Cerebral Palsy Alliance

NEWS
Cerebral Palsy Research

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Professor Nadia Badawi AM
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Interventions and Management

1. J Hand Surg Eur Vol. 2016 Jan 14. pii: 1753193415625611. [Epub ahead of print]

Wrist arthrodesis in adolescents with cerebral palsy.

Donadio J, Upex P, Bachy M, Fitoussi F.

Twenty adolescents with severe spastic deformities of the wrist (Zancolli type 3) and poor function on the House score were operated on between 2009 and 2014, and included in this retrospective cohort study. All were treated by wrist arthrodesis combining a proximal row carpectomy, curetting of the distal radius in order to imbed the capitate and a dorsal locking plate. The primary endpoint was improvement in the House score. Secondary endpoints included pre- and postoperative wrist flexion deformity, bone union, patient satisfaction regarding appearance and complications. The mean follow-up was 22 months. The mean age at the time of surgery was 16.2 years. Additional soft-tissue release was necessary in eight wrists. The mean House score improved significantly from 0.9 to 2.7. Average flexion deformity improved significantly from 66° to 10°. Bony union was achieved in all patients within 6 months. Four of the 20 patients required hardware removal because of fixed extension of the middle metacarpal. Wrist arthrodesis combining proximal row carpectomy with the use of a dorsal locking plate is a safe and reliable technique to improve function and appearance. LEVEL OF EVIDENCE: IV.

PMID: 26768218

2. Einstein (Sao Paulo). 2010 Sep;8(3):343-9. doi: 10.1590/S1679-45082010AO1678.

Two-dimensional analysis of gait asymmetry in spastic hemiplegia.

[Article in English, Portuguese]

Zonta MB, Ramalho Júnior A, Camargo RM, Dias FH, Santos LH.

OBJECTIVE: Simple measures of gait for routine clinical use could be useful when the complex gait analysis systems are not available. The aim of this study was to quantify asymmetry in children with spastic hemiplegia using a two-dimensional gait analysis by videography and to relate the asymmetry to motor function. METHODS: Twenty-four children with spastic hemiplegia (19 males, 5 females; mean age 49 months [SD 5 months], range from 39 to 60 months) were assessed with a two-dimensional gait analysis by videography and the analyzed parameters were compared with normal values and with clinical and functional data. RESULTS: There were significant differences in swing time ($p = 0.002$), stance time ($p = 0.01$) and stance/swing time ratio ($p < 0.001$). The comparison with the normal values described by Sutherland also demonstrated gait asymmetry. There was no

direct relationship between the motor function and asymmetry but a score analysis for specific Gross Motor Function Measure items could quantify it in terms of age of gait acquisition. Children with more adequate muscle tone presented longer stance time in the involved limb than those with more spasticity (p = 0.03). CONCLUSIONS: These results suggest that the best performance is associated with the smallest asymmetry in this sample. Although two-dimensional gait analysis does not provide as much data as three dimensional gait analyses, we believe it can contribute significantly to the gait assessment of children with cerebral palsy.

PMID: 26760152

3. Data Brief. 2015 Nov 10;5:967-70. doi: 10.1016/j.dib.2015.10.045. eCollection 2015.

Descriptive data on cardiovascular and metabolic risk factors in ambulatory and non-ambulatory adults with cerebral palsy.

McPhee PG, Gorter JW, Cotie LM, Timmons BW, Bentley T, MacDonald MJ.

Forty-two participants with cerebral palsy were recruited for a study examining traditional and novel indicators of cardiovascular risk (McPhee et al., 2015 [1]). Data pertaining to the prevalence of obesity, smoking, hypertension, and metabolic risk are provided. These data are presented along with the scoring methods used in evaluation of the study participants. Percentages are included for comparative purposes with the existing literature.

PMID: 26759816

4. Childs Nerv Syst. 2016 Jan 13. [Epub ahead of print]

Monosegmental laminoplasty for selective dorsal rhizotomy-operative technique and influence on the development of scoliosis in ambulatory children with cerebral palsy.

Funk JF, Haberl H.

Selective dorsal rhizotomy (SDR) reduces spasticity in children with cerebral palsy (CP) and is performed either through a lumbosacral multilevel laminectomy or a single-level laminectomy at the medullary conus. Spinal interventions generally involve the risk of subsequent instability depending on the extent of structural weakening. Destabilizing spasticity in CP might further increase this risk for both options. Laminoplasty is frequently applied to reduce instability through anatomical restoration, although the unavoidable interruption of interspinous ligaments might be a reason for inconsistent results. We report on a novel technique of laminoplasty, achieving complete restoration of the dorsal column. METHODS: One hundred sixteen ambulatory children with gross motor function classification scale (GMFCS) level I to III were submitted to SDR through a single-level approach. The lamina was reinserted with a previously unreported technique of laminoplasty. Osseous reintegration of the excised lamina was supposed, if its spinous process was located in place on late follow-up radiographs. Scoliosis was described via Cobb's angle. RESULTS: At a mean follow-up of 33 months, radiographs were available from 72 children with a mean age at surgery of 7.2 years. Sixty-two out of the 72 reinserted laminae were supposed to be vital and reintegrated. Seven children developed a predominantly mild scoliosis. No association was found between development of scoliosis and GMFCS level or age. CONCLUSIONS: This novel laminoplasty technique provides the least invasive approach for SDR. The incidence of scoliosis after this single-level approach is comparable to the natural history of ambulatory CP children.

PMID: 26759019

5. Spinal Cord. 2016 Jan 12. doi: 10.1038/sc.2015.223. [Epub ahead of print]

Validity and reliability of a locomotor stage-based functional rating scale in spinal cord injury.

Maurer-Burkhard B, Smoor I, von Reumont A, Deckstein G, Stierle I, Rupp R, Schuld C.

STUDY DESIGN: This is a prospective observational cohort study. OBJECTIVES: The objectives of this study were

to apply and adapt a rating scale based on locomotor stages (LSs) derived from cerebral palsy (CP) to spinal cord injury (SCI) and to quantify its inter-rater reliability and construct validity. METHODS: The inter-rater reliability of LSs originally developed for children with CP was tested in a chronic SCI cohort. On the basis of the distribution of the LSs for CP, Locomotor Stages in Spinal Cord Injury (LOSSCI) were defined. Their validity was then tested with the Spinal Cord Independence Measure (SCIM) in another acute SCI cohort. RESULTS: The 10-point LSs for CP were assessed by two raters in 65 chronic patients. Weighted Cohen's kappa (WCk) was 0.985 (P<0.0001). Only four mismatches were found, resulting in an accuracy of 93.4%. On the basis of the distribution of the LSs for CP in SCI, the five-point LOSSCI grading scale was developed. WCk of LOSSCI was 0.976 (P<0.0001). Only three mismatches between raters were found, resulting in an overall accuracy of 95.1%. The validity data sets consisted of 448 SCIM records from 161 patients obtained within the first year after injury. Spearman's correlation coefficients were the highest between LOSSCI and SCIM indoor mobility (room and toilet; R=0.82) and the lowest between LOSSCI and SCIM respiration and sphincter management (R=0.68). CONCLUSION: LOSSCI provides a reliable and valid clinical tool to assess locomotor function in SCI. LOSSCI not only reflects bipedal walking but also covers a wide range of key motor skills.Spinal Cord advance online publication, 12 January 2016; doi:10.1038/sc.2015.223.

PMID: 26754473

6. Spine (Phila Pa 1976). 2016 Jan;41(2):139-45. doi: 10.1097/BRS.0000000000001242.

Are S1 Screws a Useful Adjunct to Iliac Screws in Long Fusions to the Sacrum in Cerebral Palsy?

Schoenleber SJ, Asghar J, Bastrom TP, Shufflebarger HL; Harms Study Group.

STUDY DESIGN: Multicenter prospective database review of patients with cerebral palsy (CP) and spinal deformity.OBJECTIVE: To determine if the type of distal fixation is associated with improved correction of coronal deformity or pelvic obliquity (PO) at 2 years in long posterior fusions to the sacrum. SUMMARY OF BACKGROUND DATA: Multiple techniques are utilized for distal fixation in patients with CP. Although there is emerging evidence that the augmentation of iliac screws with S1 screws may be beneficial, this remains controversial. METHODS: A prospective, multicenter database was used to identify patients with CP who underwent long posterior fusions to the sacrum. Eighty-eight patients were included, 52 with iliac screws (I) and 36 with iliac and S1 screws (IS) for distal fixation. Preoperative, first erect, and 2-year follow-up radiographs and complications were analyzed. Statistical analysis was performed using ANOVA and repeated measures ANOVA with significance set at P<0.05. RESULTS: Scoliosis was the primary deformity in greater than 90% of patients in both groups (P=0.84). Preoperative coronal deformity was similar (I=83°, IS=87°, P=0.49), but correction was better with the use of S1 screws on the first erect radiograph and at 2 years (I=35°, IS=22°, P=0.001), reflecting correction of 58% and 74% for iliac and iliac-S1 screws, respectively (P<0.001). Preoperative PO was similar (I=29°, IS=30°, P=0.71) and was noted to improve more in the iliac-S1 group by 2 years (I=11°, IS=5°, P=0.004), representing correction of 60% and 77% for the iliac and iliac-S1 groups, respectively (P=0.018). There was no difference in the rate of major (P=0.27) or minor (P=0.65) complications in either group. CONCLUSION: Bilateral S1 and iliac screws are associated with improved spinal deformity and PO correction at 2 years in the CP population. Two points of distal fixation, S1, and ilium should be considered for this population. LEVEL OF EVIDENCE: 3.

PMID: 26751059

7. Einstein (Sao Paulo). 2015 Dec;13(4):555-9. doi: 10.1590/S1679-45082015AO3321.

Bone health in cerebral palsy and introduction of a novel therapy.

[Article in English, Portuguese]

Scheinberg MA, Golmia RP, Sallum AM, Pippa MG, Cortada AP, Silva TG.

Objective: To assess the bone health status of children with cerebral palsy and the therapeutic effect of denosumab in a subgroup of children with cerebral palsy and decreased bone mass. Methods Children with cerebral palsy were evaluated according to their motor disability score (classification system gross motor functions III to V), bone density and bone turnover markers. Dual X-ray energy absorption was used to measure the lumbar spine, and total

body, except the head. Thereafter a group of children with cerebral palsy and osteoporosis was treated with denosumab, a fully human monoclonal antibody. Bone turnover markers were measured before and three months after treatment. Results Reduction in bone mineral density was observed, particularly in children with greater impairment evaluated by the motor score. Decreased bone turnover markers were found in a selected group of children three months after exposure to denosumab. Conclusion Bone loss was present in children with significant impairment of motor function, as well as decreased serum levels of bone resorption markers with new forms.

PMID: 26761553

8. J Clin Endocrinol Metab. 2016 Jan 11:jc20153888. [Epub ahead of print]

Musculoskeletal and endocrine health in adults with cerebral palsy: new opportunities for intervention.

Trinh A, Wong P, Fahey MC, Brown J, Churchyard A, Strauss BJ, Ebeling PR, Fuller PJ, Milat F.

CONTEXT: Cerebral palsy (CP) increases fracture risk through diminished ambulation, nutritional deficiencies and anticonvulsant medication use. Studies examining bone mineral density (BMD) in adults with CP are limited. OBJECTIVE: To examine the relationship between body composition, BMD, and fractures in adults with CP. The effect of functional, nutritional and endocrine factors on BMD and body composition is also explored. DESIGN: Retrospective cross-sectional study. SETTING AND PARTICIPANTS: 45 adults with CP (mean age 28.3 ± 11.0 yrs) who had dual-energy x-ray absorptiometry imaging at a single tertiary hospital between 2005-2015. RESULTS: Seventeen (38%) had a past history of fragility fracture. 43% had a Z-score of ≤-2.0 at the lumbar spine (LS) and 41% at the femoral neck (FN). In non-ambulatory patients, every one unit decrease in FN Z-score increased the risk of fracture 3.2 fold (CI 1.07-9.70, P = 0.044). Stepwise linear regression revealed Gross Motor Function Classification System was the best predictor of LS Z-score (R2 = 0.550; β = -0.582, P = 0.002) and FN Z-score (R2 = 0.428; β = -0.494, P = 0.004). 35.7% of the variance in BMD was accounted for by lean tissue mass (LTM). Hypogonadism, present in 20% of patients, was associated with reduced skeletal muscle mass index and reduced LSBMD. LTM positively correlated with BMD in eugonadal, but not in hypogonadal patients. CONCLUSIONS: Low BMD and fractures are common in adults with CP. This is the first study to document hypogonadism in adults with CP with detrimental changes in body composition and BMD.

PMID: 26751195

9. Neurorehabil Neural Repair. 2016 Jan 7. pii: 1545968315624781. [Epub ahead of print]

Robotic Quantification of Position Sense in Children With Perinatal Stroke.

Kuczynski AM, Dukelow SP, Semrau JA, Kirton A.

BACKGROUND: Perinatal stroke is the leading cause of hemiparetic cerebral palsy. Motor deficits and their treatment are commonly emphasized in the literature. Sensory dysfunction may be an important contributor to disability, but it is difficult to measure accurately clinically. OBJECTIVE: Use robotics to quantify position sense deficits in hemiparetic children with perinatal stroke and determine their association with common clinical measures. METHODS: Case-control study. Participants were children aged 6 to 19 years with magnetic resonance imaging-confirmed unilateral perinatal arterial ischemic stroke or periventricular venous infarction and symptomatic hemiparetic cerebral palsy. Participants completed a position matching task using an exoskeleton robotic device (KINARM). Position matching variability, shift, and expansion/contraction area were measured with and without vision. Robotic outcomes were compared across stroke groups and controls and to clinical measures of disability (Assisting Hand Assessment) and sensory function. RESULTS: Forty stroke participants (22 arterial, 18 venous, median age 12 years, 43% female) were compared with 60 healthy controls. Position sense variability was impaired in arterial (6.01 ± 1.8 cm) and venous (5.42 ± 1.8 cm) stroke compared to controls (3.54 ± 0.9 cm, P < .001) with vision occluded. Impairment remained when vision was restored. Robotic measures correlated with functional disability. Sensitivity and specificity of clinical sensory tests were modest. CONCLUSIONS: Robotic assessment of

position sense is feasible in children with perinatal stroke. Impairment is common and worse in arterial lesions. Limited correction with vision suggests cortical sensory network dysfunction. Disordered position sense may represent a therapeutic target in hemiparetic cerebral palsy.

PMID: 26747126

10. Dev Med Child Neurol. 2016 Jan;58(1):98-9. doi: 10.1111/dmcn.12933.

Consensus classifications of gross motor, manual ability, and communication function classification systems between therapists and parents of children with cerebral palsy.

Bartlett DJ, Galuppi B, Palisano R, McCoy SW.

Letter to the Editor

PMID: 26767662

11. Child Care Health Dev. 2016 Jan 11. doi: 10.1111/cch.12309. [Epub ahead of print]

Child and youth experiences and perspectives of cerebral palsy: a qualitative systematic review.

Lindsay S

BACKGROUND: Cerebral palsy (CP) is one of the most common causes of physical disability in childhood, and many children with CP access rehabilitation services throughout their lives. The aim of this qualitative systematic review was to synthesize the experiences and perspectives of youth living with CP to inform the development of rehabilitation and social programmes. METHODS: A thematic qualitative synthesis integrating qualitative evidence was undertaken where six electronic databases (MEDLINE, Embase, Healthstar, Cumulative Index to Nursing and Allied Health Literature, Proquest and PsychInfo) were searched from 1980 to September 2014. RESULTS: Thirty-three articles involving 390 youth, aged from 2 to 25 years, across six countries were included. Themes were classified according to the International Classification of Functioning Child and Youth Version framework. Youth's accounts focused on social inclusion and the physical environment (i.e. services and supports, transportation, accessibility, accommodations, safety and weather), the role of family and peers and participation (i.e. leisure and recreation, school and civic engagement). Youth described how body structure and function (i.e. pain and physical functioning, mental health, fatigue and unpredictability of body function) affected them - often disrupting their biographies. Some youth described personal factors such as independence, coping and body image that affected their ability to cope with their condition. There was much less focus on youth's experiences of mobility, activities of daily living and assistive devices. CONCLUSIONS: Youth with CP experience pain, fatigue and impairments to body function, along with social exclusion, which can affect their biographies. However, youth had strategies to revise their biographies to maintain personal and social normalcy.

PMID: 26754030

12. Disabil Rehabil. 2016 Jan 10:1-9. [Epub ahead of print]

Child characteristics, caregiver characteristics, and environmental factors affecting the quality of life of caregivers of children with cerebral palsy.

Tseng MH, Chen KL, Shieh JY, Lu L, Huang CY, Simeonsson RJ.

Purpose: The study aimed to investigate comprehensively the determinants of the quality of life (QOL) of caregivers of children with cerebral palsy (CP) based on the International Classification of Functioning, Disability and Health for Children and Youth (ICF-CY). Methods A total of 167 children with CP (mean age 9.06 years, SD 2.61 years) and their caregivers (mean age 40.24 years, SD 5.43 years) participated in this study. The QOL of caregivers was measured with the World Health Organization Quality of Life-BREF-Taiwan version (WHOQOL-BREF-TW). The potential determinants of QOL were collected, including child characteristics, caregiver characteristics, and

environmental factors from all dimensions of the ICF-CY and analysed using multiple regression models. Results Four multiple regression models revealed that determinants of the QOL of caregivers of children with CP was multidimensional, encompassing child characteristics (age, type of CP, fine motor impairment, other diseases, behaviour and emotions, visual impairment, hearing impairment), caregiver characteristics (general mental health, parenting stress, marital status, family coping patterns, and socio-economic status), and environmental factors (child's medication, school setting, and current rehabilitation service, caregiver's spouse's age, family life impacts, and domestic helper). Conclusions Knowledge of the determinants of QOL could serve as a guide in a holistic approach to evaluation and intervention and help plan interventions targeted at these determinants to improve the QOL of caregivers of children with CP. Implications for Rehabilitation Caregivers of children with CP had lower QOL, except the environment QOL. The QOL determinants of caregivers of children with CP are multidimensional, including child characteristics, caregiver characteristics, and environmental factors. In addition to child characteristics of severity of fine motor impairments and emotional and behavioural problems, caregiver characteristics of general mental health, parenting stress, and coping patterns, and environmental factors of family life impacts, and school setting demonstrated important relationships with caregiver QOL.

PMID: 26752264

13. Eur J Paediatr Neurol. 2015 Dec 21. pii: S1090-3798(15)00212-3. doi: 10.1016/j.ejpn.2015.12.003. [Epub ahead of print]

Behavioral and emotional problems in children and adults with cerebral palsy.

Weber P, Bolli P, Heimgartner N, Merlo P, Zehnder T, Kätterer C.

OBJECTIVE: In patients with cerebral palsy (CP), psychological problems influence their participation in society. Little is known about the persistence of behavioral and social problems into adulthood. MATERIALS AND METHODS: In a two-center cross-sectional study, caregivers of 121 adults and 88 children were ask to assess behavior of the patients through the parent/caregiver forms of the Child Behavior Checklist (CBCL), the Strengths and Difficulties Questionnaire (SDQ), and the Vineland Adaptive Behavior Scale II (VABS). Questionnaires were returned from 43 adults and 39 children. RESULTS: In both groups we found the same frequency of abnormalities in attention problems (32.4 vs. 36.1%, p = 0.826) and social interaction problems (32.3 vs. 33.3%; p = 0.926) in the CBCL, and peer problems (38.9 vs. 75.7%; p = 0.115) in the SDQ. Children show a lower percentage of abnormal prosocial behavior (41.7 vs. 16.2%, p = 0.016) and lower abnormal rates of communication (88.2 vs. 61.5; p = 0.01) and daily living skills (90.0 vs. 71.8; p = 0.041), whereas the level of abnormalities in both groups in these dimensions of VABS notably high. CONCLUSION: The persistence of psychological and social problems from childhood into adulthood underlines the importance of focusing on early intervention.

PMID: 26748599

Prevention and Cure

14. Clin Genet. 2016 Jan 7. doi: 10.1111/cge.12723. [Epub ahead of print]

Association of COL4A1 gene polymorphisms with cerebral palsy in a Chinese Han population.

Bi D, Wang H, Shang Q, Xu Y, Wang F, Chen M, Ma C, Sun Y, Zhao X, Gao C, Wang L, Zhu C, Xing Q.

The basement membrane is an extracellular matrix associated with overlying cells and is important for proper tissue development, stability, and physiology. COL4A1 is the most abundant component of type IV collagen in the basement membrane, and COL4A1 variants can present with variable phenotypes that might be related to cerebral palsy (CP). We postulated, therefore, that variations in the COL4A1 gene might play an important role in the etiology of CP. In the present study, six single nucleotide polymorphisms (SNPs) in the COL4A1 gene were genotyped among 351 CP patients and 220 healthy controls from the Chinese Han population. Significant association was found for an association between CP and the rs1961495 (Allele: P = 0.008, OR = 1.387, 95% CI = 1.088-1.767) and rs1411040 (Allele: P = 0.009, OR = 1.746 95% CI = 1.148-2.656) SNPs of the COL4A1 gene. Multifactor dimensionality reduction analysis suggested that these SNPs had interactive effects on the risk of CP. Our study is the first attempt to investigate the contribution of polymorphisms in the COL4A1 gene to the

susceptibility of CP in a Chinese Han population. This study shows an association of the COL4A1 gene with CP and suggests a potential role of COL4A1 in the pathogenesis of CP.

PMID: 26748532

15. Zhonghua Er Ke Za Zhi. 2015 Sep;53(9):696-700.

[A voxel-based morphometric study on change of gray matter structures in cerebral palsy].

[Article in Chinese]

Wang Y, Wang H, Yu Y, Xu L, Chen Y, Wu D.

OBJECTIVE: To measure gray matter volume of whole brain with voxel-based morphometry (VBM) method and to study brain structures associated with gross motor function. METHOD: Forty children with cerebral palsy were recruited in the authors' hospital from Oct. 2012 to Dec. 2013 (26 male, 14 female cases, average age (3.6 ± 2.0) years). Gross motor function classification system (GMFCS) for children was used to obtain their motor function. The whole-brain three dimensional magnetic resonance imaging (MRI) was performed on a 3.0 T MRI scanner. The data were segmented by VBM 5, and the whole brain volumes of gray matter, white matter and cerebospinal fluid were produced. Correlation analysis was used to analyze the correlation of GMFCS with whole brain volumes using SPM 5 in Matalab 7.1. RESULT: The volume in left meditemporal gyrus (Z=3.57) and inferior temporal gyrus (Z=3.40), right thalamus and pallidum (Z=3.36), left thalamus and pallidum (Z=2.76), left supramarginal gyrus (Z=3.14), left precuneus gyrus (Z=3.00), right dorsolateral superior frontal gyrus (Z=3.08), right superior and medial occipital gyrus (Z=2.84) significantly increased as aggravation of gross motor dysfunction. The volume of the left medial orbitofrontal lobe and anterior cingulate (Z=3.28,3.02), left medial superior frontal gyrus (Z=3.19), left caudate (Z=3.04, 2.94, 2.92), left cerebellum (Z=2.94), right cerebellum (Z=2.97), left parahippocampal (Z=3.94), right parahippocampal (Z=3.43, 3.00), left insula (Z=3.50), right insula (Z=3.41, 3.80), left lingual (Z=3.37), right lingual (Z=3.30), left post cingulum (Z=2.73), left midioccipital gyrus (Z=2.92) and right miditemporal gyrus (Z=3.05) significantly reduced as the aggravation of gross motor dysfunction (P all<0.005). CONCLUSION: GMFCS in children with cerebral palsy is related to abnormalities of brain gray matter structure for motor, emotion, memory and default model network when examined with VBM method.

PMID: 26757971

Cerebral Palsy in Australia: A Special Supplement from the Australian Cerebral Palsy Register

1. Dev Med Child Neurol. 2016 Article first published online: 19 JAN 2016 | DOI: 10.1111/dmcn.13006

Foreword

Christine Cans and Nicole Gerrand

PMID: Not yet available

Wiley Online: http://onlinelibrary.wiley.com/doi/10.1111/dmcn.13006/abstract

2. Dev Med Child Neurol. 2016 Article first published online: 19 JAN 2016 | DOI: 10.1111/dmcn.13003 Acknowledgements

PMID: Not yet available

Wiley Online: http://onlinelibrary.wiley.com/doi/10.1111/dmcn.13003/abstract

3. Australia and the Australian Cerebral Palsy Register for the birth cohort 1996-2006

Australian Cerebral Palsy Register Group

Not yet available online

PMID: Not yet available

4. Dev Med Child Neurol. 2016 Jan 13. doi: 10.1111/dmcn.13026. [Epub ahead of print]

A special supplement: findings from the Australian Cerebral Palsy Register, birth years 1993 to 2006.

Smithers-Sheedy H, McIntyre S, Gibson C, Meehan E, Scott H, Goldsmith S, Watson L, Badawi N, Walker K, Novak I, Blair E; Australian Cerebral Palsy Register Group.

AIM: To briefly outline the strengths and limitations of cerebral palsy (CP) registers, and to report on findings of the Australian Cerebral Palsy Register (ACPR) pertaining to a population cohort of children with CP. METHOD: De-identified data were extracted from the ACPR for people with CP in birth years 1993 to 2006, from South Australia, Victoria, and Western Australia. Live birth prevalence of CP was estimated and risk factors described. RESULTS: The overall birth prevalence of CP (including those whose CP was postneonatally acquired) for the 1993 to 2006 birth cohort was 2.1 per 1000 live births (95% confidence interval [CI] 2.0-2.2). Excluding cases with a known postneonatal cause, the birth prevalence for pre/perinatally acquired CP was 2.0 per 1000 live births (95% CI 1.9-2.1). A downward trend in rates of CP in those born extremely preterm was evident over at least three consecutive periods across all three regions. Most (58.6%) children were born at term (≥37wks). Male sex, early gestational age, low birthweight, and multiple birth were risk factors for CP. INTERPRETATION: Overall rates of CP did not change during this period. The proportion of those with CP born extremely preterm decreased. The ACPR Group will investigate whether this pattern continues when data pertaining to the next birth cohort for all three regions becomes available.

PMID: 26762930

5. Dev Med Child Neurol. 2016 Jan 19. DOI: 10.1111/dmcn.12999

An international survey of cerebral palsy registers and surveillance systems

Shona Goldsmith, Sarah McIntyre, Hayley Smithers-Sheedy, Eve Blair, Christine Cans, Linda Watson, Marshalyn Yeargin-Allsopp and on behalf of the Australian Cerebral Palsy Register Group

Aim: To describe cerebral palsy (CP) surveillance programmes and identify similarities and differences in governance and funding, aims and scope, definition, inclusion/exclusion criteria, ascertainment and data collection, to enhance the potential for research collaboration. Method: Representatives from 38 CP surveillance programmes were invited to participate in an online survey and submit their data collection forms. Descriptive statistics were used to summarize information submitted. Results: Twenty-seven surveillance programmes participated (25 functioning registers, two closed owing to lack of funding). Their aims spanned five domains: resource for CP research, surveillance, aetiology/prevention, service planning, and information provision (in descending order of frequency). Published definitions guided decision making for the definition of CP and case eligibility for most programmes. Consent, case identification, and data collection methods varied widely. Ten key data items were collected by all programmes and a further seven by at least 80% of programmes. All programmes reported an interest in research collaboration.

Interpretation: Despite variability in methodologies, similarities exist across programmes in terms of their aims, definitions, and data collected. These findings will facilitate harmonization of data and collaborative research efforts, which are so necessary on account of the heterogeneity and relatively low prevalence of CP.

PMID: Not yet available

Wiley Online: http://onlinelibrary.wiley.com/doi/10.1111/dmcn.12999/abstract

6. Dev Med Child Neurol. 2016 Jan 14. doi: 10.1111/dmcn.13000. [Epub ahead of print]

Interobserver reliability of the Australian Spasticity Assessment Scale (ASAS).

Love S, Gibson N, Smith N, Bear N, Blair E; Australian Cerebral Palsy Register Group.

AIM: The aim of this paper is to present the Australian Spasticity Assessment Scale (ASAS) and to report studies of its interrater reliability. The ASAS identifies the presence of spasticity by confirming a velocity-dependent increased response to rapid passive movement and quantifies it using an ordinal scale. METHOD: The rationale and procedure for the ASAS is described. Twenty-two participants with spastic CP (16 males; age range 1y 11mo-15y 3mo) who had not had botulinum neurotoxin-A within 4 months, or bony or soft tissue surgery within 12 months, were recruited from the spasticity management clinic of a tertiary paediatric teaching hospital. Fourteen muscles in each child were assessed by each of three experienced independent raters. ASAS was recorded for all muscles. Interrater reliability was calculated using the weighted kappa statistic (quadratic weighting; κqw) for individual muscles, for upper limbs, for lower limbs, and between raters. RESULTS: The weighted kappa ranged between 0.75 and 0.92 for individual muscle groups and was 0.87 between raters. INTERPRETATION: The ASAS complies with the definition of spasticity and is clinically feasible in paediatric settings. Our estimates of interrater reliability for the ASAS exceed that of the most commonly used spasticity scoring systems.

PMID: 26762706

7. Dev Med Child Neurol. 2016 Jan 13. doi: 10.1111/dmcn.13001. [Epub ahead of print]

Temporal trends in cerebral palsy by impairment severity and birth gestation.

Reid SM, Meehan E, McIntyre S, Goldsmith S, Badawi N, Reddihough DS; Australian Cerebral Palsy Register Group.

AIM: Our aim was to build on previous research indicating that rates of cerebral palsy (CP) in the Australian state of Victoria are declining, and examine whether severity of impairments is also decreasing. METHOD: Data on individuals with CP were extracted from the Victorian Cerebral Palsy Register for birth years 1983 to 2009. The yearly rates of dichotomized categories for gross motor function, motor laterality, intellectual impairment, and epilepsy per 1000 neonatal survivors and proportions in the CP cohort were tabulated and plotted by birth gestation. Linear regression modelling was used to fit prediction curves; likelihood ratio tests were used to test for differences in trends between impairment severity groups. RESULTS: Since the mid-1990s, CP rates declined in neonatal survivors of birth at all gestations. Our data suggest that the decreasing CP rates were associated with relatively greater decreases in the rates of Gross Motor Function Classification System levels III to V, bilateral CP, epilepsy, and intellectual impairment (all $p<0.005$). Some variation was seen between birth gestation groups. INTERPRETATION: Declines in rates of CP of all levels of severity and complexity from the mid-1990s provides 'real-world' support for the effectiveness of concurrent neuroprotective strategies and continual innovation in perinatal practices.

PMID: 26762733

8. Dev Med Child Neurol. 2016 Jan 19 DOI: 10.1111/dmcn.13005

Comparing risks of cerebral palsy in births between Australian Indigenous and non-Indigenous mothers

Eve Blair, Linda Watson, Emily Okearney, Heather Dantoine, Michael Delacy and On behalf of the Australian Cerebral Palsy Register Group

Aim: To compare proportions of live births subsequently described as having cerebral palsy (CP), the distributions of associated impairments, and the causes of postneonatal CP between Aboriginal and Torres Strait Islander (Indigenous) and non-Indigenous populations in Australia. Method: Data from statutory birth records and CP registers for the 1996 to 2005 birth cohort in Queensland, Western Australia, and the Northern Territory were stratified by Indigenous status and whether the CP was acquired pre/perinatally or postneonatally. Relative risks associated with Indigenous status were estimated and the distributions of causes of postneonatal CP compared. Results: Indigenous births had a relative risk of 4.9 (95% confidence interval [CI] 3.0–7.9) for postneonatal CP but only of 1.42 (95% CI 1.2–1.7) for pre/perinatal CP. Almost half of postneonatal CP in Indigenous infants resulted from infection, whereas for non-Indigenous infants the most frequent cause was cerebrovascular accident. The impairments of Indigenous CP and of postneonatally acquired CP tended to be more numerous and more severe. Interpretation: Indigenous children are at significantly greater risk of CP, particularly postneonatal CP. The predominant cause of postneonatal CP in non-Indigenous children has shifted to cerebrovascular accident over time; however, infections followed by head injury are still the most frequent causes in Indigenous infants.

Wiley Online: http://onlinelibrary.wiley.com/doi/10.1111/dmcn.13005/abstract

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9. Dev Med Child Neurol. 2016 Jan 14. doi: 10.1111/dmcn.13021. [Epub ahead of print]

Biological sex and the risk of cerebral palsy in Victoria, Australia.

Reid SM, Meehan E, Gibson CS, Scott H, Delacy M; Australian Cerebral Palsy Register Group.

AIM: Males typically outnumber females in cerebral palsy (CP) cohorts. To better understand this 'male disadvantage' and provide insight into causal pathways to CP, this study used 1983 to 2009 Australian CP and population birth cohorts to identify associations and trends with respect to biological sex and CP. METHOD: Within birth gestation groups, sex ratios were calculated to evaluate any male excess in the CP cohort compared with livebirths, neonatal deaths, neonatal mortality and survival rates, neonatal survivors, and CP rates in survivors. Sex - and gestation-specific trends in neonatal mortality, CP rates, and CP sex ratios were assessed by plotting their annual frequencies and fitting quadratic curves. RESULTS: Over-representation of males in preterm live births partly explained the male excess in the CP cohort after preterm birth, especially at 28 to 31 weeks. Higher CP rates in male neonatal survivors also contributed to the male excess in CP, particularly at <28 and 37+ weeks. Higher neonatal mortality rates in males at all gestations had little impact on the CP sex ratio. There was no clearly discernible change over time in the CP sex ratio. INTERPRETATION: Gestation-specific associations between sex and CP provide insight into causal pathways to CP and suggest sex-specific differences in response to neuroprotective strategies.

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10. Dev Med Child Neurol. 2016 Jan 17 DOI: 10.1111/dmcn.13012

Profile of associated impairments at age 5 years in Australia by cerebral palsy subtype and Gross Motor Function Classification System level for birth years 1996 to 2005

Michael J deLacy, Susan M Reid and On behalf of the Australian cerebral palsy register group

Aim: To describe the distribution of impairments among persons with cerebral palsy (CP) in a large Australian cohort. Method: Records of persons on the Australian Cerebral Palsy Register (ACPR) (n=3466) born from 1996 to 2005 were reviewed to extract year of birth, sex, CP subtype, Gross Motor Function Classification System

(GMFCS) level, and impairments in vision, hearing, speech, intellect, and epilepsy. The distributions of GMFCS levels and CP subtype were plotted, and the proportions of each level of impairment were tabulated and presented as stacked graphs within the GMFCS and CP subtype distributions. Results:The proportions of persons with CP with each associated impairment increased with increasing GMFCS level. Compared with other spastic CP subtypes, individuals with spastic quadriplegia had higher frequencies of all associated impairments. Other than epilepsy, which was most prevalent in persons with spastic quadriplegia (53%), all impairments were most frequent in non-spastic CP subtypes. Hearing impairment was recorded for 21% of persons with dyskinesia whereas the hypotonic subtype had the highest prevalence of visual impairment (57%), intellectual impairment (90%), and speech impairment (95%). Interpretation:
Distributions of associated impairments across all GMFCS levels and CP subtypes in a large cohort are presented in formats suitable for clinical use and discussion with families.

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Wiley Online: http://onlinelibrary.wiley.com/doi/10.1111/dmcn.13012/abstract

11. Dev Med Child Neurol. 2016 Jan 14. doi: 10.1111/dmcn.13020. [Epub ahead of print] Strabismus, a preventable barrier to social participation: a short report.

Blair E, Smithers-Sheedy H; Australian Cerebral Palsy Register Group.

Isolated strabismus does not significantly impair visual functionality and has traditionally been considered a primarily cosmetic defect of little importance. However, even in the absence of strabismus amblyopia, manifest strabismus and its non-surgical treatments can render the person less socially acceptable, creating a barrier to participation and resulting in psychosocial disadvantage that has been documented in the typically developing population. The Australian Cerebral Palsy Register traditionally recorded strabismus only if it were not accompanied by visual impairment; however, even these data indicate that the proportion of cerebral palsy registrants with strabismus is many times higher than in comparable population samples, compounding their challenges to achieve participation. It is therefore inappropriate to continue to consider strabismus as merely a cosmetic defect, but one that deserves surgical correction early in life.

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12. Dev Med Child Neurol. 2016 Jan 14. DOI: 10.1111/dmcn.13013 Change in residential remoteness during the first 5 years of life in an Australian cerebral palsy cohort

Michael J DeLacy, Christalla Louca, Hayley Smithers-Sheedy, Sarah McIntyre and on behalf of the Australian Cerebral Palsy Register Group

Aim: To determine if families of children with cerebral palsy living in Australia move to less remote areas between birth and 5 years. Method: Children on the Australian Cerebral Palsy Register (n=3399) born 1996 to 2005, were assigned a remoteness value for family residence at birth and 5 years using a modification of the Australian Statistical Geography Standard. Each value at birth was subtracted from the value at 5 years yielding a positive difference if they moved more remotely, negative difference if they moved less remotely and a value of zero if they did not move or moved to an equally remote residence. Results: The small net increase in remoteness across this cohort was non-significant (p=0.43). Fifty-seven per cent of families changed postcode but only 20% changed remoteness, 11% more remotely, and 9% less remotely. There was a small trend for families with a child with more impaired gross motor function (Gross Motor Function Classification System levels IV and V) to move to a less remote area. Interpretation:
This cohort of families with children with cerebral palsy did not appear to move to less remote areas by age 5 years. Remoteness at birth and level of gross motor function seem to have little effect.

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13. Dev Med Child Neurol. 2016 Jan 19 DOI: 10.1111/dmcn.13007

The National Disability Insurance Scheme: a time for real change in Australia

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In Australia, the supports and services for persons with disabilities have long been underfunded and fragmented. Often, individuals did not receive the services they needed, but rather the services they were entitled to based on how or when they acquired their disability. As a result, there was an increasing reliance on ageing carers, a lack of permanent and respite accommodation, and reduced employment and educational opportunities. Individuals with disabilities and their families were often isolated and financially disadvantaged. In March 2013, legislation was passed in Australia to establish a National Disability Insurance Scheme, a radical new way of funding disability services. No longer would funding be directed to agencies, but rather to individuals who would make their own plan and select their preferred services and service providers, giving them more control over the services and supports they receive. The hope is that this change from a welfare-driven to an insurance-based model will improve equity of service delivery, levels of participation, and overall quality of life among Australians with disabilities and their families.

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Wiley Online: http://onlinelibrary.wiley.com/doi/10.1111/dmcn.13007/abstract

14. Dev Med Child Neurol. 2016 Jan 14. doi: 10.1111/dmcn.13015. [Epub ahead of print]

Congenital anomalies in cerebral palsy: where to from here?

McIntyre S, Blair E, Goldsmith S, Badawi N, Gibson C, Scott H, Smithers-Sheedy H; Australian Cerebral Palsy Register Group.

Proportions of cases of cerebral palsy (CP) with congenital anomalies recorded in Australian CP registers range from 15% to 40%. The anomalies seen in CP are extremely variable. We have identified that CP registers often do not have quality data on congenital anomalies, necessitating linkage with congenital anomaly registers. However, a lack of unified processes and definitions in congenital anomaly registers and data collections means that linkages are complex, need to be carefully planned, and limitations acknowledged. Historically in CP research, congenital anomalies have been classified by International Classification of Disease codes, then combined into brain and other major and minor anomalies. Systems have been developed to classify congenital anomalies into aetiologically related groups, but such a classification has yet to be trialled in CP. It is anticipated that primary prevention of a small proportion of cases of CP is possible through the primary prevention of congenital anomalies, especially those due to teratogens. Owing to the anticipated low prevalence of each subgroup, global collaboration will be required to further these lines of enquiry.

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15. Dev Med Child Neurol. 2016 Jan 14. doi: 10.1111/dmcn.13014. [Epub ahead of print]

Cerebral palsy and perinatal mortality after pregnancy-induced hypertension across the gestational age spectrum: observations of a reconstructed total population cohort.

Blair E, Watson L; Australian Cerebral Palsy Register Group.

AIM: Pregnancy-induced hypertension/pre-eclampsia (PIH/PE) is associated with cerebral palsy (CP) in term births but if sufficiently severe to necessitate preterm delivery predicts a lower risk of CP than observed in gestational peers. We investigated whether this apparent 'protection' was attributable to inappropriately chosen comparison groups and/or an increased risk of perinatal death. METHOD: Perinatal information was collected from medical records of children with CP, individually matched neonatal survivors without CP, and representative samples of perinatal deaths of Western Australian birth cohorts from 1980 to 1995. Compared with these data, the sensitivity of

statutorily collected PIH/PE data was assessed for each outcome group. Using these sensitivities, the estimated risks of death and CP in births to all women with and without PIH/PE were compared. RESULTS: Sensitivity of statutory PIH/PE data decreased with increasingly poor outcome. Reconstructed cohorts showed that PIH/PE increased the risks both of CP and of perinatal death in births at lower gestations except in births <27 weeks, where the risk of perinatal death only increased greatly. INTERPRETATION: PIH/PE does not protect against poor outcome at any gestational age. Previously reported protective effects originate from inappropriate control for gestational age and not from higher gestation-specific perinatal mortality.

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