

# Neurological ZOO

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## ABSTRACT:

Eponyms are numerous in neurology. It is estimated that there are about 450 „human” eponyms commonly used by neurologists, neuroradiologists and neuroscientists. In this short review we describe 36 „animal” eponyms and 42 „animal” clinical signs often used in neurology. If we take into account the continuous creativity of neurologists and neuroradiologists in finding animal eponyms, it is to be expected that this list will continue in the future.

**KEYWORDS:** eponyms, animals, neurology, signs and symptoms

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## SAŽETAK:

### NEUROLOŠKI ZOO

Eponimi su brojni u neurologiji. Procjenjuje se da neurolozi, neuroradiolozi i neuroznanstvenici koriste oko 450 „humanih” eponima. U ovom kratkom preglednom radu opisujemo 36 „životinjskih” eponima i 42 „životinjska” klinička znaka koja se često koriste u neurologiji. Poznavajući maštovitost neurologa i neuroradiologa za očekivati je nove eponime u budućnosti tako da i ovaj popis životinjskih eponima nije konačan.

**KLJUČNE RIJEČI:** eponimi, životinje, neurologija, znakovi i simptomi

Eponyms (Greek eponymon = the name of the god or hero after whom something is named) are numerous in medicine and refer to the names of various significant persons from the history of medicine who have described certain clinical entities, signs, symptoms. Some of the eponyms are well known to the general population (Alzheimer's, Parkinson's), while others are known only to experts in certain fields of medicine. Neurology is known for its numerous eponyms. It is estimated that there are about 450 "human" neurological eponyms. The reason for such a large number of eponyms in neurology probably lies in the fact that a

detailed neurological examination allows a topical diagnosis, that could, by recording various clinical symptoms and signs, accurately localize the lesion in the nervous system. This is especially characteristic of neurologists of the 19th and first half of the 20th century, when neurological propaedeutics was brought to perfection. Unfortunately, with the development of neuroimaging methods, the technique of neurological examination is becoming increasingly insufficient in everyday clinical practice. In this paper, inspired by the article by Beh et al. on the same topic<sup>1</sup>, we present an overview of "animal" eponyms in neurology.

Table 1. Summary of animal eponyms in neurology

ANIMAL	SIGN/SYMPTOM	DISEASE
LION-MOUSE	LEONINE-MOUSE SY.	CLUSTER HEADACHE
LION	FACIES LEONTINA	LEPROSY
BUTTERFLY	BUTTERFLY/SCHMETTERLING TUMOR	GBM
MOTH	„MOTH-EATEN APPERANCE IN BONE“	MULTIPLE MYELOMA
STORK	STORK LEGS	CMT POLYNEUROPATHY
RACCOON	RACCOON EYES	BASILAR SKULL FRACTURE
PANDA	PANDA EYES	BASILAR SKULL FRACTURE
	FACE OF THE GIANT PANDA SIGN	WILSON'S DISEASE
ANT	FORMICATION	POLYNEUROPATHY
MEDUSA	CAPUT MEDUSAE SIGN	DVA
COCK	COCK GAIT	PERONEAL NERVE PALSY
ZEBRA	ZEBRA SIGN	CEREBELLAR HEMORRHAGE
SEAHORSE	HIPPOCAMPUS	LIMBIC SYSTEM
CHAMELEON	CHAMELEON/FLY CATCHER TONGUE	HUNTINGTON'S DISEASE
MONKEY	APE-LIKE HAND	CARPAL TUNNEL SYNDROME
MONKEY	MONKEY SIGN	CPM
OWL	OWL'S EYES SIGN	CPM
PIG	PIGLET SIGN	CPM
WORM	WORM LIKE MOVEMENT	ATHETOSIS
TIGER	EYES-OF-THE-TIGER	HALLERVORDEN-SPATZ SY.
		PSP
		CBD

Table 1. (cont.)

TIGER	TIGERSTRIPE APPEARANCE	LHERMITTE-DUCLOS DISEASE
LEOPARD	LEOPARD SKIN SIGN	MLD
CROCODILE	CROCODILE TEARS	BOGORAD SY.
PENGUIN	PENGUIN SIGN	PSP
HUMMINGBIRD	HUMMINGBIRD SIGN	PSP
HORSE	CAUDA EQUINA	CAUDA EQUINA SY.
HORSE	HORSESHOE SIGN	MS
SNAKE	SERPIGINOUS VESSELS	AVM
BULL	BULL'S EYE	TOXOPLASMOSIS
		LYME DISEASE
BUFFALO	BUFFALO HUMP	CORTICOSTEROID SIDE
		EFFECTS
COW	MAD COW DISEASE	MAD COW DISEASE
OX	STEER HORN SIGN	ACC
RABBIT	RABBIT TREMOR	ANTIPSYCHOTICS SIDE
		EFFECTS
ELEPHANT	ELEPHANT MAN	NF1
ELEPHANT	DURAL (ELEPHANT) TAIL SIGN	MENINGEOMA
DUCK	DUCK GAIT	PROXIMAL MYOPATHY
FLY	MUSCAE VOLANTES	DVS
BIRD OF PREY	CLAW LIKE HAND	ULNAR NERVE PALSY
BAT	BAT WING SIGN	SY. JOUBERT
BIRD	BIRD BEAK SIGN	SY. ARNOLD-CHIARI TYPE 2
FISH	ICHTYOSIS („FISH SKIN“)	REFSUM DISEASE

Abbreviations: GBM – glioblastoma multiforme; CMT – Charcot-Marie-Tooth; DVA – developmental venous anomaly; CPM – central pontine myelinolysis; PSP – progressive supranuclear palsy; CBD – corticobasal degeneration; MLD – metachromatic leukodystrophy; MS – multiple sclerosis; AVM – arteriovenous malformation; ACC – agenesis of corpus callosum; NF1 – neurofibromatosis type 1; DVS – degenerative vitreous syndrome

### LION AND MOUSE

Cluster headache (Horton's headache) is characterized by unilateral pain, primarily in the orbital area, but also in the supraorbital and temporal areas, that lasts 150-180 minutes if left untreated and occurs in attacks ranging in frequency from once every other day to eight times daily. The pain is accompanied by ipsilateral injected conjunctiva, epiphora, nasal congestion, rhinorrhea, hyperhidrosis and redness of the forehead and face, feeling of fullness in the ear, miosis, ptosis, eyelid edema and agitation<sup>2</sup>. This type of headache is more frequently present in men. If there is a kind of constitutional predisposition for cluster headache, it probably includes "macho" appearance (athletic build, black complexion, thickened facial skin with pronounced furrows) while mentally such people are structured as ambitious and diligent. This physical-mental structure is also called "leonine-mouse" (lion-mouse) syndrome, which indicates that these are sensitive people with a rougher appearance, or mice in the body of a lion<sup>3</sup>.

### LION

Leprosy (Hansen's disease) is a chronic infection caused by the acid-resistant bacillus *Mycobacterium leprae*, which shows a unique tropism for peripheral nerves, skin and mucous membranes. It causes countless symptoms, but from the neurological point of view, the most interesting is painless peripheral neuropathy. It is not just a biblical plague because the disease is still present in endemic hotspots around the world, from Asia, Africa, Mexico, South and Central America. Changes on the skin of the face such as macules, papules, nodules or plaques, usually symmetrical distributions, give these patients a characteristic appearance of the face like a lion ("facies leontina")<sup>4</sup>.

### COCK

"Cock gait" is characteristic and easily recognizable. People with weakness of dorsiflexion of the feet when walking first touch the toes, then the middle part of the sole and the heel, producing a characteristic sound of foot tapping from the ground. There are many possible causes of this disorder, from polyneuropathy of various genesis to trauma or external compression of the peroneal nerve in the segment of the knee, i.e. the head of the fibula<sup>5</sup>.

### RACCOON

Bilateral, periorbital, posttraumatic hematomas are called "raccoon eyes" and this finding most often indicates a skull base fracture. This sign can occur 2-3 days after the trauma and can be associated with a positive Battle sign (mastoid ecchymosis)<sup>6</sup>.

### PANDA

Synonymous with "raccoon eyes" when breaking the base of the skull are "panda eyes"<sup>6</sup>. The "face of the giant panda sign" in neuroimaging refers to the appearance of the midbrain, when the red nucleus and substantia

nigra are surrounded by a high T2 signal in the tegmentum. This neuroradiological sign is typical of a disorder of copper metabolism (Wilson's disease)<sup>7</sup>.

### ZEBRA

The "zebra sign" has been termed to describe the finding of layering of blood in amongst the folia of the cerebellum<sup>8</sup>.

### SEAHORSE (LAT. HIPPOCAMPUS)

The hippocampus is part of the limbic system, and plays important roles in the consolidation of information from short-term memory to long-term memory, and in spatial memory that enables navigation. In Alzheimer's disease, the hippocampus is one of the first regions of the brain to suffer damage<sup>9</sup>. Damage to the hippocampus can also result from hypoxia, encephalitis, or medial temporal lobe epilepsy.

### CHAMELEON

Huntington's chorea is autosomally dominantly inherited disease and has a very characteristic clinical presentation with chorea and progressive cognitive decline. Diagnosis is established based on genetic testing, and therapy is only symptomatic. This disease is characterized by atrophy of the nucleus caudatus. Huntington's disease is the result of a genetic mutation (the IT15 gene, which is located at the top of the short arm of chromosome 4 and contains a number of CAG trinucleotides within its coding sequence). This gene encodes the protein huntingtin. The normal range of CAG trinucleotides in this sequence is 6-37, while in patients with Huntington's chorea it is increased. The number of CAG copies can increase from generation to generation, leading to an increasingly severe phenotype of the disease in the same family – this is a phenomenon of anticipation. Even in the early stages of Huntington's chorea, the Chameleon tongue sign is common and often goes unnoticed. The patient is not able to maintain the protrusion of the tongue for a while, since this action is constantly disturbed by involuntary movements of the head, torso and extremities<sup>10</sup>.

### MONKEY

Carpal tunnel syndrome is the most common focal compressive mononeuropathy. Compression of the median nerve in the carpal tunnel leads to hypotrophy/atrophy of the tenar muscles (*m.opponens pollicis* and *m.abductor pollicis brevis*) and consequent weakness of opposition and volar abduction of the thumb. The result of this disorder is the so-called "Ape-like hand," in which the tenar and hypotenar bones are practically in the same plane as in the chimp hand<sup>11</sup>. This sign should not be confused with the simian palmar crease on the hand that can be seen in patients with Down syndrome.

Central pontine myelinolysis is a possible complication of sudden correction of blood hyponatremia. Changes in the pons are visible on MR. The T1-axial-weighted MR image shows the hy-

pointense signal changes involving the transverse pontine fibers, with sparing of the corticospinal tracts, giving the appearance of the face of a monkey (“monkey sign”)<sup>12</sup>.

#### OWL

In central pontine myelinolysis, neuroradiologists sometimes describe an “owl’s eyes sign” where T2-axial-weighted MR images show the hyperintense signal changes involving the transverse pontine fibers, with sparing of the tracts<sup>12</sup>.

#### PIG

The “piglet sign” is seen in osmotic demyelination syndrome. It refers to the appearance of the upper pons in axial T2 and FLAIR images. The areas of coalescent T2 signal increase are reminiscent of a pig’s snout, with other features on axial MR images resembling the rest of the face of a piglet with the temporal lobes representing ears, the carotid arteries represent the eyes and the fourth ventricle as the mouth<sup>13</sup>.

#### WORM

Athetosis is characterized by “worm-shaped”, winding movements, predominantly present in distal parts of the extremities. They are the opposite of hemiballism, explosive movements of the proximal parts of the extremities. Athetosis and chorea often occur together, so we are talking about choreoathetotic movements. Chorea and athetosis are manifestations of dopaminergic hyperactivity in the basal ganglia<sup>14</sup>.

#### ANT

The term formication (“tingling”) is often used to describe the clinical problems of patients with polyneuropathy. It is a type of paresthesia, i.e. tactile hallucinations (the patient often has unpleasant sensations without real external stimuli)<sup>15</sup>.

#### TIGER

“Eyes-of-the-tiger” is a neuroradiological sign caused by reduced T2 signal on brain MR in the area of the globus pallidus (due to iron accumulation) and a longitudinal striped zone of elevated signal due to gliosis and spongiosis, which can be seen in patients with Hallervorden-Spatz syndrome, progressive supranuclear palsy and corticobasal degeneration<sup>16</sup>.

“Tigerstripe appearance” is a characteristic alternating hypo and hyperintense bands on MRI in Lhermitte-Duclos disease. This is a rare cerebellar tumor appears like the coat of a tiger<sup>17</sup>.

#### LEOPARD

The “Leopard skin sign” results from dark-spots or stripes (sparing perivascular white matter) within bright demyelinated periventricular white matter on T2W images. It is characteristically seen in metachromatic leukodystrophy<sup>18</sup>.

#### CROCODILE

Bogorad syndrome (according to F.A. Bogorad, a Russian neurologist) is also known as the syndrome of “crocodile tears”, and characterized by residual facial paralysis with profuse lacrimation during eating. This phenomenon is also known as paroxysmal lacrimation or the gusto-lacrimal reflex. It is caused by a misdirection of the regenerating autonomic fibers to the lacrimal gland instead of to the salivary gland<sup>19</sup>.

#### STORK

Charcot-Marie-Tooth neuropathy is one of the most common inherited neurological diseases and includes a clinically and genetically heterogeneous group of hereditary neuropathies. The clinical picture is manifested by involvement of the distal musculature with the progressive development of hypotrophy, areflexia and deformity of the foot. The most common symptoms are drooping foot, pes cavus, hammer deformity of the big toes, and hypotrophy and atrophy of the muscles of the lower leg and foot, with preserved muscles of the upper leg, which all lead to the characteristic appearance of “stork legs” or “champagne bottle legs”<sup>20</sup>.

#### PENGUIN AND HUMMINGBIRD

Progressive supranuclear palsy (Steele – Richardson – Olszewski syndrome) is characterized by paresis of vertical bulb movements, propensity to retropulsion, and frequent falls. Bulbar disorders and emotional incontinence are common. Characteristic is the MR finding showing a positive “hummingbird sign”, also known as the “penguin sign”. The atrophy of the midbrain results in a profile of the brainstem in which the preserved pons forms the body of the bird, and the atrophic midbrain the head, with beak extending anteriorly towards the optic chiasm<sup>21</sup>.

#### JELLYFISH (MEDUSA)

The “caput medusae sign” refers to the developmental venous anomalies of the brain, where a number of veins drain centrally towards a single drain vein. The appearance is reminiscent of Medusa, a gorgon of Greek mythology, who was encountered and defeated by Perseus. The sign is seen on both CT and MRI when contrast is administered. Angiographically the caput medusae appearance is seen only in the venous phase<sup>22</sup>.

#### HORSE

Horsetail syndrome is a condition that occurs when the bundle of nerves below the end of the spinal cord known as the cauda equina is damaged. The clinical picture includes: severe back pain, saddle anesthesia, bladder and bowel dysfunction, sciatica-type pain on one side or both sides, weakness of the muscles of the lower legs, Achilles reflex absent on both sides, sexual dys-

function, absent anal reflex and gait disturbance. The management of true cauda equina syndrome frequently involves urgent surgical decompression<sup>23</sup>.

The “horseshoe sign” or “open ring sign” is a relatively specific sign for demyelination, most commonly tumefactive multiple sclerosis, and is helpful in distinguishing between the causes of ring-enhancing lesions. The enhancing component is thought to represent the advancing front of demyelination and thus favors the white matter side of the lesion. The open part of the ring will therefore usually point towards the gray matter<sup>24</sup>.

### SNAKE

Brain arteriovenous malformations are a type of intracranial high-flow vascular malformation that includes enlarged feeding arteries, a nidus of vessels closely associated with the brain parenchyma through which arteriovenous shunting occurs, and draining veins<sup>25</sup>. The “serpiginous” (snake-like) course of blood vessels is typical of these malformations.

### BULL

Erythema migrans is a rash characteristic of Lyme disease (borreliosis). The rash occurs at the site of a tick bite, usually after 7-10 days and spontaneously fades during 3-4 weeks. It is often round or oval in appearance, with a central darker and peripheral lighter zone and resembles a shooting target or “bull’s eye”<sup>26</sup>. Neuroborreliosis should be ruled out in all patients with demyelinating changes in the CNS.

Cerebral toxoplasmosis, is an opportunistic infection caused by the parasite *Toxoplasma gondii*. It typically affects patients with HIV / AIDS and is the most common cause of cerebral abscess in these patients. Brain MRI (T2 sequence) shows a concentric target sign or “bull’s eye” sign<sup>27</sup>.

### BUFFALO

Neurologists are among the most common prescribers of corticosteroids and therefore well aware of the side effects of their long-term use. Corticosteroid-induced lipodystrophy (CIL) is a condition of abnormal fat deposition (distribution) caused by corticosteroid medications. Fat accumulates in the facial area (“moon face”), dorsocervical region (“buffalo hump”), and abdominal area with a characteristic centripetal type of fat tissue distribution<sup>28</sup>.

### COW

Creutzfeldt-Jakob disease is a rare, degenerative disease of the human brain caused by prions. If it occurs in cattle, it is called bovine spongiform encephalopathy or “mad cow disease”. The dominant clinical sign is progressive dementia, ataxia, tremor, epilepsy, etc.<sup>29</sup>.

### STEER

Agenesis of the corpus callosum (ACC) is a rare birth defect in which there is a complete or partial absence of the corpus callosum. The MRI showed a “steer horn” sign made by the frontal horns of the lateral ventricles in the coronal plane<sup>30</sup>.

### BUTTERFLY

Glioblastoma multiforme is the most malignant brain tumor. It is characterized by rapid, infiltrative growth, with zones of necrosis and bleeding and pronounced perifocal vasogenic edema on CT / MR. It often infiltrates the corpus callosum and the contralateral hemisphere of the brain, giving it its characteristic butterfly shape. Hence terms such as “butterfly-shaped tumor”, “butterfly” or “schmetterling tumor”<sup>31</sup>.

### RABBIT

It is a rare extrapyramidal side effect of antipsychotics. It is manifested by perioral tremor of 5 Hz frequency, without tongue affection - “rabbit tremor”. It is caused by long-term antipsychotic therapy, primarily haloperidol, fluphenazine, pimozide, and less frequently thioridazine, clozapine, olanzapine, and risperidone<sup>32</sup>.

### ELEPHANT

Neurofibromatosis is a well-known phacomatosis. The characteristics of type 1 neurofibromatosis are as follows: 6 or more “café au lait spots” on the skin, at least 2 neurofibromas on the skin, at least two on the iris and scoliosis. David Lynch directed in 1980. the movie „Elephant Man” in which he describes the life of Joseph Merrick who suffered from neurofibromatosis type 1, and hence the incorrect name “elephant man” for these patients<sup>33</sup>. The “Dural (elephant) tail sign” represents thickening and enhancement of the dura mater in continuity with a mass, which on MR images, gives the appearance of a tail arising from the mass. Three criteria need to be met for a „positive” dural tail sign: the tail should be seen on two successive images through the tumor, it should taper away from the tumor, and it must enhance more than the tumor. This sign has traditionally been considered as highly specific for meningioma<sup>34</sup>.

### DUCK

Weakness of the pelvic girdle muscles leads to a characteristic limping, “duck gait”. This is characteristic for proximal myopathies with consequent pelvic floor muscle hypotrophy. Advanced coxarthrosis should be ruled out differentially<sup>35</sup>.

### FLY

Floating blurred sight spots may be caused by deposits within the normally transparent vitreous body of the eye. This phenomenon is called myodesopsia. Patients usually describe them as “flying

flies" (lat. *Muscae volantes*). Fast eye movements follow and are best seen on a light or white background. The described disorders occur not only in the elderly, but also in the younger population, particularly in the short-sighted and following cataract surgery<sup>36</sup>. On the neurological side, it is important to distinguish them from photopsies in migraine with visual aura or migraine equivalents.

### MOTHS

Neurologists occasionally diagnose polyneuropathy or chronic vertebrogenic syndromes as clinical manifestation or consequence of underlying hematologic disease. One such hematologic disease is multiple myeloma, which is characterized by changes in the bones that appear to be "moth-eaten appearance in bone"<sup>37</sup>. This is particularly well seen in cranial bones. Multiple myeloma is a plasma cell cancer in which a clone of abnormal plasma cells multiplies, creates tumors in the bone marrow, and produces large amounts of abnormal antibodies that accumulate in the blood or urine.

### BIRDS OF PREY

Compressive ulnar nerve mononeuropathy is the most common consequence of its compression in the area of the cubital tunnel and the Guyon canal. The clinical picture is characteristic and includes hypotrophy of the hypothenar muscles, hypoesthesia in the ulnar nerve zone of innervation in the forearm and hand, positive Froment's sign and weakness of the abduction of the

little finger. Due to atrophy of the interosseous and lumbrical muscles of the hand, it takes on the typical appearance of a claw-like bird of prey, with the 4th and 5th fingers flexed in the interphalangeal joints and the hypotenor hypo/atrophic<sup>38</sup>.

### BAT

Joubert anomaly, also known as vermian aplasia is an autosomal recessive disorder where there is a variable degree of cerebellar vermal agenesis. The "bat wing" configuration of the 4th ventricle is typical for this anomaly<sup>39</sup>.

### BIRD

Tectal beaking ("bird beak" sign) refers to the fusion of the midbrain colliculi into a single beak pointing posteriorly and invaginating into the cerebellum. It is seen with a Chiari type II malformation<sup>40</sup>.

### FISH

Refsum disease is a rare disease caused by accumulation of phytanic acid in plasma and tissues. It is characterized by anosmia, cataract, retinitis pigmentosa, peripheral neuropathy, cerebellar ataxia, deafness and ichthyosis („fish skin“ disease)<sup>41</sup>.

If we take into account the continuous creativity of neurologists and neuroradiologists in finding animal eponyms, it is to be expected that this list will continue in the future.

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