Headshaking: - An alternative hypothesis
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Abstract:
Headshaking is recognised neurologic disorder of horses characterised by involuntary, hyper-reflexic, hyperaesthetic, up and down movements of the head during defined activities such as exercise or in defined circumstances not associated with exercise such as exposure to rain, cold, dust or wind. The condition is often reported to have an obvious abrupt onset of an apparent severe facial pain. Owners will frequently attribute the state to a “bee sting on the nose”. Affected horses are extremely distressed at the time and may be almost unmanageable. In contrast a few cases have a more insidious onset with a much milder head ‘tic’ being present for some weeks or even years before more obvious signs develop.

A high proportion of horses show the worst signs during exercise (whether ridden or free or lunged) but many have very variable symptoms. Apart from the characteristic up-down head movement, rotatory or side to side movements can be encountered. In this circumstance the condition is usually complicated and many such horses have detectable pathology in the neck or are lame (forelimb, back or hind limb lameness). Additional signs include striking at the face during exercise, rubbing the face on hard objects or commonly on the front legs. Some horses become distracted and do not respond to ridden or verbal commands – they may refuse to go forward, rear or circle when encouraged to move forward. Careful observation will often identify ‘nostril clamping’ i.e. the horse attempts to close its nostril(s) as if trying to prevent the movement of air therein.

The signs can be attributed to one side of the face or both and when both there may be significant differences in the severity of the signs between the two sides. The condition is very similar in nature to the syndromes of facial pain encountered in the human in multiple sclerosis, post herpetic neuralgia, Trigeminal Neuralgia and cluster headache syndromes. In all the conditions uncontrollable pain is the cardinal feature.

‘Headshaking’ is a well-recognised syndrome by both veterinarians and owners. It is now accepted that it is not a vice and owners now expect a more rational and less patronising investigation and assessment of prognosis.

Therapy is limited to the use of ‘steroids’, carbemazepine, hydroxyzine or management changes (including masks, contact lenses and other manipulations). Expectations for cures are high but realistically this is not achievable at the present time. Many affected horses are destroyed or retired from work. Others are sold on at times other than when the signs are evident. There is a perception that it is becoming more common and owners may blame pollution, radiation, chemical contacts or seasonal allergies.
Whatever is said about the disease in terms of cause, diagnosis or treatment, it is certainly true that the horses are noticeably distressed by it. Some horses are so badly affected that they will cause themselves serious injuries in an attempt to relieve themselves of the discomfort or pain. There are also major safety implications for riders / handlers.

**Headshaking is a serious neurological condition warranting a thorough clinical investigation and it has a significant implication for the welfare of the horse**

Currently investigations are frequently disappointing in that no useful diagnosis on possible instigatory pathology can be identified and no genuinely effective therapy has been found. This has lead to a progressive abdication of veterinary responsibility – just because it is a difficult disease it does not mean that it is a “behavioural avoidance” or “bad riding”. It has also meant that many horses are “pushed” from pillar to post – usually being sold on by what can only be described as unscrupulous owners, during periods of time when the horse shows no signs. Many cases are ascribed to “poor riding” skills. In fact the large majority of horses affected by the disease are equally affected with the best possible rider or when they are being lunged or given free exercise or lunged.

The truth is that these horses are suffering and we “thrash” them into submission at our peril. There are remarkable similarities to human Trigeminal Neuralgia / Neuropathy (TGN) – a condition characterised by excruciating persistent or intermittent facial pain. If we assume that the condition has the same aetiopathogenesis (and I admit that this is leap of faith!), we have to assume that given the chance an affected horse would prefer death. The fact that most horses presented for investigation are nice horses with nice owners simply reflects the caring attitude that they have and we should under no circumstances give irrational and cruel advice. Gratuitous advice from ill-informed lay people and even vets who actually have no idea about the disease does the horses no service. It is easy to suggest working the horse to submission, a new saddle, new stables and sundry herbal and other management measures; what is difficult is trying to understand what is happening and why!

The wide variety of suggested “causes” simply suggests that no single one is always involved and the common feature between almost all of them is the Trigeminal Nerve; this is the main sensory nerve to the face, mouth and nose. It is suggested that the condition reflects a form of hyperaesthetic hyper-reflexia, i.e. that the horse reacts inordinately to trivial stimuli (probably subliminal stimuli in normal animals) and that this response (alldynia) is mediated in the trigeminal nerve and triggers the sensation of severe sudden pain. The pain is “perceived” pain (neuropathic) rather than inflammatory pain and so analgesic drugs have no material effect at all. However, no significant pathology has so far been identified in the nerve itself, or the ganglion or the root entry zone of the nerve. There has been no serious investigation into possible central pathology and no meaningful neurophysiology has been performed that might confirm or deny this hypothesis. However, the hypothesis does explain most of the behavioural features of the disease and that these largely relate to the second division (Maxillary Branch) of the trigeminal nerve (TN). Most horses show facial and nasal discomfort and active avoidance of air turbulence on the face and in the nasal cavity. The absence of pathology suggests that the syndrome is one of (non-inflammatory) neuropathic pain.
deriving from abnormal sensory function within the peripheral or central pathways of the TN. The shaking signs largely reflect the facial nerve motor reflex pathway that derives from this stimulus.

Signs are commonly exacerbated by exercise or defined environmental conditions. Some cases are worse when exercised in bright light, others are worse in rainy, windy, warm or cold, dusty or humid conditions. At least in the initial presentation, many cases are remarkably ‘seasonal’ or circumstantial. This may suggest an allergic disorder akin to human ‘Hay Fever’. However, this is not supported by nasal biopsies and drug therapy; antihistamines and cortisone seldom resolve them (although some are helped). It seems more likely that the conditions or irritants that are responsible are simply the trigger factor required for those individual horses.

This has lead to the concept of a “loaded gun” pathogenesis i.e. the pathology exists but there are no outward sign unless and until the trigger factor (stimulus) is applied. This is largely similar to human TGN in which excruciating facial pain, either intermittent and stabbing type or persistent (mild to extreme), can be triggered by nasal, oral, dental, or facial stimuli. The trigger factor could even be a matter of intranasal air turbulence – thus as a horse breaths harder there is more nasal mucosal surface subjected to increased direct stimulus (from particles, cold, moisture, dust, pollen etc.). The relationship to exercise may also suggest a vascular or blood pressure implication – in the human circumstance vascular trigger factors are relatively common (the posterior cerebellar artery is in close juxtaposition with the root entry zone of the TN and separation by means of a Teflon / titanium plate dramatically resolves the condition at least temporarily. In the horse there is no close juxtaposition of the TN and the posterior cerebellar nerve (the artery is caudo-distant from the nerve) but the infraorbital artery runs along with the nerve within the infraorbital canal.

Some horses have defined pathology such as paranasal sinus inflammation, dental or oral or aural disease and others have a history of facial trauma. However, the large majority have no detectable disease (even when ‘scanned’ for everything using scintigraphy, radiography, endoscopy etc!). These cases are currently termed ‘idiopathic’ – not because there is no cause, but because up to now we have been unable to identify the true factors involved in either the pathology or the trigger factors or the relationship between the two.

Few suggestions as to the possible nature of the neurological problem have been proposed but hypomagnesaemia, Equine Herpes Virus latency, immune responses, inflammatory cytokine (TNF / Interleukins) and various demyelinating pathologies have been suggested. The relationship of TGN in humans to Multiple sclerosis (MS) is interesting but so far no such condition has been identified in horses. However, detection of the demyelinated plaque in human MS cases is extremely difficult and would be more so in horses. In any case there is no serological¹, histological or neurophysiological evidence of any demyelinating condition in headshakers. So far there is no know “common” factor in headshakers apart from the relatively common occurrence in dressage and pleasure horses and ponies as opposed to Thoroughbred racehorses. The proposed analogy to post herpetic neuralgia in humans has been tested in horses recently and no convincing evidence of EHV 1 at least in the brain stem was found in either headshaking or normal horses.

¹ Serological evidence of demyelination is suggested to reflect in elevation of P2-myelin sheath protein antibody in circulating blood.
The alternative hypothesis is that the condition is an autonomic disorder akin to the photic sneeze of humans. Given that the horse cannot sneeze to relieve this sensation it is suggested that nasal discomfort persists and becomes highly distressing. This hypothesis is possibly supported by the suggestion that bright light is a common trigger factor, and that anti-serotonin drugs such as cyproheptadine may resolve the condition. However, this is now largely discredited and the fact that sunshine or bright light is usually associated with many other potential triggers, simply adds to the confusion.

The prognosis for genuine headshaker horses is currently poor. Around 5-10% can be resolved and a few more can be improved significantly but this is somewhat better than the zero prognosis that existed up to recently. The major obstacle to progress seems to be the almost complete lack of any detectable neuropathology an a lack of understanding of the neurophysiology. This may be a matter of technology or perhaps that we are not looking in the correct place for it. There remains the possibility that the disease is a central ‘demyelinating’ or sclerotic vascular disease rather akin to multiple sclerosis or post herpetic neuralgia of humans in which the pathology is extremely subtle. Without high resolution magnetic resonance imaging it is almost impossible to confirm a diagnosis without biopsy or post mortem examination. Even in those circumstances usually the changes are far too subtle to detect with current technology / skill. A test that reveals some changes that are consistently present must be sought – perhaps a closer analysis of cerebrospinal fluid for oligoclonal antibodies (relating to myelin proteins and IgG) will reveal some diagnostically and pathologically significant changes.

The therapeutic options are very limited at present. Apart from the obvious avoidance strategy for the trigger factors (if these can indeed be identified!), which are often either impractical or at best inconvenient, there are no drugs that are consistently effective. The effects of steroids can be attributed to suppression of the trigger factor – not the condition. Some cases respond to some extent to hydroxyzine (a combined antihistamine and sodium channel blocker) and this is often used as a"confirmation that the disease is an allergy”. This of course is not the correct interpretation – it is far more likely that the trigger factor is an allergic response and that the benefit is due to both the resolving of the allergic “rhinitis / conjunctivitis) and some sodium channel blocking effects. Cyproheptadine (Periactin) is not effective unless combined with a strategy to protect the eyes (possibly from light but more likely form, impact and other sensory stimuli acting within the trigeminal nerve. There are no defined identified pathways in horses that link the trigeminal and optic nerves at any level within the central nervous system. Protection of the eyes / ears or nose with masks etc. probably simply acts as a counter-irritant or a means of avoiding the trigger factor contact. Given that the trigger factor MAY affect only a handful of neurones the responses are likely to be either effective or totally ineffective. The diagnostic value of carbemazapine (Tegretol) in very high doses gives credence to the hypothesis of a similar condition to TGN in humans – in the latter the response to CBZ is taken as definitive proof of the condition. The problem is that CBZ has an in-vivo half life of less than 2 hours (c.f. human at 23 hours) and so very high doses are required very frequently. This makes it almost impossible to sustain for long periods in MOST cases – a few do however, respond quite well even to low doses. The drug is also very expensive. A new approach with a drug called gabapentin has been tried in a significant number of cases at our hospital. This is a centrally acting analgesic – it is a treatment for neuropathic pain rather than inflammatory pain. A significant number of horses do benefit but many do not. It is probably best used as a diagnostic test
than a therapy. There are of course major dope-testing implications from the use of any of the drugs mentioned above and given that dressage horses are over-represented in the disease the use of drugs is not really helpful.

We have recently explored the use of trigeminal root nerve blocks and mechanisms of sclerosing or reducing the input down the maxillary branch of the TGN by partial compression of the nerve root. Compression of the infraorbital nerve is possible but not at a location that is far enough caudally to reduce the function of the posterior nasal (ethmoidal) nerve. It does seem from our work with maxillary root nerve blocks that this is a key nerve in the disease. The only consistently accessible site for accessing the nerve at present is within the infraorbital canal. This is too rostral really and so this treatment does not really tackle the best location along the pathway of the nerve. Currently we have abandoned the use of strong sclerosing agents and now favour a coiling system that causes a much slower effect. Whilst this prolongs the refractory period during which the condition may become worse, the effects are helping some horses. We have performed this procedure in around 50 cases and the results suggest that the technique is worth developing; by no means perfect. There are failures and partial successes as well as some cases that are significantly benefited by the procedure. The first signs of improvement are “not getting worse” and an improvement in the demeanour of the horse. We have had several cases where we have had to repeat the procedure but so far we have had no case that has come to necropsy to see what the coils are actually doing! We really need to perform some neurophysiological tests following the procedure but this is very problematic in horses and requires special conditions. It is important when considering this treatment to realise its limitations and complications – some horses are significantly harmed by the process – simply because we have to damage the nerve to do the procedure! We are trying to find better ways of getting at the specific branch of the nerve that we believe is most likely involved.

The anatomical differences in the horse make most of the human therapeutic options impossible with current technology. The sodium channel blocking drugs such as carbemazepine (and to a lesser extent hydroxyzine) appear to offer the best medical approach but in the end the best results are obtained by separating the trigger factors from the pathology by management or by other (surgical or medical) manipulations.

We are far behind in our understanding of this disease and its possible relationship to neuralgia states and to more generalised neurologic disease. However, we have to be aware of its welfare implications and so the current position is that the prognosis is poor at best. Research effort is a priority but the path we need to tread would be considerably facilitated if the horses could talk to us!

Further reading:


