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CP Research News

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1: *Pediatr Neurol.* 2009 Jan;40(1):19-26.

Neuropsychologic impairment in bilateral cerebral palsy.

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The lower-than-average cognitive performance of individuals with bilateral cerebral palsy found in previous studies does not always refer to an abnormal performance or clinically significant impairment. We aimed to establish the percentage of persons with bilateral cerebral palsy who present neuropsychologic impairment, and its relationship to perinatal data and motor signs. Forty children, adolescents, and adults (age range, 6-38 years; 15 females and 25 males) with bilateral cerebral palsy were neuropsychologically assessed. Vocabulary was impaired in 85% of participants, language comprehension in 13-48%, visuo-perceptual abilities in 60%, visuospatial abilities in 90%, short-term memory in 21-58%, declarative memory in 47-67%, and praxis comprehension in 20%, with executive deficits in 58-74%. Perinatal data (intrauterine growth and birth weight) contributed to explaining memory impairment. Among cerebral palsy subtypes (spastic, mixed, and dyskinetic), forms of impairment differed only in short-term verbal memory. No persons with dyskinetic cerebral palsy experienced impairment in immediate memory or working visual memory. We conclude that visuospatial deficit is the most frequent impairment in people with bilateral cerebral palsy. Moreover, short-term memory impairment seems sensitive to perinatal complications, and differs among bilateral cerebral palsy subtypes.

PMID: 19068249 [PubMed - in process]



Please join us in February 2009 at the 3rd International Cerebral Palsy Conference in Sydney, Australia. Hosted by the CP Institute, keynote speakers include some of the world's leading cerebral palsy researchers. Earl bird registrations close 10 December 2008 www.cp2009.com.au

2: Brain Dev. 2008 Dec 5. [Epub ahead of print]**Associated factors in neonatal hypoglycemic brain injury.**

Montassir H, Maegaki Y, Ogura K, Kurozawa Y, Nagata I, Kanzaki S, Ohno K.

Division of Child Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University, 36-1 Nishi-cho, Yonago 683-8504, Japan.

Although associated factors are important for the occurrence of neural damage in neonatal hypoglycemia, they are not fully understood. Sixty patients with neonatal hypoglycemia were studied through a review of their medical records in Tottori University Hospital. The patients were classified into two main groups: Group I were patients who had mental retardation, developmental delay, cerebral palsy or epilepsy while Group II were those who were normal in their follow-up. Group I consisted of 12 patients while Group II consisted of 48 patients. The median gestational age was 38 weeks in Group I and 36.7 weeks in Group II. The frequencies of small for gestational age were similar in both groups. Blood glucose levels less than 15mg/dl were more frequent in Group 1 (50.0%) than in Group 2 (14.6%) ($P=0.015$). Duration of hypoglycemia was longer in Group I (median, 14h) than in Group II (median, 1.75h) ($p<0.001$). The following factors were more frequent in Group I than in Group II: toxemia (33.3% and 8.3%, $p=0.043$), fetal distress (58.3% and 14.5%, $p=0.004$), an Apgar score of less than 5 at 1min (33.3% and 6.4%, $p=0.025$), neonatal seizure (53.8% and 4.3%, $p<0.001$) and pathological jaundice (41.7% and 6.4%, $p=0.006$). Cranial CT or MRI revealed cerebral lesions in 8 of the 9 Group I patients in follow-up examinations. This study indicates that severe and prolonged neonatal hypoglycemia can cause cerebral lesions and other perinatal risk factors, such as hypoxia, neonatal seizure and pathological jaundice, would exacerbate hypoglycemic brain injuries.

PMID: 19059741 [PubMed - as supplied by publisher]

3: Int J Qual Health Care. 2008 Dec 5. [Epub ahead of print]**Evaluating patient care communication in integrated care settings: application of a mixed method approach in cerebral palsy programs.**

Gulmans J, Vollenbroek-Hutten MM, Van Gemert-Pijnen JE, Van Harten WH.

1Roessingh Research & Development, Institute for Research in Rehabilitation Medicine and Technology, Enschede, the Netherlands.

OBJECTIVE: In this study, we evaluated patient care communication in the integrated care setting of children with cerebral palsy in three Dutch regions in order to identify relevant communication gaps experienced by both parents and involved professionals. **DESIGN:** A three-step mixed method approach was used starting with a questionnaire to identify communication links in which parents experienced gaps. In subsequent in-depth interviews with parents and focus group meetings with professionals underlying factors were evaluated. **RESULTS:** In total, 197 parents completed the questionnaire (response 67%); 6% scored negative on parent-professional communication, whereas 17% scored negative on inter-professional communication, especially between the rehabilitation physician and primary care physiotherapy (16%) and (special) education/day care (15%). In-depth interviews among a subset of 20 parents revealed various sources of dissatisfaction such as lack of cooperation and patient centeredness, inappropriate amount of information exchange and professional use of parents as messenger of information. Focus group meetings revealed that professionals recognized these gaps. They attributed them to capacity problems, lack of interdisciplinary guidelines and clear definition of roles, but also a certain hesitance for contact due to unfamiliarity with involved professionals in the care network. **CONCLUSIONS:** Parents particularly identified gaps in inter-professional communication between (rehabilitation) hospitals and primary care settings. Involved professionals recognized these gaps and primarily attributed them to organizational factors. Improvement initiatives should focus on these factors as well as facilitation of low-threshold contact across the patient's care network.

PMID: 19060037 [PubMed - as supplied by publisher]

4: J Mal Vasc. 2008 Dec 5. [Epub ahead of print]

A childhood case of antiphospholipid syndrome. [Article in French]

Chabchoub I, Ben Thabet A, Maaloul I, Aloulou H, Kamoun T, Daoued H, Mnif Z, Hachicha M.

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Many conditions can lead to cerebral strokes in children. The antiphospholipid syndrome widely described in adults in association with systemic lupus erythematosus, is rare in childhood. **CASE REPORT:** Two months after recovering from varicella and a few days after an episode of bronchitis, a 17-month-old girl developed left facial paralysis associated with right hemiplegia. Brain MRI and angio-scan showed thrombosis in the internal left carotid associated with ischemia in the superficial posterior territory of the left Sylvian artery. Echocardiography and hemoglobin electrophoresis were normal. Tests were negative for protein S, C and antithrombin III deficiencies and no resistance to activated protein C. IgM anticardiolipin antibodies were detected at high level (greater than 25IU/l) initially and six weeks later. In the absence of an evident etiology, mainly systemic lupus erythematosus (negative antinuclear antibodies), the diagnosis of primary antiphospholipid syndrome was retained. The girl was treated by heparin then by salicylate at antiaggregante doses associated with re-habilitation. Twelve months later, the patient had not developed any other thrombosis, in spite of a high level of anticardiolipin antibodies. **CONCLUSION:** In children with cerebral strokes, antiphospholipid syndrome must be discussed when the usual etiologies have been ruled out.

PMID: 19059742 [PubMed - as supplied by publisher]

5: Health Qual Life Outcomes. 2008 Dec 4;6(1):109. [Epub ahead of print]

Psychometric properties of the Child Health Assessment Questionnaire (CHAQ) applied to children and adolescents with cerebral palsy.

Morales NM, Funayama CA, Rangel VO, Frontarolli AC, Araujo RR, Pinto RM, Rezende CH, Silva CH.

ABSTRACT: BACKGROUND: Cerebral palsy (CP) patients have motor limitations that can affect functionality and abilities for activities of daily living (ADL). Health related quality of life and health status instruments validated to be applied to these patients do not directly approach the concepts of functionality or ADL. The Child Health Assessment Questionnaire (CHAQ) seems to be a good instrument to approach this dimension, but it was never used for CP patients. The purpose of the study was to verify the psychometric properties of CHAQ applied to children and adolescents with CP. **METHODS:** Parents or guardians of children and adolescents with CP, aged 5 to 18 years, answered the CHAQ. A healthy group of 314 children and adolescents was recruited during the validation of the CHAQ Brazilian-version. Data quality, reliability and validity were studied. The motor function was evaluated by the Gross Motor Function Measure (GMFM). **RESULTS:** Ninety-six parents/guardians answered the questionnaire. The age of the patients ranged from 5 to 17.9 years (average: 9.3). The rate of missing data was low (<9.3%). The floor effect was observed in two domains, being higher only in the visual analogue scales (<35.6%). The ceiling effect was significant in all domains and particularly high in patients with quadriplegia (81.8 to 90.9%) and extrapyramidal (45.4 to 91.0%). The Cronbach alphacoefficient ranged from 0.85 to 0.95. The validity was appropriate: for the discriminant validity the correlation of the disability index with the visual analogue scales was not significant; for the convergent validity CHAQ disability index had a strong correlation with the GMFM (0.77); for the divergent validity there was no correlation between GMFM and the pain and overall evaluation scales; for the criterion validity GMFM as well as CHAQ detected differences in the scores among the clinical type of CP ($p < 0.01$); for the construct validity, the patients' disability index score (mean:2.16 SD:0.72) was higher than the healthy group (mean:0.12 SD:0.23)($p < 0.01$). **CONCLUSION:** CHAQ reliability and validity were adequate to this population. However, further studies are necessary to verify the influence of the ceiling effect on the respon-

siveness of the instrument.

PMID: 19055820 [PubMed - as supplied by publisher]

6: Child Care Health Dev. 2008 Dec 3. [Epub ahead of print]

Development of a measure of family-centred care for resource-poor South African settings: the experience of using a modified version of the MPOC-20.

Saloojee GM, Rosenbaum PR, Westaway MS, Stewart AV.

School of Therapeutic Sciences, University of the Witwatersand, Johannesburg, South Africa.

Background The Measure of Processes of Care (MPOC) is a widely used tool to assess parents' self-reported experiences of family-centred behaviours of paediatric rehabilitation services. It has never been used in resource-constrained settings or in a cross-cultural environment where cultural and language differences may complicate effective implementation of family-centred services. In this study, the MPOC-20 was used as the starting point for the development of a measure of family-centred care in disadvantaged South African settings. The objective was to establish to what extent the MPOC-20 needed to be adapted for these settings. **Methods** After modifying MPOC-20 through focus groups, the adapted scale was translated into six local languages. Trained interviewers administered the scale to a convenience sample of 267 caregivers of children aged between 1 and 18 years with a diagnosis of cerebral palsy living in poorly resourced areas in two provinces in South Africa. **Results** The modified MPOC-20 was neither reliable nor valid in the new setting. Cronbach's alpha for each of the sub-scales varied between 0.30 and 0.66 while for the test-retest reliability, the Intraclass Correlation Coefficients were between 0.51 and 0.61. The first two criteria for item convergent validity were not met. Repeated multi-trait scaling identified eight items that when combined into a scale [named the MPOC-8(SA)] had acceptable reliability and validity. Factor analysis of the MPOC-8(SA) yielded two factors: an interpersonal factor and an informational factor. **Conclusions** Although extreme caution has to be used when using measures created in one socio-cultural setting in a different context, the MPOC-20 provides a useful starting point for the development of a measure of family-centred care in a poor resourced setting. Caregivers in different settings have more in common than they have differences. However, the process of asking the questions and the words used to capture caregivers' experiences needs to be different.

PMID: 19055651 [PubMed - as supplied by publisher]

7: Dev Med Child Neurol. 2008 Dec 3. [Epub ahead of print]

A classification system for hip disease in cerebral palsy.

Robin J, Graham HK, Baker R, Selber P, Simpson P, Symons S, Thomason P.

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In population-based studies, hip displacement affects approximately one-third of children with cerebral palsy (CP). Given the extreme range of clinical phenotypes in the CP spectrum, it is unsurprising that hip development varies from normality, to dislocation and degenerative arthritis. Numerous radiological indices are available to measure hip displacement in children with CP; however, there is no grading system for assessing hip status in broad categorical terms. This makes it difficult to audit the incidence of hip displacement, determine the relationship between hip displacement and CP subtypes, assess the outcome of intervention studies, and to communicate hip status between health care professionals. We developed a categorical, radiographic classification of hip morphology based on qualitative indices and measurement of the key continuous variable, the migration percentage of Reimers. One hundred and thirty-four radiographs were reviewed of 52 female and 82 male adolescents with CP who were at, or close to, skeletal maturity (mean age 16y 1mo [SD 1y 4mo] range 14y to 19y 1mo). Twenty-nine were

classified at Gross Motor Function Classification System (GMFCS) Level I, 25 at Level II, 27 at Level III, 24 at Level IV, and 29 at Level V. A classification system was developed to encapsulate the full spectrum of hip morphology in CP, with and without intervention.

PMID: 19055594 [PubMed - as supplied by publisher]

8: J Laryngol Otol. 2008 Dec 2:1-3. [Epub ahead of print]

Drooling in oro-motor dyspraxia: is there a role for surgery?

Rimmer J, Hartley BE.

Department of Otolaryngology, Great Ormond Street Hospital for Children, London, UK.

Objective:To report the successful use of a surgical approach for the treatment of drooling in a case of oro-motor dyspraxia.
Case report:A seven-year-old girl with a diagnosis of oro-motor dyspraxia was referred for management of drooling; this was her only symptom. There were no focal neurological abnormalities. Conservative measures had not helped, and her parents were keen for definitive treatment. Bilateral submandibular duct transposition was performed with no complications. The patient's drooling improved immediately.
Conclusion:There are currently no reports in the literature of surgery for persistent drooling in children with oro-motor dyspraxia who are otherwise neurologically and developmentally normal. The mainstay of treatment is speech and language therapy. We report the successful use of surgical techniques, usually reserved for children with cerebral palsy or severe neurological disorders, to treat drooling in an otherwise normal seven-year-old child with oro-motor dyspraxia.

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9: Pediatr Crit Care Med. 2008 Dec 2. [Epub ahead of print]

Antibiotic-resistant bacteria and infection in children with cerebral palsy requiring mechanical ventilation.

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INTRODUCTION: Severe and chronic illness can alter the bacterial flora carried in the oropharynx and gut. There are little data on the bacterial flora of children with chronic neurologic impairment. **OBJECTIVES:** To assess carriage of abnormal bacterial flora, antibiotic-resistant bacteria, infection, and mortality in children with cerebral palsy (CP) admitted for pediatric intensive care. **DESIGN:** Prospective observational single center cohort study. **SETTING:** Twenty-bed regional pediatric intensive care unit (PICU) in a university-affiliated tertiary referral children's hospital. **PATIENTS:** All children with an established diagnosis of CP admitted to PICU and ventilated for four or more days during a 6-yr period. **MEASUREMENTS:** Surveillance samples of throat and rectum were taken at admission to PICU and twice a week thereafter. Diagnostic samples were obtained on clinical indication. **MAIN RESULTS:** Fifty-three children with a total of 77 admissions were included. Most (90%) of the children with CP had moderate to severe functional limitations. Eighty-nine percent of the children with CP (47/53) carried abnormal bacterial flora/potential pathogens, most frequently *Pseudomonas* and *Klebsiella* species. Forty-seven percent (22/47) had antibiotic-resistant bacteria. Thirty-five children (66%) developed 86 infections during their PICU admission. Lower airways and blood were the two most commonly infected sites-*Pseudomonas aeruginosa* and coagulase-negative *Staphylococci*, the predominant infecting microorganisms. Sixty-five percent (56/86) of infections were primary endogenous infections, 21% (18/86) exogenous, and 9% (8/86) secondary endogenous. Carriage of abnormal bacterial flora, antibiotic-resistant bacteria, and infection rate was significantly higher than that of children of comparative age without CP

ventilated for four or more days on PICU. Nine (17%) of the children with CP died in PICU and four of the deaths were infection related. **CONCLUSIONS:** In children with moderate to severe chronic neurologic impairment admitted to PICU, there is a high rate of carriage of abnormal bacteria/potential pathogens, antibiotic-resistant bacteria, and infection.

PMID: 19057452 [PubMed - as supplied by publisher]

10: Arch Phys Med Rehabil. 2008 Dec;89(12):2302-8.

The effect of simulating weight gain on the energy cost of walking in unimpaired children and children with cerebral palsy.

Plasschaert F, Jones K, Forward M.

Movement Analysis Laboratory, University Hospital, Ghent, Belgium.

Plasschaert F, Jones K, Forward M. The effect of simulating weight gain on the energy cost of walking in unimpaired children and children with cerebral palsy. **OBJECTIVE:** To examine the effect of simulating weight gain on the energy cost of walking in children with cerebral palsy (CP) compared with unimpaired children. **DESIGN:** Repeated measures, matched subjects, controlled. **SETTING:** University hospital clinical gait and movement analysis laboratory. **PARTICIPANTS:** Children (n=42) with CP and unimpaired children (n=42). **INTERVENTIONS:** Addition of 10% of body mass in weight belt. **MAIN OUTCOME MEASURES:** Energy cost of walking parameters consisting of walking speed, Physiological Cost Index, Total Heart Beat Index, oxygen uptake ($Vo(2)$), gross oxygen cost, nondimensional net oxygen cost, and net oxygen cost with speed normalized to height were measured by using a breath-by-breath gas analysis system (K4b(2)) and a light beam timing gate system arranged around a figure 8 track. Two walking trials were performed in random order, with and the other without wearing a weighted belt. **RESULTS:** Children with CP and their unimpaired counterparts responded in fundamentally different ways to weight gain. The unimpaired population maintained speed and $Vo(2)$ but the children with CP trended toward a drop in their speed and an increase in their $Vo(2)$. The oxygen consumption of children with CP showed a greater dependence on mass than the unimpaired group ($P=.043$). **CONCLUSIONS:** An increase of a relatively small percentage in body mass began to significantly impact the energy cost of walking in children with CP. This result highlights the need for weight control to sustain the level of functional walking in these children.

PMID: 19061743 [PubMed - in process]

11: Dev Med Child Neurol. 2008 Dec;50(12):918-25.

Outcome tools used for ambulatory children with cerebral palsy: responsiveness and minimum clinically important differences.

Oeffinger D, Bagley A, Rogers S, Gorton G, Kryscio R, Abel M, Damiano D, Barnes D, Tylkowski C.

Shriners Hospital for Children (SHC), Lexington, KY, USA.

This prospective longitudinal multicenter study of ambulatory children with cerebral palsy (CP) examined changes in outcome tool score over time, tool responsiveness, and used a systematic method for defining minimum clinically important differences (MCIDs). Three hundred and eighty-one participants with CP (Gross Motor Function Classification System [GMFCS] Levels I-III; age range 4-18y, mean age 11y [SD 4y 4mo]; 265 diplegia, 116 hemiplegia; 230 males, 151 females). At baseline and follow-up at least 1 year later, Functional Assessment Questionnaire, Gross Motor Function Measure, Pediatric Quality of Life Inventory, Pediatric Outcomes Data Collection Instrument, Pediatric Functional Independence Measure, temporal-spatial gait parameters, and oxygen cost were collected. Adjusted standardized response means determined tool responsiveness for nonsurgical (n=292) and surgical (n=87) groups at GMFCS Levels I to III. Most scores reaching medium or large effect sizes were for GMFCS Level III.

Nonsurgical group change scores were used to calculate MCID thresholds for ambulatory children with CP. These values were verified by examining participants who changed GMFCS levels. Tools measuring function were responsive when a change large enough to cause a change in GMFCS level occurred. MCID thresholds assess change in study populations over time, and serve as the basis for designing prospective intervention studies.

PMID: 19046185 [PubMed - in process]

12: Dev Med Child Neurol. 2008 Dec;50(12):886.

Cerebral palsy and newborn care: I, II, and III (1981).

Stanley F.

Telethon Institute for Child Health Research, Centre for Child Health Research, The University of Western Australia.

PMID: 19046182 [PubMed - in process]

13: J Bone Joint Surg Am. 2008 Dec;90(12):2735-44.

Biologic characteristics of fibrous hamartoma from congenital pseudarthrosis of the tibia associated with neurofibromatosis type 1.

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BACKGROUND: Fibrous hamartoma is a key pathologic component of congenital pseudarthrosis of the tibia, a challenging and disabling bone disorder. We investigated the biologic characteristics of fibrous hamartoma cells in order to better understand the pathogenesis of this rare disease. **METHODS:** Fibrous hamartoma tissues were surgically excised at the time of osteosynthesis from seven patients with congenital pseudarthrosis of the tibia associated with neurofibromatosis type 1. Distal tibial periosteum was also harvested as control tissue during tibial derotation osteotomy from two other patients with cerebral palsy and one patient with idiopathic internal tibial torsion. Fibroblast-like cells were enzymatically dissociated and cultured from these tissues. Immunophenotypes were investigated for positive (CD44 and CD105) and negative (CD45 and CD14) mesenchymal lineage cell markers, and the mRNA expressions of bone morphogenetic protein(BMP)-2, BMP-4, and their receptors were assayed by reverse transcription-polymerase chain reaction. After rhBMP-2 treatment, the changes in alkaline phosphatase activity, and in the mRNA expressions of type-I collagen (COL1A1), alkaline phosphatase, and osteocalcin genes, were assayed with use of an RNase protection assay. The mRNA expressions of receptor activator of nuclear factor-kappa B ligand (RANKL) and osteoprotegerin (OPG) were quantitatively assayed with use of real-time RT-PCR. Osteoclastic differentiation of RAW(264.7) cells in coculture with fibrous hamartoma cells was evaluated. **RESULTS:** All fibrous hamartoma and tibial periosteal cells tested were CD44+/CD105+/CD45-/CD14- and expressed the mRNAs of BMP-2, BMP-4, and their receptors. The baseline mRNA expressions of COL1A1, alkaline phosphatase, and osteocalcin genes in the fibrous hamartoma cells were diverse. These gene expressions were upregulated by BMP treatment in tibial periosteal cells but did not change or were downregulated in fibrous hamartoma cells. Fibrous hamartoma cells expressed higher levels of RANKL and lower levels of OPG than did tibial periosteal cells. Coculture with fibrous hamartoma cells enhanced osteoclastic differentiation of RAW(264.7) cells. **CONCLUSIONS:** Fibrous hamartoma cells maintain some of the mesenchymal lineage cell phenotypes, but do not undergo osteoblastic differentiation in response to BMP. They are more osteoclastogenic than are tibial periosteal cells.

Publication Types:

Research Support, Non-U.S. Gov't

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14: J Pediatr Surg. 2008 Dec;43(12):2178-81.

Experience with a hybrid, minimally invasive gastrostomy for children with abnormal epigastric anatomy.

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PURPOSE: This communication is an analysis of the experience with a new type of gastrostomy. It was developed for patients with pronounced epigastric anatomical abnormalities, such as dense adhesions, in whom the conventional "open" gastrostomy could prove difficult and the percutaneous endoscopic, the imaging-guided, and the laparoscopically assisted methods would be unsafe. **METHOD:** A large, soft rubber catheter is inserted in the child's mouth and advanced into the stomach. A small epigastric incision is made. With the help of the catheter, the anterior gastric wall is identified and the stoma site chosen. One of the curved needles of a double-armed monofilament suture is passed through the gastric wall and through the catheter. The needle is then cut off. The other needle is passed through the abdominal wall, from the inside out at the most suitable skin stoma site. When the catheter (with the embedded suture) is withdrawn from the mouth, a tract is established. The suture is replaced by a guide wire, which allows a percutaneous endoscopic gastrostomy-type catheter to be placed by the percutaneous endoscopic gastrostomy "pull" technique. **RESULTS:** This approach was used in 15 patients (14 children-ages 1 month to 7 years and one 19 years old) with: status post (s/p) necrotizing enterocolitis and bowel loss (n = 4); s/p gastroschisis and short-gut syndrome (n = 3); cerebral palsy, s/p ventriculo-peritoneal shunt infections (n = 2); s/p complex omphalocele; dwarfism; morphologic abnormalities; repaired prune-belly syndrome; s/p duodenal atresia with malrotation; severe scoliosis with s/p multiple shunt infections (one each). There were no complications. The technique also proved useful in several other children in whom a laparotomy incision for unrelated conditions was remote from the gastrostomy site. **CONCLUSION:** Using a very small incision, this hybrid method permits safe and precise gastric and abdominal wall site selection and gastrostomy catheter placement. Gastrostomy as well as purse-string and peritoneal fixation sutures are not needed, and the danger of accidental catheter dislodgement is minimized.

PMID: 19040930 [PubMed - in process]

15: J Spinal Disord Tech. 2008 Dec;21(8):606-13.

Correction of apical axial rotation with pedicular screws in neuromuscular scoliosis.

Modi HN, Suh SW, Song HR, Lee SH, Yang JH.

Department of Orthopedics, Korea University, Guro Hospital, Seoul, Korea.

STUDY DESIGN: A retrospective study to measure the postoperative apical axial derotation with posterior pedicle screw fixation in neuromuscular scoliosis. **OBJECTIVES:** To determine whether the posterior only approach using pedicle screw fixation is able to accomplish apical axial derotation in neuromuscular scoliosis and if there is any difference according to severity of curve or type of disease. **SUMMARY OF BACKGROUND DATA:** Literature search does not reveal anything about the rectification of apical axial rotation in neuromuscular scoliosis with the pedicle screw fixation. **METHODS:** Between January 2005 and December 2006, 24 patients (9 females and 15 males, average age 19 y) with neuromuscular scoliosis (6 cerebral palsy, 9 Duchenne muscular dystrophy, 5 spinal muscular atrophy, and 4 others) underwent posterior pedicle screw construct with correction and fusion for the treatment of progressive,

symptomatic spinal deformities. Preoperative, immediate postoperative, and final follow-up radiographs were analyzed according to Cobb's angle and pelvic obliquity, whereas apical axial rotation was measured on preoperative and postoperative computerized tomography scan using Aaro-Dahlborn method from mid-sagittal plane. Twelve (9 females and 3 males) adolescent idiopathic scoliosis patients, who underwent similar operation, comprised our control group for the comparison of results. **RESULTS:** All the patients exhibited improved sitting balance after surgery. The mean preoperative Cobb's angle, pelvic obliquity, and apical rotation were 74, 14, and 42 degrees, whereas postoperative were 32, 6, and 33 degrees, respectively, showing significant correction in all 3 parameters. Comparison of results based on severity of curve did not reveal any statistically significant difference ($P=0.255$) in correction of apical axial rotation among group I, group II, and group III. A similar apical rotational correction was recorded in different disease groups ($P=0.295$). Comparing the results between neuromuscular and idiopathic scoliosis groups, we could not find any statistically significant difference. **CONCLUSIONS:** Our results indicate that apical axial derotation can be well achieved with posterior only pedicle screw fixation in patients with neuromuscular scoliosis without any need for an anterior release procedure.

PMID: 19057256 [PubMed - in process]

16: Presse Med. 2008 Dec;37(12):1793-801. Epub 2008 Sep 4.

Peripheral nerve blocks and spasticity. Why and how should we use regional blocks? [Article in French]

Viel E, Pellas F, Ripart J, Pélissier J, Eledjam JJ.

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Muscle spasticity causes pain, disability, and difficulties in the rehabilitation of patients with cerebrovascular lesions, head, brain or spine trauma, coma, or neurologic diseases such as multiple sclerosis, amyotrophic lateral sclerosis, or cerebral palsy. Regional blocks have a threefold use in patients with painful spasticity: diagnostic, prognostic, and therapeutic. Blocks are feasible on an outpatient or day-hospital basis. Blocks are applied most often to 4 peripheral sites: the pectoral nerve loop, median, obturator, and tibial nerves. The main indication is debilitating or painful spasticity. Peripheral blocks with local anesthetics are used as tests, to mimic the effects of motor blocks and determine their potential adverse effects, transiently and reversibly. Peripheral neurolytic blocks are easy to perform, effective, and inexpensive.

Publication Types:
English Abstract

PMID: 18775634 [PubMed - in process]

17: Early Hum Dev. 2008 Nov 26. [Epub ahead of print]

Intrapartum fetal heart rate patterns in infants (≥ 34 weeks) with poor neurological outcome.

Kodama Y, Sameshima H, Ikeda T, Ikenoue T.

Department of Obstetrics and Gynecology and Perinatal Center, Faculty of Medicine, University of Miyazaki, Japan.

BACKGROUND: Cases suggestive of non-acidemia related cerebral palsy (CP) are likely misdiagnosed as acidemia related CP because of the presence of nonreassuring fetal heart rate (FHR) patterns. **AIMS:** Our purpose was to compare intrapartum FHR patterns between the cases of neurological damage and the cases without disability after severe metabolic acidemia and neonatal encephalopathy, and also to

compare the FHR patterns between cases with CP due to asphyxia and cases with CP of other etiology in infants born after 34 weeks. **STUDY DESIGN:** From 1998 to 2003, our peer review conferences determined 136 infants with high-risk factors for neurological impairment in the unselected 65,197 live births. High-risk infants were chosen according to our criteria. Among them 58 were eligible infants because they were born at ≥ 34 weeks of gestation and also had legible FHR traces. **OUTCOME MEASURES:** Incidence of nonreassuring FHR patterns. **RESULTS:** Fifteen infants were acidemia related and 43 were non-acidemia related high-risk infants. Ten of the 15 acidemia infants developed CP and all had shown bradycardia ≥ 13 min with a nadir < 80 bpm. In the 43 non-acidemia infants, 35 had CP, mental retardation, epilepsy, or hearing loss and 74% (26/35) of them had shown nonreassuring FHR patterns. Incidence of severe bradycardia was significantly elevated in the acidemia related CP compared with acidotic infants without disability, and those with non-acidemia related CP. **CONCLUSIONS:** Even in infants with non-acidemia related CNS impairments, who were born at ≥ 34 weeks of gestation, 74% had shown intrapartum nonreassuring FHR patterns.

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18: J Vis Exp. 2008 Nov 19;(21). pii: 955. doi: 10.3791/955.

The hypoxic-ischemic encephalopathy model of perinatal ischemia.

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Hypoxic-Ischemic Encephalopathy (HIE) is the consequence of systemic asphyxia occurring at birth. Twenty five percent of neonates with HIE develop severe and permanent neuropsychological sequelae, including mental retardation, cerebral palsy, and epilepsy. The outcomes of HIE are devastating and permanent, making it critical to identify and develop therapeutic strategies to reduce brain injury in newborns with HIE. To that end, the neonatal rat model for hypoxic-ischemic brain injury has been developed to model this human condition. The HIE model was first validated by Vannucci et al (1) and has since been extensively used to identify mechanisms of brain injury resulting from perinatal hypoxia-ischemia (2) and to test potential therapeutic interventions (3,4). The HIE model is a two step process and involves the ligation of the left common carotid artery followed by exposure to a hypoxic environment. Cerebral blood flow (CBF) in the hemisphere ipsilateral to the ligated carotid artery does not decrease because of the collateral blood flow via the circle of Willis; however with lower oxygen tension, the CBF in the ipsilateral hemisphere decreases significantly and results in unilateral ischemic injury. The use of 2,3,5-triphenyltetrazolium chloride (TTC) to stain and identify ischemic brain tissue was originally developed for adult models of rodent cerebral ischemia (5), and is used to evaluate the extent of cerebral infarction at early time points up to 72 hours after the ischemic event (6). In this video, we demonstrate the hypoxic-ischemic injury model in postnatal rat brain and the evaluation of the infarct size using TTC staining.

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19: Epilepsia. 2008 Nov 17. [Epub ahead of print]

Clinical spectrum and medical treatment of children with electrical status epilepticus in sleep (ESES).

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Purposes: To describe the clinical spectrum and to evaluate the efficacy of different therapeutic agents in children with electrical status epilepticus in sleep (ESES). **Methods:** Clinical data of all patients with

ESES (not including patients with Landau-Kleffner syndrome) in four pediatric neurology outpatient clinics were analyzed. Thirty patients with ESES had been treated between 1994 and 2007. Results: Eleven (37%) children had benign partial epilepsies of childhood, five (17%) had cerebral palsy, five (17%) had hydrocephalus, one (3%) had schizencephaly, one (3%) had prenatal parenchymal bleeding, and the etiology was unclear in seven (23%). The duration of ESES ranged between 2 and 60 months. The antiepileptic drugs that were found to be efficacious were: levetiracetam (41%), clobazam (31%), and sulthiame (17%). Valproic acid, lamotrigine, topiramate, and ethosuximide showed no efficacy. Steroids were efficacious in 65%; immunoglobulins were efficacious in 33%. High-dose diazepam was efficacious in 37%, but all the children had temporary response. Seventeen patients (57%) had cognitive deterioration, whereas the rest presented with regression in attention, speech, communication, and behavior. Fourteen children had permanent cognitive deficit. There was a significant correlation ($p = 0.029$) between the duration of ESES and residual intellectual deficit at follow-up. Conclusions: ESES reflects an evolution of benign partial epilepsy of childhood in more than one-third of the patients, whereas there is an underlying structural brain anomaly in another one-third. The most efficacious antiepileptic drugs (AEDs) are levetiracetam and clobazam. The duration of ESES correlated significantly with residual intellectual deficit at follow-up.

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20: Dev Med Child Neurol. 2008 Nov;50(11):828-31.

Validity and reliability of the guidelines of the surveillance of cerebral palsy in Europe for the classification of cerebral palsy.

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The validity and reliability of the guidelines of the Surveillance of Cerebral Palsy in Europe (SCPE) for the classification of cerebral palsy (CP) were tested by administering 10 written case vignettes via an interactive web-based link to 30 SCPE partners. There was a moderately good level of agreement ($\kappa=0.59$) about inclusion as a CP case on the SCPE database. Classification by CP subtype differed in two main areas: assigning spastic versus dyskinetic and judgement of distribution of spastic involvement. Agreement on Gross Motor Function Classification System (GMFCS) level was less good than reported in previous studies. Twenty respondents repeated the test 5 months later and there was good repeatability for case inclusion ($\kappa=0.72$) but considerable variation in assignment of CP subtype and GMFCS level. There is a need for further collaborative work and training to improve harmonization of the classification of CP, including examination, application of SCPE guidelines, and register coding.

Publication Types:
Research Support, Non-U.S. Gov't

PMID: 19058397 [PubMed - in process]

21: Dev Med Child Neurol. 2008 Nov;50(11):854-8.

Effectiveness of sedation using nitrous oxide compared with enteral midazolam for botulinum toxin A injections in children.

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This randomized, double-blind, placebo-controlled study compared the efficacy of inhaled nitrous oxide (N₂O) with enteral midazolam for sedation of children with cerebral palsy (CP) undergoing botulinum

toxin A (BoNT-A) injections. Fifty children (29 males, 21 females; mean age 8y 2mo [SD 4y 5mo]; range 1-16y) were randomized to sedation with N(2)O (n=25) or midazolam (n=25). Groups were similar in type of CP (diplegia, 11; triplegia, three; quadriplegia, 16; hemiplegia, 16; other, three) and Gross Motor Function Classification System level (Level I, 4; II, 24; III, 4; IV, 13; V, 5). Both groups were equally sedated at time of injection (p=0.661), but those in the midazolam group were more sedated at time of discharge (p<0.001). N(2)O was more effective in reducing pain compared with midazolam as measured using the Face, Legs, Activity, Cry, Consolability (FLACC) scale (p=0.010), parental estimate of pain (p=0.009), and nursing estimate of pain (p=0.007). Parents in the N(2)O group rated it better than prior sedation with midazolam for BoNT-A injections (p=0.031). Physicians and nurses reported no difference in ease of procedure between the groups. One child in the midazolam group and eight in the N(2)O group had adverse effects, all of which resolved promptly. N(2)O appears to be an effective means of sedation for children undergoing outpatient BoNT-A injections.

PMID: 19046178 [PubMed - in process]

22: Dev Med Child Neurol. 2008 Nov;50(11):822-7.

Group aquatic aerobic exercise for children with disabilities.

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The effectiveness and safety of a group aquatic aerobic exercise program on cardiorespiratory endurance for children with disabilities was examined using an A-B study design. Sixteen children (11 males, five females) age range 6 to 11 years (mean age 9y 7mo [SD 1y 4mo]) participated in this twice-per-week program lasting 14 weeks. The children's diagnoses included autism spectrum disorder, myelomeningocele, cerebral palsy, or other developmental disability. More than half of the children ambulated independently without aids. Children swam laps and participated in relay races and games with a focus of maintaining a defined target heart rate zone. The strengthening component consisted of exercises using bar bells, aquatic noodles, and water resistance. The following outcomes were measured: half-mile walk/run, isometric muscle strength, timed floor to stand 3-meter test, and motor skills. Complaints of pain or injury were systematically collected. Significant improvements in the half-mile walk/run were observed, but not for secondary outcomes of strength or motor skills. The mean program attendance was 80%, and no injury was reported. Children with disabilities may improve their cardiorespiratory endurance after a group aquatic aerobic exercise program with a high adult:child ratio and specific goals to maintain training heart rates.

Publication Types:
Research Support, Non-U.S. Gov't

PMID: 19046177 [PubMed - in process]

23: No To Hattatsu. 2008 Nov;40(6):456-9.

Videofiberscopic study of laryngeal abnormalities in patients with severe motor and intellectual disabilities [Article in Japanese]

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A total of 43 patients with severe motor and intellectual disabilities were enrolled in videofiberscopic investigation of laryngeal abnormalities. Their ages ranged from 2 to 41 years (mean: 19.9 years) and all

of them had severe quadriplegia due to cerebral palsy or other conditions. Thirty-two out of the 43 patients (74.4%) had laryngeal abnormalities; laryngeal stenosis including that of iatrogenic origin, hypersecretion and laryngomalacia. Direct visualization of the occurrence of aspiration could be achieved in some patients. Seven (87.5%) out of 8 patients with tracheostomy and 25 (71.4%) out of 35 patients without tracheostomy had laryngeal abnormalities, showing no significant difference. The prevalence of abnormalities was significantly higher in patients with parenteral nutrition (30/33 = 90.9%) than in those with oral feeding (2/10 = 20%). Endoscopic investigation of the larynx is important, and videofiberscopy is especially effective for evaluation of laryngeal abnormalities in severely handicapped patients.

Publication Types:
English Abstract

PMID: 19039986 [PubMed - in process]

24: Zhongguo Zhen Jiu. 2008 Nov;28(11):798-800.

Observation on therapeutic effect of comprehensive therapy on optic atrophy complicated by cerebral palsy [Article in Chinese]

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OBJECTIVE: To assess therapeutic effect of combined treatment of Chinese medicine and western medicine on optic atrophy complicated by cerebral palsy. **METHODS:** One hundred and seventeen cases were divided into an observation group (n = 79) and a control group (n = 38). The control group were treated with routine western medicine treatment including neurotrophic drugs and high pressure oxygen, etc. and the observation group with acupuncture at Ganshu (BL 18), Pishu (BL 20), Chengqi (ST 1), etc. and injection of 0.2-0.3 mL Compound Danshen Injectio into Qiuhou (EX-HN 7), on the basis of the same treatment of western medicine as that in the control group. Fundus examination and the tracing body angle detection were conducted before and after treatment and the therapeutic effects were assessed in the two groups. **RESULTS:** The total effective rate was 91.1% in the observation group and 60.5% in the control group with a significant difference between the two groups (P < 0.001); after treatment the angle of tracing body significantly increased in the two groups (P < 0.01) with the observation group better than the control group (P < 0.01). **CONCLUSION:** The combined treatment of Chinese medicine and western medicine is an effective therapy for optic atrophy complicated by cerebral palsy.

Publication Types:
English Abstract

PMID: 19055282 [PubMed - in process]

25: Acta Orthop Belg. 2008 Oct;74(5):609-14.

Hip subluxation and dislocation in cerebral palsy: outcome of bone surgery in 21 hips.

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The prevalence of hip subluxation and dislocation in cerebral palsy ranges between 3% and 75% in the literature. Clinical signs are rigidity, pain and instability. We assessed functionality, stability and symptoms in 20 patients preoperatively and after follow-up. A varus derotation osteotomy was performed in cases with subluxation or dislocation, while a Chiari osteotomy was performed in the presence of a concomitant acetabular dysplasia. Results were good in 64%, fair in 22% and poor in 14% of patients

treated with a varus derotation osteotomy. In patients treated with a Chiari osteotomy, results were good in 43% of cases, fair in 43% and poor in 14%. Subluxated or dislocated hips generally show several anomalies: their severity is directly proportional to the degree of neurological impairment. In the most severe cases, correction of just one of such anomalies might not be sufficient to guarantee good results of the surgery.

PMID: 19058693 [PubMed - in process]

26: Disabil Rehabil. 2008;30(25):1910-9.

Postnatal cerebral infection leading to hemiplegic cerebral palsy: Functional limitations and disability of 13 children in Sweden.

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Purpose. To describe the motor function and disability of children with postnatal cerebral infection leading to hemiplegia, and to determine the severity of their motor disability. **Method.** The disablement process was used to describe these children. Participants included 13 children (7 girls and 6 boys, mean age 9.8 years). Traditional neurological assessment methods, together with the Movement Assessment Battery for Children and Paediatric Evaluation of Disability Inventory, were used. The children's non-hemiplegic sides were also tested. To determine severity of motor disability, new definitions were created that reflected the child's balance and fine-motor ability in relation to expected performance at his or her age. **Results.** The children had involvement of the non-hemiplegic leg and arm in all but one case. Balance and hand function was impaired in all. Hypotonicity was present in 10 children, weakness of hip muscles in 9. There was great variation in grip function. Motor difficulties dominated during pre-school years, whereas the dominant problem of all school-age children was social and communication skills. Behavioural, communicative, and chewing and swallowing problems were common. **Conclusions.** A postnatal infectious aetiology influences function on both the hemiplegic and non-hemiplegic side. In school-age children, social skills and communication difficulties are more pronounced than motor problems. The results of this study encourage the use of methods standardized for age where force dynamics and quality of movement are assessed.

PMID: 19061117 [PubMed - in process]

27: Phys Occup Ther Pediatr. 2008;28(4):309-25.

Effects of strength training aided by electrical stimulation on wrist muscle characteristics and hand function of children with hemiplegic cerebral palsy.

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Nine children with spastic hemiplegic cerebral palsy underwent 24 sessions of wrist muscles strengthening in the extended wrist range aided by electrostimulation. Isometric strength of flexors and extensors was registered in three wrist positions (30 degrees of flexion, neutral, and 30 degrees of extension) to infer on angle-torque curves. Passive stiffness of wrist flexors and wrist flexion angle during manual tasks and hand function were also documented. Significant strength gains were observed at 30 degrees of wrist extension for flexors ($p=0.029$) and extensors ($p=0.024$). No gains were observed at 30 degrees of flexion. The difference in extensor strength between the three test positions changed after intervention ($p<0.034$), suggesting a shift in the angle-torque curve. No changes were observed in passive stiffness ($p=0.506$), wrist angle ($p<0.586$), or hand function ($p=0.525$). Strength training in specific joint ranges may alter angle-torque relationships. For functional gains to be observed, however, a more ag-

gressive intervention and contextualized task training would probably be needed.

Publication Types:
Research Support, Non-U.S. Gov't

PMID: 19042474 [PubMed - in process]

28: Phys Occup Ther Pediatr. 2008;28(4):291-304; discussion 305-7.

Is waiting for rehabilitation services associated with changes in function and quality of life in children with physical disabilities?

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OBJECTIVES: To determine whether longer waiting times for rehabilitation were associated with deterioration in child functional status and/or quality of life. **METHODS:** Parents of 124 children (mean age 45 months) with physical disabilities (e.g., cerebral palsy, global developmental delay, spina bifida) who were referred to pediatric rehabilitation centers were interviewed every three months, from referral until admission into the rehabilitation program. Information from parental interviews included socio-demographics, parental empowerment (Family Empowerment Scale), function (WeeFIM: Functional Independence Measure for Children), and quality of life (PedsQL). Data on date of referral, age, gender, and diagnosis were obtained from hospital databases. **RESULTS:** Half of the sample waited more than 9.1 months (95% confidence interval: 6.5-16.1) for admission to a public rehabilitation program. A total of 42% paid for some private services while waiting. Over the waiting period, there was statistically significant improvement in WeeFIM cognition and total scores but not in mobility scores. PedsQL psychosocial summary score declined over the waiting period ($p < .05$). **CONCLUSION:** Longer wait times for rehabilitation were associated with declining psychosocial quality of life. Reducing waiting times for rehabilitation services may allow rehabilitation specialists to address psychosocial problems for these children.

Publication Types:
Research Support, Non-U.S. Gov't

PMID: 19042472 [PubMed - in process]

29: Paediatr Child Health. 2007 Dec;12(10):859-65.

Loving your child to death: Considerations of the care of chronically ill children and euthanasia in Emil Sher's Mourning Dove.

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How do parents cope when their child is ill or dying, and when he or she is experiencing constant pain or suffering? What do parents think of the contributions that medical professionals make to the care of their chronically or terminally ill child? Is it possible for a parent to love a child so much that they wish their child to be dead? The purpose of the present paper is to explore these questions and aspects of the care of chronically or terminally ill children using Mourning Dove's portrayal of one family's attempt to care for their ill daughter. Mourning Dove, a play written by Canadian playwright Emil Sher, was inspired by the case of Saskatchewan wheat farmer Robert Latimer who killed his 12-year-old daughter, Tracy, who suffered with cerebral palsy and had begun to experience tremendous pain. Rather than focusing on the medical or legal aspects of the care of a chronically ill child, the play offers a glimpse into how a family copes with the care of such a child and the effect the child's illness has on the family. The reading

and examination of nonmedical literature, such as Mourning Dove, serves as a useful means for medical professionals to better understand how illness affects and is responded to by patients and their families. This understanding is a prerequisite for them to be able to provide complete care of children with chronic or terminal illnesses and their families.

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PMCID: PMC2532575

30: Paediatr Child Health. 2007 Dec;12(10):853-8.

A randomized trial of aggressive versus conservative phototherapy for hyperbilirubinemia in infants weighing less than 1500 g: Short- and long-term outcomes.

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OBJECTIVE: Treatment regimens for hyperbilirubinemia vary for very low birth weight infants. The present study seeks to determine whether the initiation of conservative phototherapy is as effective as aggressive phototherapy in reducing peak bilirubin levels without increasing adverse effects. **STUDY DESIGN:** The present randomized, controlled study included infants with birth weights between 500 g and 1500 g, stratified into two birth weight groups. In one group, aggressive phototherapy was commenced by 12 h of age, while in the other group, conservative phototherapy was commenced if serum bilirubin levels exceeded 150 $\mu\text{mol/L}$. The primary outcome variables were peak serum bilirubin levels and hours of phototherapy. Secondary outcomes were age at peak bilirubin levels, number of infants with rebound hyperbilirubinemia, and number of adverse short- and long-term outcomes. **RESULTS:** Of 174 eligible infants, 95 consented to participate -49 in the conservative arm and 46 in the aggressive arm. Ninety-two infants completed the study. There was no significant difference in peak bilirubin levels except in infants who weighed less than 1000 g -171.2 \pm 26 $\mu\text{mol/L}$ (conservative) versus 139.2 \pm 46 $\mu\text{mol/L}$ (aggressive); $P < 0.02$. There was no difference in duration of phototherapy or rebound hyperbilirubinemia. There were no differences in short-term adverse outcomes. Of the 87 infants who survived until hospital discharge, 82 (94%) had some follow-up and 75 (86%) attended follow-up until 18 months corrected age. The incidence of cerebral palsy, abnormal mental developmental index at 18 months corrected age, or combined outcome of cerebral palsy and death did not significantly differ between the two groups. **CONCLUSIONS:** In infants weighing less than 1000 g, peak bilirubin levels were significantly higher using conservative phototherapy regimens and there was a tendency for poor neurodevelopmental outcome.

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