Cerebral Palsy INSTITUTE

CP Research News

Monday 3 May 2010

Interventions

1. BMC Pediatr. 2010 Apr 27;10(1):26. [Epub ahead of print]

Change in basic motor abilities, quality of movement and everyday activities following intensive, goal-directed, activity-focused physiotherapy in a group setting for children with cerebral palsy.

Sorsdahl AB, Moe-Nilssen R, Kaale HK, Rieber J, Strand LI.

BACKGROUND: The effects of intensive training for children with cerebral palsy (CP) remain uncertain. The aim of the study was to investigate the impact on motor function, quality of movements and everyday activities of three hours of goal-directed activity-focused physiotherapy in a group setting, five days a week for a period of three weeks. METHODS: A repeated measures design was applied with three baseline and two follow up assessments; immediately and three weeks after intervention. Twenty-two children with hemiplegia (n=7), diplegia (n=11), quadriplegia (n=2) and ataxia (n=2) participated, age ranging 3-9y. All levels of Gross Motor Function Classification System (GMFCS) and Manual Ability Classification System (MACS) were represented. Parents and professionals participated in goal setting and training. ANOVA was used to analyse change over repeated measures. RESULTS: A main effect of time was shown in the primary outcome measure; Gross Motor Function Measure-66 (GMFM-66), mean change being 4.5 ($p<0.01$) from last baseline to last follow up assessment. An interaction between time and GMFCS-levels was found, implying that children classified to GMFCS-levels I-II improved more than children classified to levels III-V. There were no main or interaction effects of age or anti-spastic medication. Change scores in the Pediatric Evaluation of Disability Inventory (PEDI) ranged 2.0-6.7, $p<0.01$ in the Self-care domain of the Functional Skills dimension, and the Self-care and Mobility domains of the Caregiver Assistance dimension. The children's individual goals were on average attained, Mean Goal Attainment Scaling (GAS) T-score being 51.3. Non-significant improved scores on the Gross Motor Performance Measure (GMPM) and the Quality of Upper Extremities Skills Test (QUEST) were demonstrated. Significant improvement in GMPM scores were found in improved items of the GMFM, not in items that maintained the same score. CONCLUSIONS: Basic motor abilities and self-care improved in young children with CP after goal-directed activity-focused physiotherapy with involvement of their local environment, and their need for caregiver assistance in self-care and mobility decreased. The individualized training within a group context during a limited period of time, was feasible and well-tolerated. The coherence between acquisition of basic motor abilities and quality of movement should be further examined.

PMID: 20423507 [PubMed - as supplied by publisher]

2. Phys Ther. 2010 Apr 29. [Epub ahead of print]

Trunk and Hip Muscle Activation Patterns Are Different During Walking in Young Children With and Without Cerebral Palsy.

Prosser LA, Lee SC, Vansant AF, Barbe MF, Lauer RT.

Rehabilitation Medicine Department, National Institutes of Health Clinical Center, Bldg 10-CRC, 1-1469, 10 Center

Dr, Bethesda MD 20892 (USA).

Background Poor control of postural muscles is a primary impairment in people with cerebral palsy (CP). Objective The purpose of this study was to investigate differences in the timing characteristics of trunk and hip muscle activity during walking in young children with CP compared with children with typical development (TD). METHODS:/b> Thirty-one children (16 with TD, 15 with CP) with an average of 28.5 months of walking experience participated in this observational study. Electromyographic data were collected from 16 trunk and hip muscles as participants walked at a self-selected pace. A custom-written computer program determined onset and offset of activity. Activation and coactivation data were analyzed for group differences. RESULTS: /b> The children with CP had greater total activation and coactivation for all muscles except the external oblique muscle and differences in the timing of activation for all muscles compared with the TD group. The implications of the observed muscle activation patterns are discussed in reference to existing postural control literature. Limitations The potential influence of recording activity from adjacent deep trunk muscles is discussed, as well as the influence of the use of an assistive device by some children with CP. CONCLUSIONS:/b> Young children with CP demonstrate excessive, nonreciprocal trunk and hip muscle activation during walking compared with children with TD. Future studies should investigate the efficacy of treatments to reduce excessive muscle activity and improve coordination of postural muscles in CP.

PMID: 20430948 [PubMed - as supplied by publisher]

3. Med Sci Monit. 2010 Apr 28;16(5):CR222-31.

Speech intelligibility in cerebral palsy children attending an art therapy program.

Wilk M, Pachalska M, Lipowska M, Herman-Sucharska I, Makarowski R, Mirski A, Jastrzebowska G.

Institute of Physiotherapy, Jan Kochanowski University, Kielce, Poland. magwilk@poczta.fm

BACKGROUND: Dysarthia is a common sequela of cerebral palsy (CP), directly affecting both the intelligibility of speech and the child's psycho-social adjustment. Speech therapy focused exclusively on the articulatory organs does not always help CP children to speak more intelligibly. The program of art therapy described here has proven to be helpful for these children. MATERIAL/METHODS: From among all the CP children enrolled in our art therapy program from 2005 to 2009, we selected a group of 14 boys and girls (average age 15.3) with severe dysarthria at baseline but no other language or cognitive disturbances. Our retrospective study was based on results from the Auditory Dysarthria Scale and neuropsychological tests for fluency, administered routinely over the 4 months of art therapy. RESULTS: All 14 children in the study group showed some degree of improvement after art therapy in all tested parameters. On the Auditory Dysarthia Scale, highly significant improvements were noted in overall intelligibility ($p<0.0001$), with significant improvement ($p<0.001$) in volume, tempo, and control of pauses. The least improvement was noted in the most purely motor parameters. All 14 children also exhibited significant improvement in fluency. CONCLUSIONS: Art therapy improves the intelligibility of speech in children with cerebral palsy, even when language functions are not as such the object of therapeutic intervention.

PMID: 20424549 [PubMed - in process]

4. Stereotact Funct Neurosurg. 2010 May 1;88(3):187-192. [Epub ahead of print]

Combined Ablative Neurosurgical Procedures in a Patient with Mixed Spastic and Dystonic Cerebral Palsy.

Sitthinamsuwan B, Chanvanitkulchai K, Nunta-Aree S, Kumthornthip W, Pisarnpong A, Ploypetch T.

Division of Neurosurgery, Department of Surgery, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand.

Background: Harmful generalized spasticity is an obstacle in rehabilitation and caregiving. Neurosurgical intervention is a therapeutic option for patients with severe spasticity who do not respond to nonoperative management. Currently, intrathecal baclofen therapy (ITB) is a good alternative treatment for such patients. However, the ITB device is costly and the intrathecal drug is not available in Thailand. Case Description: We report a combined use of ablative neurosurgical procedures in a patient with severe generalized spasticity and disabling cervical dystonia (CD). The assembled operations including selective peripheral denervation for CD, microsurgical dorsal root entry

zone lesion for upper limb spasticity, and selective dorsal rhizotomy for lower limb spasticity were conducted in a single session. Furthermore, recurrent spasticity of the upper extremities was subsequently treated by selective peripheral neurotomy. Results: The spasticity and CD totally disappeared after all operations. The patient became able to sit and perform head turning. Additionally, we also found an improvement in swallowing and the voluntary movement of the lower limbs.

PMID: 20431331 [PubMed - as supplied by publisher]

5. Gait Posture. 2010 Apr 27. [Epub ahead of print]

Repeatability of upper limb kinematics for children with and without cerebral palsy.

Reid S, Elliott C, Alderson J, Lloyd D, Elliott B.

School of Sport Science, Exercise & Health, The University of Western Australia, Perth, Australia.

BACKGROUND: There is increasing demand for a standardised and reliable protocol for the objective assessment of upper limb motion in clinical populations. This paper describes the repeatability of a three-dimensional (3D) kinematic model and protocol to assess upper limb movement for children with and without cerebral palsy (CP). METHODS: Ten typically developing (TD) children (m=10.5years+/-1.18) and seven children with CP (spastic hemiplegia) (m=11.14years+/-1.86) completed upper limb motion analysis on two occasions separated by at least one week. Participants performed three trials of four functional tasks, where 3D joint angles were calculated at the thorax, shoulder, elbow and wrist. Within and between-day repeatability was assessed using coefficients of multiple determination (CMD). FINDINGS: There were distinct kinematic patterns for both groups for each functional task. In relation to their peers, children with CP consistently displayed reduced elbow extension, and compensatory patterns at the shoulder and thorax. High within and between-day CMD scores were revealed for specific rotations, with the highest being obtained at joints with large ranges of motion. INTERPRETATION: The chosen tasks delineate the upper limb kinematic patterns of those with and without CP. The model has high within and between-day repeatability particularly where joint rotations demonstrate a large range of movement. 3D motion analysis is a feasible assessment tool for use with clinical populations.

PMID: 20430623 [PubMed - as supplied by publisher]

6. Neurorehabil Neural Repair. 2010 Apr 27. [Epub ahead of print]

Effectiveness of Modified Constraint-Induced Movement Therapy in Children With Unilateral Spastic Cerebral Palsy: A Randomized Controlled Trial.

Aarts PB, Jongerius PH, Geerdink YA, Limbeek JV, Geurts AC.

BACKGROUND: In children with unilateral spastic cerebral palsy (CP), there is only limited evidence for the effectiveness of modified constraint-induced movement therapy (mCIMT). OBJECTIVE: To investigate whether 6 weeks of mCIMT followed by 2 weeks of bimanual task-specific training (mCIMT-BiT) in children with unilateral spastic CP improves the spontaneous use of the affected limb in both qualitative and quantitative terms more than usual care (UC) of the same duration. METHODS: Children with unilateral spastic CP with Manual Ability Classification System (MACS) scores I, II, or III and aged 2.5 to 8 years were recruited and randomly allocated to either the mCIMT-BiT group (three 3-hour sessions per week: 6 weeks of mCIMT, followed by 2 weeks of task-specific training in goal-directed bimanual play and self-care activities) or to 1.5 hours of more general physical or occupational weekly plus encouragement to use the affected hand for the UC group. Primary outcome measures were the Assisting Hand Assessment and the ABILHAND-Kids. Secondary outcomes were the Melbourne Assessment of Unilateral Upper Limb Function, the Canadian Occupational Performance Measure, and the Goal Attainment Scale. RESULTS: Twenty-eight children were allocated to mCIMT-BiT and 24 to UC. Except for the Melbourne, all primary and secondary outcome measures demonstrated significant improvements in the mCIMT-BiT group. CONCLUSION: mCIMT followed by task-specific training of goal-directed bimanual play and self-care activities is an effective intervention to improve the spontaneous use of the more affected upper limb in children with relatively good baseline upper extremity function.

PMID: 20424191 [PubMed - as supplied by publisher]

7. Curr Psychiatry Rep. 2010 Apr;12(2):116-21.

Psychiatric complications in cerebral palsy.

Foster T, Rai AI, Weller RA, Dixon TA, Weller EB.

Department of Child and Adolescent Psychiatry, The Children's Hospital of Philadelphia, University of Pennsylvania, 3440 Market Street, Suite 200, Philadelphia, PA 19104, USA. dr.foster@alumni.upenn.edu

Cerebral palsy (CP) is a disorder of motor and posture impairment resulting from brain injury prior to completion of cerebral development. It affects 2 to 3 per 1000 individuals. CP is also associated with sensory, behavioral, cognitive, and emotional sequelae. Few systematic studies of psychiatric comorbidities in children and adolescents with CP have been conducted, as the main focus of concern has been on the physical disabilities. This has diverted attention from treatable psychiatric syndromes. Proper psychiatric evaluation of children with CP is an important task, as appropriate interventions can help them reach their full potential and enhance the quality of their lives and those of their families. We report the case of an individual with CP with behavioral and emotional symptoms to illustrate the diagnostic complexity involved. The case highlights the importance of engaging in a comprehensive diagnostic psychiatric evaluation process to assess and suggest treatment options for accompanying comorbid psychiatric conditions.

PMID: 20425296 [PubMed - in process]

8. Rev Neurol (Paris). 2010 Feb;166(2):142-8. Epub 2010 Feb 6.

Gait disorders: mechanisms and classification [Article in French]

Azulay JP, Vacherot F, Vaugoyeau M.

Service de neurologie et pathologie du mouvement, hôpital de la Timone, 13385 Marseille cedex 05, France. jean-philippe.azulay@ap-hm.fr

Gait disorders are important because of their prevalence, particularly among the elderly, and the associated risk of falls and injury. The main physiological characteristics of locomotion and neural networks that organize locomotion and maintain balance are briefly reviewed. A simplified classification designed clinical practice and based upon clinical characteristics and more recent data obtained from quantified gait analysis is proposed.

PMID: 20138639 [PubMed - indexed for MEDLINE]

9. Folia Phoniatr Logop. 2010;62(3):92-6. Epub 2010 Apr 29.

Studies of chinese speakers with dysarthria: informing theoretical models.

Whitehill TL.

University of Hong Kong, Hong Kong, SAR, China. tara@hku.hk

Most theoretical models of dysarthria have been developed based on research using individuals speaking English or other Indo-European languages. Studies of individuals with dysarthria speaking other languages can allow investigation into the universality of such models, and the interplay between language-specific and language-universal aspects of dysarthria. In this article, studies of Cantonese- and Mandarin-Chinese speakers with dysarthria are reviewed. The studies focused on 2 groups of speakers: those with cerebral palsy and those with Parkinson's disease. Key findings are compared with similar studies of English speakers. Since Chinese is tonal in nature, the impact of dysarthria on lexical tone has received considerable attention in the literature. The relationship between tone [which involves fundamental frequency (F(0)) control at the syllable level] and intonation (involving F(0) control at the sentential level) has received more recent attention. Many findings for Chinese speakers with dysarthria support earlier findings for English speakers, thus affirming the language-universal aspect of dysarthria. However, certain differences, which can be attributed to the distinct phonologies of Cantonese and Mandarin, highlight the lan-

guage-specific aspects of the condition. 2010 S. Karger AG, Basel.

PMID: 20424463 [PubMed - in process]

10. Indian J Orthop. 2010 Apr;44(2):177-83.

Use of vacuum assisted closure in instrumented spinal deformities for children with postoperative deep infections.

Canavese F, Krajbich JI.

Department of Orthopedics, Shriners Hospital for Children, 3101 SW Sam Jackson Park Road, Oregon 97239, Portland, USA.

BACKGROUND: Postoperative deep infections are relatively common in children with instrumented spinal deformities, whose healing potential is somewhat compromised. Children with underlying diagnosis of cerebral palsy, spina bifida and other chronic debilitating conditions are particularly susceptible. Vacuum-assisted closure (VAC) is a newer technique to promote healing of wounds resistant to treatment by established methods. This article aims to review the efficacy of the VAC system in the treatment of deep spinal infections following spinal instrumentation and fusion in children and adolescents. MATERIALS AND METHODS: We reviewed 33 patients with deep postoperative surgical site infection treated with wound VAC technique. We reviewed clinical and laboratory data, including the ability to retain the spinal hardware, loss of correction and recurrent infections. RESULTS: All patients successfully completed their wound VAC treatment regime. None had significant loss of correction and one had persistent infection requiring partial hardware removal. The laboratory indices normalized in all but three patients. CONCLUSIONS: Wound VAC technique is a useful tool in the armamentarium of the spinal surgeon dealing with patients susceptible to wound infections, especially those with neuromuscular diseases. It allows for retention of the instrumentation and maintenance of the spinal correction. It is reliable and easy to use.

PMID: 20419005 [PubMed - in process]PMCID: PMC2856393Free PMC Article

11. Brain Nerve. 2010 Feb;62(2):113-24.

Novel functional electrical stimulation for neurorehabilitation [Article in Japanese]

Hara Y.

Nippon Medical School, Chiba Hokusoh Hospital, Department of Rehabilitation Medicine, 1715 Kamagari, Inba-mura, Inba-gun, Chiba 270-1694, Japan.

Our understanding of motor learning, neuroplasticity, and functional recovery after the occurrence of brain lesions has increased considerably. New findings in basic neuroscience have provided an impetus for research in motor rehabilitation. Several prospective studies have shown that repeated motor practice and motor activity in a real world environment have a favorable effect on motor recovery in stroke patients. Electrical stimulation can be applied in a variety of ways to the hemiparetic upper extremity following a stroke. In particular, electromyography (EMG)-triggered electrical muscle stimulation improves the motor function of the hemiparetic arm and hand. Triggered electrical stimulation is reported to be more effective than non-triggered electrical stimulation in facilitating upper extremity motor recovery after stroke. Power-assisted functional electrical stimulation (FES) induces greater muscle contraction by electrical stimulation that is in proportion to voluntary integrated EMG signals. Daily power-assisted FES home-program therapy with novel equipment has been shown to effectively improve wrist, finger extension, and shoulder flexion. Combined modulation of voluntary movement, proprioceptive sensory feedback, and electrical stimulation might play an important role in improving impaired sensory-motor integration by power-assisted FES therapy. A multi-channel near-infrared spectroscopy (NIRS) studies in which the hemoglobin levels in the brain were non-invasively and dynamically measured during functional activity found that the cerebral blood flow in the injured sensory-motor cortex area is greater during a power-assisted FES session than during simple active movement or simple electrical stimulation. A novel power-assisted FES sleeve (Cyberhand) has been developed for the rehabilitation of hemiplegic upper extremities.

PMID: 20192031 [PubMed - indexed for MEDLINE]

12. Harefuah. 2010 Jan;149(1):14-7, 64, 63.

The knowledge base of Israeli pediatricians in the area of child development [Article in Hebrew]

Gabis L, Raz R.

Weinberg Child Development Center, Safra Children's Hospital, Sheba Medical Center, Tel Hashomer, Israel.

BACKGROUND: Pediatricians are required to identify and follow-up on children suspected of having developmental disorders, such as autism spectrum disorders, cerebral palsy, mental retardation or specific language impairments. This task requires basic up-to-date knowledge in the areas of child development and child neurology. The rest of the inquiry, including reaching a definite diagnosis and treatment plan, will usually be conducted in child development centers or other frameworks. METHODS: The current study examined the knowledge of Israeli pediatricians using an original questionnaire, on a sample of 86 pediatricians. RESULTS: There is high variance among Israeli pediatricians regarding the level of knowledge in the areas of child development and child neurology. This variance is not explained by different variables that were measured, such as place of work (clinic, hospital, "Tipat Halav" - "Well-Child Clinics"), seniority or use of screening tools. Most participants 164%] expressed a demand for professional enrichment in this area.

PMID: 20422834 [PubMed - in process]

13. Indian J Orthop. 2010 Apr;44(2):148-58.

Development and treatment of spinal deformity in patients with cerebral palsy.

I Tsirikos A.

Clinical Lead-Scottish National Spine Deformity Center, Royal Hospital for Sick Children, Sciennes Road, Edinburgh, EH9 1LF, UK.

Scoliosis is a common deformity in children and adolescents with cerebral palsy. This is usually associated with pelvic obliquity due to extension of the curve to the sacrum. Sagittal plane deformity is less common and often develops along with scoliosis. Spinal deformity in patients with severe neurological handicaps can affect their ability to sit and cause significant back pain or pain due to rib impingement against the elevated side of the pelvis on the concavity of the curvature. Surgical correction followed by spinal arthrodesis is indicated in patients with progressive deformities which interfere with their level of function and quality of life. Spinal deformity correction is a major task in children with multiple medical co-morbidities and can be associated with a high risk of complications including death. A well-coordinated multidisciplinary approach is required in the assessment and treatment of this group of patients with the aim to minimize the complication rate and secure a satisfactory surgical outcome. Good knowledge of the surgical and instrumentation techniques, as well as the principles of management is needed to achieve optimum correction of the deformity and balancing of the spine and pelvis. Spinal fusion has a well-documented positive impact even in children with quadriplegia or total body involvement and is the only surgical procedure which has such a high satisfaction rate among parents and caregivers.

PMID: 20419001 [PubMed - in process]PMCID: PMC2856389Free PMC Article

14. J Altern Complement Med. 2010 Apr;16(4):375-95.

Traditional Chinese Medicine for treatment of cerebral palsy in children: a systematic review of randomized clinical trials.

Zhang Y, Liu J, Wang J, He Q.

Guang'anmen Hospital, China Academy of Chinese Medical Science, Beijing, China.

OBJECTIVE: The objective of this study was to systematically evaluate the effects of Traditional Chinese Medicine (TCM) therapy including acupuncture, tu'ina, oral herbal medicine, herbal bathing, and collateral-channels conduct therapy for treating children with cerebral palsy (CP). METHODS: We included randomized controlled trials (RCTs)

on TCM for children with CP. We searched the China National Knowledge Infrastructure, Database for Chinese Technical Periodicals, Chinese Biomedical Literature Database, databases of Chinese biomedical journals/Chinese Medical Current Contents, Wan Fang Data, PubMed, MEDLINE, Embase, and the Cochrane Library until the end of July 2009, and searched the reference list of retrieved papers. Data were extracted by 1 author and checked for validation by another author, and data were analyzed using RevMan 4.3.2. Only one meta-analysis was performed due to the heterogeneity among the trials. RESULTS: Thirty-five (35) RCTs involving 3286 children with CP using TCM therapy and conventional therapy (CT) including physical, occupational, and speech therapy, hyperbaric oxygen, cranial nerves nutrition agents, or any combination of above were included. The methodological quality was generally low in terms of allocation concealment, blinding, and intention-to-treat analysis. Meta-analysis showed acupuncture combine with CT improved activities of daily living (mean difference: 6.38, 95% confidence interval 5.15-7.61; $p < 0.00001$, $n = 160$) compared with CT alone. Acupuncture plus tu'ina, or plus herbal medicine and CT showed significant beneficial effects on comprehensive function in terms of both physical and mental aspects, independence, and verbal function compared with CT alone. The combination of radix Astragali injection with CT showed significant benefit on gross motor function and social behavior adaptation comparing with CT. There are six trials reported adverse events that were not associated with acupuncture, tu'ina, and/or herbal medicine. CONCLUSIONS: Acupuncture with or without CT or other conventional therapy, tu'ina, herbal medicine, and collateral channels conduct treatment combined with CT may have benefit in children with CP. However, due to insufficient evidence, further rigorous trials are warranted.

PMID: 20423208 [PubMed - in process]

15. PM R. 2010 Apr;2(4):282-284.

Safety Considerations in the Use of Botulinum Toxins in Children With Cerebral Palsy.

Apkon SD, Cassidy D.

Department of Rehabilitation Medicine, University of Washington; and Department of Rehabilitation Medicine, Seattle Children's, M/S W-9847, PO Box 5371, Seattle, WA().

The use of botulinum toxins to decrease spasticity in children with cerebral palsy has become standard of care during the past decade. In 2008 reports of severe adverse events, including death, were reported in children who received injections of these medications. The following discussion focuses on the background of these reports, the response of the U.S. Food and Drug Administration, as well as the safety profile and pharmacokinetics of botulinum toxins. Finally, the authors will offer their perspective on the safe use of botulinum toxins.

PMID: 20430330 [PubMed - as supplied by publisher]

16. Dev Med Child Neurol. 2010 Jan;52(1):8-9.

Repeat upper limb botulinum toxin A injections: a reflection of clinical practice.

Sakzewski L.

Queensland Cerebral Palsy and Research Centre, School of Medicine, The University of Queensland, Brisbane, Queensland, Australia.

Comment on:

Dev Med Child Neurol. 2010 Jan;52(1):79-86.

PMID: 20419869 [PubMed - in process]

17. Brain. 2010 May;133(Pt 5):1292-4.

Réflexes de défense.

By J. Babinski, Brain 1922: 45; 149-184; with The physiological significance of the reflex phenomena in spastic paralysis of the lower limbs. By F. M. R. Walshe, Brain 1914: 37; 269-336; and The Babinski plantar response, its forms, and its physiological and pathological significance. By F. M. R. Walshe, Brain 1956: 79; 529-556.
van Gijn J.

PMID: 20432593 [PubMed - in process]

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.

18. Pediatr Radiol. 2010 Jun;40(6):811-8. Epub 2010 Apr 30.

Brain lesions in preterm infants: initial diagnosis and follow-up.

Argyropoulou MI.

Department of Radiology, Medical School, University of Ioannina, 45110 Ioannina, Greece. margyrop@cc.uoi.gr

Children surviving premature birth present with a wide spectrum of motor, sensory and cognitive disabilities, ranging from slight motor deficits, school difficulties and behavioural problems to cerebral palsy and mental retardation. The anatomic and functional substrate of these problems can be investigated using a variety of imaging techniques. Cranial US coupled with colour Doppler is a well-established method for the initial diagnosis of intraventricular haemorrhage, parenchymal haemorrhagic infarct and periventricular leukomalacia. MRI is useful for the follow-up study of brain maturation. Conventional T1- and T2-weighted sequences, magnetization transfer and diffusion tensor imaging coupled with sophisticated tools of tissue segmentation and analysis at a voxel level offer substantial anatomic and functional information on pathological conditions that define the prognosis of preterm infants.

PMID: 20431999 [PubMed - in process]

19. J Clin Endocrinol Metab. 2010 Apr 28. [Epub ahead of print]

Missense Mutations in the Melanocortin 2 Receptor Accessory Protein That Lead to Late Onset Familial Glucocorticoid Deficiency Type 2.

Hughes CR, Chung TT, Habeb AM, Kelestimur F, Clark AJ, Metherell LA.

Queen Mary University of London (C.R.H., T.T.C., L.A.M., A.J.L.C.), Centre for Endocrinology, William Harvey Research Institute, Barts and the London School of Medicine, London EC1M 6BQ, United Kingdom; Pediatric Endocrine Unit (A.M.H.), Maternity and Children Hospital, Madinah, Kingdom of Saudi Arabia; and Department of Endocrinology and Metabolism (F.K.), Erciyes University Medical School, 38039 Kayseri, Turkey.

Background: Familial glucocorticoid deficiency (FGD) is an autosomal recessive disorder characterized by isolated glucocorticoid deficiency. Mutations in the ACTH receptor [melanocortin 2 receptor (MC2R)] or the MC2R accessory protein (MRAP) cause FGD types 1 and 2, respectively. Typically, type 2 patients present early (median age, 0.1 yr), and no patient reported to date has presented after 1.6 yr. Aim: The aim of this study was to investigate the cause of disease in two families with late-onset FGD. Patients: The proband in family 1 was diagnosed at age 4 yr. Family review revealed two older siblings with undiagnosed FGD. One sibling was well, whereas the second had cerebral palsy secondary to hypoglycemic seizures. The proband in family 2 was diagnosed at age 18 yr with symptoms of fatigue, weight loss, and depression. Methods: The coding exons of MC2R and MRAP were sequenced.

ACTH dose-response curves were generated for MC2R when transfected with wild-type or mutant MRAP constructs using HEK293 cells. MC2R trafficking with both mutant MRAPs was investigated using immunocytochemistry. Results: MRAP gene analysis identified two novel homozygous missense mutations, c.175T>G (pY59D) in family 1 and c.76T>C (p.V26A) in family 2. In vitro analysis showed that the Y59D mutant had significant impairment of cAMP generation, and both mutants caused a shift in the dose-response curve to the right when compared to wild type. Immunocytochemistry showed normal trafficking of MC2R when transfected with both mutant MRAPs, indicating a probable signaling defect. Conclusion: These results indicate that missense MRAP mutations present with a variable phenotype of ACTH resistance and can present late in life.

PMID: 20427498 [PubMed - as supplied by publisher]

20. Am J Obstet Gynecol. 2010 Apr 22. [Epub ahead of print]

Infant death among Ohio resident infants born at 32 to 41 weeks of gestation.

Donovan EF, Besl J, Paulson J, Rose B, Iams J; Ohio Perinatal Quality Collaborative.

Child Policy Research Center, Cincinnati Children's Hospital Medical CenterThe Ohio State University, Cincinnati, OH; Ohio Perinatal Quality Collaborative, Columbus, OH.

OBJECTIVE: The aim of this study was to determine gestational age-specific, adjusted infant mortality rates for Ohio. STUDY DESIGN: Using a retrospective cohort design, all births and infant deaths from 2003-2005 were included in multivariable regression analyses. Variations in cause and timing of infant death were determined. RESULTS: Compared with births at 39 or 40 weeks, adjusted likelihood of infant death increased progressively between 38-32 weeks' gestational age. At later gestational ages, death was more likely caused by sudden infant death syndrome or intentional injury compared with congenital malformations and asphyxia or cerebral palsy at earlier gestational ages. Less mature infants tended to die earlier. CONCLUSION: The current study confirms for Ohio and extends the findings of others that infant mortality risk is increased for births at late preterm and near-term gestational ages. Decisions to deliver before 39 weeks should consider increased likelihood of infant death that may be unrelated to fetal malformations or maternal illness.

PMID: 20417495 [PubMed - as supplied by publisher]

21. Neurology. 2010 Apr 27;74(17):1386-91.

Cross-sectional comparison of periventricular leukomalacia in preterm and term children.

Lasry O, Shevell MI, Dagenais L; REPACQ Consortium.

Collaborators (10)Boucher MD, Desjardins C, Fortier J, Koclas L, Lamarre C, Malouin F, Mathieu J, Munz D, Pigeon N, Richards CL.
Division of Pediatric Neurology, Montreal Children's Hospital-McGill University Health Centre, Montreal, Quebec, Canada.

OBJECTIVE: To document and contrast the characteristics of preterm and term-born children with cerebral palsy attributed to underlying radiologic periventricular white matter injury (leukomalacia) (PVWMI/PVL). METHODS: A comprehensive cerebral palsy population-based registry (REPACQ) for a 4-year inclusive (1999-2002) birth cohort was systematically searched for all children with radiologic evidence for PVWMI/PVL. Clinical features, neurologic subtype, gross motor functional impairment, and comorbidities were compared in those children born preterm (<37 weeks) and those born at term (> or = 37 weeks). RESULTS: Of 242 children with cerebral palsy in the registry, 213 had available neuroimaging, in which 41 had PVWMI/PVL: 26 preterm born and 15 term born. Neurologic subtype differed significantly between preterm and term-born children with respect to the frequency of spastic hemiplegia (5/26 vs 8/15; $p < 0.05$) and spastic diplegia (9/26 vs 2/15; $p < 0.05$). The groups also differed significantly from a functional perspective (Gross Motor Function Classification System for Cerebral Palsy level I-II; 12/26 vs 12/15; $p < 0.05$). The comorbidity spectrum was similar between the 2 groups except for the occurrence of cortical blindness in the term-born children (3/15 vs 0/26; $p < 0.05$). CONCLUSION: Differences between preterm and term-born children with cerebral palsy with periventricular white matter injury (leukomalacia) suggest that despite a common radiologic pattern, these are different clinicopathologic entities with perhaps a different gestational timing of acquired

injury.

PMID: 20421583 [PubMed - in process]

22. Neuroradiology. 2010 Apr 27. [Epub ahead of print]

Magnetic resonance imaging of white matter diseases of prematurity.

Rutherford MA, Supramaniam V, Ederies A, Chew A, Bassi L, Groppo M, Anjari M, Counsell S, Ramenghi LA.

Robert Steiner MR Unit, MRC Clinical Sciences Centre, Imperial College, Hammersmith Hospital, Du Cane Road, W12 OHS, London, UK, m.rutherford@imperial.ac.uk.

Abstract
Periventricular leucomalacia (PVL) and parenchymal venous infarction complicating germinal matrix/intraventricular haemorrhage have long been recognised as the two significant white matter diseases responsible for the majority of cases of cerebral palsy in survivors of preterm birth. However, more recent studies using magnetic resonance imaging to assess the preterm brain have documented two new appearances, adding to the spectrum of white matter disease of prematurity: punctate white matter lesions, and diffuse excessive high signal intensity (DEHSI). These appear to be more common than PVL but less significant in terms of their impact on individual neurodevelopment. They may, however, be associated with later cognitive and behavioural disorders known to be common following preterm birth. It remains unclear whether PVL, punctate lesions, and DEHSI represent a continuum of disorders occurring as a result of a similar injurious process to the developing white matter. This review discusses the role of MR imaging in investigating these three disorders in terms of aetiology, pathology, and outcome.

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23. Neuroimage. 2010 Apr 23. [Epub ahead of print]

Atlas-based analysis of neurodevelopment from infancy to adulthood using diffusion tensor imaging and applications for automated abnormality detection.

Faria AV, Zhang J, Oishi K, Li X, Jiang H, Akhter K, Hermoye L, Lee SK, Hoon A, Stachinko E, Miller MI, van Zijl PC, Mori S.

The Russell H. Morgan Department of Radiology and Radiological Science, The Johns Hopkins University School of Medicine, Baltimore, MD, U.S.A.; Department of Radiology, University of Campinas, Campinas, SP, Brazil.

Quantification of normal brain maturation is a crucial step in understanding developmental abnormalities in brain anatomy and function. The aim of this study was to develop atlas-based tools for time-dependent quantitative image analysis, and to characterize the anatomical changes that occur from 2 years of age to adulthood. We used large deformation diffeomorphic metric mapping to register diffusion tensor images of normal participants into the common coordinates and used a pre-segmented atlas to segment the entire brain into 176 structures. Both voxel- and atlas-based analyses reported structure that showed distinctive changes in terms of its volume and diffusivity measures. In the white matter, fractional anisotropy (FA) linearly increased with age in logarithmic scale, while diffusivity indices, such as apparent diffusion coefficient (ADC), and axial and radial diffusivity, decreased at a different rate in several regions. The average, variability, and the time course of each measured parameter are incorporated into the atlas, which can be used for automated detection of developmental abnormalities. As a demonstration of future application studies, the brainstem anatomy of cerebral palsy patients was evaluated and the altered anatomy was delineated.

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24. Paediatr Perinat Epidemiol. 2010 Mar;24(2):149-55.

Socio-economic inequalities in cerebral palsy prevalence in the United Kingdom: a register-based study.

Dolk H, Pattenden S, Bonellie S, Colver A, King A, Kurinczuk JJ, Parkes J, Platt MJ, Surman G.

Public Health and Primary Care Research Group, Institute for Nursing Research, University of Ulster, Shore Road, Newtownabbey, Northern Ireland, UK. h.dolk@ulster.ac.uk

Evidence is unclear as to whether there is a socio-economic gradient in cerebral palsy (CP) prevalence beyond what would be expected from the socio-economic gradient for low birthweight, a strong risk factor for CP. We conducted a population-based study in five regions of the UK with CP registers, to investigate the relationship between CP prevalence and socio-economic deprivation, and how it varies by region, by birthweight and by severity and type of CP. The total study population was 1 657 569 livebirths, born between 1984 and 1997. Wards of residence were classified into five quintiles according to a census-based deprivation index, from Q1 (least deprived) to Q5 (most deprived). Socio-economic gradients were modelled by Poisson regression, and region-specific estimates combined by meta-analysis. The prevalence of postneonatally acquired CP was 0.14 per 1000 livebirths overall. The mean deprivation gradient, expressed as the relative risk in the most deprived vs. the least deprived quintile, was 1.86 (95% confidence interval [95% CI 1.19, 2.88]). The prevalence of non-acquired CP was 2.22 per 1000 livebirths. For non-acquired CP the gradient was 1.16 [95% CI 1.00, 1.35]. Evidence for a socio-economic gradient was strongest for spastic bilateral cases (1.32 [95% CI 1.09, 1.59]) and cases with severe intellectual impairment (1.59 [95% CI 1.06, 2.39]). There was evidence for differences in gradient between regions. The gradient of risk of CP among normal birthweight births was not statistically significant overall (1.21 [95% CI 0.95, 1.54]), but was significant in two regions. There was non-significant evidence of a reduction in gradients over time. The reduction of the higher rates of postneonatally acquired CP in the more socioeconomically deprived areas is a clear goal for prevention. While we found evidence for a socio-economic gradient for non-acquired CP of antenatal or perinatal origin, the picture was not consistent across regions, and there was some evidence of a decline in inequalities over time. The steeper gradients in some regions for normal birthweight cases and cases with severe intellectual impairment require further investigation.

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