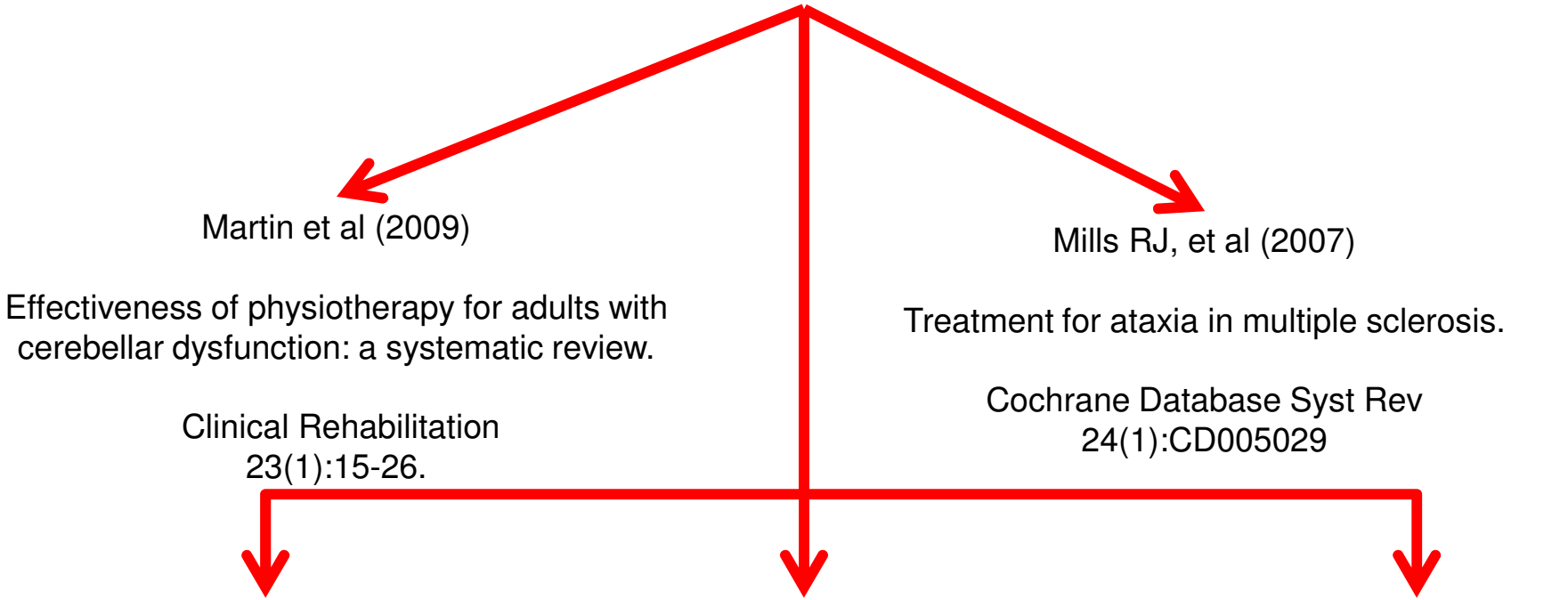
RESEARCH WITH PLYMOUTH UNIVERSITY

Rehabilitation and Cerebellar Ataxia

Prof Jon Marsden
School of Health Professions
University of Plymouth

Rehabilitation for Ataxia



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

Ataxia UK Home

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

Box et al, 2005 BMJ 331 1007-1009

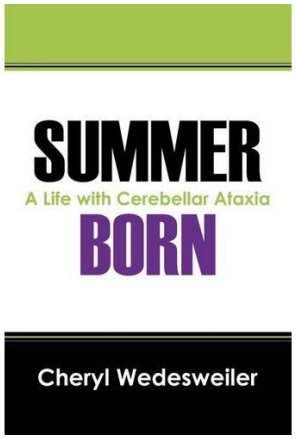
The Patient's Journey

Impact on Quality of Life

Symptoms are often not typical "cerebellar" Signs

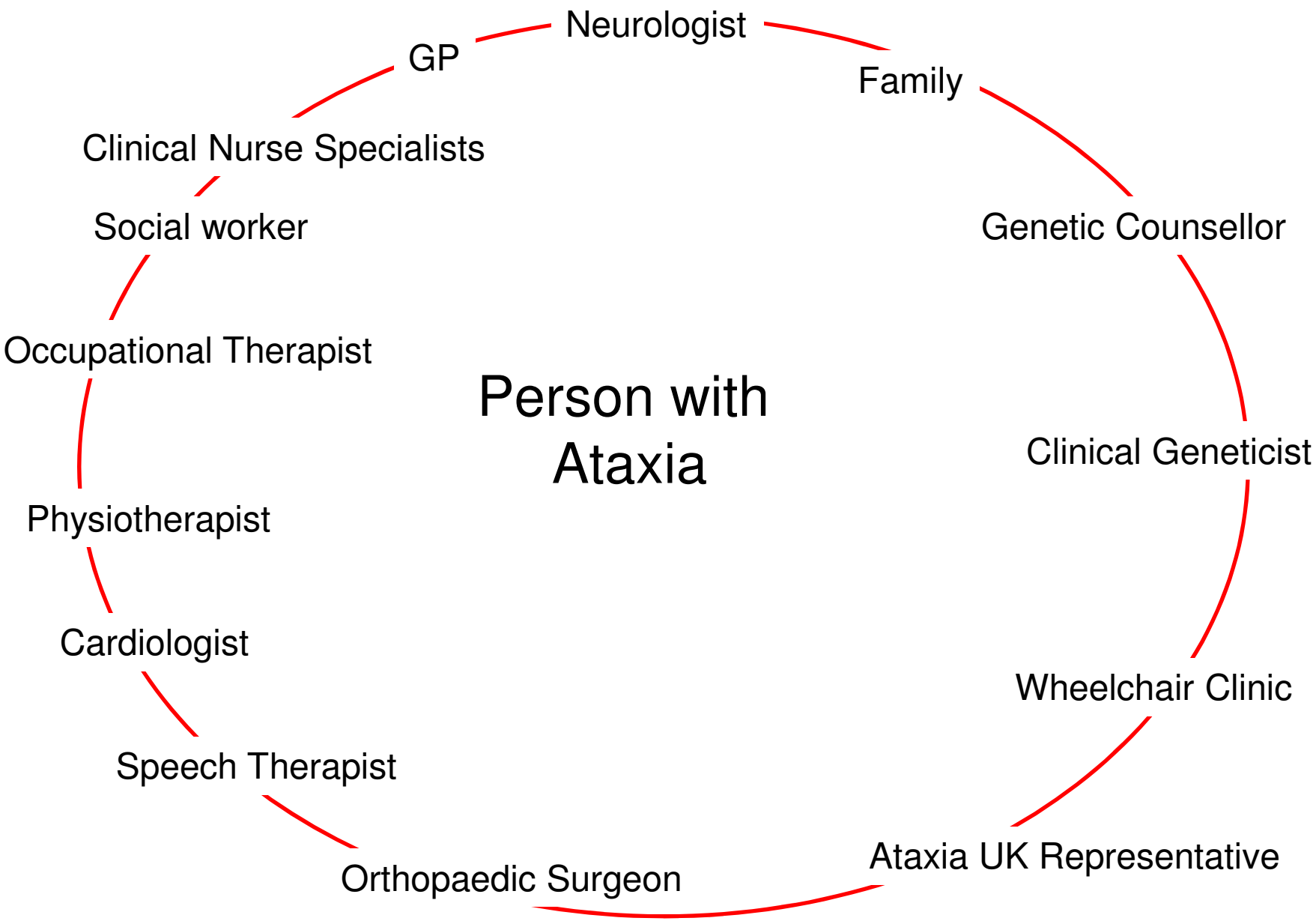
41% Sleep related compliant
38% depression

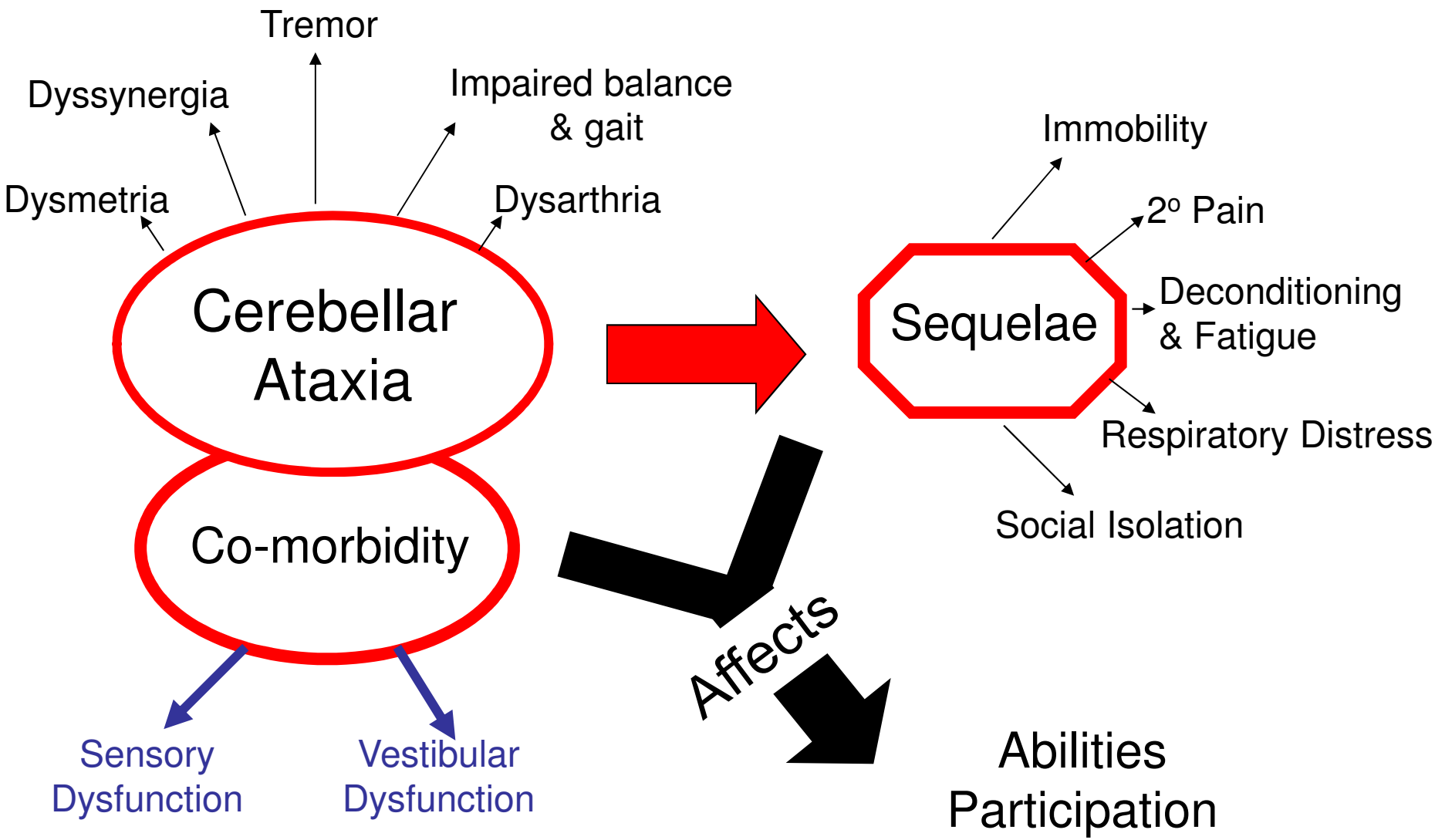
Pain
Fatigue
Bladder Control



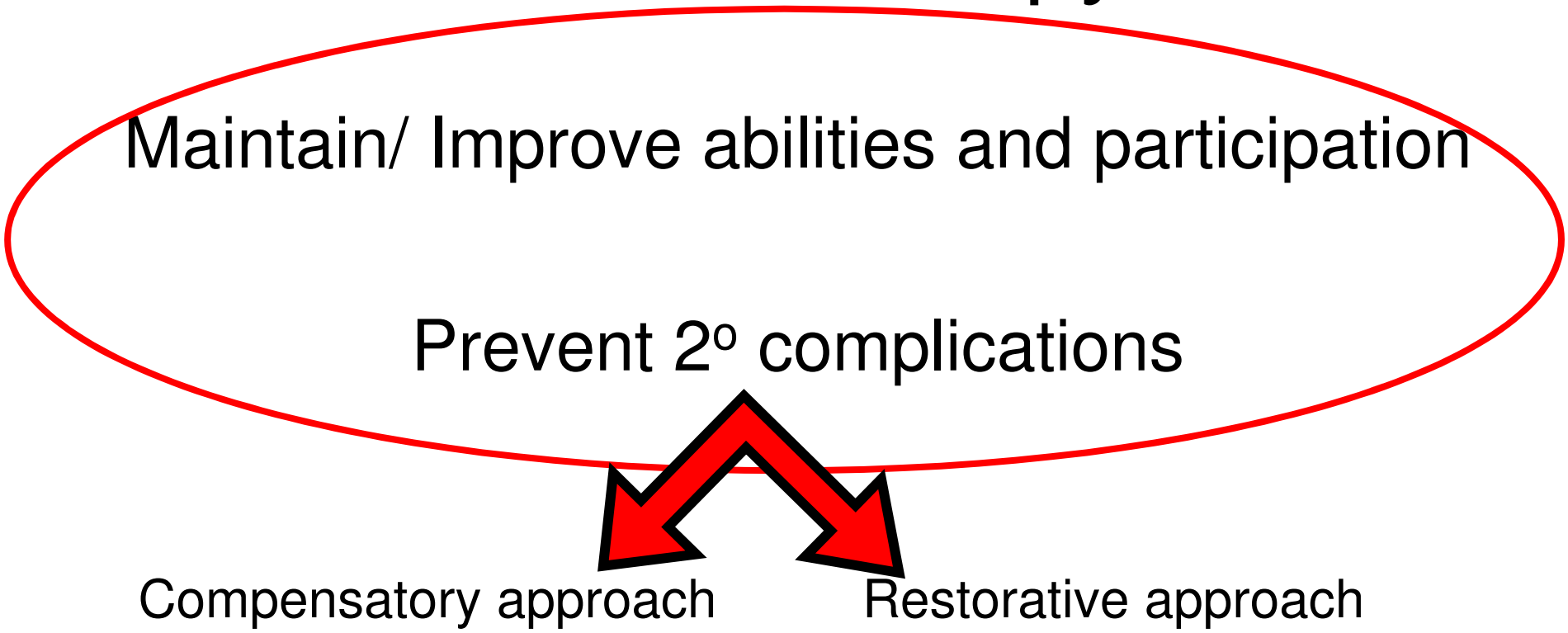
Abele et al, 2007 Movt Disord 22(3) 348-352
Wilson et al, 2007 Eur J Neurol 14 (9) 1040-1047
Epstein et al, 2008 J Neurol Sci 272 (1-2) 123-8
Lopez-Bastida et al, 2007 Movt Disord 23 (2) 212-217

The Multi-Disciplinary Team




Tremor
Dyssynergia
Impaired balance & gait
Dysmetria
Dysarthria
Cerebellar Ataxia
Co-morbidity
Sensory Dysfunction
Vestibular Dysfunction
Affects
Immobility
2º Pain
Sequelae
Deconditioning & Fatigue
Respiratory Distress
Social Isolation
Abilities
Participation

Aims of Therapy



Adaptive aids & Environmental Modification

Teach New Strategies

Compensatory Approach I

Mobility/ Accessibility

- Walking Aids
- Wheelchair
- Driving Modifications

Specialised Seating

Mobility

Pressure Relief

Stability/Posture

Upper limb Function

Swallowing

Respiratory Function

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

Food Consistency

Posture
Head flexion

Weighted cutlery

Self levelling cutlery

Adapted plates

Neater Eater

Cosmesis may be a problem

Compensatory Approach II

Use of Technology

Communication Aids

Computer Use

Lightwriter

Pictures

Environmental Modification

Mouse Trap

Uses viscous damping to Smooth out ataxic Movement

Assistive Technology
Slowing down mouse
Double click speed
Motion Filtering softwear

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

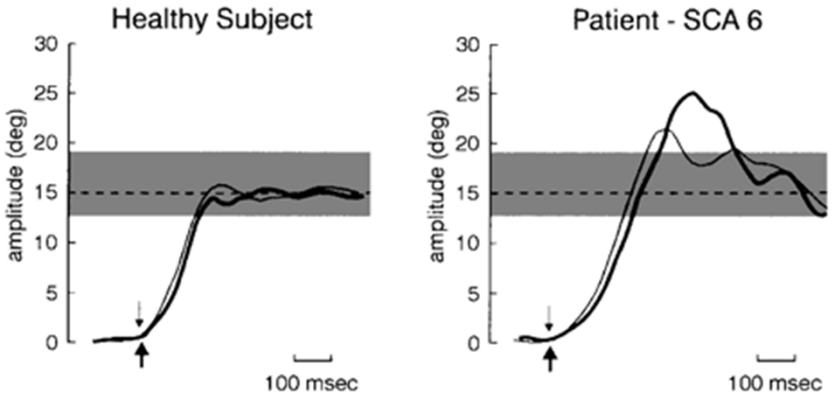
Cognitive difficulties

Compensatory Approach II

Activities of Daily Living: Using inertia and Viscosity

Use of Weights

Normal Inertia



Upper Limb

Intention Tremor: No effect

Feys et al Multiple Sclerosis 2003

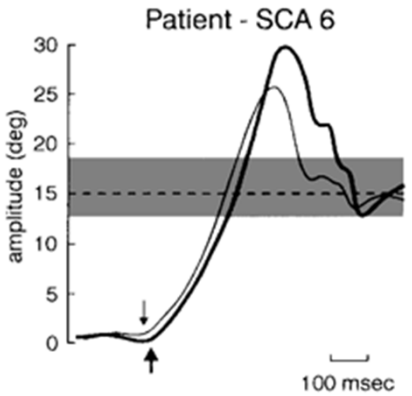
↑**Hypermetria**

Manto et al J Physiol 2003

Eating: Improved

McGrunder et al 2003 Am J Occup Ther

Add 500g



Gait → Inconsistent effects on gait parameters

Clopton et al Neurology 2003

Compensatory Approach II

Activities of Daily Living: Using inertia and Viscosity

Dynamic Lycra® Splints

Lycra Splinting

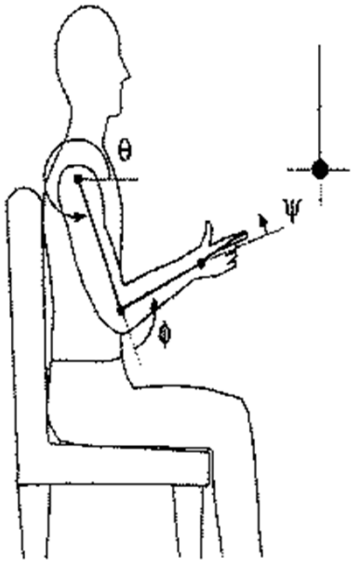
- Assistance required for don/doff
- Toileting
- Drying Time

Scott-Tatum http://www.secondskin.com.au/secondskin/PDF/scope.pdf

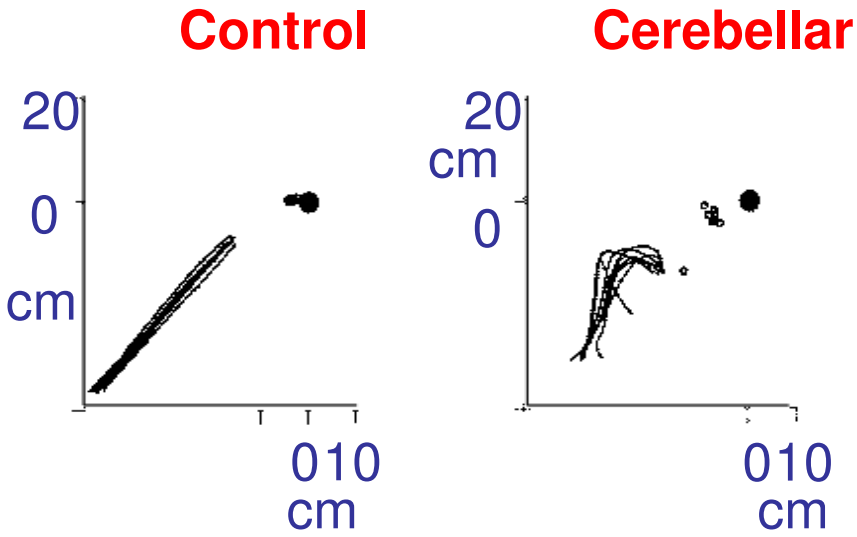
Compensatory Approach III

Learning New Strategies

Dysynergia
&
Decomposition of Movement



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

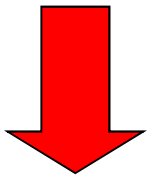


Slow accurate reaching

Compensatory Approach III

Learning New Strategies

Dysynergia
&
Decomposition of Movement

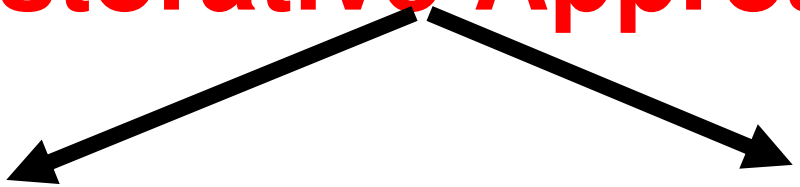


Reduce the degrees of Freedom
& use slow single joint movements
Bastain et al Phys Ther 1997 77

Use support of the table

Perform tasks in sitting not standing

Restorative Approaches



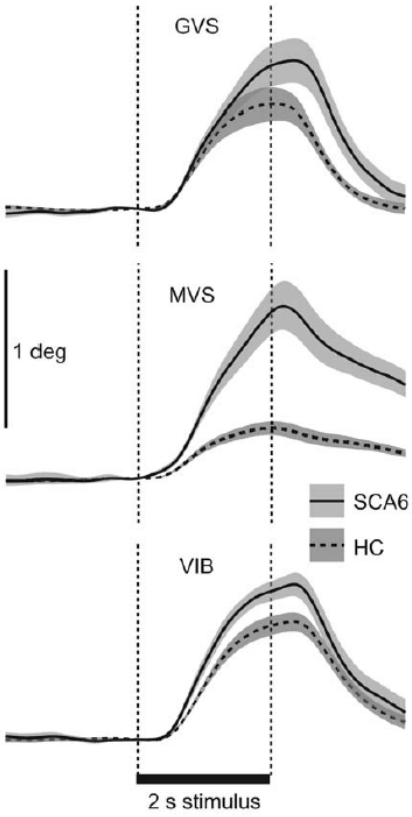
Re-train balance

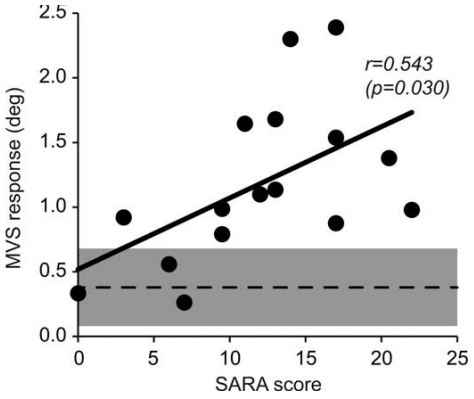
Adjuncts

Oculomotor re-training

Transcranial Magnetic Stimulation

Balance and Gait Deficits in Cerebellar Disease : Sensory Integration



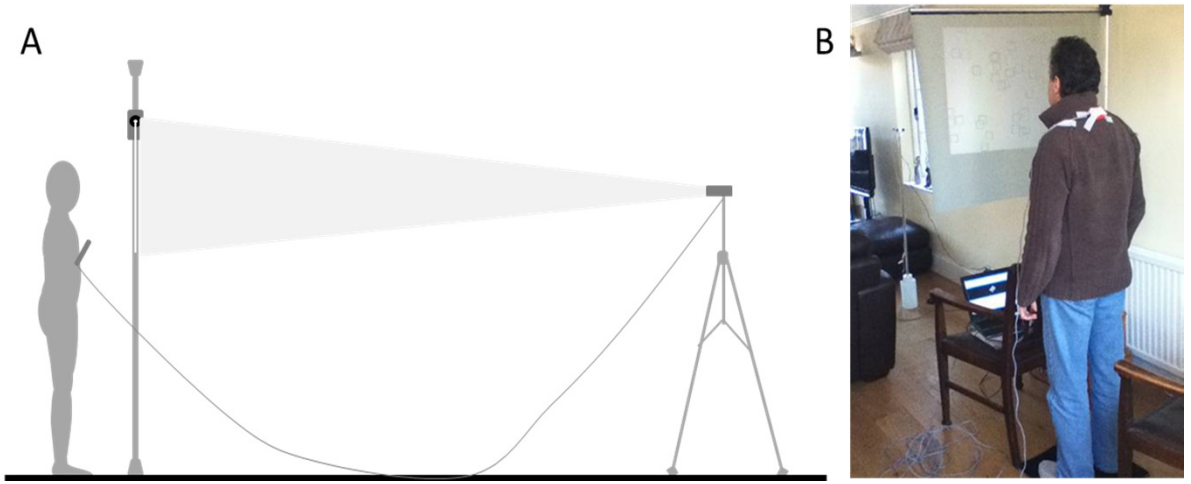


Large responses to Moving Visual Stimuli seen in SCA6 that correlate with symptom severity

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

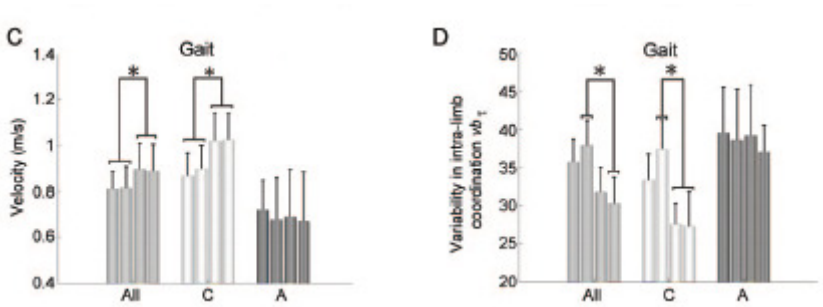
Improvements
Walking Speed
Reduction in falls
Maintained at 12 weeks

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

Intensive Co-ordination Training

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

4 weeks 3hrs / week

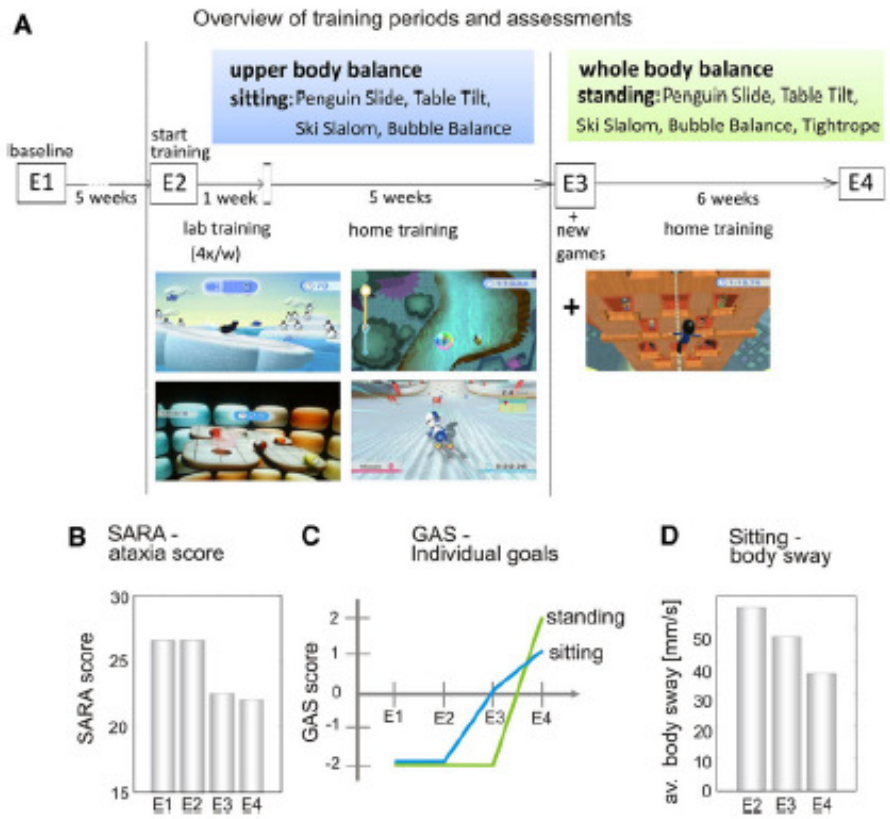


Improvements in Walking
And Co-ordination

Ilg et al 2009 Neurology 73 1823-1830

Restorative Approaches

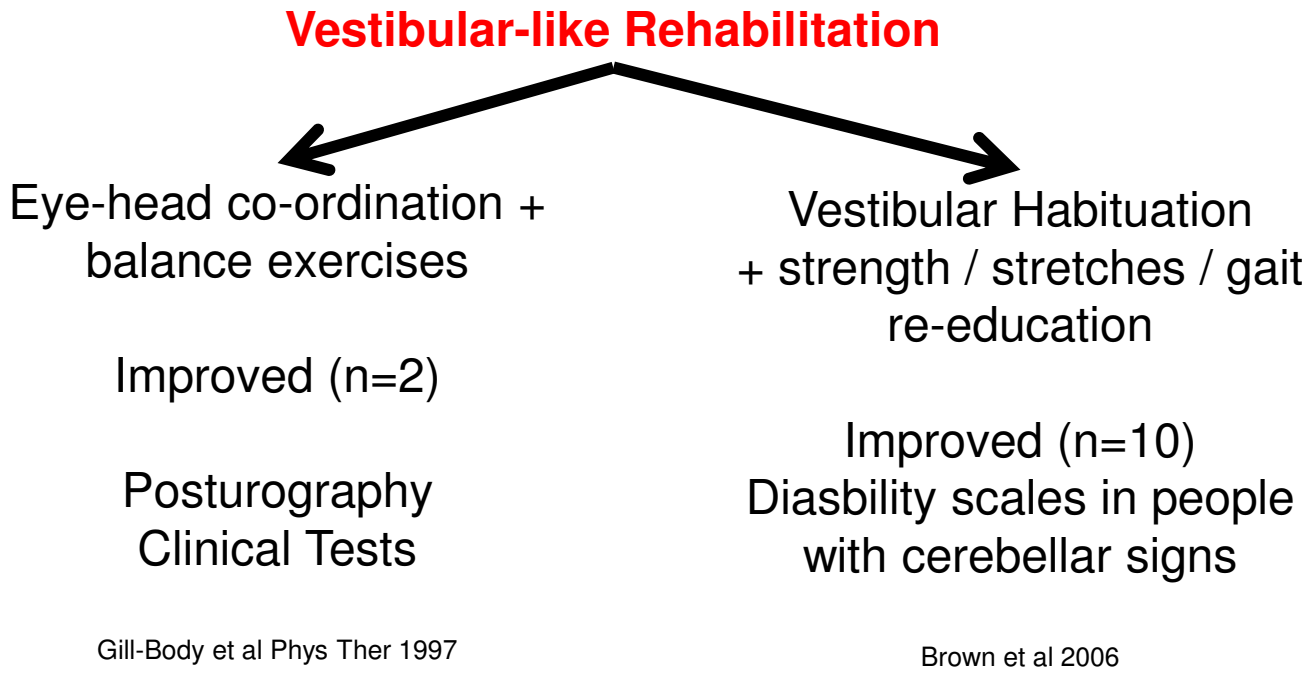
Video-gaming



N=1 – 10 Children
No control group

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

Restorative Approaches



Guidelines for Exercise Prescription

Training needs to be:

Intensive

3 hrs/ week for 4 weeks.
Boost Therapy

Maintained

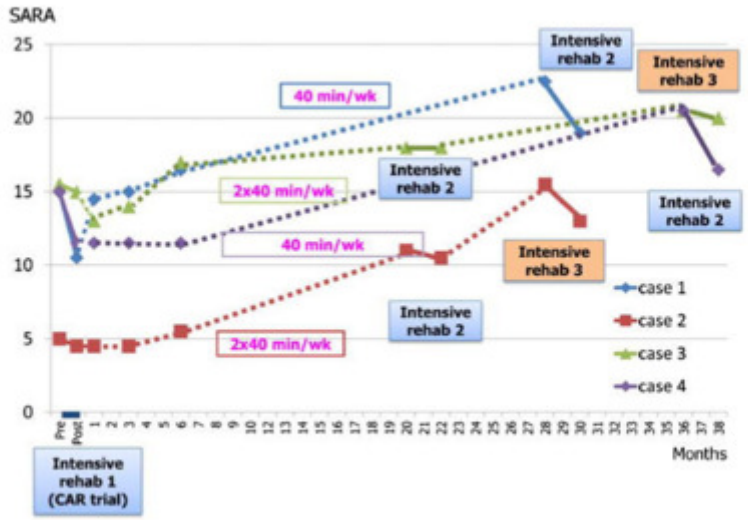
1hr/day homework? (80 mins/week ineffective)
Return to baseline in 6 months if no training

Challenging

Improvements greater in people who undertook
More challenging exercises

Individualised and Motivating

Part of overall exercise regime



Ilg, W. et al., 2014. Consensus paper: management of degenerative cerebellar disorders. Cerebellum, 13(2), pp.248–68

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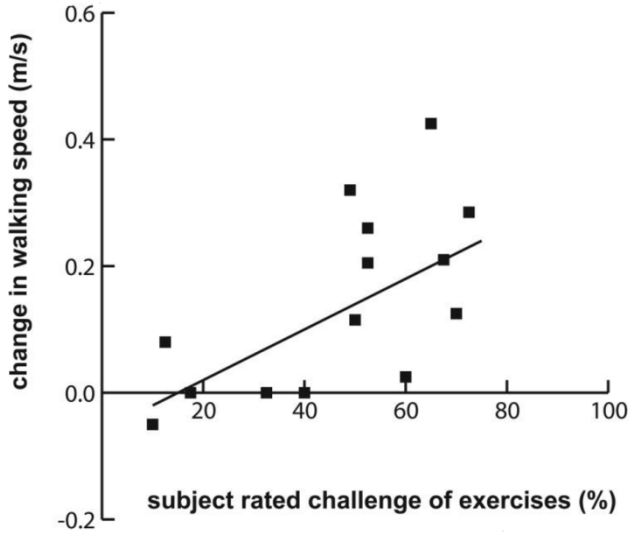


Keller, J.L. and A.J. Bastian, A home balance exercise program improves walking in people with cerebellar ataxia. Neurorehabil Neural Repair, 2014. 28(8): p. 770-8

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 **A**ctivity for people with **R**are Neurological **C**onditions

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

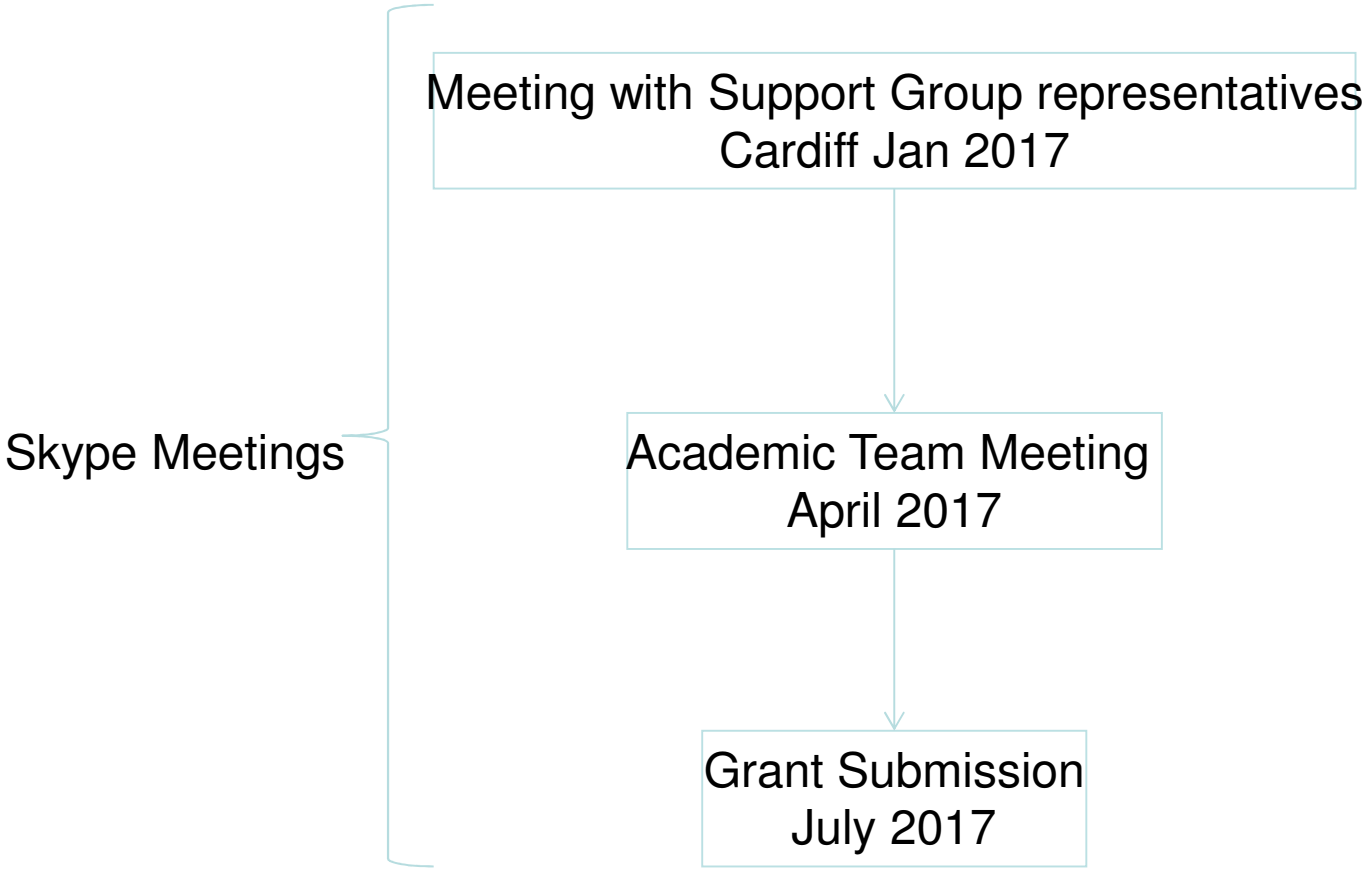
Spinocerebellar Ataxia

Hereditary Spastic Paraparesis

Neuromuscular Diseases

Progressive Supranuclear Palsy

Physical **A**ctivity for people with **R**are Neurological **C**onditions
PARC



Physical **A**ctivity for people with **R**are Neurological **C**onditions PARC

Glasgow
Lorna Paul

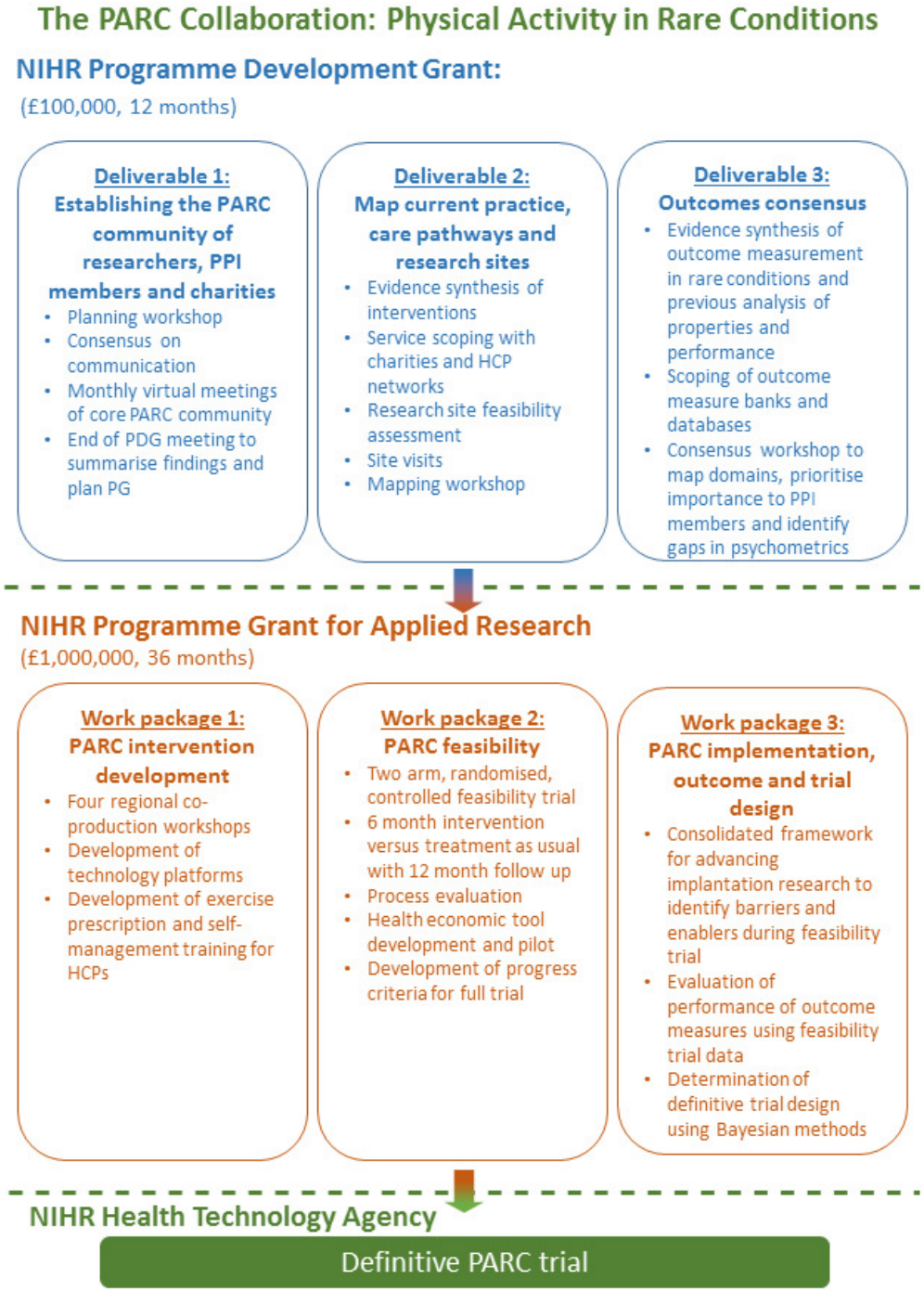
Cardiff
Monica Busse
&
Clinical Trial Unit

Newcastle
Lynn Rochester

Oxford
Helen Dawes

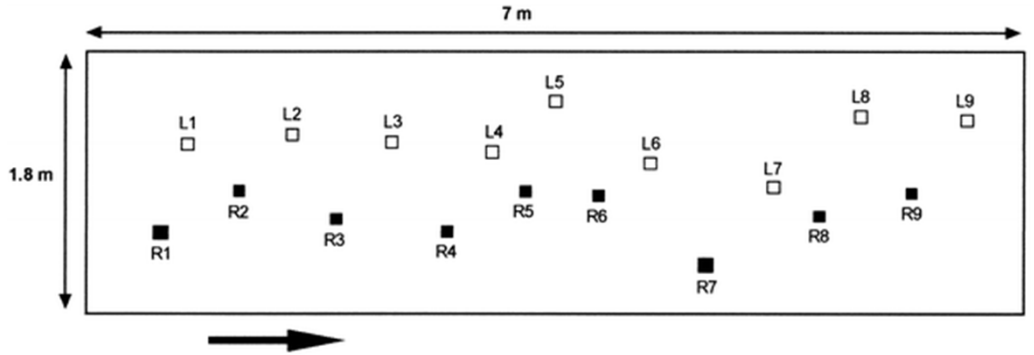
Plymouth
Jon Marsden

London
Gita Rhamdharry
(Chief Investigator)
Fiona Jones


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 scoping 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 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

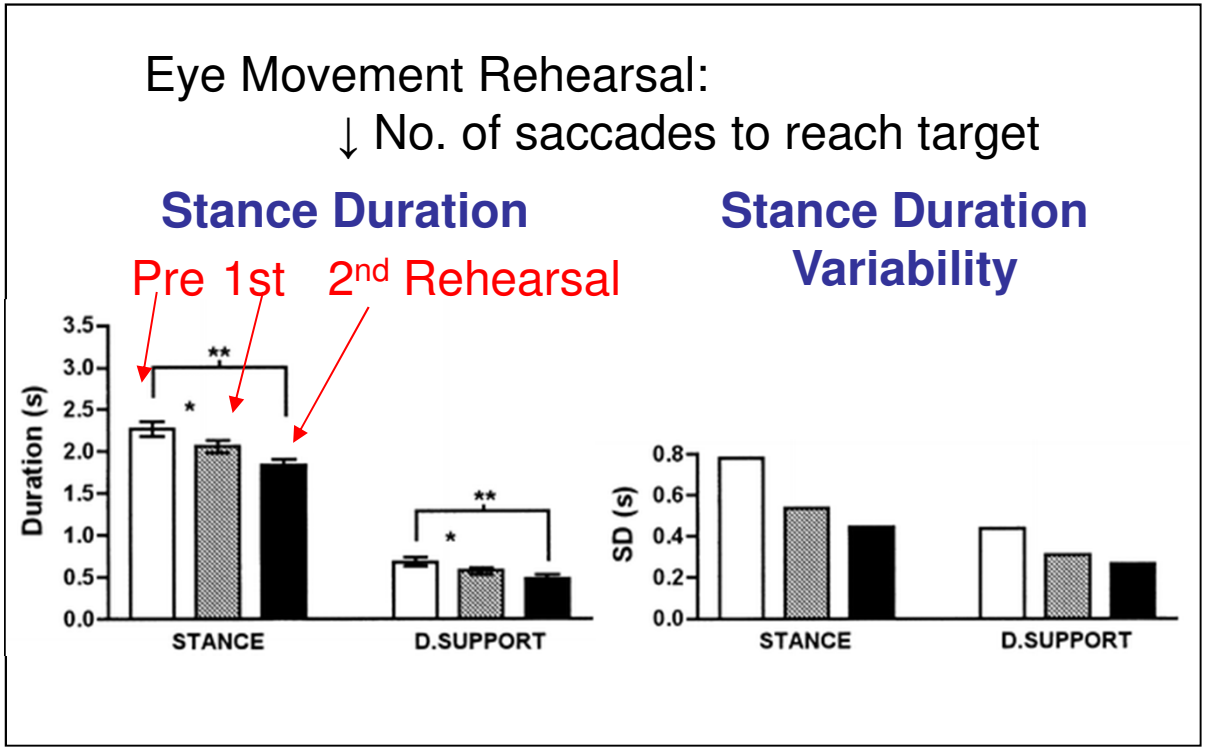


Saccade to target prior to start of swing phase

Ataxia ⟶ Several Hypometric saccades

Crowdy et al EBR 2000, 2002

Restorative Approaches
Oculomotor Retraining



Restorative Approaches
Oculomotor Retraining

OPTIMEYES

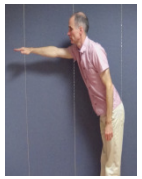
Oculomotor Re-training in Progressive MS with Cerebellar Signs

Intervention N=15 Vs n=13 Control

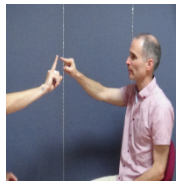
Clinical Measures

Box and Blocks

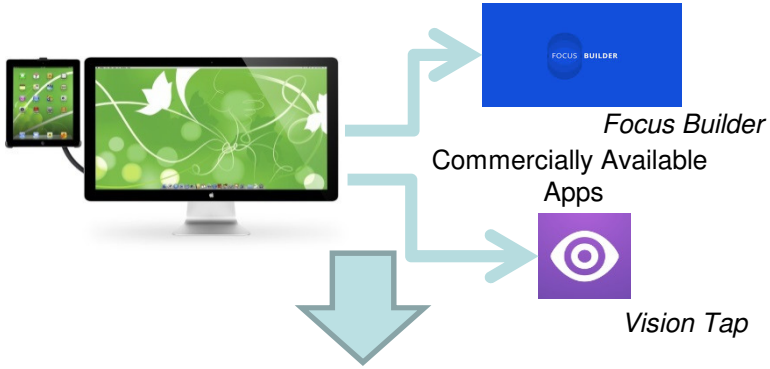
9 Hole Peg Test



Functional Reach



SARA



Focus Builder

Commercially Available Apps

Vision Tap

- Differences seen between Healthy Controls and pwMS

Significant changes in eye-hand tracking

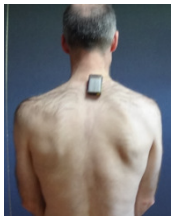
No change in clinical outcome

Subjective reports of improvements With balance in complex environments

Lab based measures of Motor Control

Eye-Hand Tracking

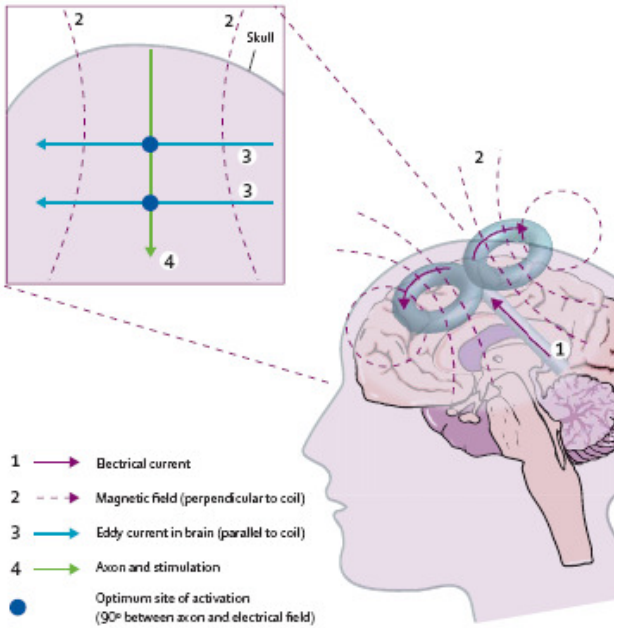
Smooth Pursuit & Saccades

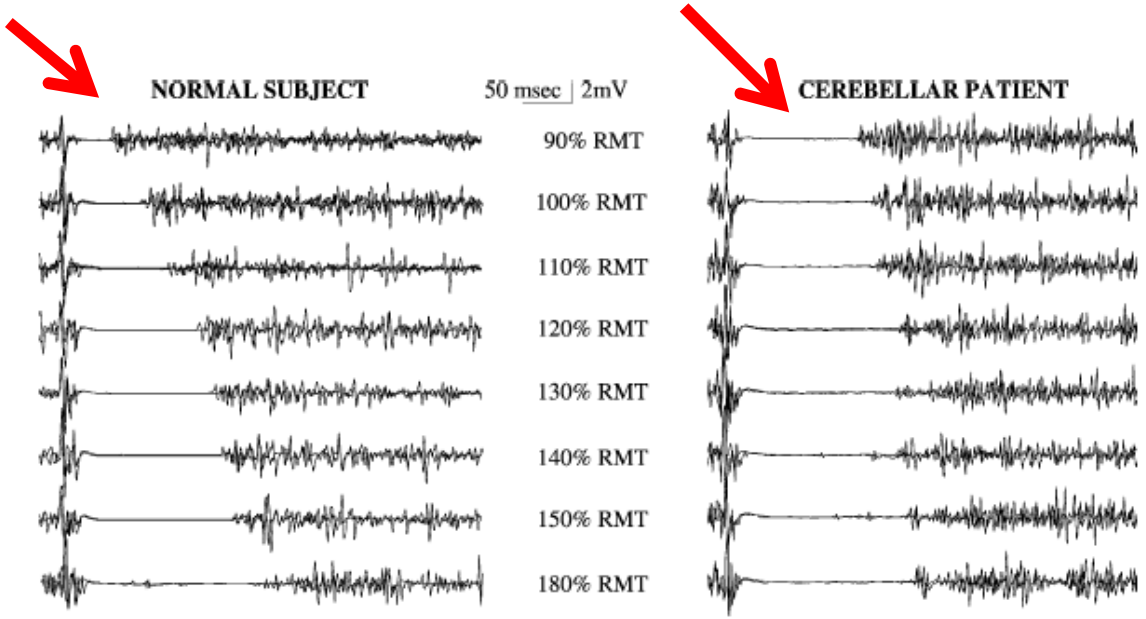


Postural Sway

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

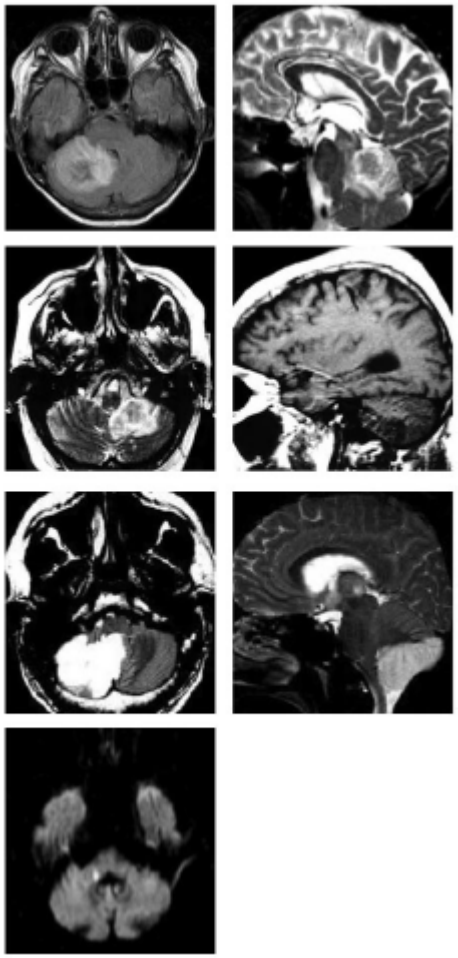
Restorative Approaches
Novel Methods

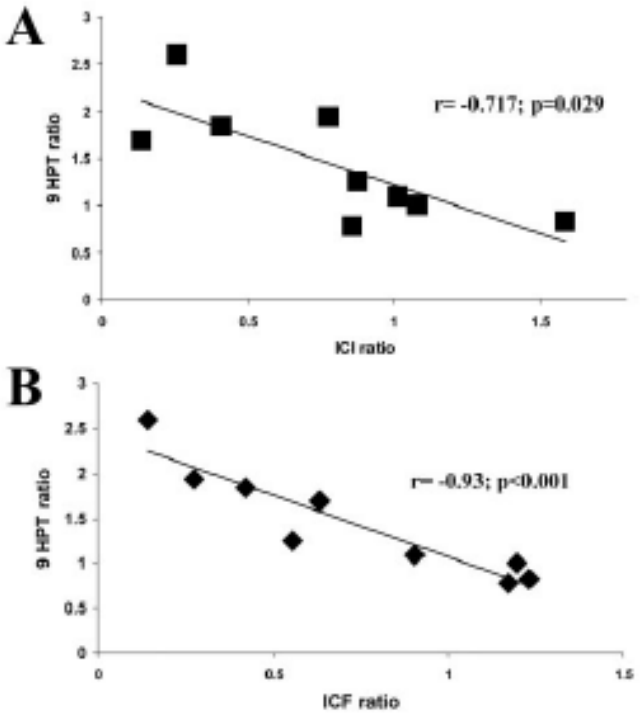
SCA Territory

AICA Territory

PICA Territory

R Dentate

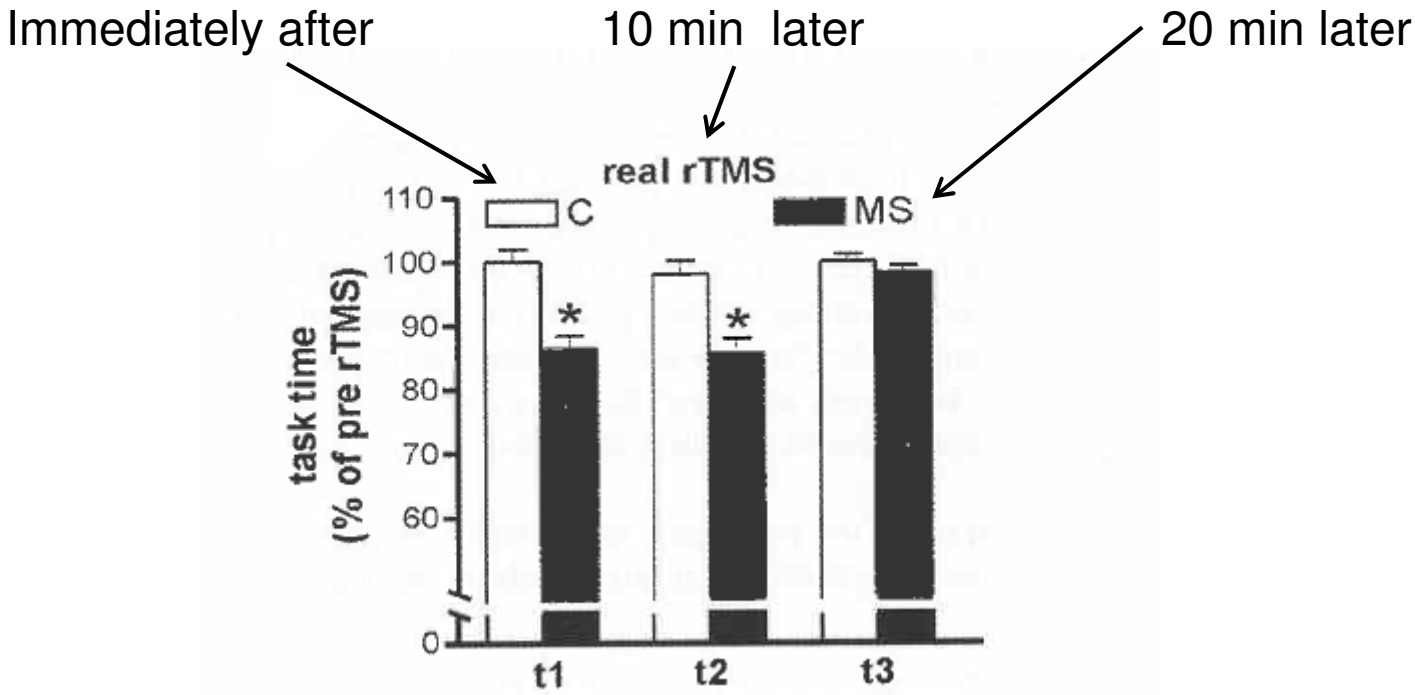




Changes in Motor cortex excitability
Correlates with changes in 9HPT following focal cerebellar lesions
Ratio of affected to unaffected hand given

Restorative Approaches
Novel Methods

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



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

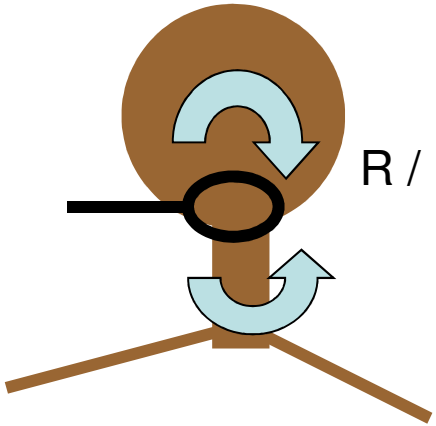
Blinded Ax

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)

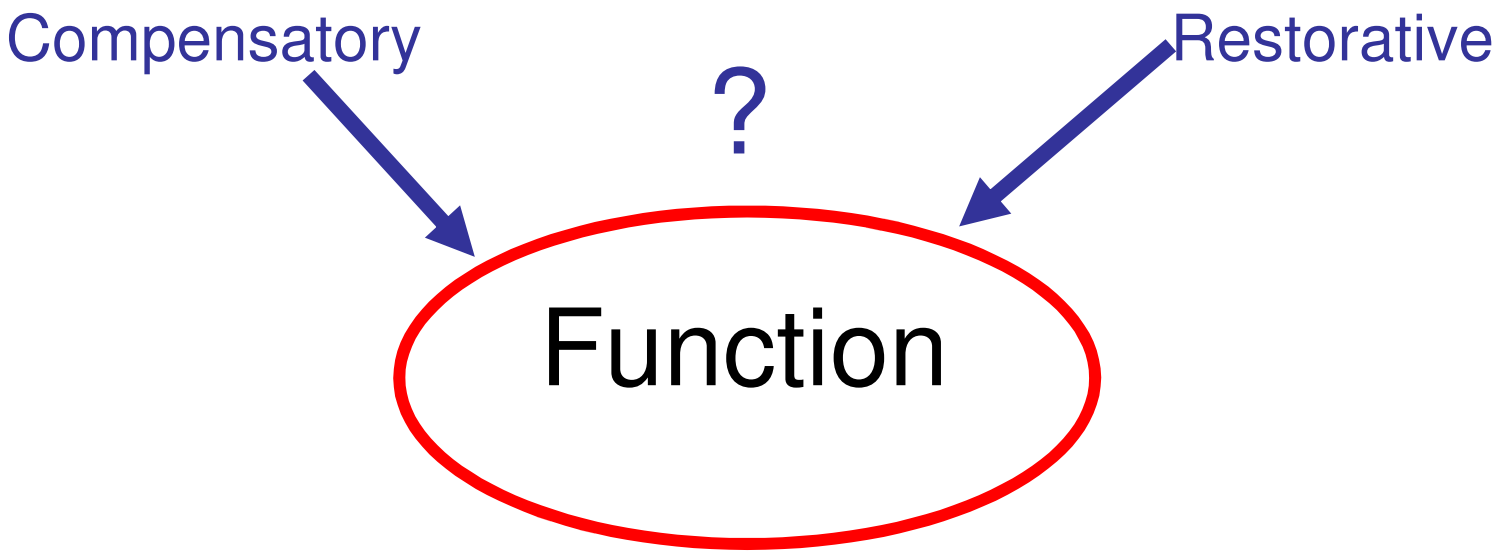
Gross Function
Walking speed
(Bennusi et al 2015, V 30, 1701-1705, Shiga et al J002 72 JNNP 124-126)
BUT
Some controlled Trials
show no effect
Kim et al 2014 J Rehabil Med 46 418-423

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



Which technique?

How long and at what intensity?

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