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Interventions and Management

1. The effects of modified constraint-induced movement therapy combined with intensive bimanual training in children with brachial plexus birth injury: a retrospective data base study.
Zielinski IM, van Delft R, Voorman JM, Geurts ACH, Steenbergen B, Aarts PBM.

Disabil Rehabil. 2019 Dec 8:1-10. doi: 10.1080/09638288.2019.1697381. [Epub ahead of print]

Background: In children with Brachial Plexus Birth Injury, a non-use of the affected upper limb despite sufficient capacity, is sometimes observed, called developmental disregard. The combination of modified constraint-induced-movement-therapy with bimanual training is frequently applied to overcome developmental disregard in unilateral Cerebral Palsy. In the current study the effects of the combination of modified constraint-induced-movement-therapy with bimanual training are investigated in children with Brachial Plexus Birth Injury in comparison to children with unilateral Cerebral Palsy. We hypothesize that the combination of modified constraint-induced-movement-therapy with bimanual training is effective in Brachial Plexus Birth Injury. Methods: Data of 19 children with Brachial Plexus Birth Injury (Mage: 4.1 years) and 18 with unilateral Cerebral Palsy (Mage: 4.5 years) were compared. The effects of modified constraint-induced-movement-therapy with bimanual training (54 h modified constraint-induced-movement-therapy, 18 h bimanual training, 8-10 weeks) was investigated by assessing spontaneous affected-upper-limb-use ("Assisting Hand Assessment"), manual abilities ("ABILHAND-kids") and subjective performance and satisfaction of problematic bimanual activities ("Canadian Occupational Performance Measure") at three time points (pre-treatment, post-treatment, follow-up). This data was analyzed using repeated-measures analysis. Results: Children with Brachial Plexus Birth Injury showed significant improvements on all outcome measures following modified constraint-induced-movement-therapy with bimanual training. These results were comparable to those observed in the group of children with unilateral Cerebral Palsy. Discussion: These results suggest that modified constraint-induced-movement-therapy with bimanual training is effective in Brachial Plexus Birth Injury. They indicate a comparable improved bimanual performance in children with Brachial Plexus Birth Injury than in unilateral Cerebral Palsy and suggests that both groups of children have affectively overcome their developmental disregard. IMPLICATIONS FOR REHABILITATION Children with Brachial Plexus Birth Injury frequently experience difficulties in activities of daily living. It has recently been suggested that children with Brachial Plexus Birth Injury may also show a non-use of the affected upper limb despite sufficient capacity, called developmental disregard. Children with Brachial Plexus Birth Injury and developmental disregard might therefore benefit from intensive therapies aimed at overcoming developmental disregard, originally developed for children with unilateral Cerebral Palsy. A combination of modified Constrained-Induced Movement Therapy with intensive Bimanual Training has shown to be affective in children with unilateral Cerebral Palsy. In a small sample, this study shows that a combination of modified Constrained-Induced Movement Therapy with intensive bimanual training is effective in children with Brachial Plexus Birth Injury, comparable and even more than in unilateral Cerebral Palsy.

PMID: 31814455

2. Fully automated image-based estimation of postural point-features in children with cerebral palsy using deep learning.
Cunningham R, Sánchez MB, Butler PB, Southgate MJ, Loram ID.

R Soc Open Sci. 2019 Nov 6;6(11):191011. doi: 10.1098/rsos.191011. eCollection 2019 Nov.

The aim of this study was to provide automated identification of postural point-features required to estimate the location and orientation of the head, multi-segmented trunk and arms from videos of the clinical test 'Segmental Assessment of Trunk Control' (SATCo). Three expert operators manually annotated 13 point-features in every fourth image of 177 short (5-10 s) videos (25 Hz) of 12 children with cerebral palsy (aged: 4.52 ± 2.4 years), participating in SATCo testing. Linear interpolation for the remaining images resulted in 30 825 annotated images. Convolutional neural networks were trained with cross-validation, giving held-out test results for all children. The point-features were estimated with error 4.4 ± 3.8 pixels at approximately 100 images per second. Truncal segment angles (head, neck and six thoraco-lumbar-pelvic segments) were estimated with error 6.4 ± 2.8°, allowing accurate classification (F 1 > 80%) of deviation from a reference posture at thresholds up to 3°, 3° and 2°, respectively. Contact between arm point-features (elbow and wrist) and supporting surface was classified at F 1 = 80.5%. This study demonstrates, for the first time, technical feasibility to automate the identification of (i) a sitting segmental posture including individual trunk segments, (ii) changes away from that posture, and (iii) support from the upper limb, required for the clinical SATCo.

PMID: 31827842

3. Do seizures compromise correction maintenance after spinal fusion in cerebral palsy scoliosis?
Abousamra O, Sullivan BT, Shah SA, Yaszay B, Samdani AF, Cahill PJ, Newton PO, Sponseller PD.

J Pediatr Orthop B. 2019 Dec 9. doi: 10.1097/BPB.0000000000000705. [Epub ahead of print]

Seizure disorder in cerebral palsy (CP) has been described as a risk factor for postoperative complications after posterior spinal fusion. However, the effect of seizures on the maintenance of curve correction has not been reported. The aim of this study is to investigate associations between seizure history and maintenance of curve correction after posterior spinal fusion in children with CP. We analyzed records of 201 children with CP who underwent posterior spinal fusion with two-year follow-up. Patients were classified as having no seizures (31%); controlled seizures (54%); or poorly controlled seizures (PCS, 15%). Perioperative data, radiographic measurements, and complications were compared between groups. Groups were similar in operative time, estimated blood loss, and rates of deep wound infection and implant-related complications. The PCS group had a higher rate of respiratory complications (27%) than the no seizures (10%; P = 0.03) and controlled seizures (12%; P = 0.04) groups. Controlled seizures and PCS groups had longer ICU stays than the no seizures group (P = 0.02 and P = 0.04). Major coronal curve and pelvic obliquity were corrected significantly in all groups, and correction was maintained at 2 years. Loss of correction during follow-up was similar between groups. Although seizures were associated with longer ICU stays and more respiratory complications, there was no association between seizure history and loss of curve correction at two years of follow-up after CP scoliosis surgery.

PMID: 31821271

4. Measuring dystonia and choreoathetosis in dyskinetic cerebral palsy.
Vanmechelen I, Dan B, Feys H, Monbaliu E.

Dev Med Child Neurol. 2019 Dec 13. doi: 10.1111/dmcn.14424. [Epub ahead of print]

AIM: To assess test-retest reliability of the Dyskinesia Impairment Scale (DIS) in children and young adults with dyskinetic cerebral palsy (CP). METHOD: Dystonia and choreoathetosis were assessed in 15 participants with dyskinetic CP (13 males, 2 females; age range 5-22y, mean 14y, SD 4y) using the DIS in two separate sessions over 7 days. Exclusion criteria were changes in muscle relaxant medication within the previous 3 months, orthopaedic or neurosurgical interventions within the previous year, and spinal fusion. Intraclass correlation coefficient, confidence intervals (CI), standard error of measurement, and the minimal detectable difference (MDD) were determined for test-retest reliability. RESULT: Intraclass correlation coefficients of the DIS, the dystonia subscale of the DIS, and the choreoathetosis subscale of the DIS were 0.98 (95% CI 0.94-

0.99), 0.97 (95% CI 0.92-0.99), and 0.96 (95% CI 0.90-0.99). The standard error of measurement and MDD were 2.6% and 7.2%. INTERPRETATION: The DIS is a reliable tool to assess dystonia and choreoathetosis; it remains stable over time in children and young adults with dyskinetic CP. These results add to the current evidence for good clinimetric properties of the DIS. WHAT THIS PAPER ADDS: The Dyskinesia Impairment Scale (DIS) shows stability in scoring dystonia and choreoathetosis. The total DIS score and dystonia and choreoathetosis subscales are clinically useful.

PMID: 31833574

5. Incidence of Pediatric Venous Thromboembolism After Elective Spine and Lower-Extremity Surgery in Children With Neuromuscular Complex Chronic Conditions: Do we Need Prophylaxis?
Shore BJ, Hall M, Matheney TH, Snyder B, Trenor CC 3rd, Berry JG.

J Pediatr Orthop. 2019 Dec 9. doi: 10.1097/BPO.0000000000001483. [Epub ahead of print]

BACKGROUND: The incidence of venous thromboembolism (VTE) after elective surgery in children with mobility impairments, including those with a neuromuscular complex chronic condition (NCCC), is unknown. Therefore, our objectives were to assess the incidence of VTE after elective spine and lower-extremity surgery in children with NCCC. METHODS: A retrospective analysis of children with NCCC undergoing elective lower-extremity and/or spinal surgeries from 2005 to 2009 included in the Pediatric Health Information Systems Plus (PHIS+) database. VTE during hospitalization for surgery was assessed through abstraction and review of ultrasound (U/S) and computed tomography results by 2 independent reviewers. VTEs related to pre-existing central venous catheters were excluded. RESULTS: There were 4,583 children with NCCC who underwent orthopaedic surgery during the study period at 6 centers. Most were male (56.3%), non-Hispanic whites (72.7%), and had private insurance (52.2%). The most common NCCC diagnoses were cerebral palsy (46.7%), brain and spinal cord malformations (31.1%), and central nervous system degenerative conditions (14.5%). Forty children (0.9%) underwent U/S to assess VTE. Eighteen children (0.4%) underwent computed tomography to assess VTE. Four children (with cerebral palsy) had a positive U/S for a lower-extremity VTE (10-18 y of age), yet 2 had their VTE before surgery. Therefore, the adjusted VTE rate for children with NCCC undergoing orthopaedic lower-extremity or spine surgery was 4 per 10,000 (2 cases per 4583 surgeries). Each of the 2 cases had a known coagulation disorder preoperatively. Only 10% of the cohort used compression devices, 3% enoxaparin, and 1.6% aspirin for prophylaxis. CONCLUSION: The rate of non-central-venous-catheter-related VTE associated with orthopaedic surgery in children with NCCC is very low and lower than rates reported in healthy children. SIGNIFICANCE: To our knowledge, this is the first multi-institutional study reporting the incidence of VTE in children with NCCCs undergoing elective hip and spine surgery. These data support no additional prophylaxis is required in children with NCCC undergoing elective hip and spine surgery unless other known risk factors are also present.

PMID: 31821246

6. Effect of task-oriented training on balance and motor function of ambulant children with cerebral palsy.
Ogwumike OO, Badaru UM, Adeniyi AF.

Rehabilitacion (Madr). 2019 Oct - Dec;53(4):276-283. doi: 10.1016/j.rh.2019.07.003. Epub 2019 Nov 19.

INTRODUCTION AND OBJECTIVES: The study evaluated the effect of task-oriented training (TOT) on the motor function (MF) and balance of ambulant children with cerebral palsy (CP). MATERIALS AND METHODS: A total of 46 children were randomised into TOT group (n=23) and Control Group (CG [n=23]), but 39 children complete the study. Balance and MF were assessed at baseline, 6th and 12th weeks and 6 weeks post-intervention. Data were analysed with repeated measures ANOVA, Friedman's, Mann-Whitney U, Student's-t and post hoc tests at $\alpha \leq 0.05$. RESULTS: The two groups were comparable in all baseline scores ($P>0.05$). At the 6th week, significant between-group difference was observed in MF only [TOT=81.9 (18.5); CG=72.8 (19.4)] ($P<0.05$). There were significant between-group differences in MF [TOT=88.8 (9.4); CG=75.5 (18.5); $P<0.05$] and balance (TOT=9.4±4.5; CG=13.6±6.9; $P<0.05$) at the 12th week ($P<0.05$) and 6 weeks post-intervention ($P<0.05$). CONCLUSION: TOT improved the balance and MF of ambulant children with CP.

PMID: 31813423

7. Spasticity.
Li S, Francisco GE.

Handb Exp Pharmacol. 2019 Dec 10. doi: 10.1007/164_2019_315. [Epub ahead of print]

Spasticity is one component of the upper motor neuron (UMN) syndrome resulting from a multitude of neurologic conditions, such as stroke, brain injury, spinal cord injury, multiple sclerosis, and cerebral palsy. It is clinically recognized as a phenomenon of velocity-dependent increase in resistance, i.e., hypertonia. Recent advances in the pathophysiology of spasticity improve our understanding of mechanisms underlying this complex phenomenon and its relations to other components of UMN syndrome (weakness and disordered motor control), as well as the resultant clinical problems. This theoretical framework provides a foundation to set up treatment goals and to guide goal-oriented clinical assessment and treatment. Among a spectrum of treatment options, botulinum toxin (BoNT) therapy is the preferred treatment for focal spasticity. The evidence is very robust that BoNT therapy effectively reduces spasticity; however, it does not improve voluntary movement. In this chapter, we highlight a few issues on how to achieve the best clinical outcomes of BoNT therapy, such as dosing, dilution, guidance techniques, adjunctive therapies, early treatment, repeated injections, and central effects, as well as the ways to improve motor function in selected subgroups of patients with spasticity. We also discuss the reasons of poor responses to BoNT therapy and when not to use BoNT therapy.

PMID: 31820170

8. Association of Knee Pain and Crouch Gait in Individuals With Cerebral Palsy.
Pelrine E, Novacheck T, Boyer E.

J Pediatr Orthop. 2019 Dec 9. doi: 10.1097/BPO.0000000000001487. [Epub ahead of print]

BACKGROUND: Crouch gait (ie, excessive knee flexion) is commonly seen in patients with cerebral palsy (CP) and has been inconsistently linked with knee pain. The definitive cause of knee pain is unknown, but may result from increased joint forces due to crouch gait kinematics. Our purpose was to determine whether knee pain is positively associated with knee flexion in gait among a large sample of ambulatory individuals with CP. We hypothesized that knee pain prevalence would increase as knee flexion increased. METHODS: In this retrospective study, pain questionnaire and 3-dimensional gait analysis data from 2015 to 2018 were extracted from the medical records of individuals with CP who had a clinical gait analysis. The pain questionnaire asked caregivers/patients to indicate the location of pain and when it occurs. A multivariate logistic regression was performed with minimum knee flexion in stance, patella alta, age, and sex as predictors of knee pain. RESULTS: Among the 729 participants included in the analysis, 147 reported knee pain (20.1%). The odds of knee pain were not associated with minimum knee flexion in stance or sex. However, the odds of knee pain increased 73.2% when patella alta was present (P=0.008) and tended to increase 2.2% as age increased (P=0.059). CONCLUSIONS: The data suggest that there is not a meaningful association between crouch gait and knee pain. Having patella alta was associated with pain. Further studies that use validated pain questionnaires are needed to understand the multifactorial etiology of knee pain within ambulatory individuals with CP. LEVEL OF EVIDENCE: Level III-case-control study.

PMID: 31821247

9. A Personalized Approach to Improve Walking Detection in Real-Life Settings: Application to Children with Cerebral Palsy.
Carcreff L, Paraschiv-Ionescu A, Gerber CN, Newman CJ, Armand S, Aminian K.

Sensors (Basel). 2019 Dec 3;19(23). pii: E5316. doi: 10.3390/s19235316.

Although many methods have been developed to detect walking by using body-worn inertial sensors, their performances decline when gait patterns become abnormal, as seen in children with cerebral palsy (CP). The aim of this study was to evaluate if fine-tuning an existing walking bouts (WB) detection algorithm by various thresholds, customized at the individual or group level, could improve WB detection in children with CP and typical development (TD). Twenty children (10 CP, 10 TD) wore 4 inertial sensors on their lower limbs during laboratory and out-laboratory assessments. Features extracted from the

gyroscope signals recorded in the laboratory were used to tune thresholds of an existing walking detection algorithm for each participant (individual-based personalization: Indiv) or for each group (population-based customization: Pop). Out-of-laboratory recordings were analyzed for WB detection with three versions of the algorithm (i.e., original fixed thresholds and adapted thresholds based on the Indiv and Pop methods), and the results were compared against video reference data. The clinical impact was assessed by quantifying the effect of WB detection error on the estimated walking speed distribution. The two customized Indiv and Pop methods both improved WB detection (higher, sensitivity, accuracy and precision), with the individual-based personalization showing the best results. Comparison of walking speed distribution obtained with the best of the two methods showed a significant difference for 8 out of 20 participants. The personalized Indiv method excluded non-walking activities that were initially wrongly interpreted as extremely slow walking with the initial method using fixed thresholds. Customized methods, particularly individual-based personalization, appear more efficient to detect WB in daily-life settings.

PMID: 31816854

10. Measures of Motor and Functional Skills for Children With Cerebral Palsy: A Systematic Review.
Ferre-Fernández M, Murcia-González MA, Barnuevo Espinosa MD, Ríos-Díaz J.

Pediatr Phys Ther. 2019 Nov 27. doi: 10.1097/PEP.0000000000000661. [Epub ahead of print]

PURPOSE: To review the level of evidence of the psychometric properties of outcome measures for motor or functional skills for children with cerebral palsy classified across I to V levels of the Gross Motor Function Classification System. METHODS: A systematic search was completed in PubMed/MEDLINE, ISI Web of Science, CINAHL, and 4 complementary databases. The COSMIN Risk of Bias checklist and the updated criteria for good measurement properties were applied to assess the quality. RESULTS: Four outcome measures were identified from 12 articles: Gross Motor Function Measure, Gross Motor Performance Measure, Pediatric Evaluation of Disability Inventory, and Functional Independence Measure for Children. Evidence levels for validity, reliability, and responsiveness varied among measures. CONCLUSIONS: Gross Motor Function Measure in all versions was the most investigated measure providing the best results, with the strongest evidence for validity and responsiveness properties. Reliability evidence should be improved to determine stability.

PMID: 31815921

11. Neural ensemble dynamics in dorsal motor cortex during speech in people with paralysis.
Stavisky SD, Willett FR, Wilson GH, Murphy BA, Rezaii P, Avansino DT, Memberg WD, Miller JP, Kirsch RF, Hochberg LR, Ajiboye AB, Druckmann S, Shenoy KV, Henderson JM.

Elife. 2019 Dec 10;8. pii: e46015. doi: 10.7554/eLife.46015. [Epub ahead of print]

Speaking is a sensorimotor behavior whose neural basis is difficult to study with single neuron resolution due to the scarcity of human intracortical measurements. We used electrode arrays to record from the motor cortex 'hand knob' in two people with tetraplegia, an area not previously implicated in speech. Neurons modulated during speaking and during non-speaking movements of the tongue, lips, and jaw. This challenges whether the conventional model of a 'motor homunculus' division by major body regions extends to the single-neuron scale. Spoken words and syllables could be decoded from single trials, demonstrating the potential of intracortical recordings for brain-computer interfaces to restore speech. Two neural population dynamics features previously reported for arm movements were also present during speaking: a component that was mostly invariant across initiating different words, followed by rotatory dynamics during speaking. This suggests that common neural dynamical motifs may underlie movement of arm and speech articulators.

PMID: 31820736

12. Co-Occurrence of Rarest Type of Dysphagia Lusoria (Type N-1) and Eosinophilic Esophagitis in a Cognitively Disabled Individual.
Kumar K, Makker J, Tariq H, Ihimoyan A, Chukwunonso C, Niazi M, Lombino M, Kamal M, Patel HK.

Case Rep Med. 2019 Nov 11;2019:2890635. doi: 10.1155/2019/2890635. eCollection 2019.

Dysphagia is an expressive symptom, described by an individual as "difficulty in swallowing." Dysphagia due to esophageal compression from an aberrant right subclavian artery is rare, and it is termed as "dysphagia lusoria." We present a rare case of co-occurrence of dysphagia lusoria with esophageal eosinophilia in a patient with cognitive disability which portends a case with diagnostic challenge and treatment dilemma. A 31-year-old man with intellectual disability, cerebral palsy, previous history of feeding difficulty, and esophageal food impaction presented with esophageal foreign body impaction. He has no known history of atopy and food allergies. There was no laboratory evidence of peripheral eosinophilia. The IgE-mediated allergic test was unremarkable. His prior presentation revealed a diagnosis of eosinophilic esophagitis. The imaging studies showed proximal esophageal dilatation with extrinsic compression at the level of the upper esophagus. The foreign bodies were removed successfully through the help of upper endoscopy. Subsequent evaluation revealed a rare type of dysphagia lusoria (type N-1) due to an aberrant left subclavian artery arising from the right-sided aortic arch. The patient's family refused further management of artery lusoria. Prolonged stasis of secretions and food in the esophagus can also lead to increased esophageal eosinophils. In our case, it remains undetermined whether increased number of esophageal eosinophils resulted from primary eosinophilic esophagitis or due to prolonged food stasis from esophageal compression caused by an aberrant subclavian artery. However, food impaction right above the compression site makes dysphagia lusoria the likely etiology.

PMID: 31814829

13. The burden of sialorrhoea in chronic neurological conditions: current treatment options and the role of incobotulinumtoxinA (Xeomin®).
Morgante F, Bavikatte G, Anwar F, Mohamed B.

Ther Adv Neurol Disord. 2019 Nov 28;12:1756286419888601. doi: 10.1177/1756286419888601. eCollection 2019.

Sialorrhoea is a frequent symptom of neurological diseases (e.g. Parkinson's disease, motor neuron disease, cerebral palsy, and stroke) and is defined as excessive saliva accumulation leading to unintentional loss of saliva from the mouth. Sialorrhoea increases the overall burden on the patient and their caregivers, the impact of which can be both physical and psychosocial. Treatments for sialorrhoea range from lifestyle and behavioural guidance, to medications, surgery or radiation. Nonpharmacological interventions include advice on posture, swallowing control, cough management, dietary changes, eating and drinking techniques, and behavioural modification; however, these conservative measures may be ineffective for people with progressive neurological conditions. The pharmacological treatment of sialorrhoea is challenging because medications licensed for this purpose are limited, but treatments can include anticholinergic drugs and botulinum toxins. Surgical treatment of sialorrhoea is typically reserved as a last resort for patients. IncobotulinumtoxinA (Xeomin®) is the first botulinum toxin type A to receive US and UK marketing authorization for the symptomatic treatment of chronic sialorrhoea due to neurological disorders in adults. In this review, we discuss and compare the frequency and method of administration, location of treatment delivery, approximate annual costs and main side effects of botulinum toxin and different anticholinergic drugs. Management of patients with chronic neurological conditions requires input from multiple specialist teams and thus a multidisciplinary team (MDT) approach is considered fundamental to ensure that care is consistent and tailored to patients' needs. To ensure that adult patients with neurological conditions receive the best care and sialorrhoea is well managed, we suggest a potential clinical care pathway for sialorrhoea with a MDT approach, which healthcare professionals could aspire to.

PMID: 31819763

14. Long-Term Gastrocolocutaneous Fistula after Endoscopic Gastrostomy: How Concerned Should We Be?
Nunes G, Paiva de Oliveira G, Cruz J, Santos CA, Fonseca J.

GE Port J Gastroenterol. 2019 Oct;26(6):441-447. doi: 10.1159/000497248. Epub 2019 Apr 3.

Percutaneous endoscopic gastrostomy (PEG) is a safe technique for long-term enteral feeding. The most common PEG-associated adverse events are minor. Gastrocolocutaneous fistula (GCCF) results from misplacement of the PEG tube through the colon. The importance of this complication is not currently defined, and there is no clearly established therapeutic algorithm. The authors report a series of 3 cases of GCCF diagnosed and treated in a tertiary center. CASE 1: An 88-year-old

man underwent PEG due to head and neck cancer. The procedure was uneventful, and the patient remained asymptomatic. After the first PEG tube substitution performed at 6 months, stool drainage through the stoma was observed. Computed tomography (CT) showed a GCCF. After tube removal, the fistula spontaneously closed, and the patient remained under nasogastric feeding until death. CASE 2: A 31-year-old man with hereditary spastic paraplegia was submitted to PEG without early complications. The patient remained asymptomatic, and 7 months later, replacement of the PEG tube was planned. Under endoscopic control, the primary tube was removed, but the balloon replacement tube, introduced through the skin, was not observed in the gastric lumen. CT displayed a GCCF that spontaneously closed after a few days. A combined laparoscopic and endoscopic approach was used to resect the fistula tracts and perform a new gastrostomy. CASE 3: A 45-year-old man with cerebral palsy was referred to PEG. Skin transillumination was only observed transiently, and the abdominal puncture was performed obliquely. The patient remained asymptomatic until the 7th month, when the primary PEG tube replacement was performed. The percutaneously placed substitution tube did not reach the stomach. GCCF was evident on CT. The fistula spontaneously closed, and the patient was referred to elective surgery for laparoscopic gastrostomy. GCCF is an uncommon complication of PEG. Its clinical course seems to be benign with patients remaining asymptomatic under ambulatory enteral feeding for long periods until PEG tube replacement. Spontaneous fistula closure is the rule in this setting. Laparoscopic gastrostomy should be considered when a new PEG is advised and cannot be safely performed due to colon interposition.

PMID: 31832501

15. Malnutrition is common in children with cerebral palsy in Saudi Arabia - a cross-sectional clinical observational study.
Almuneef AR, Almajwal A, Alam I, Abulmeaty M, Bader BA, Badr MF, Almuammar M, Razak S.

BMC Neurol. 2019 Dec 10;19(1):317. doi: 10.1186/s12883-019-1553-6.

BACKGROUND: Cerebral palsy (CP) is considered as the main cause of severe physical impairment and malnutrition in children. This cross-sectional study intended to survey the nutritional status of children cerebral palsy in Riyadh, Saudi Arabia. METHODS: We examined 74 children (age: 1-10 yrs) with CP, who attended Sultan Bin Abdulaziz Humanitarian City (SBAHC), Riyadh Saudi Arabia. Data on age, general demographics, nutritional status, and dietary intake were collected. A child was considered underweight, wasted, stunted or thin if the standard deviation scores for his/her weight for age, weight for height, height for age and body mass index for age were ≤ -2.0 standard deviation (SD) using WHO growth standards. Multivariable logistic regression identified the factors associated with nutritional indicators. RESULTS: More than half (56.4%) of the children with cerebral palsy were malnourished as they had z-score below <-2 SD in at least one of the four indicators. Thinness (50%) was the most common form of malnutrition, followed by underweight, stunting, and wasting. Arm anthropometrics gave similar results on the percent number of malnourished children. Factors that were independently associated with malnutrition with an adjusted OR (aOR) were as follow: age ≤ 5 yrs. (aOR: 4.29); presence of cognitive impairment (aOR: 4.13); presence of anemia (aOR: 3.41) and inadequate energy intake (aOR: 4.86) (p, for all trends <0.05). CONCLUSION: Children with cerebral palsy of the current study have impaired growth and nutritional status as assessed by all four common nutritional status indicators. Further large-scale community-based studies for in-depth evaluation of nutritional status and growth patterns in children with CP are needed.

PMID: 31823743

16. Factors associated with the stool characteristics of children with cerebral palsy and chronic constipation.
García Contreras AA, Vásquez Garibay EM, Sánchez Ramírez CA, Fafutis Morris M, Delgado Rizo V.

Rev Esp Enferm Dig. 2019 Dec 13;112. doi: 10.17235/reed.2019.6313/2019. [Epub ahead of print]

BACKGROUND: chronic constipation is a common gastrointestinal problem in children with cerebral palsy and several factors can influence the stool frequency, consistency and pH in these cases. AIM: to identify the association of dietary factors, use of anticonvulsants and family history of constipation with the stool characteristics of children with cerebral palsy and chronic constipation. METHODS: an analytical cross-sectional study was performed of 45 children with cerebral palsy and chronic constipation that included 19 females and 26 males, aged 37 ± 13 months. Dietary factors, the use of anticonvulsants and family history were analyzed. Stool frequency, consistency (Bristol Stool Form Scale) and pH (using a pH-meter) were also determined. RESULTS: there was a positive correlation between stool frequency and the consumption of oilseeds ($r = 0.339$, $p = 0.023$). There was a negative correlation between hard stools and fluid intake ($r = -0.336$, $p = 0.042$) and between

stool pH and the consumption of cereals rich in insoluble fiber, high soluble fiber vegetables, carrots and potatoes (r = -0.339, p = 0.030; r = -0.308, p = 0.044; r = -0.336, p = 0.027; r = -0.307, p = 0.045, respectively). An association was also identified between the use of anticonvulsant polytherapy and hard stools (OR = 14.2 [95% CI 1.16-174], p = 0.038). There was no association between family history and constipation. CONCLUSIONS: rich-fiber food consumption, fluids intake and anticonvulsant polytherapy were associated with the stool characteristics of children with cerebral palsy and chronic constipation.

PMID: 31830793

17. Development of a Data Logger for Capturing Human-Machine Interaction in Wheelchair Head-Foot Steering Sensor System in Dyskinetic Cerebral Palsy.
Gakopoulos S, Nica IG, Bekteshi S, Aerts JM, Monbaliu E, Hallez H.

Sensors (Basel). 2019 Dec 7;19(24). pii: E5404. doi: 10.3390/s19245404.

The use of data logging systems for capturing wheelchair and user behavior has increased rapidly over the past few years. Wheelchairs ensure more independent mobility and better quality of life for people with motor disabilities. Especially, for people with complex movement disorders, such as dyskinetic cerebral palsy (DCP) who lack the ability to walk or to handle objects, wheelchairs offer a means of integration into daily life. The mobility of DCP patients is based on a head-foot wheelchair steering system. In this work, a data logging system is proposed to capture data from human-wheelchair interaction for the head-foot steering system. Additionally, the data logger provides an interface to multiple Inertial Measurement Units (IMUs) placed on the body of the wheelchair user. The system provides accurate and real-time information from head-foot navigation system pressure sensors on the wheelchair during driving. This system was used as a tool to obtain further insights into wheelchair control and steering behavior of people diagnosed with DCP in comparison with a healthy subject.

PMID: 31817941

18. Developing a province-wide hip surveillance program for children with cerebral palsy: from evidence to consensus to program implementation: a mini-review.
Miller SD, Mayson TA, Mulpuri K, O'Donnell ME.

J Pediatr Orthop B. 2019 Dec 9. doi: 10.1097/BPB.0000000000000707. [Epub ahead of print]

Hip displacement is a common orthopedic problem in children with cerebral palsy (CP) that can result in significant morbidity. Hip surveillance has been shown to reduce the incidence of hip dislocations in children with CP and to reduce the need for salvage hip surgeries. Guidelines for hip surveillance have been developed and can be adapted to meet local needs. Implementation of surveillance guidelines for a population of children is complex and highly dependent upon the region, province/state, or country's system of care for children with CP. Recognizing that implementation of the evidence on hip surveillance was necessary in British Columbia, a Canadian province spanning 1 million square kilometers, a comprehensive, coordinated approach to hip surveillance was developed collaboratively by provincial stakeholders. Surveillance guidelines and a desired implementation plan were established based on the best available research evidence, current international practice, and service delivery in British Columbia. Staged implementation preceded full provincial roll out. Implementation was supported by detailed communication, knowledge translation, and evaluation plans. This province-wide hip surveillance program is the first of its kind in North America.

PMID: 31821270

19. Neurodevelopmental outcome of preterm twins at 5 years of age.
Ylijoki M, Haataja L, Lind A, Ekholm E, Lehtonen L; PIPARI study group.

Pediatr Res. 2019 Dec 12. doi: 10.1038/s41390-019-0688-x. [Epub ahead of print]

BACKGROUND: Twins are considered to be at an increased risk for perinatal mortality and morbidities, but it is unclear whether preterm twins are at an increased risk for poor developmental outcomes when compared to preterm singletons. Our aim was to compare the neurodevelopmental outcome of preterm twins vs singletons at 5 years of age. METHODS: Very low birth weight and very low gestational age infants (twins n = 66, singletons n = 157) were recruited as a part of the PIPARI project in the Turku University Hospital, covering a regional population. Cognitive development, neuropsychological performance, and neurodevelopmental impairments (including cerebral palsy, hearing deficit, visual impairment, and intellectual disability) were evaluated at 5 years of age. RESULTS: Twins and singletons had otherwise similar perinatal background factors, except for the higher proportion of preterm rupture of membranes in singletons. Twins had cognitive and neuropsychological outcomes that were otherwise comparable with singletons, but they had a slightly lower verbal intelligence quotient (estimate -5.81, 95% CI -11.14 to -0.48, p = 0.03). Being a twin was not a risk for neurodevelopmental impairments. CONCLUSIONS: Our study shows that, contrary to a common hypothesis, the overall neurodevelopment of very preterm twins does not significantly differ from that of preterm singletons.

PMID: 31830757

20. Management of Epilepsies at the Community Cottage Hospital Level in a Developing Environment.
Ekanem EE, Fajola AO, Usman R, Ogbimi RN, Ikeagwu GO, Anidima TE, Etieh MN, Umejiego CN.

Niger Med J. 2019 Jul-Aug;60(4):186-189. doi: 10.4103/nmj.NMJ_6_18. Epub 2019 Nov 25.

BACKGROUND: The epilepsy problem in much of Africa is characterized by stigmatization and neglect. This article describes the efforts at a cottage hospital level to ameliorate the epilepsy problem in a resource-limited environment. METHODS: A seizure clinic was started in a cottage hospital after targeted health talks. The International League against Epilepsy (ILEA)/World Health Organization (WHO)/International Bureau for Epilepsy (IBE) manual was adopted for the training of staff and to guide management. Patients were followed up in the clinic and with the use of simple information communication technology. RESULTS: Forty-five patients with ages ranging from 3 months to 42 years (who had lived with epilepsy for periods ranging from 3 weeks to 32 years) were registered over 12 months period. The most common seizure type was generalized tonic clonic (21 or 46.67%) followed by generalized clonic (8 or 17.78%). Ten (22.22%) had comorbidities mainly cerebral palsy (4 or 8.89%) and attention-deficit hyperactivity disorder (3 or 6.67%). Most (98.15%) were placed on carbamazepine. Twenty-three (51.11%) had complete control of seizures, 21 (46.67%) had reduced frequencies of attacks, and all 8 children who had dropped out of school resumed schooling. CONCLUSION: The epilepsy challenge in the developing world can be demystified and effectively managed at the cottage hospital level. Targeted health education, affordable management regimes, and committed follow-up are keys. A training manual based on the ILEA/WHO/IBE document should be developed for Africa.

PMID: 31831937

21. [Protocol for the etiological investigation of cerebral palsy].
Lourenço L, Campos T, Rodrigues E, Sousa R, Guardiano M, Leão M.

Rev Neurol. 2019 Dec 16;69(12):512-513. doi: 10.33588/rn.6912.2019300.

PMID: 31820821

22. [Precise diagnosis and treatment of spastic cerebral palsy].
He X, Xu Y, Yang X.

Zhongguo Xiu Fu Chong Jian Wai Ke Za Zhi. 2019 Dec 15;33(12):1584-1588. doi: 10.7507/1002-1892.201903072.

OBJECTIVE: To summarize the advancement of precise diagnosis and treatment for spastic cerebral palsy in recent years. METHODS: The literature and own experiences were reviewed, and the surgical method, precise diagnosis, and personalized treatment of spastic cerebral palsy based on the classification of spastic cerebral palsy were summarized and analyzed. RESULTS: The common classification of spastic cerebral palsy are gross motor function classification system (GMFCS) and manual ability classification system (MACS). The surgical methods of spastic cerebral palsy can be divided into soft tissue surgery, nerve surgery, and bone and joint surgery. The precise diagnosis of spastic cerebral palsy includes qualitative diagnosis, localization diagnosis, and quantitative diagnosis. Based on precise diagnosis and classification, one or more corresponding surgical methods are selected for treatment. CONCLUSION: The manifestations of spastic cerebral palsy are so diverse that it is necessary to select rational surgeries based on precise diagnosis to achieve individualized treatment.

PMID: 31823563

23. Perinatal Stroke.
Dunbar M, Kirton A.

Semin Pediatr Neurol. 2019 Dec;32:100767. doi: 10.1016/j.spen.2019.08.003. Epub 2019 Aug 7.

Perinatal strokes are a diverse but specific group of focal cerebrovascular injuries that occur early in brain development and affect an estimated 5 million people worldwide. The objective of this review is to describe the epidemiology, clinical presentations, pathophysiology, outcomes, and management for the 6 subtypes of perinatal stroke. Some perinatal strokes are symptomatic in the first days of life, typically with seizures, including neonatal arterial ischemic stroke, neonatal hemorrhagic stroke, and cerebral sinovenous thrombosis. The remaining subtypes present in the first year of life or later, usually with motor asymmetry and include arterial presumed perinatal ischemic stroke, presumed perinatal hemorrhagic stroke, and in utero periventricular venous infarction. The consequences of these injuries include cerebral palsy, epilepsy, and cognitive and behavioral challenges, in addition to the psychosocial impact on families. While there have been significant advances in understanding mechanisms of both injury and recovery, there is still a great deal to learn regarding causation and the optimization of outcomes.

PMID: 31813521

24. Daily activities, participation, satisfaction, and functional mobility of adults with cerebral palsy more than 25 years after selective dorsal rhizotomy: a long-term follow-up during adulthood.
Veerbeek BE, Lamberts RP, Fieggen AG, Verkoeijen PPJL, Langerak NG.

Disabil Rehabil. 2019 Dec 9:1-9. doi: 10.1080/09638288.2019.1695001. [Epub ahead of print]

Purpose: To determine changes in level of accomplishment and satisfaction in daily activities and social participation, and functional mobility in adults with cerebral palsy (CP) more than 25 years after selective dorsal rhizotomy (SDR).Materials and methods: This long-term observational nine-year follow-up study included 26 adults (median age 35 years) with CP and spastic diplegia, and 26 matched typically developing adults. Assessment tools used were the Life-Habits questionnaire and the Functional Mobility Scale. Results: Most of the adults with CP were independent and satisfied with accomplishing life habits and no changes were determined, except for a small change in the Housing accomplishment level. Compared to typically developing adults, the CP cohort was more dependent in accomplishing Mobility and Recreation. However, the level of satisfaction was similar for most life habits except for Mobility. Functional mobility did not change, but correlated with Life-Habits results. Conclusions: Adults with CP showed high and stable levels of accomplishment and satisfaction in daily activities and social participation more than 25 years after SDR. This is in contrast with the literature, where functional decline was shown for individuals with CP as they age. The relation with functional mobility highlights the importance to focus the rehabilitation on maintaining walking ability in order to enable high level of daily activities and social participation in adults with CP. Implications for rehabilitation: Selective dorsal rhizotomy (SDR) is a valuable treatment option for a selective group of children with cerebral palsy (CP) in order to reduce spasticity. The long-term outcomes of SDR on level of accomplishment and satisfaction in daily activities and social participation as well functional mobility in adults with CP are not clear. More than 25 years after SDR adults with CP experienced stable and lasting high levels of functioning regarding daily activities and social participation and were satisfied with the way they accomplished life habits. Functional mobility was correlated to level of accomplishment and satisfaction in daily activities and social participation, which highlights the importance to focus rehabilitation programs on maintaining functional mobility in order to enable daily activities and social participation in adults

with CP.

PMID: 31815556

25. How pain management for children with cerebral palsy in South African schools complies with up-to-date knowledge.
Johnson E, Nilsson S, Adolfsson M.

Afr J Disabil. 2019 Nov 22;8(0):575. doi: 10.4102/ajod.v8i0.575. eCollection 2019.

BACKGROUND: Pain in children with cerebral palsy (CP) has its sources in musculoskeletal problems that can influence learning in a school setting. Best pain management is essential for these children, but school staff may not keep up to date with the latest developments and interventions. Therefore, staff's perceptions of beneficial strategies may not comply with contemporary scientific knowledge about effective evidence-based interventions. OBJECTIVES: This study investigated how pain management intervention for children with CP in South African schools complied with international scientific knowledge about evidence-based interventions. The intention was to provide support for an update of knowledge on both individual level (i.e. professionals) and system level (i.e. decision makers). METHOD: Five focus groups were conducted with staff members at five schools for children with special educational needs in South Africa. Manifest and latent content analyses of professional statements identified interventions reported as beneficial and related them to higher and lower levels of intervention evidence as reported at the time of data collection. RESULTS: Most treatment strategies concerned motor functioning that fell within the framework of physiotherapists and occupational therapists. Access to orthopaedic expertise was limited, waiting times were long and medication for spasticity treatment was not offered. CONCLUSION: A discrepancy between published evidence and clinical practice for pain management in children with CP in South African school settings was noted. Suggestions for improved early intervention to identify children's hips at risk through surveillance programmes; and orthopaedic management are proposed to prevent deformities and unnecessary suffering in South African children with CP.

PMID: 31824834

26. Longitudinal Change in Common Impairments in Children with Cerebral Palsy From Age 1.5 to 11 Years.
Jeffries LM, Fiss AL, Westcott McCoy S, Avery L.

Pediatr Phys Ther. 2019 Nov 27. doi: 10.1097/PEP.0000000000000663. [Epub ahead of print]

PURPOSE: This project aimed to determine whether change occurs over time for impairments of balance, range of motion, endurance, and strength of children with cerebral palsy, by Gross Motor Function Classification System (GMFCS) levels. METHODS: Measurements were completed in 77 children at 2 sessions (T1, T2) on average 5.8 years apart. Mean ages were 2.9 years (SD = 0.9) and 8.7 years (SD = 1.1) at T1 and T2, respectively. RESULTS: There were significant differences from T1 to T2 for some children (GMFCS levels I, II, and III/IV: balance increased; GMFCS levels I and II: strength increased; and GMFCS levels III/IV and V: range of motion decreased). Endurance scores were not different and did not change. CONCLUSIONS: Longitudinal changes in most impairments occurred in children with cerebral palsy. Monitoring and targeted interventions should support each child's development.

PMID: 31815922

27. Reducing Intrathecal Baclofen Related Infections: Service Evaluation and Best Practice Guidelines.
Balaratnam MS, Donnelly A, Padilla H, Simeoni S, Bahadur S, Keenan L, Lee H, Farrell R, Curtis C, Brownstone RM, Murphy M, Grieve J, Shieff C, Nayar M, Pitceathly RDS, Christofi G, Stevenson VL.

Neuromodulation. 2019 Dec 12. doi: 10.1111/ner.13071. [Epub ahead of print]

OBJECTIVES: Intrathecal baclofen (ITB) pumps are an effective treatment for spasticity; however infection rates have been reported in 3-26% of patients in the literature. The multidisciplinary ITB service has been established at The National Hospital for Neurology and Neurosurgery, UCLH, Queen Square, London for over 20 years. Our study was designed to clarify the rate of infection in our ITB patient cohort and secondly, to formulate and implement best practice guidelines and to determine prospectively, whether they effectively reduced infection rates. METHODS: Clinical record review of all patients receiving ITB pre-intervention; January 2013-May 2015, and following practice changes; June 2016-June 2018. RESULTS: Four of 118 patients receiving ITB during the first time period (3.4%, annual incidence rate of infection 1.4%) developed an ITB-related infection (three following ITB pump replacement surgery, one after initial implant). Infections were associated with 4.2% of ITB-related surgical procedures. Three of four pumps required explantation. Following change in practice (pre-operative chlorhexidine skin wash and intraoperative vancomycin wash of the fibrous pocket of the replacement site), only one of 160 ITB patients developed infection (pump not explanted) in the second time period (0.6%, annual incidence rate 0.3%). The infection rate related to ITB surgical procedures was 1.1%. In cases of ITB pump replacement, the infection rate was reduced to 3.3% from 17.6%. CONCLUSIONS: This study suggests that a straightforward change in clinical practice may lower infection rates in patients undergoing ITB therapy.

PMID: 31828902

28. Effect of Intravenous Aminocaproid Acid on Blood Loss and Transfusion Requirements After Bilateral Varus Rotational Osteotomy: A Double-blind, Placebo-controlled Randomized Trial.
Swarup I, Nguyen J, Edmonds C, Dodwell E, Scher D.

J Pediatr Orthop. 2019 Dec 10. doi: 10.1097/BPO.0000000000001480. [Epub ahead of print]
BACKGROUND: ε-Aminocaproic acid (EACA) is an antifibrinolytic agent that has been shown to decrease blood loss and transfusion requirements in several populations undergoing various surgical procedures. However, the efficacy of EACA has not been assessed in pediatric patients with cerebral palsy undergoing bilateral varus rotational femoral osteotomies. The purpose of this study was to assess the efficacy of intravenous EACA in reducing calculated intraoperative blood loss and transfusions in this population. METHODS: Patients aged 18 years or younger were eligible. Patients were randomized to receive EACA or placebo (saline), and randomization was stratified based on sex and whether or not additional soft tissue or osseous procedures were performed. On the basis of retrospective data, the calculated sample size was 12 patients per arm to detect a difference of 250-mL blood loss. The primary outcome was calculated intraoperative blood loss. Secondary outcomes included transfusion requirements, 24-hour drain output, length of stay, and incidence of complications. RESULTS: The mean age of patients in this study was 8 years (SD: 2.4 y). There were no differences in age, sex, height, weight, type of anesthesia, operative time, and associated procedures between the EACA and placebo groups (P>0.05). Preoperative hematocrit was lower in the EACA group (37.1 vs. 40.0, P=0.04). Calculated intraoperative blood loss was 536 mL in the EACA group and 628 mL in the placebo group (P=0.45). Transfusions were required in 62% of patients in the EACA group and 67% of patients in the placebo group (P=0.68). Total 24-hour drain output was 72.5 mL in the EACA group and 103.3 mL in the placebo group (P=0.37). Length of stay was similar between both groups, and there were no drug or placebo-related complications in either group. CONCLUSIONS: There was no difference in blood loss or transfusion requirements associated with EACA compared with placebo; however, this study is underpowered to detect smaller differences in blood loss. Additional studies with larger sample sizes are needed to confirm these findings and further elucidate the indications for antifibrinolytic agents in pediatric patients. LEVEL OF EVIDENCE: Level I.

PMID: 31834239

Prevention and Cure

29. Neuroprotection Strategies in Preterm Encephalopathy.

Parikh P, Juul SE.

Semin Pediatr Neurol. 2019 Dec;32:100772. doi: 10.1016/j.spen.2019.08.008. Epub 2019 Aug 9.

Advances in neonatology have led to unprecedented improvements in neonatal survival such that those born as early as 22 weeks of gestation now have some chance of survival, and over 70% of those born at 24 weeks of gestation survive. Up to 50% of infants born extremely preterm develop poor outcomes involving long-term neurodevelopmental impairments affecting

cognition and learning, or motor problems such as cerebral palsy. Poor outcomes arise because the preterm brain is vulnerable both to direct injury (by events such as intracerebral hemorrhage, infection, and/or hypoxia), or indirect injury due to disruption of normal development. This neonatal brain injury and/or dysmaturation is called "encephalopathy of prematurity". Current and future strategies to improve outcomes in this population include prevention of preterm birth, and pre-, peri-, and postnatal approaches to protect the developing brain. This review will describe mechanisms of preterm brain injury, and current and upcoming therapies in the antepartum and postnatal period to improve preterm encephalopathy.

PMID: 31813513