Cerebellar dysfunction may result in significant functional difficulties with upper and lower limb movement, oculo-motor control, balance and walking. Such difficulties can affect employability, increase carer burden and reduce a person’s perceived quality of life.\(^1\) The economic burden of people with spinocerebellar ataxia is estimated to be around €18 776 per annum.\(^1\) This article will review the signs and symptoms and pathophysiology of cerebellar ataxia, highlighting the theoretical basis of current non-surgical and non-pharmacological rehabilitation techniques.

**Search strategy and selection criteria**

References for this review were identified from a search of PubMed (from 1966 to 2009) and the Cochrane Library with the search terms ‘cerebellum or cerebellar’ and ‘ataxia or tremor’ and ‘physiology or pathophysiology or rehabilitation or therapy’). Articles were further identified through searches of reference lists in identified papers.

**Anatomy**

**Gross anatomy**

Cerebellar ataxia arises due damage or dysfunction affecting the cerebellum and/or its input/output pathways. In a rostro-caudal direction the cerebellum can be divided into three lobes: anterior, posterior and flocculo-nodular. Some of these may take the brunt of certain pathologies. The anterior lobe, for example, is particularly affected in alcoholic cerebellar degeneration.\(^2\) The cerebellum connects to the medulla, pons and midbrain via the inferior (restiform body), middle and superior cerebellar peduncles through which pass afferent and efferent pathways.

Functionally, the cerebellum is organized in a medio-lateral fashion with different areas of the cerebellum having a distinct anatomical...
input–output relationship resulting in a specific lesion–symptom relationship. The midline vermis connects to the fastigial deep cerebellar nucleus, while the flocculonodular lobe is connected with the vestibular nucleus; lesions to these midline areas result in deficits in posture, locomotion and oculomotor control. The intermediate zone of the adjacent cerebellar hemisphere mainly connects to the interpositus deep cerebellar nucleus (globose and emboliform nuclei in humans) and lesions here result in limb tremor and impairments in limb motion and dysarthria. The lateral cerebellar hemisphere connects to the dentate deep cerebellar nucleus; lesions here result in poor visuomotor coordination. More recently, the finding of extensive connections between the lateral hemisphere and prefrontal cortex in humans and greater apes has highlighted a role of the cerebellum in non-motor functions, such as working memory as discussed later.

Blood supply

The cerebellum is supplied by three main arteries that arise from the vertebrobasilar arteries. The posterior inferior cerebellar artery supplies the dorsal medulla (including parts of the vestibular nuclei and the inferior cerebellar peduncle) and the vermis and inferior cerebellum. The anterior inferior cerebellar artery supplies the lateral pons (including the facial nucleus and parts of the vestibular nuclei and the middle cerebellar peduncle) and the cerebellar flocculus and adjacent parts of the inferior surface of the cerebellum. The anterior inferior cerebellar artery also gives rise to the internal auditory and labyrinthine arteries. The superior cerebellar artery is the largest of the three arteries; it supplies the upper pons (including superior cerebellar peduncle) and the superior part of the cerebellum.

Cytoarchitecture

The cytoarchitecture of the cerebellar cortex is very uniform throughout its extent. The output cells of the cerebellar cortex, the Purkinje cells, mainly project to the deep cerebellar nuclei with the exception of cells within the flocculus (see above). Their dendrites are oriented in a parasagittal direction. The cerebellar cortex receives two main excitatory inputs: the climbing and mossy fibres. Climbing fibres arise from defined areas of inferior olives within the brainstem and project to a small number of Purkinje cells in specific longitudinal zones; one Purkinje cell receives one climbing fibre. Mossy fibres convey inputs from all other incoming pathways, such as the spinocerebellar tracts and inputs from the cerebral cortex via the pontine nuclei. These inputs synapse with the extremely numerous granule cells (numbering between 10^10 and 10^11) whose axons bifurcate giving rise to parallel fibres. The parallel fibres run for about 2–7 mm in a medio-lateral direction, synapsing in passing on Purkinje cells (up to 80 000 parallel fibres synapse with 1 Purkinje cell).

The uniformity of the cerebellar cytoarchitecture suggests that the cerebellum may be performing an identical computational function in these different areas. Many putative, not necessarily mutually exclusive, functions of the cerebellum have been put forward and include a role in: motor learning and adaptation; coordination; modulation of sensorimotor gain; timing; internal representation of the dynamics of the limb and sensorimotor transformations. At the heart of many of these theories lies the interaction between the climbing fibres and parallel fibres at the level of the Purkinje cell.

Understanding the exact role of the cerebellum in normal motor control will aid in our interpretation and understanding of cerebellar ataxia and ultimately help to guide treatment strategies.

Aetiology and prevalence

The aetiology of cerebellar ataxia is summarized in Table 1. Of the hereditary ataxias the prevalence of Friedreich’s ataxia is estimated at 2–5 per 100 000, while the spinocerebellar ataxias have a prevalence of between 0.9 and 3.0 per 100 000 depending on the exact type. The most common spinocerebellar ataxia is SCA6, which mainly affects the cerebellum with minimal additional extracerebellar pathology. Of the non-hereditary ataxias, the most common cause of cerebellar ataxia is multiple sclerosis (approximate prevalence of 100 per 100 000); here cerebellar signs are felt to occur in between 10 and 50% of cases, depending on the age of onset.
Signs and symptoms

The classical motor signs and symptoms of cerebellar ataxia are summarized in Table 2.

Traditionally, an ipsilateral cerebellar hemisphere lesion is felt to result in ipsilateral signs. This is because the cerebellar hemispheres connect with the contralateral cerebral cortex via the contralateral ventrolateral nucleus of the thalamus (also termed ventral intermediate nucleus in humans) and the cerebral cortex output; the corticospinal tract subsequently decussates. Recent findings, however, suggest that stimulation of the interpositus nucleus in primates can result in bilateral limb movements and that a unilateral lesion in humans can affect both limbs. These bilateral symptoms may be mediated via bilateral projections to subcortical sites such as the reticular formation. These projections may be useful in mediating bimanual coordination.

Pathophysiology

Limb movement

Dyssynergia and incoordination

People with cerebellar ataxia are slow to start movements; they have an increased reaction time. The movements themselves are prolonged in duration and they show a decreased maximal velocity and an increase in spatial variability, that is, the path that is followed varies from trial to trial. Variability in the spatial path is seen early in the movement before there is any time to process visual feedback, suggesting there is a problem with movement planning. This also accounts for the prolonged reaction time. In keeping with an important role of the cerebellum in planning predictive movements fast, ballistic movements that are entirely preplanned are inaccurate. However, inaccuracies in slower movements may also be seen as without predictive control these movements now over-rely on time-delayed feedback signals.

People with cerebellar ataxia in particular show marked deficits in multi-joint movements, called dyssynergia. This, in part, results from an inability to compensate for movement-associated interaction torques. Interaction torques are turning moments brought on by movement about one joint that influence the motion of adjacent joints. The deficits in multi-joint movements mean that people with cerebellar ataxia will tend to decompose their movements into simpler, more accurate single joint movements. In addition to showing deficits in coordination between the joints in one limb, abnormalities in intralimb coordination have also been described (but see also ref. 40).

The cerebellum may coordinate the activity in different effectors such as between the eye, arm, leg or head. Deficits in ocular control and limb control, for example during a reaching or stepping task, co-vary suggesting that they may be caused by common difficulties in programming coordinated movements. Such interactions mean that the accuracy of eye and limb movements performed in isolation may be further degraded during coupled activities, as usually occurs during functional activities.

Dysmetria and tremor

A triphasic pattern of muscle activation consisting of alternating agonist–antagonist–agonist activity is normally observed with fast single joint movements. Both animal and human studies highlight that following cerebellar dysfunction/inactivation there may be a prolonged duration...
# Table 2  
Motor signs and symptoms and tests in cerebellar disease

<table>
<thead>
<tr>
<th>Signs and symptoms of cerebellar ataxia</th>
<th>Clinical tests</th>
<th>Assess</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Limb movement</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dyssynergia</td>
<td>Finger to nose</td>
<td>Ability to move joints simultaneously/ decomposition of multijoint movements</td>
</tr>
<tr>
<td></td>
<td>Heel shin</td>
<td>Speed of motion</td>
</tr>
<tr>
<td>Dysemetria</td>
<td>Finger to finger test</td>
<td>Variability of spatial path</td>
</tr>
<tr>
<td></td>
<td>Great toe–finger test</td>
<td>Over/undershoot</td>
</tr>
<tr>
<td></td>
<td>(have trunk supported to isolate any limb dysfunction)</td>
<td></td>
</tr>
<tr>
<td>Tremor</td>
<td>Holding a position, e.g. (1) arms outstretched</td>
<td>Tremor amplitude and frequency</td>
</tr>
<tr>
<td>Kinetic</td>
<td>with palms down; (2) index to index (hold</td>
<td>Assess for titubation ~3Hz tremor of the head</td>
</tr>
<tr>
<td>Intention</td>
<td>two index fingers medially)</td>
<td></td>
</tr>
<tr>
<td>Postural</td>
<td>Move to/from target (see tests above)</td>
<td></td>
</tr>
<tr>
<td>Disdiadochokinesia</td>
<td>Alternating pronation–supination</td>
<td>Rate of movement</td>
</tr>
<tr>
<td></td>
<td>Alternating wrist flexion/extension while tapping</td>
<td></td>
</tr>
<tr>
<td></td>
<td>the thigh</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ankle dorsi-plantarflexion</td>
<td></td>
</tr>
<tr>
<td>Asthenia and hypotonia</td>
<td>Passive motion of the limb</td>
<td>Assess resistance to motion</td>
</tr>
<tr>
<td></td>
<td>Test muscle strength</td>
<td>Assess strength</td>
</tr>
<tr>
<td><strong>Balance and gait dysfunction</strong></td>
<td></td>
<td>Rate, direction and amplitude of sway</td>
</tr>
<tr>
<td>Balance and gait dysfunction</td>
<td>Stand with eyes open/closed</td>
<td>(± vision) and phase between upper and lower body</td>
</tr>
<tr>
<td></td>
<td>Stand in tandem/on one foot</td>
<td>Size of response to postural perturbation</td>
</tr>
<tr>
<td></td>
<td>Balance in response to perturbation, volitional upper/lower limb</td>
<td>Walking – accuracy of foot placement/ degree of sway/time in double stance/ stride length/base of support</td>
</tr>
<tr>
<td></td>
<td>Walking</td>
<td></td>
</tr>
<tr>
<td><strong>Oculomotor</strong></td>
<td></td>
<td>Ocular alignment/presence of nystagmus</td>
</tr>
<tr>
<td>Nystagmus (e.g. gaze-evoked; downbeat; rebound nystagmus)</td>
<td>Ability to fixate into finger ~30 cm away with</td>
<td>(rhythmic oscillatory movements of the eyes) type/presence of eye movements (e.g. flutter/ocular bobbing)</td>
</tr>
<tr>
<td></td>
<td>gaze in primary position and when looking at eccentric targets and on returning gaze to midline</td>
<td></td>
</tr>
<tr>
<td>Fixation deficits (e.g. flutter; macroscopic oscillations)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Saccadic smooth pursuit</td>
<td>Follow a target moving up/down/side to side</td>
<td>Number of catch-up saccades</td>
</tr>
<tr>
<td>Poor vestibulo-ocular reflex cancellation</td>
<td>Hold arms together out in front with thumbs pointing up. Fixate gaze on the thumbs while sitting in a chair that is moved side to side</td>
<td>Ability to maintain gaze fixation</td>
</tr>
<tr>
<td>Dysmetric saccades</td>
<td>Ability to rapidly shift gaze from one eccentric target to another (up and down/side to side)</td>
<td>Latency, velocity and precision (over/undershoot of the target as seen by the need to make more than one saccade to reach the target)</td>
</tr>
<tr>
<td>Reduced velocity of divergent eye movement</td>
<td>Ability to shift gaze from targets close to and far away from subject. Ability to follow targets moving to and from target</td>
<td>Latency, velocity and precision</td>
</tr>
<tr>
<td>Abnormal vestibulo-ocular reflex and optokinetic response</td>
<td>Ability to fixate on the target Movement of the eyes in the direction of rotation and re-alignment to the midline</td>
<td></td>
</tr>
<tr>
<td><strong>Dysarthria</strong></td>
<td></td>
<td>Intelligibility; rhythm; speed; presence of hesitations, accentuation of syllables and addition/omission of pauses</td>
</tr>
<tr>
<td></td>
<td>Ability to maintain sustained vowel phonation; repeat syllables</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Repeat a sentence speech/read a standard passage of text</td>
<td></td>
</tr>
</tbody>
</table>

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of the initial agonist burst that accelerates the limb and a delay in the onset of the subsequent antagonist muscle burst.\textsuperscript{50–54} This antagonist burst normally acts to decelerate the movement. Its delay therefore results in the person overshooting the target,\textsuperscript{55} a form of dysmetria. Although delayed, activation of the antagonist muscle still occurs. However, instead of now being pre-programmed by the cerebellum it is felt to be activated through stretching via a possible transcortical stretch reflex. This antagonist contraction can in turn stretch the agonist, resulting in stretch reflex-related activation of this muscle. Thus, alternating agonist–antagonist contractions driven by proprioceptive feedback may arise, resulting in a tremor.\textsuperscript{52,56–59}

Cerebellar tremor is a type of action tremor. It is seen while maintaining a posture (postural tremor) and while moving (kinetic tremor). Tremor at rest is not seen with pure cerebellar dysfunction. Cerebellar tremor is complex and in addition to proprioceptive feedback driving tremor other causes exist. When approaching a target there is a characteristic increase in tremor amplitude (intention tremor).\textsuperscript{60} Intention tremor may be explained by the effects of the inherent delay in visuomotor processing compounding the presence of an already incoordinated movement.\textsuperscript{61,62} Visuomotor tracking tasks in people with cerebellar ataxia have shown that the smoothness of tracking is improved when vision of either the target or the operator-controlled cursor is removed.\textsuperscript{53–67} Both conditions prevent visual feedback about the error between the target and the ongoing movement. It seems that people with cerebellar ataxia use this visual feedback to try to correct any abnormalities in tracking. However, the inherent delay in the time it takes to process visual feedback and to produce a motor output (around 120 ms) results in a delayed response during which time the limb may have moved in a non-predictable direction due to the underlying ataxia. In this case visually driven corrective responses would only serve to compound the inaccuracy in the movement, resulting in intention tremor.\textsuperscript{68} Although, avoiding visual feedback during and towards the end of a movement may improve the smoothness of the movement, this strategy may decrease movement accuracy.\textsuperscript{49,69,70}

\textbf{Force generation}

Asthenia, a generalized weakness, was described by Holmes (1922). This was more marked in the acute stages and with extensive cerebellar lesions and was not universally seen; indeed normal grip force has been reported in cerebellar ataxia.\textsuperscript{71} In contrast, a reduced rate of force generation (power) has frequently been reported\textsuperscript{6,72–74} and may in part underlie the difficulty in performing rapidly alternating movements (disdiadochokinesis) and contribute to the inability to adequately compensate for the higher interaction torques that are generated with faster movements.\textsuperscript{72,75,76}

People with cerebellar ataxia also show more variability in maintaining a constant level of force.\textsuperscript{77} The deficits in force generation may affect precise manipulative tasks. This, along with poor coupling between grip and load forces and inappropriate finger placement on an object, can lead to excessive self-generated torques acting on the object.\textsuperscript{6,78–80}

\textbf{Balance and gait dysfunction}

\textit{Postural sway and balance}

People with lesions affecting the anterior lobe, vermis and the interconnected fastigial nucleus have an increase in postural sway with an associated head and truncal tremor between 2 and 5 Hz.\textsuperscript{81–86} The sway is greatest in the antero-posterior direction. Instability in the medio-lateral direction is also seen in response to alterations in the available sensory input, following a postural perturbation or while stepping.\textsuperscript{87–89}

Impairments in both postural responses and anticipatory postural adjustments have been described.\textsuperscript{90} The temporal coordination between anticipatory postural adjustments and volitional movements, for example, is impaired.\textsuperscript{91} A lack of such anticipatory adjustments will result in imbalance during self-generated movements, for example, not leaning forwards prior to standing on tiptoes will result in the person pushing themselves backwards.

In response to an unexpected perturbation of the support surface, hypermetric postural responses are observed.\textsuperscript{87,95–94} This seems to reflect an inability to set the correct size (or gain) of the response. Similar to healthy participants,
however, people with cerebellar ataxia are able to decrease the size of the postural response with repeated presentations of a predictable postural perturbation and change their response magnitude appropriately to an expected change in the size of perturbation, although the overall size of the response remains increased.92,95–98

Gait and falls

While walking, people with cerebellar dysfunction show prolonged time in double stance, poor inter-limb coordination99,100 and an increased variability in their stride length and individual joint kinematics,101–103 although their base of support may not be increased as is often presumed.104 Although primary deficits in balance can have a direct and marked impact on walking,105 dysmetria/dyssynergia affecting the lower limbs can also impact on dynamic balance while walking.99,106 When one foot is off the ground while walking, the body’s centre of mass does not usually lie over the base of support and so the body is in a state of disequilibrium; it will tend to fall away from the stance leg. The trajectory taken by the body while walking is preprogrammed and tightly coupled to the future placement of the stepping foot that acts to catch the ‘falling’ body.107,108 Incorrect foot placement due to incoordination of the lower limb, as is seen in cerebellar ataxia, can therefore result in poor dynamic balance while walking. A more medial foot placement than required, for example, can result in the body falling laterally.99

Deficits in balance and walking may contribute to the high reported incidence of falls in cerebellar ataxia.82 However, the causes of falls are often multifactorial and the relative contribution of intrinsic and extrinsic (environmental) factors in falls aetiology in cerebellar dysfunction remains to be elucidated.

Oculomotor control

Normal oculomotor function is heavily dependent on the cerebellum for adaptive control (plasticity).109,110 It helps to understand cerebellar oculomotor dysfunction with reference to three major cerebellar oculomotor areas: the flocculus/paraflocculus, the nodulus and the vermis, as each gives rise to well-recognized (and recognizable) oculomotor ‘syndromes’111 that may or may not occur together or be associated with other cerebellar signs.111–113 These syndromes can be useful diagnostic clues and may account for visual symptoms (oscillopsia, dizziness, diplopia).114 However, due to the heavy connections with the brainstem, oculomotor centres in the pontomedullary junction (horizontal eye movements) and the midbrain (vertical and torsional eye movements), it is often not possible to differentiate between brainstem and cerebellar lesions.

The flocculus/paraflocculus controls the modulation of retinal image motion (retinal slip) and maintains steady fixation in eccentric gaze directions (the neural integrator), accurate smooth pursuit, optokinetic nystagmus and normal vestibulo-ocular reflex gain. Visual input from retina and visual cortex is relayed to the inferior olive via pretectal nuclei, and uniquely, Purkinje outputs synapse with brainstem centres (bypassing the deep nuclei). Cardinal clinical signs of the ‘floccular syndrome’ are gaze-evoked nystagmus on lateral gaze and jerky (saccadic) smooth pursuit (ipsiversive to the lesion).115–121 Rebound nystagmus may also be present. Vestibulo-ocular reflex suppression is impaired which can be readily detected by the failure of a (seeing) patient to suppress post-rotatory vestibular nystagmus.122–127

Vertical eye movements may be normal, but in progressive disorders downbeat nystagmus emerges.128 Acute or chronic alcoholism may yield similar signs and may be irreversible. Downbeat nystagmus can give rise to constant oscillopsia (the illusion of oscillation of the visual surround) and poor visual acuity that is unrelated to head movement and position.113 Abnormalities in the direction and in the gain (both increases and decreases) of the angular and linear vestibulo-ocular reflex have been reported.125,129–135 Abnormalities in the vestibulo-ocular reflex can degrade visual acuity with head motion. Vertical translational head motion is especially prominent while walking, and abnormalities in the vertical vestibulo-ocular reflex may lead to falls.133
**The nodulus (and ventral uvula)**

Nystagmus is observed when people are rotated in the dark at a constant angular velocity; the slow phase being generated by the vestibulo-ocular reflex with the fast phase serving to reposition the eyes centrally in the orbit. The head velocity signal derived from vestibular nerve afferents is felt to be stored within brainstem circuits. Evidence for this storage mechanism comes from the observation that with a constant velocity rotation in the dark, the velocity of the slow phase decays exponentially with a time constant of \( t = 15–20 \text{ seconds} \), slower than the decay in vestibular nerve afferent signals.\(^{136}\) The nodulus modulates this vestibular time-constant, and lesions thereof tend to increase the vestibular time-constant beyond normal, and can lead to periodic alternating nystagmus, in which the horizontal jerk nystagmus spontaneously reverses direction with a period of tens of seconds.\(^{111}\) Periodic alternating nystagmus is often missed due to the need to observe nystagmus for some minutes. Midline cerebellar lesions can also alter the time constant of perceptual vestibular-mediated self-motion.\(^{137}\)

**The dorsal vermis and fastigial nucleus**

The dorsal vermis (lobules 6 and 7) and the fastigial nucleus are important for the calibration of saccades. The ‘vermis syndrome’ is characterized by saccade hypo- or hypermetria.\(^{122,138,139}\) Detection of dysmetria requires careful observation, but extreme hypermetria may give rise to saccadic oscillations (distinct from nystagmus) and is suggestive of a nuclear lesion. Smooth pursuit and slow vergence movements may also be affected; in particular the velocity of divergence is reduced.\(^{140}\) In children, congenital or acquired vermian lesions also lead to difficulties in triggering saccades (‘saccade initiation failure’ or ‘congenital ocular motor apraxia’) which is often associated with speech apraxia due presumably to co-involvement of vermian speech areas (see section on Dysarthria, communication and language).

Saccade speed appears not to be controlled by the cerebellum, and saccades that are clinically recognizable as being slow are usually caused by brainstem disease or drug toxicities.

Vertigo may be experienced secondary to lesions affecting the midline vermis and flocculonodular lobe.\(^{141,142}\) Prolonged isolated vertigo with imbalance that mimics symptoms of vestibular neuritis may be observed in \( \sim 10\% \) of cases of discrete cerebellar lesions. These most commonly affect the medial branch of the posterior inferior cerebellar artery territory.\(^{143}\) Vertigo may also be positional and accompanied by positional evoked nystagmus,\(^{142,144,145}\) thus mimicking benign paroxysmal positional vertigo. In contrast to benign paroxysmal positional vertigo, the nystagmus may be either vertical (up or downbeat) or torsional in direction with the eyes straight ahead, be persistent and change in direction with different head positions.\(^{146}\) Additional signs that may alert the clinician to the presence of a central cause of vertigo include the presence of skew deviation, gaze evoked nystagmus towards the affected ear (in the opposite direction to that seen with peripheral vestibular neuritis), vertical saccadic pursuit; a normal head thrust test and minimal increase in the slow phase velocity of spontaneous nystagmus in the dark compared to fixation in the light as well as the presence of associated neurological signs such as dysmetria/ dysynergia or dysarthria.\(^{147,148}\)

**Dysarthria, communication and language**

Dysarthria is commonly observed with lesions affecting the rostral paravermal region of the anterior lobes.\(^{3,149–151}\) Impairments in both articulation and prosody have been described.\(^{152}\) Commonly speech is described as scanning in nature consisting of hesitations, accentuation of some syllables and the addition of pauses or omission of appropriate pauses; in around 50\% of cases slurring may be evident.\(^{153}\) In addition, speech can be reduced in rate, and show signs of vocal instability, increased monotony, equalized stress and imprecise consonants.\(^{154}\) Acoustic analysis reveals more variable/longer syllable production and pause durations, reductions in vowel length contrasts and differences in the frequency spectrum from those in healthy controls.\(^{154–156}\) Speech intelligibility may be affected by difficulties in distinguishing the difference between plosives (e.g. \([p]\), \([t]\), \([k]\)) at the end of words.\(^{157}\)
Kinematic analysis of oro-facial movements highlights similar changes as previously described for limb movements, such as prolongation of movement duration; decreased maximal velocity and prolonged muscle bursts. Furthermore, a 3 Hz ‘postural’ tremor may be detected during sustained phonation of vowels and oral disdiadochokinesia can be detected during rapid syllable repetition, although this may not predict the syllabic rate during sentence production.

Language impairments that are not attributed to dysarthria, such as poor understanding of speech, reading and naming tasks, and agrammatism have been reported following cerebellar damage (although see ref. 166). These are felt to arise from disruption to reciprocal pathways between the right cerebellar hemisphere and left cerebral hemisphere and highlight the role of the cerebellum in pre-articulatory speech. Mutism following posterior fossa tumour resection in children has also been described in about 29% of cases. The mutism may not develop immediately and is usually self-limiting. Here damage to the vermis has been implicated, although cerebellar damage leading to a temporary reduction in activity in the interconnected cerebral cortex has also been suggested.

Non-motor symptoms

In the last two decades accumulating evidence suggests that isolated damage to the cerebellum may also result in a cerebellar cognitive affective syndrome. This syndrome includes non-motor symptoms such as poor executive function (e.g. reduced verbal fluency and working memory); impaired spatial cognition (e.g. poor visuo-spatial memory) and linguistic difficulties (e.g. dysprosodia and agrammatism) and has been described in both acquired and hereditary cerebellar dysfunction. The symptoms are more marked in the acute/subacute stages of damage/dysfunction, particularly when the posterior lobes of the cerebellum are affected bilaterally (e.g. following an infarction within the posterior inferior cerebellar artery territory). Further, affective symptoms such as apathy and disinhibited behaviour may occur with damage to the midline vermis. These symptoms are felt to reflect damage to the extensive reciprocal pathways between the cerebellum and the posterior parietal, superior temporal, prefrontal and parahippocampal cortices. Thus, the role of the cerebellum in planning and ongoing control of motor function may therefore have a corollary in non-motor behaviours.

Such non-motor symptoms may further impact on rehabilitation, for example, by affecting the ability to retain information conveyed through verbal or visual instructions; to perform abstract reasoning or to initiate activities.

Implications for rehabilitation approaches

As highlighted in recent systematic reviews of randomized controlled trials there is currently a lack of high-quality research studies into the rehabilitation of cerebellar ataxia. Despite this, the literature on the treatment of cerebellar ataxia describes some approaches that warrant further investigation. Approaches may be broadly divided into those that aim to improve functional ability by compensating for the underlying deficit and those that aim to improve function through restorative techniques that involve adaptation and recovery within the neuro-musculoskeletal system.

Compensatory approaches

Compensatory approaches include the use of strategies to encourage decomposition of movement into simpler single joint movements; visual and verbal cues to aid walking speed and stride length; the use of assistive technology to aid computer use; and aids such as customized seating and frames to help posture, balance and mobility. Increasing the visco-elastic resistance or the inertia of a limb will dampen the tremor and serve to decrease the speed and size of the stretch reflex that may drive the tremor in some cases. Thus aids that increase viscous resistance such as the Neater Eater and Mouse Trap (http://www.neater.co.uk/main.htm) have been recommended although their effectiveness has not been thoroughly investigated. The visco-elastic
resistance offered by Lycra garments\textsuperscript{195} may also underlie their proposed use in improving proximal and truncal stability and function in people with cerebellar signs. However, any functional improvements with Lycra garments in reported single-case studies to date need to be considered alongside the possible inconvenience and additional assistance required in applying the garment, leading to a potential loss in independence.\textsuperscript{196}

Increasing inertia by loading the appendicular or axial skeleton may also dampen tremor and reduce dysmetria.\textsuperscript{63,197,198} Studies into the effectiveness of weights in improving upper limb function are variable, with improvements and deterioration in function as well as a lack of effect being reported.\textsuperscript{49,199–201} This may be because the addition of a load requires adaptive scaling of agonist–antagonist activity which is affected in cerebellar disease. Thus, although an increase in agonist activity required to initially accelerate the limb may be seen, this may not be accompanied by an increase in antagonist ‘braking’ activity. Ultimately, this results in an increase in overshooting and potential deterioration in function.\textsuperscript{201} In fact the use of additional load may be beneficial as an assessment aid in revealing minimal hypermetria.\textsuperscript{202} Loading the trunk may aid balance, and here the loading may be either symmetrical or asymmetrical to counterbalance a directional impairment in balance (e.g. anterior loading is provided to counterbalance falling backwards). Although it is not always clear whether the ataxia is of pure cerebellar origin, case reports of these interventions show a favourable outcome on clinical measures of walking and balance.\textsuperscript{197,203,204}

Cooling a limb can also temporarily reduce cerebellar tremor. This may occur through a reduction in muscle thixotropy and increase in muscle stiffness as well as a reduction in nerve conduction velocity and muscle spindle afferent feedback.\textsuperscript{205,206} Cooling-related reductions in tremor resulting in functional improvements have also been reported in people with essential tremor.\textsuperscript{207,208}

Restorative approaches

The uses of defined restorative approaches targeting a specific symptom are rare and involve uncontrolled case reports. With prolonged interventions (3–12 months) improvements in interlimb coordination\textsuperscript{209,210} and a reduction in force variability have been described.\textsuperscript{191} Biofeedback of different aspects of motor control has been assessed. Biofeedback of EMG patterns during a visuomotor tracking task led to an improvement in muscle activation on the trained task after several weeks of training although the effects on functional tasks were not investigated.\textsuperscript{211} Biofeedback of muscle activity has also been described in combination with relaxation therapy to decrease tremor resulting in improved feeding.\textsuperscript{212,213} Biofeedback of the centre of pressure motion (a measure of postural sway) linked to a computer game resulted in improvement in measures of balance and falls and the use of a computer game was linked to increased practice time.\textsuperscript{214}

Other studies have assessed the effect of multicomponent approaches on balance and walking. Some of these are the subject of recent systematic reviews.\textsuperscript{185,186} Balance and ocular exercises have led to an improvement in clinical measures of postural stability and walking in people with cerebellar ataxia.\textsuperscript{215} Although this study involved small numbers ($n = 2$), the techniques utilized are based on those that are effective in treating deficits affecting the peripheral and central vestibular system with which the cerebellum has large reciprocal connections. Brown \textit{et al.}\textsuperscript{216} also used vestibular habituation training in combination with strengthening, stretching and gait re-education. Significant improvements in disability scales (Disability Handicap Inventory; Dynamic Gait Index) were seen in the subgroup of people with cerebellar ataxia ($n = 10$). Increases in walking distance following either locomotor training using body weight support on a treadmill (5 days/week for four months) or by encouraging people to decrease the amount of fixation of the arms when walking\textsuperscript{217,218} have been reported.

Improvements in standing balance have also been seen with a combination of strength and balance exercises.\textsuperscript{219,220} Both groups utilized Frenkel’s exercises that were originally developed for people with sensory ataxia secondary to tabes dorsalis (Frenkel, 1902). These exercises emphasize a reliance on visual feedback to control movement. Given that visual feedback can improve balance/postural sway in cerebellar dysfunction,\textsuperscript{81,221}
this may be a useful approach although an over-reliance on vision in cerebellar dysfunction resulting in visual vertigo has been described. It would be interesting to contrast the reliance of different sensations on balance following this approach with that seen after the vestibular habituation/rehabilitation exercises described above.

Several small-scale studies on training ocular control and dysarthria highlight the potential value of further intervention studies in these areas. In a stepping task that defined the target for foot placement, Crowdy et al. found that saccadic and foot placement accuracy could be improved by prior practice in making saccadic eye movements towards the targets. A single-case study of a person with cerebellar dysfunction secondary to thiamine deficiency assessed the effects of Lee Silverman voice treatment. This approach emphasizes loudness of phonation and is more commonly used in the treatment of people with Parkinson’s disease. After a four-week intervention period (16 sessions), improvements in acoustic measures (sound pressure level) and perceptual measures of articulation/phonation were observed and their employer reported increased satisfaction over the effectiveness of communication via the telephone.

More recently, the use of motor cortical stimulation as a treatment paradigm has been assessed. These follow animal and human studies showing that cerebellar dysfunction results in a reduction in measures of motor cortex excitability. Koch et al. investigated the effect of rapid transcranial magnetic stimulation (5 Hz) over the motor cortex. The frequency and paradigm of stimulation used had previously been shown to result in an increase in motor cortex excitability and was associated in the cerebellar ataxic group with an improvement in hand function (as determined by the 9-hole peg test) compared to healthy controls.

Direct stimulation of the cerebellum has also been investigated. Stimulation of the cerebellum using transcranial magnetic stimulation in healthy participants has also been shown to modulate cortical excitability. In people with spinocerebellar degeneration, cerebellar stimulation was applied over several sessions with treatment lasting from 21 days to eight weeks. Improvements in clinical measures of ataxia, balance and gait were reported although there were no control groups. The mechanisms underlying any functional improvements with stimulation of the cerebellum remains unclear given the fact that transcranial magnetic stimulation over the cerebellum may in part affect motor cortex excitability by stimulating adjacent peripheral nerves.

Understanding when to apply restorative or compensatory strategies remains unclear. The relative use of compensatory strategies may vary and possibly depends on disease progression and severity.

Potential mechanisms and barriers to recovery

The potential mechanisms of recovery following cerebellar damage have not been extensively studied. The results of serial experimental cerebellar lesions in primates suggest that following unilateral cerebellar lesions, important structures mediating recovery are the opposite cerebellar hemisphere, deep cerebellar nuclei, as well as extracerebellar sites within the brainstem and sensorimotor cortex, the latter depending on whether volitional limb movements or walking/balance is being investigated. Extrapolation of such findings to humans should be made with caution due to differences in neuroanatomy.

A cerebellar lesion may also cause loss of activity within the cerebral cortex due to the large interconnections between these structures. Resolution of such disachisis may also underlie symptom recovery. In humans with cerebellar dysfunction, for example, an increase in the activation of the medial premotor system (supplementary motor area) while moving has been reported; this may compensate for the lack of activation of the lateral premotor areas that receive extensive inputs from the cerebellum. Furthermore, following a unilateral lesion, imbalances in activity between left and right cerebellum and their interconnected cortical structures may also contribute to symptom presentation. Torriero et al. hypothesized that isolated damage to the left cerebellum may reduce excitatory drive to the contralateral right dorsolateral prefrontal cortex, resulting in an imbalance in activity between the left and right
cortex. Re-addressing this imbalance temporarily by inactivating the left dorsolateral prefrontal cortex using rapid transcranial magnetic stimulation resulted in an improvement in a procedural learning task.\textsuperscript{259} Such findings are in keeping with evidence from subcortical stroke, where changes in interhemispheric inhibition from the unaffected to the affected side may contribute to paresis.\textsuperscript{260}

The cause, site and extent of the lesion are important predictors of the degree of functional recovery. Functional deficits seem more marked following a haemorrhage compared to an infarct, and superior cerebellar artery strokes have a worse prognosis than strokes affecting the posterior and anterior inferior cerebellar arteries\textsuperscript{261,262} (although see ref. 263). Superior cerebellar artery strokes may be associated with damage to the dentate nucleus and the superior cerebellar peduncle, the main output pathway of the cerebellum. This is in keeping with other studies highlighting worse functional recovery with lesions affecting the output pathways of the deep cerebellar nuclei and superior cerebellar peduncle.\textsuperscript{70,264}

Extracerebellar damage seems to be a poor prognostic indicator of functional recovery.\textsuperscript{265} Functional recovery after a cerebellar stroke, for example, is worse when people initially present with global signs such as loss of consciousness/weakness, as opposed to isolated focal signs such as ataxia or vertigo.\textsuperscript{261,266,267} Following stroke, increasing age is also associated with worse functional outcome,\textsuperscript{267} although this may in part be explained by the association of age-related additional extracerebellar white matter lesions.\textsuperscript{268}

Against an effect of age on prognosis is the finding that the ability to compensate for a cerebellar tumour post resection does not seem to be better in young children (<4 years old) than in older children, adolescents and adults.\textsuperscript{264,269}

In other pathologies the presence of additional cerebellar dysfunction strongly impacts on recovery. The recovery from a peripheral vestibular nerve lesion and the recovery from a brainstem stroke is less pronounced in the presence of an additional cerebellar lesion.\textsuperscript{270,271} This may simply reflect an increasing accumulation of pathology. However, it may also reflect that the cerebellum normally plays an important role in motor learning and the recovery of symptoms following damage to other central and peripheral nervous system structures.

**Motor learning, adaptation and recovery**

The role of the cerebellum in motor learning is being investigated at a subcellular, cellular and systems level in both animals and humans and using neural networks and modelling.\textsuperscript{18} The cerebellum is part of a distributed system that is involved in motor learning and is felt to play a pivotal role in error-based motor learning and adaptation.\textsuperscript{272,273} In keeping with this, people with cerebellar ataxia may show greatly impaired learning of both simple and complex motor skills such as visuomotor adaptation,\textsuperscript{274–276} serial reaction time tasks,\textsuperscript{277,278} adaptation to prism glasses\textsuperscript{279} and force fields,\textsuperscript{280} as well as classical conditioning paradigms when the interval between the unconditioned and conditioned stimulus is short (~400 ms).\textsuperscript{281–285} Importantly, such adaptation is constantly occurring in everyday life. Tasks such as adapting the arm to its new ‘length’ and dynamics when holding an object (e.g. a pen), adapting the vestibulo-ocular reflex when glasses are put on and adapting to the effects of muscle fatigue\textsuperscript{286} with repetitive activities all seem to require cerebellar activity.

Poor recovery following a cerebellar lesion may be a consequence of damaging structures critically involved in learning and relearning of motor skills. However, following a cerebellar lesion learning and adaptation over time has been described, albeit at a lower rate and extent than that in healthy controls.\textsuperscript{73,98,287} It remains to be seen whether this reflects activity within undamaged areas of the cerebellum or activity in extracerebellar structures.

**Conclusion**

An understanding of the pathophysiology of cerebellar ataxia and the mechanisms of recovery can help to guide the development of treatments and to optimize current interventions. Interpreting studies of pathophysiology in turn crucially depends on our understanding of the healthy cerebellum in both motor and non-motor functions. Although there is currently a lack of high-quality
research studies into the rehabilitation of cerebellar ataxia, current studies highlight some potential avenues that warrant further investigation. Future studies should aim to identify the site and extent of both cerebellar and extracerebellar pathology as these may crucially determine clinical presentation and prognosis. A detailed description of the person’s presenting impairment profile should be coupled with appropriate outcome measures of the effects of intervention on a person’s ability and participation.

**Clinical messages**

- The key role of the cerebellum in motor learning and in adaptation following extracerebellar pathology may limit functional recovery in people with cerebellar dysfunction.

- Associated cognitive and affective signs may further impact on function and the rehabilitation process.

- There is limited evidence about the effectiveness of interventions in people with cerebellar dysfunction although clinical improvements in balance, walking and upper limb function have been reported. A combination of restorative and compensatory techniques may be utilized; the relative emphasis depending on the severity of cerebellar ataxia and its pattern of progression.

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