Session B: Eclectic

B:1 Atlantoaxial instability in Down syndrome

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Introduction: Joint laxity is reported as one of the main features of Down syndrome. C1–C2 instability is considered as its major and most dangerous consequence. The purpose of this paper was to undertake a thorough radiological description of C1–C2 anomalies in this disorder, and to evaluate the factors associated with an increased risk of instability and/or neurological compromise.

Material and method: As part of a national survey on the medical problems of Down syndrome we conducted an evaluation of the orthopaedic aspects of this genetic abnormality with a particular emphasis on the upper cervical spine. Four hundred and fifty-eight consecutive patients were enrolled for the purpose of the study. Exhaustive questionnaires as well as a thorough physical examination, including a neurological assessment (even the mildest signs), an analysis of the laxity profile according to Carter and Wilkinson, and a detailed orthopedic evaluation were performed by the same specialized examiner. Patients were divided into two different categories depending on the presence or absence of neurological signs, and in two groups depending on the presence or absence of a generalized laxity (as defined by Carter and Wilkinson). Lateral radiographs of C1–C2 were taken in neutral, hyperflexion and hyperextension positions for each patient, by the same x-ray technician and using the same machine, following a standardized method inspired from Singer et al.’s with modification, taking into account the magnification factor.

A simple blind interpretation of radiographs was done by the same radiologist and included screening for any congenital or degenerative abnormality, and calculation of the C1–C2 distance, the minimal sagittal diameter (MSD) (representing the space available for the spinal cord), and the C1–C2 angle (previously unreported in the literature and specially described for this study). Results were then compared to the literature values when available, and correlated to age, gender, neurological signs, and laxity profile. Seven patients were excluded from the study for lack of compliance during x-rays, and 9 for insufficient clinical or radiological data. Statistical analysis was performed on the remaining 442 patients, using the Pearson test and ANOVA parametric method with significance for p<0.05.

Results: There were 184 females and 258 males with an average age of 13.8 years. Minor neurological signs were found in 42% of the patients without any major deficit. A generalized laxity was found in 24% of the cases. Other orthopaedic problems most commonly at the feet were found in 85% of the cases. A great variation in radiological measures was found, the reason why we decided not to present averages. The C1–C2 distance was >4mm on hyperflexion films (reported limit for instability) in 34 cases. The greatest C1–C2 distance was 8mm in neutral position and 9.6mm in hyperflexion. The lowest value of MSD was 8mm in hyperflexion and 10mm in neutral position (14mm being reported as the lowest limit for spinal cord safety regardless of age). The widest range of values was found for the C1–C2 angle. Generalized laxity and C1–C2 distance are inversely proportional to the patient’s age. There is no significant relationship between C1–C2 instability (C1–C2 distance > 4mm) and gender or generalized laxity, and between C1–C2 instability or the generalized laxity and neurological deficit.

Discussion: This paper involves a large patient population, confirms some of the data already reported in the literature and brings out new information about Down syndrome. Based on a standardized radiographic technique that also takes into account the magnification factor, it offers a real assessment of the C1–C2 relationship and raises some questions about the definition, the etiology, the limit of instability, as well as its correlation with the laxity profile and the neurological picture.

B:2 Spinal deformity following selective dorsal rhizotomy

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Objectives: To determine the prevalence of spinal deformity in children with spastic cerebral palsy (CP) treated by selective dorsal rhizotomy with laminoplasty.

Design: A retrospective clinical and radiographic review.

Setting: Referral center.

Participants: Seventy-nine consecutive children (47 male, 32 females) with a mean age of 6.2±1.8 years at rhizotomy were studied. All but one was ambulatory (64 community ambulators), and 43 required no assistive devices.

Measurements and main results: A detailed chart review was completed, and upright PA and lateral radiographs were reviewed. The mean radiographic follow-up was 4.2±2.0 years, and the mean clinical follow-up was 5.8±2.4 years. Pre and post rhizotomy sagittal spine variables were analyzed for sitting films, standing films, and finally for those patients who had sitting or standing radiographs both pre and post rhizotomy.

Rhizotomy decreased spasticity (p<10^-6) and led to small but significant improvements in both lower extremity muscle strength (range for 5 muscles: p=0.003 to p<10^-6) and joint range of motion (p=10^-4) to p=10^-6). None of the patients had scoliosis or spondylolisthesis before rhizotomy. Scoliosis occurred in 13 children (17%), and the average magnitude was 16±4° (range 11–24°). There was no significant difference between preoperative (35° sitting, 38° standing) and follow-up (33° sitting, 33° standing) thoracic kyphosis, or between preoperative (17° sitting, 45° standing) and follow-up (18° sitting, 45° standing) lumbar lordosis. However, there was a significant difference in lumbar lordosis between sitting and standing radiographs (p<10^-6). No progressive or rigid hyperlordotic lumbar deformities were observed. Spondylolisthesis was identified in 9 diplegic children (12%; 8/9 grade I), one of whom required a lumbosacral arthrodesis for pain. The mean
Acknowledgement:

Results:

Method:

Participants:

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Spondylolisthesis was correlated with greater lumbar lordosis ($p=0.03$), stronger hip abductors ($p=0.01$), and increased popliteal femoral angles ($p=0.03$) preoperatively, and with stronger hip flexors postoperatively ($p=0.02$). Back pain was identified in only four patients at follow-up, two of whom had spondylolisthesis.

Conclusion: Both scoliosis and spondylolisthesis were observed following selective dorsal rhizotomy in an ambulatory population with no prior deformity, and longer term follow-up is required to determine the clinical significance of these findings. A rapidly progressive or rigid lumbar hyperlordosis was not observed. We recommend periodic radiographic screening, as spondylolisthesis may be asymptomatic. The etiology of spondylolisthesis is likely multifactorial, especially as anatomic reconstruction of the posterior elements by laminoplasty did not appear to be protective in this ambulatory population.

B:3
Outcome following simultaneous late neurolysis, shoulder reconstruction, and/or bypass grafting after upper brachial plexus birth trauma

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Objective: After 6 to 8 months of age, surgical intervention for the shoulder paralysis and deformity in children with an upper brachial plexus injury has generally been limited to contracture release and muscle transfers. This report discusses another treatment strategy.

Participants: Over a 4-year period, 25 children have been first evaluated after 10 months of age (range 10 to 22 months) for poor recovery following an upper (C5/C6) brachial plexus injury following birth trauma. Eighteen had a fixed internal rotation contracture of the shoulder of at least 45 degrees.

Method: All children underwent microsurgical neurolysis. In 18 children, the internal rotation contracture was released by subscapularis slide. Where instability existed following restoration of full passive mobility, a posterior capsular tightening was performed. In 23 children, bypass grafting into the suprascapular nerve using proximal and distal end-to-side repairs was performed. Donor nerve included both C5 and the spinal accessory nerve. All patients underwent intraoperative botulinum toxin injection to the pectoralis major and latissimus dorsi muscles.

Results: All patients have been evaluated at least 18 months following surgery. Based on the criteria of Gilbert and Tassin, all children advanced one grade or more in shoulder function. No child lost motor function. A parental questionnaire showed 100% satisfaction with outcome.

Conclusion: Children seen late (10 to 24 months) with upper brachial plexus injuries associated with both persistent paralysis and shoulder deformity benefit from late multidisciplinary reconstruction.

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functional before surgery; therefore effect size was smaller, but shown by regression equation to be greater than would have been expected without treatment. PODCI expectations were met by MT surgery with a negative trend in the Neurosurgery groups. Ceiling and/or basement effects were seen in the GMFM and PODCI respectively for some patients. Conclusion: GMFM limitations of ceiling effect and questioned relevance to activity and participation are nicely addressed by the PODCI. While GMFM total changes were similar for the MT and SDR groups, the PODCI appeared to be more responsive to the type of change in the orthopaedic surgical group, which may reflect its initial intent to measure musculoskeletal change, but may not be as useful for more impaired individuals, or those undergoing a neurosurgical procedure. The GMFM regression equation allows for a valid comparison of treatment effectiveness across non-randomized or non-matched groups of participants, and similar normative data would be a useful adjunct to interpretation of the PODCI Scores.

B:5
Electrical stimulation for muscle strengthening in cerebral palsy: a randomized placebo-controlled trial
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Objectives: To investigate the efficacy of neuromuscular electrical stimulation (NMES) and therapeutic electrical stimulation (TES) in strengthening the quadriceps muscles in children with bilateral cerebral palsy (CP).

Design: A randomized placebo-controlled trial.

Setting: Home-based treatment, assessments in a regional hospital physiotherapy department.

Participants: A volunteer sample of 63 participants, aged 5–16 years with spastic CP affecting both legs were screened. Three children did not commence treatment: one due to surgery and two due to school family commitments. Six children withdrew from the trial for medical and/or personal reasons.

Method: Sixty participants were randomized by minimization on the basis of age, sex and quadriceps strength, to one of three groups: NMES (n = 18), applied for 1 hour per day at maximum tolerable intensities; TES (n = 20), applied at a threshold intensity level (<10mA) for 8 hours at night; or placebo (n = 22) applied for 8 hours at night. Electrode placement on the quadriceps muscles and treatment parameters were standardised in the TES and NMES groups; frequency = 35Hz, pulse duration = 300ms, on time = 5s, off time = 12s. Treatment was carried out 5 days per week for 16 weeks.

Measurements and main results: Peak torque (PT) of the quadriceps, the Gross Motor Function Measure (GMFM) and the Lifestyle Assessment Questionnaire (LAQ-CP) were used as outcome measures, and recorded by a blinded assessor at baseline, end of treatment and 6 weeks follow-up. Statistical analysis utilised the Kruskal-Wallis test with non-parametric data and repeated measures ANOVA with parametric data. No statistically significant effect was evident between the groups over time. Mean differences and standard deviations for the treatment phase are shown in Table B:5. The initial sample size calculations for this study were based on an effect size (for the interaction) of 0.3, based on previous literature on resistive strengthening techniques. The actual effect sizes in the data ranged from 0.07 (for least affected leg PT) to 0.1 (for LAQ-CP).

Conclusion: No statistically significant effect was identified between the groups. Further study with greater subject numbers and comparative work with resistive strengthening is warranted.

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B:6
A study of modified Constraint Induced Movement Therapy involving play for young children with hemiplegic cerebral palsy
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Objective: To measure the change over time in hand function after intensive physiotherapy using play for the affected upper limb whilst restricting use of the unaffected upper limb in children with hemiplegic cerebral palsy (CP).

Design: Single case experimental design with the children acting as their own controls. The children were involved in the study for a 12-week period and were assessed at entry to the study and at four weekly intervals. Each child had a four-week baseline period (A) with no hand treatment to control for maturation. They then had a four-week treatment period (B) followed by a four-week follow-up period (C) with no treatment. During the treatment period, the children attended twice weekly for a one-hour session consisting of a structured programme of play activities and were given a short home programme of play activities for the non-treatment days. Verbal instruction and gentle restraint, without the use of splints, were employed to restrict use of the affected upper limb.

Participants: Nine children presenting with hemiplegic CP were involved in the study. There were six males and three females, median age 31 months, ranging from 21 months to 61 months. The ratio of right to left hemiplegia was five to...
The children were recruited from the local child development centre.

**Measurements and main results:** Changes in hand function were assessed using the Quality of Upper Extremity Skills Test (QUEST) (De Matteo et al 1993) and the researcher (CN) carried out all the assessments. The children were seen to improve throughout the study, but the greatest improvements were seen immediately following the four-week treatment period (B). Statistical significance was found using the Wilcoxon signed rank test at the 1% level immediately following treatment and at the 5% level during the baseline period and the follow-up period.

**Conclusion:** The results of this pilot study suggest that intensive physiotherapy using play for the affected upper limb, whilst restricting use of the unaffected upper limb, may be an effective way of treating children with hemiplegic CP. The children tolerated the treatment well and showed both functional improvements and increased use of the affected upper limb. Future work is planned to consolidate and develop this study.

**Acknowledgements:** This study was supported by the Royal Berkshire and Battle Hospitals NHS Trust, Reading, Berkshire, UK.

**References**
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**Session C: Botulinum Toxin A**

**C:1 Accuracy of intramuscular injection of botulinum toxin A: a comparison between manual needle placement and placement guided by electrical stimulation**

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**Objective:** To evaluate the accuracy of needle placement before injection of Botulinum Toxin A, in the management of spasticity in children with spastic cerebral palsy (CP) — a prospective, clinical trial.

**Setting:** Tertiary referral center.

**Participants:** Two hundred and twenty-six children with spastic CP, who received injections of BTX-A, in the context of focal, regional, or multilevel spasticity management.

**Method:** Children who were scheduled to receive intramuscular injection of BTX-A as part of their management of spasticity associated with CP were invited to participate in the study. Informed written consent was obtained from the parents or guardians. Target muscles for BTX-A injection were identified from presenting symptoms, findings on clinical examination, video recordings of gait and in selected participants, and instrumented motion analysis. The target muscle was identified by manual palpation and known anatomical landmarks. With the child under mask anesthesia, an insulated 27G needle (Allergan USA) was inserted into the muscle belly and the position verified by moving the distal joint through a range of motion and observing reciprocal movement of the needle in the opposite injection. The needle was then attached to a stimulator (Stimlocator, Braun). A low current was applied in a TO4 (train of four) setting and the limb was observed for contraction of the muscle belly and movement of the distal joint. The result was recorded as accurate when manual placement was confirmed by electrical stimulation and inaccurate when no contraction was observed and the needle had to be replaced into contractile tissue.

**Measurements and main results:** Two hundred and twenty-six children received 1372 injections into 492 muscles during the study period. Accuracy of needle placement was 78% in the gastrocnemius, 68% in the hip adductors, 46% in the hamstrings, and only 12% in tibialis posterior. In the hemiplegic upper limb the figures were 62% in the biceps brachii, 32% in adductor pollicis, 22% in pronator teres, 16% in FCU, and 12% in FCR.

**Conclusion:** Many variables may affect the outcome of spasticity management by intramuscular injection of BTX-A. Accuracy of needle placement has not been previously investigated or reported in this population. Our results show that needle placement after manual palpation is reasonably accurate for large subcutaneous muscles, including the gastrocnemius and biceps brachii. Manual needle placement is not accurate for smaller, deeply placed muscles. The diffusion characteristics of BTX-A in vivo are not well defined but more consistent clinical outcomes start with accurate identification and injection of the selected target muscle.