
Technique of Pronator Teres Rerouting in Pediatric Patients With Spastic Hemiparesis.

Oishi S, Butler L.

Children with spastic hemiparesis can present with forearm pronation deformities that can greatly impair function. In the appropriate setting, pronator teres rerouting can provide active supination while preserving active pronation, which may improve function in these patients. Patient selection is imperative for the success of this procedure because, in the wrong setting, pronator teres rerouting can lead to fixed supination deformity that may actually worsen position and function in these patients.

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Elvrum AG, Beckung E, Sæther R, Lydersen S, Vik T, Himmelmann K.

AIMS: To develop a revised edition of the Bimanual Fine Motor Function (BFMF 2), as a classification of fine motor capacity in children with cerebral palsy (CP), and establish intra- and interrater reliability of this edition. METHODS: The content of the original BFMF was discussed by an expert panel, resulting in a revised edition comprising the original description of the classification levels, but in addition including figures with specific explanatory text. Four professionals classified fine motor function of 79 children (3-17 years; 45 boys) who represented all subtypes of CP and Manual Ability Classification levels (I-V). Intra- and inter-rater reliability was assessed using overall intra-class correlation coefficient (ICC), and Cohen's quadratic weighted kappa. RESULTS: The overall ICC was 0.86. Cohen's weighted kappa indicated high intra-rater (κw >0.90) and inter-rater (κw >0.85) reliability. CONCLUSIONS: The revised BFMF 2 had high intra- and interrater reliability. The classification levels could be determined from short video recordings (<5 minutes), using the figures and precise descriptions of the fine motor function levels included in the BFMF 2. Thus, the BFMF 2 may be a feasible and useful classification of fine motor capacity both in research and in clinical practice.

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Bales J, Apkon S, Osorio M, Kinney G, Robison RA, Hooper E, Browd S.

**BACKGROUND/AIMS:** Selective dorsal rhizotomy for spastic cerebral palsy is an effective and well-validated surgical approach. Multiple techniques have been described in the past including multiple laminectomies and a single-level laminectomy at the level of the conus. There is considerable technical challenge involved with a single-level laminectomy approach.

**METHODS:** We report here a modification of the single-level laminectomy that selectively analyzes each individual nerve root with electromyography to separate dorsal and ventral nerve roots through comparison of stimulus responses.

**RESULTS:** In 18 children with cerebral palsy who underwent this operation there was a mean improvement in the Modified Ashworth Scale of 2.0 with no reported incidence of muscle weakness, sensory loss, or neurogenic bladder.

**CONCLUSION:** This approach allows for a modification of selective dorsal rhizotomy through a single-level laminectomy and tailors the selection of nerve root sectioning to the individual patient of interest while still maintaining its effectiveness.

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A cohort study of tibialis anterior tendon shortening in combination with calf muscle lengthening in spastic equinus in cerebral palsy.

Tsang ST, McMorran D, Robinson L, Herman J, Robb JE, Gaston MS.

The aim of this study was to evaluate the outcome of combined tibialis anterior tendon shortening (TATS) and calf muscle-tendon lengthening (CMTL) in spastic equinus. Prospectively collected data was analysed in 26 patients with hemiplegic (n=13) and diplegic (n=13) cerebral palsy (CP) (GMFCS level I or II, 14 males, 12 females, age range 10-35 years; mean 16.8 years). All patients had pre-operative 3D gait analysis and a further analysis at a mean of 17.1 months (+5.6months) after surgery. None was lost to follow-up. Twenty-eight combined TATS and CMTL were undertaken and 19 patients had additional synchronous multilevel surgery. At follow-up 79% of patients had improved foot positioning at initial contact, whilst 68% reported improved fitting or reduced requirement of orthotic support. Statistically significant improvements were seen in the Movement Analysis Profile for ankle dorsiflexion (4.15°, p=0.032), maximum ankle dorsiflexion during swing phase (11.68°, p=0.001), and Edinburgh Visual Gait Score (EVGS) (4.85, p=0.014). Diplegic patients had a greater improvement in the EVGS than hemiplegics (6.27 -vs- 2.21, p=0.024). The originators of combined TATS and CMTL showed that it improved foot positioning during gait. The present study has independently confirmed favourable outcomes in a similar patient population and added additional outcome measures, the EVGS, foot positioning at initial contact, and maximum ankle dorsiflexion during swing phase. Study limitations include short term follow-up in a heterogeneous population and that 19 patients had additional surgery. TATS combined with CMTL is a recommended option for spastic equinus in ambulatory patients with CP.

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Gastrocnemius muscle-tendon interaction during walking in typically-developing adults and children, and in children with spastic cerebral palsy.

Kalsi G, Fry NR, Shortland AP.

**BACKGROUND:** Our understanding of the interaction of muscle bellies and their tendons in individuals with muscle pathology is limited. Knowledge of these interactions may inform us of the effects of musculoskeletal pathologies on muscle-tendon dynamics and the subsequent neurological control strategies used in gait. Here, we investigate gastrocnemius muscle-tendon interaction in typically-developing (TD) adults and children, and in children with spastic cerebral palsy (SCP).

**METHODS:** We recruited six TD adults (4 female; mean age: 34 yrs. (24-54)), eight TD children (5 female; mean age: 10 yrs. (6-12)) and eight independently ambulant children with SCP (5 female; mean age 9 yrs. (6-12); 3 unilaterally-affected). A combination of 3D motion capture and 2D real-time ultrasound imaging were used to compute the gastrocnemius musculo-
tendinous unit (MTU) length and estimate muscle belly and tendon lengths during walking. For the TD subjects, the measurements were made for heel-toe walking and voluntary toe-walking. RESULTS: The gastrocnemius muscle bellies of children with SCP lengthened during single support (p = 0.003). In contrast, the muscle bellies of TD subjects did not demonstrate an increase in length over the period of single support under heel-toe or toe-walking conditions. CONCLUSION: We observed lengthening of the gastrocnemius muscle bellies in children with SCP during single support, a phase of the gait cycle in which the muscle is reported consistently to be active. Repeated lengthening of muscle bellies while they are active may lead to muscle damage and have implications for the natural history of gait in this group.

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Variability of total step activity in children with cerebral palsy: influence of definition of a day on participant retention within the study.

Wilson NC, Mudge S, Stott NS.

BACKGROUND: Activity monitoring is important to establish accurate daily physical activity levels in children with cerebral palsy (CP). However, few studies address issues around inclusion or exclusion of step count data; in particular, how a valid day should be defined and what impact different lengths of monitoring have on retention of participant data within a study. This study assessed how different 'valid day' definitions influenced inclusion of participant data in final analyses and the subsequent variability of the data. RESULTS: Sixty-nine children with CP were fitted with a StepWatch™ Activity Monitor and instructed to wear it for a week. Data analysis used two broad definitions of a day, based on either number of steps in a 24 h monitoring period or the number of hours of recorded activity in a 24 h monitoring period. Eight children either did not use the monitor, or used it for only 1 day. The remaining 61 children provided 2 valid days of monitoring defined as >100 recorded steps per 24 h period and 55 (90 %) completed 2 valid days of monitoring with ≥10 h recorded activity per 24 h period. Performance variability in daily step count was lower across 2 days of monitoring when a valid day was defined as ≥10 h recorded activity per 24 h period (ICC = 0.765) and, higher when the definition >100 recorded steps per 24 h period (ICC = 0.62). Only 46 participants (75 %) completed 5 days of monitoring with >100 recorded steps per 24 h period and only 23 (38 %) achieved 5 days of monitoring with ≥10 h recorded activity per 24 h period. Datasets of participants who functioned at GMFCS level II were differentially excluded when the criteria for inclusion in final analysis was 5 valid days of ≥10 h recorded activity per 24 h period, leaving datasets available for only 8 of 32 participant datasets retained in the study. CONCLUSION: We conclude that changes in definition of a valid day have significant impacts on both inclusion of participant data in final analysis and measured variability of total step count.

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The role of visual stimuli on standing posture in children with bilateral cerebral palsy.


BACKGROUND: In children with bilateral cerebral palsy (CP) maintaining a standing position can be difficult. The fundamental motor task of standing independently is achieved by an interaction between the visual, somatosensory, and vestibular systems. In CP, the motor disorders are commonly accompanied by sensory and perceptual disturbances. Our aims were to examine the influence of visual stimuli on standing posture in relation to standing ability. METHODS: Three dimensional motion analysis with surface electromyography was recorded to describe body position, body movement, and muscle activity during three standing tasks: in a self-selected position, while blindfolded, and during an attention-demanding task. Participants were twenty-seven typically-developing (TD) children and 36 children with bilateral CP, of which 17 required support for standing (CP-SwS) and 19 stood without support (CP-SwoS). RESULTS: All children with CP stood with a more flexed body position than the TD children, even more pronounced in the children in CP-SwS. While blindfolded, the CP-SwS group further flexed their hips and knees, and increased muscle activity in knee extensors. In contrast, the children in CP-SwoS maintained the same body position but increased calf muscle activity. During the attention-demanding task, the children in CP-SwoS stood with more still head and knee positions and with less muscle activity. CONCLUSIONS: Visual input was important for children with CP to maintain a standing position. Without visual input the children who required support dropped into a further crouched position. The somatosensory and vestibular systems alone could not provide enough information about
the body position in space without visual cues as a reference frame. In the children who stood without support, an intensified visual stimulus enhanced the ability to maintain a quiet standing position. It may be that impairments in the sensory systems are major contributors to the difficulties to stand erect in children with CP.

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'Learn From Every Patient': implementation and early results of a learning health system.

AIM: The convergence of three major trends in medicine, namely conversion to electronic health records (EHRs), prioritization of translational research, and the need to control healthcare expenditures, has created unprecedented interest and opportunities to develop systems that improve care while reducing costs. However, operationalizing a 'learning health system' requires systematic changes that have not yet been widely demonstrated in clinical practice. METHOD: We developed, implemented, and evaluated a model of EHR-supported care in a cohort of 131 children with cerebral palsy that integrated clinical care, quality improvement, and research, entitled 'Learn From Every Patient' (LFEP). RESULTS: Children treated in the LFEP Program for a 12-month period experienced a 43% reduction in total inpatient days (p=0.030 vs prior 12mo period), a 27% reduction in inpatient admissions, a 30% reduction in emergency department visits (p<0.001), and a 29% reduction in urgent care visits (p=0.046). LFEP Program implementation also resulted in reductions in healthcare costs of 210% (US$7014/child) versus a Time control group, and reductions of 176% ($6596/child) versus a Program Activities control group. Importantly, clinical implementation of the LFEP Program has also driven the continuous accumulation of robust research-quality data for both publication and implementation of evidence-based improvements in clinical care. INTERPRETATION: These results demonstrate that a learning health system can be developed and implemented in a cost-effective manner, and can integrate clinical care and research to systematically drive simultaneous clinical quality improvement and reduced healthcare costs.

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Report of a workshop on research gaps in the treatment of cerebral palsy.

Lungu C, Hirtz D, Damiano D, Gross P, Mink JW.

Cerebral palsy (CP) is heterogeneous in etiology and manifestations, making research into relevant therapies difficult and limiting the generalizability of the results. We report here on the NIH CP symposium, where stakeholders from academic, clinical, regulatory, and advocacy backgrounds discussed the major challenges and needs for moving forward with clinical research in CP, and outlined priorities and action items. New information is constantly generated through research into pathogenesis and etiology. Clinical research and new therapeutic approaches need to keep pace, through large data registry integration and new research designs. Development of standardized data collection, increasing academic focus on CP research, and iterative approaches to treatment throughout the patients' lives, have all been identified as areas of focus. The workshop identified critical gaps and areas of focus to increase the evidence base for therapeutic approaches to determine which treatments work best for which patients in the near future. These include consolidation and optimization of databases and registries, updates to the research methodology, and better integration of resources and stakeholders.

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Chorioamnionitis at birth does not increase the risk of neurodevelopmental disability in premature infants with bronchopulmonary dysplasia.


AIM: To compare preterm infants with no bronchopulmonary dysplasia (BPD), BPD with chorioamnionitis (BPDC) and BPD with no chorioamnionitis (BPDCN) for the association with neurodevelopmental disability (NDD) at 3 years corrected age.

METHODS: Demographic and outcome data of infants with birth weight (BW) ≤1250 g born during 2 epochs (1995-2000 and 2002-2007 with an interim washout period of 1 year) were compared on the basis of whether they had BPD, chorioamnionitis or both. Any NDD was considered present if there was either mild-severe cerebral palsy (CP), cognitive delay, visual or hearing impairment. Logistic regression modeling was done. RESULTS: Infants (n=1009) were included into three groups - No BPD (n=442), BPDCN (n=437) and BPDC (n=130). The adjusted odds ratios of any NDD at 3 years in infants with BPDC versus BPDCN was OR 1.37; 95% CI 0.85-2.20, and for CP the OR was 1.66; 95%CI 0.76-3.62. Infants in the BPDC group were of lower BW, gestational age and had longer length of hospital stay, duration of mechanical ventilation, more blood transfusions and sepsis compared to BPDCN and no BPD groups (all p<0.001). CONCLUSION: Chorioamnionitis was not associated with any increase in the odds of NDD in infants with BPD at 3 years corrected age. This article is protected by copyright. All rights reserved.

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Effect of storage time on gene expression data acquired from unfrozen archived newborn blood spots.

Ho NT, Busik JV, Resau JH, Paneth N, Khoo SK.

Unfrozen archived newborn blood spots (NBS) have been shown to retain sufficient messenger RNA (mRNA) for gene expression profiling. However, the effect of storage time at ambient temperature for NBS samples in relation to the quality of gene expression data is relatively unknown. Here, we evaluated mRNA expression from quantitative real-time PCR (qRT-PCR) and microarray data obtained from NBS samples stored at ambient temperature to determine the effect of storage time on the quality of gene expression. These data were generated in a previous case-control study examining NBS in 53 children with cerebral palsy (CP) and 53 matched controls. NBS sample storage period ranged from 3 to 16 years at ambient temperature. We found persistently low RNA integrity numbers (RIN=2.3±0.71) and 28S/18S rRNA ratios (~0) across NBS samples for all storage periods. In both qRT-PCR and microarray data, the expression of three common housekeeping genes-beta cytoskeletal actin (ACTB), glyceraldehyde 3-phosphate dehydrogenase (GAPDH), and peptidylprolyl isomerase A (PPIA)-decreased with increased storage time. Median values of each microarray probe intensity at log2 scale also decreased over time. After eight years of storage, probe intensity values were largely reduced to background intensity levels. Of 21,500 genes tested, 89% significantly decreased in signal intensity, with 13,551, 10,730, and 9925 genes detected within 5 years, >5 to <10 years, and >10 years of storage, respectively. We also examined the expression of two gender-specific genes (X inactivation-specific transcript, XIST and lysine-specific demethylase 5D, KDM5D) and seven gene sets representing the inflammatory, hypoxic, coagulative, and thyroidal pathways hypothesized to be related to CP risk to determine the effect of storage time on the detection of these biologically relevant genes. We found the gender-specific genes and CP-related gene sets detectable in all storage periods, but exhibited differential expression (between male vs. female or CP vs. control) only within the first six years of storage. We concluded that gene expression data quality deteriorates in unfrozen archived NBS over time and that differential gene expression profiling and analysis is recommended for those NBS samples collected and stored within six years at ambient temperature.

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