REVIEW ARTICLE

High Altitude Ataxia—Its Assessment and Relevance

Brynn A. Bird, MB; Alexander David Wright, MD; Mark H. Wilson, MD; Brian G. Johnson, MB;
Chris H. Imray, MB BS, PhD; Birmingham Medical Research Expeditionary Society

From the Medical School, University of Birmingham, Edgbaston, Birmingham, UK (Dr Wright and Birmingham Medical Research Expeditionary Society); Sandwell Hospital, Lyndon, West Midlands, UK (Dr Bird); Department of Neurosurgery, the National Hospital for Neurology and Neurosurgery, Queen Square, London (Dr Wilson); the Surgery, Chirk, Wrexham, UK (Mr Johnson); Warwick Medical School and UHCW NHS Trust, Coventry, UK (Dr Imray).

Ataxia at altitude is reviewed in relation to acute mountain sickness (AMS). The cause of ataxia occurring at altitude is unknown but may be hypoxia affecting basal ganglia and hindbrain activity. Ataxia is an important sign of high altitude cerebral edema (HACE) but is less well-established as a clinical feature of AMS. Assessment of ataxia is part of the Environmental Systems and the Lake Louise questionnaires, together with a heel-to-toe measurement. More precise measures of ataxia include the Sharpened Romberg Test (SRT) and the use of unstable platforms. Isolated ataxia at altitude may not be related to AMS or HACE. Age affects ataxia and careful baseline measurements are essential in older subjects before results at high altitude can be interpreted. Testing for ataxia needs to be standardized with sufficient learning time. Ataxia should be distinguished from weakness or fatigue occurring at altitude. Specialized tests have not been shown to be clinically important. Our results above 5000 m showed that an abnormal SRT may be specific for AMS but with relatively poor sensitivity. Wobble board results have not correlated with AMS scores consistently. Other authors using an unstable platform in a chamber and static posturography during 3 days of exposure to 4559 m also found no relationship with AMS scores. Ataxia is a common and important clinical feature of HACE but is unhelpful in the assessment of mild or even moderate AMS in the absence of an altered mental state. The simple heel-to-toe test remains a useful part of the assessment of more severe AMS bordering on HACE.

Key words: ataxia, hypoxia, high altitude, mountain sickness

Introduction

Rapid ascent to high altitude may result in clinical syndromes ranging from high altitude headache to acute mountain sickness (AMS) and high altitude cerebral edema (HACE). Current views suggest that these syndromes form a spectrum, with transition from acute mountain sickness to high altitude cerebral edema occurring with the development of serious neurological disturbances such as ataxia, changes in behavior, altered consciousness, papilledema, and focal neurological signs.¹ Traveling over dangerous, mountainous terrain wearing heavy equipment has its own inherent risks, and compounding this with an ataxic gait makes the climber particularly vulnerable to trips or falls. Indeed, ataxia is a significant feature in the analysis of causes of death above 8000 m on Mount Everest,² although profound fatigue and weakness may contribute to the unsteadiness.

Altered consciousness and ataxia are the 2 most common clinical features in established HACE;³ therefore, detection of HACE has focused around the measurement of neurological dysfunction related to these symptoms. Ataxia is also currently monitored in the assessment of AMS. Existing assessments of impaired cerebral function include tests of memory, balance, ability to learn, ability to perform arithmetic or language tests, pupil constriction times,⁴ auditory-evoked potentials, and reaction times.¹ Although many tests of cerebral function may be impaired at high altitude, they do not necessarily assist in a diagnosis of AMS. This review assesses the evidence that measurement of ataxia is possible in the field and whether such measurements hold value in predicting AMS.

What is Ataxia?

Ataxia is incoordination in the absence of significant weakness and it results from disorders of the cerebellum, or cerebellar pathways, or from loss of proprioception

Corresponding author: Alexander David Wright, MD, the Medical School, University of Birmingham, Edgbaston, Birmingham B15 2TT, UK (e-mail a.wright@bmres.org.uk).

from sensory nerves or posterior column spinal cord lesions. Therefore, there are many types of ataxia. The ataxia of hypoxia may be a general neuronal problem and is not usually associated with signs of cerebellar dysfunction. This is unlike the ataxia occurring with normal pressure hydrocephalus (magnetic gait), cerebellar ataxia (broad-based gait), or ataxia specifically caused by dorsal column disease.

Possible Mechanisms

It is not certain how the effects of high altitude cause ataxia. Hypobaric hypoxia causes neuronal hypoxia, both directly and indirectly (through compensatory hyperventilation-induced hypocapnic vasoconstriction). Hypoxia causes a generalized slowing of neuronal conduction and synaptic transmission⁵ that may result in reduced coordination (the delay in pupillary reflexes may be an example of this).⁴ More specifically, the basal ganglia are commonly affected in clinical cases of hypoxic brain injury, either through carbon monoxide poisoning⁶ or post-cardiac arrest. In these patients, coordination is similarly a major problem.

So far, MRI studies have not helped to elucidate the mechanism of ataxia in HACE although important MRI findings⁷ in 7 of 9 subjects showed an increased T₂ signal in the white matter of the brain—especially in the splenium of the corpus callosum—without any gray matter edema. The splenium is not known to have a role in control of balance and proprioception and, classically, lesions here cause disconnection syndromes; specifically, lesions in the splenium cause alexia (inability to read) without agraphia. The splenium may be affected on MRI, because it has a very high density of white matter fibers that may be more prone to vasogenic edema, and because it sits between a mass of venous channels. Alternatively, although MRI suggests that the basal ganglia are commonly affected first in hypoxia, a number of clinical syndromes that involve the hindbrain (eg, Chiari malformations, cerebellar disorders) result in ataxia; therefore, one could also speculate that HACE in some way affects the hindbrain, or the posterior circulation, prior to a more global effect.

Assessment of Ataxia

QUESTIONNAIRES

Questions on ataxia are included in the Environmental Symptoms Questionnaire (“my sense of balance is off” and “I feel clumsy”)⁸ and in the Lake Louise scoring system.⁹ In the self-reporting section of the Lake Louise scoring system, a question on unsteadiness is graded as

follows: 0 = no incoordination; 1 = mild incoordination; 2 = moderate incoordination; and 3 = severe incoordination. Hultgren¹⁰ suggested modification of the Lake Louise scoring system with self-assessment scores with a rating of zero as no problem with balance; 1 = some problem with balance (I occasionally feel unsteady); 2 = definitely a problem keeping balance (very unsteady walking); 3 = severe balance problems (I can hardly walk without help or I would fall); 4 = very unsteady (I cannot stand without help). This extended scale and better-defined degrees of ataxia have not been adopted.

Measurement

Heel-to-toe Ataxia can be exaggerated by attempting to walk heel-to-toe, and in the clinical assessment portion of the Lake Louise system, the heel-to-toe is scored as: 0 = no ataxia; 1 = maneuvers to maintain balance; 2 = steps off line; 3 = falls down; 4 = cannot stand.

Romberg From the original observations of 19th century physicians, including Moritz Romberg, patients with altered proprioception were noted to lose postural control in the dark, so-called sensory ataxia.¹¹ Modifications of the Romberg test are widely used in medicine, and the Sharpened Romberg Test (SRT) is a useful measure of ataxia as it can be easily measured without any equipment, making it applicable to field observations at altitude. It is more sensitive than the standard test and is commonly used in the assessment of divers recovering from decompression sickness.¹² In the SRT, an individual is asked to stand with feet heel-to-toe in a straight line. The subject then places both arms across the chest with hands on the opposite shoulder. The subject is asked to close his or her eyes and maintain this position for 60 seconds. A failed negative test occurs if the eyes are opened, arms are put out to regain posture, or feet are moved to regain balance. A passed test is a completed 60 seconds in this position. We have reported on its use at altitudes up to 5260 m, where the SRT scores were more often abnormal than the traditional heel-to-toe test.¹³ However, conditions need to be carefully standardized and allowances made for a learning curve for the technique in older subjects. Subsequently, we have modified (B.G.J., oral communication, 2007) the SRT with a better definition of a failed test as being any movement of the hands or feet or opening of the eyes, and have defined an appropriate learning time as measurements made over 3 days before ascent to altitude. The majority of subjects (mean age 36 years, range 22–70) completed the test at sea level on all but one occasion so did not require a learning time. Five (24%) significantly older subjects (mean age 56 years, range 40–71) failed the test on 2 or

more occasions. Of this older group, 3 improved over 3 days but 2 continued to fail before ascending to altitude.

Unstable platforms Attempts to obtain a more quantitative measure of ataxia at altitude have been made by measurements on an unstable platform or by a target-tracking test. Using an unstable platform in 19 subjects (mean age 24.6 years), Cymerman et al¹⁴ showed that tests were more unstable at 2 to 3 hours and 23 to 24 hours of exposure to 4300 m compared with baseline, both with eyes open and with eyes closed. Although absolute instability values were higher on the eyes closed test, the Romberg ratio of eyes closed/eyes open was similar. Using a similar platform and measuring sway velocity with eyes open and closed in 22 subjects (mean age 42, range 21–54) during 3 days at 4559 m, Baumgartner et al¹⁵ showed worsening of tests on all 3 days with no change in the Romberg quotient (sway velocity for ratio of eyes closed/eyes open).

We devised a test for ataxia to be used in high altitude field studies. The simple equipment was a portable wobble board (a flat board, diameter 53 cm) with a half sphere ball (diameter 15 cm) glued to the center of the under-surface.¹⁶ A metal strip was attached to the inferior surface of the circumference. The board was placed on a flat metal plate checked for horizontal position with a bubble (carpenter's) level. The metal strip and plate were connected by a single electrical circuit to a battery-powered recording box that recorded the duration of contact of the metal strip with the metal plate to the nearest 0.1 second. Every time contact was made with the metal plate, a buzzer sounded so that the person supervising the test could record the number of contacts over 2 minutes. Using the wobble board, we noted¹⁶ a significant learning curve (as with SRT) with 3.77 (SD 3.4) as the mean number of contacts at 1345 m compared with 2.58 (SD 2.97) at 1660 m, but then the number of contacts remained stable at 2.25 (SD 2.89) up to 5005 m. Thus, no change in performance on the wobble board was seen at high altitude.

Ataxia and Age

Ataxia is common in the elderly and contributes to the problem of falls. The cause is often multi-factorial but loss of proprioception and postural hypotension are common factors. Our observations with both the SRT and the wobble board in older subjects over 50 years of age suggest that, although learning the technique occurs and is important, some tests are likely to be extremely difficult in older subjects. It is essential that subjects make baseline values of steadiness before ascent to high altitude in order to reduce differences from learning and to

take into account the loss of proprioception already acquired by the individual.

Ataxia and AMS

Few studies have shown any correlation of ataxia scores with AMS. In a 24-hour exposure to 4300 m in a chamber using an unstable platform system and a test involving tracking a circular moving object, no significant correlations were found between the Environmental Symptom Questionnaire scores and any of the postural instability tests.¹⁴ Using static posturography and cerebral AMS scores from the Environmental Symptom Questionnaire, another study of postural ataxia during 3 days at 4559 m showed no difference between subjects with or without AMS¹⁵ although ataxia tended to worsen over the 3 days in those subjects with AMS. Interestingly, short-term administration of oxygen at 3L/minute for 10 minutes improved AMS scores but failed to alter ataxia measurements, suggesting different mechanisms for the different clinical features. Therefore, oxygen therapy is unlikely to reverse ataxia rapidly. In another short-term chamber study at the same altitude, a calcium channel-blocking drug, flunarizine, was associated with less severe headache but no difference in postural stance¹⁷ and again confirming that postural ataxia was the same in those with or without AMS.

Similarly, the SRT has been used at altitude by our group¹³ and, in a study of 23 subjects at 4750 m, we noted 5 subjects with an abnormal test but none had AMS. At a higher altitude of 5260 m, 10 subjects (43%) had abnormal tests and 11 had AMS with Lake Louise scores of 3 or more. This gave a sensitivity of 60% and specificity of 89% for predicting AMS. On a separate occasion at 4392 m in the Chilean Altiplano, we modified the SRT, performing it twice a day with 2 exit points at 30 seconds and at 60 seconds. In 24 subjects the SRT did not relate to AMS, with sensitivity of 21.4% and specificity of 83.5% for diagnosis of AMS (B.G.J., oral communication, 2007). No subjects were suffering from HACE.

Subjects suffering acute mountain sickness scored significantly worse on the wobble board test, although scores did not correlate with a specific question on unsteadiness.¹⁶ A positive test, defined as equal to or more than 2.5 contacts over 2 minutes, gave a predictive value for acute mountain sickness of 66.7% at 4650 m and 100% at 5005 m. Cerebral regional oxygenation, measured using infrared spectroscopy in 9 subjects at 5005 m, correlated with the wobble board test ($r = 0.73$, $P < .05$) whereas pulse oximetry did not. In a second expedition to 5260 m, using a more sensitive wobble board with a smaller diameter ball between the board and

the plate, we were unable to confirm a relationship of scores with AMS ($r = 0.3$ ns) nor with the SRT (SRT normal, AMS score 7.2 ± 3.7 SD; SRT abnormal, AMS 8.8 ± 7.6 ns) nor with regional cerebral oxygenation ($r = -0.3$ ns).

Ataxia and HACE

Disturbance of consciousness, ataxia, and focal neurological signs are important components of HACE. Early HACE is often a difficult diagnosis to make, and a high index of suspicion is important. Subtle behavioral changes, slow to develop and somewhat out of keeping with an individual's normal affect, tend to be early features. Unexplained grumpiness or bad temper in a normally affable individual or, conversely, an abnormally euphoric or excitable affect in a person who is normally subdued can be the first indicators that a problem is developing. To further complicate the diagnosis, there may actually be good reasons for the change in affect such as adverse weather, snow conditions, or a successful completion of a difficult route. Sometimes the condition results in anger and frustration and, with increasing severity, apathy, lethargy, and disinterest may develop. As HACE worsens, the behavioral abnormalities become more marked until eventually it is clear the individual is irrational and unwell. Ataxia may be an early clinical feature. The other clinical signs of HACE such as focal or global neurological disturbance or speech difficulties often develop late. A clearly ataxic person is usually very unwell and unable to descend unaided. Consequently, if one waits until the affected individual formally fails a test for ataxia before starting treatment (descent, dexamethasone, oxygen, portable hyperbaric chamber, acetazolamide), the situation not only puts the individual but also the rescuers in danger. Nevertheless ataxia was important, as noted in a large study of 54 railway workers suffering from HACE¹⁸ where ataxia was noted in 73% and was reported to have occurred earlier than the most common sign of disturbance of consciousness (79%) in the majority of patients; in a minority, ataxia and altered consciousness developed simultaneously. These authors noted that progression from ataxia and mental changes to unconsciousness took about 12 hours and commonly occurred overnight. Ataxia has been noted as a significant feature in the analysis of causes of death above 8000 m on Mount Everest.²

Conclusions

Ataxia can be measured in the field by simple tests of coordination including a modified Romberg test. Special

equipment is required for studies but is unnecessary for clinical assessment and may be difficult to manage in the field. Preferably with 2 to 3 days allowed for learning, baseline measurements are recommended no matter what test is used, as individuals vary considerably, especially in the older subject over 50 years of age.

Significant ataxia is a manifestation of HACE and, along with signs of altered consciousness, should be assessed in making a diagnosis of HACE. "If the patient seems mildly drunk at altitude, he has cerebral edema."¹⁹ However, studies show that the development of ataxia is not important in the assessment of mild AMS. Some people will have measurable ataxia if specialized tests are used, but measurement of ataxia is not sensitive or specific enough to provide a reliable indicator of AMS. We conclude that routine assessment of ataxia at altitude, in the absence of moderate or severe AMS or neurological signs, is unlikely to assist in the early diagnosis of serious altitude illness.

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