This free weekly bulletin lists the latest research on cerebral palsy (CP), as indexed in the NCBI, PubMed (Medline) and Entrez (GenBank) databases. These articles were identified by a search using the key term "cerebral palsy".

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Interventions


Use of intrathecal baclofen therapy in ambulant children and adolescents with spasticity and dystonia of cerebral origin: a systematic review.

Pin TW, McCartney L, Lewis J, Waugh MC.

Source: Kids Rehab, The Children's Hospital at Westmead, Westmead, NSW, Australia.

Aim: Studies on the use of intrathecal baclofen (ITB) for ambulant adults with spasticity and/or dystonia of cerebral origin are scarce, and are even more limited for children and adolescents. This systematic review investigates the use of ITB to improve walking, transfer ability, and gross motor activities in ambulant children and adolescents with spasticity and/or dystonia of cerebral origin. Method: Electronic databases (MEDLINE, CINAHL, PsycINFO, EMBASE, full Cochrane Library, and PEDro) were searched from the earliest date available until March 2011 using combined subject headings and free text if supported by the databases. Studies were included if they had examined individuals who: (1) received ITB therapy by any method (bolus injection, an external delivery system, or an implanted pump); (2) had spasticity and/or dystonia of cerebral origin; (3) were able to ambulate with or without a walking device, i.e. individuals with cerebral palsy (CP) who were in levels I to III of the Gross Motor Function Classification System or individuals with similar functional mobility if they did not have CP; and (4) were aged 18 years or under. Publications in English in peer-reviewed journals reporting any type of research design, except reviews and expert opinions, were included. Studies were excluded if participants had spasticity and/or dystonia of spinal origin and if baclofen was administered only orally. Studies that compared ITB with other interventions such as botulinum toxin were also excluded. Results: Two independent reviewers scored 16 studies against the guidelines for developing systematic reviews from the American Academy of Cerebral Palsy and Developmental Medicine (AACPDM). Interpretation: Fifteen studies were of levels IV or V evidence and only one of level II according to the evidence levels of the AACPDM guidelines, but all were of low quality. No study was found on the use of ITB in ambulant children or adolescents with dystonia of cerebral origin. Not all studies used objective outcome measures to assess the ambulation, transfer ability, and gross motor activities of the participants. A proportion of participants showed improvement in all these areas but adverse events were common. A proportion of participants compromised their ambulatory and transfer abilities after ITB. There was no evidence to support the clinical use of ITB in ambulant individuals with hypertonicity without further rigorous longitudinal studies.


PMID: 21635230 [PubMed - as supplied by publisher]


Source: Department of Rehabilitation Medicine, VU University Medical Center, Amsterdam, The Netherlands.

Aim: Treatment options for dystonic cerebral palsy (CP) are limited. Our aims were to determine whether intrathecal baclofen (ITB) improves daily care, decreases dystonia and decreases pain in patients with dystonic CP. Methods: Patients received randomized blinded treatment with ITB or placebo. Scores on problems of daily care were recorded and dystonia, pain and comfort were assessed. Results: Four patients (three males, average age 12 years 6 months) were included (all Gross Motor Function Classification System level V). During the trial period problem scores and dystonia scores decreased in all four patients. Conclusion: In this pilot study we report positive functional effects of ITB trial treatment in four patients with dystonic CP. A randomized trial with a larger cohort is needed to verify these results.

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Sit-to-stand movement in children with cerebral palsy: A critical review.

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Source: Department of Physiotherapy, Neuropediatrics Section, Universidade Federal de São Carlos, Rod. Washington Luis, km 235, 13565-905, São Carlos, SP, Brazil.

Sit-to-stand (STS) movement is widely performed in daily life and an important pre requisite for acquisition of functional abilities. However, STS is a biomechanical demanding task which requires high levels of neuromuscular coordination, muscle strength and postural control. As children with cerebral palsy (CP) exhibit a series of impairments in body structures and functions, STS movement performance could be impaired in this population. Thus, this article aimed to review studies that had described how STS movement is performed by children with CP, the factors that influence it and the methodological procedures adopted in it analyses. A search was performed by one reviewer in relevant databases. In all, 12 articles were identified and 9 were selected for the present review. It was detected a large variation in sample characteristics and methodological issues among studies. In fact, standardization of the method applied to STS movement analysis is not fully established. With regard to STS performance, children with CP exhibited variations among them and also when compared with their typical peers. Moreover extrinsic factors appear to influence STS movement performance in these children and its manipulation could be incorporated into rehabilitation protocols. Moreover, the relationship between STS movement and functionality in reviewed articles was not reported. Therefore the review allowed to observe that STS movement has been under-explored in children with CP, with a lack of standardized methodologies and a not well established relationship between this movement and functionality. Thus, further studies about STS movement in CP are necessary.

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Severe scoliosis in neurodevelopmental disabilities: clinical signs and therapeutic proposals.

Ferrari A, Ferrara C, Balugani M, Sassi S.

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Scoliosis is an important cause of disability in childhood, due to its incidence and limitation on ability. In neurodevelopmental disabilities, scoliosis worsens the already limited functional capacities of the child and can thwart abilities partially recovered through rehabilitation. In cerebral palsied children (CP), scoliosis is considered a consequence of damage to the central nervous system or a complication of a peripheral impairment, in particular, through obliquity of pelvis, of the unilateral dislocation of hip. In order to explain the scoliosis of central origin, primitive and pathological reflexes, reactions or automatisms must be taken into account, especially the asymmetrical ones. This contradicts the absence of scoliosis in hemiplegia. On the contrary, symmetrical patterns should act as protective factors. However, the greater incidence of scoliosis in tetraplegia than in diplegia does not confirm this idea. Scoliosis is secondary when it is linked to an unilateral hip dislocation for side of convexity, proximity, measure and timing sequence. In childhood neuromuscular diseases (NMD), scoliosis is the unavoidable consequence of muscle weakness. The only protective factor may be muscle stiffness in case of fibrotic degeneration. The main curve is reducible for longer periods, while the less reducible secondary one at cervical level can limit the correction possibilities, due to the need to keep the head aliened. In spina bifida (SB) children, lesional and supraspinal scoliosis can be present. In the former, the malformative ones directly derive from the vertebrae involved in the myelomeningocele and the secondary ones develop from the unilateral hip dislocation similar to CP, or from muscular imbalance (a typical feature of SB, especially for lower lumbar levels) or from primitive skeletal malformations of pelvic girdle or lower limbs. The acquired ones are caused by muscle weakness as in NMD. In the supraspinal scoliosis, the curve is the consequence of an impairment in neurological structures especially of ponto-cerebellar carrefour or of an acquired tethered cord and may affect any segment of spinal column, also above the primitive lesion level. The scoliosis in neurodevelopmental disabilities can be treated conservatively with corsets and postural systems, with limited results. or through a surgical approach, often made difficult due to the multiple impairments present in the same patient.

PMID: 21224789 [PubMed - indexed for MEDLINE]


The use of botulinum toxin A in children with cerebral palsy, with a focus on the lower limb.
Molenaers G, Van Campenhout A, Fagard K, De Cat J, Desloovere K.

PURPOSE: The purpose of this review is to clarify the role of botulinum toxin serotype A (BTX-A) in the treatment of children with cerebral palsy (CP), with a special focus on the lower limb. BACKGROUND: The treatment of spasticity is central in the clinical management of children with CP. BTX-A blocks the release of acetylcholine at the motor end plate, causing a temporary muscular denervation and, in an indirect way, a reduced spasticity. Children with increased tone develop secondary problems over time, such as muscle contractures and bony deformities, which impair their function and which need orthopaedic surgery. However in these younger children, delaying surgery is crucial because the results of early surgical interventions are less predictable and have a higher risk of failure and relapse. As BTX-A treatment reduces tone in a selective way, it allows a better motor control and muscle balance across joints, resulting in an improved range of motion and potential to strengthen antagonist muscles, when started at a young age. The effects are even more obvious when the correct BTX-A application is combined with other conservative therapies, such as physiotherapy, orthotic management and casts. There is now clear evidence that the consequences of persistent increased muscle tone can be limited by applying an integrated multi-level BTX-A treatment approach. Nevertheless, important challenges such as patient selection, defining appropriate individual goals, timing, dosing and dilution, accuracy of injection technique and how to measure outcomes will be questioned. Therefore, “reflection is more important than injection” remains an actual statement.

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Skeletal muscle stiffness and contracture in children with spastic cerebral palsy.
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Sensory information utilization and time delays characterize motor developmental pathology in infant sitting postural control.

Deffeyes JE, Harbourne RT, Stuberg WA, Stergiou N.

Source: Munroe-Meyer Institute, University of Nebraska Medical Center, Omaha, NE.

Sitting is one of the first developmental milestones that an infant achieves. Thus measurements of sitting posture present an opportunity to assess sensorimotor development at a young age. Sitting postural sway data were collected using a force plate, and the data were used to train a neural network controller of a model of sitting posture. The trained networks were then probed for sensitivity to position, velocity, and acceleration information at various time delays. Infants with typical development developed a higher reliance on velocity information in control in the anterior-posterior axis, and used more types of information in control in the medial-lateral axis. Infants with delayed development, where the developmental delay was due to cerebral palsy for most of the infants in the study, did not develop this reliance on velocity information, and had less reliance on short latency control mechanisms compared with infants with typical development.

PMID: 21628731 [PubMed - in process]


Delayed sternal closure after vacuum-assisted closure therapy for tracheo-innominate artery fistula repair.


Source: Department of Surgery and Clinical Science, University Graduate School of Medicine, Yamaguchi, Japan.

We report a case of successful innominate artery resection with delayed sternal closure after vacuum-assisted closure (VAC) therapy for a tracheo-innominate artery fistula (TIF). A 42-year-old woman with cerebral palsy underwent tracheostomy for respiratory assistance. On postoperative day 14, she was transferred to our hospital after an episode of massive hemoptysis. TIF was diagnosed based on the findings of multidetector computed tomography. Thus, we resected the innominate artery and started VAC therapy to control the postoperative local infection. The patient recovered uneventfully, without any infectious sequelae. Our strategy, which includes VAC therapy, for TIF repair may eliminate postoperative infective problems that could induce sequential bleeding and sternal compromise. To our knowledge, this is the first report of using VAC therapy for TIF. Keywords: Tracheo-innominate artery fistula; Tracheostomy; Vacuum-assisted closure therapy.

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Argininemia presenting with progressive spastic diplegia.

Lee BH, Jin HY, Kim GH, Choi JH, Yoo HW.

Source: Department of Pediatrics, Asan Medical Center Children's Hospital, University of Ulsan College of Medicine, Seoul, Korea.

Argininemia is caused by a deficiency of arginase 1, which catalyzes the final step in the urea cycle, i.e., the cytosolic hydrolysis of arginine to ornithine and urea. In contrast to other urea cycle disorders, hyperammonemic encephalopathy is rarely observed in patients with argininemia. Rather, most exhibit an insidious onset and progression of neurologic manifestations, including spastic diplegia. We describe the first Korean patient with argininemia, manifesting as slowly progressive spastic diplegia. Our patient carries c.[32T>C]+[913G>A] (p.[Ile11Thr]+[Gly305Arg])]
mutations in the ARG1 gene. The latter mutation was not previously reported. Although arginemia is a very rare disease, it is recognized as a pan-ethnic disorder. We conclude that arginemia should be considered more frequently in the differential diagnosis of a patient with slowly progressive neurologic manifestations, especially progressive spastic diplegia, even in a population where arginemia was previously unknown.

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Perceptions of Pediatric Physical Therapists and Physical Educators on Classifying Learning Styles of Children and Adolescents With Cerebral Palsy.

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Source: 1Centre of Excellence for Rehabilitation Medicine Utrecht, Rehabilitation Centre De Hoogstraat, Utrecht, The Netherlands.

The purpose of this study was to examine professionals' perceptions on classifying learning styles in the context of teaching motor activities to children and adolescents with cerebral palsy (CP). The participants were 21 pediatric physical therapists (PPTs) and seven physical educators (PEs) in three schools for special education in The Netherlands. All participants were introduced to the key descriptions of two existing learning style instruments (Kolb's Learning Style Inventory and Myers-Briggs Type Indicator), applied them to children and adolescents with CP, and reported their perceptions in written surveys. This study had a mixed-methods design. Quantitative and qualitative data analyses showed that PPTs and PEs are mostly positive about the idea of classifying learning styles in the context of teaching motor activities to children and adolescents with CP, giving three main reasons: individual approach, professional communication, and treatment awareness. Additionally, qualitative data analysis showed that the key descriptions of the two learning style instruments were not feasible as classifications for children and adolescents with CP. It is therefore recommended that other learning style classification instruments should be explored and that possibly a new learning style classification instrument should be developed in the context of teaching motor activities to children and adolescents with CP.

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Epidemiology / Aetiology / Diagnosis & Early Treatment


Classification of topographical pattern of spasticity in cerebral palsy: A registry perspective.

Reid SM, Carlin JB, Reddihough DS.

Source: Department of Paediatrics, University of Melbourne, Parkville, Victoria 3052, Australia; Developmental Disability Research, Murdoch Childrens Research Institute, Flemington Road, Parkville 3052, Australia; Developmental Medicine, Royal Children's Hospital, Flemington Road, Parkville 3052, Australia.

This study used data from a population-based cerebral palsy (CP) registry and systematic review to assess the amount of heterogeneity between registries in topographical patterns when dichotomised into unilateral (USCP) and bilateral spastic CP (BSCP), and whether the terms diplegia and quadriplegia provide useful additional epidemiological information. From the Victorian CP Register, 2956 individuals (1658 males, 1298 females), born 1970-2003, with spastic CP were identified. The proportions with each topographical pattern were analysed overall and by gestational age. Binary logistic regression analysis was used to assess temporal trends. For the review, data were systematically collected on topographical patterns from 27 registries. Estimates of heterogeneity were obtained, overall and by region, reporting period and definition of quadriplegia. Among individuals born <32 weeks, 48% had diplegia, whereas the proportion for children born ≥32 weeks was 24% (p<0.001). Evidence was weak for a temporal
trend in the relative proportions of USCP and BSCP (p=0.038), but much clearer for an increase in the proportion of spastic diplegia relative to quadriplegia (p<0.001). The review revealed wide variations across studies in the proportion of diplegia (range 34-90%) and BSCP (range 51-86%). These findings argue against a topographical classification based solely on laterality.

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Adaptor Protein Complex 4 Deficiency Causes Severe Autosomal-Recessive Intellectual Disability, Progressive Spastic Paraplegia, Shy Character, and Short Stature.


Source: Institute of Human Genetics, University of Erlangen, D-91054 Erlangen, Germany.

Intellectual disability inherited in an autosomal-recessive fashion represents an important fraction of severe cognitive-dysfunction disorders. Yet, the extreme heterogeneity of these conditions markedly hampers gene identification. Here, we report on eight affected individuals who were from three consanguineous families and presented with severe intellectual disability, absent speech, shy character, stereotypic laughter, muscular hypotonia that progressed to spastic paraplegia, microcephaly, foot deformity, decreased muscle mass of the lower limbs, inability to walk, and growth retardation. Using a combination of autozygosity mapping and either Sanger sequencing of candidate genes or next-generation exome sequencing, we identified one mutation in each of three genes encoding adaptor protein complex 4 (AP4) subunits: a nonsense mutation in AP4S1 (NM_007077.3: c.124C>T, p.Arg42(*)), a frameshift mutation in AP4B1 (NM_006594.2: c.487_488insTAT, p.Glu163_Ser739delinsVal), and a splice mutation in AP4E1 (NM_007347.3: c.542+1_542+4delGTAA, r.421_542del, p.Glu181Glyfs(*20)). Adaptor protein complexes (AP1-4) are ubiquitously expressed, evolutionarily conserved heterotetrameric complexes that mediate different types of vesicle formation and the selection of cargo molecules for inclusion into these vesicles. Interestingly, two mutations affecting AP4M1 and AP4E1 have recently been found to cause cerebral palsy associated with severe intellectual disability. Combined with previous observations, these results support the hypothesis that AP4-complex-mediated trafficking plays a crucial role in brain development and functioning and demonstrate the existence of a clinically recognizable syndrome due to deficiency of the AP4 complex.

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Therapeutic hypothermia for management of neonatal asphyxia: what nurses need to know.

Chirinian N, Mann N.

Birth asphyxia can induce a cascade of reactions that result in altered brain function known as hypoxic-ischemic encephalopathy. Possible outcomes for survivors of birth asphyxia vary widely, from a normal outcome to death, with a wide range of disabilities in between, including long-term neurodevelopmental disability, cerebral palsy, neuromotor delay, and developmental delay. Treatment of hypoxic-ischemic encephalopathy has centered on dampening or blocking the biochemical pathways that lead to death of neuronal cells. The reduction of body temperature by 3ºC to 5ºC less than normal body temperature can reduce cerebral injury. At Mount Sinai Hospital in Toronto, Ontario, the goal of therapeutic hypothermia is to achieve a rectal temperature of 33ºC to 34ºC, and the protocol is started within 6 hours after birth. The hypothermia is maintained for 72 hours, and then the infant is gradually warmed to normal body temperature (36.8ºC-37ºC). The protocol and nursing implications are presented.

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Exogenous glucocorticoids and adverse cerebral effects in children.

Damsted SK, Born AP, Paulson OB, Uldall P.

Source: Department of Paediatrics, Copenhagen University Hospital, Rigshospitalet, Juliane Marie Center, Blegdamsvej 9, DK-2100 Copenhagen, Denmark.

Glucocorticoids are commonly used in treatment of paediatric diseases, but evidence of associated adverse cerebral effects is accumulating. The various pharmacokinetic profiles of the exogenous glucocorticoids and the changes in pharmacodynamics during childhood, result in different exposure of nervous tissue to exogenous glucocorticoids. Glucocorticoids activate two types of intracellular receptors, the mineralocorticoid receptor and the glucocorticoid receptor. The two receptors differ in cerebral distribution, affinity and effects. Exogenous glucocorticoids favor activation of the glucocorticoid receptor, which is associated with unfavorable cellular outcomes. Prenatal treatment with glucocorticoids can compromise brain growth and is associated with periventricular leukomalacia, attentions deficits and poorer cognitive performance. In the neonatal period exposure to glucocorticoids reduces neurogenesis and cerebral volume, impairs memory and increases the incidence of cerebral palsy. Cerebral effects of glucocorticoids in later childhood have been less thoroughly studied, but apparent brain atrophy, reduced size of limbic structures and neuropsychiatric symptoms have been reported. Glucocorticoids affect several cellular structures and functions, which may explain the observed adverse effects. Glucocorticoids can impair neuronal glucose uptake, decrease excitability, cause atrophy of dendrites, compromise development of myelin-producing oligodendrocytes and disturb important cellular structures involved in axonal transport, long-term potentiation and neuronal plasticity. Significant maturation of the brain continues throughout childhood and we hypothesize that exposure to exogenous glucocorticoids during preschool and school age causes adverse cerebral effects. It is our opinion that studies of associations between exposure to glucocorticoids during childhood and impaired neurodevelopment are highly relevant.

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Apgar scores and cerebral palsy.

Kent A.

Source: Department of Obstetrics & Gynaecology, University of Cape Town Rondebosch, South Africa.

PMID: 21629499 [PubMed - in process]


Myth: Cerebral palsy cannot be predicted by neonatal brain imaging.

de Vries LS, van Haastert IC, Benders MJ, Groenendaal F.

Source: Department of Neonatology, Room KE 04.123.1, Wilhelmina Children's Hospital/University Medical Centre Utrecht, Lundlaan 6, 3584 EA Utrecht, The Netherlands.

There is controversy in the literature about the value of brain imaging in neonates regarding the prediction of cerebral palsy (CP). The aim of this review was to unravel the myth that CP cannot be predicted by neuroimaging in neonates. Major intracranial lesions in the preterm infant should be recognized with sequential cranial ultrasound and will predict those with non-ambulatory CP. Magnetic resonance imaging (MRI) at term-equivalent age will refine the prediction by assessment of myelination of the posterior limb of the internal capsule. Prediction of motor outcome in preterm infants with subtle white matter injury remains difficult, even with conventional MRI. MRI is a better
tool to predict outcome in the term infant with hypoxic-ischaemic encephalopathy or neonatal stroke. The use of diffusion-weighted imaging as an additional sequence adds to the predictive value for motor outcome. Sequential and dedicated neuroimaging should enable us to predict motor outcome in high risk newborns infants.

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