Cerebral Palsy ALLIANCE RESEARCH INSTITUTE

CP Research News

Monday 25 April 2011

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Interventions

1. Arch Phys Med Rehabil. 2011 Apr 18. [Epub ahead of print]

Validation of the Physical Activity Scale for Individuals With Physical Disabilities.

van den Berg-Emons RJ, L'ortye AA, Buffart LM, Nieuwenhuijsen C, Nooijen CF, Bergen MP, Stam HJ, Bussmann JB.

Department of Rehabilitation Medicine and Physical Therapy, Erasmus Medical Center, Rotterdam; Rijndam Rehabilitation Center, Rotterdam, The Netherlands.

OBJECTIVE: To determine the criterion validity of the Physical Activity Scale for Individuals With Physical Disabilities (PASIPD) by means of daily physical activity levels measured by using a validated accelerometry-based activity monitor in a large group of persons with a physical disability. DESIGN: Cross-sectional. SETTING: Participants' home environment. PARTICIPANTS: Ambulatory and nonambulatory persons with cerebral palsy, meningomyelocele, or spinal cord injury (N=124). INTERVENTIONS: Not applicable. MAIN OUTCOME MEASURES: Self-reported physical activity level measured by using the PASIPD, a 2-day recall questionnaire, was correlated to objectively measured physical activity level measured by using a validated accelerometry-based activity monitor. RESULTS: Significant Spearman correlation coefficients between the PASIPD and activity monitor outcome measures ranged from .22 to .37. The PASIPD overestimated the duration of physical activity measured by using the activity monitor (mean ± SD, 3.9±2.9 vs 1.5±0.9h/d; P<.01). Significant correlation (ρ=-.74; P<.01) was found between average number of hours of physical activity per day measured by using the 2 methods and difference in hours between methods. This indicates larger overestimation for persons with higher activity levels. CONCLUSIONS: The PASIPD correlated poorly with objective measurements using an accelerometry-based activity monitor in people with a physical disability. However, similar low correlations between objective and subjective activity measurements have been found in the general population. Users of the PASIPD should be cautious about overestimating physical activity levels.

PMID:21507382 [PubMed - as supplied by publisher]

2. Dev Med Child Neurol. 2011 Apr 18. doi: 10.1111/j.1469-8749.2011.03970.x. [Epub ahead of print]

Correct positioning for hip radiographs allows reliable measurement of hip displacement in cerebral palsy.

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Paediatric Radiology Department, Derbyshire Children's Hospital, Royal Derby Hospital, Derby. Ronnie Mackeith Child Development Centre, Derbyshire Children's Hospital, Royal Derby Hospital, Derby, UK.

Aim: The pelvic radiograph in children with cerebral palsy (CP) can inform the degree of hip displacement by calculation of the migration percentage. However, concerns have arisen about the reliability of this measurement. The present study examined the reliability of radiographic assessment of displacement and the importance of positioning and reporting experience. Method: Two pelvic radiographs, taken at least an hour apart, were performed in 20 children (total 40 hips) in the standard position by a trained paediatric radiographer. Children (13 males, seven females) were aged 30 months to 10 years with severe bilateral spastic CP in Gross Motor Function Classification System levels IV (n=10) and V (n=10). The migration percentage of each hip was measured on two occasions 3 months apart by two experienced radiologists independently. Comparisons of migration percentage were made in three ways by (1) the same observer at the same time, (2) the same observer 3 months apart, and (3) different observers 3 months apart. Results: Migration percentage (mean [SD]) was (1) 3.2% (3.5), (2) 3.3% (3.2), and (3) 3.7% (3.8) respectively. Interpretation: Reliable measures of migration percentage can be obtained with correct positioning and if reported by suitably experienced radiologists, making this a valid surveillance method. Clinical decisions can be made taking into account an expected error in hip displacement measurements.

PMID:21501157 [PubMed - as supplied by publisher]

3. Neuropediatrics. 2011 Apr 15. [Epub ahead of print]

Hip Lateralisation in Children with Bilateral Spastic Cerebral Palsy Treated with Botulinum Toxin Type A: A 2-Year Follow-Up.

Jung NH, Heinen F, Westhoff B, Doederlein L, Reissig A, Berweck S, Linder-Lucht M, Schandelmaier S, Mall V On On Behalf Of The German Abo Study Group For Author List See Appendix.

Division of Neuropediatrics and Muscular Disorders, Department of Pediatrics and Adolescent Medicine, University Hospital Freiburg, Freiburg, Germany.

We investigated the effect of BoNT/A injection on hip lateralisation in children with bilateral spastic cerebral palsy and bilateral adductor spasticity. Pelvic radiographs using Reimers' migration index (MI) were evaluated from 27 children (n=9 females, n=18 males; mean age 5.2±1.96 years; range: 2-10 years; initial MI <50%) with bilateral spastic cerebral palsy over a time period of 2 years. All received injections of BoNT/A (Dysport (®)) every 12 weeks with a dose of 30 Units per kilogram body weight into adductor and medial hamstring muscles on both sides. The MI was calculated before treatment and after 1 and 2 years. The mean MI increased from 25.5% (range: 0-48) to 26.7% (+1.2%, range: 0-79) on the right side and from 28.0% (range: 0-40) to 30.6% (+2.6%, range: 3-84) on the left side over 2 years, respectively. Hips of one patient dislocated bilaterally. The mean MI remained stable over 2 years. Although a specific BoNT/A effect cannot be proven because of the open design of this study, we provide strong evidence that the MI can be kept stable for a time period of 2 years under non-surgical management including therapy with BoNT/A even in CP patients with a high risk for hip dislocation.

PMID:21500143 [PubMed - as supplied by publisher]

4. Childs Nerv Syst. 2011 Apr 19. [Epub ahead of print]

Histological evidence of intraoperative monitoring efficacy in selective dorsal rhizotomy.

Fukuhara T, Nakatsu D, Namba Y, Yamadori I.

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PURPOSE: It has been advocated that intraoperative electrophysiological monitoring is mandatory in selective dor-

sal rhizotomy (SDR). However, it is still uncertain whether the monitoring procedure effectively differentiates dysfunctional rootlets. We histologically analyzed sectioned rootlets in SDR, in order to confirm the efficacy of the monitoring. METHODS: Seven children with cerebral palsy underwent SDR on the same protocol. The pieces of their sectioned nerve rootlets from L5 were examined histologically using an electron microscope. In each patient, two nerve rootlets, one with the most abnormal response to intraoperative electrical stimulation and the other with the least abnormal response, were examined. The electron microscope findings of the rootlets were compared with the electromyography (EMG) findings in the intraoperative stimulation. RESULTS: Among 14 examined nerve rootlets, definite abnormal EMG findings were seen in 5, which were 4 clonuses and one bilateral spread. All five rootlets with abnormal EMG findings showed axonal degenerations except one case, whose finding was dysmyelination. On the contrary, in the nine rootlets with normal EMG findings, eight rootlets had histologically minimum changes limited to the myelin sheath and one rootlet had dysmyelination without axonal degeneration. CONCLUSIONS: The nerve rootlets with abnormal EMG findings in the intraoperative stimulation have definite histological abnormalities. This indicates that intraoperative monitoring is a meaningful method for identifying the nerve rootlets to be sectioned. This finding should be reminded as a precaution when considering nonselective dorsal rhizotomy, especially, for children who are expected to stand up after the surgery.

PMID:21503753 [PubMed - as supplied by publisher]

5. Dev Med Child Neurol. 2011 Apr 20. doi: 10.1111/j.1469-8749.2011.03988.x. [Epub ahead of print]

A review of energy intake measures used in young children with cerebral palsy.

Walker JL, Bell KL, Caristo FM, Boyd RN, Davies PS.

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The aim of this review was to evaluate the psychometric properties and clinical utility of energy intake measures used in young children with cerebral palsy (CP). Five databases were searched for relevant literature, and measures were included if they (1) directly measured energy intake in kilojoules/kilocalories per day; (2) had published data in kilojoules/kilocalories per day for children with CP from birth to 5 years; and (3) at least 40% of participants had a diagnosis of CP. Three measures met criteria: a 3-day weighed food record, a 3-day estimated food record, and a 7-day estimated food record. Included measures were evaluated on their characteristics, intended outcome, and validity. Reliability and responsiveness were not reported for any measure. Currently there is no dietary methodology that has proven reliability or repeated validity in young children with CP. Clinicians and researchers should not rely on current methodologies until further evaluation.

PMID:21506998 [PubMed - as supplied by publisher]

6. Eur J Phys Rehabil Med. 2011 Apr 20. [Epub ahead of print]

Equinus foot classification in cerebral palsy: an agreement study between clinical and gait analysis assessment.

Benedetti MG, D'apote G, Faccioli S, Costi S, Ferrari A.

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BACKGROUND: Excessive ankle plantar flexion (equinus foot) is a common problem in cerebral palsy (CP) and several treatment options can be considered depending on the equinus type. Few attempts have been made to classify different forms of equinus foot for specific treatment. AIM: This study is aimed at defining equinus foot types in CP patients according to the Ferrari classification, integrating clinical and instrumental assessments. The hypothesis is that clinical differentiation of equinus foot can be evidenced by recurrent anomalies identifiable through

gait analysis (GA), which can make the assessment, usually based only on clinician semeiotics, more objective. DESIGN: Clinical and instrumental assessments were performed separately by a senior CP physiatrist and a senior GA physiatrist, the latter was blind to the clinical diagnosis of equinus type. SETTING: Outpatients. POPULATION: Twenty patients, 16 diplegics and 4 hemiplegics (mean age 11 years, SD 4 years 11 months). METHODS: Clinical assessment by means of Modified Ashworth Scale, Gross Motor Function Measure (GMFM), Observational Gait Analysis (OGA), and measurement of lower limb muscle strength by dynamometer were used to classify the equinus type. Gait analysis assessed the kinematics and EMG of affected lower limbs. RESULTS: Ten different equinus types were identified. Since various forms of equinus can be present in the same patient, we were able to classify a total of 61 types of equinus in 36 feet. Substantial agreement was found between Clinical and Gait Analysis equinus assignment matched in 50 out of 61 types (Index of agreement with Fleiss' Kappa 79.3 %). In some case only Gait Analysis was able to identify the equinus type, while in others it did not confirm clinical assignment. CONCLUSION: Gait analysis is able to distinguish different equinus types according to Ferrari classification, making the clinical decision less arbitrary. Clinical rehabilitation impact. Correct objective diagnosis of equinus foot in CP patients is of paramount importance when choosing suitable rehabilitative interventions.

PMID:21508920[PubMed - as supplied by publisher]

7. Eur J Phys Rehabil Med. 2011 Apr 20. [Epub ahead of print]

Cycling induced by functional electrical stimulation in children affected by cerebral palsy: case report.

Trevisi E, Gualdi S, De Conti C, Salghetti A, Martinuzzi A, Pedrocchi A, Ferrante S.

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BACKGROUND: Recently, the efficacy of functional electrical stimulation (FES) cycling have been demonstrated on the improvement of strength and motor control in adults with stroke. FES-cycling, providing a repetitive goal-oriented task, could facilitate cortical reorganization and utilization of residual cortico-spinal pathways. These benefits could be more enhanced in children because of the greater plasticity and flexibility of their central nervous system. AIM: The aim of the present case report study was to explore the feasibility of FES-cycling in children with cerebral palsy (CP) and to provide a set of instrumental measures able to evaluate the effects of this novel treatment on cycling and walking ability. DESIGN: Interventional study. SETTING AND POPULATION: Two ambulant outpatient children with diplegic CP were recruited by the "E. Medea" Scientific Institute. METHODS: Patients followed a FES-cycling treatment for 30 minutes a day, 3 days a week for 7 weeks. Pre and post treatment tests were performed, namely clinical measures and electromyographic, kinematic and oxygen expenditure analysis during gait and cycling. RESULTS: The treatment was safe, feasible and well accepted by the 2 children. After treatment both patients achieved a more symmetrical muscular strategy during voluntary cycling and gait and a significant reduction of muscle co-contractions during cycling. These improvements were corroborated by a decrease in oxygen expenditure during the post test for one of the two children, the less impaired, implying a better exploiting of bi-articular muscles. CONCLUSION AND CLINICAL REHABILITATION IMPACT: FES-cycling is feasible and safe and it may be an alternative rehabilitation method for diplegic CP patients. The set of instrumental measurements proposed seems to be a valuable tool for functional assessment to identify subclinical anomalies and improvements on cycling and gait in CP patients.

PMID:21508913 [PubMed - as supplied by publisher]

8. Dev Med Child Neurol. 2011 Apr 20. doi: 10.1111/j.1469-8749.2011.03913.x. [Epub ahead of print]

Medial gastrocnemius muscle volume and fascicle length in children aged 2 to 5 years with cerebral palsy.

Barber L, Hastings-Ison T, Baker R, Barrett R, Lichtwark G.

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Aim: The aim of this article was to compare medial gastrocnemius muscle volume, physiological cross-sectional

area (PCSA), muscle length, fascicle length, and pennation angle in children aged 2 to 5 years with spastic cerebral palsy (CP) and in typically developing children. Method: Fifteen children with spastic CP (11 males, four females; mean age 45mo [SD 15mo]; five with hemiplega; 10 with diplega; 10 classified at Gross Motor Function Classification System (GMFCS) level I, five at GMFCS level II) and 20 typically developing children (11 males, nine females; mean age 48mo [SD 14mo]) participated in the study. Individuals with spastic CP were included if they had a minimum range of motion of 0° ankle dorsiflexion with the knee extended and were excluded if they had had previous botulinum toxin treatment to the calf muscles or previous calf surgery. Typically developing children were included if they were able to walk independently and were excluded if there was a history of previous lower leg injury or other developmental disorder affecting the lower limb. Freehand two-dimensional and three-dimensional ultrasound was used to assess muscle properties of the relaxed medial gastrocnemius muscle at three ankle joint angles: maximum dorsiflexion, neutral and maximum plantarflexion. PCSA was calculated as a function of muscle volume and muscle fascicle length and pennation angle was recorded at the neutral ankle joint angle. Results: Medial gastrocnemius muscle volume was 22% lower in the group with spastic CP than in the typically developing group, which in the absence of significant group differences in neutral fascicle length gave rise to an equivalent reduction in PCSA for the group with spastic CP. Significant positive correlations were found between muscle volume and age (r=0.63-0.65) and between muscle length and age (r=0.72-0.81) in both groups. Maximum ankle dorsiflexion angle was also reduced in the group with spastic CP (8°) compared with the typically developing group (26°). Interpretation: The observed reduction in muscle PCSA in the group with spastic CP would be expected to contribute to the clinically observed muscle weakness in spastic CP and suggests the need for early intervention in order to minimize loss of muscle PCSA in spastic CP.

PMID:21506995 [PubMed - as supplied by publisher]

9. Eur J Paediatr Neurol. 2011 Apr 19. [Epub ahead of print]

Best age for surgery for Infantile esotropia.

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Infantile esotropia (IE) is defined as an esotropia before the age of 6 months, with a large angle, latent nystagmus, dissociated vertical deviation, limitation of abduction, and reduced binocular vision, without neurological disorder. Prematurity, low birth weight, and low Apgar scores are significant risk factors for IE. US standard age of first surgery is 12-18 months, in Europe 2-3 years. The only study to date with prospectively assigned early- and late-surgery groups and evaluation according to intention-to-treat, was the European Early vs. Late Infantile Strabismus Surgery Study (ELISSS). In that study 13.5% of children operated around 20 months vs. 3.9% (P = 0.001) of those operated around 49 months had gross stereopsis (Titmus Housefly) at age 6. The reoperation rate was 28.7% in children operated early vs. 24.6% in those operated late. Unexpectedly, 8% in the early group vs. 20% in the late group had not been operated at age 6, although all had been eligible for surgery at baseline at 11 SD 3.7 months. In most of these children the angle of strabismus decreased spontaneously. In a meta-regression analysis of the ELISSS and 12 other studies we found that reoperation rates were 60-80% for children first operated around age 1 and 25% for children operated around age 4. Based on these findings, the endpoints to consider when contemplating best age for surgery in an individual child with IE should be: (1) degree of binocular vision restored or retained, (2) postoperative angle and long-term stability of the angle and (3) number of operations needed or chance of spontaneous regression. IE is characterized by lack of binocular connections in the visual cortex that cannot develop, e.g. because the eyes squint, or do not develop, e.g. after perinatal hypoxia. As the cause of IE, whether motor or sensory, is a determinant of surgical outcome, a subdivision of IE according to cause is needed. As similarities exist between IE and cerebral palsy we propose to adapt the working definition formulated by the Surveillance of Cerebral Palsy in Europe and define IE as "a group of permanent, but not unchanging, disorders with strabismus and disability of fusional vergence and binocular vision, due to a nonprogressive interference, lesion, or maldevelopment of the immature brain, the orbit, the eyes, or its muscles, that can be differentiated according to location, extent, and timing of the period of development."

PMID:21511504 [PubMed - as supplied by publisher]

10. Dev Med Child Neurol. 2011 Apr 18. doi: 10.1111/j.1469-8749.2011.03977.x. [Epub ahead of print]

Botulinum toxin type B for sialorrhea in children with cerebral palsy.

Reddihough D, Graham HK.

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PMID:21501158 [PubMed - as supplied by publisher]

11. J Child Neurol. 2011 Apr 15. [Epub ahead of print]

Trihexyphenidyl Improves Motor Function in Children With Dystonic Cerebral Palsy: A Retrospective Analysis.

Ben-Pazi H.

Movement Disorder Clinic, Neuropediatric Unit, Shaare Zedek Medical Center, Jerusalem, Israel.

There are conflicting reports regarding the efficacy of trihexyphenidyl, an anticholinergic drug, for treatment of dystonia in cerebral palsy. The author hypothesized that trihexyphenidyl may be more effective in specific subgroups and performed a retrospective analysis of 31 children (8.2 ± 5.8 years) with dystonia following treatment with high-dose trihexyphenidyl (>0.5 mg/kg/day). Main outcome measure was extent of motor improvement calculated according to the body areas affected. Most (21/31) caregivers reported improvement in 1 or more areas, mainly arm, hand, and oromotor function. Improvement was greater in children without spasticity (P = .02) and in those with higher cognitive function (P = .02). While a third of caregivers (10/31) reported tone reduction, and half (15/31) noted overall functional improvement. Side effects were transient, with the exception of hyperopia (n = 1), and occurred less frequently in children with a history of prematurity (P = .02). In summary, trihexyphenidyl is effective particularly in absence of spasticity and in children with higher cognitive abilities.

PMID:21498790 [PubMed - as supplied by publisher]

12. J Med Life. 2011 Jan-Mar;4(1):36-9. Epub 2011 Feb 25.

Day case laparoscopic nephrectomy: initial experience.

Ilie CP, Luscombe CJ, Smith I, Boddy J, Mischianu D, Golash A.

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RATIONALE: Laparoscopic nephrectomy tends to become the new gold standard surgical technique in a selected population (non-functioning kidney, localised renal cell carcinoma). Day surgery is a popular pathway of care and, procedures of ever-increasing complexity are being considered. OBJECTIVE: The aim of the study was to report the postoperative complications of day case laparoscopic nephrectomy, according to the Clavien system, and, to assess the feasibility of the procedure performed as a day case. MATERIAL AND RESULTS: This study included all the patients considered for day case transperitoneal laparoscopic nephrectomy between May 2008 and November 2009. Sixteen consecutive patients were enrolled in this retrospective study. There were ten procedures on the left hand-side and six on the right hand-side. Age ranges from 22 to 77 years old. Male to female ratio was 9:7. The preoperative diagnosis was non-functioning kidney in 9 cases and kidney tumour in the other 7 cases. All but two patients have been discharged in the same day (87.5%). The readmission rate was of 12.5%. One wheel-chair bonded patient was readmitted four days after the procedure, because of adynamic ileus, and another one three days later because of wound infection. There were two grade I and one grade IV complications (Clavien system). The patient readmitted with grade IV complication, wheel-chair bonded because of cerebral palsy, was not a typical day surgery patient. DISCUSSION: The vast majority of complications were minor and resulted in no residual disability. In our small series, the day case laparoscopic nephrectomy was feasible and safe.

PMID:21505573[PubMed - in process] PMCID: PMC3056421

13. J Pediatr Gastroenterol Nutr. 2011 May;52(5):574-80.

Long-term Follow-up of Patients After Antegrade Continence Enema Procedure.

Siddiqui AA, Fishman SJ, Bauer SB, Nurko S.

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BACKGROUND: : Antegrade continence enema (ACE) has become an important therapeutic modality in the treatment of intractable constipation and fecal incontinence. There are little data available on the long-term performance of the ACE procedure in children. METHODS: : A retrospective review of patients who underwent the ACE procedure was conducted. Irrigation characteristics and complications were noted. Outcome was assessed for individual encounters based on frequency of bowel movements, incontinence, pain, and predictability. RESULTS: : One hundred seventeen patients underwent an ACE. One hundred five patients had at least 6 months of follow-up, and were included in the analysis. Diagnoses included myelodysplasia (39%), functional intractable constipation (26%), anorectal malformations (21%), nonrelaxing internal anal sphincter (7%), cerebral palsy (3%), and other diagnoses (4%). The average follow-up was 68 months (range 7-178 months). At the last follow-up, 69% of patients had successful bowel management. Of the 31% of patients who did not have successful bowel management, 20% were using the ACE despite suboptimal results, 10% required surgical removal, and 2% were not using the ACE because of behavioral opposition to it. Patients were started on normal saline, but were switched to GoLYTELY (PEG-3350 and electrolyte solution) if there was an inadequate response (61% at final encounter). Additives were needed in 34% of patients. The average irrigation dose was 23 ± 0.7 mL/kg. The average toilet sitting time was 51.7 ± 3.5 minutes, with infusions running for 12.1 ± 1.2 minutes. Stomal complications occurred in 63% (infection, leakage, and stenosis) of patients, 33% required surgical revision and 6% eventually required diverting ostomies. CONCLUSIONS: : Long-term use of the ACE gives successful results in 69% of patients, whereas 63% had a stoma-related complication and 33% required surgical revision of the stoma.

PMID:21502828 [PubMed - in process]

14. Dev Med Child Neurol. 2011 Apr 20. doi: 10.1111/j.1469-8749.2011.03987.x. [Epub ahead of print]

Robotic movement therapy in cerebral palsy.

Castelli E.

Bambino Gesù Children's Hospital, Rome, Italy.

PMID:21506997 [PubMed - as supplied by publisher]

15. Disabil Rehabil. 2011 Apr 20. [Epub ahead of print]

Psychometric evaluation of the functional walking test for children with cerebral palsy.

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Purpose. This study examined the psychometric properties of the functional walking test (FWT). Method. Fifty-six subjects with cerebral palsy (CP) (21 females and 35 males, mean age 9 years 6 months, SD 3 years 9months, range 4-17 years) were assessed on two occasions, 6 months apart, using both the FWT and the gross motor function measure (GMFM). Results. Generalisability correlation coefficients (GCC) for all 11 items were high (0.91-0.99). Inter-rater reliability was also high with excellent consensus in the scores given by the eight raters (intra-class correlation coefficient and GCC 0.99). Intra-rater reliability was equally high (GCC 0.99). The internal consistency of the FWT was estimated using Cronbach's α as 0.95 and 0.94 at Time 1 and 2, respectively. The FWT had a high

degree of correlation with the GMFM, when total scores were compared at Time 1 and 2 (Pearson's r = 0.86 and 0.87, n = 56, p < 0.01). The FWT also found statistically significant differences in total scores between the three Gross Motor Function Classification System (GMFCS) levels. The correlation between the FWT scores and GMFCS was -0.70 at Time 1 and -0.76 Time 2 (p < 0.01) indicating the construct validity of the FWT. Conclusions. This study has demonstrated that the FWT has sound psychometric properties and is valid and reliable in a sample population of ambulant children with CP.

PMID:21504407 [PubMed - as supplied by publisher]

16. Microsurgery. 2011 Apr 18. doi: 10.1002/micr.20877. [Epub ahead of print]

Contralateral C7 nerve root transfer in treatment of cerebral palsy in a child: Case report.

Xu WD, Hua XY, Zheng MX, Xu JG, Gu YD.

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A 4-year-old girl who sustained the hemiplegic cerebral palsy and subsequent spasticity in the left upper extremity underwent the C7 nerve root rhizotomy and the contralateral C7 nerve root transfer to the ipsilateral middle trunk of brachial plexus through an interpositional sural nerve graft. In a 2-year follow-up, the results showed a reduction in spasticity and an improvement in extension power of the elbow, the wrist, and the second to fifth fingers. Scores from both Quality of Upper Extremity Skills Test and Modified Ashworth Scale tests had been significantly improved during follow-up. The outcomes from this case provided the evidence that combined the C7 nerve root rhizotomy and contralateral healthy C7 nerve root transfer to the ipsilateral middle trunk of brachial plexus not only partially released flexional spasticity but also strengthened extension power of the spastic upper extremity in children with the cerebral palsy. © 2011 Wiley-Liss, Inc. Microsurgery, 2011.

PMID:21503970 [PubMed - as supplied by publisher]

Epidemiology / Aetiology / Diagnosis & Early Treatment

17. Cell Prolif. 2011 Apr;44 Suppl 1:60-9. doi: 10.1111/j.1365-2184.2010.00729.x.

The umbilical cord: a rich and ethical stem cell to advance regenerative medicine.

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Science and medicine place a lot of hope in the development of stem cell research and regenerative medicine. This review will define the concept of regenerative medicine and focus on an abundant stem cell - neonatal tissues such as the umbilical cord. Umbilical cord blood has been used clinically for over 20 years as a cell for haematopoietic stem cell transplantation. Beyond this, cord blood and umbilical cord-derived stem cells have demonstrated potential for pluripotent lineage differentiation (liver, pancreatic, neural tissues and more) in vitro and in vivo. This promising research has opened up a new era for utilization of neonatal stem cells, now used beyond haematology in clinical trials for autoimmune disorders, cerebral palsy or type I diabetes.

PMID:21481046 [PubMed - in process]

18. Fertil Steril. 2011 Apr 13. [Epub ahead of print]

Outcomes of spontaneous and assisted pregnancies in Turner syndrome: the U.S. National Institutes of Health experience.

Hadnott TN, Gould HN, Gharib AM, Bondy CA.

Developmental Endocrinology Branch, National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, Maryland; Howard Hughes Medical Institute, Chevy Chase, Maryland.

OBJECTIVE: To assess fetal and maternal outcomes of pregnancies in women with Turner syndrome (TS). DESIGN: Retrospective case series. SETTING: Clinical research center. PATIENT(S): 276 adults with cytogenetically proven TS participating in an intramural natural history protocol. INTERVENTION(S): None. MAIN OUTCOME MEASURE(S): Menstrual and obstetric histories, 50-cell karyotypes, and cardiovascular evaluation including aortic diameter measurements. RESULT(S): Our cohort included five women with spontaneous pregnancies and five with pregnancies using assisted reproduction (ART). All five women with spontaneous pregnancies had spontaneous puberty, despite 45,X in ≥90% of their 50-cell karyotype. Participants had a total of 13 pregnancies and 14 live births. One child had cerebral palsy; the others were chromosomally and developmentally normal. Delivery was by cesarean section in four out of seven spontaneous and six out of six ART-related pregnancies. One mother experienced preeclampsia in an ART-related twin pregnancy that required a preterm delivery; she has marked but stable aortic dilation years later. CONCLUSION(S): Approximately 2% of our study cohort experienced spontaneous pregnancies despite high-grade X monosomy, and a similar number achieved pregnancy via oocyte donation and ART. The potential for life-threatening cardiovascular complications warrants comprehensive screening before conception or single-embryo transfer, and caution regarding unintentional pregnancies for TS women.

PMID:21496813 [PubMed - as supplied by publisher]

19. Handb Clin Neurol. 2011;100:513-38.

Nonprimary dystonias.

Dressler D.

Dystonias can be classified as primary or secondary, as dystonia-plus syndromes, and as heredodegenerative dystonias. Their prevalence is difficult to determine. In our experience 80-90% of all dystonias are primary. About 20-30% of those have a genetic background; 10-20% are secondary, with tardive dystonia and dystonia in cerebral palsy being the most common forms. If dystonia in spastic conditions is accepted as secondary dystonia, this is the most common form of all dystonia. In primary dystonias, the dystonic movements are the only symptoms. In secondary dystonias, dystonic movements result from exogenous processes directly or indirectly affecting brain parenchyma. They may be caused by focal and diffuse brain damage, drugs, chemical agents, physical interactions with the central nervous system, and indirect central nervous system effects. Dystonia-plus syndromes describe brain parenchyma processes producing predominantly dystonia together with other movement disorders. They include dopa-responsive dystonia and myoclonus-dystonia. Heredodegenerative dystonias are dystonic movements occurring in the context of other heredodegenerative disorders. They may be caused by impaired energy metabolism, impaired systemic metabolism, storage of noxious substances, oligonucleotid repeats and other processes. Pseudodystonias mimic dystonia and include psychogenic dystonia and various orthopedic, ophthalmologic, vestibular, and traumatic conditions. Unusual manifestations, unusual age of onset, suspect family history, suspect medical history, and additional signs may indicate nonprimary dystonia. If they are suspected, etiological clarification becomes necessary. Unfortunately, potential etiologies are legion. Diagnostic algorithms can be helpful. Treatment of nonprimary dystonias, with few exceptions, does not differ from treatment of primary dystonias. The most effective treatment for focal and segmental dystonias is local botulinum toxin injections. Deep brain stimulation of the globus pallidus internus is effective for generalized dystonia. Antidystonic drugs, including anticholinergics, tetrabenazine, clozapine, and gamma-aminobutyric acid receptor agonists, are less effective and often produce adverse effects. Dopamine is extremely effective in dopa-responsive dystonia. The Bertrand procedure can be effective in cervical dystonia. Other peripheral surgery, including myotomy, myectomy, neurotomy, rhizotomy, ramizectomy, and accessory nerve neurolysis, has largely been abandoned. Central surgery other than deep brain stimulation is obsolete. Adjuvant therapies, including orthoses, physiotherapy, ergotherapy, behavioral therapy, social support, and support groups, may be helpful. Analgesics should also be considered where appropriate.

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20. Handb Clin Neurol. 2011;100:387-95.

Birth-related syndromes of athetosis and kernicterus.

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"Athetosis," from the Greek athetos, meaning "without fixed position," is a movement disorder first described by Hammond in 1871. The term described slow, irregular continual movements of the distal extremities. In 1983, Foley defined the athetoid syndrome as "a nonprogressive but evolving disorder due to damage to the basal ganglia of the full-term brain … [with] impairment of postural reflexes, arrhythmical involuntary movements, and dysarthria, [but] sparing … sensation, ocular movements and … intelligence." A decade later, "athetoid syndrome" was replaced by "dyskinetic cerebral palsy." Injury to basal ganglia by various mechanisms, including asphyxia, trauma, perinatal strokes, and kernicterus, is known to cause birth-related athetosis. Kernicterus originally described the neuropathology of bilirubin-induced brain injury, where the deep nuclei of the brain stain yellow. Kernicterus now describes the clinical features of chronic bilirubin encephalopathy, which include an extrapyramidal movement disorder, sensorineural hearing loss, impaired upward gaze, and dental enamel dysplasia. Aggressive treatment of perinatal hyperbilirubinemia has led to a decline in kernicterus so that, today, it is a rare cause of dyskinetic cerebral palsy. In this chapter, we provide a historic overview of athetosis and its formerly common cause, kernicterus. We relate earlier terminology to more recent definitions of impairments in dyskinetic cerebral palsy, including dystonia, chorea, and choreoathetosis.

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21. J Cereb Blood Flow Metab. 2011 Apr 20. [Epub ahead of print]

Therapeutic hypothermia alters microRNA responses to traumatic brain injury in rats.

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Therapeutic hypothermia promotes protection after traumatic brain injury (TBI). The mechanisms underlying hypothermic protection are multifactorial and may include the modulation of microRNA (miRNA) expression after trauma. We utilized microarrays to examine the effects of posttraumatic hypothermia on the expression of 388 rat miRNAs. Animals were subjected to sham or moderate fluid percussion brain injury, followed by 4 hours of hypothermia (33° C) or normothermia (37°C) and euthanized at 7 or 24 hours. At 7 hours, 47 miRNAs were significantly different ($P<0.05$) between TBI and sham (15 higher in TBI and 31 lower). After 24 hours, 15 miRNAs differed by $P<0.05$ (7 higher and 9 lower). The expression of miRNAs was altered by posttraumatic hypothermia. At 7 hours, seven were higher in hypothermia than normothermia and five were lower. Some miRNAs (e.g., miR-874 and miR-451) showed the most difference with hypothermia, with changes verified by quantitative reverse transcriptase-PCR. Regionally specific miRNAs also showed responses to TBI and hypothermia treatments by in situ hybridization. In addition, in vitro neuronal stretch injury studies showed similar temperature-sensitive responses to specific miRNAs. These novel data indicate that the reported beneficial effects of early hypothermia on traumatic outcome may include temperature-sensitive miRNAs involved in basic cell-processing events.Journal of Cerebral Blood Flow & Metabolism advance online publication, 20 April 2011; doi:10.1038/jcbfm.2011.33.

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22. Pediatrics. 2011 Apr 18. [Epub ahead of print]

Ethics of Resuscitation at Different Stages of Life: A Survey of Perinatal Physicians.

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Objective: We surveyed US neonatologists and high-risk obstetricians about preferences for resuscitation in ethically difficult situations to determine whether (1) their responses adhered to traditional ethical principles of best interests and patient autonomy and (2) physician specialty seemed to influence the response. Methods: In an electronic survey, we presented 8 vignettes with varying prognoses for survival and long-term outcome. Respondents were provided outcome data for mortality and morbidity in each vignette. We asked whether resuscitation was in the patient's best interest and whether the physician would accede to requests for nonresuscitation. Results: We analyzed surveys for 587 neonatologists and 108 high-risk obstetricians (15% overall response rate, 75% of web site visitors). There were no statistically significant differences in responses between the 2 physician subspecialty groups. As expected, in most cases there were inverse relationships between valuation of best interest and deferred resuscitation at the family's request. For example, for the oldest patient (an 80-year-old), 9.9% found resuscitation to be in the patient's best interest and 94.3% would allow nonresuscitation; for a 2-month-old, 93.9% found resuscitation to be in the patient's best interest and 24.5% would allow nonresuscitation. However, this pattern was not observed in the 2 newborn cases, in which resuscitation and nonresuscitation were both acceptable. In the triage scenario, the 7-year-old with cerebral palsy and acute trauma was consistently resuscitated first despite others having equivalent or better short- and long-term prognoses. Conclusions: On the basis of our results, physicians' decisions to resuscitate seem to be context-specific, rather than based on prognosis or consistent application of best-interest or autonomy principles. Despite their different professional perspectives, neonatologists and high-risk obstetricians seemed to converge on these judgments.

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23. Pain. 2011 Apr 15. [Epub ahead of print]

Chronic pain in adults with an intellectual disability: Prevalence, impact, and health service use based on caregiver report.

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This study examined chronic pain in adults with an intellectual disability (ID), in terms of its prevalence, impact on physical and psychological functioning, and treatments used. Questionnaires were distributed to 2378 primary caregivers (caregivers) of community-dwelling adults with an ID. The questionnaires were used to gather data on demographics, general health, nature of pain, impact of pain, treatment, and health-related decision making. Responses were received from 753 caregivers (31.6% response rate). Caregivers reported that 15.4% of this sample was experiencing chronic pain, for an average of 6.3years. Significantly more females than males were reported to experience chronic pain, although age, communication ability, and level of ID were not found to be associated with the presence of pain. However, the presence of pain was associated with cerebral palsy, physical disability, and reports of challenging behaviour. A significant proportion of individuals with chronic pain also experienced limitations in several aspects of daily living, and more than 78% of caregivers reported that the service user had become upset or distressed by pain. More than 80% of service users were receiving some form of treatment for their pain, with most seeing a family physician and using analgesics as the primary form of pain treatment. Results indicate that chronic pain is a significant problem for persons with an ID, with a proportion of service users living with daily pain for many years and experiencing limitations in daily functioning, emotional well-being, and quality of life. The caregivers (caregivers) of 753 persons with an intellectual disability reported that chronic pain affected 15% of those in their care, with increased prevalence of pain among females with a co-occurring physical disability. Chronic pain may significantly affect the quality of life of individuals with an intellectual disability.

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24. Zh Nevrol Psikhiatr Im S S Korsakova. 2011;111(4):8-11.

Visual disorders in children cerebral palsy. [Article in Russian]

Sal'kov VN, Levchenkova VD, Lobanova LV, Grishina TG, Sheĭnkman OG.

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A complex study of 55 patients with children cerebral palsy who complained on visual and eye movement disturbances in the age from 4 to 10 years has been carried out. Main factors in the anamnesis that have negative effect on the organism formation in antenatal and perinatal periods have been analyzed. Most of children had ophthalmologic symptoms (visual acuity decrease, disturbance of pupillary reaction to convergence, disturbance of refraction, refraction strabismus, horizontal nystagmus, restriction of the field of vision, ophtalmoparesis), movement and coordination disorders, mental and speech disorders. Plausibly, the combination of different aversive factors in the antenatal, intranatal and neonatal periods led to the brain lesion, including structures of movement and visual analyzers and their links to other analyzers, that determined the clinical picture of disease.

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