Cerebral Palsy ALLIANCE

NEWS
Cerebral Palsy Research

Monday 30 December 2013

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Interventions and Management

1. Neural Plast. 2013;2013:356275. Epub 2013 Dec 3.

Brain Reorganization following Intervention in Children with Congenital Hemiplegia: A Systematic Review.

Inguaggiato E1, Sgandurra G2, Perazza S3, Guzzetta A2, Cioni G4.

Noninvasive rehabilitation strategies for children with unilateral cerebral palsy are routinely used to improve hand motor function, activity, and participation. Nevertheless, the studies exploring their effects on brain structure and function are very scarce. Recently, structural neuroplasticity was demonstrated in adult poststroke patients, in response to neurorehabilitation. Our purpose is to review current evidence on the effects of noninvasive intervention strategies on brain structure or function, in children with unilateral cerebral palsy. The main literature databases were searched up to October 2013. We included studies where the effects of upper limb training were evaluated at neurofunctional and/or neurostructural levels. Only seven studies met our selection criteria; selected studies were case series, six using the intervention of the constraint-induced movement therapy (CIMT) and one used virtual reality therapy (VR). CIMT and VR seem to produce measurable neuroplastic changes in sensorimotor cortex associated with enhancement of motor skills in the affected limb. However, the level of evidence is limited, due to methodological weaknesses and small sample sizes of available studies. Well-designed and larger experimental studies, in particular RCTs, are needed to strengthen the generalizability of the findings and to better understand the mechanism of intervention-related brain plasticity in children with brain injury.

PMID: 24367726 [PubMed - as supplied by publisher] PMCID: PMC3866714 Free PMC Article

2. Dev Med Child Neurol. 2013 Dec 17. doi: 10.1111/dmcn.12356. [Epub ahead of print]

Mastery motivation in children with congenital hemiplegia: individual and environmental associations.

Miller L, Ziviani J, Ware RS, Boyd RN.

AIM: The aim of this study was to examine the relationship between mastery motivation and individual and environmental characteristics in school-aged children with congenital hemiplegia. METHOD: Forty-eight child-caregiver dyads (children's mean age 7y 11mo, SD 2y 4mo; 33 males, 15 females; Manual Ability Classification System [MACS] level I, n=25, and level II, n=23; predominant motor type spastic hemiplegia, n=47) were recruited to this cross-sectional study. Children were assessed using the Melbourne Assessment of Unilateral Upper Limb Function (MUUL) and the Assisting Hand Assessment. Caregivers completed the Dimensions of Mastery

Questionnaire, the Parenting Scale, and a demographic questionnaire. RESULTS: Consistent and positive parental disciplinary practices were associated with higher total motivation (p=0.006) and instrumental aspect scores (p=0.009). Children with siblings and from single-parent families experienced greater negative reactions to failure (p=0.006). Children from two-parent families (p=0.018) and with better bimanual performance (p=0.015) demonstrated greater object-oriented persistence. Age, sex, limitations in manual ability (MACS), and movement and body function of the impaired limb (MUUL) did not contribute significantly to mastery motivation. INTERPRETATION: Inconsistent, excessively lax, and verbose parenting practices may discourage children from persevering with challenging tasks. Functional parenting styles, positive discipline practices, and autonomy-supportive strategies for task engagement should be encouraged when intervening with children with cerebral palsy. Parents should be supported to engage in these practices in all aspects of daily activities.

PMID: 24341437 [PubMed - as supplied by publisher]

3. Pediatrics. 2013 Dec 23. [Epub ahead of print]

Efficacy of Upper Limb Therapies for Unilateral Cerebral Palsy: A Meta-analysis.

Sakzewski L, Ziviani J, Boyd RN.

BACKGROUND AND OBJECTIVE: Children with unilateral cerebral palsy present with impaired upper limb (UL) function affecting independence, participation, and quality of life and require effective rehabilitation. This study aims to systematically review the efficacy of nonsurgical upper limb therapies for children with unilateral cerebral palsy. METHODS: Medline, CINAHL (Cumulative Index to Nursing and Allied Health Literature), Embase, the Cochrane Central Register of Controlled Trials, and PubMed were searched to December 2012. Randomized controlled or comparison trials were included. RESULTS: Forty-two studies evaluating 113 UL therapy approaches (N = 1454 subjects) met the inclusion criteria. Moderate to strong effects favoring intramuscular injections of botulinum toxin A and occupational therapy (OT) to improve UL and individualized outcomes compared with OT alone were identified. Constraint-induced movement therapy achieved modest to strong treatment effects on improving movement quality and efficiency of the impaired UL compared with usual care. There were weak treatment effects for most outcomes when constraint therapy was compared with an equal dose (amount) of bimanual OT; both yielded similar improved outcomes. Newer interventions such as action observation training and mirror therapy should be viewed as experimental. CONCLUSIONS: There is modest evidence that intensive activity-based, goal-directed interventions (eg, constraint-induced movement therapy, bimanual training) are more effective than standard care in improving UL and individualized outcomes. There is little evidence to support block therapy alone as the dose of intervention is unlikely to be sufficient to lead to sustained changes in UL outcomes. There is strong evidence that goal-directed OT home programs are effective and could supplement hands-on direct therapy to achieve increased dose of intervention.

PMID: 24366991 [PubMed - as supplied by publisher]

4. Pediatr Phys Ther. 2014 Spring;26(1):28-37. doi: 10.1097/PEP.0000000000000008.

Effect of body-scaled information on reaching in children with hemiplegic cerebral palsy: a pilot study.

Huang HH, Ellis TD, Wagenaar RC, Fetters L.

PURPOSE: This study examined body-scaled information that specifies the reach patterns of children with hemiplegic cerebral palsy and children with typical development. METHODS: Nine children with hemiplegic cerebral palsy (3-5 years) and 9 age-matched children with typical development participated in the study. They were required to reach and grasp 10 different pairs of cubes. Reach data were coded as either a 1-handed reach or a 2-handed reach. Dimensionless ratios were calculated by dividing the cube size by the maximal aperture between the index finger and thumb. A critical ratio was used to establish the shift from a 1-handed to an exclusive 2-handed reach. RESULTS: The critical ratio was not significantly different for either preferred or nonpreferred arms within and between groups. All children used an exclusive 2-handed reach at a similar dimensionless ratio. CONCLUSION: Our study provides evidence of the "fit" between environment (cube size) and the individual's capabilities (finger aperture) for reaching for both groups.

PMID: 24356315 [PubMed - in process]

5. Dev Med Child Neurol. 2013 Dec 17. doi: 10.1111/dmcn.12353. [Epub ahead of print]

Is outcome of constraint-induced movement therapy in unilateral cerebral palsy dependent on corticomotor projection pattern and brain lesion characteristics?

Islam M, Nordstrand L, Holmström L, Kits A, Forssberg H, Eliasson AC.

AIM: The aim of the study was to explore individual variations in outcome of hand function after constraint-induced movement therapy (CIMT) in relation to the organization of corticomotor projection and brain lesion characteristics in participants with unilateral cerebral palsy (CP). METHOD: Sixteen participants (eight males, eight females; mean age 13y, [SD 2y] range 10-16y) with unilateral CP (nine right-sided; Manual Ability Classification System [MACS] level I, n=1; level II, n=15) who participated in a 2-week CIMT day camp (63h) were included in the study. Various aspects of hand function were measured by the Jebsen-Taylor Hand Function Test (JTHFT), the Assisting Hand Assessment (AHA), and the Melbourne Assessment, both before and after the day camp. Transcranial magnetic stimulation was used to explore the corticomotor organization, and brain lesion characteristics were described by visual assessment of conventional structural magnetic resonance images. RESULTS: At a group level, the training was associated with significant improvements in JTHFT (p=0.003) and AHA (p=0.046), but not in Melbourne Assessment scores. Improvements were found in all types of corticomotor projection patterns, i.e. contralateral, mixed, and ipsilateral. There was no relationship between functional improvement and brain lesion characteristics. INTERPRETATION: Individuals with CP experience improved motor outcomes after CIMT, independent of corticomotor projection pattern and lesion characteristics.

PMID: 24341408 [PubMed - as supplied by publisher]

6. Dev Med Child Neurol. 2013 Nov;55(11):977-8. doi: 10.1111/dmcn.12229. Epub 2013 Aug 13.

Using the Assisting Hand Assessment and the Mini-AHA for clinical evaluation and further research and development.

Bower E.

Comment on: Development of the Mini-Assisting Hand Assessment: evidence for content and internal scale validity.

[Dev Med Child Neurol. 2013]

PMID: 23937151 [PubMed - indexed for MEDLINE]

7. Int J Rehabil Res. 2013 Dec 21. [Epub ahead of print]

Impaired predictive and reactive control of precision grip in chronic stroke patients.

Dispa D, Thonnard JL, Bleyenheuft Y.

Skilled hand movements require a precise coordination between the grip force and the load force. To coordinate those forces, we rely on both a predictive and a reactive control. On the basis of specific impairments observed previously in children with hemiplegic cerebral palsy, we aimed to assess the predictive or reactive nature of hand deficits in stroke patients. This case-control study was carried out with eight stroke patients and eight control participants. The load of a handheld object was rapidly increased by dropping a mass attached to the object. We tested predictive and reactive aspects of the movement in the same task as the drop was triggered either unexpectedly by the examiner (reactive condition) or by the patient himself (predictive condition). Deficits observed in the paretic hand were similar to those highlighted previously in children with hemiplegic cerebral palsy. Under predictive conditions, temporal deficits were observed after impact. Under reactive conditions, the reflex latency

was slightly increased in the paretic hand. The nonparetic hand showed similar results to controls. The predictive mechanism is present but altered in the paretic hand. These alterations suggest an inability to anticipate the consequences of dynamic perturbations in the paretic hand only.

PMID: 24365783 [PubMed - as supplied by publisher]

8. Phys Occup Ther Pediatr. 2013 Dec 17. [Epub ahead of print]

Efficacy and Acceptability of Reduced Intensity Constraint-Induced Movement Therapy for Children Aged 9 -11 years with Hemiplegic Cerebral Palsy: A Pilot Study.

McConnell K, Johnston L, Kerr C.

Objective: Assess efficacy and acceptability of reduced intensity constraint-induced movement therapy (CIMT) in children with cerebral palsy (CP). Methods: Single-subject research design and semi-structured interviews. Children (9-11y) with hemiplegia underwent five baseline assessments followed by two weeks CIMT. Six further assessments were performed during treatment and follow-up phases. The primary outcome was the Melbourne Assessment of Unilateral Upper Limb Function (MUUL). Quantitative data were analysed using standard single-subject methods and qualitative data by thematic analysis. Results: Four of the seven participants demonstrated statistically significant improvements in MUUL (3-11%, $p < .05$). Two participants achieved significant improvements in active range of motion but strength and tone remained largely unchanged. Qualitative interviews highlighted limitations of the restraint, importance of family involvement, and coordination of treatment with education. Conclusions: Reduced intensity CIMT may be effective for some children in this population; however it is not suitable for all children with hemiplegia.

PMID: 24341455 [PubMed - as supplied by publisher]

9. Pediatr Phys Ther. 2014 Spring;26(1):37. doi: 10.1097/PEP.0000000000000009.

Commentary on "effect of body-scaled information on reaching in children with hemiplegic cerebral palsy: a pilot study".

Charles J, Gordon AM.

PMID: 24356316 [PubMed - in process]

10. Res Dev Disabil. 2013 Dec 12;35(2):393-399. doi: 10.1016/j.ridd.2013.11.017. [Epub ahead of print]

Motion analysis of throwing Boccia balls in children with cerebral palsy.

Huang PC1, Pan PJ2, Ou YC3, Yu YC4, Tsai YS5.

Boccia is a sport suitable for children with cerebral palsy (CP). Throwing Boccia balls requires upper extremity and torso coordination. This study investigated the differences between children with CP and normally developed children regarding throwing patterns of Boccia balls. Thirteen children with bilateral spastic CP and 20 normally developed children participated in this study. The tests in this study were a pediatric reach test and throwing of Boccia balls. A 3D electromagnetic motion tracking system and a force plate were synchronized to record and analyze biomechanical parameters of throwing Boccia balls. The results of the pediatric reach test for participants with CP were significantly worse than those for normally developed participants. The 2 groups of participants did not significantly differ regarding the distance between a thrown Boccia ball and a target ball (jack). Participants with CP demonstrated significantly longer movement duration, smaller amplitude of elbow movement, greater amplitudes of shoulder abduction and flexion, slower maximal velocity of torso flexion and the linear velocity of moving the wrist joint forward, faster maximal velocity of head flexion, and smaller sway ratio compared with normally developed participants when throwing Boccia balls. Participants with CP seemed to mainly use head and shoulder movements to bring the Boccia balls forward with limited torso movement. Normally developed participants brought the Boccia ball forward with faster torso and greater elbow movement while stabilizing head and shoulder movements. Nevertheless, participants with CP did not demonstrate significantly worse performance

in the throwing accuracy of Boccia balls.

PMID: 24334228 [PubMed - as supplied by publisher]

11. Res Dev Disabil. 2013 Dec 23;35(2):498-505. doi: 10.1016/j.ridd.2013.12.001. [Epub ahead of print]

Outpatient physical therapy utilization for children and adolescents with intellectual disabilities in Taiwan: A population-based nationwide study.

Chang YC1, Lin JD2, Tung HJ1, Chiang PH3, Hsu SW4.

This study analyzed the utilization and utilization determinants of outpatient physical therapy (PT) among children and adolescents with intellectual disabilities (ID) in Taiwan. A cross-sectional study was conducted to analyze 2007 national health insurance (NHI) claim data from 35,802 eighteen-year-old and younger persons with intellectual disabilities. A total of 3944 (11.02%) claimants received outpatient physical therapy. Variables that affected PT utilization included age, residence urbanization level, ID level, copayment status and major co-morbidity. The average annual PT visit frequency was 25.4±33.0; pre-school children, claimants suffering from catastrophic disease and ID co-occurring with cerebral palsy had a higher mean cost per visit. Age, ID level, copayment status and co-morbidity were factors that influenced expenditure. Pre-school children, males, individuals who resided in the lowest urbanization areas and individuals with a catastrophic disease tended to use hospital services. The point prevalence of epilepsy and cerebral palsy were 12.10% and 19.80%, respectively. Despite the NHI program and government regulations to provide special services, the use of physical therapy for children and adolescents with intellectual disabilities was low, and the utilization decreased as the subjects aged.

PMID: 24370652 [PubMed - as supplied by publisher]

12. Gait Posture. 2013 Nov 27. pii: S0966-6362(13)00682-6. doi: 10.1016/j.gaitpost.2013.11.010. [Epub ahead of print]

The effects of surgical lengthening of hamstring muscles in children with cerebral palsy - The consequences of pre-operative muscle length measurement.

Laracca E1, Stewart C2, Postans N3, Roberts A3.

Children with cerebral palsy often undergo multiple orthopaedic surgical procedures in a single episode. Evidence of the effectiveness of individual components within the overall package is sparse. The introduction of musculoskeletal modelling in Oswestry has led to a more conservative management approach being taken with hamstring muscles for children walking in a degree of crouch. Muscles which were shown to be of at least normal length at initial contact were not surgically lengthened, as would have been the case previously. A retrospective review of 30 such patients was therefore possible, comparing 15 patients treated before the policy change who had their hamstrings lengthened with 15 treated after who did not. All patients had pre and post operative gait assessments and significant changes were observed for each group separately and for the two groups when compared. The comparison revealed that preserving the hamstrings does tend to reduce, and therefore normalize, the dynamic muscle length. Examination of the two patient groups separately, however, reveals a more complex picture with more global gait improvements seen when the hamstrings were lengthened. No absolute recommendation can be made to inform the clinical management of all children with normal to long hamstring muscles during gait. The final decision of whether to include a hamstring lengthening will need to take into account the characteristics of the individual child.

PMID: 24332744 [PubMed - as supplied by publisher]

13. BMC Musculoskelet Disord. 2013 Dec 23;14(1):365. [Epub ahead of print]

Movement within foot and ankle joint in children with spastic cerebral palsy: a 3-dimensional ultrasound analysis of medial gastrocnemius length with correction for effects of foot deformation.

Huijing PA, Bénard MR, Harlaar J, Jaspers RT, Becher JG.

BACKGROUND: In spastic cerebral palsy (SCP), a limited range of motion of the foot (ROM), limits gait and other activities. Assessment of this limitation of ROM and knowledge of active mechanisms is of crucial importance for clinical treatment. METHODS: For a comparison between spastic cerebral palsy (SCP) children and typically developing children (TD), medial gastrocnemius muscle-tendon complex length was assessed using 3-D ultrasound imaging techniques, while exerting externally standardized moments via a hand-held dynamometer. Exemplary X-ray imaging of ankle and foot was used to confirm possible TD-SCP differences in foot deformation. RESULTS: SCP and TD did not differ in normalized level of excitation (EMG) of muscles studied. For given moments exerted in SCP, foot plate angles were all more towards plantar flexion than in TD. However, foot plate angle proved to be an invalid estimator of talocrural joint angle, since at equal foot plate angles, GM muscle-tendon complex was shorter in SCP (corresponding to an equivalent of 1 cm). A substantial difference remained even after normalizing for individual differences in tibia length. X-ray imaging of ankle and foot of one SCP child and two typically developed adults, confirmed that in SCP that of total footplate angle changes (0-4 Nm: 15[degree sign]), the contribution of foot deformation to changes in foot plate angle (8) were as big as the contribution of dorsal flexion at the talocrural joint (7[degree sign]). In typically developed individuals there were relatively smaller contributions (10 -11%) by foot deformation to changes in foot plate angle, indicating that the contribution of talocrural angle changes was most important. Using a new estimate for position at the talocrural joint (the difference between GM muscle-tendon complex length and tibia length, GM relative length) removed this effect, thus allowing more fair comparison of SCP and TD data. On the basis of analysis of foot plate angle and GM relative length as a function externally applied moments, it is concluded that foot plate angle measurements underestimate angular changes at the talocrural joint when moving in dorsal flexion direction and overestimates them when moving in plantar flexion direction, with concomitant effects on triceps surae lengths. CONCLUSIONS: In SCP children diagnosed with decreased dorsal ROM of the ankle joint, the commonly used measure (i.e. range of foot plate angle), is not a good estimate of rotation at the talocrural joint. since a sizable part of the movement of the foot (or foot plate) derives from internal deformation of the foot.

PMID: 24364826 [PubMed - as supplied by publisher] Free full text

14. Chirurgia (Bucur). 2013 Nov-Dec;108(6):866-73.

Herbert capsuloplasty and burnei tenomyoplasty for the correction of genu flexum in cerebral palsy, arthrogryposis and posttraumatic.

Gavriliu S, Georgescu I, Ulici A, Ghita R, Japie E, Pandea N, Pârvan A, Burnei C, Tiripa I, Martiniuc A, Hamei S, Draghici I.

Introduction: Studies of gait dynamics revealed the complex motions that the knee must undergo in sync with the hip and ankle, in both the swing and support phase of walking. If these motions are restricted, usually as a consequence of cerebral palsy or arthrogryposis, normal gait is hindered; the patient may be able to walk for very short distances or, eventually, not at all. Children with knee extension limited by 10 - 30 degrees, especially those with cerebral palsy, exhibit a stance compatible with walking. Walking is difficult and the gait pattern, â crouch gaitâ, is considered typical for this degree of limitation. Aim: This paper is meant as an update regarding the usefulness of Herbert knee capsuloplasty, conceived in 1938 and introduced in Romania in 1956 by Clement Baciu, and Burneidistal medial hamstring tenomyoplasty, invented in 1993.Materials and methods: Herbert knee capsuloplasty, although initially intended for ailments other than spasticity or arthrogryposis, became known, in time, as a useful operation for spastic genu flexum with a 15 to 30 degree limitation of extension. Severing the posterior cruciate ligament (PCL) in children less than 10 years old often results in genu recurvatumor joint instability. In order to avoid these complications, PCL transection has been phased out and our clinic started touse, preferentially for spastic genu flexum rather than arthrogryposis, the Burnei tenomyoplasty. When applied in the same operative session, the two techniques complement each other and act in synergy. Results: Herbert capsuloplasty can achieve only partialcorrection of genu flexum ranging between 30 and 60 degrees of extension deficit. Full extension is opposed by the PCL, contracture of the hamstrings and vascular retraction. Burneitenomyoplasty used by itself is useful for genu flexum with less than 30 degrees of extension deficit. For children with 30 to 60 degrees of knee

extension deficit, combining the Herbertand Burnei procedures achieves the best results. Conclusions: The simultaneous application of Herbert capsuloplastyand Burnei tenomyoplasty allows for the correction of stiff genu flexum and enables the patient to resume walking, with or without support. This course of treatment also avoids the progression of genu flexum beyond 60 degrees, which would require an osteotomy. This combined procedure avoids the cartilage lesions which may develop when patients with 30- 60 degree genu flexum undergo Herbert capsuloplasty alone. Not in the least, the risk of postoperative knee dislocation is significantly reduced.

PMID: 24331328 [PubMed - in process] Free full text

15. Yonsei Med J. 2014 Jan 1;55(1):191-6. doi: 10.3349/ymj.2014.55.1.191.

The effect of obturator nerve block on hip lateralization in low functioning children with spastic cerebral palsy.

Park ES, Rha DW, Lee WC, Sim EG.

Purpose: Hip adductor spasticity has a great impact on developing hip displacement in children with cerebral palsy (CP). Obturator nerve (ON) block is less invasive intervention rather than soft tissue surgery for reduction of hip adductor spasticity. The aim of this study is to investigate the effect of ON block on hip lateralization in low functioning children with spastic CP. Materials and Methods: The study was performed by retrospective investigation of the clinical and radiographic follow-up data of low functioning children [gross motor function classification system (GMFCS) level III to V] with spastic cerebral palsy whose hip was subluxated. Migration percentage (MP) was measured on hip radiographs and its annual change was calculated. In intervention group, ON block was done with 50% ethyl alcohol under the guidance of electrical stimulation. Results: The data of 49 legs of 25 children for intervention group and the data of 41 legs of 23 children for nonintervention group were collected. In intervention group, the MP were significantly reduced at 1st follow-up and the MPs at 2nd and last follow-up did not show significant differences from initial MP. Whereas in nonintervention group, the MPs at 1st, 2nd and last follow-up were all significantly increased compared to initial MPs. Conclusion: ON block with ethyl alcohol is useful as an early effective procedure against progressive hip displacement in these children with spastic CP.

PMID: 24339306 [PubMed - in process] Free full text

16. Pediatrics. 2013 Dec 23. [Epub ahead of print]

Importance for CP Rehabilitation of Transfer of Motor Improvement to Everyday Life.

Taub E, Uswatte G.

PMID: 24366987 [PubMed - as supplied by publisher]

17. Bull Hosp Jt Dis (2013). 2013;71(4):272-7.

Hospital cost analysis of neuromuscular scoliosis surgery.

Diefenbach C, Ialenti MN, Lonner BS, Kamerlink JR, Verma K, Errico TJ.

Study Design: A retrospective review of 74 consecutive, surgical patients with neuromuscular scoliosis (NMS). Objective: This study evaluates the distribution of hos- pital and operating room costs incurred during surgical correction of NMS. Background Data: Recent studies have demonstrated that surgical treatment improves both medical outcomes and the quality of life in patients with progressive NMS. Characterization of the costs incurred at the time of surgery and hospitalization will facilitate the identification of opportunities for cost reduction. Methods: Demographic data collected included gender, age, preoperative height, weight, and BMI. Major coronal curvatures and T5-T12 kyphosis were assessed from radiographs. Construct type and number of screws, hooks, and wires implanted were recorded. Surgical costs were calculated based on cost of surgical correction, hospital stay, and postoperative care. Results: Mean age was 15.8 ± 7.3 years; 57% were male. Comorbidities included cerebral palsy (28%) and familial dysautonomia (14%). The mean preoperative major curve magnitude was 60°; minor curve magnitude was 33°. Posterior approach (76%) and pedicle screws (75%) were predominantly utilized. The average

length of hospitalization was 8 days (range: 3 to 47). There were six major complications (8%). The total surgical cost was $50,096 ± $23,998. The highest individual cost was for implants ($13,916; 24% of total costs). The second highest was inpatient room and ICU costs ($12,483; 22%); bone grafts were the third ($6,398; 11%). Increased major and minor structural curve, increased total (A/P) levels fused, and increased length of hospital stay predicted an increase in total cost. Conclusions: Major contributors to cost in NMS surgery are implants, inpatient room and ICU costs, and bone grafts. Independent predictors of higher cost are the degree of major and minor structural curve, total number of A/P levels fused, and length of hospital stay. These conclusions provide insight into costs associated with care for a medically fragile and challenging patient population.

PMID: 24344619 [PubMed - in process]

18. Eur Spine J. 2013 Dec 15. [Epub ahead of print]

Long-term surgical outcomes of cervical myelopathy with athetoid cerebral palsy.

Kim KN, Ahn PG, Ryu MJ, Shin DA, Yi S, Yoon DH, Ha Y.

PURPOSE: To understand the long-term surgical outcomes and prognostic factors for the operative treatment of cervical myelopathy (CM) in patients with athetoid cerebral palsy (ACP). METHODS: We retrospectively reviewed 24 patients with ACP who underwent surgery for CM at our hospital between March 2002 and June 2008. All patients had more than 5 years follow-up. Anterior fusion (11 patients), posterior fusion (1 patient), or combined anterior and posterior (AP) fusion (7 patients) and C1-2 fusion (5 patients) surgeries were performed. Surgical outcomes (average follow-up 102 months), as assessed using modified JOA (mJOA) scores, the Neck Disability Index (NDI), and a visual analog scale (VAS) were compared between the preoperative and postoperative states. RESULTS: Preoperative cervical kyphosis decreased mJOA scores significantly. Long-term follow-up clinical outcomes demonstrated that 10 patients showed favorable (excellent and good) outcomes and 11 patients had non-favorable (fair and worse) outcomes. According to the mJOA scores, patients showed postoperative improvement (7.10-10.45). NDI decreased from 68.46 to 31.66. A second operation was done in seven cases due to instrument failure, progressive kyphotic deformities and adjacent segment degeneration. A preoperative botulinum toxin injection significantly decreased ($p < 0.05$) the incidence of a second operation. CONCLUSIONS: Patients with ACP have high incidence of instrument failure. Strong surgical fixation, bone fusion and perioperative immobilizations using botulinum toxin injection should be carefully planned preoperatively.

PMID: 24337359 [PubMed - as supplied by publisher]

19. PM R. 2013 Dec 16. pii: S1934-1482(13)01226-4. doi: 10.1016/j.pmrj.2013.12.005. [Epub ahead of print]

Long-term Intrathecal Baclofen: Outcomes after Greater than 10 Years of Treatment.

Mathur SN1, Chu SK1, McCormick Z2, Chien GC1, Marciniak CM1.

OBJECTIVE: No study has reported outcomes of intrathecal baclofen (ITB) therapy for spasticity in a cohort of patients who have received this treatment for least 10 years. This study explored long term outcomes of ITB for spasticity management. Cross-sectional survey and retrospective chart review SETTING: Academic rehabilitation outpatient clinic PARTICIPANTS: Adult patients with spasticity treated with ITB for at least 10 years INTERVENTIONS: n/a MAIN OUTCOME MEASURES: Surveys included the Brief Pain Inventory (BPI), Penn Spasm Frequency Scale (PSFS), Epworth Sleepiness Scale (ESS), Fatigue Severity Scale (FSS), Diener Satisfaction with Life (SWLS), Life Satisfaction Questionnaire (LSQ), and Intrathecal Baclofen Survey (IBS). RESULTS: 24 subjects completed surveys. Subjects had been treated with ITB from 10.0-28.4 years with a mean of 14.7 years and standard deviation (SD) of 4.2. The mean dose of ITB was 627.9 micrograms/day (SD 306.7), with only 6 subjects averaging daily dose changes of greater than 10% compared to 3 years prior. The mean scores on outcomes surveys were: 2.6 (SD 2.3) for pain severity on the BPI, 1.4 (SD 0.7) for spasm severity on the PSFS, 7.9 (SD 5.4) on the ESS, 4.1 (SD 1.6) on the FSS, 19.4 (SD 8.1) on the SWLS, 3.9 (SD 0.9) on the LSQ, and 8.8 (SD 1.9) for overall satisfaction with ITB on the IBS. On the BPI, mean scores for pain severity and interference of pain with life showed moderate inverse correlations with ITB dose ($r = -0.386$, $p = .115$ and $r = -0.447$, $p = .062$ respectively). On the LSQ, mean scores for life satisfaction showed statistically significant positive correlation with ITB dose ($r = 0.549$, $p = .021$). CONCLUSIONS: Subjects reported low levels of pain, moderate levels of life satisfaction, normal levels of sleepiness, low to moderate levels of fatigue, infrequent spasms at mild to

moderate severity, and high levels of satisfaction. The efficacy and favorable side effect profile of ITB therapy was sustained in this cohort of subjects with over a decade of treatment.

PMID: 24355547 [PubMed - as supplied by publisher]

20. Physiother Theory Pract. 2013 Dec 16. [Epub ahead of print]

Aquatic aerobic exercise for children with cerebral palsy: a pilot intervention study.

Fragala-Pinkham MA, Smith HJ, Lombard KA, Barlow C, O'Neil ME.

Purpose: The primary purpose of this pilot study was to evaluate the effectiveness of a14-week aquatic exercise program on gross motor function and walking endurance in children with cerebral palsy (CP). The secondary purpose was to evaluate changes in functional strength, aerobic capacity and balance. Method: A prospective time series group design consisting of four measurement sessions (two baseline, one post intervention, and 1-month follow-up) was used. Eight ambulatory children ages 6-15 years with CP and classified at Gross Motor Function Classification System Level I or Level III participated in an aquatic aerobic exercise program. Results: Significant improvements were observed for the primary outcomes of gross motor function and walking endurance. No significant differences between any of the secondary measures were observed, although all of the measures demonstrated trends of improvement after intervention. Conclusion: Ambulatory children with CP may improve their gross motor skills and walking endurance after an aquatic exercise program held twice per week for 14 weeks, utilizing moderate-to-vigorous exercise intensity and consisting of functional activities.

PMID: 24328930 [PubMed - as supplied by publisher]

21. Physiother Theory Pract. 2013 Dec 11. [Epub ahead of print]

Effects of a recreational ice skating program on the functional mobility of a child with cerebral palsy.

Walsh SF, Scharf MG.

Purpose: The purpose of this study was to describe the effects of an ice skating program on the ambulation, strength, posture and balance of a child with cerebral palsy (CP). Description: The subject was a five-year-old female with a diagnosis of CP and a Gross Motor Classification System level of III. The subject was a slow and labored household ambulator on level surfaces with bilateral forearm crutches and bilateral ankle foot orthoses. She was unable to transfer to and from the floor to stand independently, stand unsupported or take steps independently. Until the initiation of this study she was receiving physical therapy services 2×/week. For the purpose of this study she participated in a 1h/week local ice skating program for people with disabilities for a period of four months. Outcomes: The subject displayed clinically significant improvements in functional mobility including: improved standing posture; independent transfer to and from the floor to stand; maintenance of independent standing for 3 min; independent walking for 10 feet; increased ability to isolate extremity musculature; increased strength; improved Gross Motor Function Measure-88 scores and increased endurance. A subsequent testing session four months after the ice skating program had ended displayed declines but not to pre-intervention levels in muscle strength; ability to transfer to and from the floor to stand; functional mobility and standing balance. Discussion: The results appear to suggest that the participation in an ice skating program clinically improved this child's functional mobility. Further research needs to be done with regard to physical recreational programs and the benefit they can have on the function of children with activity limitations.

PMID: 24328904 [PubMed - as supplied by publisher]

22. Dev Med Child Neurol. 2013 Dec 18. doi: 10.1111/dmcn.12352. [Epub ahead of print]

Development and reliability of a system to classify the eating and drinking ability of people with cerebral palsy.

Sellers D, Mandy A, Pennington L, Hankins M, Morris C.

AIM: The aim of this study was to develop a valid classification system to describe eating and drinking ability in people with cerebral palsy (CP), and to test its reliability. METHOD: The Eating and Drinking Ability Classification System (EDACS) was developed in four stages in consultation with individuals with CP, parents, and health professionals: Stage 1, drafting informed by literature and clinical experience; Stage 2, modification by nominal groups; Stage 3, refinement in an international Delphi survey; and Stage 4, testing of agreement and reliability between classifications made by speech and language therapists (SaLTs), and between SaLTs and parents. RESULTS: Seven nominal groups involved 56 participants; 95 people participated in two rounds of the Delphi survey. Using the version of EDACS produced from this process, SaLTs in pairs classified 100 children. The rate of absolute agreement was 78% (kappa=0.72; intraclass correlation coefficient [ICC]=0.93; 95% confidence interval [CI] 0.90-0.95). Any disagreement was only by one level, with one exception. SaLTs and parents classified 48 children. The rate of absolute agreement was 58% (kappa=0.45, ICC=0.86; 95% CI 0.76-0.92). Parents either agreed with SaLTs or rated their children as more able by one level. INTERPRETATION: The EDACS provides a valid and reliable system for classifying eating and drinking performance of people with CP, for use in both clinical and research contexts.

PMID: 24344767 [PubMed - as supplied by publisher]

23. Eur J Clin Nutr. 2013 Dec;67 Suppl 2:S1-2. doi: 10.1038/ejcn.2013.221.

An introduction to the supplement 'A practical approach to the nutritional management of children with cerebral palsy'.

Gottrand F, Sullivan PB.

PMID: 24301002 [PubMed - in process]

24. Eur J Clin Nutr. 2013 Dec;67 Suppl 2:S3-4. doi: 10.1038/ejcn.2013.222.

Nutrition and growth in children with cerebral palsy: setting the scene.

Sullivan PB.

PMID: 24301006 [PubMed - in process]

25. Eur J Clin Nutr. 2013 Dec;67 Suppl 2:S13-6. doi: 10.1038/ejcn.2013.225.

Nutritional management of children with cerebral palsy.

Bell KL, Samson-Fang L.

Children with severe cerebral palsy and particularly those with oropharyngeal dysfunction are at risk of poor nutritional status. Determining the need and the mode of nutritional intervention is multifactorial and requires multiple methodologies. First-line treatment typically involves oral nutritional support for those children who are safe to consume an oral diet. Enteral tube feeding may need to be considered in children with undernutrition where poor weight gain continues despite oral nutritional support, or in those with oropharyngeal dysphagia and an unsafe swallow. Estimates for energy and protein requirements provide a starting point only, and ongoing assessment and monitoring is essential to ensure nutritional needs are being met, that complications are adequately managed and to avoid over or under feeding.

PMID: 24301003 [PubMed - in process]

26. Eur J Clin Nutr. 2013 Dec 18. doi: 10.1038/ejcn.2013.281. [Epub ahead of print]

Nutritional management of children with cerebral palsy: a practical guide.

Kuperminc MN, Gottrand F, Samson-Fang L, Arvedson J, Bell K, Craig GM, Sullivan PB.

PMID: 24346472 [PubMed - as supplied by publisher]

27. Eur J Clin Nutr. 2013 Dec;67 Suppl 2:S9-S12. doi: 10.1038/ejcn.2013.224.

Feeding children with cerebral palsy and swallowing difficulties.

Arvedson JC.

Children with cerebral palsy (CP) are at risk for aspiration with oral feeding with potential pulmonary consequences, and commonly have reduced nutrition/hydration status and prolonged stressful meal times. There is considerable variability in the nature and severity of swallowing problems in these children whose needs change over time. Children with generalized severe motor impairment (for example, spastic quadriplegia) are likely to experience greater swallowing deficits than those with diplegia, but oropharyngeal dysphagia is prevalent even in children with mild CP. This review is focused on dysphagia in children with CP: types of deficits, clinical and instrumental evaluation, management decision making and evidence of effectiveness of interventions.

PMID: 24301008 [PubMed - in process]

28. Eur J Clin Nutr. 2013 Dec;67 Suppl 2:S5-8. doi: 10.1038/ejcn.2013.223.

Assessment of growth and nutrition in children with cerebral palsy.

Samson-Fang L, Bell KL.

This manuscript provides an update on the assessment of growth and nutrition in children with cerebral palsy and children with similar neurodevelopmental disabilities. Topics include the assessment of linear growth using segmental measures, avoidance of commonly used tools to assess nutritional status in typically developing children that are not valid in this population of children and how to use other nutritional assessment tools that have been developed specific to this population of children.

PMID: 24301007 [PubMed - in process]

29. Dev Med Child Neurol. 2013 Dec 17. doi: 10.1111/dmcn.12342. [Epub ahead of print]

Accuracy of skinfold and bioelectrical impedance assessments of body fat percentage in ambulatory individuals with cerebral palsy.

Oeffinger DJ, Gurka MJ, Kuperminc M, Hassani S, Buhr N, Tylkowski C.

AIM: This study assessed the accuracy of measurements of body fat percentage in ambulatory individuals with cerebral palsy (CP) from bioelectrical impedance analysis (BIA) and skinfold equations.
METHOD: One hundred and twenty-eight individuals (65 males, 63 females; mean age 12y, SD 3, range 6-18y) with CP (Gross Motor Function Classification System [GMFCS] levels I (n=6), II (n=46), and III (n=19) participated. Body fat percentage was estimated from (1) BIA using standing height and estimated heights (knee height and tibial length) and (2) triceps and subscapular skinfolds using standard and CP-specific equations. All estimates of body fat percentage were compared with body fat percentage from dual-energy X-ray absorptiometry (DXA) scans. Differences between DXA, BIA, and skinfold body fat percentage were analyzed by comparing mean differences.

Agreement was assessed by Bland-Altman plots and concordance correlation coefficients (CCC). RESULTS: BMI was moderately correlated with DXA (Pearson's r=0.53). BIA body fat percentage was significantly different from DXA when using estimated heights (95% confidence intervals [CIs] do not contain 0) but not standing height (95% CI -1.9 to 0.4). CCCs for all BIA comparisons indicated good to excellent agreement (0.75-0.82) with DXA. Body fat percentage from skinfold measurements and CP-specific equations was not significantly different from DXA (mean 0.8%; SD 5.3%; 95% CI -0.2 to 1.7) and demonstrated strong agreement with DXA (CCC 0.86). INTERPRETATION: Accurate measures of body fat percentage can be obtained using BIA and two skinfold measurements (CP-specific equations) in ambulatory individuals with CP. These findings should encourage assessments of body fat in clinical and research practices.

PMID: 24344745 [PubMed - as supplied by publisher]

30. PM R. 2013 Dec;5(12):1077-80. doi: 10.1016/j.pmrj.2013.07.002.

Deleterious cognitive and motoric effects of haloperidol in an adolescent with cerebral palsy: a case report.

Mortimer D1, Gelfius CD2, Potts MA3.

This case report describes a 15-year-old male patient with spastic diplegic cerebral palsy, Gross Motor Function Classification System Level III, who developed severe new cognitive and motoric impairments after the administration of haloperidol. He received this dopamine antagonist and typical antipsychotic medication for an acute postoperative episode of agitation. He improved when he received the dopamine agonists amantadine and carbidopa/levodopa. This case suggests that dopamine blockade may be deleterious for individuals with cerebral palsy. Potential explanations for the events observed in this case are also presented.

PMID: 24332231 [PubMed - in process]

31. Dev Med Child Neurol. 2013 Dec 21. doi: 10.1111/dmcn.12343. [Epub ahead of print]

Developmental trajectories of social participation in individuals with cerebral palsy: a multi-centre longitudinal study.

Tan SS, Wiegerink DJ, Vos RC, Smits DW, Voorman JM, Twisk JW, Ketelaar M, Roebroeck ME; the PERRIN+ study group.

AIM: This study aimed to determine the developmental trajectories of social participation, by level of gross motor function and intellectual disability, in a Dutch population of individuals with cerebral palsy (CP) aged 1 to 24 years. METHOD: As part of the Pediatric Rehabilitation Research in the Netherlands (PERRIN+), 424 individuals with CP (261 males, 163 females; mean age [SD] 9y 6mo [6y 2mo]; Gross Motor Function Classification [GMFCS] levels I-V [50% level I]; 87% with spastic CP; 26% with intellectual disability) were longitudinally followed for up to 4 years between 2002 and 2007. Social participation was assessed with the Vineland Adaptive Behavior Scales survey. Effects of age, GMFCS level and intellectual disability were analysed using multilevel modelling. RESULTS: The developmental trajectories for individuals in GMFCS levels I to IV did not significantly differ from each other. For individuals without intellectual disability, the degree of social participation increased with age and stabilized at about 18 years. These individuals reached social participation levels similar to typically developing individuals. The trajectories were significantly less favourable for individuals in GMFCS level V and individuals with intellectual disability. INTERPRETATION: Intellectual disability is more distinctive for the development of social participation than GMFCS level. The developmental trajectories will support individuals with CP and their families in setting realistic goals and professionals in optimizing the choice of interventions at an early age.

PMID: 24359158 [PubMed - as supplied by publisher]

32. Autism. 2013 Dec 18. [Epub ahead of print]

Access to services, quality of care, and family impact for children with autism, other developmental disabilities, and other mental health conditions.

Vohra R, Madhavan S, Sambamoorthi U, St Peter C.

This cross-sectional study examined perceived access to services, quality of care, and family impact reported by caregivers of children aged 3-17 years with autism spectrum disorders, as compared to caregivers of children with other developmental disabilities and other mental health conditions. The 2009-2010 National Survey of Children with Special Health Care Needs was utilized to examine the association between child's special needs condition and three outcomes (N = 18,136): access to services (difficulty using services, difficulty getting referrals, lack of source of care, and inadequate insurance coverage), quality of care (lack of care coordination, lack of shared decision making, and no routine screening), and family impact (financial, employment, and time-related burden). Multivariate logistic regressions were performed to compare caregivers of children with autism spectrum disorders to caregivers of children with developmental disabilities (cerebral palsy, Down syndrome, developmental delay, or intellectual disability), mental health conditions (attention deficit hyperactivity disorder, anxiety, behavioral/conduct problems, or depression), or both developmental disabilities and mental health conditions. Caregivers of children with autism spectrum disorders were significantly more likely to report difficulty using services, lack of source of care, inadequate insurance coverage, lack of shared decision making and care coordination, and adverse family impact as compared to caregivers of children with developmental disabilities, mental health conditions, or both.

PMID: 24353274 [PubMed - as supplied by publisher]

33. Res Dev Disabil. 2013 Dec 16;35(2):414-422. doi: 10.1016/j.ridd.2013.11.021. [Epub ahead of print]

Household task participation of children and adolescents with cerebral palsy, Down syndrome and typical development.

Amaral MF1, Drummond AD2, Coster WJ3, Mancini MC4.

This cross-sectional study compared patterns of household task participation (e.g., performance, assistance and independence) of youth with cerebral palsy (CP), Down syndrome (DS) and typical development (TD). Parents of 75 children and adolescents were interviewed to report on their youths' active engagement in daily self-care and family-care tasks, using the children helping out: responsibilities, expectations and supports (CHORES) questionnaire. Groups were equivalent in age (mean=9.3 years; SD=2.2 years), sex (male=39; female=36), respondent education, presence of maid, and number of siblings at home, but differed on child cognitive function and family socioeconomic status, with the DS and the CP groups scoring lower than the TD group but not different from each other. ANOVA revealed group differences on CHORES performance of self-care tasks ($p=0.004$), on total participation score ($p=0.04$) and on assistance scores ($p<0.02$). Post hoc comparisons showed that TD group scored higher than CP and DS groups on performance and assistance in self-care tasks and total assistance; TD and CP groups were similar on total performance and assistance in family-care tasks. The groups also differed on independence indices; the TD index was greater than the CP and DS, and the CP index was greater than the DS. Parents from the three groups did not differ on ratings of importance regarding their children's household participation ($p=0.416$). In spite of observed differences, children and adolescents with CP and DS are actively engaged in daily self-care and family-care tasks; their participation at home is not prevented by the presence of their disabilities.

PMID: 24355162 [PubMed - as supplied by publisher]

34. Int J Nephrol. 2013;2013:937268. doi: 10.1155/2013/937268. Epub 2013 Nov 25.

Prevalence and Predictors of Urinary Tract Infections among Children with Cerebral Palsy in Makurdi, Nigeria.

Anígilájé EA1, Bitto TT2.

Background. Children with cerebral palsy (CP) are prone to urinary tract infection (UTI). Methods/Objectives. The prevalence and the predictors of UTI among children with CP were compared to age- and sex-matched children without CP at Federal Medical Centre, Makurdi, Nigeria, from December 2011 to May 2013. Results. The age range was between 2 and 15 years with a mean age of 8.63 ± 3.83 years including 30 males and 22 females. UTI was confirmed in 20 (38.5%) CP children compared to 2 children (3.8%) without CP (P value 0.000). Among CP children, Escherichia coli was the commonest organism isolated in 9 (9/20, 45.0%), Streptococcus faecalis in 4 (20.0%), and Staphylococcus aureus in 3 (15%), while both Proteus spp. and Klebsiella spp. were isolated in 2 children (10.0%) each. Escherichia coli was also found in the 2 children without CP. All the organisms were resistant to cotrimoxazole, nalidixic acid, nitrofurantoin, and amoxiclav, while they were 100% sensitive to ceftriaxone and the quinolones. In a univariate regression analysis, only moderate to severe gross motor dysfunction predicted the risk of UTI (OR = 54.81, 95% CI, 2.27-1324.00, P value 0.014). Conclusion. Efforts should be put in place to aid mobility among CP children in order to reduce the risk of UTI.

PMID: 24371524 [PubMed] PMCID: PMC3858867 Free full text

35. Ann Phys Rehabil Med. 2013 Dec 2. pii: S1877-0657(13)01327-4. doi: 10.1016/j.rehab.2013.11.002. [Epub ahead of print]

Analysis of the medical causes of death in cerebral palsy.

Duruflé-Tapin A1, Colin A2, Nicolas B1, Lebreton C3, Dauvergne F4, Gallien P5.

AIM: To investigate causes of death and age at death in cerebral palsy subjects compared with the general population. METHOD: Analysis of data supplied by the centre of epidemiology on the medical causes of death within the National Institute of Health and Medical Research in France was conducted. Three thousand and thirty-one death certificates indicating a diagnosis of cerebral palsy (ICD-10 code G80) were reported between 2000 and 2008. RESULTS: Median age at death was between 45-54 years and principal cause of death (24%) comprised the category "symptoms, signs, and abnormal results of clinical and laboratory tests, not classified elsewhere". Of these, 66% were related to the circulatory and respiratory systems. "Diseases of the respiratory system" were the second most common cause of death (19% compared with 6% in the French general population). The third most common cause of death was "diseases of the circulatory system" (15% compared with 29% in the French general population). While deaths caused by tumour pathologies in the general population are the most common cause of deaths, these represented only 7% of deaths in subjects with cerebral palsy. INTERPRETATION: These results concur with other published data, i.e. subjects with cerebral palsy die younger than the French general population, and the principal causes of death are respiratory and circulatory problems. This study emphasises the importance of access to epidemiological data about the French cerebral palsy population.

PMID: 24341986 [PubMed - as supplied by publisher]

36. Neuropsychiatr Dis Treat. 2013;9:1853-8. doi: 10.2147/NDT.S52947. Epub 2013 Nov 29.

Influence of cerebellar stereotactic stimulation on left-right electrodermal information transference in a patient with cerebral palsy.

Bob P1, Galanda T2, Jombik P3, Raboch J4, Galanda M2.

BACKGROUND AND OBJECTIVES: Recent evidence indicates that cerebral palsy is connected to specific autonomic dysregulation between sympathetic and parasympathetic efferent pathways, likely linked to hemispheric influences. These findings suggest a hypothesis that contralateral interhemispheric disinhibition, which may occur

on various levels of brain processing including motor functions, could be linked to specific functional dysregulation and structural lesions, which may play a specific role in the modulation of autonomic functions and lead to autonomic dysregulation in cerebral palsy. METHOD: With the aim of comparing autonomic functions as they relate to interhemispheric modulatory influences during therapeutically indicated stereotactic cerebellar stimulation, we have performed bilateral electrodermal activity measurement and calculations of pointwise transinformation (PTI) in a patient with cerebral palsy. Measurement was performed during therapeutic deep cerebellar stimulation in two cerebellar areas in anterior cerebellar lobe-culmen (left electrode) and central lobule-superior cerebellar peduncle (right electrode). RESULTS: The results indicate that information transference (PTI) is able to distinguish the states related to specific cerebellar stimulations and that lowest levels of the PTI have been found during stimulation of the central lobule-superior cerebellar peduncle (electrode deepest contact 1), indicating a significantly increased level of inhibition between the left and right sides. CONCLUSION: The results may present potentially useful clinical findings indicating that increased PTI calculated from electrodermal activity could indirectly indicate disinhibitory activity as a possible indicator of a failure of interhemispheric communication that could explain some specific pathogenetic mechanisms in cerebral palsy. Nevertheless, these results need detailed confirmation in further research, as well as reliable clinical evaluation of their usefulness in the therapy of cerebral palsy.

PMID: 24348041 [PubMed] PMCID: PMC3849142 Free PMC Article

37. J Neurosci Rural Pract. 2013 Oct;4(4):408-12. doi: 10.4103/0976-3147.120241.

Intelligence quotient is associated with epilepsy in children with intellectual disability in India.

Lakhan R.

BACKGROUND: Epilepsy is a disorder that is commonly found in people with intellectual disability (ID). The prevalence of epilepsy increases with the severity of ID. The objective of this study was to determine if there is an association between intelligence quotient (IQ) and epilepsy in children with ID. MATERIALS AND METHODS: A total of 262 children, aged 3-18 years, with ID were identified as part of a community-based rehabilitation project. These children were examined for epilepsy and diagnosed by a psychiatrist and physicians based on results of electroencephalogram tests. A Spearman's correlation (ρ) was used to determine if there was an association between IQ scores and the occurrence of epilepsy. X(2) statistics used to examine the relationship of epilepsy with gender, socioeconomic status, population type, severity of ID, family history of mental illness, mental retardation, epilepsy, and coexisting disorder. RESULTS: Spearman's rho -0.605 demonstrates inverse association of IQ with epilepsy. X(2) demonstrates statistically significant association ($P < 0.05$) with gender, severity of ID, cerebral palsy, behavior problems, and family history of mental illness, mental retardation, and epilepsy. CONCLUSIONS: Lower IQ score in children with ID has association with occurrence of epilepsy. Epilepsy is also found highly associated with male gender and lower age.

PMID: 24347947 [PubMed] PMCID: PMC3858759 Free PMC Article

Prevention and Cure

38. BMJ Open. 2013 Dec 23;3(12):e004141. doi: 10.1136/bmjopen-2013-004141.

Antenatal melatonin as an antioxidant in human pregnancies complicated by fetal growth restriction--a phase I pilot clinical trial: study protocol.

Alers NO, Jenkin G, Miller SL, Wallace EM.

BACKGROUND: Fetal growth restriction complicates about 5% of pregnancies and is commonly caused by placental dysfunction. It is associated with increased risks of perinatal mortality and short-term and long-term morbidity, such as cerebral palsy. Chronic in utero hypoxaemia, inflammation and oxidative stress are likely culprits contributing to the long-term neurological sequelae of fetal growth restriction. In this regard, we propose that melatonin, a powerful antioxidant, might mitigate morbidity and/or mortality associated with fetal growth restriction. Melatonin has an excellent biosafety profile and crosses the placenta and blood-brain barrier. We present the

protocol for a phase I clinical trial to investigate the efficacy of maternal oral melatonin administration in women with a pregnancy complicated by fetal growth restriction. METHODS AND ANALYSIS: The proposed trial is a single-arm, open-label clinical trial involving 12 women. Severe, early onset fetal growth restriction will be diagnosed by an estimated fetal weight =10th centile in combination with abnormal fetoplacental Doppler studies, occurring before 34 weeks of pregnancy. Baseline measurements of maternal and fetal well-being, levels of oxidative stress and ultrasound and Doppler measurements will be obtained at the time of diagnosis of fetal growth restriction. Women will then start melatonin treatment (4 mg) twice daily until birth. The primary outcomes are the levels of oxidative stress in the maternal and fetal circulation and placenta. Secondary outcomes are fetoplacental Doppler studies (uterine artery, umbilical artery middle cerebral artery and ductus venosus), fetal biometry, fetal biophysical profile and a composite determination of neonatal outcome. A historical cohort of gestational-matched fetal growth restriction and a healthy pregnancy cohort will be used as comparators. ETHICS AND DISSEMINATION: Ethical approval has been obtained from Monash Health Human Research Ethics Committee B (HREC12133B). Data will be presented at international conferences and published in peer-reviewed journals.

TRIAL REGISTRATION NUMBER: Clinical Trials, protocol registration system: NCT01695070.

PMID: 24366583 [PubMed] Free full text

39. Dev Med Child Neurol. 2013 Dec 21. doi: 10.1111/dmcn.12355. [Epub ahead of print]

Etiology of impaired selective motor control: emerging evidence and its implications for research and treatment in cerebral palsy.

Cahill-Rowley K, Rose J.

Selective motor control (SMC) impairment involves movement patterns dominated by flexor or extensor synergies that interfere with functional movements in children with cerebral palsy (CP). Emerging evidence on neural correlates of impaired SMC has important implications for etiology and for the treatment for children with CP. Early evidence on the microstructure of brain white matter assessed with diffusion tensor imaging in adult patients after stroke suggests that the rubrospinal tract may compensate for injury to the corticospinal tract. Furthermore, the observed changes on diffusion tensor imaging corresponded to the degree of SMC impairment. The rubrospinal tract may provide imperfect compensation in response to corticospinal tract injury, resulting in diminished SMC. Cortical mapping evidence in stroke patients indicates that loss of SMC is also associated with increased overlap of joint representation in the sensorimotor cortices. The severity of SMC impairment can be assessed with the recently developed Selective Control Assessment of the Lower Extremity, a validated observation-based measure designed for children with spastic CP. Recent advances in neuroimaging and assessment of SMC provide an opportunity to better understand the etiology and impact of impaired SMC, which may ultimately guide strategic treatment for children with CP.

PMID: 24359183 [PubMed - as supplied by publisher]

40. Am J Med Genet A. 2013 Dec 19. doi: 10.1002/ajmg.a.36329. [Epub ahead of print]

Severe clinical presentation in monozygotic twins with 10p15.3 microdeletion syndrome.

Vargiami E, Ververi A, Kyriazi M, Papathanasiou E, Gioula G, Gerou S, Al-Mutawa H, Kambouris M, Zafeiriou DI.

Submicroscopic deletion of 10p15.3 is a rare genetic disorder, currently reported in 21 unrelated patients. It is mainly associated with cognitive deficits, speech disorders, motor delay and hypotonia. The size of the deleted region ranges between 0.15 and 4Mb and does not generally correlate with phenotype. A monozygotic female twin pair with a de novo 2.7Mb deletion of 10p15.3 is herein reported. The girls presented at the age of 8 months with severe developmental delay and failure to thrive since the first month of life. Their perinatal and family history was unremarkable. On admission they both exhibited generalized dystonia, microcephaly, complete absence of voluntary movements and visual/auditory unresponsiveness. Their brain MRIs demonstrated dilatation of ventricles, subarachnoid spaces and anterior interhemispheric fissure and sylvian fissures bilaterally. Cranial radiography revealed partial fusion of both coronal sutures. Visual and brainstem auditory evoked potentials were markedly

abnormal, indicating severe visual and sensorineural hearing impairment. The electroencephalogram, as well as a screening for inborn errors of metabolism, were unremarkable. Both patients required gastrostomy and tracheostomy before the age of 1 year. They were, additionally, managed with physical therapy, as well as baclofen and low-dose haloperidol. Their current state at the age of 2 years is relatively stable. The index patients' phenotype includes features, such as dystonic cerebral palsy, visual and sensorineural hearing impairment or craniosynostosis, which have not been previously reported in individuals with 10p15.3 deletion. It is necessary to consider these novel clinical features and investigate their possible relationship with the recently recognized syndrome. © 2013 Wiley Periodicals, Inc.

PMID: 24357427 [PubMed - as supplied by publisher]

41. J Pediatr. 2014 Jan;164(1):9-11. doi: 10.1016/j.jpeds.2013.10.020.

Stopping the swinging pendulum of postnatal corticosteroid use.

Demauro SB1, Dysart K1, Kirpalani H2.

PMID: 24359899 [PubMed - in process]

42. BMC Health Serv Res. 2013 Dec 19;13(1):527. doi: 10.1186/1472-6963-13-527.

Magnesium sulphate for fetal neuroprotection: a cost-effectiveness analysis.

Bickford CD, Magee LA, Mitton C, Kruse M, Synnes AR, Sawchuck D, Basso M, Senikas VM, von Dadelszen P; MAG-CP Working Group.

BACKGROUND: The aim of this study was to assess the cost-effectiveness of administering magnesium sulphate to patients in whom preterm birth at<32+0 weeks gestation is either imminent or threatened for the purpose of fetal neuroprotection. METHODS: Multiple decision tree models and probabilistic sensitivity analyses were used to compare the administration of magnesium sulphate with the alternative of no treatment. Two separate cost perspectives were utilized in this series of analyses: a health system and a societal perspective. In addition, two separate measures of effectiveness were utilized: cases of cerebral palsy (CP) averted and quality-adjusted life years (QALYs). RESULTS: From a health system and a societal perspective, respectively, a savings of $2,242 and $112,602 is obtained for each QALY gained and a savings of $30,942 and $1,554,198 is obtained for each case of CP averted when magnesium sulphate is administered to patients in whom preterm birth is imminent. From a health system perspective and a societal perspective, respectively, a cost of $2,083 is incurred and a savings of $108,277 is obtained for each QALY gained and a cost of $28,755 is incurred and a savings of $1,494,500 is obtained for each case of CP averted when magnesium sulphate is administered to patients in whom preterm birth is threatened. CONCLUSIONS: Administration of magnesium sulphate to patients in whom preterm birth is imminent is a dominant (i.e. cost-effective) strategy, no matter what cost perspective or measure of effectiveness is used. Administration of magnesium sulphate to patients in whom preterm birth is threatened is a dominant strategy from a societal perspective and is very likely to be cost-effective from a health system perspective.

PMID: 24350635 [PubMed - in process] Free full text

43. Glia. 2013 Dec 19. doi: 10.1002/glia.22616. [Epub ahead of print]

Astrocytes play a key role in EAE pathophysiology by orchestrating in the CNS the inflammatory response of resident and peripheral immune cells and by suppressing remyelination.

Brambilla R, Morton PD, Ashbaugh JJ, Karmally S, Lambertsen KL, Bethea JR.

Astrocytes respond to insult with a process of cellular activation known as reactive astrogliosis. One of the key signals regulating this phenomenon is the transcription factor nuclear factor-kappa B (NF-?B), which is responsible for modulating inflammation, cell survival, and cell death. In astrocytes, following trauma or disease, the expression of NF-B-dependent genes is highly activated. We previously demonstrated that inactivation of astroglial NF-?B in vivo (GFAP-I Ba-dn mice) leads to improved functional outcome in experimental autoimmune encephalomyelitis

(EAE), and this is accompanied by reduction of pro-inflammatory gene expression in the CNS. Here we extend our studies to show that recovery from EAE in GFAP-I Ba-dn mice is associated with reduction of peripheral immune cell infiltration into the CNS at the chronic phase of EAE. This is not dependent on a less permeable blood-brain barrier, but rather on a reduced immune cell mobilization from the periphery. Furthermore, once inside the CNS, the ability of T cells to produce pro-inflammatory cytokines is diminished during acute disease. In parallel, we found that the number of total and activated microglial cells is reduced, suggesting that functional improvement in GFAP-I?Ba-dn mice is dependent upon reduction of the overall inflammatory response within the CNS sustained by both resident and infiltrating cells. This results in preservation of myelin compaction and enhanced remyelination, as shown by electron microscopy analysis of the spinal cord. Collectively our data indicate that astrocytes are key players in driving CNS inflammation and are directly implicated in the pathophysiology of EAE, since blocking their pro-inflammatory capability results in protection from the disease. GLIA 2013;.

PMID: 24357067 [PubMed - as supplied by publisher]

44. J Child Neurol. 2013 Dec 11. [Epub ahead of print]

Is Prepregnancy Obesity Associated With Risk of Cerebral Palsy and Epilepsy in Children?

Pan C, Deroche CB, Mann JR, McDermott S, Hardin JW.

We conducted a retrospective cohort study to investigate the association between prepregnancy obesity in women and risk of cerebral palsy and epilepsy in their children using data from the South Carolina Medicaid program. The cohort included 83,901 maternal-child pairs; 100 cases of cerebral palsy were initially identified, followed by 53 cases that had at least 2 cerebral palsy diagnoses. For confirmed epilepsy, diagnosed on at least 5 occasions or by more than 1 provider, 83,472 observations were included with 338 cases. There was no association between maternal body mass index and risk of childhood epilepsy. A significant association between increasing maternal body mass index and any diagnosis of cerebral palsy was found, and morbid obesity was associated with increased risk of any and confirmed cerebral palsy. In conclusion, there appears to be an association of maternal body mass index with cerebral palsy, but there is no evidence to support an association with epilepsy.

PMID: 24334343 [PubMed - as supplied by publisher]

45. J Dev Behav Pediatr. 2013 Dec 18. [Epub ahead of print]

Disparities in Canadian Indigenous Health Research on Neurodevelopmental Disorders.

Di Pietro NC, Illes J.

OBJECTIVE: To map the landscape of research on autism (ASD), cerebral palsy (CP), and fetal alcohol spectrum disorder (FASD) in Canadian Aboriginal children. METHOD: The authors used a detailed search strategy to identify and access publications on ASD, CP, and FASD involving Canadian Aboriginal children, families, and communities from online databases. They analyzed these materials for the type of research, stated objectives, methodologies, and the level of engagement of Aboriginal Peoples. RESULTS: The authors found a total of 52 reports published since 1981 relevant to Aboriginal children. Of these, 51 focused exclusively on FASD. They also found a near-complete failure to acknowledge community involvement in research decisions or dissemination of results in any of the publications. CONCLUSIONS: The focus on FASD in Aboriginal children and the absence of research on the other 2 major childhood disorders are at odds with rates of these disorders across Canadian children. The authors argue that this trend violates fundamental principles ensuring equitable representation of all children regardless of background in research and access to benefits of research in health care and perpetuates stigma in an already marginalized population.

PMID: 24356498 [PubMed - as supplied by publisher]

46. J Neuroinflammation. 2013 Dec 17;10:153. doi: 10.1186/1742-2094-10-153.

LPS and TNF alpha modulate AMPA/NMDA receptor subunit expression and induce PGE2 and glutamate release in preterm fetal ovine mixed glial cultures.

Weaver-Mikaere L, Gunn AJ, Mitchell MD, Bennet L, Fraser M.

BACKGROUND: White matter injury (WMI) is the major antecedent of cerebral palsy in premature infants, and is often associated with maternal infection and the fetal inflammatory response. The current study explores the therapeutic potential of glutamate receptor blockade or cyclooxygenase-2 (COX-2) inhibition for inflammatory WMI. METHODS: Using fetal ovine derived mixed glia cultures exposed to tumour necrosis factor-a (TNF-a) or lipopolysaccharide (LPS), the expression of alpha-amino-3-hydroxy-5-methyl-4-isoxazole-propionate (AMPA) and N -methyl D-aspartate (NMDA) glutamate receptors and their contribution to inflammation mediated pre-oligodendrocyte (OL) death was evaluated. The functional significance of TNF-a and COX-2 signalling in glutamate release in association with TNF-a and LPS exposure was also assessed. RESULTS: AMPA and NMDA receptors were expressed in primary mixed glial cultures on developing OLs, the main cell-type present in fetal white matter at a period of high risk for WMI. We show that glutamate receptor expression and configuration are regulated by TNF-a and LPS exposure, but AMPA and NMDA blockade, either alone or in combination, did not reduce pre-OL death. Furthermore, we demonstrate that glutamate and prostaglandin E2 (PGE2) release following TNF-a or LPS are mediated by a TNF-a-COX-2 dependent mechanism. CONCLUSIONS: Overall, these findings suggest that glial -localised glutamate receptors likely play a limited role in OL demise associated with chronic inflammation, but supports the COX-2 pathway as a potential therapeutic target for infection/inflammatory-mediated WMI.

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Coagulase-negative staphylococcus sepsis in preterm infants and long-term neurodevelopmental outcome.

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Objective: The objective of this study was to examine the impact of Coagulase-negative staphylococcus (CoNS) sepsis in preterm infants on the neurodevelopmental outcomes at 30 to 42 months corrected age (CA). Study Design: This is a retrospective cohort study. All preterm infants born at <29 weeks gestational age between 1995 and 2008 and had a neurodevelopmental assessment at 30 to 42 months CA were eligible. The neurodevelopmental outcomes of infants exposed to CoNS sepsis were compared with infants unexposed to any type of neonatal sepsis. Result: A total of 105 eligible infants who were exposed to CoNS sepsis were compared with 227 infants with no neonatal sepsis. In univariate analysis, infants with CoNS sepsis were more likely to have total major disability (odds ratio (OR)=1.9; 95% CI: 1.07 to 3.38) and cognitive delay (OR=2.53; 1.26 to 5.14).There was no significant difference in the incidence of cerebral palsy, blindness and deafness between the two groups. After correcting for potential confounders, CoNS sepsis was associated with increased risk of cognitive delay (adjusted odds ratio (aOR)= 2.23; 95% CI 1.01 to 4.9), but not with the total major disability (aOR=1.14; 95% CI: 0.55 to 2.34).Conclusion:Our study suggests that CoNS sepsis in preterm infants might be associated with increased risk for cognitive delay at 36 months CA.

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Our experience with the aetiological diagnosis of global developmental delay and intellectual disability: 2006-2010. [Article in English, Spanish]

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INTRODUCTION: Global developmental delay (GDD) and intellectual disability (ID) are common reasons for consultation in paediatric neurology. Results from aetiological evaluations of children with GDD/ID vary greatly, and

consequently, there is no universal consensus regarding which studies should be performed MATERIAL AND METHOD: We review our experience with determining aetiological diagnoses for children with GDD/ID who were monitored by the paediatric neurology unit over the 5-year period between 2006 and 2010 RESULTS: During the study period, 995 children with GDD/ID were monitored. An aetiological diagnosis was established for 309 patients (31%), but not in 686 (69%), despite completing numerous tests. A genetic cause was identified in 142 cases (46% of the total aetiologies established), broken down as 118 cases of genetic encephalopathy and 24 of metabolic hereditary diseases. Our data seem to indicate that diagnosis is easier when GDD/ID is associated with cerebral palsy, epilepsy, infantile spasms/West syndrome, or visual deficit, but more difficult in cases of autism spectrum disorders. Genetic studies provide an increasing number of aetiological diagnoses, and they are also becoming the first step in diagnostic studies. Array CGH (microarray-based comparative genomic hybridisation) is the genetic test with the highest diagnostic yield in children with unexplained GDD/ID DISCUSSION: The cost-effectiveness of complementary studies seems to be low if there are no clinically suspected entities. However, even in the absence of treatment, aetiological diagnosis is always important in order to provide genetic counselling and possible prenatal diagnosis, resolve family (and doctors') queries, and halt further diagnostic studies.

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Astrocytes and microglia in acute cerebral injury underlying cerebral palsy associated with preterm birth.

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Cerebral palsy is one of the most devastating consequences of brain injury around the time of birth, and nearly a third of cases are now associated with premature birth. Compared with term babies, preterm babies have an increased incidence of complications that may increase the risk of disability, such as intraventricular hemorrhage, periventricular leukomalacia, sepsis, and necrotizing enterocolitis. The response to injury is highly dependent on brain maturity, and although cellular vulnerability is well documented, there is now evidence that premyelinating axons are also particularly sensitive to ischemic injury. In this review, we will explore recent evidence highlighting a central role for glia in mediating increased risk of disability in premature infants, including excessive activation of microglia and opening of astrocytic gap junction hemichannels in spreading injury after brain ischemia, in part likely involving release of adenosine triphosphate (ATP) and overactivation of purinergic receptors, particularly in white matter. We propose the hypothesis that inflammation-induced opening of connexin hemichannels is a key regulating event that initiates a vicious circle of excessive ATP release, which in turn propagates activation of purinergic receptors on microglia and astrocytes. This suggests that developing effective neuroprotective strategies for preterm infants requires a detailed understanding of glial responses.

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