The menagerie of neurology

Animal signs and the refinement of clinical acumen

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Summary

Neurology is a field known for “eponymophilia.” While eponym use has been a controversial issue in medicine, animal-related metaphoric descriptions continue to flourish in neurologic practice, particularly with the advent of neuroimaging. To provide practicing and trainee neurologists with a useful reference for all these colorful eponyms, we performed a literature review and summarized the various animal eponyms in the practice of neurology (and their etiologic implications) to date. We believe that the ability to recognize animal-like attributes in clinical neurology and neuroradiology may be attributed to a visual phenomenon known as pareidolia. We propose that animal eponyms are a useful method of recognizing clinical and radiologic patterns that aid in the diagnostic process and therefore are effective aides-mémoire and communicative tools that enliven and improve the practice of neurology.

While medicine and science have a tradition of eponyms (naming discoveries after scientists, physicians, patients, geographic locations, mythical beings, etc.), neurology is particularly famous (or infamous, depending on your views) for “eponymophilia.”¹

While most agree on repudiating eponyms that honor war criminals,¹ the debate for and against eponym use continues. Opponents prefer more descriptive names (e.g., extensor plantar response, rather than the Babinski sign). Supporters argue that eponyms are useful, linguistically economical representations of convoluted, unwieldy terminology (e.g., Sneddon syndrome, instead of idiopathic livedo reticularis with cerebrovascular accidents).² Some propose that eponyms add character, color, flavor, and dimension to the practice of neurology; others malign “eponymophilia” for engendering an overly complicated and cabalistic image of the specialty.^{2,3} Despite this controversy, zoologically inspired metaphorical descriptions continue to flourish in the luxuriant “eponymic forest of neurologic disorders,”³ and even more so with the advent of neuroimaging (table).

A word of caution: while animal-inspired eponyms are a pithy way of describing clinical and neuroradiologic signs, discretion should be exercised when using these terms in front of

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Table Summary of diagnostic implications of animal-inspired eponyms in neurology

Sign	Diagnoses
Ape hand	Median neuropathy
Arachnodactyly	Fibrillinopathies (e.g., Marfan syndrome, Ehlers-Danlos syndrome)
Barking tics	Some of the tics of Tourette syndrome
Bat-wing fourth ventricle	Joubert syndrome
Bat-wing sylvian fissure	Glutaric acidemia type I
Bat-wing tremor	Cerebellar ataxia, Wilson disease, multiple sclerosis, dystonia, essential tremor, parkinsonism
Bovine cough	Unilateral vocal cord paralysis
Butterfly lesion	High-grade glioma, lymphoma, demyelinating disease, Susac syndrome, neuronal ceroid lipofuscinosis
Butterfly medulla	Horizontal gaze palsy and progressive scoliosis
Cat-like cries	Cri-du-chat (5p minus) syndrome
Cock/equine gait	Footdrop
Dromedary gait	Axial dystonia (Huntington disease)
Duck gait	Proximal lower extremity weakness
Elephant sign	Hippocampal atrophy in dementing diseases
Eye-of-the-tiger sign	Neurodegeneration with brain iron accumulation, corticobasal degeneration, multiple system atrophy, HARP syndrome, progressive supranuclear palsy
Face of the giant panda	Wilson disease
Face of the panda cub	Wilson disease (double panda sign if coexisting with face of the giant panda)
Fish (or codfish) vertebra	Sickle cell disease, osteoporosis
Fishtail nystagmus	Paraneoplastic brainstem encephalitis, paraneoplastic syndromes, metabolic-toxic causes, idiopathic
Fly catcher tongue	Huntington disease
Gallop tongue	Following head/neck trauma
Hatchet face	Myotonic dystrophy
Hummingbird (or penguin) sign	Progressive supranuclear palsy
Inverted rabbit ear sign	Vitamin B ₁₂ deficiency
Leopard skin changes	Metachromatic leukodystrophy
Monkey sign	Central pontine myelinolysis
Panda sign	Sarcoidosis (on gallium scintigraphy)
Piglet sign	Reported in a case of central pontine myelinolysis
Rabbit syndrome	Adverse effect of dopamine antagonist therapy
Raccoon (or panda) eyes	Basilar skull fracture, amyloidosis, malignancies, vigorous sneezing/coughing, thrombocytopenia; continuous positive airway pressure, migraine, trigeminal autonomic cephalgia, rhinoplasty

Continued

Table Continued

Sign	Diagnoses
Scotty dog sign	Normal lumbar spine on x-ray; spondylosis causes a “broken neck”
Simian crease	Classically associated with trisomy 21; may be normal or associated with other syndromes
Snake eyes (or owl's eyes) sign	Compressive myelopathy, spinal cord infarctions, amyotrophic lateral sclerosis, paraneoplastic myelopathy
Swan neck	Myotonic dystrophy (not the swan neck deformity of rheumatoid arthritis)
Tadpole brainstem atrophy	Alexander disease
Tiger-stripping (cerebellum)	Lhermitte-Duclos disease
Tigroid fundus	Normal finding
Tigroid white matter changes	Metachromatic leukodystrophy
White rabbit syndrome	Orthostatic tremor

patients. Most understand that we intend no insult, but there are those who would perceive that we are making sport of their illness. As such, the honest, open discussion of such strategies that are utilized in order to augment clinical acumen cannot be overemphasized.

Animals at the bedside

Hands The *simian crease* (a single transverse palmar crease) may be a normal finding or may be found in various genetic syndromes (classically trisomy 21). The *ape hand* may be seen in median neuropathy, where thenar atrophy leads to recession of thumb-associated metacarpals to the plane of the other metacarpals, as a consequence of the unopposed actions of the extensor pollicis longus and adductor pollicis.⁴ Arachnodactyly (*spider fingers*) refers to abnormally long and slender fingers; it is typically found in fibrillinopathies like Marfan syndrome, Ehlers-Danlos syndrome, and homocystinuria.⁵

Gait Animal-inspired metaphors are especially popular when characterizing disorders of locomotion. Proximal lower extremity weakness results in the appearance of waddling, hence the term *duck gait*.⁶ In contradistinction, weakness of the anterior tibialis and peroneal muscle groups culminates in footdrop, compelling the patient to excessively flex the hip and lift the knee to keep the foot clear of the ground (termed the *cock* or *equine gait*).^{6,7}

Axial dystonia may cause increasing lumbar lordoscoliosis while walking, inducing bizarre truncal postures with the hip bulging backwards and sideways accompanied by compensatory turning of the shoulders and neck with the head held vertically; this progressive lordotic dysbasia is succinctly referred to as the *dromedary gait*.⁶

Head and neck The *tigroid fundus* is a normal fundoscopic finding in which lesser amounts of pigment in the retinal pigment epithelium allow visualization of streaks of underlying normal choroidal pigmentation. When a saccade terminates with a burst of flutter, this is called flutter dysmetria or *fishtail nystagmus*; causes include paraneoplastic disorders or metabolic-toxic states.⁷

Almost all clinicians are familiar with *raccoon eyes* (bilateral periorbital ecchymoses) signifying the presence of a basilar skull fracture.^{8–10} Not many, however, are aware that raccoon eyes have been observed in many other conditions, including amyloidosis,^{11,12} neuroblastoma,^{13–16} multiple myeloma,¹⁷ Kaposi sarcoma,¹⁸ following vigorous sneezing or coughing,^{19,20,e1} continuous positive airway pressure use,^{e2} dengue fever–induced thrombocytopenia,^{e3} trigeminal autonomic cephalgia,^{e4} migraine,^{e5} and following rhinoplasty.^{e6}

Supplemental Data

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Figure 1 Animal prints

The tigroid pattern (A) in metachromatic leukodystrophy is due to T2-hypointense radial stripes within the hyperintense subcortical white matter, resembling tiger stripes. T2-hypointense (B) or T1-hyperintense (C) spots in the subcortical white matter resemble a leopard's skin. Reproduced from Nandhagopal R, Krishnamoorthy SG. Neurological picture. Tigroid and leopard skin pattern of dysmyelination in metachromatic leucodystrophy. *J Neurol Neurosurg Psychiatry* 2006;77:344, with permission from BMJ Publishing Group Ltd.

The unmistakable physiognomy of myotonic dystrophy imparts an uncanny resemblance to the hatchetfish (*Argyropelecus* species), hence the term *hatchet face*. This conspicuous morphologic manifestation may be combined with sternomastoid weakness and atrophy associated with exaggerated forward neck curvature, reminiscent of a *swan neck*.⁷

Tongue The term *fly catcher tongue* aptly describes the recurrent back-and-forth darting movements of the tongue in Huntington disease.^{e7} Substantially more inconspicuous is the *galloping tongue* phenomenon, most commonly associated with head and neck trauma and characterized by longer latency, rhythmically stereotyped, paroxysmally focal and centrifugal tongue contractions, the pathophysiologic pattern of which has been measured at a frequency of 3 Hz.^{e8}

Vocalizations In some disorders, nonvolitional patient-transmitted sounds resemble animal vocalizations. For instance, cri-du-chat syndrome received its name from the characteristic high-pitch cry (strikingly similar to a mewling cat) of the afflicted children. Some of the vocal tics of Tourette syndrome are colloquially referred to as *barking tics*.⁶

Unilateral vagal nerve lesions can preclude the normal matched and synchronous opposition of the vocal cords, thereby causing a *bovine cough*.⁸ Notwithstanding the unusual nature of the bovine cough, pathoetiologic caution is advised given that the absence of collateral signs of vagal dysfunction may implicate psychogenic underpinnings.^{e9}

Movement disorders The widely recognized *rabbit syndrome* describes a perioral tremor (involuntary, rhythmic, and often vertical mouth movements without tongue involvement) arising from the use of dopamine antagonist therapy (e.g., antipsychotic and certain antiemetic agents).^{e10} It may perhaps be better stated as the *rabbit sign* since a syndrome refers to a collection of signs and symptoms that share a common pathophysiology. This should not be confused with the *white rabbit syndrome* of orthostatic tremor, a peculiar condition in which patients prefer either sitting down or moving about,⁸ resulting in almost constant motion reminiscent of the white rabbit in *Alice's Adventures in Wonderland*.

Another animal-inspired tremor neurologists are familiar with is the *bat-wing tremor*: a severely disabling, low-frequency, proximal-dominant upper extremity postural/kinetic tremor that results in an abduction-adduction movement of the arm. It may be seen in cerebellar ataxia, Wilson disease, multiple sclerosis, dystonia, essential tremor, and parkinsonism.^{e11}

Figure 2 Tadpole

In Alexander disease, a T2-hypointense periventricular rim (A) may be discernible, but more conspicuously, marked cervicomedullary atrophy with an intact pons contributes to the appearance of the tadpole sign (B) on sagittal MRI sections. Reproduced from Schmidt H, Kretzschmar B, Lingor P, et al. Acute onset of adult Alexander disease. *J Neurol Sci* 2013;331:152-154, with permission from Elsevier.

The beasts of neuroradiology and pathology

Basal ganglia The *eye-of-the-tiger* sign on brain MRI (pallidal T2 hypointensity with a central zone of hyperintensity) is characteristic of, but not pathognomonic for, pantothenate kinase-associated neurodegeneration, formerly called Hallervorden-Spatz syndrome (an eponym shunned for its Nazi connections). This popular sign has also been reported in multiple system atrophy, corticobasal degeneration, HARP syndrome (hypoprebetalipoproteinemia, acanthosytosis, retinitis pigmentosa, and pallidal degeneration), and progressive supranuclear palsy (PSP).^{e12-e16}

White matter The distinct colorations of animal fleeces have also found their way into neurologic descriptions. Neuropathologists (and those preparing for the neurology boards) would be familiar with *zebra bodies*, broad transversely stacked myelinoid membranes found in the nervous tissue of patients with Fabry disease, metachromatic leukodystrophy, Niemann-Pick disease, GM2 gangliosidosis, and mucopolysaccharidoses.^{e17} Some leukodystrophies (metachromatic leukodystrophy, Pelizaeus-Merzbacher disease, and globoid cell leukodystrophy) result in white matter T2 hyperintensity on MRI with occasional hypointense abnormalities within the demyelinated white matter; linear, radiating hypointensities are described as *tigroid*, while punctate hypointensities take on the appearance of *leopard skin* (figure 1).^{e18-e20}

A lesion that involves both frontal lobes and crosses the corpus callosum is commonly referred to as a *butterfly lesion*, classically suggesting a high-grade glioma or lymphoma, but has also been observed in demyelinating disease, Susac syndrome, and neuronal ceroid lipofuscinosis.^{e21-e22}

Ventricles and hippocampi Bat wings have enjoyed a prominent role in radiologic eponyms. Medical students would be familiar with the bat-wing appearance of pulmonary edema on chest radiographs. In Joubert syndrome, dilation of the fourth ventricle has been described as bat-wing shaped.^{e23} Additionally, bat-wing dilation of the sylvian fissures on CT or MRI is characteristic of glutaric acidemia type I.^{e24-e25}

Deep to the sylvian fissure lie the hippocampi, which bear an uncanny resemblance to their Greek namesake, seahorses. With progressive hippocampal atrophy in disorders like Alzheimer

Figure 3 Butterfly

MRI images from 2 patients with familial horizontal gaze palsy and scoliosis (A, B and C, D) compared with a healthy control (E, F), at 2 representative anatomical levels. In both patients, the fourth ventricle is enlarged (A and C) and lacks the normal prominence due to the underlying abducens nuclei (short arrows in E). The butterfly-shaped medulla (B and D) is the result of medullary dysplasia with a prominent deep ventral sulcus (long arrows) and relative prominence of the inferior olives in comparison to the pyramids. Reproduced from Pieh C, Lengvel D, Neff A, et al. Brainstem hypoplasia in familial horizontal gaze palsy and scoliosis. *Neurology* 2002;59:462-463, with permission from Lippincott Williams & Wilkins/Wolters Kluwers Health.

disease and frontotemporal dementia, these “seahorses” resemble elephants (hence, *elephant sign*).^{e26}

Brainstem and cerebellum Tegmental mesencephalic atrophy with relative preservation of pontine volume in PSP gives the midbrain the silhouette of a hummingbird on midsagittal MRI (the *hummingbird sign*).^{e27–e28} More ornithologically inclined neurologists have also called it the *penguin sign*.^{e29} Adult-onset Alexander disease (resulting from glial fibrillary acidic protein mutation and pathologically characterized by Rosenthal fibers) demonstrates medullary and cord atrophy with a relatively well-preserved, plump pons; on sagittal MRI, this has been described as *tadpole brainstem atrophy*.^{e30} (figure 2).

Horizontal gaze palsy and progressive scoliosis is a rare genetic disease due to *ROBO3* mutation; the dysplastic medulla with its deep ventral sulcus and relative prominence of the inferior olives in comparison to the pyramids has been likened to a butterfly.^{e31–e32} (figure 3) Changes in brainstem structures resulting from osmotic demyelination have given rise to various zoologically related radiologic signs including the piglet^{e33} and monkey^{e34} signs. The unique cerebellar foliation pattern in Lhermitte-Duclos disease results in distinct parallel linear striations called *tiger-stripping* on MRI and CT.^{e35}

Vertebrae and spinal cord The *Scotty dog sign* refers to the normal appearance of the lumbar spine on oblique radiographs; spondylosis results in a Scotty dog with a “collar” or “broken neck.”^{e36–e37} The *fish vertebra* (or *codfish vertebra*) sign is characterized by biconcave vertebral bodies occurring as a result of squared-off depression of the vertebral endplates and compression by adjacent intervertebral discs; it is seen in osteoporosis and sickle cell disease^{e38} (figure 4).

The bundle of lumbosacral spinal roots originating from the conus medullaris is called the *cauda equina*, after its resemblance to a horse’s tail. MRI T2 hyperintensity of the dorsal columns of the spinal cord in cyanocobalamin-deficiency myelopathy has been called the *inverted rabbit ear sign*.^{e39} *Snake eyes* or *owl’s eyes sign* refers to the appearance of 2 small,

Figure 4 Fish vertebra

The *fish vertebra* (or *codfish vertebra*) sign is the result of exaggeration of the normal concavity of the superior and inferior surfaces of the vertebral bodies. It may be seen in sickle cell disease (where microinfarctions of the central vertebral growth plate result in impaired vertebral growth) or in osteoporosis. Reproduced from Ntagiopoulos PG, Moutzouris DA, Manetas S, et al. The “fish-vertebra” sign. *Emerg Med J* 2007;24:674–675, with permission from BMJ Publishing Group Ltd.

symmetric, T2-hyperintense spots in the spinal cord parenchyma; it is the result of bilateral tract-specific involvement (the affected tracts depend on the underlying pathoetiologic process). Such changes have been reported in compressive myelopathy,^{e40} spinal cord infarctions,^{e41} amyotrophic lateral sclerosis,^{e42} and paraneoplastic myelottractopathy.^{e43}

Those adorable pandas The cute pandas have attracted immense public interest and adoration, in addition to their status as an eponym and acronym. *Panda eyes* are sometimes used interchangeably with raccoon eyes.⁸ The *face of the giant panda sign* is a result of midbrain tegmental T2 hyperintensity sparing the red nuclei, in conjunction with normal or low signal intensity of the substantia nigra pars reticulata and superior colliculi hypointensity; it is classically associated with Wilson disease^{e44} but has been reported in a case of multiple sclerosis.^{e45} The *double panda sign* is the result of the appearance of the face of the giant panda sign in the midbrain, accompanied by the *face of the panda cub* or *miniature panda sign* in the dorsal pons.^{e46–e47}

The *miniature panda face* is a result of relative hypointensity of the central tegmental tracts and the medial longitudinal fasciculi, in contrast to the hyperintensity of the most caudal extent of the cerebral aqueduct as it opens into the fourth ventricle. In this circumstance, the apparent panda pattern is bounded inferiorly by the superior medullary velum and laterally by the superior cerebellar peduncles.^{e46–e47}

Focal accumulation of gallium-67 citrate in the nasopharynx, parotid glands, and lacrimal glands (the *panda sign*) when performing gallium scintigraphy is highly characteristic of sarcoidosis.^{e48}

In its application as an acronym, PANDAS refers to the controversial constellation of syndromic features (an acquired movement disorder, typically tics, in association with behavioral manifestations, classically obsessive-compulsive disorder) derivative of streptococcal infection; Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections.

Animal-inspired eponyms and neurologic practice

Pareidolia, an apophenic phenomenon characterized by misperceptions from random stimuli (e.g., observing faces in the clouds or religious iconography in food), most likely accounts for the ability to recognize a resemblance between clinical phenomena and animal-like attributes.^{e49} Despite the negative associations of such illusions (e.g., seeing Jesus Christ on toast), pareidolia may actually be a very useful asset to neurologists.

Pattern recognition and effective communication are indispensable skills to the diagnostic capabilities of any clinician. For neurologists, these skills are particularly important since the intricacies of the nervous system often give rise to recognizable and eloquent syndromes.

For example, the stooped, shuffling, elderly patient with a pill-rolling tremor immediately brings the eponym Parkinson disease to mind. Instead of being lost in complex clinical phenomenology and radiologic descriptions, recognizing a peculiar pattern of abnormalities would direct a neurologist's diagnostic deliberations to a set of differential diagnoses. Further, animal eponyms can be used to succinctly summarize and communicate a complex abnormality without resorting to a litany of awkward, tongue-twisting designations.

CONCLUSION

Identifying and concisely communicating clinical or radiologic patterns of abnormalities is a cardinal component of the practice of neurology. Animal-inspired eponyms should not be dismissed as cabalistic or pejorative but should instead be considered as colorful, useful aides-mémoire that enliven and improve the diagnostic process, facilitate communication, and reinforce understanding of the entity itself, with patently obvious dividends for those we serve.

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