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Interventions and Management

1. Weight-supported training of the upper extremity in children with cerebral palsy: a motor learning study.

Keller JW, van Hedel HJA.

J Neuroeng Rehabil. 2017 Aug 30;14(1):87. doi: 10.1186/s12984-017-0293-3.

BACKGROUND: Novel neurorehabilitation technologies build upon treatment principles derived from motor learning studies. However, few studies have investigated motor learning with assistive devices in children and adolescents with Cerebral Palsy (CP). The aim of this study was to investigate whether children with CP who trained with weight support in a playful, virtual environment would improve upper extremity task performance (i.e. skill acquisition), transfer, and retention, three aspects that indicate whether motor learning might have occurred or not. **METHODS:** Eleven children with CP (mean age 13.3 years, standard deviation 3.4 years), who were mildly to moderately impaired, participated. They played in the Armeo® Spring the exergame Moorhuhn with their more affected arm during 3 days (70 min pure play time). For this within-subject design, kinematic assessments, the Box and Block Test, and five items of the Melbourne Assessment were administered twice during a baseline week (one week before the intervention), directly before and after the intervention, and one day after the training phase (retention). **RESULTS:** The average exergame score improved from 209.55 to 339.73 ($p < 0.001$, Cohen's $d = 1.80$), indicating skill acquisition. The change in the Box and Block test improved from 0.45 (baseline week) to 3.95 (intervention week; $p = 0.008$, $d = 1.59$) indicating skill transfer. The kinematic assessments and the Melbourne items did not change. Improvement in game score and Box and Block Test persisted one day later (retention). **CONCLUSIONS:** We found evidence indicating the successful acquisition, transfer, and retention of upper extremity skills in children with CP. We therefore infer that motor learning occurred when children with CP trained their more affected arm with weight-support in a playful, virtual environment.

[PMID: 28806711](#)

2. Effect of diagnosis, body site and experience on text entry rate of individuals with physical disabilities: a systematic review.

Koester HH, Arthanat S.

Disabil Rehabil Assist Technol. 2017 Aug 28:1-11. doi: 10.1080/17483107.2017.1369588. [Epub ahead of print]

OBJECTIVE: This study systematically reviewed the research on computer text entry by people with physical disabilities, and conducted a quantitative synthesis of text entry rates associated with individuals' diagnosis, body site used with the interface and their level of experience. **METHOD:** We searched 10 databases and included studies in which: typing speed was reported; the access interface was available for public use; and individuals with physical impairments were in the study population. For quantitative synthesis, we used only the text entry rates (TER) reported for individuals with physical impairments; studies also had to report the sample size, and the average and standard deviation for the text entry rates. **RESULTS:** Thirty-nine studies

involving 248 subjects met the criteria for quantitative synthesis. Cerebral palsy was associated with significantly slower TER, at 5.5 wpm, than muscular dystrophy (12.5 wpm), spina bifida (10.4 wpm), SCI high cervical (10.1 wpm) and SCI low cervical (13.3 wpm). Among the 19 body sites represented, the Fingers bilateral category had the highest average, at 17.72 wpm. Head (2.92 wpm) and Hand (non-typing) (3.95 wpm) were each associated with significantly slower TER than Hands unspecified, Fingers bilateral, Hand with control enhancer, Voice and Mouth. The three levels of experience examined, LowPlus, Medium and High, provided very similar TER. **CONCLUSION:** This study contributes external evidence for service providers who provide computer access interventions for individuals with disabilities. The analyzed text entry performances serve as benchmarks to be used as guidelines during interface selection and training. **Implications for Rehabilitation** The median text entry rate (TER) across the literature for individuals with physical disabilities is 7.0 wpm. TER is affected by a user's diagnosis and the body site used for typing. These TER data can serve as diagnostic norms and benchmarks to be used as guidelines during interface selection and training. We recommend that practitioners measure text entry rate with clients, to place their performance in the context of these results and provide a baseline against which to measure effectiveness of an intervention.

[PMID: 28845735](#)

3. Alleviation of Motor Impairments in Patients with Cerebral Palsy: Acute Effects of Whole-body Vibration on Stretch Reflex Response, Voluntary Muscle Activation and Mobility.

Krause A, Schönau E, Gollhofer A, Duran I, Ferrari-Malik A, Freyler K, Ritzmann R.

Front Neurol. 2017 Aug 16;8:416. doi: 10.3389/fneur.2017.00416. eCollection 2017.

INTRODUCTION: Individuals suffering from cerebral palsy (CP) often have involuntary, reflex-evoked muscle activity resulting in spastic hyperreflexia. Whole-body vibration (WBV) has been demonstrated to reduce reflex activity in healthy subjects, but evidence in CP patients is still limited. Therefore, this study aimed to establish the acute neuromuscular and kinematic effects of WBV in subjects with spastic CP. **METHODS:** 44 children with spastic CP were tested on neuromuscular activation and kinematics before and immediately after a 1-min bout of WBV (16-25 Hz, 1.5-3 mm). Assessment included (1) recordings of stretch reflex (SR) activity of the triceps surae, (2) electromyography (EMG) measurements of maximal voluntary muscle activation of lower limb muscles, and (3) neuromuscular activation during active range of motion (aROM). We recorded EMG of m. soleus (SOL), m. gastrocnemius medialis (GM), m. tibialis anterior, m. vastus medialis, m. rectus femoris, and m. biceps femoris. Angular excursion was recorded by goniometry of the ankle and knee joint. **RESULTS:** After WBV, (1) SOL SRs were decreased ($p < 0.01$) while (2) maximal voluntary activation ($p < 0.05$) and (3) angular excursion in the knee joint ($p < 0.01$) were significantly increased. No changes could be observed for GM SR amplitudes or ankle joint excursion. Neuromuscular coordination expressed by greater agonist-antagonist ratios during aROM was significantly enhanced ($p < 0.05$). **DISCUSSION:** The findings point toward acute neuromuscular and kinematic effects following one bout of WBV. Protocols demonstrate that pathological reflex responses are reduced (spinal level), while the execution of voluntary movement (supraspinal level) is improved in regards to kinematic and neuromuscular control. This facilitation of muscle and joint control is probably due to a reduction of spasticity-associated spinal excitability in favor of giving access for greater supraspinal input during voluntary motor control.

[PMID: 28861038](#)

4. Effects of multilevel surgery on a flexed knee gait in adults with cerebral palsy.

Putz C, Wolf SI, Mertens EM, Geisbüsch A, Gantz S, Braatz F, Döderlein L, Dreher T.

Bone Joint J. 2017 Sep;99-B(9):1256-1264. doi: 10.1302/0301-620X.99B9.BJJ-2016-1155.R1.

AIMS: A flexed knee gait is common in patients with bilateral spastic cerebral palsy and occurs with increased age. There is a risk for the recurrence of a flexed knee gait when treated in childhood, and the aim of this study was to investigate whether multilevel procedures might also be undertaken in adulthood. **PATIENTS AND METHODS:** At a mean of 22.9 months (standard deviation 12.9), after single event multi level surgery, 3D gait analysis was undertaken pre- and post-operatively for 37 adult patients with bilateral cerebral palsy and a fixed knee gait. **RESULTS:** There was a significant improvement of indices and clinical and kinematic parameters including extension of the hip and knee, reduction of knee flexion at initial contact, reduction of minimum and mean knee flexion in the stance phase of gait, improved range of movement of the knee and a reduction of mean flexion of the hip in the stance phase. Genu recurvatum occurred in two patients ($n = 3$ legs, 4%) and an increase of pelvic tilt ($> 5^\circ$) was found in 12 patients ($n = 23$ legs, 31%). **CONCLUSION:** Adult patients with bilateral cerebral

palsy and a flexed knee gait benefit from multilevel surgery including hamstring lengthening. The risk of the occurrence of genu recurvatum and increased pelvic tilt is lower than has been previously reported in children. Cite this article: Bone Joint J 2017;99-B:1256-64.

[PMID: 28860409](#)

5. Experience with jumping mechanography in children with cerebral palsy.

Duran I, Martakis K, Stark C, Alberg E, Bossier C, Semler O, Schoenau E.

J Musculoskelet Neuronal Interact. 2017 Sep 1;17(3):237-245.

OBJECTIVES: Jumping mechanography provides robust motor function indicators among healthy children. The aim of the study was to assess the reproducibility and validity of jumping mechanography conducted as single two-legged jump (S2LJ) in children with cerebral palsy (CP). **METHODS:** 215 S2LJ investigations from a sample of 75 children with CP were eligible for evaluation. For the estimation of the reproducibility, only the baseline set of data per patient were used. Gross motor function was evaluated by the Gross Motor Function Measure (GMFM-66). In 135 S2LJ investigations, GMFM-66 was assessed within a week in the same child. This data was used for validity assessment. **RESULTS:** Coefficients of variation for the main outcome parameters ranged between 6.15-9.71%, except for jump height (CV%=27.3%). The intraclass correlation coefficients for peak velocity (Vmax) and peak power relative to body weight (Pmax/mass) was 0.927 and 0.931. Vmax and Pmax/mass were also the test parameters with the strongest correlation to the GMFM-66 score (>0.7). **CONCLUSIONS:** S2LJ assessed in the present study provided reproducible outcome measures particularly for Vmax and Pmax/mass in children with CP. Further, Vmax and Pmax/mass showed the strongest correlation with the GMFM-66 score and seem to be the most relevant evaluation criteria.

[PMID: 28860426](#)

6. Trajectory of phantom limb pain relief using mirror therapy: Retrospective analysis of two studies.

Griffin SC, Curran S, Chan AWY, Finn SB, Baker CI, Pasquina PF, Tsao JW.

Scand J Pain. 2017 Apr;15:98-103. doi: 10.1016/j.sjpain.2017.01.007. Epub 2017 Mar 7.

BACKGROUND AND PURPOSE: Research indicates that mirror therapy reduces phantom limb pain (PLP). Objectives were to determine when mirror therapy works in those who respond to treatment, the relevance of baseline PLP to when pain relief occurs, and what pain symptoms respond to mirror therapy. **METHODS:** Data from two independent cohorts with unilateral lower limb amputation were analyzed for this study (n=33). Mirror therapy consisted of 15-min sessions in which amputees performed synchronous movements of the phantom and intact legs/feet. PLP was measured using a visual analogue scale and the Short-Form McGill Pain Questionnaire. **RESULTS:** The severity of PLP at the beginning of treatment predicted when pain relief occurred. Those with low baseline PLP experienced a reduction ($p<0.05$) in PLP by session 7 of treatment, those with medium baseline PLP experienced pain relief by session 14 of treatment, and those with high baseline PLP experienced pain relief by session 21 of treatment. Mirror therapy reduced throbbing, shooting, stabbing, sharp, cramping, aching, tender, splitting, tiring/exhausting, and punishing-cruel pain symptoms. **CONCLUSION:** The degree of PLP at baseline predicts when mirror therapy relieves pain. **IMPLICATIONS:** This article indicates that the degree of baseline PLP affects when mirror therapy relieves pain: relief occurs by session 7 in patients with low PLP but by session 21 in patients with high PLP. Clinicians should anticipate slower pain relief in patients who begin treatment with high levels of pain. ClinicalTrials.gov numbers: NCT00623818 and NCT00662415.

[PMID: 28850360](#)

7. Does power mobility training impact a child's mastery motivation and spectrum of EEG activity? An exploratory project.

Kenyon LK, Farris JP, Aldrich NJ, Rhodes S.

Disabil Rehabil Assist Technol. 2017 Aug 30;1-9. doi: 10.1080/17483107.2017.1369587. [Epub ahead of print]

PURPOSE: The purposes of this exploratory project were: (1) to evaluate the impact of power mobility training with a child who has multiple, severe impairments and (2) to determine if the child's spectrum of electroencephalography (EEG) activity changed during power mobility training. **STUDY DESIGN:** A single-subject A-B-A-B research design was conducted with a four-week duration for each phase. Two target behaviours were explored: (1) mastery motivation assessed via the dimensions of mastery questionnaire (DMQ) and (2) EEG data collected under various conditions. Power mobility skills were also assessed. **METHODS:** The participant was a three-year, two-month-old girl with spastic quadriplegic cerebral palsy, gross motor function classification system level V. Each target behaviour was measured weekly. During intervention phases, power mobility training was provided. **RESULTS:** Improvements were noted in subscale scores of the DMQ. Short-term and long-term EEG changes were also noted. Improvements were noted in power mobility skills. **CONCLUSIONS:** The participant in this exploratory project demonstrated improvements in power mobility skill and function. EEG data collection procedures and variability in an individual's EEG activity make it difficult to determine if the participant's spectrum of EEG activity actually changed in response to power mobility training. Additional studies are needed to investigate the impact of power mobility training on the spectrum of EEG activity in children who have multiple, severe impairments. Implications for Rehabilitation Power mobility training appeared to be beneficial for a child with multiple, severe impairments though the child may never become an independent, community-based power wheelchair user. Electroencephalography may be a valuable addition to the study of power mobility use in children with multiple, severe impairments. Power mobility training appeared to impact mastery motivation (the internal drive to solve complex problems and master new skills) in a child who has multiple, severe impairments.

[PMID: 28853621](#)

8. Muscle-Bone Interactions in Pediatric Bone Diseases.

Veilleux LN, Rauch F.

Curr Osteoporos Rep. 2017 Aug 30. doi: 10.1007/s11914-017-0396-6. [Epub ahead of print]

PURPOSE: Here, we review the skeletal effects of pediatric muscle disorders as well as muscle impairment in pediatric bone disorders. **RECENT FINDINGS:** When starting in utero, muscle disorders can lead to congenital multiple contractures. Pediatric-onset muscle weakness such as cerebral palsy, Duchenne muscular dystrophy, spinal muscular atrophy, or spina bifida typically are associated with small diameter of long-bone shafts, low density of metaphyseal bone, and increased fracture incidence in the lower extremities, in particular, the distal femur. Primary bone diseases can affect muscles through generic mechanisms, such as decreased physical activity or in disease-specific ways. For example, the collagen defect underlying the bone fragility of osteogenesis imperfecta may also affect muscle force generation or transmission. Transforming growth factor beta released from bone in Camurati Engelman disease may decrease muscle function. **FUTURE DIRECTIONS:** Considering muscle-bone interactions does not only contribute to the understanding of musculoskeletal disorders but also can identify new targets for therapeutic interventions.

[PMID: 28856575](#)

9. Isolated neonatal MRI punctate white matter lesions in very preterm neonates and quality of life at school age.

Arberet C, Proisy M, Fausser JL, Curt M, Bétrémieux P, Tréguier C, Rozel C, Pladys P.

J Neonatal Perinatal Med. 2017 Aug 23. doi: 10.3233/NPM-1691. [Epub ahead of print]

OBJECTIVE: To study the quality of life at school age of very preterm infants presenting isolated punctate periventricular white matter lesions (IPWL) on late-preterm or term magnetic resonance imaging (MRI). **METHODS:** In 1996-2000, 16 of the 131 very preterm neonates explored by MRI were found to have IPWL. At the age of 9-14, 12 children from the IPWL group were compared with 54 children born preterm but with a normal MRI (no lesion). Quality of life (Health Status Classification System Pre School questionnaire), school performance, and motor outcome were investigated. **RESULTS:** Overall quality of

life did not differ between the groups (classified as perfect in 2/12 of the IPWL vs 20/54 in the no-lesion). The sub-items mobility and dexterity differed significantly between the two groups, with impairment in the IPWL group ($p < 0.001$ and $p < 0.05$). This group also displayed higher levels of motor impairment: they began walking later [20(4) vs. 15(3) months], $p < 0.01$, had higher frequencies of cerebral palsy (6/12 vs. 2/54, $p < 0.05$), and dyspraxia (4/12 vs. 0/54, $p < 0.001$). The rate of grade retention did not differ between the groups (3/12 in the IPWL group vs. 17/54 in the no-lesions group) but, as expected, was higher than that of the French general population (17.4%) during the study period. **CONCLUSION:** This long-term follow-up study detected no increase in the risk of subsequent cognitive impairment in very preterm infants with IPWL, but suggests that these children may have a significantly higher risk of dyspraxia, and motor impairment.

[PMID: 28854519](#)

10. Parents and Physiotherapists Recognition of Non-Verbal Communication of Pain in Individuals with Cerebral Palsy.

Riquelme I, Pades Jiménez A, Montoya P.

Health Commun. 2017 Aug 29;1-6. doi: 10.1080/10410236.2017.1358243. [Epub ahead of print]

Pain assessment is difficult in individuals with cerebral palsy (CP). This is of particular relevance in children with communication difficulties, when non-verbal pain behaviors could be essential for appropriate pain recognition. Parents are considered good proxies in the recognition of pain in their children; however, health professionals also need a good understanding of their patients' pain experience. This study aims at analyzing the agreement between parents' and physiotherapists' assessments of verbal and non-verbal pain behaviors in individuals with CP. A written survey about pain characteristics and non-verbal pain expression of 96 persons with CP (45 classified as communicative, and 51 as non-communicative individuals) was performed. Parents and physiotherapists displayed a high agreement in their estimations of the presence of chronic pain, healthcare seeking, pain intensity and pain interference, as well as in non-verbal pain behaviors. Physiotherapists and parents can recognize pain behaviors in individuals with CP regardless of communication disabilities.

[PMID: 28850264](#)

11. Enteral feeding and its impact on family mealtime routines for caregivers of children with cerebral palsy: A mixed method study.

Russell M, Jewell V, Poskey GA, Russell A.

Aust Occup Ther J. 2017 Aug 29. doi: 10.1111/1440-1630.12418. [Epub ahead of print]

BACKGROUND/AIM: Enteral feedings are part of the daily mealtime experience for many caregivers of children with cerebral palsy. The scope of occupational therapy practice incorporates multiple aspects of the enteral feeding process. Yet, the research in this area is very limited. The purpose of this study was to provide practitioners with better understanding of the impact enteral feedings of children with cerebral palsy have on family mealtime routines. **METHODS:** Using a complimentary mixed method approach, data were obtained through an online survey containing the Satisfaction Questionnaire with Gastrostomy Feeding (SAGA-8) and supplementary questions, and qualitative semi-structured phone interviews. Participants were caregivers of children with cerebral palsy who receive their primary nutrition through a gastrostomy tube. **RESULTS:** This study's cohort consisted of $n = 36$, SAGA-8, and $n = 6$ in-depth interviews. The mean age of children of was 9.4 (6.94 SD) with a mean age of 3.4 (5.35 SD) when enteral feeding was introduced. While families' overall situations positively changed after the gastrostomy tube placement, environmental barriers and length of feeding time continued to present a challenge to mealtime routines. The mixed methods data analysis revealed that successful adjustment to having a child with a gastrostomy tube and problem solving are closely linked and a consistent part of mealtime experience. **CONCLUSION:** Findings highlighted the necessity of comprehensive support from health professionals in achieving positive mealtime experience. Themes in this study indicated that caregivers would benefit from a professional with knowledge in the development and integration of rituals and routines to support positive outcomes.

[PMID: 28850693](#)

12. Low cost assistive technology to support educational activities for adolescents with cerebral palsy.

da Silva AP, Bulle Oliveira AS, Pinheiro Bezerra IM, Pedrozo Campos Antunes T, Guerrero Daboin BE, Raimundo RD, Dos Santos VR, de Abreu LC.

Disabil Rehabil Assist Technol. 2017 Aug 30;1-7. doi: 10.1080/17483107.2017.1369590. [Epub ahead of print]

INTRODUCTION: The concept of assistive technology covers several areas of action; one of them is communication with the elaboration of accessible solutions to overcome daily difficulties. It contributes to the resumption of functional abilities, expanding and facilitating inclusion and independent living. **OBJECTIVE:** To analyze the usability of a low cost prototype device to support educational activities of adolescents with cerebral palsy. **METHODS:** A descriptive observational study. The evaluation of a prototype device was made through a validated questionnaire, Quest Version 2.0, on the level of the user's satisfaction with an assistive technology, composed of 12 evaluation items. The questionnaire was filled out by the educator based on the observation of four wheelchair-bound participants diagnosed with cerebral palsy according to the international classification of diseases and health-related problems, ICD-10, who attend a coexistence and teaching institution in the state of São Paulo, Brazil. **RESULTS:** The device developed was considered an assistive technology, which provided an experience with a positive level of satisfaction for the participants. **CONCLUSION:** The tested prototype contributes to communication and interaction allowing adolescents with cerebral palsy to participate in educational activities. Implications for Rehabilitation The device assists the individual in the educational activities and can positively influence their development, observe the individual number 5, who has an important limitation in coordination and fine movements, placing the role of the task in the vertical position offers a new perspective to perform the task, this stimulates him to try to perform the work, so the challenge was adjusted to the demands of each individual which can contribute to its neuromotor development, the amplitude of the distal movements and the manual ability, since it must look for alternatives to complete the task requested.

[PMID: 28853616](#)

13. Eye-gaze control technology for children, adolescents and adults with cerebral palsy with significant physical disability: Findings from a systematic review.

Karlsson P, Allsop A, Dee-Price BJ, Wallen M.

Dev Neurorehabil. 2017 Sep 1:1-9. doi: 10.1080/17518423.2017.1362057. [Epub ahead of print]

PURPOSE: The primary objective of this systematic review was to examine the effectiveness of eye-gaze control technology for facilitating communication across different social contexts for people with cerebral palsy and significant physical disability. **METHODS:** Systematic review. **RESULTS:** The search identified 756 potentially eligible articles, of which two, low level articles were eligible. One study reported positive results for achieving communication goals for children with cerebral palsy. The second concluded that eye-gaze control technology resulted in greater quality of life and less depression for adults with late stage amyotrophic lateral sclerosis when compared to non-users. **DISCUSSION:** Research regarding the effectiveness of eye-gaze control technology used to access a laptop, tablet or computer on communication outcomes, participation, quality of life and self-esteem in children, adolescents and adults with cerebral palsy and significant physical disability is sparse. A scoping review to fully identify issues to inform clinical practice and future research is required.

[PMID: 28862491](#)

14. Understanding participation of children with cerebral palsy in family and recreational activities.

Alghamdi MS, Chiarello LA, Palisano RJ, McCoy SW.

Res Dev Disabil. 2017 Aug 23;69:96-104. doi: 10.1016/j.ridd.2017.07.006. [Epub ahead of print]

AIMS: The primary aim of this study was to determine the effect of age, sex, gross motor, manual ability, and communication functions on the frequency and enjoyment of children's participation in family and recreational activities. The secondary aim was to determine the relationships between motor and communication functions and participation. **METHODS:** Participants were 694 children, 1.5-12 years old, with cerebral palsy (CP) and their parents across the US and Canada. Parents rated children's frequency and enjoyment of participation using the Child Engagement in Daily Life measure. Parents and therapists identified children's level of function using Gross Motor Function Classification System (GMFCS), Manual Ability Classification System (MACS), and Communication Function Classification System (CFCS). **RESULTS:** Differences in

frequency and enjoyment of participation were found based on children's GMFCS, MACS, and CFCS levels but not age or sex. Children with higher gross motor, manual, and communication functions had higher frequency and enjoyment of participation, compared to children with lower functions. Frequency of participation was associated with GMFCS and CFCS levels whereas enjoyment of participation was only associated with CFCS level. **IMPLICATIONS:** Knowledge of child's gross motor, manual ability, and communication functions of children with CP is important when setting goals and planning interventions for participation.

[PMID: 28843215](#)

15. Exploring perceptions of health caregivers on the causes of caregivers' occupational burnout in institutes of children with cerebral palsy: A qualitative study.

Dehghan A, Hosseini SA, Rassfiani M, Dalvand H.

Electron Physician. 2017 Jun 25;9(6):4516-4523. doi: 10.19082/4516. eCollection 2017 Jun.

BACKGROUND: Providing care for children with cerebral palsy (CP) is hard, energy-consuming, and long-term. Consequently, occupational burnout is highly probable for caregivers. **OBJECTIVE:** This study aimed to explore the perception of health caregivers regarding the causes of caregivers' occupational burnout in institutes of children with CP. **METHODS:** This qualitative study was conducted using content analysis methodology during an eight-month period in 2016. Nine caregivers, two managers, and one physiotherapist participated in the study. Purposive sampling method was used to select participants. In depth, semi-structure interviews were used to gather the data. All interviews were conducted at their workplace, in Tehran, Iran. The interviews were recorded, transcribed verbatim, and overviewed. Constant comparative analyses were used to analyze the interviews. **RESULTS:** The results were categorized into three main categories and nine sub-categories. The main categories were as follows: care-related stress, nature of caring occupation, and organizational demands and resources. **CONCLUSION:** Results of this study showed that several factors are involved in creating burnout among caregivers. The exploration of these factors may help us in designing appropriate interventions for caregivers to correctly implement caring activities so that they less suffer from care-related pressures and also become able to allocate some time for their own activities of interest and for recreational, social, and family activities.

[PMID: 28848625](#)

16. Gap analysis of service needs for adults with neurodevelopmental disorders.

Burke SL, Wagner E, Marolda H, Quintana JE, Maddux M.

J Intellect Disabil. 2017 Jan 1:1744629517726209. doi: 10.1177/1744629517726209. [Epub ahead of print]
In Florida, the Agency for Persons with Disabilities provides waivers for adults with the following types of disabilities: intellectual disability, autism spectrum disorder, cerebral palsy, spina bifida, Down syndrome, and Prader-Willi syndrome. This review examined the peer-reviewed literature to indicate and assess the common needs for individuals with intellectual and developmental disabilities. Current models of service delivery, the efficacy of these services, and remaining gaps in the need fulfillment of individuals within the six diagnostic categorizations of interest were examined. Severity level within each diagnostic category was plotted on a matrix according to whether the needs of individuals were minimal, moderate, severe, or universal. The study found that sexual health education, socialization, and adult-focused medical care are universal needs among the six conditions. The study indicates that health-care professionals must work toward addressing the many unmet needs in comprehensive life span care services for adult individuals with neurodevelopmental disorders.

[PMID: 28847208](#)

Prevention and Cure

17. Reactive astrocyte COX2-PGE2 production inhibits oligodendrocyte maturation in neonatal white matter injury.

Shiow LR, Favrais G, Schirmer L, Schang AL, Cipriani S, Andres C, Wright JN, Nobuta H, Fleiss B, Gressens P, Rowitch DH.

Glia. 2017 Aug 30. doi: 10.1002/glia.23212. [Epub ahead of print]

Inflammation is a major risk factor for neonatal white matter injury (NWMI), which is associated with later development of cerebral palsy. Although recent studies have demonstrated maturation arrest of oligodendrocyte progenitor cells (OPCs) in NWMI, the identity of inflammatory mediators with direct effects on OPCs has been unclear. Here, we investigated downstream effects of pro-inflammatory IL-1 β to induce cyclooxygenase-2 (COX2) and prostaglandin E2 (PGE2) production in white matter. First, we assessed COX2 expression in human fetal brain and term neonatal brain affected by hypoxic-ischemic encephalopathy (HIE). In the developing human brain, COX2 was expressed in radial glia, microglia, and endothelial cells. In human term neonatal HIE cases with subcortical WMI, COX2 was strongly induced in reactive astrocytes with "A2" reactivity. Next, we show that OPCs express the EP1 receptor for PGE2, and PGE2 acts directly on OPCs to block maturation in vitro. Pharmacologic blockade with EP1-specific inhibitors (ONO-8711, SC-51089), or genetic deficiency of EP1 attenuated effects of PGE2. In an IL-1 β -induced model of NWMI, astrocytes also exhibit "A2" reactivity and induce COX2. Furthermore, in vivo inhibition of COX2 with Nimesulide rescues hypomyelination and behavioral impairment. These findings suggest that neonatal white matter astrocytes can develop "A2" reactivity that contributes to OPC maturation arrest in NWMI through induction of COX2-PGE2 signaling, a pathway that can be targeted for neonatal neuroprotection.

[PMID: 28856805](#)

18. Biology of Microglia in the Developing Brain.

Kaur C, Rathnasamy G, Ling EA.

J Neuropathol Exp Neurol. 2017 Sep 1;76(9):736-753. doi: 10.1093/jnen/nlx056.

Microglia exist in different morphological forms in the developing brain. They show a small cell body with scanty cytoplasm with many branching processes in the grey matter of the developing brain. However, in the white matter such as the corpus callosum where the unmyelinated axons are loosely organized, they appear in an amoeboid form having a round cell body endowed with copious cytoplasm rich in organelles. The amoeboid cells eventually transform into ramified microglia in the second postnatal week when the tissue becomes more compact with the onset of myelination. Microglia serve as immunocompetent macrophages that act as neuropathology sensors to detect and respond swiftly to subtle changes in the brain tissues in pathological conditions. Microglial functions are broadly considered as protective in the normal brain development as they phagocytose dead cells and sculpt neuronal connections by pruning excess axons and synapses. They also secrete a number of trophic factors such as insulin-like growth factor-1 and transforming growth factor- β among many others that are involved in neuronal and oligodendrocyte survival. On the other hand, microglial cells when activated produce a plethora of molecules such as proinflammatory cytokines, chemokines, reactive oxygen species, and nitric oxide that are implicated in the pathogenesis of many pathological conditions such as epilepsy, cerebral palsy, autism, and perinatal hypoxic-ischemic brain injury. Although many studies have investigated the origin and functions of the microglia in the developing brain, in-depth in vivo studies along with analysis of their transcriptome and epigenetic changes need to be undertaken to elucidate their full potential be it protective or neurotoxic. This would lead to a better understanding of their roles in the healthy and diseased developing brain and advancement of therapeutic strategies to target microglia-mediated neurotoxicity.

[PMID: 28859332](#)

19. Cerebral palsy and asphyxia in 32-35 week preterm infants.

Pillers DM.

J Perinatol. 2017 Aug;37(8):899-900. doi: 10.1038/jp.2017.78.

The development of hypothermia as a therapy to reduce the impact of hypoxic-ischemic encephalopathy has been one of the great achievements in neonatal medicine of the twenty-first century. The approach, however, has been limited in its application to the term infant, as it has been discouraged in infants who are <35 weeks of gestational age (GA) at birth, pending further study. This has raised the following questions: (1) What is the impact of HIE on the premature infant?

[PMID: 28855708](#)

20. [PRINCIPLES OF CAUSALITY BETWEEN PERIPARTUM EVENTS AND CEREBRAL PALSY].

[Article in Hebrew]

Blickstein I.

Harefuah. 2017 Aug;156(8):498-501.

INTRODUCTION: Understanding the intrapartum development of cerebral palsy (CP) may improve obstetrical management, would decrease the incidence of CP, and would help the court in judging whether CP is a result of negligence. In 1999, the Australian task force, and in 2003 the American task force, published their consensus regarding the criteria to establish intrapartum development of CP (recently reaffirmed by the American task force). The rationale is that CP is the end product of a continuum starting with an intrapartum hypoxic event that continues with the birth of an asphyxiated neonate, that exhibit specific neurological manifestations, and ends with a specific type of CP. The more studies that are performed over time on the consensus criteria, the more valid they appear to be.

[PMID: 28853525](#)**21. Neurodevelopmental outcomes for high-risk neonates in a low-resource setting.**

Milner KM, Duke T, Steer AC, Kado JH, Koyamaibole L, Kaarira R, Namudu K, Woolfenden S, Miller AE, O'Heir KE, Neal EFG, Roberts G.

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Worldwide, most neonates who survive prematurity and serious illness reside in low-resource settings where developmental outcome data and follow-up care are limited. This study aimed to assess in Fiji, a low-resource Pacific setting, prevalence and risk factors for moderate to severe neurodevelopmental impairment (NDI) in early childhood among high-risk neonates compared with controls. Retrospective cohort study comparing long-term outcomes for high-risk neonatal intensive care unit patients (n=149) compared with matched term, normal birth weight neonates (n=147) discharged from Colonial War Memorial Hospital between November 2008 and April 2010. NDI was defined as one or more of cerebral palsy, moderate to severe hearing or visual impairment, or global developmental delay using Bayley Scales of Infant and Toddler Development Third Edition (ie, score <70 in ≥1 of cognitive, language or motor domains). At median (IQR) age 36.1 (28.3, 38.0) months, prevalence of moderate to severe NDI % (95% CI, n) in high-risk and control groups was 12 (5 to 17, n=13) and 5 (2 to 12, n=5), respectively, an increased risk ratio (95% CI) of 2.7 (0.8 to 8.9). Median gestational age (weeks (median, IQR)) in the high-risk group was 37.5 (34-40) weeks. Among high-risk neonates, gestational age, birth weight, asphyxia, meningitis and/or respiratory distress were significantly associated with risk of NDI. Prevalence of NDI was high among this predominantly term high-risk neonatal cohort compared with controls. Results, including identified risk factors, inform efforts to strengthen quality of care and models of follow-up for high-risk neonates in this low-resource setting.

[PMID: 28847882](#)**22. Neuroprotective effects of estrogen in CNS injuries: insights from animal models.**

Raghava N, Das BC, Ray SK.

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Among the estrogens that are biosynthesized in the human body, 17β-estradiol (estradiol or E2) is the most common and the best estrogen for neuroprotection in animal models of the central nervous system (CNS) injuries such as spinal cord injury (SCI), traumatic brain injury (TBI), and ischemic brain injury (IBI). These CNS injuries are not only serious health problems, but also enormous economic burden on the patients, their families, and the society at large. Studies from animal models of these CNS injuries provide insights into the multiple neuroprotective mechanisms of E2 and also suggest the possibility of translating the therapeutic efficacy of E2 in the treatment SCI, TBI, and IBI in humans in the near future. The pathophysiology of these injuries includes loss of motor function in the limbs, arms and their extremities, cognitive deficit, and many other serious consequences including life-threatening paralysis, infection, and even death. The potential application of E2 therapy to treat the CNS injuries may become a trend as the results are showing significant therapeutic benefits of E2 for neuroprotection

when administered into the animal models of SCI, TBI, and IBI. This article describes the plausible mechanisms how E2 works with or without the involvement of estrogen receptors and provides an overview of the known neuroprotective effects of E2 in these three CNS injuries in different animal models. Because activation of estrogen receptors has profound implications in maintaining and also affecting normal physiology, there are notable impediments in translating E2 therapy to the clinics for neuroprotection in CNS injuries in humans. While E2 may not yet be the sole molecule for the treatment of CNS injuries due to the controversies surrounding it, the neuroprotective effects of its metabolite and derivative or combination of E2 with another therapeutic agent are showing significant impacts in animal models that can potentially shape the new treatment strategies for these CNS injuries in humans.

[PMID: 28845391](#)