Rehabilitation and Cerebellar Ataxia

Prof Jon Marsden
School of Health Professions
University of Plymouth
Rehabilitation for Ataxia

Martin et al (2009)
Effectiveness of physiotherapy for adults with cerebellar dysfunction: a systematic review.

Treatment for ataxia in multiple sclerosis.
Cochrane Database Syst Rev 24(1):CD005029

Marquer et al 2014
The assessment and treatment of postural disorders in cerebellar ataxia: A systematic review
Annals Of Physical Medicine and Rehabilitation 57 67-78

Ilg et al 2014
Consensus paper: management of degenerative cerebellar disorders
Cerebellum 13(2) 248-268

Marsden and Harris 2011
Cerebellar Ataxia: pathophysiology and Rehabilitation
Clinical Rehabilitation 25(3) 195-216
The Patient’s Journey

Suddenly we had a diagnosis, and we found it opened doors….a lot of people never get a diagnosis and have to deal with that grey area for years

Some people presume I’ve been drinking even in the morning

“Patients have also identified the following as helpful”

• Getting a Diagnosis

• Sensitive Health Professionals

• Contact with others in a similar position (support group)

• Carrying a card explaining the symptoms and that they are not drunk

• Adopting wheelchair use

• Counselling

http://www.ataxia.org.uk/

Box et al, 2005 BMJ 331 1007-1009
The Patient’s Journey

Impact on Quality of Life

Symptoms are often not typical “cerebellar” Signs

41% Sleep related compliant
38% depression

Pain
Fatigue
Bladder Control

Abele et al, 2007 Movt Disord 22(3) 348-352
Wilson et al, 2007 Eur J Neurol 14 (9) 1040-1047
Epstein et al, 2008 J Neurol Sci 272 (1-2) 123-8
Lopez-Bastida et al, 2007 Movt Disord 23 (2) 212-217
The Multi-Disciplinary Team

Person with Ataxia

GP  Neurologist  Family
Clinical Nurse Specialists
Social worker
Occupational Therapist
Physiotherapist
Cardiologist
Speech Therapist
Orthopaedic Surgeon
Wheelchair Clinic
Ataxia UK Representative
Clinical Geneticist
Genetic Counsellor
Cerebellar Ataxia

Co-morbidity

Dysmetria
Dyssynergia
Dysarthria
Tremor
Impaired balance & gait

Sensory Dysfunction
Vestibular Dysfunction

Sequelae

Immobility
2° Pain
Deconditioning & Fatigue
Respiratory Distress
Social Isolation

Abilities Participation

Affects
Aims of Therapy

Maintain/ Improve abilities and participation

Prevent $2^{nd}$ complications

Compensatory approach  Restorative approach

Adaptive aids & Environmental Modification

Teach New Strategies
Compensatory Approach I

Mobility/ Accessibility

- Walking Aids
- Wheelchair
- Driving Modifications
Compensatory Approach I

Mobility/Accessibility

- Walking Aids
- Wheelchair
- Driving Modifications

Specialised Seating

Standardised Vs Specialised
Clark et al Int J Ther Rehabil 2004

Friedreich’s Ataxia 6-21Yrs

Sitting Posture (Motion analysis)

Respiratory Function (spirometry)

Jebsen Hand Function =
Compensatory Approach I
Swallowing / Feeding

Utensils
- Weighted cutlery
- Self levelling cutlery
- Adapted plates

Food Consistency

Posture
- Head flexion

Cosmesis may be a problem

Neater Eater
Compensatory Approach II

Use of Technology

Communication Aids

Computer Use

Lightwriter

Pictures

Assistive Technology

Slowing down mouse
Double click speed
Motion Filtering software

Environmental Modification

Mouse Trap

Uses viscous damping to
Smooth out ataxic Movement

Upper limb support

Feys et al J Rehabil Res & Dev 2001
Compensatory Approach II

Use of Technology

Communication Aids
- Computer Use
- Lightwriter
- Pictures

Environmental Modification
- Ataxia
- Vision / Visuo-spatial activity
- Cognitive difficulties
Compensatory Approach II
Activities of Daily Living: Using inertia and Viscosity

Use of Weights

Normal Inertia

Intention Tremor: No effect
Feys et al Multiple Sclerosis 2003

Upper Limb

Hypermetria
Manto et al J Physiol 2003

Eating: Improved
McGruder et al 2003 Am J Occup Ther

Gait
Inconsistent effects on gait parameters
Clopton et al Neurology 2003

Add 500g
Compensatory Approach II
Activities of Daily Living: Using inertia and Viscosity

Dynamic Lycra® Splints

Assistance required for don/doff

Toileting

Drying Time

Compensatory Approach III
Learning New Strategies

Dysynergia
&
Decomposition of Movement

(Bastian et al, 1996, J. Neurophysiol.)

Slow accurate reaching

Control

Cerebellar
Compensatory Approach III
Learning New Strategies

Dysynergia
&
Decomposition of Movement

Reduce the degrees of Freedom
& use slow single joint movements
Bastain et al Phys Ther 1997 77

Use support of the table

Perform tasks in sitting not standing
Restorative Approaches

- Re-train balance
- Oculomotor re-training
- Transcranial Magnetic Stimulation
- Adjuncts
Balance and Gait Deficits in Cerebellar Disease: Sensory Integration

Large responses to Moving Visual Stimuli seen in SCA6 that correlate with symptom severity

Bunn et al Mov Dis 30(9) 1259-1266
Restorative Approaches: Sensory Re-training

Large responses to Moving Visual Stimuli seen that correlate with symptom severity

Feasibility trial of Balance re-training with Moving visual stimuli

SCA6 N=6 intervention Vs n=6 Control
15 mins 5 days/week for 4 weeks
Suggests improvements in balance (bal SARA) similar in effect size to that seen with disease progression over 1 year

Bunn et al 2015 Clin Rehabil 29(2) 143-153
Restorative Approaches
Intensive Exercise

**N=42 Degenerative Diseases**

RCT

**Therapy 4 weeks (6hrs / week)**
- Strength exercises
- Range of Motion
- Static and Dynamic Balance
- Walking outside / Stair Climbing

**Improvements**
- Walking Speed
- Reduction in falls
- Maintained at 12 weeks

**Intensive Co-ordination Training**

- Cerebellar (n=10) or Afferent pathway degeneration (n=6)

**4 weeks 3hrs / week**

**Improvements in Walking And Co-ordination**

Miyai et al 2012 Neurorehab and Neural Repair 26(5) 515-522

Ilg et al 2009 Neurology 73 1823-1830
Restorative Approaches

Video-gaming

N=1 – 10 Children
No control group

Ilg et al 2012 Neurology 79(20) 2056-2060
Synofzik et al 2014 Biomed Rres Inter 10.1155/2014/583507
Restorative Approaches

Vestibular-like Rehabilitation

Eye-head co-ordination + balance exercises
- Improved (n=2)
- Posturography
- Clinical Tests

Vestibular Habituation
+ strength / stretches / gait re-education
- Improved (n=10)
- Disability scales in people with cerebellar signs

Gill-Body et al Phys Ther 1997
Brown et al 2006
Guidelines for Exercise Prescription

Training needs to be:

- Intensive
  - 3 hrs/week for 4 weeks.
- Boost Therapy

- Maintained
  - 1 hr/day homework? (80 mins/week ineffective)
  - Return to baseline in 6 months if no training

- Challenging
  - Improvements greater in people who undertook
  - More challenging exercises

- Individualised and Motivating

- Part of overall exercise regime

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Physiotherapy and Exercise

Value
Collaborative
Long term
Therapeutic relationships

Self devised Exercise results in multiple Psychosocial benefits That are absent from Physiotherapy-prescribed Home exercise programs

Cassidy et al 2017 Disability and Rehabil
http://dx.doi.org/10.1080/09638288.2016.1277400
Physical Activity for people with Rare Neurological Conditions 
PARC

Focus Groups with patients, carers and health professionals to identify:
• PA experiences
• impact of HD,
• carer’s experience
• enablers.

Engage -HD

Pilot Study
• Physical Activity Coaching
• Physical Activity Workbook
• Physical Activity DVD

Huntington’s Chorea
Spinocerebellar Ataxia
Hereditary Spastic Paraparesis
Neuromuscular Diseases
Progressive Supranuclear Palsy
Physical Activity for people with Rare Neurological Conditions
PARC

Meeting with Support Group representatives
Cardiff Jan 2017

Skype Meetings

Academic Team Meeting
April 2017

Grant Submission
July 2017
Physical Activity for people with Rare Neurological Conditions PARC

Cardiff
Monica Busse & Clinical Trial Unit

Newcastle
Lynn Rochester

Oxford
Helen Dawes

London
Gita Rhamdharry (Chief Investigator) Fiona Jones

Glasgow
Lorna Paul

Plymouth
Jon Marsden
The PARC Collaboration: Physical Activity in Rare Conditions

NIHR Programme Development Grant:
(£100,000, 12 months)

Deliverable 1: Establishing the PARC community of researchers, PPI members and charities
- Planning workshop
- Consensus on communication
- Monthly virtual meetings of core PARC community
- End of PDG meeting to summarise findings and plan PG

Deliverable 2: Map current practice, care pathways and research sites
- Evidence synthesis of interventions
- Service profiling with charities and HCP networks
- Research site feasibility assessment
- Site visits
- Mapping workshop

Deliverable 3: Outcomes consensus
- Evidence synthesis of outcome measurement in rare conditions and previous analysis of properties and performance
- Scoping of outcome measure banks and databases
- Consensus workshop to map domains, prioritise importance to PPI members and identify gaps in psychometrics

NIHR Programme Grant for Applied Research
(£1,000,000, 36 months)

Work package 1: PARC intervention development
- Four regional co-production workshops
- Development of technology platforms
- Development of exercise prescription and self-management training for HCPs

Work package 2: PARC feasibility
- Two arm, randomised, controlled feasibility trial
- 6 month intervention versus usual treatment as usual with 12 month follow up
- Process evaluation
- Health economic tool development and pilot
- Development of progress criteria for full trial

Work package 3: PARC implementation, outcome and trial design
- Consolidated framework for advancing implantation research to identify barriers and enablers during feasibility trial
- Evaluation of performance of outcome measures using feasibility trial data
- Determination of definitive trial design using Bayesian methods

NIHR Health Technology Agency
Definitive PARC trial
Potential Adjuncts To Treatment
Restorative Approaches
Oculomotor Retraining

Saccade to target prior to start of swing phase

Ataxia → Several Hypometric saccades

Crowdy et al EBR 2000, 2002
Restorative Approaches
Oculomotor Retraining

Eye Movement Rehearsal:
↓ No. of saccades to reach target

Stance Duration
Pre 1st 2nd Rehearsal

Stance Duration Variability

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Restorative Approaches
Oculomotor Retraining

OPTIMEYES
Oculomotor Re-training in Progressive MS
with Cerebellar Signs

Intervention N=15 Vs n=13 Control

Clinical Measures
- Box and Blocks
- 9 Hole Peg Test
- Functional Reach
- SARA

Lab based measures of Motor Control
- Focus Builder
- Commercially Available Apps
- Vision Tap
- Eye-Hand Tracking
- Postural Sway
- Smooth Pursuit & Saccades

• Differences seen between Healthy Controls and pwMS

Significant changes in eye-hand tracking
No change in clinical outcome
Subjective reports of improvements
With balance in complex environments
Restorative Approaches
Novel Methods

Transcranial Magnetic Stimulation

Prolonged cortical silent period in response to TMS in pw Cerebellar Ataxia

Tamburin et al 2004 Clin Neurophysiol  115 348-355
Restorative Approaches
Novel Methods

Changes in Motor cortex excitability
Correlates with changes in 9HPT following focal cerebellar lesions
Ratio of affected to unaffected hand given
Restorative Approaches
Novel Methods

Rapid TMS (5HZ Stimulation 10 stimuli x 18) Primary Motor Cortex
100% Resting motor Threshold
8 pw MS + cerebellar Signs

Immediately after 10 min later 20 min later

Percentage improvement in 9HPT

Koch et al, 2008 MS 14 995-998
Restorative Approaches

Novel Methods

Effects of Direct Cerebellar Stimulation

SCA degeneration

39 Active

35 Sham

Blinded Ax

Improved

10 m Timed walk (time/steps)
Nominal scale of walking / standing

No change in regional blood flow

Inion
R / L lateral to Inion

100% output x 30 stimuli (10 ea location)
21 consecutive days

Shiga et al JNNP 2002
Restorative Approaches
Brain Stimulation

TMS and tDCS
Groups: Progressive and Stroke
Case studies, Pre-post and RCTs

Changes reported in:

Motor Cortex Neurophysiology
(eg Cerebellar Brain Inhibition)

Gross Function
Walking speed
(Bennusi et al 2015, V 30, 1701-1705, Shiga et al J002 72 JNNP 124-126)

BUT
Some controlled Trials show no effect
Kim et al 2014 J Rehabil Med 46 418-423

Unclear:

• Effect of paradigm (e.g. 1 Hz, theta burst, ANODAL) and site of stimulation
  • Effect of lesion site Cerebellar Vs Medulla

• Target: Purkinje Cells Vs Interneurons Vs antidromic stimulation of Corticospinal Tract

• Effect of stimulation on motor control Vs gross function
Compensatory

Function

Restorative

Which technique?

How long and at what intensity?
Acknowledgments

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• Brian Day

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• Lisa Bunn