
Locomotor tests predict community mobility in children and youth with cerebral palsy.

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Ambulatory children and youth with cerebral palsy have limitations in locomotor capacities and in community mobility. The ability of three locomotor tests to predict community mobility in this population (N = 49, 27 boys, 6-16 years old) was examined. The tests were a level ground walking test, the 6-min-Walk-Test (6MWT), and two tests of advanced locomotor capacities, the 10-meter-Shuttle-Run-Test (10mSRT) and the Timed-Up-and-Down-Stairs-Test (TUDS). Community mobility was measured with the Assessment of Life Habits mobility category. After age and height were controlled, regression analysis identified 10mSRT and TUDS values as significant predictors of community mobility. They explained about 40% of the variance in the Life Habits mobility category scores. The 10mSRT was the strongest predictor (standardized Beta coefficient = 0.48, p = 0.002). The 6MWT was not a significant predictor. Thus, advanced locomotor capacity tests may be better predictors of community mobility in this population than level ground walking tests.

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Quantification of patellar tendon shortening in a patient with cerebral palsy.

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Purpose: In the last few years the use of patellar tendon shortening procedure appears to give good results for the treatment of crouch gait in Cerebral Palsy (CP) patients. It offers a secondary approach in cases where conventional hamstrings lengthening failed. The evidence for the effectiveness of knee extensor shortening is poor and only one study quantitatively assessed the effects of this technique performed in addition to other procedures.
in CP patients. In this case study we used Gait Analysis (GA) to quantify the effects of patellar tendon shortening for the management of crouch gait persisting after hamstring lengthening. Methods: Crouch persisted seven years after a first multilevel procedure including hamstring lengthening in a 15-year old male. Gait was assessed over time (before and from six to 18 months after knee extensor shortening) with Gait Analysis (GA). Results: Before treatment the patient walked slowly and with a high duration stance phase; sagittal kinematics revealed excessive hip and knee flexion and ankle dorsiflexion during the whole gait cycle. After surgery, data showed progressive significant improvements at all lower limb levels during the follow-ups considered; gait became more physiologic and lower limb joint kinematics improved overall. Conclusions: The results demonstrated that addressing the other side and shortening the antagonists may be a valuable option to treat crouch gait in patients with CP especially if this persists after hamstring and knee extension surgery.

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Beneficial Functional Outcomes of Selective Dorsal Rhizotomy (SDR) Are Long Lasting and Alter the Natural History of Motor Development in Spastic Cerebral Palsy.


INTRODUCTION: In spastic cerebral palsy (CP), damage to the immature brain is non-progressive, but the clinical expression of the disorder changes over time along with the growth and development of the child. Large-scale, population based studies describing the natural history of gross motor development in spastic CP have indicated that most patients plateau in terms of motor development in childhood, and more severely affected children decline through adolescence. Selective dorsal rhizotomy (SPR) is a well-recognized treatment for children with spastic CP, and randomized control trials have revealed the benefits of this procedure, at least in the short term. However, no study has reported long-term follow-up outcomes on large numbers of patients. Thus, the impact of SDR on the natural history of CP remains unresolved. METHODS: Here we analyzed long-term follow-up data obtained from the McGill University Rhizotomy Database to assess the durability of functional outcomes after SDR. Children were evaluated by a multidisciplinary team preoperatively, and at 1 year, 5 years, 10 years, and 15 years postoperatively. Quantitative, standardized assessments of lower-limb spasticity, gross motor function, and activities of daily living were evaluated. In addition, we assessed the need for adjuvant orthopedic procedures for spasticity following SDR. RESULTS: Of 102 patients that had formal preoperative evaluations more than ten years earlier, 97, 62, 57, and 14 patients completed formal postoperative assessments at 1-year, 5-years, 10-years, and 15-years, respectively. We found statistically significant, durable improvements in spasticity, Gross Motor Function Measure (GMFM), activities of daily living up to 10 and 15 years after SDR (ie, though adolescence and into early adulthood). When children were stratified according to severity by the Gross Motor Function Classification System (GMFCS) we found long-lasting improvements through adolescence, which were not seen in the previously reported natural history curves for untreated patients. Furthermore, following SDR, our patients had half as many orthopedic procedures as reported in the literature for untreated patients. CONCLUSION: Therefore, we conclude that the benefits of SDR are long-lasting and positively alter the natural history of spastic CP.

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Does the level of proximal femur rotation osteotomy influence the correction results in patients with cerebral palsy?

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Proximal femur external rotation osteotomy is a common procedure used for the correction of increased femur anteversion and hip internal rotation in cerebral palsy (CP). Different levels of osteotomy have been used at the
proximal femur, but there are no studies in the literature comparing the results in CP. Patients with spastic CP, Gross Motor Function Classification System (GMFCS) I-III, who had undergone a femoral rotational osteotomy from August 1998 to August 2007, and with complete documentation at gait laboratory were included in the study. Patients were divided into two groups according to the level of osteotomy at the proximal femur. Group A [Dynamic Compression Plate (DCP) group] included 24 patients (36 osteotomies), and the osteotomy in this group was performed below the lesser trochanter. In Group B (Blade Plate group), 29 patients (35 osteotomies) were included and the level of osteotomy was above the lesser trochanter. Age at surgery, sex distribution, follow-up time, previous surgical procedures, surgical procedures performed in the same session as femur osteotomy, GMFCS level, topographic classification, clinical findings (internal and external hip rotation, and femur anteversion), and hip rotation at kinematics were analyzed and the results were compared between groups. Groups A and B were matched in terms of the sex distribution, follow-up time, GMFCS levels, and severity of clinical findings and hip internal rotation at kinematics before surgery. The mean age of the patients at surgery was 9.24 years in group A and 12 years in group B, and this difference was significant on performing statistical analysis (P=0.004). The number of patients who had undergone previous hip adductors’ tenotomy was higher in group B (P=0.036). Improvements in clinical and kinematics parameters were observed in both groups after femur osteotomy (P<0.001). The increase in hip external rotation at clinical examination and the reduction in hip internal rotation at kinematics did not show differences between groups A and B on performing statistical analysis. However, reduction of femoral anteversion (P=0.032) and hip internal rotation (P=0.002) were more remarkable in group B. In conclusion, reduction of hip internal rotation and femur anteversion at physical examination were more significant in patients with intertrochanteric osteotomies; however, improvement in kinematics was observed in both groups after surgical procedures.

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The Effect of Bilateral Globus Pallidus Internus Deep Brain Stimulation plus Ventralis Oralis Thalamotomy on Patients with Cerebral Palsy.

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Objective: We compared bilateral globus pallidus internus (GPI) deep brain stimulation (DBS) with bilateral GPI DBS plus ventralis oralis (Vo) thalamotomy to analyze the effect of the combined Vo thalamotomy. Methods: Between March 2003 and December 2008, 10 patients underwent DBS and/or Vo thalamotomy for treatment of cerebral palsy in our institute of neurosurgery and rehabilitation medicine. Four patients received bilateral posteroventral GPI DBS as group I and 6 patients received GPI DBS plus unilateral thalamotomy as group II. Results: The movement and disability scores of group I improved by 32 and 14.3%, respectively, at the last follow-up compared with baseline. The movement and disability scores of group II improved by 31.5 and 0.18%. The BFMDRS-movement subscores of group II demonstrated statistically significant improvement in the contralateral arm compared to group I (p = 0.042). Body pain, vitality and mental health seemed to improve in group II, in terms of health-related quality of life. Conclusions: Contrary to our expectations, we were unable to demonstrate clear improvements in overall BFMDRS scores between group I and group II. However, movements of the contralateral upper extremities improved and health-related quality of life in group II showed satisfactory results.

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'Remind to move': a pilot study on the effects of sensory cueing treatment on hemiplegic upper limb functions in children with unilateral cerebral palsy.

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Objective: To investigate the effects of sensory cueing with repetitive practice on hemiplegic arm functions in children with unilateral cerebral palsy. Design: A single-group pre/post comparison. Setting: A special school for children with physical disabilities. Subjects: Eight children with unilateral cerebral palsy aged 6-18 years. Interventions: Participants were required to wear for three weeks a sensory cueing, non-activated wristwatch device and complete 5 hours of conventional therapy per week (treatment A). This was followed by three weeks of continuing conventional therapy and wearing the now activated wristwatch, which prompted the children to do predetermined exercises on the hemiplegic arm for 6 hours daily, 5 days per week, for three weeks (treatment B), and three weeks follow-up. Main measures: Assessments of arm efficiency, functional hand use and arm impairments were carried out at baseline (day before treatment A), posttest 1 (day after treatment A), posttest 2 (day after treatment B) and follow-up (three weeks after treatment B). Results: Arm efficiency as revealed by the Jebsen-Taylor Hand Function Test and the Bruininks-Oseretsky Test of Motor Proficiency - but not actual arm use and grip strength - showed significant improvement after treatment B to follow-up (from 286.0 ± 73.9 to 191.9 ± 73.5, P = 0.002, and from 15.5 ± 5.0 to 18.1 ± 7.7, P = 0.021).Conclusion: This pilot study provides proof-of-concept data showing that a wearable device might be used to remind children with cerebral palsy to perform a set of predetermined arm exercises in order to promote hemiplegic arm function.

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Chronic pain, fatigue, and depressive symptoms in adults with spastic bilateral cerebral palsy.

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Aim: To investigate the prevalence and co-occurrence of chronic pain, fatigue, and depressive symptoms in adults with spastic bilateral cerebral palsy (SBCP) and explore associations of chronic pain and fatigue with depressive symptoms and daily functioning. Method: Fifty-six adults with SBCP without severe cognitive impairment participated (35 males, 21 females; mean age 36y 5mo, SD 5y 10mo; Gross Motor Function Classification System level I [13], II [28], III [11], IV [4]). Chronic pain (>3mo), severity and nature of fatigue (Fatigue Severity Scale; Multidimensional Fatigue Inventory), and depressive symptoms (Center for Epidemiological Studies Depression Scale) were assessed. Associations were explored using multivariable logistic regression analyses. Results: The study sample had a higher prevalence of chronic pain (75% vs 39%; p<0.001), mean fatigue (Fatigue Severity Scale, 4.4 [SD 1.3] vs 2.9 [SD 1.1]; p<0.001), and prevalence of depressive symptoms (25% vs 12%; p=0.004) than Dutch healthy reference samples. Chronic pain and severe fatigue co-occurred in 34% and in combination with depressive symptoms in 16% of the participants. Severity of fatigue was associated with depressive symptoms (OR 3.38; p<0.01). Chronic pain and fatigue were not associated with limitations in daily functioning. Interpretation: These findings suggest that adults with SBCP are severely affected by chronic pain, fatigue, and depressive symptoms, in addition to their spastic paresis.


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Unraveling the sources of chronic pain in cerebral palsy.

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Neurodevelopmental Outcomes Following Two Different Treatment Approaches (Early Ligation and Selective Ligation) for Patent Ductus Arteriosus.

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OBJECTIVE: To examine whether a change in the approach to managing persistent patent ductus arteriosus (PDA) from early ligation to selective ligation is associated with an increased risk of abnormal neurodevelopmental outcomes. STUDY DESIGN: In 2005, we changed our PDA treatment protocol for infants born at ≤27 6/7 weeks’ gestation from an early ligation approach, with prompt PDA ligation if the ductus failed to close after indomethacin therapy (period 1: January 1999 to December 2004), to a selective ligation approach, with PDA ligation performed only if specific criteria were met (period 2: January 2005 to May 2009). All infants in both periods received prophylactic indomethacin. Multivariate analysis was used to compare the odds of a composite abnormal neurodevelopmental outcome (Bayley Mental Developmental Index or Cognitive Score <70, cerebral palsy, blindness, and/or deafness) associated with each treatment approach at age 18-36 months (n = 224). RESULTS: During period 1, 23% of the infants in follow-up failed indomethacin treatment, and all underwent surgical ligation. During period 2, 30% of infants failed indomethacin, and 66% underwent ligation after meeting prespecified criteria. Infants treated with the selective ligation strategy demonstrated fewer abnormal outcomes than those treated with the early ligation approach (OR, 0.07; P = .046). Infants who underwent ligation before 10 days of age had an increased incidence of abnormal neurodevelopmental outcome. The significant difference in outcomes between the 2 PDA treatment strategies could be accounted in part for in the earlier age of ligation during period 1. CONCLUSION: A selective ligation approach for PDAs that fail to close with indomethacin therapy is not associated with worse neurodevelopmental outcomes at age 18-36 months.

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Cerebral palsy in children: Movements and postures during early infancy, dependent on preterm vs. full term birth.


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BACKGROUND: A deviant motor behaviour at age 3 to 5 months is predictive of cerebral palsy (CP). Particular features of the early motor repertoire even proved predictive of the degree of functional limitations as classified on the Gross Motor Function Classification System (GMFCS) in children with CP, born preterm. AIMS: We aimed to
determine whether an association between the early motor repertoire and the GMFCS also holds true for children born at term. STUDY DESIGN: Longitudinal study. SUBJECTS: 79 infants (60 boys and 19 girls; 47 infants born at term; video recorded for the assessment of movements and posture at age 9 to 20 weeks postterm age) who developed CP. OUTCOME MEASURES: The GMFCS was applied at age 2 to 5 years. RESULTS: Motor optimality at age 3 to 5 months showed a significant correlation with functional mobility and activity limitation as classified on the GMFCS at age 2 to 5 years in both children born at term (Spearman rho = -0.66, p < 0.001) and born preterm (rho = -0.37, p < 0.05). Infants born preterm were more likely to show normal movement patterns than infants born at term. A normal posture and an abnormal, jerky (yet not monotonous) movement character resulted in better levels of function and mobility. With the exception of one, none of the infants showed fidgety movements. A cramped-synchronised movement character, repetitive opening and closing of the mouth, and abnormal finger postures characterised children who would show a poor self-mobility later. CONCLUSIONS: Assessing the quality of motor performance at 9 to 20 weeks postterm age (irrespective of the gestational age) improves our ability to predict later functional limitations in children with CP.

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Interrater reliability study of cerebral palsy diagnosis, neurological subtype, and gross motor function.

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Aim: To evaluate the interrater reliability of the inclusion in registries and classification of children with cerebral palsy (CP). Method: Two studies were conducted. In study 1, 12 paediatricians from 11 countries viewed video sequences of 12 children with or without CP (nine males, three females; median age 6 y; range 2-16). In study 2, 19 professionals from eight countries participated in an online exercise. They had to classify the same children but based on written vignettes. All participants had to evaluate whether the child had CP, the neurological subtype (Surveillance of Cerebral Palsy in Europe classification system), and gross motor function level (Gross Motor Function Classification System [GMFCS]). Kappa (κ) coefficients were calculated for categorical variables and intraclass correlation coefficients (ICCs) for ordinal data. Results: Reliability was excellent in assessing whether or not a child had CP in study 1 (κ = 1.00) and substantial in study 2 (κ = 0.73); 95% confidence interval [CI] 0.58-0.87). For the neurological subtype, overall κ between paediatricians was 0.85 (95% CI 0.68-0.98), with full agreement observed for eight children. In study 2, overall κ was 0.78 (95% CI 0.61-0.91) with full agreement seen for five children. For the GMFCS, the ICC was 0.88 (95% CI 0.78-0.95) in study 1 and 0.80 (95% CI 0.64-0.91) in study 2. Interpretation: Reliability was excellent for all characteristics classified by paediatricians viewing the videos and substantial for professionals reading vignettes.


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Evidence that nuclear factor IA inhibits repair after white matter injury.

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OBJECTIVE: Chronic demyelination can result in axonopathy and is associated with human neurological conditions such as multiple sclerosis (MS) in adults and cerebral palsy in infants. In these disorders, myelin regeneration is inhibited by impaired differentiation of oligodendrocyte progenitors into myelin-producing oligodendrocytes. However, regulatory factors relevant in human myelin disorders and in myelin regeneration remain poorly understood. Here we have investigated the role of the transcription factor nuclear factor IA (NFIA) in oligodendrocyte progenitor differentiation during developmental and regenerative myelination. METHODS: NFIA expression patterns in human neonatal hypoxic-ischemic encephalopathy (HIE) and MS as well as developmental expression in mice were evaluated. Functional studies during remyelination were performed using a lysolecithin model, coupled with lentiviral misexpression of NFIA. The role of NFIA during oligodendrocyte lineage development was characterized using chick and mouse models and in vitro culture of oligodendrocyte progenitors. Biochemical mechanism of NFIA function was evaluated using chromatin immunoprecipitation and reporter assays. RESULTS: NFIA is expressed in oligodendrocyte progenitors, but not differentiated oligodendrocytes during mouse embryonic development. Examination of NFIA expression in white matter lesions of human newborns with neonatal HIE, as well active MS lesions in adults, revealed that it is similarly expressed in oligodendrocyte progenitors and not oligodendrocytes. Functional studies indicate that NFIA is sufficient to suppress oligodendrocyte progenitor differentiation during adult remyelination and embryonic development through direct repression of myelin gene expression. INTERPRETATION: These studies suggest that NFIA participates in the control of oligodendrocyte progenitor differentiation and may contribute to the inhibition of remyelination in human myelin disorders.

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Survival of individuals with cerebral palsy born in Victoria, Australia between 1970 and 2004 - Reid et al. reply.
Reid SM, Carlin JB, Reddihough DS.
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Improved survival in cerebral palsy in recent decades?
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Clinical features of abruptio placentae as a prominent cause of cerebral palsy.
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BACKGROUND: Although abruptio placentae causes hypoxia in the infant and thus leading to cerebral palsy (CP), its incidence and clinical features at a nationwide level have not been demonstrated. AIMS: To determine the proportion of abruptio placentae among antenatal and intrapartum causative factors leading to cerebral palsy (CP) and clinical features of such abruptio placentae. STUDY DESIGN: A review was conducted in 107 infants with CP
in whom CP was determined to be due to antenatal and or intrapartum hypoxic conditions by the Japan Council for Quality Health Care until April 2012. RESULTS: Abruptio placenta was responsible for 28 (26%) of the 107 CP infants, and was the single leading causative factor of CP. Of these 28 women, 22 (79%) exhibited non-reassuring fetal status on admission to obstetric facilities at 36.2±2.6 weeks of gestation and had neonates with umbilical cord arterial blood pH (base excess) of 6.728±0.164 (-25±5.4mmol/L). In these 22 women, strong abdominal pain and/or profuse vaginal bleeding occurred 159±99min prior to admission to an obstetric facility, and the interval until delivery after admission was 47±31min. Hypertension or isolated proteinuria preceded clinical events in one (4.5%) and five (23%) of these 22 women, respectively. CONCLUSIONS: Abruptio placentae was responsible for CP in one quarter of all cases determined to be due to antenatal and/or intrapartum hypoxic conditions in Japan. New strategies to shorten the interval until admission to an obstetric facility after onset of symptoms are urgently needed.

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Preterm twin and triplet pregnancies are at increased risk for the development of cystic periventricular leukomalacia.

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BACKGROUND: An increased risk of cerebral palsy in multiples has been reported. AIMS: To determine the risk for the development of periventricular leukomalacia (PVL) of twin and triplet pregnancy. STUDY DESIGN: Retrospective single-centre study at a tertiary care university hospital. SUBJECTS: Infants ≤35 weeks gestational age born between 1988 and 2008. OUTCOME MEASURES: Risk of twin and triplet compared to singleton pregnancy regarding development of PVL in one offspring. RESULTS: Of 6195 infants 117 singletons and 39 multiples were diagnosed as having cystic PVL. Perinatal data did not differ as did not ultrasonographic findings and neurologic outcome. The relative risk (RR) of a twin pregnancy resulting in at least one infant with PVL when born prior to 36 weeks was 2.181 (CI 95% 1.474-3.228, p < .0001), and 6.793 (CI 95% 2.470-13.108, p < .0001) of a triplet pregnancy. In-vitro fertilisation was present in 3% of affected twins compared to 100% in triplets (p < .001).
CONCLUSION: We found an increased risk for PVL in preterm twin and triplet pregnancies.

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Influence of gestational age on death and neurodevelopmental outcome in premature infants with severe intracranial hemorrhage.

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Objective: To determine whether death and/or neurodevelopmental impairment (NDI) after severe intracranial hemorrhage (ICH; grade 3 or 4) differs by gestational age (GA) at birth in extremely low birth weight (ELBW) infants. Study Design: Demographic, perinatal and neonatal factors potentially contributing to NDI for ELBW infants (23 to 28 weeks gestation) were obtained retrospectively; outcome data came from the ELBW Follow-up Study. NDI was defined at 18 to 22 months corrected age as moderate/severe cerebral palsy, Bayley Scales of Infant Development II cognitive or motor score <70, and/or blindness or deafness. Characteristics of younger versus older infants with no versus severe ICH associated with death or NDI were compared. Generalized linear mixed models predicted death or NDI in each GA cohort. Result: Of the 6638 infants, 61.8% had no ICH and 13.6% had severe ICH; 39% of survivors had NDI. Risk-adjusted odds of death or NDI and death were higher in the lower GA group.
Lower GA increased the odds of death before 30 days for infants with severe ICH. Necrotizing enterocolitis (particularly surgical NEC), late onset infection, cystic periventricular leukomalacia and post-natal steroids contributed to mortality risk. NDI differed by GA in infants without ICH and grade 3, but not grade 4 ICH. Contributors to NDI in infants with severe ICH included male gender, surgical NEC and post-hemorrhagic hydrocephalus requiring a shunt. Conclusion: GA contributes to the risk of death in ELBW infants, but not NDI among survivors with severe ICH. Male gender, surgical NEC and need for a shunt add additional risk for NDI.

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A Systematic Review and Meta-Analysis of Long-Term Development of Early Term Infants.

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Background: Births between 37 0/7 and 38 6/7 weeks of gestation are newly defined as early term infants (ETIs), and are increasingly considered to be at higher risk of adverse outcomes than infants born at 39-41 weeks’ gestation. To date, the long-term development of ETIs has not been systematically reviewed. Objective: To assess the effect of being born early term on long-term developmental outcomes. Methods: The literature of MEDLINE, EMBASE, Cochrane Database of Systematic Reviews and Cochrane Central Register of Controlled Trials, and s from the Society of Pediatric Research were searched. If two or more studies regarding the same outcome were retrieved, a meta-analysis was conducted by RevMan 5. A sensitivity analysis was performed to assess the heterogeneity. Results: Eleven studies involving 4 categories of long-term development were included. Compared with infants born at 39-41 weeks’ gestation, ETIs had poorer outcomes in school performance, neurodevelopment, behavior and emotional status and long-term social outcomes. Meta-analyses showed that being born early term significantly increased the risk of cerebral palsy [relative risk, RR, 1.75 (95% confidence interval, CI, 1.32, 2.31)] and mathematical difficulties [RR 1.13 (95% CI 1.04, 1.21)]. The statistical test of heterogeneity for cerebral palsy was significant. Sensitivity analysis demonstrated that variations in follow-up periods were associated with heterogeneity. Conclusion: Emerging evidence suggests that ETIs are at risk of adverse long-term outcomes. Due to paucity and heterogeneity of the existing data, future research is needed to clarify the long-term risk of being born early term.

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