Cerebral Palsy ALLIANCE

NEWS
Cerebral Palsy Research

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Interventions and Management

1. Evaluation of cervical posture improvement of children with cerebral palsy after physical therapy based on head movements and serious games.

Velasco MA, Raya R, Muzzioli L, Morelli D, Otero A, Iosa M, Cincotti F, Rocon E.

Biomed Eng Online. 2017 Aug 18;16(Suppl 1):74. doi: 10.1186/s12938-017-0364-5.

BACKGROUND: This paper presents the preliminary results of a novel rehabilitation therapy for cervical and trunk control of children with cerebral palsy (CP) based on serious videogames and physical exercise. MATERIALS: The therapy is based on the use of the ENLAZA Interface, a head mouse based on inertial technology that will be used to control a set of serious videogames with movements of the head. METHODS: Ten users with CP participated in the study. Whereas the control group (n = 5) followed traditional therapies, the experimental group (n = 5) complemented these therapies with a series of ten sessions of gaming with ENLAZA to exercise cervical flexion-extensions, rotations and inclinations in a controlled, engaging environment. RESULTS: The ten work sessions yielded improvements in head and trunk control that were higher in the experimental group for Visual Analogue Scale, Goal Attainment Scaling and Trunk Control Measurement Scale (TCMS). Significant differences (27% vs. 2% of percentage improvement) were found between the experimental and control groups for TCMS ($p < 0.05$). The kinematic assessment shows that there were some improvements in the active and the passive range of motion. However, no significant differences were found pre- and post-intervention. CONCLUSIONS: Physical therapy that combines serious games with traditional rehabilitation could allow children with CP to achieve larger function improvements in the trunk and cervical regions. However, given the limited scope of this trial (n = 10) additional studies are needed to corroborate this hypothesis.

PMID: 28830552

2. Postural orientation and standing postural alignment in ambulant children with bilateral cerebral palsy.

Domagalska-Szopa M, Szopa A.

Clin Biomech (Bristol, Avon). 2017 Aug 16;49:22-27. doi: 10.1016/j.clinbiomech.2017.08.005. [Epub ahead of print]

BACKGROUND: Standing postural alignment in children with cerebral palsy is usually altered by central postural control disorders. The primary aim of this study is to describe body alignment in a quiet standing position in ambulatory children with bilateral cerebral palsy compared with children with typical development. METHODS: Fifty-eight children with bilateral cerebral palsy (aged 7-13years) and 45 age-matched children with typical development underwent a surface topography examination based on Moiré topography and were classified according to their sagittal postural profiles. FINDINGS: The following eight grouping variables were extracted using a data reduction technique: angle of trunk inclination, pelvic tilt, and lordosis, the difference between kyphosis and lordosis, angle of vertebral lateral curvature, shoulder inclination, and shoulder and pelvic rotation. According to the cluster analysis results, 25% of the participants were classified into Cluster 1, 9% into Cluster 2, 49% in Cluster 3, and 17% in Cluster 4. INTERPRETATION: Three different postural patterns emerged in

accordance with the sagittal postural profiles in children with bilateral cerebral palsy and were defined as follows: 1) a lordotic postural pattern corresponding to forward-leaning posture; 2) a swayback postural pattern corresponding to backward-leaning posture; and 3) a balanced postural pattern corresponding to balanced posture.

PMID: 28830044

3. Outcomes of intrathecal baclofen therapy in patients with cerebral palsy and acquired brain injury.

Yoon YK, Lee KC, Cho HE, Chae M, Chang JW, Chang WS, Cho SR.

Medicine (Baltimore). 2017 Aug;96(34):e7472. doi: 10.1097/MD.0000000000007472.

Intrathecal baclofen (ITB) has been known to reduce spasticity which did not respond to oral medications and botulinum toxin treatment. However, few results have been reported comparing the effects of ITB therapy in patients with cerebral palsy (CP) and acquired brain injury. This study aimed to investigate beneficial and adverse effects of ITB bolus injection and pump therapy in patients with CP and to compare outcomes to patients with acquired brain injury such as traumatic brain injury and hypoxic brain injury. ITB test trials were performed in 37 patients (19 CP and 18 acquired brain injury). Based on ambulatory function, CP patients were divided into 2 groups: 11 patients with nonambulatory CP and 8 patients with ambulatory CP. Change of spasticity was evaluated using the Modified Ashworth Scale. Additional positive or negative effects were also evaluated after ITB bolus injection. In patients who received ITB pump implantation, outcomes of spasticity, subjective satisfaction and adverse events were evaluated until 12 months post-treatment. After ITB bolus injection, 32 patients (86.5%) (CP 84.2% versus acquired brain injury 88.9%) showed a positive response of reducing spasticity. However, 8 patients with CP had negative adverse effects. Particularly, 3 ambulatory CP patients showed standing impairment and 1 ambulatory CP patient showed impaired gait pattern such as foot drop because of excessive reduction of lower extremity muscle tone. Ambulatory CP patients received ITB pump implantation less than patients with acquired brain injury after ITB test trials (P=.003 by a chi-squared test). After the pump implantation, spasticity was significantly reduced within 1 month and the effect maintained for 12 months. Seventeen patients or their caregivers (73.9%) were very satisfied, whereas 5 patients (21.7%) suffered from adverse events showed no subjective satisfaction. In conclusion, ITB therapy was effective in reducing spasticity in patients with CP and acquired brain injury. Before ITB pump implantation, it seems necessary to perform the ITB bolus injection to verify beneficial effects and adverse effects especially in ambulatory CP.

PMID: 28834868

4. Analysis of gait patterns pre- and post- Single Event Multilevel Surgery in children with Cerebral Palsy by means of Offset-Wise Movement Analysis Profile and Linear Fit Method.

Ancillao A, van der Krogt MM, Buizer AI, Witbreuk MM, Cappa P, Harlaar J.

Hum Mov Sci. 2017 Aug 19;55:145-155. doi: 10.1016/j.humov.2017.08.005. [Epub ahead of print]

Gait analysis is used for the assessment of walking ability of children with cerebral palsy (CP), to inform clinical decision making and to quantify changes after treatment. To simplify gait analysis interpretation and to quantify deviations from normality, some quantitative synthetic descriptors were developed over the years, such as the Movement Analysis Profile (MAP) and the Linear Fit Method (LFM), but their interpretation is not always straightforward. The aims of this work were to: (i) study gait changes, by means of synthetic descriptors, in children with CP that underwent Single Event Multilevel Surgery; (ii) compare the MAP and the LFM on these patients; (iii) design a new index that may overcome the limitations of the previous methods, i.e. the lack of information about the direction of deviation or its source. Gait analysis exams of 10 children with CP, pre- and post-surgery, were collected and MAP and LFM were computed. A new index was designed asa modified version of the MAP by separating out changes in offset (named OC-MAP). MAP documented an improvement in the gait pattern after surgery. The highest effect was observed for the knee flexion/extension angle. However, a worsening was observed as an increase in anterior pelvic tilt. An important source of gait deviation was recognized in the offset between observed tracks and reference. OC-MAP allowed the assessment of the offset component versus the shape component of deviation. LFM provided results similar to OC-MAP offset analysis but could not be considered reliable due to intrinsic limitations. As offset in gait features played an important role in gait deviation, OC-MAP synthetic analysis was proposed as a novel approach to a meaningful parameterisation of global deviations in gait patterns of subjects with CP and gait changes after treatment.

PMID: 28829950

5. Team Approach: Single-Event Multilevel Surgery in Ambulatory Patients with Cerebral Palsy.

Georgiadis AG1, Schwartz MH, Walt K, Ward ME, Kim PD, Novacheck TF.

JBJS Rev. 2017 Aug 22. doi: 10.2106/JBJS.RVW.16.00101. [Epub ahead of print]

[No abstract available]

PMID: 28832346

6. Comment on Silverio et al: Proximal femur prosthetic interposition arthroplasty for painful dislocated hips in children with cerebral palsy.

El-Sobky TA, Mahran M.

J Child Orthop. 2017 Jun 1;11(3):237-238. doi: 10.1302/1863-2548.11.170007.

[Commentary on Silverio et al]

PMID: 28828069

7. Response to Comment on Silverio et al: Proximal femur prosthetic interposition arthroplasty for painful dislocated hips in children with cerebral palsy.

Silverio AL, Nguyen SV, Schlechter JA, Rosenfeld SR.

J Child Orthop. 2017 Jun 1;11(3):239. doi: 10.1302/1863-2548.11.170034.

We respect and appreciate the readers' letter to the editor regarding our study. They make valid points. With that being said providing hip stability for these children was not a goal of this procedure nor was there any attempt to reduce the endoprosthesis into the acetabulum. The endoprosthesis was placed as an interposition material with imbricated capsule with an ultimate goal of providing more painless range of motion and was intended to decrease migration. The endoprosthesis also helped prevent heterotopic ossification, a recognized complication of resection arthroplasty of the hip. In addition, we noted one of our article's limitations was its retrospective nature and use on a non-validated outcome score modeled after that of the one utilized in the article by Wright et al. We feel that we were forth coming with our results albeit mixed and reporting of our outcomes and complications in this fragile population. Again we thank the readers for their comments and hopefully this response clarifies some of their concerns.

PMID: 28828070

8. Combined pelvic and femoral reconstruction in children with cerebral palsy.

Alassaf N, Saran N, Benaroch T, Hamdy RC.

J Int Med Res. 2017 Jan 1:300060517723797. doi: 10.1177/0300060517723797. [Epub ahead of print]

Objective The primary aim of this study was to determine the effect of age, femoral head migration, and ambulatory status on radiographic outcomes after combined pelvic and femoral reconstruction in children with cerebral palsy. The secondary aim was to evaluate the fate of the opposite hip after unilateral reconstruction. Methods A retrospective cohort study design of consecutive patients from 1995-2009 was used. The records were screened for patients who underwent varus derotational osteotomy and modified Dega osteotomy. Results Eighty-five hips in 71 patients were included. The mean age was 8.4 ± 3.2 years and the mean follow-up was 6.6 ± 3.1 years. The final measures were a mean migration index of 20% ± 15.58%, centre edge angle of 28.45° ± 15.98°, and Sharp's angle of 40.75° ± 8.5°. Those values were not correlated with age and the initial migration index. Nonambulatory status did not negatively affect hip stability. Final measurements of the contralateral hips were similar to the reconstructed hips, and the cumulative incidence for later reconstruction was 5.67%. Conclusions Regardless of

age, preoperative displacement, and ambulation, the combined procedure provides durable radiographic improvement. In unilateral cases, there is a low risk of later deterioration of the opposite side.

PMID: 28823214

9. Transcranial direct current stimulation as a motor neurorehabilitation tool: an empirical review.

Sánchez-Kuhn A, Pérez-Fernández C, Cánovas R, Flores P, Sánchez-Santed F.

Biomed Eng Online. 2017 Aug 18;16(Suppl 1):76. doi: 10.1186/s12938-017-0361-8.

The present review collects the most relevant empirical evidence available in the literature until date regarding the effects of transcranial direct current stimulation (tDCS) on the human motor function. tDCS in a non-invasive neurostimulation technique that delivers a weak current through the brain scalp altering the cortical excitability on the target brain area. The electrical current modulates the resting membrane potential of a variety of neuronal population (as pyramidal and gabaergic neurons); raising or dropping the firing rate up or down, depending on the nature of the electrode and the applied intensity. These local changes additionally have shown long-lasting effects, evidenced by its promotion of the brain-derived neurotrophic factor. Due to its easy and safe application and its neuromodulatory effects, tDCS has attracted a big attention in the motor neurorehabilitation field among the last years. Therefore, the present manuscript updates the knowledge available about the main concept of tDCS, its practical use, safety considerations, and its underlying mechanisms of action. Moreover, we will focus on the empirical data obtained by studies regarding the application of tDCS on the motor function of healthy and clinical population, comprising motor deficiencies of a variety of pathologies as Parkinson's disease, stroke, multiple sclerosis and cerebral palsy, among others. Finally, we will discuss the main current issues and future directions of tDCS as a motor neurorehabilitation tool.

PMID: 28830433

10. Transient Changes in Brain Metabolites after Transcranial Direct Current Stimulation in Spastic Cerebral Palsy: A Pilot Study.

Auvichayapat P, Aree-Uea B, Auvichayapat N, Phuttharak W, Janyacharoen T, Tunkamnerdthai O, Boonphongsathian W, Ngernyam N, Keeratitanont K.

Front Neurol. 2017 Jul 31;8:366. doi: 10.3389/fneur.2017.00366. eCollection 2017.

BACKGROUND: Muscle spasticity is a disability caused by damage to the pyramidal system. Standard treatments for spasticity include muscle stretching, antispastic medications, and tendon release surgeries, but treatment outcomes remain unsatisfactory. Anodal transcranial direct current stimulation (tDCS) in patients with muscle spasticity is known to result in significant improvement in spastic tone ($p < 0.001$). However, the mechanism of action by which tDCS treatment affects spasticity remains unclear. This pilot study aimed to investigate the effect of anodal tDCS upon brain metabolites in the left basal ganglia and ipsilateral primary motor cortex (M1) in children with spastic cerebral palsy (CP). MATERIALS AND METHODS: This study consisted of three steps: a baseline evaluation, a treatment period, and a follow-up period. During the treatment period, patients were given 20 min of 1 mA anodal tDCS over the left M1 for five consecutive days. Outcomes were compared between pre- and immediate posttreatment in terms of brain metabolites, Tardieu scales, and the quality of upper extremity skills test. RESULTS: Ten patients with spastic CP were enrolled. Following tDCS, there were significant increases in the ratio of N-acetylaspartate (NAA)/creatine (Cr) ($p = 0.030$), choline (Cho)/Cr ($p = 0.043$), and myoinositol (mI)/Cr ($p = 0.035$) in the basal ganglia. Moreover, increased glutamine-glutamate (Glx)/Cr ratio in the left M1 ($p = 0.008$) was found. In addition, we also observed improvements in the extent of spasticity and hand function ($p = 0.028$). CONCLUSION: Five consecutive days of anodal tDCS over the left M1 appeared statistically to reduce the degree of spasticity and increase NAA, Cho, mI, and Glx. Future research studies, involving a larger sample size of spastic CP patients undergoing tDCS is now warranted.

PMID: 28824525

11. Basics of bone metabolism and osteoporosis in common pediatric neuromuscular disabilities.

Yaşar E, Adigüzel E, Arslan M, Matthews DJ.

Eur J Paediatr Neurol. 2017 Aug 10. pii: S1090-3798(17)30236-2. doi: 10.1016/j.ejpn.2017.08.001. [Epub ahead of print]

Bone modeling is a process that starts with fetal life and continues during adolescence. Complex factors such as hormones, nutritional and environmental factors affect this process. In addition to these factors, physical conditioning and medications that have toxic effects on bony tissue should be carefully considered in patient follow-up. Osteoporosis is a significant problem in pediatric population because of ongoing growth and development of skeletal system. Two types of osteoporosis are primary and secondary types and children with neuromuscular disabilities constitute a major group with secondary osteoporosis. Low bone mass in patients with cerebral palsy, spina bifida, and Duchenne muscular dystrophy cause increased bone fragility in even slight traumas. Maximizing peak bone mass and prevention of bone loss are very important to reduce the fracture risk in neuromuscular diseases. This article aims to review the determinants of bone physiology and bone loss in children with cerebral palsy, spina bifida, and Duchenne muscular dystrophy.

PMID: 28830650

12. A lower-extremity exoskeleton improves knee extension in children with crouch gait from cerebral palsy.

Lerner ZF, Damiano DL, Bulea TC.

Sci Transl Med. 2017 Aug 23;9(404). pii: eaam9145. doi: 10.1126/scitranslmed.aam9145. Epub 2017 Aug 23.

The ability to walk contributes considerably to physical health and overall well-being, particularly in children with motor disability, and is therefore prioritized as a rehabilitation goal. However, half of ambulatory children with cerebral palsy (CP), the most prevalent childhood movement disorder, cease to walk in adulthood. Robotic gait trainers have shown positive outcomes in initial studies, but these clinic-based systems are limited to short-term programs of insufficient length to maintain improved function in a lifelong disability such as CP. Sophisticated wearable exoskeletons are now available, but their utility in treating childhood movement disorders remains unknown. We evaluated an exoskeleton for the treatment of crouch (or flexed-knee) gait, one of the most debilitating pathologies in CP. We show that the exoskeleton reduced crouch in a cohort of ambulatory children with CP during overground walking. The exoskeleton was safe and well tolerated, and all children were able to walk independently with the device. Rather than guiding the lower limbs, the exoskeleton dynamically changed the posture by introducing bursts of knee extension assistance during discrete portions of the walking cycle, a perturbation that resulted in maintained or increased knee extensor muscle activity during exoskeleton use. Six of seven participants exhibited postural improvements equivalent to outcomes reported from invasive orthopedic surgery. We also demonstrate that improvements in crouch increased over the course of our multiweek exploratory trial. Together, these results provide evidence supporting the use of wearable exoskeletons as a treatment strategy to improve walking in children with CP.

PMID: 28835518

13. Longitudinal change in foot posture in children with cerebral palsy.

Church C, Lennon N, Alton R, Schwartz J, Niiler T, Henley J, Miller F.

J Child Orthop. 2017 Jun 1;11(3):229-236. doi: 10.1302/1863-2548.11.160197.

PURPOSE: Foot deformities are common in children with cerebral palsy (CP), yet the evolution of such deformities is not well documented. We aimed to observe and analyse changes in foot posture during growth in children with CP. Methods We followed 51 children (16 unilateral, 35 bilateral; 37 Gross Motor Function Classification Scale (GMFCS) I/II, 14 III/IV) aged two to 12 years in this level II, IRB-approved prospective longitudinal study. Data after bony foot corrections were excluded. Outcome measures included coronal plane pressure index (CPPI) and pressure impulses from the heel, medial midfoot and medial forefoot. Data were LOESS smoothed and resulting models were compared for significant differences across time using a derived FANOVA method. RESULTS: The GMFCS I/II group had more foot valgus than typically developing (TD) children until seven years which normalised thereafter. From two to 12 years, GMFCS III/IV children had more foot valgus than TD children. Heel impulse was significantly reduced in both GMFCS groups compared with TD children, and the III/IV group had less heel contact than the I/II group. CONCLUSIONS: Due to early variability and the tendency for resolving valgus foot posture in children with CP, conservative management of coronal plane foot deformity is suggested in early childhood, especially for children classified as GMFCS I and II.

PMID: 28828068

14. Efficacy of Repeated Botulinum Toxin Type A Injections for Spastic Equinus in Children with Cerebral Palsy-A Secondary Analysis of the Randomized Clinical Trial.

Hong BY, Chang HJ, Lee SJ, Lee S, Park JH, Kwon JY.

Toxins (Basel). 2017 Aug 21;9(8). pii: E253. doi: 10.3390/toxins9080253.

Botulinum toxin A is considered an important tool to control spasticity in children with cerebral palsy. Several factors are known to affect the efficacy of botulinum toxin, such as dosage, appropriate muscle selection and application, age, and accompanying therapy. A multicenter, double-blind, randomized, prospective phase III clinical trial of botulinum toxin A for the treatment of dynamic equinus in 144 children with cerebral palsy was performed to compare the efficacies of letibotulinumtoxin A and onabotulinumtoxin A. Secondary analyses were performed to evaluate factors that affected the outcome, focusing on the number of times injections were repeated. Effectiveness was defined as a change of 2 or more in the physician's rating scale. Multivariate regression analyses were performed with multiple variables. The first injection of botulinum toxin A significantly improved D subscale of Gross Motor Function Measure-88 scores at 3 months compared to repeated injections ($p < 0.05$). After 6 months, patients who had one injection or none before the study showed significantly better outcomes than those who had more than one injection in terms

PMID: 28825663

15. The effect of a running intervention on running ability and participation in children with cerebral palsy: a randomized controlled trial.

Gibson N, Chappell A, Blackmore AM, Morris S, Williams G, Bear N, Allison G.

Disabil Rehabil. 2017 Aug 21:1-9. doi: 10.1080/09638288.2017.1367426. [Epub ahead of print]

PURPOSE: The purpose of this study is to evaluate effects of a running intervention on running ability and participation in children with cerebral palsy. MATERIALS AND METHOD: Children with cerebral palsy (9-18 years) with Gross Motor Function Classification System levels I-III were randomly assigned to a 12-week running intervention or usual care. Primary outcomes included improvement in running ability (assessed by Goal Attainment Scaling, high level mobility (assessed by the High-Level Mobility Assessment Tool) and participation (assessed by the Participation and Environment Measure for Children and Youth). Secondary outcomes were aerobic and anaerobic fitness and agility. Blinded assessments took place at baseline and 12 weeks. Regression analysis adjusting for baseline differences was used to determine between group differences. RESULTS: Forty-two participants (mean age 12.5 years, SD 2.8 years; 15 female) completed the study. Statistically significant group differences at 12-weeks were found for improvements in running ability (86% treatment group versus 0% control group achieved or exceeded their running goals, $p < 0.001$), and participation in the school environment (Participation and Environment Measure mean difference 1.18: 95%CI 1.00-1.39, $p = 0.045$). CONCLUSION: A 12-week individualized running training skills intervention results in achievement of running ability goals and participation in the school environment in children with cerebral palsy. Implications for Rehabilitation Children with cerebral palsy who can walk unaided demonstrate impairments in higher level mobility such as running. Running is a motor skill that can be trained in children with cerebral palsy. Individually tailored running skills intervention, delivered in a group context can improve goal-identified running ability and translate into a higher frequency of participation in school activities.

PMID: 28826274

16. Physical activity interventions for children and young people with cerebral palsy.

Gorter JW.

Dev Med Child Neurol. 2017 Aug 21. doi: 10.1111/dmcn.13550. [Epub ahead of print]

[This commentary is on the systematic reviews by Reedman et al.]

PMID: 28832995

17. Emergency department visits for children with cerebral palsy: what triggers the call for help?

Murphy N.

Dev Med Child Neurol. 2017 Aug 23. doi: 10.1111/dmcn.13531. [Epub ahead of print]

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

PMID: 28832991

18. Parenting stress in caregivers of children with chronic physical condition-A meta-analysis.

Pinquart M.

Stress Health. 2017 Aug 18. doi: 10.1002/smi.2780. [Epub ahead of print]

On the basis of the parenting stress model we compared levels of parenting stress in families with and without a child with a chronic physical condition and analysed correlates of parenting stress in families with a child with a chronic condition. A systematic search through electronic databases identified 547 relevant studies that were included in a random-effects meta-analysis. Parents of children with a chronic condition showed small to moderate elevations of general parenting stress and stress related to the parent-child relationship in particular. They showed moderate to large elevations in health-related parenting stress. Parents of children with cancer, cerebral palsy, HIV infection or AIDS, and spina bifida showed the highest levels of parenting stress. Stress levels also varied by illness severity and duration, child age, parental gender and mental health, marital status, marital quality, and levels of perceived support. Behaviour problems of the child and low parental mental health were the strongest correlates of parenting stress. The present results assist with identifying parents at highest needs for interventions aimed at reducing parenting stress. These interventions should address the reduction of child behaviour problems, the promotion of parental mental health, the increase in marital quality and social support in general, and skills for dealing with stressors.

PMID: 28834111

19. Pre-surgery evaluations by telephone decrease travel and cost for families of children with cerebral palsy.

Robinson JD, Prochaska JD, Yngve DA.

SAGE Open Med. 2017 Jul 23;5:2050312117720046. doi: 10.1177/2050312117720046. eCollection 2017.

INTRODUCTION: Children with cerebral palsy need highly specialized care. This can be very burdensome for families, particularly in large rural states, due to the need for long-distance travel to appointments. In this study, children undergoing the selective percutaneous myofascial lengthening surgery utilized a telephone-based telemedicine evaluation to assess for surgical eligibility. The goal was to avoid a separate preoperative clinic visit weeks before the surgery. If possible, eligibility was determined by telephone, and then, the patient could be scheduled for a clinic visit and possible surgery the next day, saving the family a trip. The purposes of the study were to calculate estimated reductions in miles traveled, in travel expenses, and in carbon emissions and to determine whether the telephone assessment was accurate and effective in determining eligibility for surgery. METHODS: From 2010 to 2012, 279 patients were retrospectively reviewed, and of those, 161 mailed four-page questionnaire and anteroposterior pelvis X-ray followed by a telephone conference. Geographic information system methods were used to geocode patients by location. Savings in mileage and travel costs were calculated. From 2014 to 2015, 195 patients were additionally studied to determine accuracy and effectiveness. RESULTS: The telephone prescreening method saved 106,070 miles in transportation over 3 years, a 38% reduction with US$55,326 in savings. Each family saved an average of 658 (standard deviation = 340) miles of travel and US$343.64 (standard deviation = US$178) in travel expenses. For each increase of 10 miles in distance from the health center, the odds of a patient utilizing telephone screening increased by 10% (odds ratio: 1.101, 95% confidence interval: 1.073-1.129, $p < 0.001$). After the telephone prescreening, 86% were determined to be likely candidates for the procedure. For 14%, a clinic visit only was scheduled, and they were not scheduled for surgery. CONCLUSION: Families seeking specialized surgical care for their disabled children particularly benefited from this approach.

PMID: 28839936

20. Health self-management, transition readiness and adaptive behavior in persons with cerebral palsy or myelomeningocele.

Warschausky S, Kaufman JN, Schutt W, Evitts M, Hurvitz EA.

Rehabil Psychol. 2017 Aug;62(3):268-275. doi: 10.1037/rep0000157.

PURPOSE/OBJECTIVE: This study was conducted to examine the associations between generic and condition-specific health self-management and levels of adaptive behavior in 2 groups of transition-age youth with congenital neurodevelopmental conditions. METHOD: The sample included 43 adolescents/young adults diagnosed with cerebral palsy (CP) and 36 with spina bifida/myelomeningocele (SBM), ages 13-29, mean age 18.96 (4.77), 51.9% female. Health self-management was assessed with the Transition Readiness Assessment Questionnaire (TRAQ) completed by the child, and the Kennedy Krieger Independence Scales-Spina Bifida (KKIS-SB) completed by the parent/guardian. The Adaptive Behavior Assessment System (ABAS-II) completed by the parent/guardian was used to assess levels of adaptive behavior. RESULTS: There were significant group differences in condition-specific health self-management, including lower KKIS-SB Initiation of Routines and KKIS-SB Prospective Memory scores in the group with SBM. Those differences were no longer significant when scoring was modified to account for item applicability. Group differences in generic health self-management and adaptive behavior were not significant. There were significant differences in the correlations between health self-management instruments and ABAS-II composite scores. CONCLUSIONS: For youth with congenital neurodevelopmental conditions who are in the transition to adulthood, there are important condition-specific self-management needs that are not captured by measuring generic transition readiness or adaptive behavior. Findings highlight the need for clinicians to assess health self-management needs from multiple perspectives, utilizing generic and condition-specific measures that can inform targeted interventions and supports for optimal independence. (PsycINFO Database Record

PMID: 28836807

21. Multiple stakeholder perceptions of assistive technology for individuals with cerebral palsy in New Zealand.

Taherian S, Davies C.

Disabil Rehabil Assist Technol. 2017 Aug 23:1-10. doi: 10.1080/17483107.2017.1369585. [Epub ahead of print]

BACKGROUND: This study sought to gain an understanding of the experiences and perspectives of assistive technology from different stakeholders in technology adoption, in the New Zealand context. METHODS: A focus group was held with individuals with cerebral palsy (n = 5), service providers (n = 4), caregivers (n = 3) and a biomechanical engineer. The data recordings from the focus group were transcribed and coded using thematic analysis. RESULTS: Themes emerged around barriers imposed by the assessment process and training in assistive technology procedures, the influence of family members, the environment that assistive technology is used in, and psychosocial aspects of being able to participate and integrate into society. CONCLUSION: The results are similar to other literature, suggesting new innovations and changes are in dire need, to improve assistive technology experiences for all stakeholders. Implications for Research Service providers for assistive technology desire more effective training and support of existing and emerging technologies. Although the set procedure for acquiring assistive technology in New Zealand is comprehensive, incorporating multiple perspectives, it is difficult to follow through in practice. More innovative procedures are needed. The movement of Universal Design is significantly improving the perception of individuals with disabilities, and has enabled greater social inclusion. More assistive technology developers need to ensure that they incorporate these principles in their design process.

PMID: 28830261

Prevention and Cure

22. Adjuvant treatment with monosialoganglioside may improve neurological outcomes in neonatal hypoxic-ischemic encephalopathy: A meta-analysis of randomized controlled trials.

Sheng L, Li Z.

PLoS One. 2017 Aug 23;12(8):e0183490. doi: 10.1371/journal.pone.0183490. eCollection 2017.

BACKGROUND: Ganglioside has a neuroprotective role in neonatal hypoxic-ischemic encephalopathy (HIE). This study aimed to evaluate the neurological outcomes of monosialoganglioside as adjuvant treatment for neonatal HIE by conducting a meta-analysis. METHODS: A comprehensive literature search was made in the Pubmed, EMBASE, Cochrane Library, Wanfang, CNKI, VIP databases through October 2016. Randomized controlled trials comparing monosialoganglioside with the usual treatment for newborns having HIE deemed eligible. Weighted mean difference (WMD) and risk ratio (RR) with 95% confidence interval (CI) were calculated for continuous and dichotomous data, respectively. RESULTS: Ten trials consisting of 787 neonates were included. Adjuvant treatment with monosialoganglioside significantly reduced major neurodevelopmental disabilities (RR = 0.35; 95% CI = 0.21-0.57), cerebral palsy (RR = 0.32; 95% CI = 0.12-0.87), mental retardation (RR = 0.31; 95% CI = 0.11-0.88) as well as improved the mental (WMD = 14.95; 95% CI = 7.44-22.46) and psychomotive (WMD = 13.40; 95% CI = 6.69-20.11) development index during the follow-up. Also, monosialoganglioside significantly improved Neonatal Behavioral Neurological Assessment scores (WMD = 2.91; 95% CI = 2.05-3.78) compared with the usual treatment. However, adverse effects associated with monosialoganglioside were poorly reported in the included trials. CONCLUSION: Adjuvant treatment with monosialoganglioside had beneficial effects in improving neurological outcomes in neonatal HIE. However, these findings should be interpreted with caution because of methodological flaws in the included trials. Furthermore, safety of monosialoganglioside use should also be further evaluated.

PMID: 28832625

23. Survival and neurodevelopmental outcomes in extremely preterm infants: 22-24 weeks of gestation born in Western Australia.

Sharp M, French N, McMichael J, Campbell C.

J Paediatr Child Health. 2017 Aug 24. doi: 10.1111/jpc.13678. [Epub ahead of print]

AIM: The management of births at borderline viability continues to present dilemmas for health professionals and parents. The aim of the study was to review local outcomes of infants born between 22 and 24 weeks of gestation between 2004 and 2010 in Western Australia (WA) to aid perinatal counselling. METHODS: Survival data for the study were sourced retrospectively from the Neonatal Clinical Care Unit and Department of Health records of births in WA. Neurodevelopmental follow-up outcomes were assessed using the most recent standardised assessment (Griffiths, Bayley-III and Wechsler Preschool and Primary Scale of Intelligence, 3rd Ed) and medical examination of infants/children 12 months to 8 years from follow-up clinic appointments. RESULTS: At these gestations, 159 survivors represented 72% of neonatal intensive care unit admissions, 53% of WA live births and 26% of WA live and still births; 5% of live births survived at 22 weeks, 46% at 23 weeks and 77% at 24 weeks. Of the 14 outborn/retrieved infants, 4 survived (29%). At a median age of 59 months, disabilities were severe in 13% of children (22-23w = 19%; 24w = 11%). The median test quotient was 90. Moderate and severe cognitive disability was found in 16%, cerebral palsy was found in 7% (n = 11), and 55% of children were free from impairment as defined in this study. CONCLUSION: At these gestations, survival figures varied markedly with the chosen population denominator. Regional data are essential for valid population comparison. While many developmental difficulties occurred in these children, 78% were free from moderate or severe impairment at ages 3-5 years.

PMID: 28836705

24. Postnatal hypoxia evokes persistent changes within the male rat's dopaminergic system.

Decker MJ, Jones KA, Keating GL, Rye DB.

Sleep Breath. 2017 Aug 22. doi: 10.1007/s11325-017-1558-6. [Epub ahead of print]

PURPOSE: Hypoxic insults occurring during the perinatal period remain the leading cause of permanent brain impairment. Severe cognitive and motor dysfunction, as seen in cerebral palsy, will occur in 4-10% of post-hypoxic newborns. Subtle

cognitive impairment, apparent in disorders of minimal brain dysfunction will occur in > 3 million post-hypoxic newborns. Analyses of post-hypoxic rodent brains reveal reduced extracellular levels of dopamine, a key neurotransmitter of vigilance, execute function, and behavior. The purpose of this study was to assess whether synaptic levels of dopamine could be enhanced in post-hypoxic, hypodopaminergic rats. METHODS: Newborn male rats were exposed to subtle, repetitive hypoxic insults for 4-6 h per day, during postnatal days 7-11. During adolescence, we quantified dopamine content within the caudate nuclei. We then determined whether extracellular dopamine levels could be increased by injecting the psychostimulant d-amphetamine. We next assessed whether the post-hypoxic rat's response to d-amphetamine would differentially impact place preference behavior when compared with littermate controls. RESULTS: Total tissue content of dopamine was significantly higher in post -hypoxic rats. Injection of d-amphetamine liberated that dopamine which subsequently enhanced extracellular levels. Post-hypoxic rats acquired conditioned place preference for d-amphetamine during the training days. During the testing day, total time spent in the amphetamine-pairing box did not differ between post-hypoxic and control littermates. CONCLUSION: Postnatally occurring hypoxic insults promote remodeling of the dopaminergic system resulting in increased intracellular sequestering of this monoamine. That sequestered dopamine can be released using the psychostimulant d-amphetamine, which did not promote a conditioned place preference any greater than was observed in non-hypoxic littermate controls.

PMID: 28828549

25. Epilepsy, ataxia, sensorineural deafness, tubulopathy syndrome in a European child with KCNJ10 mutations: A case report.

Papavasiliou A, Foska K, Ioannou J, Nagel M.

SAGE Open Med Case Rep. 2017 Jul 27;5:2050313X17723549. doi: 10.1177/2050313X17723549. eCollection 2017.

BACKGROUND: Epilepsy, ataxia, sensorineural deafness, tubulopathy syndrome is a multi-organ disorder that links to autosomal recessive mutations in the KCNJ10 gene, which encodes for the Kir4.1 potassium channel. It is mostly described in consanguineous, non-European families. CASE REPORT: A European male of non-consanguineous birth, with early-onset, static ataxic motor disorder, intellectual disability and epilepsy, imitating cerebral palsy, presented with additional findings of renal tubulopathy, sensorineural deafness and normal neuroimaging leading to the diagnosis of epilepsy, ataxia, sensorineural deafness, tubulopathy syndrome. The patient was heterozygous for two KCNJ10 mutations: a missense mutation (p.R65C) that is already published and a not yet published duplication (p.F119GfsX25) that creates a premature truncation of the protein. Both mutations are likely damaging. Parental testing has not been performed, and therefore, we do not know for certain whether the mutations are on different alleles. This young man presents some clinical and laboratory features that differ from previously reported patients with epilepsy, ataxia, sensorineural deafness, tubulopathy syndrome. CONCLUSION: The necessity of accurate diagnosis through genetic testing in patients with static motor disorders resembling cerebral palsy phenotypes, atypical clinical features and noncontributory neuroimaging is emphasized.

PMID: 28835827

26. Neonatal cerebral lesions predict 2-year neurodevelopmental impairment in children treated with laser surgery for twin-twin transfusion syndrome.

Chmait RH, Chon AH, Schrager SM, Llanes A, Hamilton AH, Vanderbilt DL.

J Matern Fetal Neonatal Med. 2017 Aug 24:1-10. doi: 10.1080/14767058.2017.1371694. [Epub ahead of print]

OBJECTIVE: To assess whether postnatally detected cerebral abnormalities are predictive of neurodevelopmental impairment (NDI) in survivors of twin-twin transfusion syndrome (TTTS) that underwent laser surgery. MATERIALS AND METHODS: 99 children treated for TTTS had neurodevelopmental assessment at age 2-years (± 6 weeks). "High-risk survivors" had cerebral imaging in the neonatal period. "High-risk survivors" were defined as: 1) delivered at < 32 weeks; or 2) cerebral imaging clinically indicated. NDI was a composite outcome of: Battelle Developmental Inventory 2nd edition (BDI-2) score < 70, cerebral palsy, blindness, and/or deafness. Multilevel logistic regression with robust standard errors was used to evaluate associations between cerebral lesions and NDI. RESULTS: Fifty-six children were "high-risk survivors" and had neonatal cerebral imaging. Ten twins (18%) had at least one cerebral lesion, including grade 1-2 intraventricular hemorrhage (8), cystic periventricular leukomalacia (2), ventriculomegaly (1), and bilateral subependymal cyst (1). The risk of NDI in the "high-risk survivors" was 7% (4/56) compared to 0% (0/43) in the remaining group. Among "high-risk survivors", cerebral lesions were a significant risk factor for NDI (OR = 19.28, $p < 0.001$). CONCLUSIONS: Among "high-risk survivors" of TTTS treated with laser surgery, cerebral lesions identified on neonatal imaging were associated with NDI at 2-years.

PMID: 28835143

27. Neural substrate and clinical significance of general movements: an update.

Hadders-Algra M.

Dev Med Child Neurol. 2017 Aug 23. doi: 10.1111/dmcn.13540. [Epub ahead of print]

General movements are present from early fetal life to 3 to 5 months corrected age. Atypical general movements, especially in the last, so-called fidgety general movement phase, are predictive of cerebral palsy (CP). This review updates knowledge on the neural substrate and clinical significance of typical and atypical general movements. Typical general movements are primarily characterized by complexity and variation. Presumably these core characteristics are initially induced by modulating activity of the cortical subplate. When the subplate gradually dissolves between 3 months before term and 3 months corrected age the cortical plate takes over. This coincides with the fidgety general movement phase. Conceivably, fidgety activity reflects 'sparsification', i.e. fragmentation of cortical network activity. The quintessential feature of atypical general movements is reduced complexity and variation. This is attributed to impaired integrity of extensive cortical-subcortical networks, in which the subplate and periventricular white matter play a prominent role. The most serious forms of network impairment are associated with absent fidgety movements.

PMID: 28832987

28. Missense mutation in the ITPR1 gene presenting with ataxic cerebral palsy: Description of an affected family and literature review.

Das J, Lilleker J, Shereef H, Ealing J.

Neurol Neurochir Pol. 2017 Jul 8. pii: S0028-3843(17)30009-9. doi: 10.1016/j.pjnns.2017.06.012. [Epub ahead of print]

The inositol 1,4,5-triphosphate receptor type 1 (ITPR1) gene on chromosome 3 belongs to a family of genes encoding intracellular calcium channel proteins. Such channels are located primarily within the endoplasmic reticular membrane and release Ca2+, an intracellular messenger, which governs numerous intracellular and extracellular functions. We report a family with infantile-onset cerebellar ataxia with delayed motor development and intellectual disability caused by a heterozygous c.805C>T, p.Arg269Trp missense mutation in ITPR1. Both affected family members had postural tremor, hypotonia and dysarthria, but neither had pyramidal signs. Their neuroimaging revealed cerebellar atrophy. Several neurological conditions have been associated with ITPR1 mutations, such as spinocerebellar ataxia type 15 and Gillespie syndrome, and the phenotype may vary according to the location and type of mutations. Spinocerebellar ataxia type 15 is an autosomal dominant disorder, which causes late onset pure cerebellar ataxia. Gillespie syndrome is characterised by bilateral iris hypoplasia, congenital hypotonia, non-progressive ataxia and cerebellar atrophy. In this report, we provide a detailed phenotypic description of a family with a missense mutation in ITPR1. This mutation has only been reported once before. We also provide a literature review of the various phenotypes associated with ITPR1 gene.

PMID: 28826917

29. The Ethics of Teaching Physicians Electronic Fetal Monitoring: And Now for the Rest of the Story.

Sartwelle TP, Johnston JC, Arda B.

Surg J (N Y). 2017 Mar 20;3(1):e42-e47. doi: 10.1055/s-0037-1599229. eCollection 2017 Jan.

Electronic fetal monitoring (EFM) does not predict or prevent cerebral palsy (CP), but this myth remains entrenched in medical training and practice. The continued use of this ineffectual diagnostic modality increases the cesarean section rate with concomitant harms to mothers and babies alike. EFM, as it is used in defensive medical practice, is a violation of patient autonomy and raises serious ethical concerns. This review addresses the need for improved graduate medical education so that physicians and medical residents are taught both sides of the EFM-CP story.

PMID: 28825019

30. [Predictive effect of neonatal morbidities on the poor outcomes at 12 months corrected age in very low birth weight premature infants].

[Article in Chinese; Abstract available in Chinese from the publisher]

Rao YB, Yang J, Cao B, Chen DM, Gao PM, Zhong Q, Li MX, Gao JH, Chen YJ, Zhong XM, Ren ZX.

Zhonghua Er Ke Za Zhi. 2017 Aug 2;55(8):608-612. doi: 10.3760/cma.j.issn.0578-1310.2017.08.012.

Objective: To investigate the prognostic effect of neonatal morbidities on poor outcomes at 12 months corrected age in very low birth weight (VLBW) premature infants . Method: From November 2013 to October 2014, a multi-center retrospective study was conducted in 8 tertiary Maternal and Children's hospitals in Guangdong, Hunan and Fujian. The premature infants survived to a postmenstrual age (PMA) of 36 weeks with birth weight less than 1 500 g and without congenital diseases were included, and divided into two groups according to poor outcomes. The birth weight, gestational age, morbidities and poor outcomes (death, cerebral palsy, cognitive delay, et al) were recorded. Data were analyzed with Chi-square test to investigate the relationship between morbidities and poor outcomes. And the predictive effect of the top three morbidities were analyzed by Logistic regression analysis. Result: Total of 834 VLBW premature infants (473 boys and 361 girls) finished the follow-up, whose average gestational age and birth weight were (30.6±1.8) weeks and (1 189±159)g. The incidences of BPD, severe ROP, NEC, brain injury and sepsis were 207 (24.8%), 119 (14.3%), 58 (7.0%), 281 (33.7%) and 124 (14.9%), respectively. There were significant differences between the two groups in the incidences of BPD, severe ROP, NEC, brain injury and sepsis($\chi(2)$ =42.10, 47.20, 4.81, 44.28, 18.63, all $P<0.01$), which had significant correlation with poor outcomes at 12 months corrected age. The three top morbidities were severe ROP, BPD and brain injury(OR=3.82, 2.90, 2.80). Combined morbidities with BPD, severe ROP and brain injury correlated with higher risk of poor outcomes (one morbidity, OR=3.14, β=1.15; two morbidities, OR=7.31, β=1.99; three morbidities, OR=22.41, β=3.11; all $P<0.01$). Conclusion: BPD, severe ROP, NEC, brain injury and sepsis were the risk factors of poor outcomes at 12 months corrected age in VLBW infants. And the more combined morbidities with severe ROP, BPD and brain injury, the higher risk of poor outcomes in this population. Trial registration Clinical Trails, NCT03104946.

PMID: 28822437