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Cerebral palsy research news

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Interventions and Management

1. Contra-Lateral Unintended Upper Arm Movement during Unimanual Tasks in Children with Cerebral Palsy.
Kim SK, Park HK, Park ES.

Yonsei Med J. 2020 Mar;61(3):235-242. doi: 10.3349/ymj.2020.61.3.235.

PURPOSE: To characterize associated reactions (ARs) in the contralateral arm across multiple muscles during unimanual tasks and to identify factors related to ARs in children with cerebral palsy (CP). MATERIALS AND METHODS: This was a prospective, cross-sectional study of 35 children with CP. The extent of ARs of the contra-lateral, non-task hand was assessed while performing three unimanual tasks (opening and clenching the fist, a finger opposition task, and tapping fingers). The occurrence of ARs in each trial was evaluated separately for each task using a four-point scale (total scores ranged from 0 to 12). Surface electromyography (SEMG) was used to measure the firing activity of the muscles of the opposite arm during the task. The Manual Ability Classification System and Melbourne Assessment 2 (MA-2) were used to evaluate upper limb function. RESULTS: AR scores were higher in the more-affected limb than in the less-affected limb. SEMG data on the non-task hand showed motor overflow up to the elbow muscles in the more-affected limb. Root mean square ratios of EMG signals were significantly higher in children with ARs than in children without ARs. Multiple regression analysis showed both age and MA-2 to be significant factors related to ARs in the more-affected limb. CONCLUSION: Children with visible ARs showed motor overflow in the non-task limb during unimanual hand tasks. Age and upper limb function were significantly related to the extent of ARs in the more-affected limb of children with CP.

PMID: 32102124

2. Effect of botulinum toxin type-A in spasticity and functional outcome of upper limbs in cerebral palsy.
Yadav S, Chand S, Majumdar R, Sud A.

J Clin Orthop Trauma. 2020 Mar-Apr;11(2):208-212. doi: 10.1016/j.jcot.2020.01.002. Epub 2020 Jan 8.

INTRODUCTION: Spasticity has been considered to be a main contributor to both the impairment of function as well as posture in children with cerebral palsy (CP). Patterns of upper limb motor involvement in CP vary with resultant limitations in daily independence, participation, and quality of life. Botulinum Toxin-A (BTX-A) is a potent neurotoxin which acts by preventing the release of acetylcholine (Ach) from presynaptic axon at motor end plate reducing focal spasticity. With literature established role of BTX-A available for lower limb spasticity in CP, the purpose of this study was to present an objective analysis of the effect of a single i.m. injection of BTX-A in reduction of spasticity in the upper limb as well as functional outcome in children (4-12yrs) with spastic CP. METHODS: A total of 28 patients (30 upper limbs) of spastic CP with minimum follow up of 6months were included in the study. Modified Ashworth Scale (MAS) and Modified Tardieu Scale (MTS) were used to measure the spasticity. Surface landmarks were used to give I.m. Botox in selected spastic muscles followed by targeted rehabilitation. Functional outcomes were measured by MACS (Manual Ability Classification System)

and Canadian Occupational Performance Measure (COPM) before treatment, at 3 and 6 months follow up. RESULTS: Pronator teres was the most frequently injected muscle followed by FCU and Adductor pollicis. MAS scores at all joints and MTS scores at forearm deteriorated between 3 and 6 months. However, MACS and COPM showed sustained improvement at 3months and 6months with statistically significant change. CONCLUSION: I.m. BTX-A injected using anatomical landmarks had significant improvement in both clinical and functional outcome measures. We noticed significant improvement in MACS and COPM at 6 months despite return of local spasticity. It is safe and effective for spasticity of upper limbs in cerebral palsy and capable of improving function without major side effects. MACS & COPM are easy to use, less time consuming & easily adjusted to local needs. Randomized control trials with long follow up are required in future with special focus on dosing and timing, scoring system for functional outcome as per regional needs and issue for antibody formation for repeat injections of BTX-A.

PMID: 32099281

3. Bilateral Symmetry in Leg and Joint Stiffness in Children with Spastic Hemiplegic Cerebral Palsy During Gait.
Huang HP, Kuo CC, Lu TW, Wu KW, Kuo KN, Wang TM.

J Orthop Res. 2020 Feb 22. doi: 10.1002/jor.24635. [Epub ahead of print]

Deviations are often identified at individual joints in the gait analysis of patients with cerebral palsy. Previous gait studies on hemiplegic cerebral palsy (HCP) have focused mainly on deviations of the affected side. The current study aimed to quantify and compare the joint and leg stiffness, the contributions of skeletal and muscular components, and the associated joint angles and moments of the affected and non-affected lower limbs during level walking in children with spastic HCP. Twelve children with spastic HCP and 12 healthy controls walked at a self-selected speed in a gait laboratory while their kinematic and forceplate data were measured and analyzed during loading response, mid-stance, terminal stance and pre-swing. The altered joint kinematics and kinetics in the non-affected limb in the HCP group appeared to be mainly a compensatory strategy to minimize the bilateral asymmetry in leg stiffness during the double-limb support phase and joint stiffness during the entire stance phase. The current results suggest that therapeutic planning and decision-making for children with HCP should consider not only the mechanics of the affected side, but also the control of the non-affected side.

PMID: 32086827

4. Intraoperative electrophysiology during single-level selective dorsal rhizotomy: technique, stimulation threshold, and response data in a series of 145 patients.
De Vloo P, Huttunen TJ, Forte D, Jankovic I, Lee A, Hair M, Cawker S, Chugh D, Carr L, Crowe BHA, Pitt M, Aquilina K.

J Neurosurg Pediatr. 2020 Feb 28:1-10. doi: 10.3171/2019.12.PEDS19372. [Epub ahead of print]

OBJECTIVE: Selective dorsal rhizotomy (SDR) is effective at permanently reducing spasticity in children with spastic cerebral palsy. The value of intraoperative neurophysiological monitoring in this procedure remains controversial, and its robustness has been questioned. This study describes the authors' institutional electrophysiological technique (based on the technique of Park et al.), intraoperative findings, robustness, value to the procedure, and occurrence of new motor or sphincter deficits. METHODS: The authors analyzed electrophysiological data of all children who underwent SDR at their center between September 2013 and February 2019. All patients underwent bilateral SDR through a single-level laminotomy at the conus and with transection of about 60% of the L2-S2 afferent rootlets (guided by intraoperative electrophysiology) and about 50% of L1 afferent roots (nonselectively). RESULTS: One hundred forty-five patients underwent SDR (64% male, mean age 6 years and 7 months, range 2 years and 9 months to 14 years and 10 months). Dorsal roots were distinguished from ventral roots anatomically and electrophysiologically, by assessing responses on free-running electromyography (EMG) and determining stimulation thresholds ($\geq$ 0.2 mA in all dorsal rootlets). Root level was determined anatomically and electrophysiologically by assessing electromyographic response to stimulation. Median stimulation threshold was lower in sacral compared to lumbar roots ($p < 0.001$), and 16% higher on the first operated (right) side ($p = 0.023$), but unrelated to age, sex, or functional status. Similarly, responses to tetanic stimulation were consistent: 87% were graded 3+ or 4+, with similar distributions between sides. This was also unrelated to age, sex, and functional status. The L2-S2 rootlets were divided (median 60%, range 50%-67%), guided by response to tetanic stimulation at threshold amplitude. No new motor or sphincter deficits were observed, suggesting sparing of ventral roots and sphincteric innervation, respectively. CONCLUSIONS: This electrophysiological technique appears robust and reproducible, allowing reliable identification of afferent nerve roots, definition of root levels, and guidance for rootlet division. Only a direct comparative study will establish whether intraoperative electrophysiology during SDR

minimizes risk of new motor or sphincter worsening and/or maximizes functional outcome.

PMID: 32109863

5. Correction: Association between Gait Deviation Index and Physical Function in Children with Bilateral Spastic Cerebral Palsy: A Cross-Sectional Study. J. Clin. Med. 2020, 9, 28.
Ito T, Noritake K, Sugiura H, Kamiya Y, Tomita H, Ito Y, Sugiura H, Ochi N, Yoshihashi Y.

J Clin Med. 2020 Feb 19;9(2). pii: E569. doi: 10.3390/jcm9020569.

The authors wish to make the following corrections to this paper [...]. Erratum for Association between Gait Deviation Index and Physical Function in Children with Bilateral Spastic Cerebral Palsy: A Cross-Sectional Study. [J Clin Med. 2019]

PMID: 32093130

6. Evaluation of the multidimensional effects of adaptive seating interventions for young children with non-ambulatory cerebral palsy.
Inthachom R, Prasertsukdee S, Ryan SE, Kaewkungwal J, Limpaninlachat S.

Disabil Rehabil Assist Technol. 2020 Feb 25:1-9. doi: 10.1080/17483107.2020.1731613. [Epub ahead of print]

Purpose: To investigate the short-term effectiveness of the first adaptive seating system received by children with non-ambulatory cerebral palsy (CP) who are classified as level IV or V according to the Gross Motor Function Classification System. Materials and methods: A trained clinical assessor examined 20 children with non-ambulatory CP (mean age: 4.5 years) for their trunk control ability in static, active, and reactive tasks using the Segmental Assessment of Trunk Control. Their primary caregivers were also interviewed about their child's activity and participation using the Paediatric Evaluation of Disability Inventory Computer - Adaptive Test in daily activity and social/cognitive domains and Family Impact of Assistive Technology Scale for Adaptive Seating in child and family functioning domains. Data for each measure were collected 3 times: at baseline (pre-intervention) and then 6 weeks and 3 months after children received their first adaptive seating system.Results and conclusion: The static and active trunk control scores between baseline and 6 weeks, and baseline and 3 months significantly improved. Daily activity scaled scores significantly improved between baseline and 3 months, and 6 weeks and 3 months. Significant, large gains in child and family functioning overall were detected between baseline and 6 weeks, and baseline and 3 months. These findings provide emerging evidence of multidimensional effects associated with the introduction of a first adaptive seating system into the lives of young children with non-ambulatory CPIMPLICATIONS FOR REHABILITATIONThe introduction of an adaptive seating system into the wheelchair of children with non-ambulatory cerebral palsy may be associated with short-term gains in body function, activities, participation and aspects of the child's environment.

PMID: 32096423

7. Calcaneal lengthening for the pes planovalgus foot deformity in children with cerebral palsy.
Aboelenein AM, Fahmy ML, Elbarbary HM, Mohamed AZ, Galal S.

J Clin Orthop Trauma. 2020 Mar-Apr;11(2):245-250. doi: 10.1016/j.jcot.2018.07.021. Epub 2018 Jul 29.

PURPOSE: The objective of this study was to evaluate the operative management of pes planovalgus deformity in ambulatory cerebral palsy (CP) children by calcaneal lengthening osteotomy described by Evans. METHOD: Fifteen children (10 girls and 5 boys) with average age 11 years 6 months (range, 8 years 4 months-14 years 6 months) with 22 feet with pes planovalgus (PPV) deformity were included in this study. Clinical evaluation was made according to Dogan's scale and graded as perfect, good, fair and poor. Preoperative and postoperative radiological assessment of anteroposterior talo-first metatarsal angle (AP-T1MT), anteroposterior talo-calcaneal angle (AP-TC), laterl Talo-first metatarsal angle (Lat. T1MT), lateral Talo-calcaneal

angle (Lat. TC), and lateral Calcaneal pitch angle (Lat. CP) had been done for all feet. All feet were corrected with the modification of the calcaneal lengthening osteotomy described by Mosca. RESULT: Clinical results were perfect in 18 feet (82%), good in 2 feet (9%) and fair in 2 feet (9%). Radiological results showed improvement in 20 feet, while 2 feet showed no improvement. The improvement was significant in Lat. T1MT (P ‹ 0.001), AP-T1MT (P < 0.05)., AP-TC and Lat. CP (P < 0.001, <0.001 respectively) whereas it was insignificant in Lat. TC (P > 0.05). CONCLUSION: The results of the present study showed that the procedure reliably relieves pain in PPV foot in CP children and proved effective in addressing all components of the deformity in both hindfoot and forefoot clinically and Radiologically.

PMID: 32099288

8. Investigation of the validity and reliability of the L test in children with cerebral palsy.
Cetin SY, Erel S.

Physiother Theory Pract. 2020 Feb 24:1-7. doi: 10.1080/09593985.2020.1731894. [Epub ahead of print]

Background: The L test is a modified version of the Timed Up and Go Test (TUG), with a walking path that is L-shaped. The L test is a more comprehensive test since it includes a longer walking path than TUG and turning in both directions.Objective: This study aimed to examine the reliability and validity of the L test, and the minimal detectable change (MDC) in children with cerebral palsy (CP).Methods: The study included 80 children with CP at levels 1 and 2 according to the Gross Motor Function Classification System (GMFCS). The Intraclass Correlation Coefficient (ICC) was used to assess the reliability of the L test according to GMFCS level. MDC estimates were calculated using baseline data. The correlations of the L test with TUG and the Timed up and Down Stairs Test (TUDS) were assessed for convergent validity.Results: Intra-rater (ICC 0.903-0.985 for level 1-2) and inter-rater (ICC 0.806-0.937 for level 1-2) reliability of the L test were determined to be excellent. A moderate correlation was found between the L test and TUG (r: 0.691) and TUDS (r: 0.546) for level 1; a moderate correlation was found between the L test and TUG (r: 0.625) and a high correlation was found between the L test and TUDS (r: 0.734) for level 2. The MDC values in terms of intra-rater were 1.44-2.21 s for level 1-2, and 1.30-1.57 s for level 2 in terms of inter-rater, respectively.Conclusion: The results of this study showed that the L test is a valid and reliable method in the assessment of functional mobility in children with CP.

PMID: 32090672

9. Walking on uneven ground: How do patients with unilateral cerebral palsy adapt?
Romkes J, Freslier M, Rutz E, Bracht-Schweizer K.

Clin Biomech (Bristol, Avon). 2020 Feb 8;74:8-13. doi: 10.1016/j.clinbiomech.2020.02.001. [Epub ahead of print]

BACKGROUND: Children with cerebral palsy experience movement disorders that influence gait stability. It is likely that gait stability further decreases when walking on uneven compared to even ground. Therefore, the aim of this study was to investigate gait on uneven ground in children with unilateral cerebral palsy. METHODS: Twenty children with unilateral cerebral palsy and twenty typically developing children performed a three-dimensional gait analysis when walking on even and uneven ground. Spatio-temporal parameters, full-body joint kinematics and centre of mass displacements were compared. FINDINGS: On uneven versus even ground, both groups showed decreased cadence, increased stance phase and double support time, increased toe clearance height, and increased knee and hip flexion during swing phase. Whereas only the typically developing children walked slower and had increased dorsiflexion and external foot progression during stance phase, only the patients showed increased stride width, increased elbow flexion (affected and non-affected side), and kept the centre of mass more medial when standing on the affected leg. INTERPRETATION: Patients and healthy children use similar adaptation mechanisms when walking on uneven ground. Both groups increased the toe clearance height by increasing knee and hip flexion during swing. However, whereas patients enlarge their base of support by increasing stride width, healthy children do so by increasing their external foot progression angle. Furthermore, patients seem to feel more insecure and hold their arms in a position to prepare for falls on uneven ground. They also do not compensate with their non-affected side for their affected side on uneven ground.

PMID: 32086046

10. Sleep problems and solution seeking for children with cerebral palsy and their parents.
Petersen S, Francis KL, Reddihough DS, Lima S, Harvey A, Newall F.

J Paediatr Child Health. 2020 Feb 26. doi: 10.1111/jpc.14830. [Epub ahead of print]

AIM: Sleep problems are common in school-aged children with cerebral palsy (CP). Despite the significant impact of sleep disturbance and deprivation, there is a paucity of research in the area. The aims of this study were to (i) investigate the frequency of sleep problems in children with CP and their parents and (ii) understand what happens when parents ask for help with sleep problems from their health-care professionals. METHODS: This was a prospective cohort study using an online survey sent to parents/primary care givers of children with CP aged 6-12 years recruited through the Victorian Cerebral Palsy Register. The following sleep assessment tools: The Children's Sleep Habits Questionnaire, the Pittsburgh Sleep Quality Index and the Gross Motor Function Classification System Parent Rating Tool were administered along with custom-designed questions that were informed by a preceding qualitative scoping study. RESULTS: Complete data sets were received from 126 parents/care givers. Almost half (46%) of the parents reported their child had sleep problems. Of the 64 parents who reported seeking help for a child's sleep problem, only 21 indicated that their attempt was successful. If a child had poor sleep, the parent was more likely to have a sleep problem than parents who did not report poor child sleeping. CONCLUSION: Sleep problems are common in children with CP and their parents. Parents do not always seek help, and those who do may not find an effective solution. Future research should explore how sleep problems can be effectively prioritised for children with CP and their parents/care givers.

PMID: 32100418

11. Cognitive and academic profiles in children with cerebral palsy: a narrative review.
Fluss J, Lidzba K.

Ann Phys Rehabil Med. 2020 Feb 19. pii: S1877-0657(20)30036-1. doi: 10.1016/j.rehab.2020.01.005. [Epub ahead of print]

BACKGROUND: Considerable effort has recently been made to improve the accurate diagnosis of cerebral palsy (CP) in childhood and to establish early intervention aiming to improve functional outcome. Besides the visible motor impairments, cognitive abilities are frequently affected but might remain unrecognised in children with mild forms. On the other hand, some severely disabled children with presumed intellectual disabilities might demonstrate normal-range reasoning capacities. Most studies on this topic have emphasized a variety of cognitive profiles (cognitive level) related to the type of cerebral palsy and the underlying brain lesions (biological level). However, little is known at the behavioural level, namely learning skills and educational achievement. OBJECTIVE: This narrative review aimed to discuss cognitive and scholastic skills typically affected in children with CP. METHODS: Online literature research for studies of cerebral palsy, cognition and academic achievement, extracting all relevant articles regardless of article type. RESULTS: In children with CP, intellectual disability is frequent and correlated with the degree of motor impairment and early epilepsy. Speech and language problems are prevalent in all forms of CP and might hamper everyday participation on varying levels depending on the degree of motor disability. Most children with CP have neuropsychological deficits affecting predominantly visuospatial functions, attention, and/or executive functions. These problems relate to academic performance and social participation. DISCUSSION: An adequate interdisciplinary follow-up of children with CP requires a sensitization of clinicians to the complex topic of cognitive and academic problems in this population and a better synergy between the medical and educational worlds.

PMID: 32087307

12. Mechanical Design of a New Device to Assist Eating in People with Movement Disorders.
Turgeon P, Dubé M, Laliberté T, Archambault PS, Flamand V, Routhier F, Campeau-Lecours A.

Assist Technol. 2020 Feb 27. doi: 10.1080/10400435.2020.1734111. [Epub ahead of print]

Many people living with neurological disorders, such as cerebral palsy, stroke, muscular dystrophy or dystonia, experience upper limb impairments (muscle spasticity, loss of selective motor control, muscle weakness or tremors) and are unable to eat independently. This article presents the development of a new device to assist with eating, aimed at stabilizing the movement of people who have movement disorders. The design was guided by insights gathered through focus groups, with occupational

therapists and engineers, about the challenges faced by individuals who have movement disorders and difficulty in eating autonomously. The proposed assistive device prototype is designed to be fixed on a table and to support a spoon. The mechanism is designed so that the spoon maintains a position parallel to the ground for the user. Dampers and inertia allow stabilizing the user's motion. A preliminary trial with five individuals living with cerebral palsy is presented to assess the prototype's performance and to guide future iterations of the prototype. Task completion time generally decreased and movement fluidity generally improved when using the assistive device prototype. The prototype showed good potential in stabilizing the spoon for the user and improving movement fluidity.

PMID: 32105199

13. Effect of feedback and target size on eye gaze accuracy in an off-screen task.
Sakamaki I, Adams K, Tavakoli M, Wiebe SA.

Disabil Rehabil Assist Technol. 2020 Feb 26:1-11. doi: 10.1080/17483107.2020.1729874. [Epub ahead of print]

Purpose: Eye gaze interfaces have been used by people with severe physical impairment to interact with various assistive technologies. If used to control robots, it would be beneficial if individuals could gaze directly at targets in the physical environment rather than have to switch their gaze between a screen with representations of robot commands and the physical environment to see the response of their selection. By using a homogeneous transformation technique, eye gaze coordinates can be mapped between the reference coordinate frame of eye tracker and the coordinate frame of objects in the physical environment. Feedback about where the eye tracker has determined the eye gaze is fixated is needed so users can select targets more accurately. Screen-based assistive technologies can use visual feedback, but in a physical environment, other forms of feedback need to be examined.Materials and methods: In this study, an eye gaze system with different feedback conditions (i.e., visual, auditory, vibrotactile, and no-feedback) was tested when participants received visual feedback on a display (on-screen) and when looking directly at the physical environment (off-screen). Target selection tasks in both screen conditions were performed by ten non-disabled adults, three non-disabled children, and two adults and one child with cerebral palsy.Results: Tasks performed with gaze fixation feedback modalities were accomplished faster and with higher success than tasks performed without feedback, and similar results were observed in both screen conditions. No significant difference was observed in performance across the feedback modalities, but participants had personal preferences.Conclusion: The homogeneous transformation technique enabled the use of a stationary eye tracker to select target objects in the physical environment, and auditory and vibrotactile feedback enabled participants to be more accurate selecting targets than without it.Implications for RehabilitationBeing able to select target objects in the physical environment by eye gaze could make it easier for children with disabilities to control assistive robots, because in this way they do not have to change their focus between a computer screen with commands and the robot.Providing auditory or vibrotactile feedback when using an eye gaze system made it faster and easier to know if a target was being gazed upon.Being able to select targets in the environment using eye gaze could be beneficial for other assistive technology, too, such as destination selection for power wheelchairs.

PMID: 32100583

14. Reliability and validity of the pediatric feeding and swallowing disorders family impact scale for Turkish children with cerebral palsy by endoscopic evaluation.
Umay E, Gündoğdu İ, Öztürk EA.

Turk J Pediatr. 2019;61(5):741-748. doi: 10.24953/turkjped.2019.05.013.

Umay E, Gündoğdu İ, Öztürk EA. Reliability and validity of the pediatric feeding and swallowing disorders family impact scale for Turkish children with cerebral palsy by endoscopic evaluation. Turk J Pediatr 2019; 61: 741-748. The caregivers of children with cerebral palsy (CP) have high mood disorders and stress levels. This study was aimed to conduct validity and reliability of Turkish version of The Pediatric Feeding and Swallowing Disorders Family Impact Scale (PFSDFIS) by using an objective method. This study was performed in our physical medicine and rehabilitation (PMR) clinic between July 2016 and July 2018. This study was performed with 251 children with CP who had complaint of swallowing and/or feeding problems, and their primer caregivers. Cronbach`s alpha and corrected item-total correlations were used to assess internal consistency. Test and retest reliability studies were also conducted. The construct validity was assessed using the dysphagia level defined with flexible fiberoptic endoscopic evaluation of swallowing and Impact on Family Scale (IFS). Total score of T-PFSDFIS was correlated to the dysphagia level by using FEES. Results showed, Cronbach's alpha value of the scale to be 0.821. Corrected item-to-total correlations ranged from 0.729 to 0.911. Test-retest reliability coefficients was calculated with intra-class

correlation coefficient (ICC), the total score was 0.989. A negative significant good level correlation was found between the dysphagia level by using endoscopic evaluation and the T- PFSDFIS total score as well as between total scores of IFS and T-PFSDFIS. In subgroup analysis; the lowest value was in normal swallowing and significantly different from all dysphagia levels. In conclusion; this scale is effective in reflecting the influence of caregivers on the severity of dysphagia measured objectively and T-PFSDFIS is a valid and reliable scale for Turkish children with CP.

PMID: 32105006

15. Quality of life after selective dorsal rhizotomy: an assessment of family-reported outcomes using the CPQoL questionnaire.
Robins JMW, Boyle A, McCune K, Lodh R, Goodden JR.

Childs Nerv Syst. 2020 Feb 24. doi: 10.1007/s00381-020-04546-1. [Epub ahead of print]

BACKGROUND: Selective dorsal rhizotomy (SDR) is widely accepted as an effective procedure for management of lower limb spasticity in children with cerebral palsy. However, effects of the procedure on quality of life are not widely reported and less so using a structured and validated quality of life tool such as Cerebral Palsy Quality of Life Questionnaire (CPQoL). Here, we present complete data for CPQoL outcomes for SDR patients operated in a single institution at 2 years follow-up. METHODS: Patients were operated over a 5-year period by the same surgeon using the same technique in a single institution. CPQoL questionnaires were completed by patients and families pre-operatively and at 6 months, 1 year and 2 years post-operatively. Data was collected prospectively. RESULTS: A total of 78 patients (58 male, 20 female), age range 2.6-13.8 years (median 6.33) were included whom underwent SDR between October 2012-November 2017. All had complete follow-up up to 2 years post-procedure (most recent November 2019). Four patients were excluded due to incomplete follow-up data. Statistically significant improvement was seen across five out of seven CPQoL domains and this was sustained to 2 years post-SDR. CONCLUSIONS: We demonstrate using a validated Quality of Life Tool that SDR has a beneficial effect on the quality of life for patients with cerebral palsy at this length of follow-up.

PMID: 32095868

16. Cerebral palsy: managing expectations and optimising outcomes.
Stirrups R.

Lancet Child Adolesc Health. 2020 Mar;4(3):179-180. doi: 10.1016/S2352-4642(20)30033-X.

PMID: 32087140

17. Exploring the contributing factors that influence the volition of adolescents with cerebral palsy: A directed content analysis.
Dehghanizadeh M, Khalafbeigi M, Akbarfahimi M, Yazdani F, Zareiyan A.

Scand J Occup Ther. 2020 Feb 23:1-11. doi: 10.1080/11038128.2020.1723686. [Epub ahead of print]

Background: Communication, behavioural disturbances and low motivation influence the functional potential and the effectiveness of interventions in adolescents with cerebral palsy (CP). While the model of human occupation (MOHO) is a conceptual model in occupational therapy, no research on the volition of adolescents with CP in daily activities has been undertaken. Aim: To explore the elements contributing to the volition of adolescents with CP based on their own experience. Methods: A qualitative approach using directed content analysis with volition of MOHO as a framework was applied. Semi-structured in-depth interviews were carried out with five adolescents with CP and five parents of the same adolescents. Results: In this study primary codes were categorized into eight categories: family and community-related values, individual values, individual perceptions of ability level, sense of control over conditions, enjoying performing activities, enjoying interpersonal relationships, physical context features and social context features. Conclusions: Personal causation in interaction with environmental features is strongly influenced by adolescents' motivation. Significance: Occupational therapists could improve

the motivation of adolescents with CP in interaction with the individual's volition with focussing on self-efficacy for promoting personal causation. Adapting to the physical environment and changing the attitudes of others to these adolescents is necessary.

PMID: 32089013

18. Disabled and immigrant, a double minority challenge: a qualitative study about the experiences of immigrant parents of children with disabilities navigating health and rehabilitation services in Norway.
Arfa S, Solvang PK, Berg B, Jahnsen R.

BMC Health Serv Res. 2020 Feb 22;20(1):134. doi: 10.1186/s12913-020-5004-2.

BACKGROUND: Immigrants and their Norwegian-born children make up approximately 18% of the total population in Norway. While several studies have been conducted on immigrants' utilization of healthcare services, immigrant families are systematically underrepresented in international studies of children with disabilities. By focusing on experiences of immigrant parents of children with disabilities navigating health and rehabilitation services in Norway, this study generated knowledge of how accessible and tailored the services were from their point of view. METHODS: This study took a qualitative approach, using semi-structured interviews to explore the experiences of immigrant parents of children with disabilities from non-Western countries. The interviews were transcribed, coded, and analyzed via an inductive thematic analytic approach. RESULTS: The findings show how the "immigrant experience" influenced the way the parents looked at, experienced, and even praised the services. The parents appreciated the follow-up services provided by the pediatric rehabilitation centers, which they experienced as predictable and well-organized. While navigating the services, they experienced several challenges, including the need for information, support, and timely help. They felt exhausted because of years of struggle in the healthcare system to access the help and services they needed. They expressed how this struggle had affected their own health. The feeling of being treated differently from the majority was another challenge they experienced while navigating the services. The findings also show how parents' experiences of communication with healthcare providers were influenced not only by their own language and communication skills but also by the healthcare providers' intercultural communication skills and dominant organizational culture. CONCLUSIONS: The parents' experiences show that there is still a gap between the public ideal of equal healthcare services and the reality of the everyday lives of immigrant families of children with disabilities. By exploring immigrant parents' experiences, this study highlights the importance of mobilization at both the individual and systemic levels to fill the current gap and provide tailored and accessible services to the entire population.

PMID: 32087730

19. Long-term health of children conceived after assisted reproductive technology.
Bergh C, Wennerholm UB.

Ups J Med Sci. 2020 Feb 26:1-6. doi: 10.1080/03009734.2020.1729904. [Epub ahead of print]

The aim of this narrative review is to summarize the present knowledge on long-term outcome of children born after assisted reproductive technologies (ART). The main outcomes covered are neurodevelopment including cerebral palsy, cognitive development, attention deficit hyperactivity disorder and autism spectrum disease, growth, cardiovascular function, diabetes type 1, asthma, malignancies, and reproductive health. Results have mainly been obtained from systematic reviews/meta-analyses and large registry studies. It has been shown that children born after ART, when restricted to singletons, have a similar outcome for many health conditions as their spontaneously conceived peers. For some outcomes, particularly cardiovascular function and diabetes, studies show some higher risk for ART singletons or subgroup of ART singletons. The fast introduction of new ART techniques emphasizes the importance of continuous surveillance of children born after ART.

PMID: 32101068

20. Conference Report on Contractures in Musculoskeletal and Neurological Conditions.
Nuckolls GH, Kinnett K, Dayanidhi S, Domenighetti AA, Duong T, Hathout Y, Lawlor MW, Lee S, Magnusson SP, McDonald CM, McNally EM, Miller NF, Olwin BB, Raghavan P, Roberts T, Rutkove S, Sarwark JF, Senesac C, Vogel LF, Walter GA, Willcocks R, Rymer WZ, Lieber RL.

Muscle Nerve. 2020 Feb 28. doi: 10.1002/mus.26845. [Epub ahead of print]

Limb contractures are debilitating complications associated with various muscle and nervous system disorders. This report summarizes a conference at the Shirley Ryan AbilityLab in Chicago, IL on April 19-20, 2018 involving researchers and physicians from diverse disciplines convened to discuss current clinical and preclinical understanding of contractures in Duchenne muscular dystrophy, stroke, cerebral palsy and other conditions. Presenters described changes in muscle architecture, activation, extracellular matrix, satellite cells and muscle fiber sarcomeric structure that accompany or predispose muscles to contracture. Participants identified ongoing and future research directions that may lead to understanding of the intersecting factors that trigger contractures. These include additional studies of changes in muscle, tendon, joint and neuronal tissues during contracture development using imaging, molecular and physiologic approaches. Participants identified the need for improved biomarkers and outcome measures to identify patients likely to develop contractures and to accurately measure efficacy of treatments currently available and under development.

PMID: 32108365

21. Loss of TNR causes a nonprogressive neurodevelopmental disorder with spasticity and transient opisthotonus.
Wagner M, Lévy J, Jung-Klawitter S, Bakhtiari S, Monteiro F, Maroofian R, Bierhals T, Hempel M, Elmaleh-Bergès M, Kitajima JP, Kim CA, Salomao JG, Amor DJ, Cooper MS, Perrin L, Pipiras E, Neu A, Doosti M, Karimiani EG, Toosi MB, Houlden H, Jin SC, Si YC, Rodan LH, Venselaar H, Kruer MC, Kok F, Hoffmann GF, Strom TM, Wortmann SB, Tabet AC, Opladen T.

Genet Med. 2020 Feb 26. doi: 10.1038/s41436-020-0768-7. [Epub ahead of print]

PURPOSE: TNR, encoding Tenascin-R, is an extracellular matrix glycoprotein involved in neurite outgrowth and neural cell adhesion, proliferation and migration, axonal guidance, myelination, and synaptic plasticity. Tenascin-R is exclusively expressed in the central nervous system with highest expression after birth. The protein is crucial in the formation of perineuronal nets that ensheath interneurons. However, the role of Tenascin-R in human pathology is largely unknown. We aimed to establish TNR as a human disease gene and unravel the associated clinical spectrum. METHODS: Exome sequencing and an online matchmaking tool were used to identify patients with biallelic variants in TNR. RESULTS: We identified 13 individuals from 8 unrelated families with biallelic variants in TNR sharing a phenotype consisting of spastic para- or tetraparesis, axial muscular hypotonia, developmental delay, and transient opisthotonus. Four homozygous loss-of-function and four different missense variants were identified. CONCLUSION: We establish TNR as a disease gene for an autosomal recessive nonprogressive neurodevelopmental disorder with spasticity and transient opisthotonus and highlight the role of central nervous system extracellular matrix proteins in the pathogenicity of spastic disorders.

PMID: 32099069

22. Neuroprem: the Neuro-developmental outcome of very low birth weight infants in an Italian region.
Lugli L, Pugliese M, Plessi C, Berardi A, Guidotti I, Ancora G, Grandi S, Gargano G, Braibanti S, Sandri F, Soffritti S, Ballardini E, Arena V, Stella M, Perrone S, Moretti S, Rizzo V, Ferrari F; Neuroprem Working Group, Picciolini O, Bellù R, Turoli D, Corvaglia LT, Garani G, Paoletti V, Biasucci G, Biasini A, Benenati B, Stagi P, Magnani C, Dallaglio S, DellaCasa Muttini E, Roversi MF, Bedetti L, Lucaccioni L, Bertoncelli N, Boncompagni A.

Ital J Pediatr. 2020 Feb 22;46(1):26. doi: 10.1186/s13052-020-0787-7.

INTRODUCTION: The survival of preterm babies has increased worldwide, but the risk of neuro-developmental disabilities remains high, which is of concern to both the public and professionals. The early identification of children at risk of neuro-developmental disabilities may increase access to intervention, potentially influencing the outcome. AIMS: Neuroprem is an area-based prospective cohort study on the neuro-developmental outcome of very low birth weight (VLBW) infants that aims to define severe functional disability at 2 years of age. METHODS: Surviving VLBW infants from an Italian network of 7 neonatal intensive care units (NICUs) were assessed for 24 months through the Griffiths Mental Developmental Scales (GMDS -R) or the Bayley Scales of Infant and Toddler Development (BSDI III) and neuro-functional evaluation according to the International Classification of Disability and Health (ICF-CY). The primary outcome measure was severe functional disability at 2 years of age, defined as cerebral palsy, a BSDI III cognitive composite score < 2 standard deviation (SD) or a GMDS-R global quotients score < 2 SD, bilateral blindness or deafness. RESULTS: Among 211 surviving VLBW infants, 153 completed

follow-up at 24 months (72.5%). Thirteen patients (8.5%) developed a severe functional disability, of whom 7 presented with cerebral palsy (overall rate of 4.5%). Patients with cerebral palsy were all classified with ICF-CY scores of 3 or 4. BSDI III composite scores and GMDS-R subscales were significantly correlated with ICF-CY scores ($p < 0.01$). CONCLUSION: Neuroprem represents an Italian network of NICUs aiming to work together to ensure preterm neuro-developmental assessment. This study updates information on VLBW outcomes in an Italian region, showing a rate of cerebral palsy and major developmental disabilities in line with or even lower than those of similar international studies. Therefore, Neuroprem provides encouraging data on VLBW neurological outcomes and supports the implementation of a preterm follow-up programme from a national network perspective.

PMID: 32087748

23. [Rehabilitation strategy and recommendation for motor dysfunction in children with cerebral palsy]. [Article in Chinese; Abstract available in Chinese from the publisher]
Subspecialty Group of Rehabilitation, the Society of Pediatrics, Chinese Medical Association.

Zhonghua Er Ke Za Zhi. 2020 Feb 2;58(2):91-95. doi: 10.3760/cma.j.issn.0578-1310.2020.02.005.

PMID: 32102143

24. Neural activation within the sensorimotor cortices during bimanual tasks in children with unilateral cerebral palsy.
Surkar SM.

Dev Med Child Neurol. 2020 Feb 26. doi: 10.1111/dmcn.14505. [Epub ahead of print]

PMID: 32100282

25. State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy.
Novak I, Morgan C, Fahey M, Finch-Edmondson M, Galea C, Hines A, Langdon K, Namara MM, Paton MC, Popat H, Shore B, Khamis A, Stanton E, Finemore OP, Tricks A, Te Velde A, Dark L, Morton N, Badawi N.

Curr Neurol Neurosci Rep. 2020 Feb 21;20(2):3. doi: 10.1007/s11910-020-1022-z.

PURPOSE OF REVIEW: Cerebral palsy is the most common physical disability of childhood, but the rate is falling, and severity is lessening. We conducted a systematic overview of best available evidence (2012-2019), appraising evidence using GRADE and the Evidence Alert Traffic Light System and then aggregated the new findings with our previous 2013 findings. This article summarizes the best available evidence interventions for preventing and managing cerebral palsy in 2019. RECENT FINDINGS: Effective prevention strategies include antenatal corticosteroids, magnesium sulfate, caffeine, and neonatal hypothermia. Effective allied health interventions include acceptance and commitment therapy, action observations, bimanual training, casting, constraint-induced movement therapy, environmental enrichment, fitness training, goal-directed training, hippotherapy, home programs, literacy interventions, mobility training, oral sensorimotor, oral sensorimotor plus electrical stimulation, pressure care, stepping stones triple P, strength training, task-specific training, treadmill training, partial body weight support treadmill training, and weight-bearing. Effective medical and surgical interventions include anti-convulsants, bisphosphonates, botulinum toxin, botulinum toxin plus occupational therapy, botulinum toxin plus casting, diazepam, dentistry, hip surveillance, intrathecal baclofen, scoliosis correction, selective dorsal rhizotomy, and umbilical cord blood cell therapy. We have provided guidance about what works and what does not to inform decision-making, and highlighted areas for more research.

PMID: 32086598

Prevention and Cure

26. Treatment of Neonatal Hypoxic-Ischemic Encephalopathy with Erythropoietin Alone, and Erythropoietin Combined with Hypothermia: History, Current Status, and Future Research.
Oorschot DE, Sizemore RJ, Amer AR.

Int J Mol Sci. 2020 Feb 21;21(4). pii: E1487. doi: 10.3390/ijms21041487.

Perinatal hypoxic-ischemic encephalopathy (HIE) remains a major cause of morbidity and mortality. Moderate hypothermia (33.5 °C) is currently the sole established standard treatment. However, there are a large number of infants for whom this therapy is ineffective. This inspired global research to find neuroprotectants to potentiate the effect of moderate hypothermia. Here we examine erythropoietin (EPO) as a prominent candidate. Neonatal animal studies show that immediate, as well as delayed, treatment with EPO post-injury, can be neuroprotective and/or neurorestorative. The observed improvements of EPO therapy were generally not to the level of control uninjured animals, however. This suggested that combining EPO treatment with an adjunct therapeutic strategy should be researched. Treatment with EPO plus hypothermia led to less cerebral palsy in a non-human primate model of perinatal asphyxia, leading to clinical trials. A recent Phase II clinical trial on neonatal infants with HIE reported better 12-month motor outcomes for treatment with EPO plus hypothermia compared to hypothermia alone. Hence, the effectiveness of combined treatment with moderate hypothermia and EPO for neonatal HIE currently looks promising. The outcomes of two current clinical trials on neurological outcomes at 18-24 months-of-age, and at older ages, are now required. Further research on the optimal dose, onset, and duration of treatment with EPO, and critical consideration of the effect of injury severity and of gender, are also required.

PMID: 32098276