NEUROLOGY SECRETS

SIXTH EDITION

QUESTIONS YOU WILL BE ASKED

TOP 100 SECRETS ■ KEY POINTS ■ WEBSITES

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NEUROLOGY

SIXTH EDITION

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PREFACE

The first edition of *Neurology Secrets* was published over 20 years ago. The subsequent editions have tracked the dramatic advances in the field of neurology. In this 6th edition, each chapter has been revised and updated to reflect the current state of the art and science of the topic. New chapters have been added that emphasize the multidisciplinary nature of the practice of neurology. The purpose of this book is to focus on the fundamental issues of the field of neurology. This edition follows the now familiar *Neurology Secrets* format with each chapter organized as a series of questions and answers. Key points are highlighted in each chapter. The chapters are developed to form a basis for further discussion with directed references and reading for more in-depth review. Each chapter is designed to provide the reader with a concise and accurate review which crystallizes the essential features of each topic.

We are indebted to the contributors of *Neurology Secrets* 6th edition. Most of these authors have some academic relationship to Baylor College of Medicine in Houston, Texas, either as current or former faculty members or trainees. This is a tradition begun with the first edition and has continued through the subsequent editions. Some are new to this edition; however, there are a number who have been contributing from the start.

The founding editor of *Neurology Secrets*, Loren A. Rolak, MD, who also edited the subsequent editions, relinquished that role for this 6th edition—although he continues as a contributor. He began this effort within the context of his role as member of the faculty and Neurology Residency Program Director, Department of Neurology, at Baylor College of Medicine in Houston, Texas. Throughout the years, Dr Rolak, who is now at Marshfield Clinic in Marshfield, Wisconsin, has been committed to medical education. The previous editions of *Neurology Secrets* are, in part, evidence of his dedication and skill as an educator, and we trust that the 6th edition does justice to the rich tradition he established.

Joseph S. Kass, MD, JD

Eli M. Mizrahi, MD

DEDICATION

To Loren A. Rolak, MD, founding editor of Neurology Secrets, expert and compassionate clinician, and skilled and dedicated medical educator.

Joseph S. Kass, MD, JD

Eli M. Mizrahi, MD

TOP 100 SECRETS

These secrets are 100 of the top board alerts. They summarize the concepts, principles, and most salient details of neurology.

1. Long-term potentiation is the synaptic mechanism of learning and memory.
2. If the facial nerve is damaged (such as from Bell's palsy), the entire side of the face is weak. If the cortical input to the facial nerve is damaged (such as from a stroke), only the lower half of the face will be weak.
3. A dilated or "blown" pupil implies compression of the III nerve.
4. Noncommunicating hydrocephalus is a medical emergency because the obstructed cerebrospinal fluid (CSF) will cause the intracranial pressure to rise.
5. To distinguish between a common peroneal neuropathy at the popliteal fossa and a L5 radiculopathy, examine for weakness in hip abduction and ankle inversion. Weakness in these muscles indicates L5 radiculopathy.
6. The first step in treating patients with neurologic disease is to localize the lesion.
7. Peripheral neuropathies often produce distal weakness, atrophy, fasciculations, sensory loss, and pain.
8. Spinal cord diseases often produce pyramidal tract deficits, sphincter problems, and a sensory level.
9. Brainstem lesions often produce cranial nerve deficits accompanied by weakness or numbness on the contralateral body.
10. Myopathies usually cause proximal symmetric weakness, with or without other symptoms.
11. Myotonic dystrophy is the most common muscular dystrophy in adults.
12. Myasthenia gravis typically presents with subacute to chronic, fatigable, proximal arm and leg weakness, ptosis, and diplopia.
13. Myasthenia gravis can cause rapid onset neuromuscular respiratory failure, which can be fatal and is therefore a neurological emergency.
14. Lambert–Eaton myasthenic syndrome is associated with cancer (typically small-cell carcinoma of the lung) in approximately 60% of cases, and may be the first manifestation of malignancy.
15. The most common causes of peripheral neuropathy are diabetes and alcoholism.
16. The most often overlooked cause of peripheral neuropathy is genetic.
17. The spinal fluid of patients with Guillain–Barré syndrome has high protein but normal cell counts (cytoalbuminologic dissociation).
18. Compression of the C6 nerve root causes radicular pain in the lateral side of the forearm and thumb, C7 compression causes pain in the index and middle fingers, and C8 compression causes symptoms in the fourth and fifth fingers.
19. Ninety-five percent of lumbar disc herniations occur at the L4/5 or L5/S1 disc spaces.
20. The dermatome for the nipple line is at T4 and the umbilicus is at T10.

21. The first seven cervical nerves exit above the vertebral body with the eight exiting below C7, and the remainder of the spinal roots exit below their corresponding vertebral body.
22. Transverse myelitis is an inflammatory process that is localized over several segments of the cord functionally transects the cord.
23. The most common metastatic tumors to the spinal cord are breast, lung, gastrointestinal tract, lymphoma, myeloma, and prostate.
24. A unilateral lesion within the brainstem often causes "crossed syndromes," in which ipsilateral dysfunction of one or more cranial nerves is accompanied by hemiparesis and/or hemisensory loss on the contralateral body.
25. Symptoms of brainstem ischemia are usually multiple, and isolated findings (such as vertigo or diplopia) are more often caused by peripheral lesions affecting individual cranial nerves.
26. Brainstem glioma is the most frequent brainstem neoplasm. Other brainstem neoplasms include ependymomas that occur in the fourth ventricle and metastatic lesions that may originate from malignant melanomas or carcinomas of the lung and breast.
27. Ménière's disease presents with the symptomatic triad of episodic vertigo, tinnitus, and hearing loss. It is caused by an increased amount of endolymph in the scala media. Pathologically, hair cells degenerate in the macula and vestibule.
28. Central pontine myelinolysis (osmotic demyelination syndrome) occurs primarily in patients suffering from malnutrition or alcoholism complicated by hyponatremia. Rapid correction of the hyponatremia has been implicated as a cause of the pathologic abnormality.
29. Cerebellar strokes and hemorrhage may result in a neurological/neurosurgical emergency by causing obstructive hydrocephalus.
30. Episodic ataxia type 2 is caused by mutations in the same gene (*CACN1A4*) as familial hemiplegic migraine and spinocerebellar ataxia type 6.
31. Differential diagnosis of cerebellar/ataxic conditions can vary by age: (1) adults are more likely to have autosomal dominant spinocerebellar ataxias, degenerative forms of ataxia, extra-axial tumors, paraneoplastic syndromes, and vascular insults to the cerebellum; (2) pediatric patients are more likely to manifest with autosomal recessive cerebellar ataxias, intra-axial cerebellar tumors, infections, or congenital/developmental abnormalities.
32. Levodopa remains the most effective therapy for Parkinson's disease, but management of levodopa-related complications continues to be a challenging probelm that often requires treatment with multiple medications and deep brain stimulation.
33. Essential tremor is a familial disorder, but the genes responsible for this alcohol-responsive action tremor have not yet been identified.
34. Cardinal symptoms of autonomic insufficiency include orthostatic hypotension, bowel and bladder dysfunction, impotence, and sweating abnormalities.
35. Autonomic failure can be seen in the setting of systemic peripheral neuropathies, the most common being diabetic neuropathy, or can be seen without involvement of the sensorimotor neurons, such as pure autonomic failure. Some dysautonomias have autoimmune etiology.
36. The diagnosis of multiple sclerosis requires lesions disseminated in time and in space: two separate symptoms at two separate times.
37. Faulty interpretation of magnetic resonance imaging is the most common error leading to the misdiagnosis of multiple sclerosis.
38. No treatment has yet been proven to alter the level of long-term disability in multiple sclerosis.

39. Dementia is a category, not a diagnosis, and Alzheimer's disease is the most common form of dementia.
40. Most causes of dementia are treatable even if not curable.
41. Frontotemporal dementia (FTD) can present with either behavioral symptoms or primary progressive aphasia.
42. Progressive supranuclear palsy and corticobasal syndrome are parkinsonian disorders associated with dementia and tau aggregation.
43. Cerebral amyloid angiopathy can cause dementia and has manifestations apart from classic lobar hemorrhage.
44. Ten to 15% of patients with amnestic mild cognitive impairment progress to development of Alzheimer's disease each year.
45. Repetitive mild traumatic brain injury can lead to a syndrome of progressive cognitive decline, behavioral and mood changes, and motor/parkinsonian symptoms known as chronic traumatic encephalopathy. The diagnosis can only be confirmed postmortem by identification of hyperphosphorylated tau protein deposits in the sulci.
46. The diagnosis of posttraumatic stress disorder requires exposure to trauma with development of symptoms (intrusive thoughts, re-experiencing, avoidance, negative alterations in cognition and mood, and marked alterations in arousal and reactivity) lasting >1 month and causing significant distress and/or impairment in functioning.
47. An acute onset of cognitive decline with fluctuations in orientation and level of alertness is the hallmark of delirium rather than indicative of dementia.
48. The main clinical feature of stroke is sudden onset of a focal neurological deficit.
49. The only Food and Drug Administration-approved treatment for acute ischemic stroke is intravenous (IV) tissue plasminogen activator, administered within 3 hours of the time the patient was last seen normal.
50. Hemorrhagic strokes often present with a diminished level of consciousness; ischemic strokes rarely do.
51. A depressed level of consciousness (Glascow Coma Scale <8) is the greatest risk factor for airway obstruction and aspiration.
52. Think of cerebral amyloid angiopathy as the most likely cause of spontaneous lobar intracerebral hemorrhage in patients age >55 years.
53. Nimodipine in aneurismal subarachnoid hemorrhage is neuroprotective as it improves outcome but has not shown to reduce vasospasm.
54. Early decompressive hemicraniectomy after large hemispheric ischemic stroke within 48 hours for patients <60 years of age improves survival and functional outcome.
55. Steroids are not recommended in Guillain–Barré syndrome.
56. Treat convulsive status epilepticus early and aggressively with benzodiazepines
57. Brain death is a clinical diagnosis.
58. Central nervous system (CNS) tumors are classified into four grades according to the World Health Organization (WHO) grading system.
59. Glioblastoma (Grade IV astrocytoma) is the most common and malignant primary brain tumor in adults.
60. Brain metastases are the most common intracranial tumors in adults, occurring nearly 10 times more often than primary brain tumors.

61. Intrathecal methotrexate use has been associated with aseptic meningitis, transverse myelopathy, encephalopathy, and leukoencephalopathy.
62. Do not prescribe opioid or butalbital-containing medications as first-line treatment for recurrent headache disorders.
63. The risk of postdural puncture headache can be greatly reduced by use of an atraumatic needle. Bedrest following the procedure is not preventive.
64. Headache or neck pain is the only symptom of cervical artery dissection in 8% of individuals.
65. A seizure is a single event, while epilepsy refers to (1) recurrent unprovoked seizures, (2) a single seizure with a high risk for recurrent seizures, or (3) recurrent reflex seizures.
66. Epilepsy surgery has the best outcomes in patients with a structural lesion (80%) or those with temporal lobe epilepsy (60% to 70%).
67. Medication-refractory (pharmacoresistant) epilepsy is diagnosed when two or three antiepileptic medications at appropriate doses fail to control seizures.
68. Sleep problems occur frequently in individuals with uncomplicated medical histories (20% to 40%) and very frequently in children and adults with complicated medical histories (40% to 80%).
69. All patients with a stroke must have screening for cardiovascular disease.
70. Patients with uremia often develop a metabolic encephalopathy with signs of neuronal depression such as lethargy as well as excitation such as myoclonus.
71. Most patients with Cushing's disease have frank weakness with myopathic findings on electromyography.
72. Decompensated hypothyroidism can cause myxedema coma with mortality rates as high as 25% to 60%.
73. Cerebrovascular ischemic events occur 13 times more frequently in pregnant women than in age-matched nonpregnant women.
74. Headache, jaw claudication, and constitutional symptoms compose the triad of clinical symptoms often found in temporal arteritis.
75. Patients suspected of having bacterial meningitis should receive adjunctive dexamethasone along with empiric antibiotics. The dosing regimen is dexamethasone 4 mg IV every 6 hours for 4 days with the first dose given either 30 minutes prior to the first dose of antibiotics or concomitantly with the first dose of antibiotics. If the CSF cultures indicate the pathogen is not *Streptococcus pneumoniae*, then dexamethasone may be discontinued.
76. Herpes simplex virus-1 (HSV-1) encephalitis should be considered in a patient presenting with fever, behavioral changes, and/or seizures and should be treated empirically with IV acyclovir. HSV polymerase chain reaction in CSF can be negative in the first few days of infection, necessitating repeating lumbar puncture 3 or more days after infection onset.
77. The differential diagnosis for a ring-enhancing lesion in a person with Acquired Immunodeficiency Syndrome includes most commonly *Toxoplasma gondii* and primary CNS lymphoma but also includes tuberculoma, cryptococcoma, histoplasmoma, and other fungal infections, bacterial brain abscess, metastatic disease, and primary brain tumor.
78. Neurocysticercosis is the most common infectious cause of epilepsy with treatment strategies varying depending on cyst life cycle stage and location within the nervous system.
79. The possibility of multiple mutation mechanisms should be considered in ordering and interpreting diagnostic test results for many neurogenetic diseases.

80. The expanding use of genome-level technologies is revealing a previously unsuspected degree of phenotypic variability in many neurogenetic diseases.

81. Causes of intrauterine infection include toxoplasmosis and other agents, such as rubella, cytomegalovirus, and herpes simplex virus.

82. Simple febrile seizures are generalized tonic or tonic–clonic seizures that typically occur between 3 months and 5 years of age. They are often accompanied by a fever greater than 38° C not associated with a CNS infection, and last less than 15 minutes with no focal features and no recurrence within 24 hours. No postictal neurologic abnormalities typically occur.

83. Headaches concerning for an intracranial mass include a recent onset of headaches or change in character of chronic headaches, headaches that awaken the patient from sleep or are present on awakening in the morning, and headaches in association with altered mental status, vomiting, constriction of visual fields, or focal neurologic deficits.

84. Psychiatric and neurologic disorders are highly comorbid and bidirectionally related, and many neurologic disorders may present first with psychiatric symptoms; neurological and medical etiologies should be considered for patients presenting with psychiatric symptoms.

85. Psychiatric disorders are debilitating but are highly treatable; all patients should be screened for common disorders and suicidality.

86. Somatic symptom disorders (including functional neurologic symptom disorder) are commonly comorbid with focal neurologic disorders and can improve with close follow-up with a single provider and psychotherapy.

87. The cardinal feature of delirium is impaired environmental awareness and ability to direct, sustain, or appropriately shift attention. No other psychiatric disorder can be diagnosed in the context of delirium.

88. Acute and chronic disorders of the vestibular system are characterized by disturbances in the behaviors mediated by the vestibular system, e.g., blurred vision, vertigo, impaired balance, nausea, and temporary changes in cardiovascular measures.

89. Most vestibular disorders can be treated. Some disorders are best treated with exercise and other aspects of rehabilitation; other disorders are best treated with medication and/or surgery.

90. The normal adult electroencephalogram (EEG), relaxed with eyes closed, is characterized by 9 to 11 cycles per second activity in the back of the brain (occipital lobes) called the alpha rhythm.

91. Periodic lateralizing epileptiform discharges on an EEG imply an acute, large lesion involving one hemisphere, such as a stroke or focal encephalitis.

92. The generalized three per second spike and wave pattern on an EEG is usually seen in patients with absence seizures.

93. The finding on an EEG that is most suggestive of focal epilepsy is a very brief (less than 70 ms) transient deflection called a spike.

94. The most common compression neuropathies are carpal tunnel syndrome (median nerve compression at the wrist) and cubital tunnel syndrome (ulnar nerve compression at the elbow).

95. The earliest conduction abnormalities in Guillain–Barré syndrome (acute inflammatory demyelinating polyneuropathy) are absent H reflexes.

96. Motor conduction abnormalities in chronic inflammatory demyelinating polyneuropathy are abnormal F waves, distal latencies delay, conduction block with dispersion of distal potential, and motor velocities >20% of normal.

97. A patient with parkinsonism, ataxia, dysautonomia, and alpha-synuclein-positive glial cytoplasmic inclusions at autopsy has multiple system atrophy.
98. *C9orf72* hexanucleotide repeat expansion is the most frequent genetic alteration underlying familial amyotrophic lateral sclerosis (ALS) including ALS with frontotemporal dementia (ALS/FTD).
99. An infiltrative (diffuse) glioma with astrocytic cytology, mitotic activity, microvascular proliferation, and necrosis is a WHO Grade IV glioblastoma.
100. Tuberculous meningitis and neurosarcoidosis both have a predilection for the basilar meninges.