, Chau T, Fehlings DL, Ameratunga S, Stott NS.

Department of Surgery, University of Auckland, Auckland, New Zealand.

Davies TC, Chau T, Fehlings DL, Ameratunga S, Stott NS. Youth with cerebral palsy with differing upper limb abilities: how do they access computers?

OBJECTIVE: To identify the current level of awareness of different computer access technologies and the choices made regarding mode of access by youth with cerebral palsy (CP) and their families. **DESIGN:** Survey. **SETTING:** Two tertiary-level rehabilitation centers in New Zealand and Canada. **PARTICIPANTS:** Youth (N=60) with CP, Manual Ability Classification Scale (MACS) levels I to V, age 13 to 25 years. **INTERVENTIONS:** Not applicable. **MAIN OUTCOME MEASURE:** Questionnaire.

RESULTS: Fifty (83%) of the 60 youth were aware of at least 1 available assistive technology (AT), such as touch screens and joysticks. However, only 34 youth (57%) were familiar with the accessibility options currently available in the most common operating systems. Thirty-three (94%) of 35 youth who were MACS I and II used a standard mouse and keyboard, while few chose to use assistive technology or accessibility options. In contrast, 10 (40%) of 25 youth who were MACS III to V used a variety of assistive technologies such as touch screens, joysticks, trackballs, and scanning technologies. This group also had the highest use of accessibility options, although only 15 (60%) of the 25 were aware of them. **CONCLUSION:** Most youth with CP were aware of, and used, assistive technologies to enhance their computer access but were less knowledgeable about accessibility options. Accessibility options allow users to modify their own computer interface and can thus enhance computer access for youth with CP. Clinicians should be knowledgeable enough to give informed advice in this area of computer access, thus ensuring that all youth with CP can benefit from both AT and accessibility options, as required.

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PMID: 21112440 [PubMed - in process]

3. Arch Phys Med Rehabil. 2010 Dec;91(12):1897-903.

Dynamic versus fixed equinus deformity in children with cerebral palsy: how does the triceps surae muscle work?

Svehlík M, Zwick EB, Steinwender G, Kraus T, Linhart WE.

Paediatric Orthopaedic Unit, Department of Paediatric Surgery, Medical University of Graz, Graz, Austria; Department of Children and Adult Orthopaedics and Traumatology, 2nd Faculty of Medicine, Charles University Prague, Prague, Czech Republic.

Švehlík M, Zwick EB, Steinwender G, Kraus T, Linhart WE. Dynamic versus fixed equinus deformity in children with cerebral palsy: how does the triceps surae muscle work?

OBJECTIVES: To detect outcome measures that could help differentiate between dynamic and fixed equinus (FEQ) deformities in children with cerebral palsy, and secondary, to describe the function of the gastrocnemius and soleus (SOL) muscles when either dynamic triceps surae tightness or FEQ contracture is present. **DESIGN:** A group-comparison study. **SETTING:** Gait analysis laboratory. **PARTICIPANTS:** Children (N=23; 31 limbs) with cerebral palsy; 12 limbs showed a fixed contracture (FEQ group) and 19 limbs showed dynamic tightness of the triceps muscle (dynamic equinus group). Healthy children (N=12) without a neurologic or orthopedic disorder served as the control group. **INTERVENTIONS:** Not applicable. **MAIN OUTCOME MEASURES:** Time-distance, kinematic and kinetic gait variables, muscle-tendon length, and velocity parameters. **RESULTS:** Maximal ankle dorsiflexion angles were decreased in both equinus groups compared with the control group. Ankle range of motion, maximal power generation of the plantar flexors, and its timing during the gait cycle were different among groups. The ankle slope parameter showed substantial differences among groups. Muscle-tendon length parameters for the SOL and the medial (MGAC) and lateral gastrocnemius muscles were abnormal in both equinus groups compared with the control group. Maximal muscle lengths of the MGAC and SOL were longer in the dynamic equinus than FEQ group.

Peak lengthening velocity of the triceps surae muscle was significantly slower for all triceps surae muscles in the FEQ group than in the dynamic equinus group and occurred in the early swing phase. **CONCLUSIONS:** The presented results indicate that peak lengthening velocity of the triceps surae muscle might be one of the discriminating factors between FEQ and dynamic equinus deformity in children with cerebral palsy. This could help clinical decision making for treatment of an equinus gait pattern.

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PMID: 21112432 [PubMed - in process]

4. Dev Med Child Neurol. 2011 Jan;53(1):68-73. doi: 10.1111/j.1469-8749.2010.03777.x.

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

Kim WH, Park EY.

Department of Physical Therapy, Ulsan College, Ulsan, South Korea. Department of Secondary Special Education, College of Education, Jeonju University, Jeonju, South Korea.

Aim: This study examined the causal relation between spasticity, weakness, gross motor function, and functional outcome (expressed as activity limitation) in children with cerebral palsy (CP) and tested models of functional outcome mediated by gross motor function. **Method:** Eighty-one children (50 males, 31 females) with CP were recruited for this cross-sectional study. Their mean age was 10 years 4 months (SD 1y 9mo). Strength was assessed using the Manual Muscle Test. Spasticity was assessed by the Modified Ashworth Scale. The Gross Motor Function Measure assessed gross motor function. The Functional Skills domain of the Pediatric Evaluation of Disability Inventory assessed functional outcome. Twenty-eight children (34.6%) had quadriplegia, 44 children (54.3%) had diplegia, and nine children (11.1%) had hemiplegia. Children were classified using the Gross Motor Function Classification System with 14 (17.3%) in level I, 9 (11.1%) in level II, 13 (16.0%) in level III, 5 (6.2%) in level IV, and 40 (49.4%) in level V. **Results:** The proposed path model showed good fit indices. The direct effects were significant between spasticity and gross motor function ($\beta=-0.339$), between strength and gross motor function ($\beta=0.447$), and between gross motor function and functional outcome ($\beta=0.708$). Spasticity had a significant negative indirect effect ($\beta=-0.240$) and strength had a significant positive indirect effect ($\beta=0.317$) on functional outcome through effects on gross motor function. **Interpretation:** Activity-based rather than impairment-based intervention is more important for reducing activity limitation in children with CP. The study established a base from which researchers can further develop a causal model between motor impairments and functional outcome.

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PMID: 21126242 [PubMed - in process]

5. Disabil Health J. 2010 Apr;3(2):86-92. Epub 2009 Nov 18.

Promoting physical activity in an adolescent and a young adult with physical disabilities.

Buffart LM, van den Berg-Emons RJ, van Mechelen W, van Meeteren J, van der Slot W, Stam HJ, Roebroek ME.

Department of Rehabilitation Medicine, Erasmus MC, University Medical Centre, 3000 CA Rotterdam, the Netherlands.

BACKGROUND: We sought to describe the design of the Active Lifestyle and Sports Participation (ALSP) intervention for adolescents and young adults with physical disabilities, and to present the first 2 cases. **METHODS:** A 17-year-old boy with myelomeningocele and hydrocephalus and a 23-year-old woman with unilateral cerebral palsy were enrolled into the ALSP intervention, a personalized intervention designed to improve physical activity and fitness levels. Main outcome measures were self-reported physical activity and aerobic fitness. Fitness was determined by submaximal 6-minute walk or wheel test and by maximal cycle or arm ergometer-exercise test. Participants rated satisfaction with the intervention on a Likert-type numeric scale from 1 to 10. **RESULTS:** Improvements in self-reported physical activity were 51% and 75% for the male and female participant, respectively. Respective

improvements in submaximal exercise were 16% and 9%. Maximal exercise increased 39% in the male participant but did not increase in the female participant. Satisfaction with the intervention was rated moderate-good to excellent. CONCLUSION: Data for the first 2 cases suggested that ALSP intervention seemed feasible to offer in an outpatient rehabilitation department, and the effectiveness may be promising. Future studies should determine the short- and long-term effectiveness of the intervention.

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PMID: 21122773 [PubMed - in process]

6. Disabil Rehabil. 2010 Nov 26. [Epub ahead of print]

A hand rehabilitation system with force feedback for children with cerebral palsy: two case studies.

Choi KS, Lo KH.

School of Nursing, The Hong Kong Polytechnic University, Hung Hom, Kowloon, Hong Kong, China.

Purpose. To investigate whether the provision of force feedback as guidance in a computer-assisted training environment was feasible for improving handwriting ability of children with cerebral palsy. **Method.** Two children at the age of 7, diagnosed with dystonia and dyskinesia, respectively, took part in a 2-week pre-post test study. A desktop computer system equipped with a haptic device was developed to provide visual and haptic cues for practising Chinese handwriting. The system was used by the subjects two times a week. Average writing time and path length of 10 test characters and the trajectory of the pen tip were used to evaluate their performance. Paper-based test on handwriting legibility before and after the intervention were also conducted and video taped for subjective comparison. **Results.** The subjects were able to reduce the writing time through repeated practice. Path length also appeared to decrease, suggesting improvement in fine motor control ability and handwriting accuracy. One subject showed slight progress in legibility, while both of them developed a better sense of the proper ways of handwriting. **Conclusion.** Provision of haptic feedback in a virtual environment appears to be a feasible approach to improve cerebral palsy children's handwriting skills.

PMID: 21110729 [PubMed - as supplied by publisher]

7. Disabil Rehabil Assist Technol. 2010 Nov 26. [Epub ahead of print]

Responsiveness of the Seated Postural Control Measure and the Level of Sitting Scale in children with neuromotor disorders.

Field DA, Roxborough LA.

Therapy Department, Sunny Hill Health Centre for Children, Vancouver, British Columbia, Canada.

Purpose. Responsiveness of the Seated Postural Control Measure (SPCM) and the Level of Sitting Scale (LSS) was explored for children with neuromotor disorders. Total change scores for alignment (SPCM-A), function (SPCM-F) and sitting ability (LSS) were compared with a criterion change measure, the Global Change Scale (GCS). The a priori hypotheses predicted moderate correlations ($r > 0.40$). **Method.** Both SPCM and LSS were administered twice, 6 months apart. Parents and two therapists rated changes in alignment and function, and indicated importance of those changes on the GCS. Participants ($n = 114$) were divided into two groups: those whose posture was expected to change, (with a range of diagnoses) and those who were expected to remain stable (with a diagnosis of cerebral palsy). Ages ranged from 1 to 18 years. **Results.** Fair-to-moderate significant correlations ($p \leq 0.01$) were found between SPCM-F and LSS change scores and parents' and therapists' rating of change and importance of change on the GCS. Correlations for SPCM-A change scores were insignificant. The standardised response mean values for SPCM-F and LSS confirmed a minimal clinically important difference. **Conclusions.** SPCM-F shows promise as a responsive outcome measure, however; SPCM-A requires further work. LSS may be useful for evaluative purposes, in addition to its role as a classification index.

PMID: 21110727 [PubMed - as supplied by publisher]

8. Gait Posture. 2010 Nov 29. [Epub ahead of print]**Tibialis anterior tendon shortening in combination with Achilles tendon lengthening in spastic equinus in cerebral palsy.**

Rutz E, Baker R, Tirosh O, Romkes J, Haase C, Brunner R.

Pediatric Orthopaedic Department, University Children's Hospital Basle UKBB, Switzerland; Murdoch Childrens Research Institute, The Royal Children's Hospital, Victoria, Australia; Laboratory for Movement Analysis, University Children's Hospital Basle UKBB, Switzerland.

Equinus is the commonest deformity in cerebral palsy (CP). Many different surgical procedures have been described for the treatment of spastic equinus. In long standing equinus deformities the tibialis anterior muscle becomes elongated which is one reason for muscle weakness. Surgical tendon shortening of the tibialis anterior tendon was therefore introduced to rebalance muscle strength. All patients with CP who had a tibialis anterior tendon shortening (TATS) in combination with a tendo Achilles lengthening (TAL) were included in this study. A total of 29 patients had 30 surgical interventions (21 hemiplegic patients: 14 boys/7 girls, age 9-22 years; mean 15.2 years; 5 diplegics and 3 quadriplegics; 5 boys/3 girls, age 7-37.5 years; mean 14.8 years). Fifteen patients had additional surgery (soft tissue or bony procedures). The TATS was performed at the distal insertion with transosseous tendon fixation in the medial cuneiform bone at the original place. Movement Analysis Profile (MAP) for ankle dorsi-/plantarflexion, Gait Profile Score (GPS), Gait Deviation Index (GDI), and Gillette Gait Index (GGI) improved significantly for all patients compared pre- to postoperatively. In 93% of the patients active dorsiflexion of the ankle was possible postoperatively. We conclude that TATS in combination with TAL in spastic equinus in CP is a safe procedure and improves but not completely corrects foot positioning during gait. For the treatment of spastic equinus in CP we recommend shortening of the elongated antagonist (TATS) in combination with lengthening of the short agonist (TAL) for achieving optimal postoperative function.

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PMID: 21123070 [PubMed - as supplied by publisher]

9. J Neurosurg Spine. 2010 Dec;13(6):672-85.**Surgical management of spinal deformities in cerebral palsy.**

Jones-Quaidoo SM, Yang S, Arlet V.

Department of Orthopaedic Surgery, University of Virginia, Charlottesville, Virginia.

Cerebral palsy (CP) spinal deformities encompass a spectrum of deformities that are often initially treated nonoperatively, only to result in progression of scoliotic curves and further morbidity. Various surgical interventions have been devised to address the progressive curvature of the spine. This endeavor cannot be taken lightly and at times can be encumbered by prior treatments such as the use of baclofen pumps or dorsal rhizotomies. Care of these patients requires a multidisciplinary approach and comprehensive preoperative and postoperative management, including nutritional status, orthopedic assessment of functional level with specific emphasis on the hips and pelvic obliquity, and wheelchair modifications. The surgical techniques in CP scoliosis have progressively evolved from the classic Luque-Galveston fixation methods, the use of unit rods, and lately the use of pedicle screws, to modern sacropelvic fixation. With the latter method, the spinal deformity in patients with CP can usually be almost completely corrected.

PMID: 21121743 [PubMed - in process]

10. Clin Rehabil. 2010 Dec;24(12):1144-7.**Letter to the editor: Botulinum type A treatment: no evidence of increased risk of seizures in juvenile cerebral palsy.**

Brin MF, Pogoda JM, Boodhoo T, Bowen B, Albavera-Hernández C, Idrovo AJ.

Global Drug Development and Chief Scientific Officer, BOTOX, Allergan, LLC; Professor of Neurology, University of California, Irvine, CA, USA. brin_mitchell@allergan.com.

PMID: 21115685 [PubMed - in process]

11. Disabil Health J. 2010 Jul;3(3):179-85. Epub 2009 Dec 10.

Quality of diabetes care for adults with developmental disabilities.

Shireman TI, Reichard A, Nazir N, Backes JM, Greiner KA.

Department of Preventive Medicine and Public Health, University of Kansas School of Medicine, Kansas City, KS 66160, USA.

BACKGROUND: Given that individuals with developmental disabilities have a history of difficulty accessing appropriate health care, possess numerous risk factors for diabetes, and frequently have unique needs within the health care setting, it is important to conduct surveillance research to determine the quality of their diabetes care. **OBJECTIVE/HYPOTHESIS:** We assessed the quality of diabetes care for adults with developmental disabilities enrolled in Kansas Medicaid. Developmental disability was defined in accordance with Kansas Medicaid program eligibility and included individuals with intellectual disability, cerebral palsy, autism, and/or seizure disorder. **METHODS:** We identified a retrospective cohort of persons with developmental disabilities who were also diabetic and continuously enrolled in Kansas Medicaid. We tracked their quality of care measures (Hb(A1c)/glucose testing, cholesterol testing, eye examinations, microalbuminuria screening, and primary care visits) across the subsequent 12 months. Quality care measures were evaluated in relation to basic demographic variables and comorbid hypertension using unconditional logistic regression. **RESULTS:** Among 5,960 adults with developmental disability, 666 had diabetes (11.2%). Annual testing rates were Hb(A1c)/glucose testing, 51.7%; cholesterol, 44.3%; eye examinations, 29.3%; and microalbuminuria, 18.5%. Nearly all (93.5%) had contact with a primary care provider during the period. Comorbid hypertension was associated with higher rates of Hb(A1c), cholesterol testing, and primary care visits. Dual eligibility was associated with lower Hb(A1c)/glucose testing and cholesterol testing rates but comparable rates for other measures. Caucasians were more likely to have had an eye examination but less likely to have had their microalbumin checked. **CONCLUSIONS:** Adults with developmental disabilities and diabetes who were enrolled in the Kansas Medicaid Program were screened at lower frequency than published national figures for key quality indicators of diabetes care. These results call for action to find approaches to improve their quality of care. Further work is needed to understand the barriers to appropriate care and incentives that will remedy these gaps. In addition, research is needed to determine the accuracy of diabetes identification, treatment, and monitoring of adults with developmental disabilities.

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PMID: 21122783 [PubMed - in process]

12. Disabil Rehabil. 2010 Dec 2. [Epub ahead of print]

Responding to the challenges of parenting a child with cerebral palsy: a focus group.

Whittingham K, Wee D, Sanders M, Boyd R.

Queensland Cerebral Palsy and Rehabilitation Research Centre, The School of Medicine Faculty of Health Sciences.

Purpose. To explore the unique parenting challenges that parents of children with cerebral palsy (CP) face and to explore the feasibility of a new parenting intervention, Stepping Stones Triple P (SSTP), for this population. **Method.** Focus groups were conducted with parents of children with CP (n=8) and health professionals with experience in working with families of children with CP (n=5). The discussion was transcribed verbatim and a descriptive thematic analysis was performed. **Results.** Parents stated that knowing whether a particular behaviour is the result of CP or a behavioural issue is challenging. Parents were also keen to promote communication, independence and socialisation in their child. In addition, parents also discussed the challenges of parenting under time pressure, with addi-

tional parenting tasks, under public scrutiny and with grief. Both parents and professionals found SSTP to be a feasible and appropriate intervention for parents of children with CP. Conclusions. The parents of children with CP in this study faced a range of parenting challenges that may be effectively targeted by a parenting intervention. In addition, parents and health professionals found SSTP content acceptable and feasible for use with parents of children with CP.

PMID: 21126162 [PubMed - as supplied by publisher]

13. Disabil Rehabil Assist Technol. 2010 Nov 26. [Epub ahead of print]

Using Lego robots to estimate cognitive ability in children who have severe physical disabilities.

Cook AM, Adams K, Volden J, Harbottle N, Harbottle C.

Faculty of Rehabilitation Medicine, University of Alberta, Edmonton, Alberta, Canada.

Purpose. To determine whether low-cost robots provide a means by which children with severe disabilities can demonstrate understanding of cognitive concepts. **Method.** Ten children, ages 4 to 10, diagnosed with cerebral palsy and related motor conditions, participated. Participants had widely variable motor, cognitive and receptive language skills, but all were non-speaking. A Lego Invention [1] 'roverbot' was used to carry out a range of functional tasks from single-switch replay of pre-stored movements to total control of the movement in two dimensions. The level of sophistication achieved on hierarchically arranged play tasks was used to estimate cognitive skills. **Results.** The 10 children performed at one of the six hierarchically arranged levels from 'no interaction' through 'simple cause and effect' to 'development and execution of a plan'. Teacher interviews revealed that children were interested in the robot, enjoyed interacting with it and demonstrated changes in behaviour and social and language skills following interaction. **Conclusions.** Children with severe physical disabilities can control a Lego robot to perform un-structured play tasks. In some cases, they were able to display more sophisticated cognitive skills through manipulating the robot than in traditional standardised tests. Success with the robot could be a proxy measure for children who have cognitive abilities but cannot demonstrate them in standard testing.

PMID: 21110730 [PubMed - as supplied by publisher]

14. Phys Occup Ther Pediatr. 2010 Dec 1. [Epub ahead of print]

Parent Experience of Implementing Effective Home Programs.

Novak I.

Cerebral Palsy Institute, Darlinghurst, Sydney, Australia.

The aim of this preliminary study was to describe parent views about implementing effective home programs to inform practice recommendations. Semi-structured interviews were conducted with 10 parents of children with cerebral palsy (2 fathers and 8 mothers) who had participated in a home program by using a partnership-based approach. Transcripts were analyzed using grounded theory to the level of open coding of categories. Parents believed practice of home program activities was a part of life, to maximize progress, gain guidance, and manage time. Partnership-based home programs provided benefits including support, realism, flexibility, motivation, generalisable activities, practice reminders, progress updates, and role clarification. Parents advised other parents to accept their child's disability, never refuse help, be honest, develop routines, and consider programs essential. Parents advised professionals that parents want support, interdisciplinary coordination, and prognostic information, without pressure to comply. The findings suggest that parents experienced benefits using partnership home programs. Parents used these programs to help parent their child. Provision of ongoing support to parents was vital for motivation.

PMID: 21121708 [PubMed - as supplied by publisher]

Epidemiology / Aetiology / Diagnosis & Early Treatment

15. *Disabil Health J.* 2009 Jan;2(1):45-8.

Prevalence of cerebral palsy: Autism and Developmental Disabilities Monitoring Network, three sites, United States, 2004.

Arneson CL, Durkin MS, Benedict RE, Kirby RS, Yeargin-Allsopp M, Van Naarden Braun K, Doernberg NS.

University of Wisconsin-Madison, WI 53705, USA.

BACKGROUND: Cerebral palsy (CP) is the most common cause of motor disability in children and an important public health issue in the United States. The Autism and Developmental Disabilities Monitoring (ADDM) Network is a multisite program funded by the Centers for Disease Control and Prevention to determine trends in the prevalence of children with developmental disabilities, including CP, in the United States. This report describes population-based estimates of CP prevalence among 8-year-old children in three sites in the United States. **METHODS:** The ADDM Network conducted surveillance of CP among 8-year-old children living in north central Alabama, metropolitan Atlanta, and south central Wisconsin in 2004 (N = 68,272). This multisite collaboration involved the retrospective collection, linking, and analysis of data from multiple service providers and the population census estimates. **RESULTS:** The average prevalence of CP in 2004 across the three sites was 3.3 per 1,000 (95% confidence interval, 2.9-3.8). The prevalence was significantly higher in boys than in girls overall (male/female ratio, 1.4:1). The most common subtype across all three sites was spastic CP, ranging from 85% in Georgia to 89% in Alabama and Wisconsin. **CONCLUSIONS:** Ongoing, systematic, population-based surveillance in different areas of the United States is needed to describe and monitor CP prevalence. In addition, enhancing the surveillance system to include information about functional abilities is needed to better understand the public health impact of CP and strategies for improving quality of life and participation in activities at home and in the community.

PMID: 21122742 [PubMed - in process]

16. *Acta Paediatr.* 2010 Dec 3. doi: 10.1111/j.1651-2227.2010.02109.x. [Epub ahead of print]

Transient idiopathic dystonia in infancy.

Calado R, Monteiro JP, Fonseca MJ.

Hospital do Espírito Santo, Évora -EPE Hospital Garcia de Orta, Almada Hospital Garcia de Orta, Almada.

Aim: Review of transient idiopathic dystonia cases in order to improve knowledge on this entity, in relation to frequency, characterization and evolution. **Methods:** Retrospective review and characterization of clinical cases seen in Pediatric Neurology Consultation, diagnosed with transient idiopathic dystonia, between February 2001 and June 2009, using clinical files complemented with photographic records and updated information through the physician. **Results:** 13 infants who were referred to the Pediatric Neurology Consultation over a period of eight years, for asymmetric tone, posture and movements of the upper limb with onset before six months, with spontaneous favorable evolution and disappearance without sequelae, although the reason for referral was, in most cases, the suspicion of a hemiplegic cerebral palsy. **Conclusion:** Transient changes of tone, posture and movement can be observed during the first months of life. Differential diagnosis is extensive and complex, based on a careful history and neurological examination. Distinction between neurological, neuromuscular and orthopedic pathology is difficult, particularly at the onset of clinical manifestations. The cases presented are similar to those previously reported by Willemse and Deonna, classified as transient idiopathic dystonia of childhood. Pathophysiology is unknown, some findings support a genetic susceptibility to functional imbalance in brain neurotransmitters and synaptogenesis.

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PMID: 21126264 [PubMed - as supplied by publisher]

17. BMC Pregnancy Childbirth. 2010 Dec 1;10(1):77. [Epub ahead of print]**Long Term follow up after intra-Uterine transfusionS; the LOTUS study.**

Verduin EP, Lindenburg IT, Smits-Wintjens VE, van Klink JM, Schonewille H, van Kamp IL, Oepkes D, Walther FJ, Kanhai HH, Doxiadis II, Lopriore E, Brand A.

BACKGROUND: The Leiden University Medical Center (LUMC) is the Dutch national referral centre for pregnancies complicated by haemolytic disease of the fetus and newborn (HDFN) caused by maternal alloimmunization. Yearly, 20-25 affected fetuses with severe anaemia are transfused with intra-uterine blood transfusions (IUT). Mothers of whom their fetus has undergone IUT for HDFN are considered high responders with regard to red blood cell (RBC) antibody formation. Most study groups report high perinatal survival, resulting in a shift in attention towards short- and long-term outcome in surviving children. **METHODS:** We set up a large long-term observational follow-up study (LOTUS study), in cooperation with the Sanquin Blood Supply Foundation and the LUMC departments of Obstetrics, Neonatology and ImmunoHematology & Bloodtransfusion. The first part of this study addresses several putative mechanisms associated with blood group alloimmunization in these mothers. The second part of this study determines the incidence of long-term neurodevelopment impairment (NDI) and associated risk factors in children treated with IUT. All women and their life offspring who have been treated with IUT for HDFN in the LUMC from 1987-2008 are invited to participate and after consent, blood or saliva samples are taken. RBC and HLA antigen profile and antibodies are determined by serologic or molecular techniques. Microchimerism populations are tested by real time polymerase chain reaction (RT PCR). All children are tested for their neurological, cognitive and psychosocial development using standardised tests and questionnaires. The primary outcome is neurodevelopmental impairment (NDI), a composite outcome defined as any of the following: cerebral palsy, cognitive or psychomotor development < 2 standard deviation, bilateral blindness and/or bilateral deafness.

DISCUSSION: The LOTUS study includes the largest cohort of IUT patients ever studied and is the first to investigate post-IUT long-term effects in both mother and child. The results may lead to a change in transfusion policy, in particular future avoidance of certain incompatibilities. Additionally the LOTUS study will provide clinicians and parents better insights in the long-term neurodevelopmental outcome in children with HDFN treated with IUTs, and may improve the quality of antenatal counselling and long-term guidance.

PMID: 21122095 [PubMed - as supplied by publisher]Free Article

18. Dev Med Child Neurol. 2010 Dec 1. doi: 10.1111/j.1469-8749.2010.03840.x. [Epub ahead of print]**Anisotropy of transcallosal motor fibres indicates functional impairment in children with periventricular leukomalacia.**

Koerte I, Pelavin P, Kirmess B, Fuchs T, Berweck S, Laubender RP, Borggraefe I, Schroeder S, Danek A, Rummeny C, Reiser M, Kubicki M, Shenton ME, Ertl-Wagner B, Heinen F.

Institute of Clinical Radiology, Ludwig-Maximilians-University, Munich, Germany, Psychiatry Neuroimaging Laboratory, Department of Psychiatry, Brigham and Women's Hospital, Harvard Medical School, Boston, MA, USA, Pediatric Neurology and Developmental Medicine, Dr von Hauners Children's Hospital, Ludwig-Maximilians-University, Munich, Germany, Institute of Medical Informatics, Biometry and Epidemiology, Ludwig-Maximilians-University Munich, Germany, Department of Neurology, Ludwig-Maximilians-University, Munich, Germany, Clinical Neuroscience Division, Laboratory of Neuroscience, Department of Psychiatry, VA Boston Healthcare System, Harvard Medical School, Boston, MA, USA.

Aim: In children with bilateral spastic cerebral palsy (CP), periventricular leukomalacia (PVL) is commonly identified on magnetic resonance imaging. We characterized this white matter condition by examining callosal microstructure, interhemispheric inhibitory competence (IIC), and mirror movements. **Method:** We examined seven children (age range 11y 9mo-17y 9mo, median age 15y 10mo, four females, three males) with bilateral spastic CP/PVL (Gross Motor Function Classification System level I or II, Manual Ability Classification System level I) and 12 age-matched controls (age range 11y 7mo-17y 1mo, median age 15y 6mo, seven females, five males). Fractional anisotropy of the transcallosal motor fibres (TCMF) and the corticospinal tract (CST) of both sides were calculated. The parameters of IIC (transcranial magnetic stimulation) and mirror movements were measured using a standardized clinical examination and a computer-based hand motor test. **Results:** Fractional anisotropy was lower in children with bilateral spastic CP/PVL regarding the TCMF, but not the left or right CST. Resting motor threshold was elevated in children with bilateral spastic CP/PVL whereas measures of IIC tended to be lower. Mirror movements were markedly

elevated in bilateral spastic CP/PVL. Interpretation: This study provides new information on different aspects of motor function in children with bilateral spastic CP/PVL. Decreased fractional anisotropy of TCMF is consistent with impairment of hand motor function in children with bilateral spastic CP/PVL. The previously overlooked microstructure of the TCMF may serve as a potential indicator for hand motor function in patients with bilateral spastic CP/PVL.

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19. *Disabil Health J.* 2008 Jul;1(3):157-62.

Does race influence age of diagnosis for children with developmental delay?

Mann JR, Crawford S, Wilson L, McDermott S.

University of South Carolina, School of Medicine, Department of Family and Preventive Medicine, Columbia, SC 29208, USA.

BACKGROUND: Early identification of developmental delay is important for ensuring that children receive the early intervention services they need. Racial disparities exist for a number of childhood conditions, but it is not known whether there are racial disparities in the age of diagnosis with developmental delay. **OBJECTIVE/HYPOTHESIS:** This study aimed to determine the mean age of diagnosis with developmental delay for children ensured by South Carolina Medicaid. We hypothesized that African American children would be diagnosed later than white children. **METHODS:** A retrospective cohort study design explored South Carolina Medicaid claim records to determine the age when 5358 children with developmental delay (DD) were first diagnosed and whether there were racial disparities in age of diagnosis. **RESULTS:** The mean age at diagnosis was 4.08 years for African American children and 4.27 years for white children. For children diagnosed with DD and mental retardation, the average age of first diagnosis was 2.6 years, and for children with DD plus cerebral palsy, the average age was 2.1 years. African American race was significantly associated with younger diagnosis with DD in a multivariable model, but the overall model explained little of the variation in age at diagnosis. **CONCLUSIONS:** There were no clinically significant racial differences in the mean age of diagnosis with developmental delay. However, in general the age of diagnosis was undesirably late for both groups. Additional efforts are needed to ensure that children with DD, living in South Carolina, are identified near the beginning of early intervention services.

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20. *Lancet Neurol.* 2010 Nov 25. [Epub ahead of print]

Clinical and molecular characterisation of hereditary dopamine transporter deficiency syndrome: an observational cohort and experimental study.

Kurian MA, Li Y, Zhen J, Meyer E, Hai N, Christen HJ, Hoffmann GF, Jardine P, von Moers A, Mordekar SR, O'Callaghan F, Wassmer E, Wraige E, Dietrich C, Lewis T, Hyland K, Heales SJ, Sanger T, Gissen P, Assmann BE, Reith ME, Maher ER.

Department of Medical and Molecular Genetics, University of Birmingham School of Medicine, Institute of Biomedical Research, and Centre for Rare Diseases and Personalised Medicine, University of Birmingham, Birmingham, UK; Department of Paediatric Neurology, Birmingham Children's Hospital, Birmingham, UK.

BACKGROUND: Dopamine transporter deficiency syndrome is the first identified parkinsonian disorder caused by genetic alterations of the dopamine transporter. We describe a cohort of children with mutations in the gene encoding the dopamine transporter (SLC6A3) with the aim to improve clinical and molecular characterisation, reduce diagnostic delay and misdiagnosis, and provide insights into the pathophysiological mechanisms. **METHODS:** 11 children with a biochemical profile suggestive of dopamine transporter deficiency syndrome were enrolled from seven paediatric neurology centres in the UK, Germany, and the USA from February, 2009, and studied until June, 2010. The syndrome was characterised by detailed clinical phenotyping, biochemical and neuroradiological studies, and SLC6A3 mutation analysis. Mutant constructs of human dopamine transporter were used for in-vitro functional

analysis of dopamine uptake and cocaine-analogue binding. FINDINGS: Children presented in infancy (median age 2.5 months, range 0.5-7) with either hyperkinesia (n=5), parkinsonism (n=4), or a mixed hyperkinetic and hypokinetic movement disorder (n=2). Seven children had been initially misdiagnosed with cerebral palsy. During childhood, patients developed severe parkinsonism-dystonia associated with an eye movement disorder and pyramidal tract features. All children had raised ratios of homovanillic acid to 5-hydroxyindoleacetic acid in cerebrospinal fluid, of range 5.0-13.2 (normal range 1.3-4.0). Homozygous or compound heterozygous SLC6A3 mutations were detected in all cases. Loss of function in all missense variants was recorded from in-vitro functional studies, and was supported by the findings of single photon emission CT DaTSCAN imaging in one patient, which showed complete loss of dopamine transporter activity in the basal nuclei. INTERPRETATION: Dopamine transporter deficiency syndrome is a newly recognised, autosomal recessive disorder related to impaired dopamine transporter function. Careful characterisation of patients with this disorder should provide novel insights into the complex role of dopamine homeostasis in human disease, and understanding of the pathophysiology could help to drive drug development. FUNDING: Birmingham Children's Hospital Research Foundation, Birth Defects Foundation Newlife, Action Medical Research, US National Institutes of Health, Wellchild, and the Wellcome Trust.

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Correlation between dental maturation and chronological age in patients with cerebral palsy, mental retardation, and Down syndrome.

Diz P, Limeres J, Salgado AF, Tomás I, Delgado LF, Vázquez E, Feijoo JF.

Stomatology Department, School of Medicine and Dentistry, Santiago de Compostela University, c/ Entrerriós s/n, 15782 Santiago de Compostela, Spain.

Determining a child's chronological age and stage of maturation is particularly important in fields such as paediatrics, orthopaedics, and orthodontics, as well as in forensic and anthropological studies. Some systemic conditions can cause abnormal physiological maturation, and skeletal maturation is usually more delayed than dental maturation. The aim of this study was to determine dental age in a group of patients with the most prevalent congenital or perinatally occurring physical and mental disabilities. The study group comprised 155 white Spanish children aged 3-17 years (35 with cerebral palsy, 83 with mental retardation and no associated syndromes or systemic conditions, and 37 with Down syndrome). The dental maturation indices described by Nolla and Demirjian were used to generate regression lines for the dental age of individuals in a control group (688 white Spanish children aged 3-17 years) and the formulae were then used to determine the dental age of patients in the study group. No significant differences were found between dental and chronological age in boys with cerebral palsy, mental retardation, or Down syndrome. In contrast, dental age (calculated from the linear regression model that included values for the Demirjian index) was significantly delayed compared with chronological age in girls with cerebral palsy or Down syndrome.

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