Interventions and Management

1. Hand-Arm Bimanual Intensive Therapy Improves Prefrontal Cortex Activation in Children With Hemiplegic Cerebral Palsy.

Surkar SM, Hoffman RM, Willett S, Flegle J, Harbourne R, Kurz MJ.


PURPOSE: To determine the changes in the prefrontal cortical (PFC) activation following hand-arm bimanual intensive therapy (HABIT) in children with hemiplegic cerebral palsy (HCP). METHODS: Nine children with HCP and 15 children who were developing typically participated in the study. Children with HCP received 50 hours of HABIT. We assessed pre- and post-HABIT PFC activation using functional near-infrared spectroscopy neuroimaging. Bimanual coordination and motor task performance were assessed using the Assisting Hand Assessment (AHA), the average number of shapes matched, the shape matching errors, the reaction time, the 9-hole peg test, and the box and blocks test. RESULTS: The PFC activation decreased following HABIT and became similar to what was seen in the children who were developing typically. Post-HABIT PFC activation improvements paralleled with the improvements seen in the AHA and the behavioral outcomes. CONCLUSION: HABIT potentially improves the PFC's involvement in the action planning of the upper extremity movements in children with HCP.

PMID: 29578992


OBJECTIVE: To introduce the Windmill-task, a new objective assessment to quantify the presence of mirror movements (MMs) in children with unilateral cerebral palsy (uCP). In children with uCP MMs are frequently observed. They are typically assessed with the observation-based Woods and Teuber scale (W&T). However, due to its subjective nature and variable administration, interpretation of data across studies is problematic. DESIGN: Prospective, observational, cohort pilot study. SETTING: Children with uCP were recruited from Monash Children's Hospital, Melbourne, Australia as a convenience sample from a cohort of children previously recruited for a larger study. PARTICIPANTS: Prospective cohort of 23 children with uCP (age range: 6-15y, mean age=10y5m, SD=2y7m). Inclusion criteria were diagnosis of uCP with a Manual Ability Classification System (MACS)20 level I-III. INTERVENTIONS: Not applicable. MAIN OUTCOME MEASURE(S): The concurrent validity of the Windmill-task is assessed and sensitivity and specificity for MM detection is compared between both assessments. To assess the concurrent validity, Windmill-data are compared to W&T-data using Spearman-rank (rho) correlations for two conditions (affected-hand-moving vs. less-affected-hand-moving). Sensitivity and specificity are compared by presenting the mean percentage of children being assessed inconsistently across both assessments. RESULTS: Outcomes of both assessments correlated significantly (affected-hand-moving: rho=.520; p=.005; less-affected-hand-moving: rho=.488; p=.009). However, many children displayed MMs on the Windmill-task, but not on the W&T (sensitivity: affected-
hand-moving:27.5%; less-affected-hand-moving:40.6%). Only two children displayed MMs on the W&T, but not on the Windmill-task (specificity: affected-hand-moving:2.9%; less-affected-hand-moving:1.4%). CONCLUSIONS: The Windmill-task seems to be a valid tool to assess MMs in children with uCP and has additional advantage of sensitivity to detect MMs.

PMID: 29580937

3. Commentary on "Hand-Arm Bimanual Intensive Therapy Improves Prefrontal Cortex Activation in Children With Hemiplegic Cerebral Palsy".

Harpster K, Foci F.
[No abstract available]

PMID: 29578993


Trac J, Dawe J, Likitlersuang J, Musselman KE, Zariffa J.

Previous studies showed success using wrist-worn accelerometers to monitor upper-limb activity in adults and children with hemiparesis. However, a knowledge gap exists regarding which specific joint movements are reflected in accelerometry readings. We conducted a case series intended to enrich data interpretation by characterizing the influence of different pediatric upper-limb movements on accelerometry data. Approach: The study recruited 6 typically developing children and 5 children with hemiparetic cerebral palsy. The participants performed unilateral and bilateral activities, and their upper limb movements were measured with wrist-worn accelerometers and the Microsoft Kinect, a markerless motion-capture system that tracks skeletal data. The Kinect data were used to quantify specific upper limb movements through joint angle calculations (trunk, shoulder, elbow and wrist). Correlation coefficients (r) were calculated to quantify the influence of individual joint movements on accelerometry data. Regression analyses were performed to examine multi-joint patterns and explain variability across different activities and participants. Main results: Single-joint correlation results suggest that pediatric wrist-worn accelerometry data are not biased to particular individual joint movements. Rather, the accelerometry data could best be explained by the movements of the joints with the most functional relevance to the performed activity. Significance: This case series provides deeper insight into the interpretation of wrist-worn accelerometry data, and supports the use of this tool in quantifying functional upper-limb movements in pediatric populations.

PMID: 29578452

5. Impaired muscle growth precedes development of increased stiffness of the triceps surae musculotendinous unit in children with cerebral palsy.

Willerslev-Olsen M, Choe Lund M, Lorentzen J, Barber L, Kofoed-Hansen M, Nielsen JB.

AIM: If increased muscle stiffness and contractures in children with cerebral palsy (CP) are related to impaired muscle growth, reduced muscle growth should precede or coincide with increased muscle stiffness during development. Here, we compared the volume of the medial gastrocnemius muscle and the passive (non-neural) stiffness of the triceps surae musculotendinous unit in typically developing children and children with CP from birth until 4 years of age. METHOD: Forty-one children with CP and 45 typically developing children were included. Freehand three-dimensional ultrasound was used to evaluate the volume of the medial gastrocnemius muscle. Biomechanical and electrophysiological measures were used to determine passive and reflex mediated stiffness of the triceps surae musculotendinous unit. RESULTS: Medial gastrocnemius muscle volume increased with the same rate in typically developing and children with CP until 12 months of age, when a significant smaller rate of growth was observed in children with CP. Passive stiffness of the triceps surae musculotendinous unit showed a linear increase with age in typically developing children. Children with CP older than 27 months showed a significant increase in passive stiffness. Reflex mediated stiffness was only pathologically increased in four children with CP. INTERPRETATION: The deviation of medial gastrocnemius muscle volume, earlier than musculotendinous unit stiffness, is consistent with the hypothesis. The data also point out that muscle atrophy and muscle stiffness already develops within the first 1 to 2 years. This emphasizes the
6. Use of Hybrid Assistive Limb (HAL®) for a postoperative patient with cerebral palsy: a case report.


BACKGROUND: The Hybrid Assistive Limb (HAL®) is an exoskeleton wearable robot suit that assists in voluntary control of knee and hip joint motion. There have been several studies on HAL intervention effects in stroke, spinal cord injury, and cerebral palsy. However, no study has investigated HAL intervention for patients with cerebral palsy after surgery. CASE PRESENTATION: We report a case of using HAL in a postoperative patient with cerebral palsy. A 15-year-old boy was diagnosed with spastic diplegia cerebral palsy Gross Motor Function Classification System level IV, with knee flexion contracture, equinus foot, and paralysis of the right upper extremity with adduction contracture. He underwent tendon lengthening of the bilateral hamstrings and Achilles tendons. Although the flexion contractures of the bilateral knees and equinus foot improved, muscle strength decreased after the soft tissue surgery. HAL intervention was performed twice during postoperative months 10 and 11. Walking speed, stride, and cadence were increased after HAL intervention. Post HAL intervention, extension angles of the knee in stance phase and hip in the pre-swing phase were improved. In the gait cycle, the proportion of terminal stance in the stance and swing phase was increased. CONCLUSIONS: Hybrid Assistive Limb intervention for postoperative patients with cerebral palsy whose muscle strength decreases can enhance improvement in walking ability. Further studies are needed to examine the safety and potential application of HAL in this setting.

PMID: 29587833

7. Head Control Changes After Headpod Use in Children With Poor Head Control: A Feasibility Study.

Brown JE, Thompson M, Brizzolara K.


PURPOSE: To determine the feasibility of Headpod use to improve head control in children with cerebral palsy classified as Gross Motor Function Classification System level V. METHODS: Children (3-11 years) with poor head control were video recorded before and after 3 and 6 months of Headpod use. Head control without Headpod was measured by active time children could hold head upright and number of head bobs in 5 minutes. RESULTS: Fourteen children enrolled with 43% attrition rate. While average active time increased, average head bobs decreased and there were significant improvements in active time, but not head bobs. Active times were higher at 6 months compared with 3 months and baseline. Surveyed parents reported that the changes were apparent, supporting the large quantitative treatment effect. CONCLUSIONS: Six months of Headpod use, 45 minutes per day, appears to be feasible, has a large effect on active head control, and merits a larger randomized controlled trial.

PMID: 29579004

8. Targeted Training in Managing Children With Poor Trunk Control: 4 Case Reports.

Pin TW, Butler PB, Shum SL.


PURPOSE: This study investigated whether targeted training was feasible with young children younger than 2 years with poor trunk control due to cerebral palsy or developmental delay without using specialized equipment and the overall change in trunk control and functional ability, with the addition of targeted training to standard care physical therapy. SUMMARY OF KEY POINTS: Four children younger than 24 months with motor disorders were recruited. No difficulty was identified in using targeted training with this group. Within 3 months of commencing intervention, the 4 children had increased trunk control and functional abilities including independent sitting and bilateral hand use in play. CONCLUSIONS AND
RECOMMENDATIONS FOR CLINICAL PRACTICE: This form of targeted training was feasible for young children with motor disorders and augmented standard care therapy in enhancing both trunk control and motor skills. Targeted training should be further investigated as a potentially effective treatment.

**PMID: 29579009**


Gmelig Meyling C, Ketelaar M, Kuijper MA, Voorman J, Buizer AI.


PURPOSE: To review the existing literature on the effects of postural management on hip migration in children with cerebral palsy. METHODS: A systematic literature search was performed using 5 databases. Quality of articles was assessed and study designs were appraised according to the American Academy of Cerebral Palsy and Developmental Medicine Systematic Review Method. RESULTS: Eight of 655 identified studies were included, reporting postural management in children with cerebral palsy. Seven of 8 studies reported positive effects on hip migration after postural management interventions. However, level of evidence and quality of the articles were low. CONCLUSION: The evidence for postural management to prevent or reduce hip migration in children with cerebral palsy is limited by the lack of high-quality studies. Strong recommendations for clinical practice are not possible. Future high-quality research is crucial to improve our understanding of the effects of postural management to prevent hip migration in children with cerebral palsy.

**PMID: 29578990**

10. Commentary on "Effects of Postural Management on Hip Migration in Children With Cerebral Palsy: A Systematic Review".

Zipp GP, O'Connell L.


[No abstract available]

**PMID: 29578991**

11. Selective percutaneous muscle lengthening in cerebral palsy: when there is little or no evidence.

Chambers HG.


[No abstract available]

**PMID: 29574713**


Thomason P, Tan A, Donnan A, Rodda J, Graham HK, Narayanan U.


AIM: We investigated the validity of the Gait Outcomes Assessment List (GOAL), as an assessment of gait function in children with cerebral palsy (CP). METHOD: We studied a prospective cohort of 105 children with CP (Gross Motor Function Classification System [GMFCS] levels I-III; 65 males, 40 females; mean [SD] age 11y 11mo [3y 5mo], range 6-20y), who attended gait assessment over a 10-month period. Parents completed the GOAL, Functional Mobility Scale (FMS), and Functional Assessment Questionnaire (FAQ) during their child's gait evaluation. Ninety children completed instrumented gait analysis (IGA). Total GOAL and domain scores, Gait Profile Score (GPS), and Gait Variable Scores were calculated. RESULTS: The total GOAL discriminated between GMFCS levels (mean [SD] GMFCS level I, 72.5 [12.7]; GMFCS level II,
61.4 [13.0]; GMFCS level III, 38.8 [10.6]; [F2,97 =42.4, p<0.001]). Moderate correlations were found between total GOAL and FMS (5m and 50m r=0.59; 500m r=0.66) and FAQ walking (r=0.77) and activities list (r=0.75, p<0.01). There was a moderate negative correlation between total GOAL and GPS (r=-0.59) and gait appearance domain and GPS (r=-0.52, p<0.01).

INTERPRETATION: The GOAL is a valid assessment of gait function in ambulant children with CP. It has the potential to improve understanding of the child's and parents' priorities and thus, in conjunction with IGA, provide a more balanced assessment across the domains of the World Health Organization's International Classification of Functioning, Disability and Health. WHAT THIS PAPER ADDS: The Gait Outcomes Assessment List (GOAL) can discriminate between Gross Motor Function Classification System levels. The GOAL correlates with standard functional assessments and gait analysis. Used with gait analysis, the GOAL provides comprehensive assessment across all International Classification of Functioning, Disability and Health domains.

PMID: 29573409


Davids J.
[This commentary is on the original article by Thomason et al.]
PMID: 29574696

14. Gait research in cerebral palsy: should we zoom in or zoom out?

Givon U
[No abstract available]
PMID: 29600510

15. A Pediatric Service-Learning Program in Physical Therapy Education.

Hou YJ, Liu WY, Lin YH, Lien HY, Wong AMK, Chen CM.
PURPOSE: The purpose of this report was to describe a pediatric service-learning program in entry-level physical therapy (PT) education in Taiwan. KEY POINTS: To meet the needs of the local community and provide preclinical service-learning experience to PT students with people with physical disability, a service-learning program of a 2-day camp for children with cerebral palsy (CP) was developed in 2012. To date, 356 entry-level PT students have participated in this program, serving 286 children and their families. Important professional attributes, identified by the World Confederation for Physical Therapy guideline, such as altruism, compassion and caring, cultural competence, personal and professional development, professional duty, social responsibility and advocacy, and teamwork, were in the reflective reports of some of the PT students. CONCLUSIONS: The experiences provided by this pediatric PT service-learning program appear to have the potential to foster the development of professional attributes in entry-level PT students.

PMID: 29579005


Swinnen E.
[No abstract available]
PMID: 29573390
17. Threshold values of ankle dorsiflexion and gross motor function in 60 children with cerebral palsy.

Rasmussen HM, Svensson J, Thornig M, Pedersen NW, Overgaard S, Holsgaard-Larsen A.


Background and purpose - Threshold values defining 3 categories of passive range of motion are used in the Cerebral Palsy follow-Up Program to guide clinical decisions. The aim of this study was to investigate the threshold values by testing the hypothesis that passive range of motion in ankle dorsiflexion is associated with gross motor function and that function differs between the groups of participants in each category. Patients and methods - We analyzed data from 60 ambulatory children (aged 5-9 years) with spastic cerebral palsy. Outcomes were passive range of motion in ankle dorsiflexion with flexed and extended knee and gross motor function (Gait Deviation Index, Gait Variable Score of the ankle, peak dorsiflexion during gait, 1-minute walk, Gross Motor Function Measure, the Pediatric Quality of Life Inventory Cerebral Palsy Module, and Pediatric Outcomes Data Collection Instrument). Results - Significant (p < 0.05) and moderate correlations were documented for range of motion versus Gait Variable Score of the ankle (r = -0.37 and r = -0.37) and range of motion versus peak dorsiflexion (r = 0.49 and r = 0.55). Differences between the groups formed by the categories were shown for Gait Variable Score of the ankle and peak dorsiflexion (p < 0.05). No other significant correlations or differences between the categories were observed. Interpretation - The results suggest that threshold values for ankle dorsiflexion used in the Cerebral Palsy follow-Up Program are of limited clinical value in assessing overall gross motor function, but may be used to identify deviations in ankle-specific gait function.

PMID: 29589480

18. "Appropriate Treatment" and Therapeutic Window in Spasticity Treatment with IncobotulinumtoxinA: From 100 to 1000 Units.

Ianieri G, Marvulli R, Gallo GA, Fiore P, Megna M.


Many neurological diseases (ischemic and hemorrhagic stroke, multiple sclerosis, infant cerebral palsy, spinal cord injuries, traumatic brain injury, and other cerebrovascular disorders) may cause muscle spasticity. Different therapeutic strategies have been proposed for the treatment of spasticity. One of the major treatments for tone modulation is botulinum toxin type A (BTX-A), performed in addition to other rehabilitation strategies based on individualized multidisciplinary programs aimed at achieving certain goals for each patient. Therapeutic plans must be precisely defined as they must balance the reduction of spastic hypertonia and retention of residual motor function. To perform and optimize the treatment, an accurate clinical and instrumental evaluation of spasticity is needed to determine how this symptom is invalidating and to choose the best doses, muscles and times of injection in each patient. We introduce an "appropriate treatment" and no "standard or high dosage treatment" concept based on our retrospective observational study on 120 patients lasting two years, according to the larger Therapeutic Index and Therapeutic Window of Incobotulinumtoxin A doses from 100 to 1000 units. We studied the efficiency and safety of this drug considering the clinical spasticity significance for specialist physicians and patients.

PMID: 29597251

19. The feasibility of assessing speech and non-speech function of the speech apparatus in adults with cerebral palsy.

Schölderle T, Staiger A, Ziegler W.


This short note reports on observations concerning the feasibility of a set of speech and non-speech assessment tasks in an investigation of dysarthria in 21 adults (15 males/6 females; median 23 years) with cerebral palsy and concomitant cognitive impairment. The participants were assessed with nine tasks representing standard components of clinical dysarthria assessment (i.e. six speech and three non-speech tasks). The tasks were evaluated for their feasibility on the basis of common clinical criteria. Our results indicated that, overall, speech tasks were more feasible than non-speech tasks. Several participants showed signs of dysexecutive behaviour on some of the non-speech tasks, but not on the speech tasks. The current note provides tentative clues regarding the impact of cognitive deficits on the feasibility of assessment tasks in the diagnosis of dysarthria.

PMID: 29580108

Brunton LK.

PURPOSE: To describe the effect of fatigue and self-management practices for adolescents and young adults with cerebral palsy. METHODS: A survey of 124 people with cerebral palsy with the Fatigue Impact and Severity Self-Assessment. RESULTS: Participants in Gross Motor Function Classification System (GMFCS) level I experienced little effect of fatigue, with high proportions of this group disagreeing to statements about fatigue impacting their general activities, mobility, and social activities. Participants in GMFCS levels II to V reported effect of fatigue on activities. Differences between groups were evident in questions related to fatigue interference with length of time for physical activity and with motivations to participate in social activities. All other items related to management of fatigue were not significantly different between groups. CONCLUSIONS: Fatigue effect is greater for participants with more functional limitations. The lack of significant differences between groups, on the Management and Activity Modification subscale, indicates more research is needed regarding strategies for fatigue management.

PMID: 29579002


Goldsbury C, Greve K.
[No abstract available]
PMID: 29579003

22. Commentary on "Linking the Pediatric Evaluation of Disability Inventory-Computer Adaptive Test (PEDI-CAT) to the International Classification of Function".

Fragala-Pinkham M, Fehlner A.
[No abstract available]
PMID: 29578998

23. The risk, burden, and management of non-communicable diseases in cerebral palsy: a scoping review.

Ryan JM, Allen E, Gormley J, Hurvitz EA, Peterson MD.

AIM: To examine the risk, burden, and management of non-communicable diseases (NCDs) among people with cerebral palsy (CP). METHOD: Databases (Ovid MEDLINE, Embase Ovid, CINAHL Plus) were systematically searched up to August 2017. Data on the prevalence of risk factors for, and the burden and management of, cardiovascular diseases, diabetes, cancers, and respiratory diseases were extracted. RESULTS: Thirty-six studies that examined the prevalence of risk factors among people with CP were identified. There was inconsistent evidence that people with CP had higher prevalence of metabolic risk factors such as hypertension, hyperlipidaemia, and obesity, but strong evidence that they participated in low levels of physical activity, compared with people without CP. Seven studies reported on the burden of NCDs. Adults with CP had a higher risk of NCDs, including stroke, chronic obstructive pulmonary disease, and other heart conditions, and death due to NCDs, including cancers, chronic obstructive pulmonary disease, stroke, and ischaemic heart disease, compared with the general population. Only one study reported on the management of NCD, specifically the uptake of breast cancer screening among females. INTERPRETATION: The burden of NCDs is higher among adults with CP compared with the general population. Further research is required to determine the prevalence of metabolic risk factors and management of NCDs among people with CP. WHAT THIS PAPER ADDS: Adults with cerebral palsy (CP) have an increased risk of non-communicable diseases (NCDs) and increased risk of death because of NCDs. Evidence is inconsistent about the elevated prevalence of metabolic risk factors.
for NCDs. Evidence is consistent that people with CP participate in reduced physical activity. Only one study reported on management of NCD among people with CP. Available evidence suggests people with CP are less likely to receive preventive medicine.

PMID: 29572812


INTRODUCTION: The diagnostic proof of fungal infection in preterm infants is difficult. Antifungal treatment (AFT) is often initiated empirically when infants with suspected infection do not improve despite broad spectrum antibiotic therapy. It was the aim of our study to determine the rate of exposure to empirical AFT in a large cohort of very low birth weight infants (VLBW) of the German Neonatal Network (GNN) and to address associated risks and outcomes. METHODS: The epidemiological database consisted of n= 13,343 VLBWI born in 54 GNN centers between 2009 and 2015. AFT was defined as number of neonates who got any dose of at least one of the following antifungal drugs: Fluconazole, Amphotericin B, Voriconazole and Caspofungin (denominator: number of infants enrolled in GNN) for treatment (not prophylaxis) of (suspected) fungal infection. Univariate and logistic regression analyses were used to identify risk factors for exposure to AFT as well as associated short- and long-term morbidities and long-term outcomes at 5 year-follow-up. RESULTS: In our cohort, 724/13,343 (5.4%) VLBWI were exposed to empiric antifungal treatment, and had a mean gestational age of 25.7 (+ 2.1) weeks. 44/13,343 (0.3%) had proven bloodstream infection with Candida spp. The main risk factors for exposure to AFT were gestational age, postnatal steroid treatment, need for abdominal surgery and use of carbapenems. Notably, antifungal treatment was associated with adverse outcomes such as bronchopulmonary dysplasia (BPD; adjusted OR 1.9, 95% CI 1.6-2.3, p < 0.001) and Retinopathy of prematurity requiring intervention (ROP; adjusted OR 1.69, 95% CI 1.3-2.3, p <0.001) but not mortality. In the subgroup of infants available for 5-year-follow-up (n= 895), exposure to antifungal treatment was associated with a risk for cerebral palsy (CP; adjusted OR 2.79, 95% CI 1.11-7.04, p = 0.049) and IQ < 85 (adjusted OR 2.07, 95% CI 1.01-4.28, p = 0.049). CONCLUSION: A significant proportion of VLBWI is exposed to AFT, specifically those born < 26 weeks. Exposed infants were found to have a higher risk for adverse outcomes which may reflect their significant vulnerability in general. Given the observational design of our study, it remains unclear whether potential side effects of empirical or target antifungal treatment itself contribute to adverse outcome. Future studies need to include risk-based strategies and stewardship programs to restrict the use of antifungal management in VLBWI.

PMID: 29601449

25. Risk Factor of Mortality in Indonesian Children with Cerebral Palsy.

Prastiya IG, Risky VP, Mira I, Retno AS, Darto S, Erny P.


OBJECTIVE: Individuals with CP have a lower life expectancy than the general population. The objective of the study is to investigate the risk factor of mortality in Indonesian children with CP. METHODS: An observational analytic study was conducted using medical records at pediatric ward Dr. Soetomo Hospital, Surabaya, Indonesia, from January 2014 to December 2016. Inclusion criteria was all of CP patients in that were hospitalized from 6 months to 14 year of age. Mortality information was obtained from annual computer and matched against the subjects with CP on the basis of name, date of birth, type of disability, degree of disability, nutritional status and the outcomes. Risk factors were counted using logistic regression. RESULT: Fifty five children were enrolled. The outcome revealed 12 patients (21.8%) were death. Pneumonia was the most common underlying disease cause of death (OR=5.185;95% CI 1.249 to 21.520;Psize 8 < 0.007). Other risk factors of mortality that significant were acute kidney injury (AKI) (OR=3.333;95% CI 1.317 to 8.436;Psize 8 < 0.03), and Gross Motor Functioning Classification System (GMFCS) more than level IV (OR=1.480;95% CI 1.184 to 1.850;Psize 8 < 0.006). CONCLUSION: Pneumonia, AKI and severe GMFCS level were risk factors of death in patients with CP. J. Med. Invest. 65:18-20, February, 2018.

PMID: 29593188
26. Epidemiology of Cerebral Palsy in Northeastern Switzerland.

Forni R, Stojicevic V, van Son C, Lava SAG, Kuenzle C, Beretta-Piccoli M.


PURPOSE: Cerebral palsy (CP) is the most common motor disability of childhood. Less is known about its prevalence and associated factors in Switzerland, so we aimed to fill this knowledge gap in one Swiss canton, evaluating the feasibility to build up a CP registry. METHODS: A retrospective review of medical records was undertaken. Children born during 1995 and 2009 in the canton of Saint Gallen diagnosed with CP were eligible for inclusion. RESULTS: A total of 140 patients formed the study cohort. The prevalence of CP was 0.19%, with 85% of children affected by spastic CP. CONCLUSIONS: The results of the present study were comparable with data of the Surveillance of Cerebral Palsy in Europe network. Moreover, this pilot study demonstrated the feasibility and usefulness of creating a registry of children affected by CP.

PMID: 29579006

27. A sustainable solution for the activities of the European network for surveillance of congenital anomalies: EUROCAT as part of the EU platform on rare diseases registration.


The Joint Research Centre (JRC) in collaboration with the Directorate-General for Health and Food Safety (DG SANTE) is developing the European Platform on Rare Diseases Registration (EU RD Platform) with the objective to set European-level standards for data collection and data sharing. In the field of rare diseases the EU RD Platform will be a source of information on available rare disease patient data with large transnational European coverage. One main function of the EU RD Platform is to enable interoperability for the >600 existing RD registries in Europe. The second function is to offer a sustainable solution for two large European surveillance networks: European Surveillance of Congenital Anomalies (EUROCAT) and Surveillance of Cerebral Palsy in Europe (SCPE). EUROCAT is European network of population-based registries for the epidemiological surveillance of congenital anomalies. It covers about one third of the European birth population. The Central Database contains about 800,000 cases with congenital anomalies among livebirths, stillbirths and terminations of pregnancy, reported using the same standardised classification and coding. These high quality data enables epidemiological surveillance of congenital anomalies, which includes estimating prevalence, prenatal diagnosis and perinatal mortality rates and the detection of teratogenic exposures among others. The network also develops recommendations for primary prevention in the Rare Diseases National Plans for medicinal drugs, food/nutrition, lifestyle, health services, and environmental pollution. The network has received the European Commission's (EC) support since its inception. In order to offer a sustainable solution for the continuation of EUROCAT activities, it was agreed that EUROCAT would become part of the EU RD Platform. In 2015, the European level-coordination activities and the Central Database were transferred to the JRC, where the JRC-EUROCAT Central Registry is now located. This paper describes the functioning of EUROCAT in the new setting, and gives an overview of the activities and the organisation of the JRC-EUROCAT Central Registry.

PMID: 29597096


Broström L, Vollmer B, Bolk J, Eklöf E, Ådén U.


AIM: To study the prevalence of minor neurological dysfunction (MND) at 6 years of age in a cohort of children born extremely preterm without cerebral palsy (CP) and to investigate associations with motor function, cognitive abilities, and behaviour. METHOD: This study assessed 80 children born at less than 27 weeks of gestation and 90 children born at term age between 2004 and 2007 at a mean age of 6 years 6 months. The assessments included a simplified version of the Touwen Infant Neurological Examination, the Movement Assessment Battery for Children, Second Edition (MABC-2), Wechsler Intelligence Scale for Children, Fourth Edition (WISC-IV), the Strengths and Difficulties Questionnaire (SDQ), and the parent version of the Five to Fifteen questionnaire. RESULTS: Fifty-one of the children born preterm had normal neurology, 23 had simple MND, and six had complex MND compared with 88 who had normal neurology and two simple MND in the term-born group (p=0.001). There were significant differences between the children with normal neurology and MND in the preterm group in MABC-2-assessed motor function (p≤0.001), general cognitive abilities with WISC-IV (p=0.005), and SDQ overall behavioural problems and peer problems reported by the parents (p=0.021 and p=0.003 respectively). SDQ teacher-reported
overall behavioural and hyperactivity problems were significantly different between children with normal and simple MND (p=0.036 and p=0.019). INTERPRETATION: Children born extremely preterm, in the absence of CP, are at risk of MND and this is associated with motor function, cognitive ability, and behaviour. WHAT THIS PAPER ADDS: Extremely preterm birth carries a risk of minor neurological dysfunction (MND). MND in children born extremely preterm is associated with impaired motor function and cognitive abilities, and behavioural problems. Male sex is associated with MND in children born extremely preterm.

PMID: 29573402

29. Muscle contractures in patients with cerebral palsy and acquired brain injury are associated with extracellular matrix expansion, pro-inflammatory gene expression, and reduced rRNA synthesis.


INTRODUCTION: Children with cerebral palsy (CP) and acquired brain injury (ABI) commonly develop muscle contractures with advancing age. An underlying growth defect contributing to skeletal muscle contracture formation in CP/ABI has been suggested. METHODS: The biceps muscles of children and adolescents with CP/ABI (n=20) and typically developing controls (n=10) were investigated. We used immunohistochemistry, qRT-PCR and western blotting to assess gene expression relevant to growth and size homeostasis. RESULTS: Classical pro-inflammatory cytokines and genes involved in extracellular matrix production were elevated in skeletal muscle of children with CP/ABI. Intramuscular collagen content was increased and satellite cell number decreased and this was associated with reduced levels of RNA polymerase (POL) I transcription factors, 45s pre-rRNA and 28S rRNA. DISCUSSION: The present study provides novel data suggesting a role for pro-inflammatory cytokines and reduced ribosomal production in the development/maintenance of muscle contractures; possibly underlying stunted growth and perimysial extracellular matrix expansion. This article is protected by copyright. All rights reserved.

PMID: 29572878