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Management of the Ataxias: towards best Clinical Practice

Physiotherapy supplement

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Introduction

This document aims to provide considered guidance to physiotherapists working with people with progressive ataxias. It is a supplement to the Ataxia Guidelines produced by Ataxia UK 'Management of the ataxias: towards best clinical practice', to which readers are referred for more general information on progressive ataxias and their management. This document may also be of value for physiotherapists working with people with non-progressive ataxias.

The authors have systematically examined the published literature in developing this guidance. There are few randomised controlled trials to draw upon to inform the treatment and management of ataxia, or the role of the physiotherapist. The majority of intervention studies reviewed were small case studies or case series designs. Furthermore, little research has been conducted with people with spinocerebellar or Friedreich's Ataxia (FA). Most research about ataxia rehabilitation has been carried out with people who have multiple sclerosis (MS), or extrapolated from heterogeneous studies of participants with MS, brain injury or children with cerebral palsy.

Seven databases (CINAHL, Psych Info, Pubmed Central, British Nursing Index, AMED, EMBASE, SCOPUS), the Web of Knowledge and the Cochrane Data Base of Systematic Reviews were searched (1980-2009) using uniform terms (physiotherapy or physical therapy and ataxia; rehabilitation or exercise or training and ataxia). Reference lists of identified papers were hand searched. The criteria for inclusion were intervention studies (including case studies), opinion pieces or reviews primarily about ataxia and the role or efficacy of physiotherapy. With heterogeneous groups of participants, the papers were sifted to determine whether findings from those with ataxia could be extrapolated from group findings. Forty studies were included in the review. No qualitative studies were identified. Supplemental information pertinent to the role of the physiotherapist but not identified through the literature search was included by consensus. Three independent physiotherapists developed the following clinical practice guidance based on their review of the identified papers and clinical experience. Disagreements were resolved and consensus achieved through discussion.

Guidance rarely produces definitive answers (Scalzitti, 2001) and perhaps in this case, can only help to reduce some of the uncertainty involved in clinical decision-making process. It is anticipated that readers will integrate information provided in this document with their clinical experience and expertise in conjunction with patients' preferences and goals. To help determine the similarity between individual patients and study participants, evidence tables of reviewed papers have been produced (see Appendix A). Summary scores of methodological quality are also available (Appendix B). All papers were scored for methodological quality based on the quantitative review form produced by Law et al (1998) and following the system used by Martin et al (2009). The reviewers scored papers independently and the final agreed score was achieved through consensus. Each paper was rated according to the hierarchy of evidence used in the NICE MS clinical guideline (2003) and adapted from Eccles and Mason (2001) (see Appendix A).

Framework: Compensatory and Restorative Approaches

Anecdotally, rehabilitation for people with ataxia seems to have been largely underpinned by a compensatory approach. Bastian et al (1996) suggested individuals with cerebellar lesions have difficulty controlling interaction torques during multijoint movements. Bastian (1997) went on to reason that treatment should focus on teaching strategies to simplify movement by reducing the number of moving joints and by stabilising against inertial effects of limb movement. In other words, ataxia may be treated by teaching individuals to avoid rapid multi-joint movements and to favour slow, single joint movement, a so called compensatory approach. In accordance with this understanding of ataxia it would seem some people with ataxia (perhaps particularly those with severe upper limb tremor, see for example Gillen, 2000) may benefit from reducing the degrees of freedom and adopting compensatory techniques to maintain functional performance.

However Panturin (1997) has questioned whether such strategies would be as valuable for patients recovering from acute cerebellar injury. Indeed recent years has seen a growing body of evidence from the field of neurophysiology that has advanced the understanding of function and dysfunction of the cerebellum in motor control and motor learning (for example Molinari et al, 1997; Doyon et al, 2003; Stolze et al, 2002; Morton and Bastian, 2003, 2007; Ioffe et al, 2007; Ilg et al, 2007). Building on this work, theories have developed about possible responses of the cerebellum at a neural level (neuroplasticity) to respond to rehabilitation interventions (for example Lacourse et al, 2004). This evidence has lent support to the clinical impression that motor learning is possible following damage to the cerebellum (Boyd and Winstein, 2004; Ioffe et al, 2006) and thus gives weight to a more restorative approach to the rehabilitation of people with ataxia.

Improvements in people with chronic and progressive conditions have been reported in some of the higher quality studies reviewed in this guidance (Armutlu et al, 2001; Bailliet et al, 1987; Brown et al, 2006, Brown et al, 2005; Cernak et al, 2008; Gialanella et al, 2005; Gill-Body et al, 1997; Smedal et al, 2006 and Vaz et al, 2008); thus lending support to a more optimistic view of the potential response of the cerebellum to rehabilitation.

It is envisaged that physiotherapists will employ a combination of restorative and compensatory approaches guided by the patient's clinical presentation and context. This guidance aims to assist physiotherapists in this clinical decision making process.

Guidance

1. Systematic Reviews

One systematic review of nine studies investigating the effectiveness of physiotherapy for adults with cerebellar dysfunction was identified (Martin et al, 2009). The authors commented that the lack of detailed descriptions of interventions made it impossible to accurately document the treatment strategies used. A wide range of outcome measures were used which did not discuss validity and reliability for the population under investigation, long term effects were not investigated in the majority of papers and most studies were small case studies which limited the generalisability of results. All papers (Armutlu et al, 2001, Balliet et al, 1987; Brown et al, 2006; Foltz and Sinaki, 1995; Gialanella et al, 2005; Gill-Body et al, 1997; Jones et al, 1996; Karakaya et al, 2000; Perlmutter and Gregory, 2003) from this systematic review are included in this guidance, and appear under the relevant section.

The methodological quality of the systematic review was scored at 29/42 using the Quality of Reporting of Meta-analyses checklist (Moher et al, 1999). Limitations of the review were identified in the search strategy and review process.

Summary

- *The authors concluded there was modest evidence to support the effectiveness of physiotherapy with respect to gait, trunk control and activity limitations but insufficient evidence to support the efficacy of one specific intervention.*
- *Further research based on a robust understanding of neuronal plasticity is required and which addresses the methodological limitations identified in the review.*

2. Rehabilitation Approaches

Thirteen papers were identified for review (Armutlu et al, 2001; Balliet et al 1987; Brown et al, 2006; Dordal, 1989; Gialanella et al, 2005; Gill-Body et al, 1997; Gillen, 2000; Harris-Love et al, 2004; Jones et al, 1996; Karakaya et al, 2000; Perlmutter and Gregory, 2003; Smedal et al, 2006 and Stoykov et al, 2005). Findings supported those reported by Martin et al (2009). Most studies (7) were either small case studies or single case experimental designs with only one randomised controlled trial (Armutlu et al, 2001). The methodological quality score ranged from 4 to 11 out of a maximum 16. Participants had wide ranging cerebellar pathology including multiple sclerosis, head

injury, cerebellar stroke, brain tumour, cerebellar degeneration and central vestibular dysfunction; one child had Friedreich's ataxia.

Interventions were individually tailored for all studies except Balliet et al (1987), varying in type, intensity, duration and frequency. Commonly reported interventions included PNF, Frenkel's exercises, dynamic training of postural stability with task and activity focus, gait and balance training, along with strengthening and flexibility. Therapeutic equipment was often provided to support function. With the exception of Balliet et al (1987), Gill-Body et al (1997) and Gillen (2000) the majority of studies did not describe the intervention in detail and thus would be difficult to replicate in practice. A wide range of outcome measures were used and none of the papers fully reported the validity and reliability for this patient population. The long-term outcome was not consistently reported.

As with Martin and colleagues (2009) studies with a methodological quality score of ≥ 8 were considered of sufficient rigor to draw limited conclusions about the efficacy of physiotherapy. Four studies fell into this category (Balliet et al, 1987; Brown et al, 1987; Gill-Body et al, 1997 and Stoykov et al, 2005), Gialanella et al, (2005) was excluded because this was not an intervention study. Although they were small studies they provide some evidence in support of physiotherapy being able to improve gait, balance and trunk control for people with ataxia and reduce activity limitations and support increased participation.

Summary

- *Dynamic task practice that challenges stability and explores stability limits and aims to reduce upper limb weight bearing seems an important intervention for people with cerebellar dysfunction to improve gait and balance.*
- *Strength and flexibility training may be indicated in conjunction with the above.*
- *A compensatory approach (which includes orthotics and devices, movement retraining, reducing the degrees of freedom and optimising the environment) seems valuable for teaching people practical, every day strategies and ways of managing the condition and may be particularly important for those with severe upper limb tremor.*
- *Documentation of a full taxonomy of contemporary approaches for treating people with ataxia would provide a useful basis for further investigation in this field.*
- *Development / adoption of valid and reliable outcome measures for this population would improve methodological rigor and interpretation of research.*

3. Specific Interventions for Gait

3.1 Treadmill Training

Three studies investigated the effect of treadmill training for individuals at least one year post brain injury and presenting with ataxia. This intervention has not been explored for people with progressive ataxias. All studies were of good methodological quality (range 10-13/16) and included one RCT (Brown et al, 2005) and two case studies (Cernak et al, 2008 and Vaz et al, 2008). Training varied in duration, frequency, intensity (minimum 20 minutes three times a week for four weeks, to a maximum of daily training for 5 months). One study (Cernak et al, 2008) combined treadmill and over ground training. All studies used a combination of reliable gait parameters and functional outcome measures. Improvements were reported for all studies, the most functionally meaningful improvements were described by Cernak et al (2008) who combined over ground training with body weight support treadmill training at an intensity and duration significantly greater than the other studies.

Summary

- *Findings from treadmill training studies present encouraging evidence of the efficacy of this intervention for people with ataxia due to brain injury. Intensity and duration of training seem to be significant factors.*

- *Consistent intensive training over many months combined with over ground training may be required.*
- *Further research required to determine whether treadmill training is beneficial to those who have ataxia as part of a long-term progressive condition.*

3.2 Visually Guided Stepping

Oculomotor and locomotor control systems interact during visually guided stepping i.e. the locomotor system depends on information from the oculomotor system during functional mobility for accurate foot placement (Crowdy et al, 2000).
Crowdy and colleagues (2002) demonstrated, in two participants with mild cerebellar degeneration, marked improvements in oculomotor and locomotor performance following eye movement rehearsal. The authors suggest that rehearsal of intended steps through eye movement alone i.e. looking at foot target placement for each step, before negotiating a cluttered room, might improve performance and safety.

Summary

- *This simple strategy, which although task specific and short lived in nature, is promising and relatively quick and easy to apply in a functional setting.*

3.3 Balance and Mobility Aids

No studies have specifically evaluated the role of balance and mobility aids for people with ataxia. Clinical experience suggests walking aids should be considered on a case-by-case basis. Jeka (1997) reviewed a series of studies on postural control using light touch contact of fingertips or a walking aid as a means of balance. Results showed somatosensory cues from the fingertips provided a powerful reference orientation even when contact force levels were inadequate to provide physical support for the body. Clinical observation suggests that some individuals with ataxia find light touch contact more useful as a strategy than a conventional walking aid. This may explain why some people prefer to use Nordic poles, which help encourage light touch contact, rather than traditional walking sticks that tend towards force contact and a reduction in muscular forces acting through the lower limbs. Decreasing dependency of weight bearing through the upper limbs in people with ataxia is also supported by Balliet et al (1987). Furthermore individuals with cerebellar hemisphere lesions, who are more likely to have dysmetria and tremor, may find balance and mobility aids hard to use because placing and controlling a stick can be as difficult as trying to accurately place legs during swing phase. In a small study of healthy young adults walking aids have been found to compromise the ability to respond to balance disturbances through impeding lateral compensatory stepping and thus can affected safety (Bateni et al, 2004).

Summary

- *Light touch as a balance aid may be helpful for postural orientation and stability*
- *Upper extremity weight bearing during ambulation may perpetuate a deterioration or worsening of gait parameters*
- *Careful assessment is required for those with dysmetria*

3.4 Axial Weighting

The effect of weighting the axial skeleton been studied in five subjects with ataxic gait of unreported aetiology (Clopton et al, 2003) and 19 participants with mixed CNS pathology five of whom had ataxic gait (Foltz and Sinaki, 1995). Folz and Sinaki (1995) reported subjective improvements in gait and posture plus feelings of steadiness, but low methodological quality means findings should be viewed with caution. Conversely Clopton et al (2003) reported gait characteristics changed unpredictably with axial weights, worsening more often than improving, and concluded that use of axial weights to improve gait for patients with ataxia was not supported (Clopton et al 2003).

A case study by Perlmutter and Gregory (2003) reported that a 5lb weighted vest, used as part of a general rehabilitation programme for a woman with severe cerebellar ataxia, was a useful adjunct

when the patient was carrying out reaching activities in sitting. It is unclear how much the weighted vest contributed to overall improvements, therefore similar use would need to be evaluated on an individual basis.

Summary

- *A very limited theoretical basis for axial weighting and no evidence to support use of axial weights to improve gait in people with ataxia.*

3.5 Lycra Garments

Adults

A pilot proof of principle single case experimental design ($n = 6$) was undertaken at the University of East Anglia and led by Dr Martin Watson. A six week base line period was followed by a six week intervention phase using custom made lycra shorts, worn for between eight to ten hours a day. Repeated measures of postural sway, walking effort and speed were taken during all phases including a six week follow up. Results were mixed; some participants benefitted in certain aspects of functional movements, which indicates the need for further studies to identify whether findings can be replicated and if the garments are more useful for some individuals than others. Full findings are yet to be published but a summary can be found at www.ataxia.org.uk. For further information contact M.Watson@uea.ac.uk

Children

Three studies were reviewed that investigated the effect of lycra garments in children (Blair et al, 1995; Corn et al, 2003; Nicholson et al, 2001). Two studies had good methodological quality (Corn et al, 2003 and Nicholson et al, 2001) but lacked a control group and conclusions should therefore be viewed with caution. Likewise studies were small, different types of lycra garment were compared within and across studies with samples of heterogeneous participants. As such, results of these studies are highly variable across participants and beneficial effects were slight or of limited duration. Blair et al (1995) and Nicholson et al (2001) reported significant adverse effects.

Summary

- *Insufficient data is available to support the use of lycra garments for children or adults with ataxia.*

3.7 Biofeedback

Baram and Miller (2007) investigated the effect of auditory biofeedback for 14 people with MS and gait dysfunction due to cerebellar ataxia. Positive results were reported in terms of speed and step length but the real world benefits were not addressed and it seems that some participants benefitted whilst others deteriorated. The findings were therefore inconclusive and further investigation is warranted.

4. Specific Interventions for Upper Limb Tremor

Lesions affecting the cerebellar hemispheres give rise to ipsilateral limb symptoms including tremor in addition to dysynergia, disdiadochokinesia and rebound phenomenon. An *action tremor* occurs during movement i.e. is produced by voluntary contraction of muscle and includes; *postural tremor* (occurs when voluntarily maintaining a position against gravity e.g. holding an arm out straight) and k*inetic tremor* (occurs during any type of voluntary movement). Kinetic tremor is further subdivided into: simple kinetic tremor: occurs during voluntary movements that are not target-directed (e.g. flexion/extension or pronation/supination), and intention tremor, which occurs during target directed, visually guided movements (e.g. finger-nose test), and worsens at the terminal phase of the movement as the target is approached (Deuschl et al, 1998).

In addition to affecting activities of daily living (Feys, et al, 2004) the psychosocial consequences of upper limb tremor can be significant (McGruder et al, 2003). The treatment of upper limb tremor via the action of pharmacological agents and physiotherapy remains wanting.

4.1 Manipulation of Visual Information

A clinical observation by Pope (2007) that closure of eyes whilst eating may assist in the control of upper limb ataxia has some support from experimental evidence. These studies with the exception of Beppu et al (1987) were conducted with people who had intention tremor as a result of MS. Results suggest that kinetic tremor improves if movement is not visually guided (Beppu et al, 1987; Sanes et al, 1988; Quintern et al, 1999), and dysmetria improves if visual feedback is manipulated (Feys et al, 2005a, 2006). Saccadic dysmetria was noted to frequently coexist with intention tremor and inaccurate eye movements are likely to impair accurate motor performance of the hand (Feys et al, 2003) and individuals with intention tremor or other cerebellar deficits had difficulty using visual information to control arm and hand movements (Feys et al, 2003).

Summary

- *Tremor amplitude may be reduced if target directed movements are performed from memory rather than under direct visual guidance (Sanes et al, 1988) or if the primary saccade and the hand movement to reach the object are performed separately (Feys et al, 2005a).*

4.2 Cold Therapy

Two studies (Quintern et al, 1999 and Feys et al, 2005b) reported functionally significant reductions in upper limb tremor following cooling of the upper limb in people with MS. Although both studies report improvements, there were differences in effect which might be related to the duration of cooling; 15 minutes compared to one minute. Several mechanisms have been suggested that may contribute to reported effects; a temperature dependent decrease in muscle spindle sensitivity causing a reduction in Ia afferent discharge and thus a reduction in response of the long latency stretch reflex (Quintern et al, 1999; Feys et al, 2005b), and a decrease in nerve conduction velocity with an increase in stiffness of cooled muscles (Feys et al, 2005b).

Summary

- *Transient tremor control using cooling could have important functional implications when performing discrete functional activities such as intermittent self catheterisation, signing documents, working a PC and taking a meal (Feys et al, 2005b).*
- *Deep cooling may be more effective than moderate cooling in individuals with severe tremor. Upper limb cooling in general may not be as useful for individuals who also have significant proximal tremor (Feys et al, 2005b).*
- *Further studies to assess the effects of cooling on functional tasks are warranted.*

4.3 Wrist Weighting

Investigation of wrist weighting as an intervention to reduce upper limb tremor stretches back several decades (McGruder et al, 2003; Feys et al, 2003; Manto et al, 1994; Sanes et al, 1988; Morgan et al, 1975; Langton-Hewer et al, 1972), there has also been one study that has used a potentially more sophisticated mechanical damping devise which as yet lacks clinical utility (Aisen et al, 1993). Findings from these studies are inconclusive partly due to methodological issues (quality score ranges from 3-9); inclusion of heterogeneous tremors in the same cohort, the use of differing weights and weighting systems plus various outcome measures without reports of reliability or validity. Beneficial, (Aisen et al, 1993; Langton Hewer et al, 1972, Morgan et al, 1975; McGruder et al, 2003), detrimental (Manto et al, 1994), and mixed (Sanes et al, 1988) effects were reported, along with findings of no difference except slowing of the transport phase of movement (Feys et al, 2003).

Summary

- *Evidence in this area is equivocal; it seems weighted wrist cuffs (of different weights) and weighted cutlery may be useful for some individuals under specific circumstances and should be assessed on a case-by-case basis.*

- *Based on McGrunder et al (2003) and clinical experience, exploration of the optimal weight required to support function should be assessed using different weights; under-weighted cuffs will not dampen tremor and over-weighted cuffs may be no better than no weight at all. Thus weighted wrist cuffs may be more suitable than weighted cutlery (refer to OT section in main guidance document) and should be assessed on an individual basis.*
- *As some individuals show exaggerated tremor for a short time on removal of weights, it is suggested that specific functions such as eating or writing are targeted. The long-term effects are not known; clinical observation suggests some people accommodate to the weight.*
- *Weighted cuffs may be too fatiguing or cumbersome to confer any functional or psychosocial benefit for some individuals, thus patient goals and perspectives are critical in assessing the value of this intervention.*

4.4 Relaxation and Biofeedback

Guercio et al (1997, 2001) describe two case studies of individuals with severe ataxic kinetic tremor following a TBI that used behavioural relaxation training with EMG auditory biofeedback to reduce tremor severity and improve functional performance. Findings reported benefits in functional performance and reduction of tremor.

Summary

- *This approach cannot be recommended for people with ataxia without further evidence but presents an interesting avenue for further research, in particular the development of task orientated biofeedback (Huang et al, 2006).*

5. Wheelchair Seating

Wheelchairs rank among the most important therapeutic devices used in rehabilitation and can make the difference between an active and efficient alignment and a postural catastrophe. Few studies have investigated the physiological and functional impact of postural supports such as specialist wheelchairs, which can present significant methodological challenges (Sprigle, 2004). This guidance document reviewed one paper of low methodological quality (Clark et al, 2004) that included four participants with FA. Findings were equivocal and further research is required.

Despite the lack of research studies, clinical observation suggests that power wheelchair mobility with appropriate postural support is an option to provide people with ataxia with a means of independent mobility. Power chairs may also help conserve energy that can then be used outside the wheelchair for carrying out activities of daily living in antigravity postures. Additionally an appropriate posture in the power chair may facilitate respiration and swallow in those patients who may be compromised in these areas. In the absence of other evidence, clinical experience and patients' needs should be used to guide clinical reasoning (Huhn and Guarrera-Bowlby, 2007).

6. Exercise

In general people with ataxia should be encouraged to exercise as part of health promotion and as long as risk factors and health and safety considerations have been assessed. Exercise should be tailored towards what appeals most to participants and may involve exploring several different options as well as building motivation and sustainability into the exercise prescription (Dean, 2009; Rhodes and Fiala, 2009)

Note of caution: Cardiac abnormalities are a common occurrence in people with Friedreich's Ataxia. Before embarking on an exercise programme potential cardiac problems should be excluded. Please refer to the cardiac problems section in the medical guidance of themain guidance document.

6.1 Endurance/aerobic Training

Fillyaw and Ades (1989) investigated the physiological adaptation to aerobic training in a 38 year old man with Friedreich's Ataxia. Training took place over a period of nine weeks and consisted of 27 ECG monitored, bicycle ergometer sessions of 20-25 minutes of continuous cycling at training level intensities. Prior to commencement of training the participant, who had no history of cardiac symptoms, underwent a cardiovascular examination and echocardiogram that confirmed normal cardiac size and function. Increases in cardiorespiratory and work measures demonstrated clinically important physiological adaptations to aerobic conditioning in this individual case. The authors suggest that for people without cardiomyopathy, aerobic training offers a means of promoting activity and reducing deconditioning which may provoke functional as well as other health benefits. However, these case study findings cannot be extrapolated to clinical practice without further controlled trials.

6.1 Other Forms of Exercise

6.1.1 Therapeutic Riding (Hippotherapy)

No studies have evaluated the effect of therapeutic riding (TR) specifically for people with ataxia. Hammer et al, (2005) investigated therapeutic riding using a single case experimental design (ABA) in 11 individuals with MS. Overall balance and SF-36 Role-Emotion dimension showed most improvements although TR seemed to have individual effects as would be expected with the heterogeneous nature of the participants. Similarly Silkwood-Sherer and Warmbier (2007) investigated the effect of TR on balance in 9 people with MS and reported significant improvements on the Berg Balance Scale and Tinetti Performance Oriented Mobility Assessment. Several studies have explored the effect of TR for children with cerebral palsy (for example Sterba et al, 2002, Sterba, 2007 and Snider et al, 2007), however these papers look at the effect of TR in heterogeneous groups of children with cerebral palsy.

Summary

- *No studies have investigated the effects of therapeutic riding for people with ataxia as the primary impairment.*
- *Therapeutic riding provided outside of the usual rehabilitation environment may have a positive impact on health related quality of life as well as physical functioning.*
- *Future work should investigate the physical as well as psychosocial effects of this potential therapeutic intervention for people with ataxia as the primary impairment.*

6.1.2 Hydrotherapy and Swimming

No studies directly evaluate the efficacy of hydrotherapy for people with ataxia. However anecdotal evidence supports the value of hydrotherapy for people with ataxia as a form of exercise. Cook (2007) advocates the use of hydrotherapy and swimming for people with ataxia because water activities offer risk and challenge, provide freedom of movement often not available on land and may be beneficial for speech. Hydrotherapy is also considered to offer beneficial effects on health related quality of life. Further studies are required to investigate assumptions concerning physiological and functional benefits.

6.1.3 General fitness training, Yoga and Pilates

Anecdotal evidence advocates the benefits of general fitness training, yoga and Pilates for people with ataxia to help maintain strength, flexibility and balance. Psychosocial benefits have also been reported. No studies directly investigating Pilates or yoga or similar forms of exercise were identified in the literature review but further investigation is warranted.

7. Specific Impairments

People with ataxia can experience a number of specific impairments which physiotherapists should be aware of. Clinical experience and feedback from people with ataxia indicates that fatigue can be a common and at times an overwhelming issue. Spasticity, contractures and dystonia, are also symptoms that can occur. Clinicians are referred to Section 4 of the Ataxia Guidelines main document and the MS Society Guidance for Physiotherapists (2008) for further direction about managing these symptoms. Bladder and bowel problems (such as frequency, urgency and incontinence) can also be a feature of the ataxias. For specialist advice and assessment referral to a gynaecologist or urologist may be required (see Section 4 of Ataxia Guidelines main document). For further advice refer to the Association of Chartered Physiotherapists in Women's Health (ACPWH) who provide assessment and treatment for men and women with bladder and bowel impairment. A referral to a continence nurse may be useful. Finally, neuropathic pain can be a feature of the ataxias (see section 4 of the Ataxia guidelines main document).

For information on palliative care see section 7 of the Ataxia Guidelines main document.

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Appendix A

Table 1.0 Rehabilitation

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Armutlu et al, 2001	RCT n=26 adults with PP or SPMS, predominantly ataxic, slight muscle weakness, walk unassisted. Control n =13 Intervention n = 13 Age range 23-45 years	Rx 3 days a week for 4 weeks Control: PNF (rhythmic stabilisations), Frenkel co-ordination exercises (prone, supine sitting, standing), PNF mat activities (crawling, kneeling, ½ kneeling), approximation antagonists/agonists, static & dynamic balance training (external perturbation, weight transfer both extremities, & to posterior parts of the feet in standing and feet together), training semi-tandem and tandem, single limb stance on balance board, Cawthorne-Cooksey, walking on uneven ground. Progression from eyes open to eyes closed. Intervention: a/a + 20 min JPS to both legs before physiotherapy and during crawling, kneeling, ½ kneeling & single limb stance.	Sensory assessment, SLST, step width, walking velocity (3m), Ambulation Index, anterior balance (Lovet-Reynold method), ECT, NECT, SSEP, MEPs.	Both groups improved sensory evoked potentials, ECT, NECT, gait parameters and EDSS.	MQS: 7 The only RCT in this field of research, it is of low methodological quality but demonstrated that people with ataxia as part of living with MS improved in balance and gait parameters following a short course of rehabilitation. JPS demonstrated no additional benefit compared to 'conventional therapy'.
Balliet et al, 1987	Case study n=5 men with cerebellar dysfunction 4=TBI, 1=cerebellar dysfunction following drug treatment for leukaemia. Age range 22-63 years 1-12 years post onset Able to stand and weight bear	Rx 1 hour twice a week for 3/12, progressing to x1 every two months for the remaining period. Case study III more intensive and described in detail. Prescribed programme to regain walking independence through reducing upper extremity weight bearing; progressing from simulated gait in sitting, to dynamic balance in standing and walking with reducing upper limb support. Targets set for each stage before progressing. + self-training programme which constituted 95% of total training.	Functional Rating Scale 4 categories: 1. Assistive device (8 point scale) 2. Amount of upper limb weight bearing (5 point scale) 3. Level of assistance walking (7 point scale) 4. Distance walked (m)	Improvements in all categories; progressed to a single assistive device (e.g. stick) or better to walk, stand-by level of assistance to walk or better, walk at least 300m.	MQS: 9 Intervention well described. Progressive balance and gait retraining may take many months if not years following TBI, intensive physiotherapy may not be required but ongoing supported practice may be important. Practice organisation and structure not described. Reliability and validity of outcome measures not addressed.

Abbreviations: MQS: methodological quality score, JPS: Johnstone Pressure Splints, SLST: single limb stance time, DLST; double limb support time, ECT: equilibrium coordination test (no. of footfalls outside a 10cm wide walkway), NECT: non-equilibrium coordination test (knee-heel test, pendular limb movement), SSEPs: somatosensory evoked potential, MEPs: motor evoked potentials, DHI: dizziness handicap inventory, ABC; activities-specific balance confidence, TUG; timed up and go, DGI; dynamic gait index, 5xSTS; five times sit to stand, PP; primary progressive, SP; secondary progressive, FA; Friedreich's ataxia, LE; lower extremity, UE; upper extremity, JTHF; Jebsen test hand function, FSS; Functional Systems Scale, FIM; functional independence measure, SCED; single case experimental design, BBS; Berg balance scale, SL; step length, RVGA; Rivermead visual gait assessment, VAS visual analogue scale, RMI; Rivermead mobility index, PGIC; patient global impression of change, CGIC; clinician global impression of change, FAC; functional ambulatory index, FTRS; Fahn's tremor rating scale; ICARS; international cooperative ataxia rating scale, CFSS; cerebellar functional systems scale, PEDI: paediatric evaluation of disability inventory, MAS; modified Ashworth scale, CRTS; clinical rating of tremor scale.

Table 1.0 Rehabilitation – *continued*

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Brown et al, 2006	Retrospective case series n=48, adults with central vestibular dysfunction, age range 17-90 years, duration of symptoms before intervention mean 33.6 mo, 22 patients used an assistive device.	Individually tailored, mean of 5 sessions, (range 2-12), Rx inc 1 or more of; balance and gait training, general strengthening and flexibility exercises, vestibular adaptation training for those with mixed peripheral and central disorders.	DHI, ABC, TUG, DGI, 5xSTS	Significant differences on all tests comparing pre and post test scores. Participants remained at risk of falling and had reduced confidence in their balance abilities.	MQS: 11 Participants with cerebellar dysfunction improved the least, but did improve. Good account of clinical importance of change. PT may be valuable for those with severe cerebellar problems.
Dordal, 1989	Case study n=2, age 22 and 23 years, TBI 6 & 10 years previously. One severe ataxia. Pt with ataxia unable to stand or walk without support, difficulty using wheelchair	Individually tailored 1: 7/12, 225 hrs of training ~ 8 hrs a week, 2: 10/52193 hrs of training ~ 20 hours a week. Training: dynamic graded exercise, stretching, indoor sports, 'special' walking exercises, mobility training (floor to stand, climbing over obstacles, moving objects, falling exercises), swimming, static bike, trike outdoors, task specific practice.	Video analysis, photographic anthropometric assessment of posture, bicycle ergonometry (heart rate), light track measure of simple movements, descriptions of functional tasks, quality of movement and posture.	Improvements in standing without support, functional walk (3m), 20m independent walk, floor-stand-walk, improved swimming and diving, improved quality and speed of movement.	MQS: 4 Demonstrated that 2 people post TBI could make long term improvements in functional activities with activity programmes that surpass usual physiotherapy dose. Low methodological quality.
Gialanella et al, 2005	Retrospective case series comparing the recovery following isolated cerebellar stroke to cerebellar stroke with extra-cerebellar involvement, n = 43, mean age 67 yrs, mean interval between stroke and admission to rehabilitation unit 27 days.	Bobath therapy mean of 300 minutes per week (5 days a week) until discharge.	Trunk control test (TCT), Lindmark scale (walking) and Rankin scale (disability). No significant differences between groups prior to intervention.	Functional recovery present in both groups, those with isolated cerebellar stroke improved more than those with cerebellar and extracerebellar involvement.	MQS: 9 Functional gains demonstrated in both groups, supporting previous studies.
Gill-Body et al, 1997	Case study n=2, female (1), 36 yrs old, 7/12 post surgical resection pilocystic astrocytoma, dizziness, reliant on vision, difficulty selecting appropriate sensory input for postural control, independent basic ADL. Male (2) 48 yrs, 10 year history cerebellar atrophy and cerebrotendinous xanthamatosis, difficulty with standing balance and walking.	Rx x1 week for 6/52, individually tailored programme emphasising practice of activities that challenged stability. Rx described in detail and reasoned based on assessment findings related to cerebellar dysfunction.	DHI, TUG, kinematic indicators of stability (gait speed, BOS, tandem stand, Romberg's test, stance duration, eyes open, eyes shut).	Improvement for both participants; decrease in frequency and intensity of disequilibrium, DHI improved (1), walking outdoors independently, driving (1), improved on some indicators or stability.	MQS: 8 Good case studies providing detailed description of key assessment findings and clinical reasoning supporting chosen interventions. Reliability and validity of outcome measures not described. People with long term cerebellar dysfunction can improve postural stability.

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Gillen, 2000	Case study, 31 yr old male with MS, admitted for rehabilitation with severe ataxia, unable to care for himself, severe upper limb tremor.	Custom OT and PT 90'/day, for 5/52, plus 10 days iv steroids. Tailored programme focussing on task specific training, with orthotics, environmental adaptation, adaptive equipment prescription and movement retraining.	FIM items and personal goals; Feeding, grooming, bathing, upper body dressing, lower body dressing, bladder and bowel management, sexual activity, instrumental activities of daily living.	Feeding, grooming, bathing, bladder management improved from 1-2 to 6 (FIM) indicative of less assistance, sexual activity and IADL goals were achieved.	MQS: 7 Detailed description of key assessment findings and clinical reasoning supporting interventions. Concluded that steroids dampened the tremor but techniques and devices were required to support and maintain function.
Harris-Love et al, 2004	Case study, 14 yr old girl with FA, assistance of 1 to stand, used powered wheelchair, walking frame	PT x1/12 for 60', plus PT x1 per quarter for 60', + 20-30' adapted PE/day. Rx: 'accommodative/restorative' UE bimanual task oriented training, LE stretching, functional strengthening (hip and trunk), gait training. Replaced rollator wheeled walking with U-step walking stabiliser (USWS: reverse break system and tension controlled wheels).	Manual muscle testing, passive ROM, 9HPT, SLST, gradation of force, gait speed, DLST, step length asymmetry, step time asymmetry	9HPT, SLST, manual muscle testing showed minimal changes. Gait speed decreased by 69.4% concomitant with a 43.7% increase in force variability. Provision of the USWS improved gait performance (speed and reduced falls).	MQS: 7 Concluded that force variability may be a clinically useful measure of ataxia in this population, USWS was a useful aid to extend walking ability for this child with FA.
Jones et al, 1996	Wait list controlled comparison study, n=62 (intervention n=36), adults with MS, mean age 36 yrs, moderate to severe ataxia UE and trunk. EDSS mean 7 (intervention), 6.5 (control).	30' OT and PT /day for 8 days Rx to promote normal posture & movement, weight bearing, joint approximation, co-contraction and compression, proximal stabilisation and dynamic balance (gym ball), damping and weighting, ADL equipment and advice,	FSS, EDSS, JTHF, Northwick Park ADL Index, VAS (patients and assessor) – fatigue and 3 areas of performance.	JTHF, EDSS and FSS did not change across groups. Northwick Park ADL Index significant changes between groups, VAS: significant improvement reported.	MQS: 7 Short course of PT and OT compared to no treatment improves functional activity.
Karakaya et al, 2000	Before and after comparison study, 2 groups, age 19-63 yrs, n=20 posterior fossa tumour, n=20 cerebellopontine angle tumour, score >40 points on Karnofsky performance scale, balance and co-ordination problems.	5 days/week for at least 2 weeks until discharge. Individually tailored Frenkel exercises, PNF, dysmetria, dysdiadochokinesia, balance, walking and stair training.	Mokken's 4 point rating scale of functional independence; sitting, crawling, standing both feet, standing one foot (R&L), Bohannon's standing balance score 5 point ordinal scale.	Both groups improved overall balance and standing balance ($p<0.05$). Those with cerebellopontine angle tumours made more improvement than the posterior fossa tumour group.	MQS: 7 Provides some evidence that balance and co-ordination improved in this population with rehabilitation. Those with more aggressive tumours (posterior fossa) had a less favourable rehabilitation prognosis.
Perlmutter and Gregory, 2003	Case study, 75 yr old woman, with paraneoplastic cerebellar degeneration. Unable to sit independently, wheelchair dependent, limited sitting tolerance. Severe upper limb ataxia.	3 weeks in patient individually tailored Rx, provision of tilt-in-space wheelchair seating system, sitting balance training with weighted vest, dynamic balance training, reaching with weighted cuff, PNF, tilt table standing.	FIM; bed mobility, sitting balance, sitting tolerance, transfers, standing balance, wheelchair management, ambulation.	FIM improved from 2 to 4 for transfers and from 1 to 4 for standing, from 1 to 5 for bed mobility and sitting balance. Discharged home instead of a nursing home.	MQS: 5 Case study demonstrating meaningful improvements following a relatively short intervention period.

Table 1.0 Rehabilitation – *continued*

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Smedal et al, 2006	SCED (ABAA) n=2, RRMS, able to walk for 10 minutes, EDSS 6.0: 48 yr old woman (1), diagnosed 20 yrs previously, ataxia and spastic paraparesis; 32 yr old man (2), diagnosed aged 17yrs, spastic paraparesis.	Individually tailored treatment one hour/day, five times a week for 3 weeks in the B phase. 1: postural stability and orientation, dynamic activities designed to experience movement through postural adaptation. 2: mobilisation of LE in preparation for weight bearing and facilitation of trunk control and activity.	BBS, gait parameters; velocity, SL, double stance phase as a % of gait cycle (at 3 speeds), TUG, RVGA, self report (VAS) perceived gait problem, Borg exertion scale, RMI, PGIC, CGIC. All assessments were conducted once a week in the four three week periods (ABAA); follow up was at six weeks after completion of the second A phase.	Both improved on TUG, BERG and RVGA and reported improvements in balance and gait, and scored their condition as much improved.	MQS: 11 Patient with ataxia: gait speed and other parameters improved but remained slow and did not approach norm values, improvements in the BERG and TUG were clinically important and clinically significant. Preliminary evidence.
Stoykov et al, 2005	Case study, 68 yr old woman three years post left midbrain haemorrhage, dependent for all ADL. Right hemiplegia, severely ataxic. Barthel ADL index = 0.	1 hour OT 3x week for four weeks. Progressive tailored programme included; training for sitting balance, passive ROM, rolling, dynamic balance activities in sitting, scapular strengthening exercises caregiver training. Lumbar support provided for the wheelchair.	Fugl-Meyer Upper Extremity Motor Scale (FMUEMS) Postural Assessment Scale for Stroke Patients (PASS)	FMUEMS improved from 35/66 to 53/66 PASS improved from 2/36 to 7/36 Barthel remained at 0 Increased time of comfortable sitting sufficient to support participation in chosen activities.	MQS: 6 Four week intervention period led to improvements in upper limb control 3 years post brain haemorrhage, no effect on level of assistance required for ADL. Preliminary level evidence only.

Table 2.0 Gait: Treadmill Training

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Brown et al, 2005	RCT n=20, aged 20-57 yrs, >6yrs post TBI, all able to stand or walk for 20', 15/19 minimal to severe ataxia and were evenly distributed to the intervention or control group.	Rx gait training twice a week for 14 weeks, 15' of walking practice during a 30' session. Control: conventional over ground training Intervention: body weight support treadmill training, body weight support gradually reduced from 30% to 10%, +/- physical assistance from up to 3 physiotherapists, gait speed increased as tolerated. No over ground practice.	Gait parameters – gait velocity, stride width, step length, functional reach, FAC, TUG.	No significant difference for either group for speed, FAC. Both groups improved on TUG and functional reach but not significantly, control significantly improved step length differential, both groups narrowed step width not significantly, no significant differences between groups. Trends towards significant improvement favoured the control group.	MQS: 12 Good quality study. Both groups improved but over ground training was more effective for improving gait symmetry. Authors suggested a longer training period and over ground training as a supplement BWSTT to promote carry over might be important considerations for future research.

Table 2.0 Treadmill Training – *continued*

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Cernak et al, 2008	Case study, nonambulatory 13 yr old girl with severe cerebellar ataxia following a brain haemorrhage 16 months previously.	Partial body weight support treadmill training (30%-10%), with overground practice, 40' 5 days/week for four weeks, followed by PWSTT daily practice at home and x2/week 90' PT at home for 4/12. Progressively increased speed 0.18-0.8m/s and reduced assistance.	Gillette Functional Walking Scale, Paediatric FIM transfers and mobility subscale, number of unassisted steps.	Gillette improved from some stepping with assistance to walking for household distances. Transfers improved from moderate assistance to modified independence Walking improved from maximum assistance to supervision No. of unassisted steps improved from 0-200-no assistance.	MQS: 10 Good case study, significant improvements were not seen in the short term. Months of consistent training and practice might be required. Overground training was combined with treadmill training from the start and may have been an important factor in functional carry over
Vaz et al, 2008	SCED (ABA) n=2, chronic ataxia following TBI, able to walk at least 10m independently; (1) 25yrs, ICARS (ataxia severity) 22/100, gait speed 0.51m/s; (2) 53 yrs, ICARS 60/100, gait speed 0.33m/s	4/52 treadmill training, 20' 3x a week for 4 weeks. Progressive increases in velocity and step length and reduced upper limb support.	Gait parameters (speed, cadence, step length), TUG, customised balance assessment, RVGA.	Improvements in gait speed leg speed, step length, rising from am chair and balance.	MQS:13 All gains started in the baseline phase and therefore could not be attributed to the introduction of treadmill training. Practice of functional tasks in during measurements in the baseline phase may have contributed to improved performance. A longer baseline period would be necessary to counter this effect.

Table 2.1 Gait: Visually Guided Stepping

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Crowdy et al, 2002	Before and after proof of principle study. N=2, cerebellar degeneration, aged 54 and 44 yrs.	Baseline recordings of eye movements and locomotor performance (5 walks to 18 footfall targets along a walk way). Intervention: verbal instruction to concentrate on making accurate eye movements to the footfall target rather than accurate steps plus eye movement rehearsal to first 6 targets. Followed by 3 further test walks.	Step phase duration (stance time, double support time), saccadic eye movement, % of double, triple or quadruple saccades.	Marked improvement in oculomotor and locomotor performance; increased regularity and accuracy of stepping and increased proportion of single saccades i.e. reduced saccadic dysmetria.	MQS: 9 Preliminary evidence that rehearsal of eye movements to pre-determined footfalls improved visuomotor performance in people with cerebellar dysfunction. Potential functional implications discussed with reference to everyday tasks that require visually guided stepping.

Table 2.2 Gait: Auditory Biofeedback

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Baram and Miller, 2007	Before and after study, n=14 people with MS, EDSS 3.5-6.0, CFSS 2.5-4.0 (mild to severe). N=11 controls.	Baseline 4x 10m walk Device on: a/a auditory cue responsive to step pattern via a closed loop motion sensor, 'make the auditory cue as rhythmic as possible' Device off: repeat baseline	Gait parameters; walking speed, stride length.	Device on: walking speed improved by 12.84% (SD 18.74%) Stride length improved 8.3% (SD 11.87%) Controls did not demonstrate a trend for either measure.	MQS: 7 Majority of patients fell below the mean change in speed and stride length due to larger effects in small numbers of participants. Findings should be interpreted with caution.

Table 3.0 Upper Limb Tremor: manipulation of visual information

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Beppu et al, 1987	Control comparison study with two conditions. 11 participants with cerebellar degeneration, duration of condition 2-11 yrs, aged 45-66 yrs, and 9 age matched healthy controls.	To determine the role of visual information on a visually guided slow ramp elbow tracking task. The tracking task was undertaken under two conditions; with visual information about the moving limb and performance or without this information.	Trajectory, signal, velocity, error, EMG (BB, B, TB), weave ratios = degree of undulation	Controls: withdrawal of the visual cue had negligible effects on tracking pattern and precision. People with ataxia: withdrawal of the visual cue significantly improved the smoothness of the trajectory close to that of controls, independent of severity of the condition.	MQS: 10 Demonstrated that repeated visually guided error correction responses were responsible for slow ramp elbow tracking errors. With deprivation of visual cues people with ataxia used memory and proprioceptive information and improved performance.
Feys et al, 2003	A comparison study of eye and hand movements. N=16 people with moderate intention tremor as part of MS, EDSS 5.5-8.0, n=16 age matched healthy controls.	To examine the characteristics of intention tremor and simultaneously produced eye movements during rapid goal directed movement. To examine the effect of loading the wrist (75g) on the above.	Performance measures (ms) - duration of initiation, ballistic and target phase of the task Kinematics - peak velocity (m/s) - time to peak velocity (%) Tremor measures (mm) - ballistic curvature index - end point error - target curvature index - amplitude of directional changes - number of directional changes	Patients demonstrated impaired limb coordination. Differences between patients and controls were most pronounced at the end of the movement ('homing in') in line with the diagnosis of intention tremor. Patients: decreased peak velocity, longer time to peak velocity with target overshoot. Indicative of prolonged agonist activity, delayed onset antagonist activation insufficient to brake the ballistic phase, causing overshoot Eye movement: inefficient compared to controls. Loading no effect on intention tremor.	MQS: 9 Further evidence that impaired limb coordination is coupled with unsteady eye fixation on the target in people with intention tremor during rapid goal directed movement; suggesting that the command structures for eye and hand movements are closely related. Intention tremor was a robust feature independent of the application of inertial loads.

Table 3.0 Upper Limb Tremor: manipulation of visual information - *continued*

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Feys et al, 2005a	A comparison study of the interaction of eye and hand movement. N=16 people with MS, EDSS 6-8, slight to severe intention tremor (FTRS) n=15 age matched healthy controls.	Experiments conducted to record eye and hand movements simultaneously in both groups using a wrist step tracking task. Wrist selected because action tremor is more apparent distally and step tracking task is similar to a two phase pointing task. Three conditions: a) eye and hand track target as usual b) eye only c) hand only	Transport phase: - duration (ms), peak velocity (ms), initial error (mm), mean end point error, mean amplitude primary saccade and hand movement Target phase: - additional path length ~/= tremor amplitude, number of directional changes	Participants with MS: Coupling between primary eye saccade and hand movement was preserved but abnormal compared to controls, because the primary saccade was performed more slowly and the peak velocity of the hand was delayed. Spatial coupling between primary saccade and hand movement was preserved. Amplitude of the primary hand movement exceeded that of the primary saccade (as is usual) but the amplitude of both the eye and the hand movement was greater than the controls, which could be related to the initially large primary saccade. Intention tremor amplitude was reduced when there was no preceding saccadic eye movement.	MQS: 11 Findings suggest that eye movement influence hand movements in people with intention tremor during a wrist step tracking task. Tremor amplitude may be reduced if the primary saccade and the hand movement to reach for an object are performed separately.
Feys et al, 2006	A comparison study of the interaction of eye and hand movement. N=13 people with MS, median EDSS 6, slight to severe intention tremor (FTRS), duration MS mean 13.9 yrs, n=14 aged matched controls.	Experiments conducted to record eye and hand movements simultaneously in both groups using a wrist step tracking task. Visual representation of the actual hand position was displayed immediately or averaged over time windows of 150, 250 and 350ms.	Transport phase: - duration (ms), peak velocity (ms), initial error (mm) Target phase: - additional path length ~/= tremor amplitude	Amplitude and error of tremor was significantly reduced by the delayed visual display in the patient group.	MQS: 11 Severity of hand tremor seems to be dependent on visual feedback with reference to position of the hand and movement errors.

Table 3.1 Upper Limb Tremor: peripheral cooling

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Feys et al, 2005b	Before and after study to determine the effects of peripheral cooling in intention tremor, n=18, people with MS, mean age 44yrs, range 18-63 yrs, duration of MS range 2-36 yrs, EDSS 5.5-7.5. FTRS range slight to severe.	Deep (18ºC) n=20 and moderate (25ºC) cooling n=8 for 15', reducing skin temperature by 13.5ºC and 7ºC respectively, using cooling fluid continuously circulating through a cryomanchet wrapped around the forearm.	Measures taken before, after and up to 30' after. Tremor amplitude and frequency using a step track task. Finger tap test, heart rate, body temperature, nerve conduction velocity.	HR and central body temperature – unchanged. Reduction of tremor amplitude and frequency reduced in both conditions but more so in the deeper cooling condition for up to 30'. Significantly worse on the FTT immediately after cooling but after 10' performance similar to pre-cooling (deep cooling only). Movement speed during step track task significantly reduced (deep cooling only). Nerve conduction was decreased after deep cooling.	MQS: 9 No adverse effects reported, authors advocated careful assessment to exclude those with contraindications. Overall reduction in the amplitude of tremor proportional to the intensity of the cooling. Effect possibly caused by cooling effect on nerve conduction, changed muscle properties and reduced muscle spindle activity, muscle spindle discharge is temperature dependent. Peripheral cooling might support upper limb function.
Quintern et al, 1999	Experimental study with healthy control comparison. Effect of cooling investigated with reference to a fast goal directed pointing activity in 11 people with MS (EDSS 5.5-7.5) and 11 age matched controls.	Rapid pointing task analysed with video motion before and after cooling for 1 minute in ice water up to the shoulder. Three conditions: - visual guidance and visual trigger - visual guidance and auditory trigger - memory guided and auditory trigger	Amplitude, peak velocity, 3D finger sway, frequency of postural tremor; before and after, no follow up.	Cooling had no effect on amplitude or peak velocity of the pointing movement under all conditions. Cooling significantly reduced finger sway.	MQS: 8 Outcome of finger sway attributed to reduction in the activity of group I spindle afferents. Cooling affected postural tremor but not dysmetria.

Table 3.2 Upper Limb Tremor: Mechanical Damping

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Aisen et al, 1993	Lab-based experimental study with healthy age matched control comparison (n=5) and n=10 with chronic moderate severe cerebellar ataxia secondary to TBI or MS. Age 21-50 yrs. Tremor affected the elbow or shoulder.	Investigated the effect of a mechanical tremor damper (produces opposing force proportional to velocity) in 3 degrees of freedom during a computer mediated pursuit tracking task at different damping levels in 2-5 trials.	Functional performance using a 6 task clinical assessment scale (estimate of tremor amplitude, handwriting Archimedes spiral, pouring water, using a spoon, pointing to keys on a key board. O-32 (0 = no tremor). + n=5 with and without inertial weight.	Control: no significant difference. Experimental: reduced tremor without impairing functional performance for all participants. Effect of added inertial was variable – no effect, better and worse.	MQS: 7 Lab based study with equipment that as yet has limited clinical utility. Interesting results re reducing involuntary tremor whilst preserving voluntary movement through damping rather than weighting.

Table 3.3 Upper Limb Tremor: wrist weighting

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Langton Hewer et al, 1972	Before and after study, n=50, age range 8-79 yrs, intention tremor due to idiopathic or essential tremor, MS, FA, cerebellar degeneration, stroke, PD, post-traumatic, cirrhosis of the liver, congenital dyskinesia, cerebellar tumour.	Assessment of upper limb tremor with application of wrist weights 240-720g, some up to 1kg and some with up 2kg on the upper arm.	Photography of action tremor, accelerometry, tremor rating scale, finger nose test, finger tap test, handwriting, figure drawing, spiral drawing, maze test. Questionnaire about social disability.	Tremor reduced in 29 participants, therapeutic effect for n=18, most of these continued to wear the splint for at least 6 months. Variable optimum weight requirement (480-600g for most participants), further increases not beneficial.	MQS: 3 Not clear if certain pathologies had a better outcome with weights than others. Those with weakness and fatigue possibly had fewer benefits. Low methodological quality.
McGruder et al, 2003	Single Case Experimental Design. N=5 aged 30-81, acute and chronic post TBI, brain tumour, multiple strokes and encephalitis; all with intention tremor that interfered with self feeding.	8 or 16 meal sessions with weighted cuff or sham cuff.	Time to acquire food Amount consumed No. of spills and compensations Participant self rating severity of tremor (0-10) Investigator rating of tremor (6 point ordinal scale)	All participants demonstrated improvements in one or more of the measured variables.	MQS: 9 Weights were self-selected and not reported individually. Clinical or functional significance was not clearly reported.
Morgan et al, 1975	Before and after study, n=31, aged 8-69 yrs people with intention tremor (very severe to mild) due to MS, essential tremor, cerebellar degeneration, PD, FA, cerebellar trauma, congenital lesions to the cerebellum and basal ganglia, stroke and cerebellar tumour.	10 randomly ordered trials of movement towards a button for each patient with and without a 600g wrist weight.	Accelerometry yielding a numerical index of tremor.	20/31 showed significant reduction in tremor. No association between cause of tremor and outcome (numbers in each category were low). More beneficial for moderate to severe tremor.	MQS: 5 Early study demonstrating some improvements due to wrist weighting but of low methodological quality.
Manto et al, 1994	Before and after study. N=11 controls, n=8 participants with cerebellar dysfunction (cerebellar stroke, neonatal hypoxia, cerebellar atrophy, pontocereballar angle tumour, glioma) with mild to moderate arm tremor, age range 14-73, duration of condition 20 days to 20 yrs.	Pre and post measures of dysmetria using wrist flexion under no weight, weighted 200g or 400g. Investigated dysmetria under conditions of fast accurate movement.	EMG of FCR ECR	Healthy controls adapted to weighted conditions by increasing agonist activity (launch force) and antagonist (braking force). Participants with cerebellar dysfunction could increase agonist but not antagonist activity. Onset of braking also was also delayed compared to controls.	MQS: 8 Weighting increased overshoot in people with cerebellar dysfunction and decreased it in controls.
Sanes et al, 1988	Before and after study. N=5, aged 35-70, with postural and kinetic tremor due to cerebellar infarct, olivopontocerebellar degeneration, MS, vascular tumour in the red nucleus.	An investigation of the influence of visual guidance and three types of mechanical loading (viscous, opposing and inertial) on postural and kinetic tremor.	Accelerometry, EMG torque motor studies. .	Postural maintenance and horizontal movement both improved with eyes shut. All mechanical loads that opposed wrist flexion reduced tremor. Postural tremor not observed during isometric contraction	MQS: 8 Further evidence that inaccurate processing of visual information contributes to UE tremor. Somaethestic inputs via mechanical loads can influence postural tremor.

Table 3.4 Upper Limb Tremor: Relaxation and Biofeedback

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Guercio et al, 1997	Single case experimental design (ABCAC), n=1, 23 yr old man, 3 yrs post TBI, upper limb kinetic tremor, difficulty feeding, wheelchair for mobility.	Relaxation and biofeedback to reduce upper limb tremor and improve functional performance. A: baseline measures B: behavioural relaxation training (BRT) C: as above plus auditory biofeedback from EMG forearm (static and dynamic conditions) A: return to baseline C: as above 1 year follow up ABCAC took place over 33 sessions, two sessions per week.	Behavioural Relaxation Scale EMG (resting and during functional tasks)	Large percentage improvement in BRS rated relaxation of posture during the BRT phases which continued into the follow up period. EMG levels reduced during the intervention phases. Anecdotal reporting of improvements in everyday function.	MQS: 5 Low methodological quality, reductions in EMG were not clearly interpreted with reference to severity of tremor.
Guercio et al, 2001	Before and after study n=1, 21 yr old man >3 months following a TBI. Right hemiplegia, severe left upper limb ataxic kinetic tremor, wheelchair user, used a spell board to communicate.	Relaxation and biofeedback to reduce upper limb tremor and improve performance in using a letter board. Intervention sequence: - Baseline data - Relaxation training (BRT) - BRT + EMG auditory biofeedback (3 levels of sensitivity) - Follow up at 2 years Over 23 sessions.	Behavioural Relaxation Scale EMG (during a spelling task) Self-rating of relaxation (0-7 scale) CRTS	Improved skills in relaxation over time. EMG: as skills in relaxation improved EMG during the letter spelling task reduced and CRTS scores fell indicating an improvement in the severity of tremor. Improvement from baseline was maintained at follow up.	MQS: 7 Interesting case study suggesting that improvements in tremor were associated with behavioural interventions to promote general relaxation and EMG reduction during a task. Preliminary evidence only.

Table 4.0 Axial Weighting

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Clopton et al, 2003	Single case experimental design (ABCA), n=5, community ambulators without orthosis or gait device, ataxic gait, diagnosed with cerebellar ataxia.	Baseline: 5 walks of 9.76m (3.66m on a Gaitrite mat) no weight. Intervention: as above with shoulder weights (10% body weight) Intervention: as above but with waist weights Return to baseline.	Gait parameters measured by Gaitrite; Velocity, cadence, step length deviation, step time deviation, width base of support, double stance time.	Not clearly reported but no findings were consistent across participants and it seems that gait deteriorated rather than improved.	MQS: 11 No clear support for axial weighting in this group of participants.
Folz and Sinaki, 1995	Prospective case series, n=19, range of pathologies including amyotrophic lateral sclerosis, Parkinsonism, cerebellar ataxia, degeneration of the CNS with ataxic gait. All had static or dynamic postural abnormality.	A postural training support device which consisted of anteriorally or posteriorally positioned weights on the trunk (range 1.75-2.5 pounds depending on individual need) to counter a posterior inclined lean or an anteriorally inclined lean respectively. 3 month follow up via telephone call.	Photographic comparisons were made before and after application of the device. Participants were asked to wear the device during walking activities if they found it helpful.	Subjective improvements reported in posture and gait and all participants except one reported using the device at 3 month follow up. No adverse effects reported. People with ataxia reported feeling more stable.	MQS: 6 Low methodological quality varied outcome. No firm conclusions can be drawn.

Table 5.0 Lycra Garments (paediatrics)

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Blair et al, 1995	Cross-over trial, n=16 in the intervention group, n=8 matched controls, plus 8 other controls. Age range unclear, severity of motor impairment ranged from mild to profound. Five participants were described as having ataxia as the predominant motor impairment.	24 children assigned to wear a full body lycra suit in an ABAB design study.	Observer rating 14 functional activity items Progress towards motor goals Positional stability Quality of movement Grip strength Sit ups Spirometry	Complex reporting. Dynamic function was generally positively reported. Video observations reported positive effect on postural stability, quality of upper limb movement, involuntary movement reduced. Respiratory function decreased sufficient to contraindicate use for one participant.	MQS = 5 Low methodological quality and complex reporting of the method, intervention and results. Significant adverse effects reported.
Corn et al, 2003	Single case experimental design (AB), n=4, age range 8-16 yrs, CP or ABI (one with ataxic hemiplegia).	Upper limb lycra garment fitted following a baseline phase on reported measures.	Melbourne Assessment on Unilateral Upper Limb Function.	Highly variable between individuals.	MQS: 12 No firm conclusions can be drawn. Limitations addressed.

Table 5.0 Lycra Garments (paediatrics) – *continued*

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Nicholson et al, 2001	Before and after study, n=12; aged 2-17 yrs (mean 6.8yrs) with various types of CP (one boy with ataxia),	Full body suit or adapted full body suit – 2 weeks gradual exposure following a minimum of wear 6 hrs a day for 6 weeks.	PEDI before and after application of lycra garment. N=5 evaluated using motion analysis for reach and grasp. Carer questionnaire regarding practicalities of using the garment.	All children wore the garment for 6 weeks but daily use ranged from 2.7-10.2 hrs. 11 children significantly improved on at least one of the three functional items of the PEDI; 6 improved on all items. All of these changes were slight although noted by carers. Motion analysis was mixed, some improvements and two showed deterioration. Significant adverse effects were reported. Only one parent wanted to continue use.	MQS: 10 Good quality study, demonstrated varied effects that offer some indication of benefit but this seems to be outweighed by significant adverse effects. No control group therefore results should be interpreted with caution.

Table 6.0 Hippotherapy (therapeutic riding)

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Hammer et al, 2005	SCED (ABA); n=11 participants with MS, mean age 47.9 yrs (range 35-61) time since diagnosis mean 10yrs, mean EDSS 5, described in terms of social status, previous riding experience, medication, walking aids, mood and depression.	10 weekly sessions of hippotherapy of 30' each in the intervention phase, individually tailored, included activities on the horse; trunk rotation, balance exercises, with and without visual input, as well activities of the horse e.g. weaving in an out of cones riding diagonals and circles.	BERG, figure of 8 walk, TUG, 10m walk, spasticity (MAS), Index of Muscle Function, the Birgitta Lindmark motor assessment part B, coordination, self rated pain, muscle tension, patient specific functional scale, SF-36.	Variable across individuals, the majority of participants improved in one or more of the variables, mainly balance. Eight participants improved in the role-emotional section of the SF-36.	MQS: 11 Good SCED demonstrating individual differences but a trend towards improved balance. However, as a SCED these findings cannot be generalised and further research is required for people with ataxia.

Table 7.0 Wheelchair Seating

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Clark et al, 2004	Prospective, two periodsrandomised within subject cross over study. N=19 (n=15 DMD, n=4 FA). Aged 6-21 yrs.	Random order provision of adapted wheelchair or standard wheelchair; 15 minute interval between tests.	CODA Motion Analysis System (7 variables), JTHF, lung function (FVC, FEV, PEF, slow VC)	No significant difference in lung function. Improvement in one of the subtests of the JTHF. Several postural measures improved.	MQS: 6 Low methodological quality, specialist seating may improve as well as adversely affect posture therefore longitudinal studies with more sensitive outcome measures would be useful.

Table 8.0 Endurance Training

Source	Design and Participants	Intervention	Outcome Measures	Results	Quality and Comment
Fillyaw and Ades, 1989	Case report n=1, 38 year old man with FA, onset of neurological signs as a teenager, ataxic gait, walked without assistance, no cardiac symptoms and normal cardiac size and function.	27 supervised and ECG monitored exercise sessions over a nine week period, 10' warm up, 20-25' cycling at 50rpm HR maintained at 70-80% max.	Oxygen consumption (peak VO_2, peak ventilation, peak tidal volume), maximum work in Watts and exercise time, body weight.	Physiological adaptations consistent with aerobic conditioning were reported. Improvements in peak VO_2, peak ventilation, tidal volume. Exercise time increased from 23 to 28 minutes reflected a 50W increase in maximum work.	MQS: 9 Good quality study demonstrating potential benefits of endurance training that require further investigation. These findings are preliminary and should be interpreted with caution for this population.

Appendix B

Comparison

	Hierarchy of Evidence Level	Purpose clearly stated	Relevant lit review	Appropriate design	Biases accounted for	Sample described in detail	Sample size justified	Informed consent	Outcomes reliable	Outcomes valid	Intervention described in detail	Statistical significance reported	Analysis appropriate	Clinical importance reported	Conclusions appropriate	Clinical implications reported	Limitations reported	Score
Rehabilitation																		
Armutlu et al, 2001	Ib	Y	N	Y	Y	Y	N	N	N	N	N	Y	N	Y	N	N	Y	7
Balliet et al, 1987	III	Y	Y	Y	N	Y	N	N	N	N	Y	Y	N	Y	Y	Y	N	9
Brown et al, 2006	III	Y	Y	Y	N	Y	N	Y	N	N	N	Y	Y	Y	Y	Y	Y	11
Dordal, 1989	III	Y	N	Y	N	Y	N	N	N	N	N	N	N	N	Y	N	N	4
Gialanella et al, 2005	III	Y	Y	Y	N	Y	N	N	N	N	N	Y	Y	Y	Y	Y	N	9
Gill-Body et al, 1997	III	Y	Y	Y	N	Y	N	N	N	N	Y	N	N	Y	Y	Y	N	8
Gillen, 2000	III	Y	Y	Y	N	Y	N	N	N	N	Y	N	N	Y	N	Y	N	7
Harris-Love et al, 2004	III	Y	Y	Y	N	Y	N	N	N	N	N	N	N	Y	Y	Y	N	7
Jones et al, 1996	IIa	Y	Y	Y	N	N	N	Y	N	N	N	Y	N	Y	N	N	Y	7
Karakaya et al, 2000	IIb	Y	Y	Y	N	Y	N	N	N	N	N	Y	Y	Y	N	N	N	7
Perlmutter and Gregory, 2003	III	Y	N	Y	N	Y	N	N	N	N	N	N	N	Y	Y	N	N	5
Smedal et al, 2006	IIb	Y	Y	Y	N	Y	N	Y	N	N	N	Y	Y	Y	Y	Y	Y	11
Stoykov et al, 2005	III	Y	Y	Y	N	Y	N	N	N	N	N	N	N	Y	N	Y	N	6
Visual Tracking																		
Beppu et al, 1987	IIb	Y	Y	Y	N	Y	N	Y	N	N	Y	Y	Y	Y	Y	N	N	10
Crowdy et al, 2002	IIb	Y	Y	Y	N	N	N	Y	N	N	Y	Y	Y	N	Y	Y	N	9
Feys et al, 2003	IIb	Y	Y	Y	N	Y	N	Y	N	N	Y	Y	Y	N	Y	N	N	9
Feys et al, 2005a	IIb	Y	Y	Y	N	Y	N	Y	N	N	Y	Y	Y	N	Y	Y	Y	11
Feys et al, 2006	IIb	Y	Y	Y	N	Y	N	Y	N	N	Y	Y	Y	N	Y	Y	Y	11
Peripheral Weighting																		
Langton Hewer et al, 1972	IIb	Y	N	N	N	N	N	N	N	N	N	N	N	Y	N	Y	N	3
Manto et al, 1994	IIb	Y	N	Y	N	Y	N	Y	N	N	Y	Y	Y	N	N	Y	N	8
McGruder et al, 2003	IIb	Y	Y	Y	N	Y	N	Y	N	N	Y	Y	N	N	N	Y	Y	9
Morgan et al, 1975	IIb	Y	N	Y	N	Y	N	N	N	N	N	Y	Y	N	N	N	N	5
Sanes et al, 1988	Iib	Y	Y	Y	N	N	N	N	N	N	Y	Y	Y	N	Y	Y	N	8
Mechanical Damping																		
Aisen et al, 1993	IIa	Y	N	Y	N	N	N	Y	N	N	N	Y	Y	Y	N	Y	N	7
Peripheral Cooling																		
Feys et al, 2005b	IIb	Y	Y	Y	N	Y	N	Y	N	N	Y	Y	Y	N	Y	N	N	9

Quintern et al, 1999	IIa	Y	Y	Y	N	Y	N	N	N	N	Y	Y	Y	N	Y	N	N	8
Biofeedback																		
Baram and Miller, 2007	IIb	Y	Y	Y	N	Y	N	Y	N	N	Y	Y	N	N	N	N	N	7
Guercio et al, 1997	IIb	Y	Y	Y	N	N	N	N	Y	N	Y	N	N	N	N	N	N	5
Guercio et al, 2001	IIb	Y	Y	Y	N	N	N	N	Y	N	Y	Y	Y	N	N	N	N	7
Treadmill Training																		
Brown et al, 2005	Ib	Y	Y	Y	N	N	N	Y	Y	N	Y	Y	Y	Y	Y	Y	Y	12
Cernak et al, 2008	III	Y	Y	Y	N	Y	N	N	Y	Y	Y	N	N	Y	Y	Y	N	10
Vaz et al, 2008	IIb	Y	Y	Y	N	Y	N	Y	Y	N	Y	Y	Y	Y	Y	Y	Y	13
Axial Weighting																		
Clopton et al, 2003	IIb	Y	Y	Y	N	N	N	Y	Y	N	Y	Y	Y	N	Y	Y	Y	11
Folz and Sinaki, 1995	IIb	Y	Y	Y	N	Y	N	N	N	N	N	N	N	Y	N	Y	N	6
Endurance Training																		
Fillyaw and Ades, 1989	III	Y	Y	Y	N	Y	N	Y	N	N	Y	N	N	Y	Y	Y	N	9
Seating																		
Clark et al, 2004	IIb	Y	Y	Y	N	N	N	N	N	N	N	N	N	Y	Y	Y	N	6
Therapeutic Riding																		
Hammer et al, 2005	IIb	Y	Y	Y	N	Y	N	Y	N	N	N	Y	Y	Y	Y	Y	Y	11
Lycra																		
Blair et al, 1995	IIb	Y	Y	N	N	Y	N	Y	N	N	N	Y	N	N	N	N	N	5
Corn et al, 2003	IIb	Y	Y	Y	N	Y	N	Y	Y	Y	N	Y	Y	N	Y	Y	Y	12
Nicholson et al, 2001	IIb	Y	Y	Y	N	N	N	N	Y	Y	Y	Y	Y	N	Y	Y	N	10
Systematic Review																		
Martin et al, 2009	Ib	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a

Valid Inputs

Y

N

Hierarchy of Evidence: Grading Scheme

Adapted from the National Collaborating Centre for Chronic Conditions (2003) Multiple Sclerosis: Management of multiple sclerosis in primary and secondary care, clinical guideline 8, National Institute for Clinical Excellence which was adapted from Eccles, M and Mason, J (2001) How to develop cost-conscious guidelines. *Health Technology Assessment*, 5, 16.

Evidence Category	Source
Ia	Evidence from meta-analysis of randomised controlled trials
Ib	Evidence from a randomised control trial /at least one RCT
IIa	Evidence from a controlled study without randomisation
IIb	Evidence from a type of quasi-experimental study
III	Evidence from non-experimental descriptive studies
IV	Evidence from expert committee reports, opinions of clinical experience of respected authorities