


Playing a musical instrument demands the integration of sensory and perceptual information with motor processes in order to produce a harmonic musical piece. The diversity of brain mechanisms involved and the joyful character of playing an instrument make musical instrument training a potential vehicle for neurorehabilitation of motor skills in patients with cerebral palsy (CP). This clinical condition is characterized by motor impairments that can affect, among others, manual function, and limit severely the execution of basic daily activities. In this study, adolescents and adult patients with CP, as well as a group of typically developing children learned to play piano for 4 consecutive weeks, having completed a total of 8 hours of training. For ten of the participants, learning was supported by a special technical system aimed at helping people with sensorimotor deficits to better discriminate fingers and orient themselves along the piano keyboard. Potential effects of piano training were assessed with tests of finger tapping at the piano and tests of perception of vibratory stimulation of fingers, and by measuring neuronal correlates of motor learning in the absence of and after piano training. Results were highly variable especially among participants with CP. Nevertheless, a significant effect of training on the ability to perceive the localization of vibrations over fingers was found. No effects of training on the performance of simple finger tapping sequences at the piano or on motor-associated brain responses were registered. Longer periods of training are likely required to produce detectable changes.

PMID: 29123403

2. Understanding the relationship between brain and upper limb function in children with unilateral motor impairments: A multimodal approach.


Atypical brain development and early brain injury have profound and long lasting impact on the development, skill acquisition, and subsequent independence of a child. Heterogeneity is present at the brain level and at the motor level; particularly with respect to phenomena of bilateral activation and mirrored movements (MMs). In this multiple case study we consider the feasibility of using several modalities to explore the relationship between brain structure and/or activity and hand function: Electroencephalography (EEG), both structural and functional Magnetic Resonance Imaging (sMRI, fMRI), diffusion tensor imaging (DTI), transcranial magnetic stimulation (TMS), Electromyography (EMG) and hand function.
assessments. METHODS: 15 children with unilateral CP (ages: 9.4 ± 2.5 years) undertook hand function assessments and at least two additional neuroimaging and/or neurophysiological procedures: MRI/DTI/ fMRI (n = 13), TMS (n = 11), and/or EEG/EMG (n = 8). During the fMRI scans and EEG measurements, a motor task was performed to study cortical motor control activity during simple hand movements. DTI tractography analysis was used to study the corpus-callosum (CC) and cortico-spinal tracts (CST). TMS was used to study cortico-spinal connectivity pattern. RESULTS: Type and range of severity of brain injury was evident across all levels of manual ability with the highest radiological scores corresponded to children poorer manual ability. Evidence of MMs was found in 7 children, mostly detected when moving the affected hand, and not necessarily corresponding to bilateral brain activation. When moving the affected hand, bilateral brain activation was seen in 6/11 children while 3/11 demonstrated unilateral activation in the contralateral hemisphere, and one child demonstrated motor activation predominantly in the supplementary motor area (SMA). TMS revealed three types of connectivity patterns from the cortex to the affected hand: a contralateral (n = 3), an ipsilateral (n = 4) and a mixed (n = 1) connectivity pattern; again without clear association with MMs. No differences were found between children with and without MMs in lesion scores, motor fMRI laterality indices, CST diffusivity values, and upper limb function. In the genu, midbody, and splenium of the CC, higher fractional anisotropy values were found in children with MMs compared to children without MMs. The EEG data indicated a stronger mu-restoration above the contralateral hemisphere in 6/8 children and above the ipsilateral hemisphere in 2/8 children.

CONCLUSION: The current results demonstrate benefits from the use of different modalities when studying upper-limb function in children with CP; not least to accommodate to the variations in tolerance and feasibility of implementation of the differing methods. These exposed multiple individual brain-reorganization patterns corresponding to different functional motor abilities. Additional research is warranted to understand the transactional influences of early brain injury, neuroplasticity and developmental and environmental factors on hand function in order to develop targeted interventions.

PMID: 29111113


D’Aquino D, Moussa AA, Ammar A, Ingale H, Vloeberghs M.


BACKGROUND: Selective dorsal rhizotomy (SDR) has been established as an effective surgical treatment for spastic diplegia. The applicability of SDR to the full spectrum of spastic cerebral palsy and the durability of its therapeutic effects remain under investigation. There are currently limited data in the literature regarding efficacy and outcomes following SDR in Gross Motor Function Classification System (GMFCS) IV and V patients. Intrathecal baclofen has traditionally been the surgical treatment of choice for these patients. When utilised primarily as a treatment for the relief of spasticity, it is proposed that SDR represents a rational and effective treatment option for this patient group. We report our outcomes of SDR performed on children with severe cerebral palsy (GMFCS grade IV and V). The commensurate improvement in upper as well as lower limb spasticity is highlighted. Apparent benefit to urological function following SDR in this patient group is also discussed. METHOD: A retrospective review of prospectively collected data for 54 paediatric patients with severe cerebral palsy (GMFCS IV-V) who received SDR plus specialised physiotherapy. Mean age was 10.2 years (range, 3.0-19.5). SDR guided by electrophysiological monitoring was performed by a single experienced neurosurgeon. All subjects received equivalent physiotherapy. The primary outcome measure was change to the degree of spasticity following SDR. Spasticity of upper and lower limb muscle groups were quantified and standardised using the Ashworth score. Measures were collected at baseline and at 2-, 8- and 14-month postoperative intervals. In addition, baseline and 6-month postoperative urological function was also evaluated as a secondary outcome measure. RESULTS: The mean lower limb Ashworth score at baseline was 3.2 (range, 0–4). Following SDR, significant reduction in lower limb spasticity scores was observed at 2 months and maintained at 8 and 14 months postoperatively (Wilcoxon rank, p < 0.001). The mean reduction at 2, 8 and 14 months was 3.0, 3.2 and 3.2 points respectively (range, 1–4), confirming a sustained improvement of spasticity over a 1-year period of follow-up. Significant reduction in upper limb spasticity scores following SDR was also observed (mean, 2.9; Wilcoxon rank, p < 0.001). Overall, the improvement to upper and lower limb tone following SDR—generally to post-treatment Ashworth scores of 0— was clinically and statistically significant in GMFCS IV and V patients. Urological assessment identified pre-existing bladder dysfunction in 70% and 90% of GMFCS IV and V patients respectively. Following SDR, improvement in urinary continence was observed in 71% of affected GMFCS IV and 42.8% of GMFCS V patients. No serious postoperative complications were identified. CONCLUSIONS: We conclude that SDR is safe and—in combination with physiotherapy—effectively reduces spasticity in GMFCS grade IV and V patients. Our series suggests that spastic quadriplegia is effectively managed with significant improvements in upper limb spasticity that are commensurate with those observed in lower limb muscle groups. These gains are furthermore sustained more than a year postoperatively. In light of these findings, we propose that SDR constitutes an effective treatment option for GMFCS IV and V patients and a rational alternative to intrathecal baclofen.

PMID: 29116382


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STUDY DESIGN: Grand Round case report. OBJECTIVE: We report a pancreatic fracture associated with Wirsung duct disruption, following a scoliosis surgery in a cerebral palsy adolescent. Spinal fusion surgery is the standard treatment for severe neuromuscular scoliosis. Many complications such as digestive ones account for its complexity. Postoperative acute pancreatitis is well described, although its pathophysiology remains unclear. To our knowledge, pancreatic fracture following scoliosis correction has never been described to date. Clinical presentation is not specific, and management is not consensual.

CASE REPORT: A 14-year-old adolescent had posterior spinal fusion for neuromuscular scoliosis due to cerebral palsy. During the postoperative course, she developed progressive nonspecific abdominal symptoms. The abdominal CT scan demonstrated a pancreatic fracture and a surgical exploration was decided as perforations of the bowel were highly suspected. Drains were placed around the pancreatic area as the retrogastric region was out of reach due to local inflammation. Conservative management led to the occurrence of a pseudocyst in the following weeks as the pancreatic leakage progressively dropped.

DISCUSSION: Two hypotheses have been proposed: direct iatrogenic trauma from lumbar pedicle screws and pancreatic rupture related to the correction of the spinal deformity. As the latter seems the most likely, spinal surgeons should be aware of this occurrence following severe scoliosis correction. CONCLUSION: Spinal fusion for severe neuromuscular scoliosis is a difficult procedure, with a high rate of complications. Among them, pancreatic fracture should be considered when abdominal pain persists in the postoperative period. Conservative management is advocated especially in case of a poor general condition.

PMID: 29101470


Kiernan D, O'Sullivan R, Malone A, O'Brien T, Simms CK.


BACKGROUND: Increased loading at the lumbar spine, particularly in the coronal plane, has been reported in children with cerebral palsy (CP). As pelvic and trunk movements associated with Trendelenburg and Duchenne type gait are most significant in the coronal plane, the potential exists for lower lumbar spinal loading to be negatively affected in children with CP and these types of movement patterns. OBJECTIVE: The objective of this study was to assess trunk and pelvic kinematics and lower lumbar spinal loading patterns in children with CP and Trendelenburg and Duchenne type gait. DESIGN: This was a cross-sectional study. METHODS: Three-dimensional kinematic (lower limb and thorax) and L5-S1 kinetic data were recorded. Children were divided according to clinical presentation of Trendelenburg or Duchenne type gait. Several discrete kinematic and kinetic parameters were assessed between groups. RESULTS: Three distinct pelvic and trunk movement patterns were identified for children with CP: Trendelenburg, Duchenne, and complex Trendelenburg-Duchenne. Peak L5-S1 lateral bending moments were increased by 62% in children with CP and Duchenne type gait. Children with CP and complex Trendelenburg-Duchenne gait demonstrated the largest deviations from normal, with increased peak ipsilateral and contralateral directed moments of 69% and 54%, respectively, compared to children with typical development.

LIMITATIONS: A test-retest reliability analysis or measure of minimal detectable change was not conducted as part of this study. Results suggest that measures of minimal detectable change may be high for some of the reported variables. In addition, the inverse dynamic approach determines only the net intersegmental reactive forces that reflect the effect of external loads. Previous studies have shown that spinal loads may be larger than the net intersegmental force. CONCLUSIONS: Trendelenburg and Duchenne type movements were not always distinct, and a third type of movement, a combination of the two, was the most common in this study. Clinicians should be aware that children with CP and the Duchenne type or the complex Trendelenburg-Duchenne type of gait pattern experience abnormal loading that may have significant implications for the lower spine in the long term.

PMID: 29106655

Khamis S, Herman T, Krimus S, Danino B.


BACKGROUND: Functional electrical stimulation (FES) is a well-known intervention used during walking to improve walking abilities and correct gait deviations by facilitating the proper muscle group at the appropriate timing in the gait cycle. Our aim was to study the types of surface FES currently used in a cerebral palsy (CP) population and examine the evidence of its ability to improve gait deviations, functional ability and therapeutic effects. METHODS: A computerized database search was conducted from inception until 6/2016. Included were all clinical trials performing gait analysis of children with CP applying surface FES to any lower leg muscles evaluating the efficiency of the stimulation and any carry-over effect. RESULTS: Fifteen studies met the inclusion criteria. The most common FES stimulated the dorsi flexors muscles with a positive orthotic effect, improved dorsi flexion during the swing phase and enhanced the foot contact pattern. A smaller positive effect was found for knee extensors stimulation facilitating knee extension during the stance phase and for hip abductors stimulation improving frontal plane knee alignment. No evidence was found to support the use of plantar flexors stimulation in correcting gait deviations. There is scarce evidence of any retention effect. CONCLUSION: We encourage the clinician to evaluate the use of FES on a case to case basis. Controlled investigations with larger numbers of participants are warranted to determine the orthotic and therapeutic efficacy of FES.

PMID: 29102346

7. Should the Gross Motor Function Classification System be used for children who do not have cerebral palsy?

Towns M, Rosenbaum P, Palisano R, Wright FV.


This literature review addressed four questions. (1) In which populations other than cerebral palsy (CP) has the Gross Motor Function Classification System (GMFCS) been applied? (2) In what types of study, and why was it used? (3) How was it modified to facilitate these applications? (4) What justifications and evidence of psychometric adequacy were used to support its application? A search of PubMed, MEDLINE, and Embase databases (January 1997 to April 2017) using the terms: 'GMFCS' OR 'Gross Motor Function Classification System' yielded 2499 articles. 118 met inclusion criteria and reported children/adults with 133 health conditions/clinical descriptions other than CP. Three broad GMFCS applications were observed: as a categorization tool, independent variable, or outcome measure. While the GMFCS is widely used for children with health conditions/clinical description other than CP, researchers rarely provided adequate justification for these uses. We offer recommendations for development/validation of other condition-specific classification systems and discuss the potential need for a generic gross motor function classification system.

PMID: 29105760

8. Effects of scalp electroacupuncture on the PI3K/Akt signalling pathway and apoptosis of hippocampal neurons in a rat model of cerebral palsy.


Background Substantial evidence from clinical reports has established that most cerebral palsy (CP) patients benefit from a comprehensive rehabilitation exercise training programme. Such advances are enhanced when scalp electroacupuncture (EA), applied at a location corresponding to the projection of the motor area, is combined with rehabilitation exercise training. However, little information exists regarding the mechanistic basis for these effects. Objective To examine whether EA stimulation within the scalp projection location of the motor area can inhibit apoptosis of hippocampal neurons by regulating the PI3K/Akt signalling pathway in a rat model of CP. Methods Fifty male Sprague-Dawley rats underwent surgical modelling of CP. Five were used to confirm successful establishment of the model and the remaining 45 rats were randomly divided into one of three groups that remained untreated (CP group, n=15) or received EA treatment alone (CP+EA group, n=15) or EA in combination with a PI3K/Akt inhibitor (CP+EA+LY294002 group, n=15). An otherwise healthy negative control group of rats undergoing sham surgery was also included (Control group, n=15). In the CP+EA and CP+EA+LY294002 groups, EA was
applied to the scalp surface at allocation corresponding to the projection of the motor area. Basso, Beattie and Bresnahan (BBB) locomotor scores, hippocampal protein expression of Akt and p-Akt (by Western blot analysis) and neuronal apoptosis in hippocampal tissue (by histopathology) were assessed at 7, 14 and 21 days post-CP induction. Results CP rats receiving scalp EA treatment demonstrated improved behavioural scores, less hippocampal neuronal apoptosis and higher expression levels of Akt and p-Akt (p<0.05) at all time points studied compared with untreated CP rats. There were no significant differences observed between CP+EA+LY294002 and untreated CP model groups. Conclusions The effects of scalp EA on the PI3K/ Akt signalling pathway may represent one of the mechanisms involved in the inhibition of hippocampal neuronal apoptosis and improvement of deficits associated with CP in a rat model.

PMID: 29102966


BACKGROUND: Lower-limb alignment in children is classically assessed clinically or based on conventional radiography, which is associated with projection bias. Low-dose biplanar radiography was described recently as an alternative to conventional imaging. The primary objective of this study was to assess the reliability of length and angle values inferred from 3D reconstructions in children seen in everyday practice. The secondary objective was to obtain reference values for goniometry parameters in children. HYPOTHESIS: 3D reconstructions can be used to assess the lower limbs in children. MATERIAL AND METHODS: The paediatric reliability study was done in 18 volunteers who were divided into three groups based on whether they were typically developing (TD) children, had skeletal development abnormalities, or had cerebral palsy. The reference data were obtained in 129 TD children. Each study participant underwent biplanar radiography with 3D reconstruction performed by experts and radiology technicians. Goniometry parameters were computed automatically. Reproducibility was assessed based on the intra-class coefficient (ICC) and the ISO 5725 standard (standard deviation of reproducibility, SDR). RESULTS: For length parameters, the ICCs ranged from 0.94 to 1.00 and the SDR from 2.1 to 3.5mm. For angle parameters, the ICC and SDR ranges were 0.60-0.95 and 0.9°-4.6°, respectively. No significant differences were found across experts or radiology technicians. Age-specific reference data are reported. DISCUSSION: These findings confirm the reliability of low-dose biplanar radiography for assessing lower-limb parameters in children seen in clinical practice. In addition, the study provides reference data for commonly measured parameters.

PMID: 29122688


BACKGROUND: Cerebral palsy (CP) is a disorder of motor function often accompanied by cognitive impairment. There is a paucity of research focused on cognition in dyskinetic CP and on the potential effect of related factors. AIM: To describe the cognitive profile in dyskinetic CP and to assess its relationship with motor function and associated impairments. METHOD: Fifty-two subjects with dyskinetic CP (28 males, mean age 24 y 10 mo, SD 13 y) and 52 typically-developing controls (age- and gender-matched) completed a comprehensive neuropsychological assessment. Gross Motor Function Classification System (GMFCS), Communication Function Classification System (CFCS) and epilepsy were recorded. Cognitive performance was compared between control and CP groups, also according different levels of GMFCS. The relationship between cognition, CFCS and epilepsy was examined through partial correlation coefficients, controlling for GMFCS. RESULTS: Dyskinetic CP participants performed worse than controls on all cognitive functions except for verbal memory. Milder cases (GMFCS I) only showed impairment in attention, visuo-perception and visual memory. Participants with GMFCS II–III also showed impairment in language-related functions. Severe cases (GMFCS IV-V) showed impairment in intelligence and all specific cognitive functions but verbal memory. CFCS was associated with performance in receptive language functions. Epilepsy was related to performance in intelligence, visuospatial abilities, visual memory, grammar comprehension and learning. CONCLUSION: Cognitive performance in dyskinetic CP varies with the different levels of motor impairment, with more cognitive functions impaired as motor severity increases. This study also demonstrates the relationship between communication and epilepsy and cognitive functioning, even controlling for the effect of motor severity.

PMID: 29108712

Harvey A, Reddihough D, Scheinberg A, Williams K.


AIM: To examine current clinical practice for prescribing medications for children with dystonic cerebral palsy (CP) by medical doctors working at a tertiary care centre. METHODS: Rehabilitation and developmental paediatric specialists completed: (i) a custom-designed online cross-sectional survey capturing their usual prescribing patterns; and (ii) one-page questionnaires detailing medication prescription for each child with CP who they started on a new medication for dystonia over a 12-month period. RESULTS: Eleven participating doctors ranged in experience in managing children with CP from less than 5 years to more than 20 years. The cross-sectional survey showed that most doctors used more than one medication, with six making choices taking into account four or more different medications. Oral baclofen was used by all doctors and was the first choice of 10 of 11. Prospective surveys from 57 children showed that medication was prescribed mainly for children aged 3-10 years (n = 35/57), classified within Gross Motor Function Classification System levels IV and V (n = 40/57) and with a mixed movement disorder (n = 38/57). Gabapentin and baclofen were the most frequently prescribed (n = 21/57 and 19/57, respectively), with other drugs used less frequently. Dosage regimens varied between and within doctors, particularly for the use of gabapentin and diazepam. CONCLUSIONS: Oral medication prescribing practices varied among doctors managing children with dystonic CP at a tertiary care hospital, particularly with respect to dosing. There is a need for clinical guidelines for medication prescription to be developed based on best evidence and consensus by clinical experts.

PMID: 29105865

12. Multiple Nutritional Deficiencies in Cerebral Palsy Compounding Physical and Functional Impairments.

Hariprasad PG, Elizabeth KE, Valamparampil MJ, Kalpana D, Anish TS.


INTRODUCTION: Cerebral palsy (CP) refers to a spectrum of disorders causing physical and intellectual morbidity. Macro and micro nutrient deficiencies often contribute to the subnormal physical and mental capabilities of them. OBJECTIVES: To assess the growth, nutritional status, physical and functional ability and quality of life in cerebral palsy children and to determine any relation with their gross motor and functional capabilities. METHOD: The study was conducted at a Tertiary Care Centre, with the participants in the age group 1-16 years. A pretested evaluation tool was prepared which included Anthropometric measurements, tests for hemoglobin and Vitamin D estimation, evidence of micronutrient deficiencies, Dietary patterns, Epidemiological factors, Functional assessment using GMFM (Gross Motor Function Measure ) and FIM (Functional Independent Measurement) scales and Quality of life (QOL) assessment. The data was statistically analyzed. RESULTS: Out of the 41 children, 30 had quadriplegia, 3 had hemiplegia and 8 had spastic diplegia. 34 (82.9%) were severely underweight, 35 (85.4%) had severe stunting and 38 (92.7%) had severe wasting. Micronutrient deficiencies were noted like vitamin B complex deficiency in 37 (90.2%), vitamin A deficiency in 31 (75.6%), low vitamin D levels in 27 (65.9%) and insufficient levels in 9 (22%), severe anemia in 5 (12.2%) and moderate anemia in 26 (63.4%). The gross motor and functional scores were suboptimum in the majority of patients and the care givers had significant impairment in the quality of life. CONCLUSION: Majority of children with cerebral palsy had multiple nutritional deficiencies, gross motor and functional disabilities. QOL of the children and their care givers were suboptimum. A comprehensive package that address dietary intake, correction of micronutrient deficiencies especially anemia and vitamin D deficiency, physical and emotional support is recommended for the wellbeing of the affected children.

PMID: 29123343


Mutch AM.


INTRODUCTION: This article presents an additional case of concomitant topiramate and phenobarbital administration that resulted in 8 hospital admissions for hypothermia that resolved after discontinuation of phenobarbital. CASE: A 56-year-old white female with cerebral palsy and quadriplegia, epilepsy, and hypothyroidism was admitted to a community teaching hospital multiple times with documented hypothermia. These admissions followed a subsequent dose increase of topiramate in December 2014. In February 2015, the patient was admitted with 35°C rectal temperature. Her 2 admissions in April were for
hypothermia with temperatures of 34.6°C and 33.6°C, respectively. The patient had 5 other admissions with hypothermia through December 2015. All other causes of hypothermia were ruled out. The hypothermia resolved when phenobarbital was discontinued. DISCUSSION: A recent case series noted an association between phenobarbital and topiramate causing hypothermia. The patient's hypothermia developed while on concomitant phenobarbital and topiramate but only after an increase in topiramate. No other causes for hypothermia were found based upon physical examination or lab work. The Naranjo nomogram noted a probable causation. CONCLUSION: This case report points to an association of hypothermia with concomitant topiramate and phenobarbital with resolution after phenobarbital discontinuation. Improvement after discontinuation of phenobarbital seems to support a drug-effect relationship.

PMID: 29117788

14. Do adolescents with cerebral palsy agree with their caregivers on their participation and quality of life?

Büğüşan S, Kahraman A, Elbasan B, Mutlu A.


BACKGROUND: It is important to determine the quality of life (QoL) and level of participation in children with Cerebral Palsy (CP). Previous research has used reports from adolescents or caregivers, but there is no evidence that caregivers' reports accurately reflect the experiences of the adolescents they are interested in. OBJECTIVE/HYPOTHESIS: The aim of this study was to investigate whether a difference was present in the views of the adolescents and their caregivers regarding the participation and the quality of life of adolescents with CP, and to reveal the parameters creating such differences. METHODS: The participation levels and QoL of the adolescents were evaluated separately by the caregiver and the adolescent using the Pediatric Outcomes Data Collection Instrument (PODCI). RESULTS: A statistically significant difference was found in terms of caregivers and adolescents' scores of PODCI upper extremity (Z = -2.560, p = 0.008), transfer&basic mobility (Z = -3.839, p = 0.000), sports/physical functioning (Z = -3.103, p = 0.002), happiness (Z = -2.420, p = 0.016) and global functioning (Z = -3.639, p = 0.001). The children's scores were statistically significantly higher than caregivers'. It was found that there was a poor consistence in terms of caregivers and adolescents' scores of upper extremity (ICC = 0.373, p = 0.012), transfer/basic mobility (ICC = 0.289, p = 0.016), sport/physical functioning (ICC = 0.359, p = 0.009); moderate consistence in terms of those of global functioning (ICC = 0.421, p = 0.003). CONCLUSION: It was determined that caregivers and children's answers were not compatible with one another especially in terms of subjective assessments such as happiness and pain, which suggests that the consideration of caregivers or children in the assessment of subjective situations will change the results.

PMID: 29100958

15. Contextual Factors and Mastery Motivation in Young Children with and without Cerebral Palsy: A Systematic Review.

Huang HH, Sun TH, Lin CI, Chen YR.


BACKGROUND: Mastery motivation is the driving force behind children's desire to explore the surrounding world and their comprehensive development. However, disease factors may lower a child's motivation and hamper development. The aim of this review is to examine mastery motivation in preschool children with cerebral palsy (CP) and the impact of contextual factors on mastery motivation. METHODS: Six electronic databases were searched (PubMed, ScienceDirect, Scopus, PsycINFO, Medline, and Airiti Library) using the keywords "Activity," "Cerebral Palsy," "Preschool," "Motivation," "Mastery motivation," "Gross motor," and "Toddler." We reviewed six observational studies and one interventional study for the following features: (1) participants' characteristics; (2) assessment, observation, and intervention methods; (3) findings. RESULTS: Of the seven studies, three were individual cohort studies and four were individual case-control studies. There were two types of motivation-related measures, standardized measurements and observations of structured tasks or free play. Three studies showed no significant difference in mastery motivation between children with and those without CP when given mental -age-appropriate tasks of moderate difficulty. However, environmental factors including social experience, family interaction, and caregivers' perceptions may affect motivation in preschool children with CP. CONCLUSION: Current studies on mastery motivation in preschool children with CP are very limited, and the lack of a universal, theory-based definition of mastery motivation and common assessment frameworks makes it difficult to draw clear conclusions on mastery motivation in children with CP. Future studies should investigate mastery motivation with rigorous study designs to identify ideal activities and environments for preschool children with CP.

PMID: 29124053
16. [Psychological Health of Children with Chronic Physical Illness and their Parents - Results from Meta-Analyses].

Pinquart M.


Psychological Health of Children with Chronic Physical Illness and their Parents - Results from Meta-Analyses The present paper summarizes results from meta-analyses on psychological well-being of children with chronic physical illnesses and their parents. At the beginning, we discuss potential reasons for psychological effects of a chronic physical illness on children and adolescents as well as their parents. We then summarize results of meta-analyses of studies that compared aspects of mental health of children with a chronic physical illness and their parents with families of healthy children. Depressive symptoms, anxiety, and internalizing symptoms in general were most elevated in children with chronic fatigue syndrome and chronic headache while externalizing symptoms were most elevated in young people with epilepsy, chronic headache, and cerebral palsy. Depression and anxiety was less elevated in the ill children than in their parents. Parents of children with HIV-infection/AIDS and cerebral palsy reported the highest levels of distress, followed by parents of children diagnosed with cancer and spina bifida. Conclusions are drawn for future research and practice.

PMID: 29111893

Prevention and Cure

17. microRNAs participate in the regulation of oligodendrocytes development in white matter injury.

Xiao D, Qu Y, Pan L, Li X, Mu D.


White matter injury (WMI) often results in cognitive impairment, behavioral disorders, and cerebral palsy and thus imposes a tremendous burden on society. The cells in brain white matter mainly comprise oligodendrocytes (OLs), astrocytes, and microglia. The dysregulation of OLs development is the pathological hallmark of WMI. Recent studies have demonstrated that microRNAs (miRNAs or miRs) participate in the regulation of OLs development, and the dysregulation of this process represents the pathogenesis of WMI. This review summarizes the progress made in this field that will help clinicians and researchers understand the molecular etiology of WMI and develop miRNAs as new agents for the prevention and treatment of WMI.

PMID: 29120862

18. The panorama of cerebral palsy in Sweden part XII shows that patterns changed in the birth years 2007-2010.

Himmelmann K, Uvebrant P.


AIM: This was the 12th population-based study to explore the epidemiology of cerebral palsy (CP) in western Sweden. METHODS: From 2007-2010 there were 104,713 live births in the area. We analysed the birth characteristics, aetiology and neuroimaging findings, calculated the prevalence and compared the results with previous study cohorts. RESULTS: CP was found in 205 children, corresponding to a crude prevalence of 1.96 per 1,000 live births. The gestational age-specific prevalence for < 28 gestational weeks was 59.0 per 1,000 live births, 45.7 for 28-31 weeks, 6.0 for 32-36 weeks and 1.2 for > 36 weeks. Hemiplegia accounted for 44%, diplegia for 34%, tetraplegia for 5%, dyskinetic CP for 12% ataxia for 3%. Neuroimaging showed maldevelopment in 12%, white-matter lesions in 49%, cortical/subcortical lesions in 15% and basal ganglia lesions in 11%. The aetiology was considered prenatal in 38%, peri/neonatal in 38% and remained unclassified in 24%. CP due to term or near-term asphyxia had decreased. CONCLUSION: A non-significant decrease in CP prevalence was seen in term-born children. Hemiplegia was still the most prevalent CP type, while the prevalence of dyskinetic CP had decreased. One in two children had white matter lesions, indicating late second or early third trimester timing. This article is protected by copyright. All rights reserved.

PMID: 29121418
19. Cerebral palsy after in vitro fertilization.

Källén B.


[This commentary is on the original article by Goldsmith et al.]

PMID: 29105749

20. Fibrinogen Activates BMP Signaling in Oligodendrocyte Progenitor Cells and Inhibits Remyelination after Vascular Damage.


Blood-brain barrier (BBB) disruption alters the composition of the brain microenvironment by allowing blood proteins into the CNS. However, whether blood-derived molecules serve as extrinsic inhibitors of remyelination is unknown. Here we show that the coagulation factor fibrinogen activates the bone morphogenetic protein (BMP) signaling pathway in oligodendrocyte progenitor cells (OPCs) and suppresses remyelination. Fibrinogen induces phosphorylation of Smad 1/5/8 and inhibits OPC differentiation into myelinating oligodendrocytes (OLs) while promoting an astrocytic fate in vitro. Fibrinogen effects are rescued by BMP type I receptor inhibition using dorsomorphin homolog 1 (DMH1) or CRISPR/Cas9 activin A receptor type I (ACVR1) knockout in OPCs. Fibrinogen and the BMP target Id2 are increased in demyelinated multiple sclerosis (MS) lesions. Therapeutic depletion of fibrinogen decreases BMP signaling and enhances remyelination in vivo. Targeting fibrinogen may be an upstream therapeutic strategy to promote the regenerative potential of CNS progenitors in diseases with remyelination failure.

PMID: 29103804


Dan B, Paneth N.


Much of what we know about cerebral palsy—including its effect on motor functioning, quality of life, families, and society; the importance of associated disabilities; the effectiveness of management strategies; and even fundamental epidemiological features, such as prevalence and risk factors—is derived from observations made in high-income countries (HICs). Yet, the burden of cerebral palsy is thought by many to be greater in low-income countries (LICs), for which little information is available.

PMID: 29102351


BACKGROUND: Few population-based studies of cerebral palsy have been done in low-income and middle-income countries. We aimed to examine cerebral palsy prevalence and subtypes, functional impairments, and presumed time of injury in children in Uganda. METHODS: In this population-based study, we used a nested, three-stage, cross-sectional method (Iganga-Mayuge Health and Demographic Surveillance System [HDSS]) to screen for cerebral palsy in children aged 2-17 years in a rural
eastern Uganda district. A specialist team confirmed the diagnosis and determined the subtype, motor function (according to the Gross Motor Function Classification System [GMFCS]), and possible time of brain injury for each child. Triangulation and interviews with key village informants were used to identify additional cases of suspected cerebral palsy. We estimated crude and adjusted cerebral palsy prevalence. We did \( \chi^2 \) analyses to examine differences between the group screened at stage 1 and the entire population and regression analyses to investigate associations between the number of cases and age, GMFCS level, subtype, and time of injury. **FINDINGS:** We used data from the March 1, 2015, to June 30, 2015, surveillance round of the Iganga-Mayuge HDSS. 31,756 children were screened for cerebral palsy, which was confirmed in 86 (19%) of 442 children who screened positive in the first screening stage. The crude cerebral palsy prevalence was 2.7 (95% CI 2.2–3.3) per 1000 children, and prevalence increased to 2.9 (2.4–3.6) per 1000 children after adjustment for attrition. The prevalence was lower in older (8–17 years) than in younger (<8 years) children. Triangulation added 11 children to the cohort. Spastic unilateral cerebral palsy was the most common subtype (45 [46%] of 97 children) followed by bilateral cerebral palsy (39 [40%] of 97 children). 14 (27%) of 51 children aged 2–7 years had severe cerebral palsy (GMFCS levels 4–5) compared with only five (12%) of 42 children aged 8–17 years. Few children (two [2%] of 97) diagnosed with cerebral palsy were born preterm. Post-neonatal events were the probable cause of cerebral palsy in 24 (25%) of 97 children. **INTERPRETATION:** Cerebral palsy prevalence was higher in rural Uganda than in high-income countries (HICs), where prevalence is about 1.8–2.3 cases per 1000 children. Children younger than 8 years were more likely to have severe cerebral palsy than older children. Fewer older children than younger children with cerebral palsy suggested a high mortality in severely affected children. The small number of preterm-born children probably resulted from low preterm survival. About five times more children with post-neonatal cerebral palsy in Uganda than in HICs suggested that cerebral malaria and seizures were prevalent risk factors in this population.

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23. [Developmental neurology - networked medicine and new perspectives].


Developmental neurology is one of the major areas of neuropediatrics and is among other things (legally) responsible for monitoring the motor, cognitive and psychosocial development of all infants using standardized monitoring investigations. The special focus is on infants born at risk and/or due to premature birth before 32 weeks of gestation or a birth weight less than 1500 g. Early diagnosis of deviations from normal, age-related development is a prerequisite for early interventions, which may positively influence development and the long-term biopsychosocial prognosis of the patients. This article illustrates the available methods in developmental neurology with a focus on recent developments. Particular attention is paid to the predictive value of general movements (GM). The current development of markerless automated detection of spontaneous movements using conventional depth imaging cameras is demonstrated. Differences in spontaneous movements in infants at the age of 12 weeks are illustrated and discussed exemplified by three patients (healthy versus genetic syndrome versus cerebral palsy).

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