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## Interventions and Management

**1. J Neuroeng Rehabil. 2013 Jun 15;10(1):57. [Epub ahead of print]**

**Contribution of hip joint proprioception to static and dynamic balance in cerebral palsy: a case control study.**

Damiano DL, Wingert JR, Stanley CJ, Curatalo L.

**BACKGROUND:** Balance problems are common in cerebral palsy (CP) but etiology is often uncertain. The classic Romberg test compares ability to maintain standing with eyes open versus closed. Marked instability without vision is a positive test and generally indicates proprioceptive loss. From previous work showing diminished hip joint proprioception in CP, we hypothesized that static and dynamic balance without vision (positive Romberg) would be compromised in CP. **METHODS:** Force plate sway and gait velocity data were collected using 3D motion capture on 52 participants, 19 with diplegic CP, 13 with hemiplegic CP, and 20 without disability. Center of mass (COM) and center of pressure (COP) velocity, excursion, and differences between COM and COP in AP and ML directions were computed from static standing trials with eyes open and closed. Mean gait velocity with and without dribble glasses was compared. Hip joint proprioception was quantified as the root mean square of magnitude of limb positioning errors during a hip rotation task with and without view of the limb. Mixed model repeated measures analysis of variance (ANOVA) was performed with condition as within-subject (EO, EC) and group as between-subject factors (hemiplegia, diplegia, controls). Sway characteristics and gait speed were correlated with proprioception values. Groups with CP had greater sway in standing with eyes open indicating that they had poorer balance than controls, with the deficit relatively greater in the ML compared to AP direction. Contrary to our hypothesis, the decrement with eyes closed did not differ from controls (negative Romberg); however, proprioception error was related to sway parameters particularly for the non-dominant leg. Gait speed was related to proprioception values such that those with worse proprioception tended to walk more slowly. **CONCLUSIONS:** Postural instability is present even in those with mild CP and is yet another manifestation of their motor control disorder, the specific etiology of which may vary across individuals in this heterogeneous diagnostic category.

[PMID: 23767869](https://pubmed.ncbi.nlm.nih.gov/23767869/) [PubMed - as supplied by publisher]

**2. Disabil Health J. 2013 Jul;6(3):244-52. doi: 10.1016/j.dhjo.2013.01.013. Epub 2013 Mar 17.**

**Gait and participation outcomes in adults with cerebral palsy: A series of case studies using mixed methods.**

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**BACKGROUND:** There is a paucity of information on long-term outcomes of adults with cerebral palsy (CP) who received orthopedic interventions in childhood. Clinical effectiveness research requires assessment of outcomes that account for personal and environmental factors that may mediate the effects of treatment, in addition to body structures & function, activity, and participation. **OBJECTIVE/HYPOTHESIS:** The purpose of this study is to provide a descriptive analysis of characteristics associated with gait and participation outcomes in a series of case studies of adults with CP. **METHODS:** Participants had follow up gait analysis and clinical evaluation in adulthood and assessment of outcomes with the FIM(®) instrument, the SF-36(®) Health survey, the Canadian Occupational Performance Measure, and semi-structured questions. **RESULTS:** Twenty-two out of 26 participants (mean age = 25 years; GMFCS level I (n = 9); II (n = 3); III (n = 11); IV (n = 3)) maintained or improved childhood gait abilities, with levels of participation in society similar to age matched peers. Higher level of severity and personal choices impacted gait abilities in the four who declined. Majority of participants lost range of motion in hip flexion and knee extension, had pain, reported a fitness program, and increased in weight status. Personal factors and environmental factors played a role in both gait and participation outcomes. **CONCLUSION:** Promotion of fitness activities and social advocacy are warranted for adults with CP. Clinical effectiveness research of long-term impact of orthopedic interventions should account for treatment effects on body structures & function, activity, participation, and modifying effects of personal, and environmental factors.

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[PMID: 23769484](#) [PubMed - in process]

**3. Gait Posture. 2013 Jun 14. pii: S0966-6362(13)00236-1. doi: 10.1016/j.gaitpost.2013.05.006. [Epub ahead of print]**

**A new method for measuring AFO deformation, tibial and footwear movement in three dimensional gait analysis.**

Ridgewell E, Sangeux M, Bach T, Baker R.

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Solid ankle-foot orthoses (AFOs) are designed to immobilise the ankle but numerous studies have measured a considerable ankle range of motion (ROM) in AFO users. Measurement of ankle kinematics may be affected by soft-tissue artefact (STA) of the knee marker, deformation of the AFO or tibial movement within the AFO. A new model based on the Conventional Gait Model (CGM) was developed to calculate these effects. Although movement of the AFO within the shoe should not affect the measured ankle joint angle the model also allows an estimation of this movement. Seven children (13 limbs) with spastic diplegic cerebral palsy were assessed to present the benefits of the new model compared to the CGM. STA of the knee marker was estimated to result in a 1.5° overestimation of total ankle ROM (from 8.2° to 9.7°). STA error was strongly related to angle of knee flexion ( $r=0.82$ ) with an average maximum error of 3.8°. AFO deformation contributed approximately two thirds of the ankle ROM ( $6.0\pm 4.3^\circ$ ) with the remaining third from tibial movement relative to the AFO ( $2.8\pm 0.9^\circ$ ). Movement of the AFO within the footwear was very small ( $1.8\pm 0.8^\circ$ ). A strong positive relationship ( $r=0.9$ ) was found between body mass (kg) and AFO deformation which was statistically significant ( $p<0.001$ ). This is the first model to attempt to quantify different contributions to ankle dorsiflexion measured during gait analysis of people wearing AFOs.

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**4. Clin Neuropharmacol. 2013 Jun 17. [Epub ahead of print]****Dysport (Botulinum Toxin Type A) in Routine Therapeutic Usage: A Telephone Needs Assessment Survey of European Physicians to Evaluate Current Awareness and Adherence to Product Labeling Changes.**

Hubble J, Schwab J, Hubert C, Abbott CC.

\*Ipsen Biopharmaceuticals, Inc, Basking Ridge, NJ; and †Stethos, Sevres, France.

**BACKGROUND:** Botulinum neurotoxin type A is a well-established treatment for a number of conditions involving muscle hyperactivity. Dysport (Ipsen Ltd, Wrexham, United Kingdom) is a botulinum neurotoxin type A preparation that has been available for a number of therapeutic uses for over 20 years in the European Union (EU). This survey was part of the EU botulinum toxin risk management plan to identify potential educational needs of injectors by collecting data on their routine practice administration of Dysport and their awareness of potential adverse events (AEs) that are included in the current product labeling. **METHODS:** Dysport-experienced injectors in 5 EU countries were surveyed via telephone about their experience of Dysport in patients with cervical dystonia, adult upper and lower limb spasticity, pediatric cerebral palsy, and blepharospasm/hemifacial spasm. **RESULTS:** The reconstitution dilution volume most often used was 2.5 mL per 500 U for all indications. The mean total dose ranged from 387 to 530 U for cervical dystonia, 508 to 773 U for upper limb spasticity, 600 to 832 U for lower limb spasticity, 375 to 700U for pediatric cerebral palsy, and 54 to 213U for blepharospasm/hemifacial spasm. The potential AEs most commonly mentioned by surveyed physicians were dysphagia for cervical dystonia, arm muscle weakness for upper limb spasticity, leg muscle weakness for lower limb spasticity, and pediatric cerebral palsy and ptosis for blepharospasm/hemifacial spasm. **CONCLUSIONS:** The results indicate that product-labeling recommendations are generally applied in clinical practice and that there is a good familiarity with potential AEs based on clinical condition. Nevertheless, the survey shows that experienced injectors do sometimes deviate from the manufacturers labeling recommendations, highlighting the importance of ongoing education.

[PMID: 23783005](#) [PubMed - as supplied by publisher]

**5. Dev Med Child Neurol. 2013 Jun;55(6):492. doi: 10.1111/dmcn.12131. Epub 2013 Mar 14.****All in the hope of saving a few minutes: improving efficiency of the GMFM-66.**

White H.

Motion Analysis Laboratory, Shriners Hospitals for Children, Lexington, KY, USA.

Comment on: Criterion validity of the GMFM-66 item set and the GMFM-66 basal and ceiling approaches for estimating GMFM-66 scores. [Dev Med Child Neurol. 2013]

[PMID: 23489063](#) [PubMed - indexed for MEDLINE]

**6. J Paediatr Child Health. 2013 Jun 18. doi: 10.1111/jpc.12289. [Epub ahead of print]****'Every picture tells a story': Interviews and diaries with children with cerebral palsy about adapted cycling.**

Pickering D, Horrocks LM, Visser KS, Todd G.

Department of Physiotherapy, School of Healthcare Studies, Cardiff University, Cardiff.

**AIMS:** The study aims to explore the effect of participation in adapted cycling on quality of life for children with cerebral palsy (CP). **METHODS:** The study used a qualitative approach developing creative mosaic methods using interviews and diaries. This study had two groups of children with CP: those who participated in adapted cycling and a control group who had not yet started. Children with CP aged 2-17 years who were already cycling were invited to take part in two interviews at the beginning and end of a 6-week period and keep a cycling diary during this time. The control group of children with CP aged 2-17 years were asked to keep a diary of physical activities over 6 weeks and to take part in one interview at the end. Welsh, English, Bengali and Arabic were the languages spoken, and some children used communication aids. **RESULTS:** Results presented here include the first 25

children's interviews and diaries. The emergent themes from the analysis are: learning a new skill, the impact on wider family and friends, social participation and future aspirations. The diaries added an emotional dimension, by illustrations drawn by the children. **CONCLUSION:** The children who took part in adapted cycling enjoyed this experience and it improved their sense of well-being. Some in the control group went on to participate in adapted cycling. Physiotherapists can carry out creative research to hear the voices of children and young people with CP and incorporate their views and ideas into the development of service model delivery and treatment programmes.

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## **7. Disabil Rehabil Assist Technol. 2013 Jun 19. [Epub ahead of print]**

### **Patient and equipment profile for wheelchair seating clinic provision.**

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**Purpose:** To characterise the provision of wheelchair seating both pre- and post-clinical intervention and compare and contrast the two largest diagnostic groups. **Method:** The case notes of those attending a wheelchair seating clinic for adults over a defined period were reviewed retrospectively. A classification system was devised that delineates between the complexity and type of equipment to gain a better understanding of provision. **Results:** 146 patients were included; mean age 45 years (SD 16); 53.4% male. The two most prevalent primary medical diagnoses were cerebral palsy (CP) and multiple sclerosis (MS); 48.6% and 20.5%, respectively. The MS group, in comparison to the CP group, were significantly more likely to be older, new to seating provision, have been seen more recently, have a powered wheelchair, self-propel their manual wheelchair, have low rather than high complexity equipment and have their equipment changed following assessment. **Conclusions:** The equipment classification system will allow results from different studies to be readily compared. The results for those with CP and MS reflect the respective stable and progressive nature of these conditions. Referrals for those with MS should be prioritised. Wheelchair seating users with MS should be reassessed ~18 months after provision. **Implications for Rehabilitation** A detailed classification of wheelchair seating equipment based on a recognised standard vocabulary, such as the one proposed, is required to gain a better understanding of provision. Wheelchair seating equipment budget and staffing levels should reflect the diagnostic make up of a service's patient population. Referrals for people with MS should be prioritised as their current wheelchair seating provision is more likely not to be meeting their needs. People with MS should have a clinical review 18 months after wheelchair seating provision.

[PMID: 23782226](#) [PubMed - as supplied by publisher]

## **8. J Neuroeng Rehabil. 2013 Jun 15;10:56. doi: 10.1186/1743-0003-10-56.**

### **Femoral anteversion and tibial torsion only explain 25% of variance in regression analysis of foot progression angle in children with diplegic cerebral palsy.**

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**BACKGROUND:** The relationship between torsional bony deformities and rotational gait parameters has not been sufficiently investigated. This study was to investigate the degree of contribution of torsional bony deformities to rotational gait parameters in patients with diplegic cerebral palsy (CP). **METHODS:** Thirty three legs from 33 consecutive ambulatory patients (average age 9.5 years, SD 6.9 years; 20 males and 13 females) with diplegic CP who underwent preoperative three dimensional gait analysis, foot radiographs, and computed tomography (CT) were included. Adjusted foot progression angle (FPA) was retrieved from gait analysis by correcting pelvic rotation from conventional FPA, which represented the rotational gait deviation of the lower extremity from the tip of the

femoral head to the foot. Correlations between rotational gait parameters (FPA, adjusted FPA, average pelvic rotation, average hip rotation, and average knee rotation) and radiologic measurements (acetabular version, femoral anteversion, knee torsion, tibial torsion, and anteroposterior-orto-first metatarsal angle) were analyzed. Multiple regression analysis was performed to identify significant contributing radiographic measurements to adjusted FPA. **RESULTS:** Adjusted FPA was significantly correlated with FPA ( $r=0.837$ ,  $p<0.001$ ), contralateral FPA ( $r=0.492$ ,  $p=0.004$ ), pelvic rotation during gait ( $r=-0.489$ ,  $p=0.004$ ), knee rotation during gait ( $r=0.376$ ,  $p=0.031$ ), and femoral anteversion ( $r=0.350$ ,  $p=0.046$ ). In multiple regression analysis, femoral anteversion ( $p=0.026$ ) and tibial torsion ( $p=0.034$ ) were found to be the significant contributing structural deformities to the adjusted FPA ( $R^2=0.247$ ). **CONCLUSIONS:** Femoral anteversion and tibial torsion were found to be the significant structural deformities that could affect adjusted FPA in patients with diplegic CP. Femoral anteversion and tibial torsion could explain only 24.7% of adjusted FPA.

[PMID: 23767833](#) [PubMed - in process] [PMCID: PMC3686583](#)

**9. Res Dev Disabil. 2013 Jun 11;34(9):2684-2693. doi: 10.1016/j.ridd.2013.05.002. [Epub ahead of print]**

**Identification of gait patterns in individuals with cerebral palsy using multiple correspondence analysis.**

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Great importance has been placed on the development of gait classification in cerebral palsy (CP) to assist clinicians. Nevertheless, gait classification is challenging within this group because the data is characterized by a high-dimensionality and a high-variability. Thus, the aim of this study was to analyze without a priori, a database of clinical gait analysis (CGA) of CP patients, using multiple correspondence analysis (MCA). A retrospective search, including biomechanical and clinical parameters was done between 2006 and 2012. One hundred and twenty two CP patients were included in this study (51 females and 71 males, mean age $\pm$ SD: 14.2 $\pm$ 7.5 years). Sixteen biomechanical spatio-temporal and kinematic parameters were included in the analysis. This data was transformed by a fuzzy window coding based on the distribution of each parameter in three modalities: low, average and high. Afterward, a MCA was used to associate parameters and to define classes. From this, seven most explicative gait parameters used to characterize gait of CP patients were identified: maximal hip extension, hip range, knee range, maximal knee flexion at initial contact, time of peak knee flexion, and maximal ankle dorsiflexion in stance phase and in swing phase. Moreover, four main profiles of CP patients have been defined from the multivariate approach: an apparent equinus gait group (the most similar of the control group with diplegic and hemiplegic patients with a GMFCS 1), a true equinus gait group (the youngest group with diplegic and some hemiplegic patients with a GMFCS 1), a crouch gait group (the oldest group with a majority of diplegic and rare hemiplegic patients with a GMFCS 2) and a jump knee gait group (the greatest level of global spasticity of the lower limbs with a majority of diplegic and rare hemiplegic patients with a GMFCS 2). Thus, this study showed the feasibility of the MCA in order to characterize and classify a large database of CP patients.

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**10. J Pediatr Orthop. 2013 Jul-Aug;33 Suppl 1:S29-32. doi: 10.1097/BPO.0b013e3182860034.**

**Pathologic hip morphology in cerebral palsy and down syndrome.**

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The pathogenesis and clinical course of hip dysplasia in cerebral palsy and Down syndrome is different than idiopathic developmental dysplasia of the hip. Unlike idiopathic developmental hip dysplasia, hip development in cerebral palsy and Down syndrome is typically normal in utero and instability develops after birth secondary to

musculoskeletal disorders associated with the disease condition. For this reason, treatment protocols in Down syndrome and cerebral palsy hip dysplasia differ greatly from protocols designed to treat idiopathic hip dysplasia. The purpose of this review is to describe the pathologic hip morphology that results from cerebral palsy and Down syndrome.

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## 11. Pediatrics. 2013 Jun 17. [Epub ahead of print]

### **Osteopathic Manipulative Treatment for Pediatric Conditions: A Systematic Review.**

Posadzki P, Lee MS, Ernst E.

Medical Research Division, Korea Institute of Oriental Medicine, Daejeon, South Korea; and.

**BACKGROUND AND OBJECTIVES:** Most osteopaths are trained in pediatric care, and osteopathic manipulative treatment (OMT) is available for many pediatric conditions. The objective of this systematic review was to critically evaluate the effectiveness of OMT as a treatment of pediatric conditions. **METHODS:** Eleven databases were searched from their respective inception to November 2012. Only randomized clinical trials (RCTs) were included, if they tested OMT against any type of control in pediatric patients. Study quality was critically appraised by using the Cochrane criteria. **RESULTS:** Seventeen trials met the inclusion criteria. Five RCTs were of high methodological quality. Of those, 1 favored OMT, whereas 4 revealed no effect compared with various control interventions. Replications by independent researchers were available for 2 conditions only, and both failed to confirm the findings of the previous studies. Seven RCTs suggested that OMT leads to a significantly greater reduction in the symptoms of asthma, congenital nasolacrimal duct obstruction (posttreatment), daily weight gain and length of hospital stay, dysfunctional voiding, infantile colic, otitis media, or postural asymmetry compared with various control interventions. Seven RCTs indicated that OMT had no effect on the symptoms of asthma, cerebral palsy, idiopathic scoliosis, obstructive apnea, otitis media, or temporomandibular disorders compared with various control interventions. Three RCTs did not perform between-group comparisons. The majority of the included RCTs did not report the incidence rates of adverse effects. **CONCLUSIONS:** The evidence of the effectiveness of OMT for pediatric conditions remains unproven due to the paucity and low methodological quality of the primary studies.

[PMID: 23776117](#) [PubMed - as supplied by publisher]

## 12. Z Orthop Unfall. 2013 Jun;151(3):248-56. doi: 10.1055/s-0032-1328492. Epub 2013 Jun 14.

### **Determination of activity levels in patients with cerebral palsy [Article in German]**

Lampe R, Mitternacht J, Merdanovic E, Salzmann M, Pilge H.

Orthopädische Klinik, Technische Universität München.

**Aim:** The aim of this study is the determination of physical activity in everyday life of handicapped persons due to cerebral palsy using standardised testing procedures. **Method:** The subjects were examined according to the Conconi test on a treadmill at increasing speeds for their fitness. We used a continuous heart rate monitoring, 3-axis acceleration sensors, lactate measurements and pedography. **Results:** Three groups of subjects could be differentiated. In one group, a rapid rise in heart rate even at lower walking speed was observed. In a second group of persons with poor motor coordination, the maximum walking speed was limited. Single subjects were not limited neither in their physical performance nor in their condition. **Conclusion:** In the physical therapy for patients with cerebral palsy one should not lose sight of possible cardiovascular limitations additional to the physical disabilities.

[PMID: 23771330](#) [PubMed - in process]

**13. Int J Pediatr Otorhinolaryngol. 2013 Jun 15. pii: S0165-5876(13)00262-0. doi: 10.1016/j.ijporl.2013.05.025. [Epub ahead of print]**

**Pathological consideration of tracheo-innominate artery fistula with a case report.**

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**OBJECTIVES:** Tracheo-innominate artery fistula (TIF) is a rare but life-threatening complication of tracheostomy. There are many reports about TIF but the mechanism of TIF formation remains poorly documented. Our objective is to investigate the TIF pathologically and suggest the possible mechanism of TIF formation. **PATIENT AND METHODS:** The patient was an 11-year old boy with a history of severe childhood epilepsy, cerebral palsy, and psychomotor retardation who died from TIF. We investigated the TIF histopathologically through his autopsy and reviewed bibliographical considerations. **RESULTS:** Autopsy revealed massive blood aspiration from a large TIF and histopathological findings showed the following: (1) fibroepithelial polyps around the fistula; (2) squamous metaplasia of tracheal mucosa; (3) disappearance of tracheal cartilages and tracheal glands; and (4) arteritis (infiltration of neutrophils into the mesarterium and endarterium). Probably, (1) and (2) indicate chronic and repetitive irritations caused by cannula. (3) and (4) indicate intense inflammation caused by local infection. In this case, it is suggested that the chronic damage by cannula and the local infection resulted in the fistula formation and rupture of innominate artery. **CONCLUSION:** The present case suggests a pathogenesis in which chronic damage to the anterior tracheal wall by the cannula and consequent infection led to fistulation and rupture of the innominate artery.

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**14. Dent Assist. 2013 Mar-Apr;82(2):36, 38, 48.**

**Dental assistants and the care of individuals with disabilities: Part II.**

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[PMID: 23691606](#) [PubMed - indexed for MEDLINE]

**15. Disabil Rehabil. 2013 Jun 20. [Epub ahead of print]**

**Social support and adaptation outcomes in children and adolescents with cerebral palsy.**

Carona C, Moreira H, Silva N, Crespo C, Canavarro MC.

Faculty of Psychology and Education Sciences, The University of Coimbra, Coimbra, Portugal.

**Objectives:** This study had two main objectives: first, to describe the social support and psychological maladjustment of children and adolescents with cerebral palsy (CP); and second, to test a mediation model where psychological maladjustment was hypothesized to mediate the link between social support and health-related quality of life (HRQL). In addition, the moderating role of gender and age was examined for this mediation model. **Methods:** Self- and proxy-report questionnaires on the aforementioned variables were administered to a sample of 96 children/adolescents with CP and 118 healthy controls, as well as one of their parents. Univariate and multivariate analyses of covariance were conducted to examine differences in social support and psychological maladjustment, respectively. PROCESS computational tool was used for path analysis-based mediation, moderation and moderated mediation analyses. **Results:** Children/adolescents with CP reported lower levels of social support than their healthy peers, but no significant differences emerged in terms of their psychological maladjustment. For children/adolescents with CP, internalizing and externalizing problems were found to mediate

the link between social support and HRQL, and these indirect effects were not conditional upon age or gender. Discussion: Children and adolescents with CP are likely have more negative perceptions of social support, but not necessarily more psychological adjustment problems than their healthy, able-bodied peers. Results further suggest that interventions targeting social support perceptions may positively affect HRQL outcomes in children/adolescents with CP, through the improvement of internalizing and externalizing dimensions of their psychological adjustment. Implications for Rehabilitation Social support perceptions are important intervention targets in psychosocial rehabilitation with children and adolescents with CP. Children and adolescents with CP do not necessarily present increased psychological maladjustment. Interventions targeting these children and adolescents' social support may promote their psychological adjustment and health-related quality of life. Developmental specificities, such as age and gender differences, should be considered when planning and implementing psychosocial interventions.

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**16. Res Dev Disabil. 2013 Jun 17;34(9):2725-2733. doi: 10.1016/j.ridd.2013.05.017. [Epub ahead of print]**

**Comparison of health-related quality of life between children with cerebral palsy and spina bifida.**

Tezcan S, Simsek TT.

Special Ilgim Special Education and Rehabilitation Center, Düzce, Turkey.

This study has two aims-the first is to compare the Health Related Quality of Life (HRQoL) between children with cerebral palsy (CP) and children with spina bifida (SB); the second is to investigate the relationship between HRQoL and age, sex, body mass index (BMI), level of ambulation, cooperation, family income and the mother's education level in both groups of children. The study included 96 children with CP and 70 children with SB (aged 5-18) who attended a physiotherapy and rehabilitation program at an institute of special training and rehabilitation. Socio-demographic information was obtained within the study. The Child Health Questionnaire (CHQ-PF50) was used to evaluate HRQoL. A significant difference was found in terms of age and BMI between children with CP and SB ( $p < 0.05$ ). HRQoL was lower for children with CP. There was a significant difference between the two groups in terms of role/social limitations - emotional behavioral, behavior, global behavior, parental impact-emotional and parental impact-time ( $p < 0.05$ ). A positive correlation was found between BMI and self-esteem in children with SB, unlike children with CP. The HRQoL of children with CP was lower than children with SB. The parameters of behavior and parental impact were particularly affected in the children with CP. Minimizing behavioral problems (which can improve with advancing age) of the children with CP and reducing parental impact are important for improving the HRQoL of both the child and parents. There is a need for further studies on this issue.

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**17. Res Dev Disabil. 2013 Jun 12;34(9):2694-2700. doi: 10.1016/j.ridd.2013.05.029. [Epub ahead of print]**

**Assistive technology for promoting choice behaviors in three children with cerebral palsy and severe communication impairments.**

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A technology-based program to promote independent choice behaviors by three children with cerebral palsy and multiple disabilities was assessed. The program was based on learning principles and assistive technology (i.e., customized input devices/sensors, personal computers, screening of preferred stimuli according to a binomial criterion). The first purpose of the present study was to provide the participants with a new set-up of assistive technology and to allow them to choose among three categories (i.e., food, beverage and leisure), and to request a specific item out of four in each category, adopting a procedure that minimized (according to a conditional probability criterion) unintentional choices. The second aim of the study was to carry out the effects of the program on detectable mood signs (i.e., happiness index). The study was conducted according to an ABAB sequence with a subsequent post intervention check for each participant. The results showed an increase of engagement and of the

happiness index during intervention phases. Psychological as well as educational implications were discussed.

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## Prevention and Cure

### 18. J Registry Manag. 2013 Spring;40(1):9-13.

#### **Collaborating with consumers: the key to achieving statutory notification for birth defects and cerebral palsy in Western Australia.**

Bower C, McKenzie A, Watson L, Charles A.

**Introduction:** The Western Australian Birth Defects Registry and the Western Australian Cerebral Palsy Register used multiple sources of voluntary notification without consent and have a high level of case ascertainment, but there were concerns over privacy and a call for statutory notification. **Objective:** To seek consumer consensus on whether notification to the registers should be statutory or only with consent. **Methods:** Two facilitated workshops for consumer and community members of groups representing people with birth defects, cerebral palsy and disability, and the Western Australian Health Consumers' Council. **Results:** Parent groups and the Health Consumers' Council were unanimous in their support for statutory notification, with 3 conditions: that comprehensive and open information be provided to consumer groups and community; that consumers have input into the development of statutory notification; and that an opt-out clause be included. A Consumer Reference Group was established. They decided on a name for the new register (Western Australian Register of Developmental Anomalies), developed an opt-out clause and reviewed drafts of the regulations for statutory notification. The regulations came into effect in January 2011. **Conclusions:** Consumers were key to achieving statutory notification. We encourage others to engage with their consumers and community in equal partnership for mutual benefit.

[PMID: 23778692](#) [PubMed - in process]

### 19. J Paediatr Child Health. 2013 Jun 11. doi: 10.1111/jpc.12279. [Epub ahead of print]

#### **Genetic and clinical contributions to cerebral palsy: A multi-variable analysis.**

O'Callaghan ME, Maclennan AH, Gibson CS, McMichael GL, Haan EA, Broadbent JL, Baghurst PA, Goldwater PN, Dekker GA; Australian Collaborative Cerebral Palsy Research Group.

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**AIM:** This study aims to examine single nucleotide polymorphism (SNP) associations with cerebral palsy in a multi-variable analysis adjusting for potential clinical confounders and to assess SNP-SNP and SNP-maternal infection interactions as contributors to cerebral palsy. **METHODS:** A case control study including 587 children with cerebral palsy and 1154 control children without cerebral palsy. Thirty-nine candidate SNPs were genotyped in both mother and child. Data linkage to perinatal notes and cerebral palsy registers was performed with a supplementary maternal pregnancy questionnaire. History of known maternal infection during pregnancy was extracted from perinatal databases. **RESULTS:** Both maternal and fetal carriage of inducible nitric oxide synthase SNP rs1137933 were significantly negatively associated with cerebral palsy in infants born at less than 32 weeks gestation after adjustment for potential clinical confounders and correction for multiple testing (odds ratio 0.55, 95% confidence interval 0.38-0.79; odds ratio 0.57, 95% confidence interval 0.4-0.82, respectively). Analysis did not show any statistically significant SNP-SNP or SNP-maternal infection interactions after correction for multiple testing. **CONCLUSIONS:** Maternal and child inducible nitric oxide synthase SNPs are associated with reduced risk of cerebral palsy in infants born very preterm. There was no evidence for statistically significant SNP-SNP or SNP-maternal infection interactions as modulators of cerebral palsy risk.

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Australasian College of Physicians).

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**20. Acta Paediatr. 2013 Jun 17. doi: 10.1111/apa.12320. [Epub ahead of print]**

**Movement Outcomes Of Infants Born Moderate And Late Preterm.**

Odd DE, Lingam R, Emond A, Whitelaw A.

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**AIM:** To investigate if children born between 32-36 weeks gestation have an increased risk of motor coordination difficulties or cerebral palsy (CP) at age 7 years. **METHODS:** A cohort study based on the Avon Longitudinal Study of Parents and Children (ALSPAC). The primary outcomes were poor motor coordination, defined as an ALSPAC coordination test score <5th centile, or the presence of CP. Exposure groups were defined as moderate or later preterm (32-36 weeks gestation) or term (37-42 weeks). Regression models were used to investigate the association between gestational age and outcomes. Multiple imputation was used to account for missing covariate data. **RESULTS:** In the fully adjusted model there was strong evidence that children born at moderate or late preterm had worse coordination (OR 1.41 (1.14-1.74)) and higher risk of CP (OR 6.38 (2.28-17.76)) than term peers. However restricting the analysis to well grown infants born vaginally, in good condition, the associations attenuated substantially. **CONCLUSIONS:** Moderate or late preterm infants were at increased risk of developing coordination problems and cerebral palsy. After restricting the analysis to 'well' infants the associations of gestation with the coordination measures and CP reduced substantially, suggesting antenatal, intrapartum and neonatal causal pathways are likely to be involved. This article is protected by copyright. All rights reserved.

[PMID: 23772915](#) [PubMed - as supplied by publisher]

**21. Am J Obstet Gynecol. 2013 Jun 12. pii: S0002-9378(13)00615-7. doi: 10.1016/j.ajog.2013.06.007. [Epub ahead of print]**

**The association of cerebral palsy and death with small-for-gestational age birth weight in preterm neonates by individualized and population-based percentiles.**

Grobman WA, Lai Y, Rouse DJ, Spong CY, Varner MW, Mercer BM, Leveno KJ, Iams JD, Wapner RJ, Sorokin Y, Thorp JM Jr, Ramin SM, Malone FD, O'sullivan MJ, Hankins GD, Caritis SN.

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**OBJECTIVE:** To determine whether an individualized growth standard (IS) improves identification of preterm small-for-gestational-age (SGA) neonates at risk of developing moderate/severe cerebral palsy (CP) or death. **STUDY DESIGN:** Secondary analysis of data from a randomized trial of MgSO<sub>4</sub> for prevention of CP or death among anticipated preterm births. Singleton non-anomalous liveborns delivered before 34 weeks' were classified as SGA (< 10th % for their GA) by a population standard (PS) or an IS (incorporating maternal age, height, weight, parity, race/ethnicity, and neonatal gender). The primary outcome was prediction of moderate or severe CP or death by age 2. **RESULTS:** Of 1588 eligible newborns, 143 (9.4%) experienced CP (N=33) or death (N=110). Forty-four (2.8%) were SGA by the PS and 364 (22.9%) by the IS. All PS-SGA newborns also were identified as IS-SGA. SGA newborns by either standard had a similarly increased risk of CP or death (PS: RR 2.4, 95% CI 1.3-4.3 vs. IS: RR 1.8, 95% CI 1.3-2.5, respectively). The similarity of RRs remained after stratification by MgSO<sub>4</sub> treatment group. The IS was more sensitive (36% vs. 6%, p <.001), but less specific (78% vs. 98%, p <.001) for CP or death. ROC curve analysis revealed a statistically lower AUC for the PS, although the ability of either method to predict which neonates would subsequently develop CP or death was poor (PS: 0.55, 95% CI 0.49-0.60 vs. IS: 0.59, 95% CI 0.54-0.64, p<.001). **CONCLUSION:** An individualized SGA growth standard does not improve the association with, or prediction of, CP or death by age 2.

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**22. BMJ. 2013 Apr 30;346:f2799. doi: 10.1136/bmj.f2799.**

**Encouraging outcomes for Sweden's extremely preterm babies [No authors listed]**

[PMID: 23637140](#) [PubMed - indexed for MEDLINE]

**23. Indian J Endocrinol Metab. 2013 Jan;17(1):19-30. doi: 10.4103/2230-8210.107822.**

**The molecular mechanisms, diagnosis and management of congenital hyperinsulinism.**

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Congenital hyperinsulinism (CHI) is the result of unregulated insulin secretion from the pancreatic  $\beta$ -cells leading to severe hypoglycaemia. In these patients it is important to make an accurate diagnosis and initiate the appropriate management so as to avoid hypoglycemic episodes and prevent the potentially associated complications like epilepsy, neurological impairment and cerebral palsy. At a genetic level abnormalities in eight different genes (ABCC8, KCNJ11, GLUD1, GCK, HADH, SLC16A1, HNF4A and UCP2) have been reported with CHI. Loss of function mutations in ABCC8/KCNJ11 lead to the most severe forms of CHI which are usually medically unresponsive. At a histological level there are two major subgroups, diffuse and focal, each with a different genetic etiology. The focal form is sporadic in inheritance and is localized to a small region of the pancreas whereas the diffuse form is inherited in an autosomal recessive (or dominant) manner. Imaging using a specialized positron emission tomography scan with the isotope fluoroine-18 L-3, 4-dihydroxyphenylalanine (18F-DOPA-PET-CT) is used to accurately locate the focal lesion pre-operatively and if removed can cure the patient from hypoglycemia. Understanding the molecular mechanisms, the histological basis, improvements in imaging modalities and surgical techniques have all improved the management of patients with CHI.

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**24. J Nucl Med. 2013 Jun 19. [Epub ahead of print]**

**Increased GABA-A Receptor Binding and Reduced Connectivity at the Motor Cortex in Children with Hemiplegic Cerebral Palsy: A Multimodal Investigation Using 18F-Fluoroflumazenil PET, Immunohistochemistry, and MR Imaging.**

Park HJ, Kim CH, Park ES, Park B, Oh SR, Oh MK, Park CI, Lee JD.

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$\gamma$ -aminobutyric acid (GABA)-A receptor-mediated neural transmission is important to promote practice-dependent plasticity after brain injury. This study investigated alterations in GABA-A receptor binding and functional and anatomic connectivity within the motor cortex in children with cerebral palsy (CP). METHODS: We conducted 18F-fluoroflumazenil PET on children with hemiplegic CP to investigate whether in vivo GABA-A receptor binding is altered in the ipsilateral or contralateral hemisphere of the lesion site. To evaluate changes in the GABA-A receptor subunit after prenatal brain injury, we performed GABA-A receptor immunohistochemistry using rat pups with a diffuse hypoxic ischemic insult. We also performed diffusion tensor MR imaging and resting-state functional MR imaging on the same children with hemiplegic CP to investigate alterations in anatomic and functional connectivity at the motor cortex with increased GABA-A receptor binding. RESULTS: In children with hemiplegic CP, the 18F-fluoroflumazenil binding potential was increased within the ipsilateral motor cortex. GABA-A receptors with the  $\alpha 1$  subunit were highly expressed exclusively within cortical layers III, IV, and VI of the motor cortex in rat pups. The motor cortex with increased GABA-A receptor binding in children with hemiplegic CP had reduced thalamocortical and corticocortical connectivity, which might be linked to increased GABA-A receptor distribution in cortical layers in

rats. CONCLUSION: Increased expression of the GABA-A receptor  $\alpha 1$  subunit within the ipsilateral motor cortex may be an important adaptive mechanism after prenatal brain injury in children with CP but may be associated with improper functional connectivity after birth and have adverse effects on the development of motor plasticity.

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## 25. J Trop Pediatr. 2013 Jun 19. [Epub ahead of print]

### Changes in the Clinical Spectrum of Cerebral Palsy over Two Decades in North India--An Analysis of 1212 Cases.

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Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh-160012, India.

Background: Clinical spectrum of cerebral palsy (CP) is different in developing and developed countries. We evaluated the clinical profile, etiological factors and co-morbidities of children with CP in North India, and compared with our previous study. Methods: 1212 children with CP registered in last 10 years in our rehabilitation center were compared with our previous study of 1000 children from same center. Results: Spastic quadriplegia is the commonest type of CP (51.5%) although lesser than previous decade (61%). Birth asphyxia remains the main (51.98%) etiological factor as earlier (45.3%). CP due to CNS infections decreased from 63.5% to 57.4%, due to bilirubin-encephalopathy remained same (~30%). Speech problems (83.7%), microcephaly (64.27%), seizures (44.5%) and intellectual disability (38.61%) are common co-morbidities. Common neuroimaging findings include hypoxic-ischemic changes and periventricular leucomalacia. Conclusion: The spectrum of CP is evolving in the developing countries with an increase in diplegic and a decrease in quadriplegic CP.

[PMID: 23783583](#) [PubMed - as supplied by publisher]

## 26. Ugeskr Laeger. 2013 Jun 17;175(25):1784-1785.

### Brain damage in a child after outpatient induction of labour [Article in Danish]

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A 32-year-old woman had labour induced (25 + 25 microgram misoprostol vaginally) at 291 days gestation for post-term pregnancy. A cardiotocography (CTG) showed normal conditions. The woman went home to await regular contractions in line with hospital policy. She awoke at 3.15 a.m. with abdominal pain and came to the labour ward at 3.45 a.m. with less pain but a desire to push. The cervix was dilated but the foetal head sat high in the pelvis. Internal CTG showed a pre-terminal pattern. A girl was born at 4.27 a.m. She had hypoxic-ischaemic encephalopathy (APGAR at 1, 5 and 10 min.: 1, 3, 3; umbilical cord arterial blood pH 6.71, base excess -19.9 mmol/l). At the age of three years she had severe cerebral palsy.

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