What's the time? – numerical eponyms and brainstem syndromes

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Abstract:

Brainstem syndromes and their eponyms are well known and numerous, but represent only a drop in the ocean of approximately 450 neurological eponyms. Unlike these "human" eponyms dedicated to famous neurologists, "numerical" eponyms that describe various disorders at the anatomical level of the brainstem, are less known and are rarely used in clinical practice. In this short review, we will give an overview of "human" eponyms and present in more detail the "numerical" eponyms related to brainstem syndromes. The availability of sophisticated neuroradiological and other diagnostics methods cannot replace the importance of neurological examination nor diminish the joy of the neurologist when recognizing a set of signs named after an eponym, or to describe a new one. In any case, neurology is a joy and valuable opportunity for lifelong learning, but also for contribution to continuously growing knowledge about the hidden universe in our nervous system.

Keywords:

eponyms, brainstem, neurology, signs and symptoms

Sažetak:

Koliko je sati? Brojčani eponimi i sindromi moždanog debla

Sindromi moždanog debla i njihovi eponimi dobro su poznati i brojni, ali predstavljaju samo kap u moru od približno 450 neuroloških eponima. Za razliku od ovih „humanih" eponima posvećenih poznatim neurolozima, "numerički" eponimi koji opisuju razne poremećaje na anatomskoj razini moždanog debla, manje su poznati i rijetko se koriste u kliničkoj praksi. U ovom kratkom osvrtu dati ćemo pregled "humanih" eponima i detaljnije prikazati "numeričke" eponime povezane sa sindromima moždanog debla. Dostupnost sofisticiranih neuroradioloških i drugih dijagnostičkih metoda ne može zamijeniti važnost neurološkog pregleda niti umanjiti radost neurologa kada prepoznaju bilo koji od ovih sindroma ili opišu novi. U svakom slučaju, neurologija je radost i dragocjena prilika za cjeloživotno učenje, ali i za doprinos kontinuirano rastućem znanju o skrivenom svemiru u našem živčanom sustavu.

Ključne riječi:

eponimi, moždano deblo, neurologija, znakovi i simptomi

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Brainstem syndromes and their eponyms are well known and numerous, but represent only a drop in the ocean of approximately 450 neurological eponyms. Their application throughout history is a consequence of the development of neurological propaedeutics and consequently topical diagnostics long before the development of neuroimaging techniques. A precise and focused anamnestic data joined with neurological examination, even before the era of modern diagnostic tools, results in the identification of various clinical symptoms and signs. The sum of these symptoms and signs leads to the precise localization of the nervous system damage or dysfunction.

Unlike these "human" eponyms dedicated to famous neurologists, "numerical" eponyms that describe various disorders at the anatomical level of the brainstem, are less known and are rarely used in clinical practice. In this short review, we will give an overview of "human" eponyms and present in more detail the "numerical" eponyms related to brainstem syndromes. In literature we can find 24 brainstem syndromes that carry „human" eponyms and additional 4 that do not have eponyme. Among the syndromes that are named by eponyms are: 4 mesencephalic syndromes (Claude[1], Benedict[2], Nothnagel[3], Weber[4]), 9 pontine (Brissaud-Sicard[5], Gasperini[6], Gellé[7], Grenet[8], Foville[9], Marie-Foix[10], Raymond[11], Raymond-Cestan[12], Millard-Gubler[13-14]) and 11 medulla oblongata related syndromes (Avellis[15], Babinski-Nageotte[16], Cestane-Chenais[17], Reinhold[18], Jackson[19], Wallenberg[20], Dejerine[21], Schmidt[22], Spiller[23], Tapia[24], Vernet[25]). Syndromes that do not carry eponyms are: "facial colliculus"[26], "locked-in"[27], "top of the basilar"[28] and Ondine curse[29].

The first described brainstem syndrome with a "human" eponym dates from 1856 (Millard and Gubler) [13-14], and the last from 1922 (Marie, Foix, Alajouanine)[10]. 10. This period of the second half of the 19th and the beginning of the 20th century is also the time of the renaissance of European, primarily French neurology. The causes of these syndromes are most often vascular (ischemic and hemorrhagic strokes), demyelinating disease, and much less common brainstem neoplasms or inflammatory processes (bacterial, viral, fungal, or granulomatous inflammation).

Brainstem syndromes with „numerical eponyms"

Clinical features of these syndromes are shown in Table 1.

"Half and half" syndrome

Randhawa et al.[30] described a patient with left-side internuclear ophtalmoplegia (INO) (the "half") and moderate underaction of abduction (the "half") in the left eye caused by focal haemorrhagic lesion in the floor of the aqueduct in the region of the dorsal pons with consequent lesion of the left medial longitudinal fasciculus (MLF) and the CN VI fasciculus (sparing the CN VI nucleus). The result is "half and half" syndrome (0,5+0,5).

One-and-a-half syndrome

The "one-and-a-half" syndrome is disturbance of horizontal eye movements in which patients have lateral gaze palsy in one direction, and INO in the other direction. This syndrome is caused by damage of the pontine tegmentum: the MLF, the ipsilateral paramedian pontine reticular formation (RF) or the ipsilateral CN VI nucleus. Multiple sclerosis, brainstem infarction or tumors are the most common causes. It means that one eye cannot move laterally at all (the „one"), and the other can move only in outward direction (the „half"). This is a combination of conjugate horizontal gaze palsy in one direction and INO in the other[31]. The result is "one-and-a-half" syndrome (1+0,5=1,5).

Seven-and-a-half syndrome

Sadaka et al.[32] described a case of a patient with right INO (the „half") and ipsilateral peripheral CN VII palsy (the „seven") caused by small localized right hemipons infarct involving CN VII motor nucleus and facial genu as well as the right MLF. They introduced „seven-and-a-half" syndrome as a new clinico-radiologic syndrome. The result is "seven-and-a-half" syndrome (0,5+7=7,5).

Eight-and-a-half syndrome

Duffy et al.[33] described patient with "one-and-a-half" syndrome (combination of conjugate horizontal gaze palsy in one direction and INO in the other) (the „one-and-a-half") and ipsilateral peripheral CN VII palsy (the "seven"). This clinical findings are called "eight-and-a-half" syndrome. It is caused by damage of the dorzal tegmentum of the caudal pons in the region of the facial colliculus. The lesion is unilateral at the midpontine level with damage to both the paramedian pontine RF and MLF. These structures are intimiately related to the CN VII nucleus and intraaxial fasciculus of the CN VII. Brainstem infarction, and hemorrhage or demyelination lesion are the most common causes. When this occurs, a "one-and-a-half" syndrome plus a CN VII palsy, it is termed an "eight-and-a-half" syndrome. The result is "eight-and-a-half" syndrome (1,5+7=8,5).

Nine syndrome

Mahale et al.[34] described two patients with „nine" syndrome. They present clinical features of the „eight-and-a-half" syndrome: "One-and-a-half" syndrome (combination of conjugate horizontal gaze palsy in one direction and INO in the other) (the „one-and-a-half") and ipsilateral peripheral CN VII palsy (the "seven") associated with hemiataxia (the "half"). The first patient had left "eight-and-a-half" syndrome due to caudal pontine tegmental demyelinating lesion with involvement of inferior cerebellar peduncle responsible for left hemiataxia, and the second patient had right "eight-and-a-half" syndrome due to bleeding in the right caudal pontine tegmentum with extension into midbrain tegmentum/red nucleus responsible for contralateral hemiataxia. The result is "nine" syndrome (1,5+7+0,5=9).

Table 1. Summary of the brainstem syndromes („numerical eponyms")

Syndrome	Signs and symptoms
Half and half syndrome	internuclear ophthalmoplegia (INO) (the "half") and ipsilateral CN VI palsy (the "half"): 0,5+0,5
One-and-a-half syndrome	ipsilateral conjugate gaze palsy (the "one") and INO (the "half"): 1+0,5=1,5
Seventh-and-a-half syndrome	ipsilateral INO (the „half"); and peripheral CN VII palsy (the „seven"): 0,5+7=7,5
Eight-and-a-half syndrome	ipsilateral conjugate horizontal gaze palsy (the "one"); and ipsilateral (INO) (the "half"); and ipsilateral peripheral CN VII palsy (the "seven"): 1+0,5+7=8,5
Nine syndrome	one and a half syndrome (the „one and a half"); and an ipsilateral peripheral CN VII palsy („the seven"); and contralateral hemiparesis, hemihypesthesia or hemiataxia (the „half"): 1,5+7+0,5=9
Thirteen-and-a-half syndrome	ipsilateral conjugate horizontal gaze palsy (the "one"); and an ipsilateral INO (the "half"); and an ipsilateral peripheral CN VII palsy (the "seven"); and an ipsilateral CN V palsy (the "five"): 1+0,5+7+5=13,5
Fifteen-and-a-half syndrome	ipsilateral conjugate gaze palsy (the "one"); and an INO (the "half"); and bilateral peripheral CN VII palsy (the "fourteen"): 1,5+(2x7)=15,5.
Sixteen syndrome	ipsilateral conjugate gaze palsy (the "one"); and an INO (the "half"); and bilateral CN VII palsy (the "fourteen"); and partial hemiparesis (the "half"): 1+0,5+(2x7)+0,5=16
Sixteen-and-a-half syndrome	ipsilateral conjugate horizontal gaze palsy (the "one"); and an ipsilateral INO (the "half"); and an ipsilateral peripheral CN VII palsy (the "seven"); and an impaired ipsilateral auditory nerve (CN VIII) (the "eight"): 1+0,5+7+8=16,5
Twenty-and-a-half syndrome	ipsilateral conjugate gaze palsy (the "one"); and an INO (the "half"); and bilateral CN VII palsy (the "fourteen"); and an unilateral CN V palsy (the "fifth"): 1+0,5+(2x7)+5=20,5
Twenty-four syndrome	bilateral facial palsy (the "fourteen"); and bilateral horizontal gaze palsy (the "two"); and contralateral sensorineural hearing loss (CN VIII) (the "eight"): (2x7)+(2x1)+8=24
Twenty-four-and-a-half syndrome	ipsilateral conjugate gaze palsy (the "one"); and an INO (the "half"); and an ipsilateral CN VII palsy (the "seven"); and bilateral CN VIII palsy (the "sixteen"): 1+0,5+7+(2x8)=24,5

Abbreviations: INO – internuclear ophtalmoplegia; CN – cranial nerve

Thirteen-and-a-half syndrome

Allbon et al.[35] described patient with "eight-and-a-half" syndrome, including "one-and-a-half" syndrome - combination of conjugate horizontal gaze palsy in one direction and INO in the other (the „one-and-a-half") and ipsilateral peripheral CN VII palsy (the "seven") associated with an ipsilateral CN V palsy (the "five") because of a post-transplant lymphoproliferative disorder. When this occurs, a "one-and-a-half" syndrome plus CN VII palsy, and ipsilateral CN V palsy it is termed as "thirteen-and-a-half" syndrome. The result is "thirteen-and-a-half" syndrome (1,5+7+5=13.5).

Fifteen-and-a-half syndrome

Bae et Song[36] first described "fifteen-and-a-half" syndrome, including "one-and-a-half" syndrome - combination of conjugate horizontal gaze palsy in one direction and INO in the other (the „one-and-a-half") and bilateral CN VII palsy (the "fourteen"). The dorsal tegmentum of the caudal pons including the MLF, the paramedian pontine RF, CN VI nucleus, and the adjacent CN VII is the anatomical basis of this rare syndrome. The result is "fifteen-and-a-half" syndrome (1,5+(2x7)=15,5).

Sixteen syndrome

Li et al.[37] first described "sixteen" syndrome, including "one-and-a-half" syndrome – combination of conjugate horizontal gaze palsy in one direction and INO in the other (the "one-and-a-half"), bilateral CN VII palsy (the "fourteen"), and partial hemiparesis (the "half"). In this case, the axons of internuclear neurons from the CN VI nucleus were affected nearly as they passed through the midline in the MLF rising up to the level of the CN III subnucleus that controls the medial rectus. The result is "sixteen" syndrome (1,5+(2x7)+0,5=16).

Sixteen-and-a-half syndrome

Borgman and Jackson[38] described a case of "sixteen-and-a-half" syndrome, including "one-and-a-half" syndrome – combination of conjugate horizontal gaze palsy in one direction and INO in the other (the "one-and-a-half"), ipsilateral CN VII palsy (the "seven"), and sensorineural hearing impairment consistent with CN VIII involvement (the "eight"). The syndrome is caused by ischemic lesion of the dorsal tegmentum of the caudal pons. The result is "sixteen-and-a-half" syndrome (1,5+7+8=16,5).

Twenty-and-a-half syndrome

Dube et al.[39] described a case of "twenty-and-a-half" syndrome, including "one-and-a-half" syndrome – combination of conjugate horizontal gaze palsy in one direction and INO in the other (the "one-and-a-half"), bilateral CN VII palsy (the "fourteen") and CN V palsy (the "five"). The syndrome is caused by ischemic lesion of right posterolateral aspect of pons and medulla. The result is "twenty-and-a-half" syndrome (1,5+(2x7)+5=20,5).

Twenty-four syndrome

Karadan et al.[40] described a case of "twenty-four" syndrome, including "one-and-a-half" syndrome – combination of conjugate horizontal gaze palsy in one direction and INO in the other (the "one-and-a-half"), bilateral CN VII palsy (the "fourteen"), contralateral sensorineural hearing loss (CN VIII) (the "eight") and hemiparesis (the "half"). The syndrome is caused by pontine hemorrhage. The result is "twenty-four" syndrome (1,5+(2x7)+8+0,5=24).

Twenty-four-and-a-half syndrome

Man et al.[41] described a case of "twenty-four-and-a-half" syndrome, including "one-and-a-half" syndrome – combination of conjugate horizontal gaze palsy in one direction and INO in the other (the "one-and-a-half"), ipsilateral CN VII palsy (the "seven"), contralateral hemifacial spasm and ataxia, and bilateral hearing loss (CN VIII) (the "sixteen"). The syndrome is caused by pontine cavernoma with perilesional oedema. The result is "twenty-four-and-a-half" syndrome (1,5+7+(2x8)=24,5).

Theoretical knowledge and clinical experience in neurological propaedeutics and its neuroanatomical background is crucial in recognizing the localization of a brainstem lesion and diagnosing these "human" or "numerical" brainstem syndromes. Personally, we prefer "human" eponyms of brainstem syndrome because behind each of them lies an interesting story related to their origin and the work of top neurologists who described them based on their clinical observations, without the use of, at that time, unavailable supportive diagnostic methods. If we take into account all possible numerous combinations of clinical signs and symptoms caused by brainstem lesions still not described in clinical practice, we could expect future reports and papers describing new "numerical" syndromes. Perhaps, someone in the future, will describe the case of „seventy-eight" syndrome with unilateral affection of all 12 CN (result: 1 + 2 + 3 + 4 + 5 + 6 + 7 + 8 + 9 + 10 + 11 + 12 = 78) caused by Garcin's (hemibase) syndrome. Eponyms in neurology are the result of skilful neurological examination and knowledge about the neuroanatomical basis of neurological symptoms and signs. The eponyms are a valuable thread that begins in the past and leads us through the present to the future. The availability of sophisticated neuroradiological and other diagnostics methods cannot replace the importance of neurological examination nor diminish the joy of the neurologist when recognizing a set of signs named after an eponym, or to describe a new one. In any case, neurology is a joy and valuable opportunity for lifelong learning, but also for contribution to continuously growing knowledge about the hidden universe in our nervous system.

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