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Interventions and Management

1. Phys Occup Ther Pediatr. 2013 Jul 10. [Epub ahead of print]

Motor Training and Physical Activity Among Preschoolers with Cerebral Palsy: A Survey of Parents' Experiences.

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Objective: To describe motor training and physical activity among preschoolers with cerebral palsy (CP) in Norway, and assess associations between child, parent, and motor intervention characteristics, and parent-reported child benefits from interventions. Method: Survey of 360 parents and data from the Norwegian CP follow-up program. The response rate was 34%. Results: During the six months preceding the time of the survey, 75% of the children performed gross-motor training, 73% fine-motor training, 80% manual stretching, and 67% participated regularly in physical activities. The training was highly goal-directed, intensive, frequently incorporated in daily routines, and often with a high level of parental involvement. The use of goals was associated with higher parent-reported child benefits for all types of interventions. Moreover, the positive relationship, which was indicated between frequency of training, parent education, and parent-reported child benefits of gross-motor training, was not seen for fine-motor training. Conclusion: Parent-reported child benefits support goal-directed motor interventions, and the use of everyday activities to increase practice of motor skills.

[PMID: 23841711](#) [PubMed - as supplied by publisher]

2. Dev Med Child Neurol. 2013 Jul 9. doi: 10.1111/dmcn.12199. [Epub ahead of print]

Postural asymmetries in young adults with cerebral palsy.

Rodby-Bousquet E, Czuba T, Hägglund G, Westbom L.

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AIM: The purpose was to describe posture, ability to change position, and association between posture and contractures, hip dislocation, scoliosis, and pain in young adults with cerebral palsy (CP). METHODS: Cross-sectional data of 102 people (63 males, 39 females; age range 19-23y, median 21y) out of a total population with

CP was analysed in relation to Gross Motor Function Classification System (GMFCS) levels I (n=38), II (n=21), III (n=13), IV (n=10), and V (n=20). The CP subtypes were unilateral spastic (n=26), bilateral spastic (n=45), ataxic (n=12), and dyskinetic CP (n=19). The Postural Ability Scale was used to assess posture. The relationship between posture and joint range of motion, hip dislocation, scoliosis, and pain was analysed using logistic regression and Spearman's correlation. RESULTS: At GMFCS levels I to II, head and trunk asymmetries were most common; at GMFCS levels III to V postural asymmetries varied with position. The odds ratios (OR) for severe postural asymmetries were significantly higher for those with scoliosis (OR=33 sitting), limited hip extension (OR=39 supine), or limited knee extension (OR=37 standing). Postural asymmetries correlated to hip dislocations: supine (rs =0.48), sitting (rs =0.40), standing (rs =0.41), and inability to change position: supine (rs =0.60), sitting (rs =0.73), and standing (rs =0.64). CONCLUSIONS: Postural asymmetries were associated with scoliosis, hip dislocations, hip and knee contractures, and inability to change position.

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3. Journal of Children's Orthopaedics. 2013 June.

The effects of Botulinum toxin injection frequency on calf muscle growth in young children with spastic cerebral palsy: A 12 month prospective study.

Barber L, Hastings-Ison T, Baker R, Graham HK, Barrett R & Lichtwark G

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Purpose: This study was a 12-month prospective investigation of changes in the medial gastrocnemius (MG) muscle morphology in children aged 2–5 years with spastic cerebral palsy (CP) who had received no previous intramuscular injections of botulinum neurotoxin type-A (BoNT-A) and were randomised to receive either single or multiple (three) BoNT-A injections to the gastrocsoleus. MG morphological changes were compared to age-matched typically developing (TD) peers. **Methods:** Thirteen children with spastic CP with a mean age of 45 (15) months and 18 TD children with a mean age of 48 (14) months participated in the study. The principal outcome measures were MG muscle volume, fascicle length, pennation angle and physiological cross-sectional area (PCSA), which were obtained using 2D and 3D ultrasound. **Results:** The single and multiple injection frequency groups significantly increased MG muscle volume at 12 months relative to the baseline by 13 and 15 %, respectively. There were no significant differences in the MG muscle volume 28.5 (12.3) versus 30.3 (3.8) ml, fascicle length 48.0 (10.4) versus 44.8 (1.2) mm or PCSA 7.0 (1.2) versus 6.6 (1.7) cm² between the single and multiple injection groups, respectively, at 12 months follow-up. The change in MG muscle volume in the single and multiple injection groups was significantly lower than the TD peers by 66 and 60 %, respectively. **Interpretation:** In young children with spastic CP, naive to BoNT-A treatment, MG muscle growth over 12 months does not appear to be influenced by intramuscular BoNT-A injection frequency. However, MG muscle growth in the spastic CP groups was significantly lower than the age-matched TD peers. It is unclear whether this is an effect of intramuscular BoNT-A injections or reduced growth rates in children with spastic CP in general. Controlled investigations and longitudinal studies with multiple measurement time points are required in order to determine the influence of BoNT-A treatment on muscle physiological and mechanical growth factors in young children with spastic CP.

Available: [SpringerLink](http://link.springer.com/article/10.1007/s11832-013-0503-x) <http://link.springer.com/article/10.1007/s11832-013-0503-x>

4. Foot Ankle Int. 2013 Jul 11. [Epub ahead of print]**Realignment Surgery for Severe Talar Tilt Secondary to Paralytic Cavovarus.**

Lee WC, Ahn JY, Cho JH, Park CH.

Department of Orthopaedic Surgery, Seoul Paik Hospital, Institute for Research of Foot and Ankle Diseases, Inje University, Seoul, South Korea.

BACKGROUND: Realignment surgeries for mild to moderate ankle osteoarthritis with minimal talar tilt have been reported to be effective. However, there has been no report on joint-sparing surgery of ankle osteoarthritis in patients with paralytic disorders who have severe talar tilt. We therefore investigated whether ankle osteoarthritis with severe talar tilt caused by paralytic disorders can be improved after operative treatment. **METHODS:** This study included 12 ankles (11 patients) with varus ankle osteoarthritis from paralytic disorders with cavovarus deformity of the foot. Mean follow-up period was 3.0 years (range, 2-4.5 years). Causes of paralysis were residual polio in 7 ankles (6 patients), cerebral palsy in 2 ankles, and idiopathic in 3 ankles. Preoperative and postoperative clinical assessments were performed using the American Orthopaedic Foot & Ankle Society (AOFAS) ankle-hindfoot score and a visual analogue scale (VAS). The Ankle Osteoarthritis Scale (AOS) was used for postoperative assessment. Pre- and postoperative radiographic parameters were compared. **RESULTS:** Mean AOFAS score improved from 39.1 (range, 32-57) preoperatively to 77.9 (range, 72-85) postoperatively. Mean talar tilt improved from 17.4 degrees (range, 9.5-33.5 degrees) to 1.4 degrees (range, 0-4 degrees). Degree of osteoarthritis according to Takakura classification improved in all ankles except two. Mean heel alignment angle was reduced from 40.4 degrees (range, 2-65 degrees) of varus preoperatively to 11.2 degrees (range, -3 to 25.5 degrees) of varus postoperatively. **CONCLUSION:** Medial varus ankle osteoarthritis from paralytic cavovarus may be improved even in cases of severe talar tilt.

LEVEL OF EVIDENCE: Level IV, retrospective case series.

[PMID: 23832713](#) [PubMed - as supplied by publisher]

5. Dev Neurorehabil. 2013 Jul 11. [Epub ahead of print]**A new virtual reality tool for unilateral cerebral palsy rehabilitation: Two single-case studies.**

Dinomais M, Veaux F, Yamaguchi T, Richard P, Richard I, Nguyen S.

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Objective: To examine the "clinical utility" of a new virtual rehabilitation tool in order to treat upper-limb motor deficit in cerebral palsy (CP) patient. **Methods:** Single-case experimental designs. Virtual reality intervention was performed in two left unilateral spastic CP adolescent patients. The virtual reality intervention was given for 60 minutes per session, two sessions a day, and 5 days a week over 2 weeks. **Results:** For each patient and for both hands, the number of blocks transported within one minute (box and block test scores) is increased. The nonoverlap of all pairs indices for the paretic hand were calculated as 0.95 for subject 1 and 0.93 for subject 2, and the nonoverlap of all pairs indices for the nonparetic hand were calculated, respectively, as 0.92 and 1. **Conclusion:** We provide empirical evidence in support of a new simple Virtual Rehabilitation system in CP patient to improve upper-limb motor function.

[PMID: 23845037](#) [PubMed - as supplied by publisher]

6. J Plast Surg Hand Surg. 2013 Jul 8. [Epub ahead of print]**New chest tube management maintained with negative pressure therapy.**

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Management of a chest tube, especially when lasting for a long period, becomes difficult. The orifice of thoracostomy where a tube is inserted is maintained sterile usually with water-tight sutures. The longer and tighter the sutures are placed, the more severely skin becomes sore by pressure. Recently not only pressure sores but also other various open wounds were successfully treated by negative pressure therapy. This study presents two cases of intrathoracic abscess which were successfully treated by chest tube maintained with negative pressure therapy. One case was a bed-ridden cerebral palsy patient with empyema thoracis and the other was a premature baby with lung abscess. Both needed a chest tube longer than a month. Negative pressure was applied not only in the abscess cavity but also in the wound of tube insertion, thus simultaneously draining the abscess and accelerating the wound healing of the tube insertion. It was gradually elevated from 20 to 50 H₂O (14.7 to 36.7 mmHg), which is below the level of the physiologic forced inspiration, with a 10 cm H₂O (7.4 mmHg) increase every other day. Both abscesses disappeared without a complication.

[PMID: 23829503](#) [PubMed - as supplied by publisher]

7. Acta Odontol Scand. 2013 Jul 9. [Epub ahead of print]**Inter-rater and intra-rater agreement on the Nordic Orofacial Test-Screening examination in children, adolescents and young adults with cerebral palsy.**

Edvinsson SE, Lundqvist LO.

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Objective. To evaluate inter-rater and intra-rater agreement on the Nordic Orofacial Test-Screening (NOT-S) examination applied to children, adolescents and young adults with cerebral palsy (CP). **Materials and methods.** Using the NOT-S examination, two speech and language pathologists independently assessed video recordings of 48 subjects with CP aged 5-22 years and representing all CP sub-diagnoses and levels of gross motor function and manual ability. Thirty-one subjects were reassessed. Fifteen out of 17 items in the NOT-S examination domains (1) Face at rest, (2) Nose breathing, (3) Facial expression, (4) Masticatory muscle and jaw function, (5) Oral motor function and (6) Speech were rated using a 'yes' (dysfunction observed)/'no' format, generating an overall score of 0-6. **Results.** Inter-rater agreement: Twelve out of 15 items and five out of six domains showed acceptable unweighted kappa values ($\kappa = 0.46-1.00$). The lowest kappa value was found for domain 4 ($\kappa = -0.04$), although it had high inter-rater agreement (92%). The linear weighted kappa value for the overall NOT-S examination score was 0.65 (95% CI = 0.49-0.82). Intra-rater agreement: All items and domains showed acceptable unweighted kappa values (items 0.58-1.00 and 0.59-1.00, domains 0.81-1.00 and 0.62-0.89) for both raters. The linear weighted kappa value for the overall NOT-S examination score was 0.81 (95% CI = 0.63-0.99) for rater A and 0.54 (95% CI = 0.25-0.82) for rater B. **Conclusions.** The NOT-S examination has acceptable inter-rater and intra-rater agreement when used in young individuals with CP.

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8. Logoped Phoniatr Vocol. 2013 Jul 10. [Epub ahead of print]**Living through a computer voice: A personal account.**

Martin A, Newell C.

Alan Martin, the first author of this paper, has cerebral palsy and uses a voice output communication aid (VOCA) to speak, and this paper describes the personal experience of living 'through' a computer voice (or VOCA) in the form

of an interview of Mr Martin conducted by Dr Newell. The interview focuses on the computerized voice output rather than other features of the VOCA. In presenting a first-hand account of the experience of actually using VOCA, the intention is that both everyday, practical issues of the technology and broader imaginative, philosophical, and sociological implications will be explored. Based upon the interview, the authors offer an informal set of design requirements and recommendations for the development of future VOCAs.

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9. Surg Endosc. 2013 Jul 9. [Epub ahead of print]

Disease-based mortality after percutaneous endoscopic gastrostomy: utility of the enterprise data warehouse.

Poulose BK, Kaiser J, Beck WC, Jackson P, Nealon WH, Sharp KW, Holzman MD.

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BACKGROUND: Percutaneous endoscopic gastrostomy (PEG) remains a mainstay of enteral access. Thirty-day mortality for PEG has ranged from 16 to 43 %. This study aims to discern patient groups that demonstrate limited survival after PEG placement. The Enterprise Data Warehouse (EDW) concept allows an efficient means of integrating administrative, clinical, and quality-of-life data. On the basis of this concept, we developed the Vanderbilt Procedural Outcomes Database (VPOD) and analyzed these data for evaluation of post-PEG mortality over time. **METHODS:** Patients were identified using the VPOD from 2008 to 2010 and followed for 1 year after the procedure. Patients were categorized according to common clinical groups for PEG placement: stroke/CNS tumors, neuromuscular disorders, head and neck cancers, other malignancies, trauma, cerebral palsy, gastroparesis, or other indications for PEG. All-cause mortality at 30, 60, 90, 180, and 360 days was determined by linking VPOD information with the Social Security Death Index. Chi-square analysis was used to determine significance across groups. **RESULTS:** Nine hundred fifty-three patients underwent PEG placement during the study period. Mortality over time (30-, 60-, 90-, 180-, and 360-day mortality) was greatest for patients with malignancies other than head and neck cancer (29, 45, 57, 66, and 72 %) and least for cerebral palsy or patients with gastroparesis (7 % at all time points). Patients with neuromuscular disorders had a similar mortality curve as head and neck cancer patients. Stroke/CNS tumor patients and patients with other indications had the second highest mortality, while trauma patients had low mortality. **CONCLUSIONS:** PEG mortality was much higher in patients with malignancies other than head and neck cancer compared to previously published rates. PEG should be used with great caution in this and other high-risk patient groups. This study demonstrates the power of an EDW-based database to evaluate large numbers of patients with clinically meaningful results.

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10. Arch Phys Med Rehabil. 2013 Jul 5. pii: S0003-9993(13)00512-1. doi: 10.1016/j.apmr.2013.06.020. [Epub ahead of print]

Capacity to Participation in Cerebral Palsy: Evidence of an Indirect Path via Performance.

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OBJECTIVE: To examine the hypothesis that the influence of physical activity capacity on participation is mediated through activity performance. **DESIGN:** Secondary analysis of a prospective cross-sectional study sample. **SETTING:** Regional pediatric specialty care hospital. **PARTICIPANTS:** Participants included 128 children with cerebral palsy (CP) with 59% male, ages 2-9 years, Gross Motor Function Classification System levels I-III, 49% had hemiplegia and 72% with spasticity. **INTERVENTION:** Not Applicable. **MAIN OUTCOME MEASURES:** Activity capacity was measured with the Gross Motor Function Measure-66 (GMFM-66) and performance with the Activities Scale for Kids (ASKp). Participation was measured with the Assessment of Life Habits (Life-H). Children's Assessment of Participation and Enjoyment (CAPE) and the Assessment of Preschool Children's Participation

(APCP) assessed diversity participation. Regression equations and Sobel's z test were employed to examine the mediated effect via performance. RESULTS: Physical activity performance mediates 74.9% ($\beta=.83$, $p<0.001$) of the effect of activity capacity on total participation levels and 52.8% ($\beta=.47$, $p=0.001$) of the effect of capacity on diversity participation. CONCLUSIONS: The relationship between what an ambulatory child with CP is 'able' to perform in a clinic setting and their participation in life is significantly mediated by what they 'actually' do motorically in day to day life. Results suggest that interventions focusing on improving what they actually do every day regardless of capacity to perform (what they can do when tested) may positively influence participation.

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Prevention and Cure

11. BMJ. 2013 Jul 9;347:f4089. doi: 10.1136/bmj.f4089.

Mediators of the association between pre-eclampsia and cerebral palsy: population based cohort study.

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OBJECTIVE: To test the hypothesis that pre-eclampsia is a risk factor for cerebral palsy mediated through preterm birth and being born small for gestational age. DESIGN: Population based cohort study. SETTING: Clinical data from the Norwegian Cerebral Palsy Registry were linked with perinatal data prospectively recorded by the Medical Birth Registry of Norway. PARTICIPANTS: All singleton babies who survived the neonatal period during 1996-2006 (849 children with cerebral palsy and 616 658 control children). MAIN OUTCOME MEASURES: Cerebral palsy and cerebral palsy subtypes. RESULTS: Children exposed to pre-eclampsia had an excess risk of cerebral palsy (unadjusted odds ratio 2.5, 95% confidence interval 2.0 to 3.2) compared with unexposed children. Among children born at term (≥ 37 weeks), exposure to pre-eclampsia was not associated with an excess risk of cerebral palsy in babies not born small for gestational age (1.2, 0.7 to 2.0), whereas children exposed to pre-eclampsia and born small for gestational age had a significantly increased risk of cerebral palsy (3.2, 1.5 to 6.7). Non-small for gestational age babies born very preterm (< 32 weeks) and exposed to pre-eclampsia had a reduced risk of cerebral palsy compared with unexposed children born at the same gestational age (0.5, 0.3 to 0.8), although the risk was not statistically significantly reduced among children exposed to pre-eclampsia and born small for gestational age (0.7, 0.4 to 1.3). Exposure to pre-eclampsia was not associated with a specific cerebral palsy subtype. CONCLUSIONS: Exposure to pre-eclampsia was associated with an increased risk of cerebral palsy, and this association was mediated through the children being born preterm or small for gestational age, or both. Among children born at term, pre-eclampsia was a risk factor for cerebral palsy only when the children were small for gestational age.

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12. Ann Neurol. 2013 Jul 9. doi: 10.1002/ana.23971. [Epub ahead of print]

Mutations in gamma adducin are associated with inherited cerebral palsy.

Kruer MC, Jepperson T, Dutta S, Steiner RD, Cottenie E, Sanford L, Merkens M, Russman BS, Blasco PA, Fan G, Pollock J, Green S, Woltjer RL, Mooney C, Kretschmar D, Paisán-Ruiz C, Houlden H.

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OBJECTIVE: Cerebral palsy is estimated to affect nearly 1 in 500 children, and although prenatal and perinatal

contributors have been well-characterized, at least 20% of cases are believed to be inherited. Previous studies have identified mutations in the actin-capping protein KANK1 and the adaptor protein -4 complex in forms of inherited cerebral palsy, suggesting a role for components of the dynamic cytoskeleton in the genesis of the disease. METHODS: We studied a multiplex consanguineous Jordanian family by homozygosity mapping and exome sequencing, then used patient-derived fibroblasts to examine functional consequences of the mutation we identified in vitro. We subsequently studied the effects of adducin loss of function in drosophila. RESULTS: We identified a homozygous c.1100G>A [p.G367D] mutation in ADD3, encoding gamma adducin in all affected members of the index family. Follow-up experiments in patient fibroblasts found that the p.G367D mutation, which occurs within the putative oligomerization critical region, impairs the ability of gamma adducin to associate with the alpha subunit. This mutation impairs the normal actin capping function of adducin, leading to both abnormal proliferation and migration in cultured patient fibroblasts. Loss of function studies of the Drosophila adducin orthologs confirmed a critical role for adducin in locomotion. INTERPRETATION: Although likely a rare cause of cerebral palsy, our findings indicate a critical role for adducins in regulating the activity of the actin cytoskeleton, suggesting that impaired adducin function may lead to neuromotor impairment and further implicating abnormalities of the dynamic cytoskeleton as a pathogenic mechanism contributing to cerebral palsy. ANN NEUROL 2013. © 2013 American Neurological Association.

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13. J Vasc Surg. 2013 Jul 2. pii: S0741-5214(13)00869-0. doi: 10.1016/j.jvs.2013.04.047. [Epub ahead of print]

A peroxisome proliferator-activated receptor gamma agonist attenuates neurological deficits following spinal cord ischemia in rats.

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OBJECTIVE: Neuroprotective effects of the peroxisome proliferator-activated receptor gamma (PPAR γ) agonist in cerebral ischemia have been reported, but the effect of a PPAR γ agonist on spinal cord ischemia has not been investigated. The objective of this study was to investigate the effect of a PPAR γ agonist on spinal cord ischemia. Pioglitazone, a PPAR γ agonist, was administered in a rat model of spinal cord ischemia, and the extent of neurological damage and histological alterations were assessed. METHODS: Forty-five rats were randomly enrolled into one of the three groups: (1) pioglitazone group (group PIO): rats were treated with pioglitazone 24 hours before ischemia; (2) control group (group C): rats were treated with the same volume of saline 24 hours before ischemia; and (3) sham group (group sham): rats were treated with the same volume of saline 24 hours before the sham surgery. Spinal cord ischemia was induced using a balloon-tipped catheter placed on the proximal descending aorta. Neurologic function was assessed using the motor deficit index (0 = normal, 6 = complete paralysis) during the 48 hours after reperfusion. Histological and biochemical evaluations were then performed. RESULTS: Compared with group C, group PIO presented with lower motor deficit index 48 hours after reperfusion (5.0 [4.0-6.0] vs 3.0 [2.0-3.0]; group C vs group PIO, respectively; P < .001). Group PIO presented with a higher number of normal motor neurons (10.7 [8.1-11.9] vs 14.7 [14.0-15.3]; group C vs group PIO, respectively; P = .009) and a smaller area of infarcts (48.4% [46.3%-54.0%] vs 16.8% [11.5%-18.3%]; group C vs group PIO, respectively; P = .009) when compared with group C. The degree of inflammatory reactions, assessed by microglia activities, was significantly reduced in group PIO. Oxidative stress level, assessed using malonyldialdehyde assay, was significantly reduced in group PIO relative to group C (192.21% [173.5%-206.4%] of sham vs 141.1% [131.7%-152.1%] of sham; group C vs group PIO, respectively; P = .007). The sham group exhibited no abnormality upon neurological or histological examination. CONCLUSIONS: PPAR γ agonist pioglitazone pretreatment significantly reduces infarct volume and attenuates neurological deficits following spinal cord ischemia. The possible mechanism of neuroprotection by PPAR γ agonist may involve modulation of inflammatory reaction and oxidative stress.

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14. *Pediatr Res.* 2013 Jul 8. doi: 10.1038/pr.2013.117. [Epub ahead of print]

Single nucleotide polymorphism (SNP) associations with preterm delivery: a case-control replication study and meta-analysis.

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Background: To replicate single nucleotide polymorphism (SNP) associations with preterm birth (PTB; birth <37 completed weeks of gestation) and synthesise currently available evidence using meta-analysis. **Methods:** Spontaneous PTB cases and controls were selected from an existing cohort. Candidate SNPs were taken from an existing genotype panel. A systematic review was conducted for each SNP in the panel to determine suitability as a PTB candidate. Those with significant associations previously reported in Caucasians were selected for replication. Candidate SNPs were already genotyped in cases and controls and clinical data accessed from state perinatal and cerebral palsy databases. Association analysis was conducted between each SNP and PTB and meta-analysis conducted if there were \geq three studies in the literature. Maternal and fetal SNPs were considered as separate candidates. **Results:** A cohort of 170 cases and 583 controls was formed. Eight SNPs from the original panel of genotyped SNPs were selected as PTB candidates and replication based on systematic literature review results. In our cohort, fetal Factor V Leiden (FVL) was significantly associated with PTB (OR 2.6, 95% CI 1.31-5.17) and meta-analysis confirmed this association (OR 2.71, 95% CI 1.15-6.4). **Conclusion:** Replication and meta-analysis support an increased risk of PTB in Caucasians with the fetal FVL mutation.

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