You will have 120 minutes to complete this 13-page examination of 49 questions. The test is divided into four sections based on the type of question: Single best answer, Single or multiple best answers, Matching, Short Answer. Please use the answer sheet to record your answers. Please print your name on the top of each page of the answer sheet. Good Luck!

Part I. Choose the single best answer to the following questions (1 point each question)

1. An 85-year-old male trips and falls, hitting his head. He gets up quickly and appears to have suffered no injury. Ten days later, he complains of headache and has progressive confusion and falls to the right. CT scan demonstrates a concave-shaped area of hyperdensity compressing the left parietal lobe. What is the most likely source of bleeding in this patient?

   A. bridging veins
   B. middle meningeal artery
   C. middle cerebral artery
   D. anterior cerebral artery
   E. posterior cerebral artery

2. Arnold Chiari Malformation may include all of the following but

   A. syringomyelia
   B. cerebellar tonsil herniation
   C. noncommunicating hydrocephalus
   D. myelomeningocele
   E. Dandy-Walker malformation

3. Which of the following is NOT true of hypertensive disease of the small penetrating arteries in the brain:

   A. It is associated with lacunar infarctions
   B. It is associated with intracerebral hemorrhage
   C. It occurs commonly in the pons
   D. It occurs commonly in the cerebral cortex
   E. It occurs commonly in the basal ganglia

4. Which of the following has the highest bleeding risk:

   A. Cavernous angioma
   B. Venous malformation
   C. Arteriovenous malformation
   D. Capillary telangiectasia
   E. Carotid stenosis
5. Primary HIV infection produces all of the following EXCEPT

A. acute aseptic meningitis
B. subacute encephalitis
C. vacuolar myelopathy
D. inflammatory myopathy
E. demyelinating polyneuropathy

6. Regarding neuropharmacology of Alzheimer's disease (AD), which of the following is true?

A. Drugs inhibiting acetylcholinesterase dramatically reverse the cognitive deficits in AD
B. Serotonergic drugs are useful in the treatment of AD
C. Anticholinergic drugs produce improvements in cognitive function in AD
D. Loss of cholinergic function in the brain correlates with the degree of cognitive impairment in AD
E. Dopamine agonists produce improvements in cognitive function in AD.

7. A 46 year old man has had progressive, bilateral weakness of arms and legs for several months. Motor examination reveals diffuse weakness, atrophy and fasciculations in the arms and legs. Deep tendon reflexes are increased. Sensory examination is normal.

7a. The most likely diagnosis is

A. Multiple sclerosis
B. Amyotrophic lateral sclerosis
C. Myasthenia gravis
D. Cerebral astrocytoma
E. Muscular dystrophy

7b. The pathology in this disorder is most prominent in

A. The dorsal columns of the spinal cord
B. The ventral horn of the spinal cord
C. The lateral columns of the spinal cord
D. Peripheral nerve myelin
E. Cerebellar vermis

7c. The most useful test in making this diagnosis would be:

A. EEG
B. MRI brain
C. Spect
D. Tensilon test
E. EMG-NCS
8. Muscle fiber type grouping seen on muscle biopsy is characteristic of

A. muscular dystrophy
B. polyneuropathy
C. myasthenia gravis
D. amyotrophic lateral sclerosis
E: B and D

9. Which is NOT true about the neurologic manifestations of normal aging

A. Cognitive function begins to decline after the age of 30
B. Brain weight starts decreasing with age in one's 20's
C. Reaction time, fine coordination, muscular power are diminished in elderly individuals
D. Neuronal loss is present in many individuals over the age of 80
E. Granulovacuolar degeneration, neuritic plaques and neurofibrillary tangles are seen in the hippocampus in elderly individuals.

10. Clinical features of Parkinson's disease include all BUT

A. resting tremor
B. spasticity
C. impaired balance
D. hypokinesia
E. slowed gait

11. Chorea may be produced by all BUT

A. rheumatic fever
B. systemic lupus erythematosus
C. infarction of the caudate nucleus
D. L-Dopa

12. Which of the following is true about Huntington's disease?

A. X-linked inheritance
B. Neuronal loss in the caudate nucleus
C. Neuronal loss in the midbrain tegmentum
D. Autosomal recessive inheritance
E. Damage to the subthalamic nucleus
13. A 12 year old child is found to have a cerebellar tumor. The most likely diagnosis is:

A. metastatic tumor  
B. medulloblastoma  
C. ependymoma  
D. glioblastoma multiforme  
E. choroid plexus papilloma

14. What is true about meningiomas:

A. They are more common in men than women  
B. Recurrent tumors are very successfully treated with chemotherapy  
C. Pregnancy or estrogen replacement is associated with tumor recession.  
D. Most meningiomas are asymptomatic.  
E. The incidence peaks in early adulthood

15. Spinal cord metastasis

A. is often painless  
B. appropriate treatment may be either surgery or radiation therapy  
C. produce bilateral motor deficits, with atrophy, fasciculations and reflex loss.  
D. is a medical emergency  
E. B and D

16. What is true about chronic meningitis?

A. Tubercular meningitis generally produces hypoglycorachia (low blood glucose level in the CSF).  
B. Inflammatory response is usually most prominent over the convexities, sparing the basal cisterns.  
C. Extension of the inflammation to the arteries in the subarachnoid space leads to subarachnoid hemorrhage  
D. Candida and aspergillus usually spread to the CNS from local infection in the sinuses.  
E. A and B

17. Developmental milestones can be delayed by the following:

A. dysmyelination  
B. maternal smoking  
C. neonatal muscle disease  
D. coexistent medical disease such as cardiac disease  
E. all of the above may contribute to a delay in the acquisition of developmental milestones.
18. Of the following normal reflex behaviors, which of the following disappears over time?

A. propping reflex  
B. parachute reflex  
C. stepping reflex  
D. Moro reflex  
F. supporting reaction (neonate)

19. Which is the following statements is true:

A. In the adult patient, the spinal cord ends at about L4-5.
B. In the lumbosacral region, the most common site of disc herniation is the L1-2 level.
C. Diminished knee jerk may be found with compression of either the L1 or L2 nerve root.
D. Disc bulges and herniations may be present without symptoms referable to the spine or nerve roots.
E. A, C

20. With respect to herniations of the intervertebral disc:

A. Lateral disc herniation at the C5-6 level may cause C6 radiculopathy.
B. Extreme or far lateral disc herniation at the L4-5 level may cause L4 radiculopathy.
C. Medially located lumbar disc herniations may impinge on roots that are still within the dural sac.
D. Thoracic radiculopathy is not usually due to disc herniations.
E. A-D are all correct responses.

21. With respect to cauda equina syndrome, which of the following statements is true:

A. It is most often caused by trauma or tumor.
B. It causes radicular pain, paresthesias, and sensory loss referable to multiple bilateral roots.
C. Bowel and bladder dysfunction may occur early.
D. Bilateral leg weakness and loss of lower extremity reflexes can be observed.
E. All of the above statements are true.

22. Complete cord transaction at the level of C5 would be expected to cause all of the following EXCEPT:

A. incontinence  
B. paraplegia  
C. paralysis of the diaphragm  
D. spastic paralysis below lesion  
E. complete anesthesia below lesion
23. A 34 year old woman with epilepsy develops waxing and waning confusion over 2 weeks. No motor convulsions were observed. In the ER she is afebrile and appears lethargic and her responses are slow. She is receiving valproic acid and phenobarbital to prevent seizures and citalopram to treat depression. No focal motor, sensory, reflex deficits are found.

23a. The differential diagnosis for confusion in this patient includes

A. Medication toxicity
B. Confusional migraine
C. Non-convulsive seizures
D. Herpes encephalitis
E. A, B, C

23b. The least important of the following tests to order in the ER is

A. Brain MRI
B. Toxicology screen and alcohol level
C. Valproic acid and phenobarbital levels
D. Electrolytes
E. Liver function tests and ammonia level

24. Which of the following are causes of epilepsy?

A. Head trauma
B. Hyponatremia
C. Cerebral infarction
D. A, B, C
E. A, B, C

25. Which of the following is NOT true of a patient with grand mal seizures?

A. the seizure reflects an abnormal neuronal discharge
B. the seizures are associated with loss of consciousness
C. Tonic clonic movements are invariable in the unassisted patient
D. epileptic spike on EEG may be seen interictally (between seizures)
E. postictal confusion is generally present

26. Consequences of transtentorial herniation include

A. pontine hemorrhage
B. PCA compression and occipital infarction
C. CNIII compression
D. A and C
E. A, B, C
27. Drug induced parkinsonism may be distinguished from Parkinson's disease by:

A. absence of resting tremor
B. presence of action tremor
C. abnormal mental status examination
D. A, C
E. these cannot be distinguished

28. Tardive dyskinesia

A. occurs after prolonged use of dopamine receptor blocking drugs
B. may include chorea, dystonia and athetosis
C. may be relieved temporarily by increasing the offending agent
D. is believed to reflect increased sensitivity of dopamine receptors
E. all of the above

29. Which of the following is true of Spina Bifida?

A. It is associated with mental retardation
B. Myelomeningoceles occur most commonly in the cervical region
C. Maternal levels of alphafetoprotein levels may detect this malformation antenally
D. It is associated with maternal pyridoxine deficiency
E. A and C

30. MS plaques usually demonstrate the following pathologic features EXCEPT

A. periventricular location
B. sharp demarcation
C. myelin loss
D. axon loss
E. early inflammatory changes

31. Myasthenia gravis is characterized by:

A. Fatigable weakness
B. Increment in motor response to repetitive nerve stimulation.
C. Improvement with use of anticholinergic medications.
D. Autoimmune attack on pre-synaptic membrane.
E. All of the above.
32. Which differentiates polyneuropathy from myopathy?

A. Neuropathy typically affects proximal more than distal muscles.
B. Myopathies will have abnormal sensory nerve conduction studies.
C. Muscle biopsy of neuropathy will show fiber type grouping; myopathy will not.
D. Muscle fibers undergo Wallerian degeneration, nerve fibers do not.
E. Myopathies typically show upper motor neuron signs.

33. A radiculopathy will demonstrate which of the following characteristics?

A. bilateral, symmetric weakness and sensory loss.
B. Dermatomal pattern of sensory loss.
C. Incremental response to higher rates of repetitive nerve stimulation.
D. A regional distribution of weakness, sensory loss and reflex loss.

34. Parkinsonism may be due to which one of the following?

A. loss of pars compacta nigral neurons
B. loss of pre-synaptic dopamine
C. loss of post synaptic neurons in the putamen
D. loss of post synaptic dopamine receptors
E. all of the above

35. Each the following statements about the differential diagnosis of delirium are true EXCEPT,

A. Delirium may be exacerbated by medications, illicit drug use, or alcohol.
B. Systemic infection may precipitate delirium.
C. Most cases of delirium are caused by brain injury or disease.
D. In an older patient, delirium should trigger an evaluation for underlying dementia.
E. Sleep deprivation and unfamiliar environment may contribute to delirium

36. Choose the single best answer concerning seizure that occurs after alcohol withdrawal:

A. Most seizures are focal in onset.
B. Status epilepticus is a common manifestation of “rum fits”.
C. There is a characteristic pattern of focal slowing on EEG associated with alcohol withdrawal seizures.
D. Peak incidence of occurrence is about 24 hours after cessation of drinking.
E. Other symptoms of withdrawal, delirium tremens, usually precede seizure(s).
37. Infectious agents may obtain access to the CNS via

A. hematogenous spread  
B. via peripheral nerves  
C. direct extension from adjacent structures  
D. A, C  
E. A, B, C

**Part II. Choose the correct statement(s) or answer; one or more statement may be correct. (2 points each question)**

38. Regarding vitamin B1 or thiamine:

A. Thiamine is an essential co-factor in several enzymes involved in protein metabolism.  
B. Thiamine deficiency may be associated with signs of abnormal cardiac dysfunction, such as tachycardia or orthostatic hypotension.  
C. Dextrose should be administered prior thiamine in cases of suspected thiamine deficiency.  
D. In patients with thiamine deficiency, pathological changes may be seen in the dorsomedial thalamus, periaqueductal grey matter, and vestibular nuclei.  
E. Alcohol consumption has a direct effect on thiamine metabolism in the brain.

39. Which of the following represent potential neuropathological complications of perinatal ischemic brain injury?

A. Status marmoratus  
B. Periventricular leukomalacia  
C. Lacunar infarction  
D. Multicystic encephalopathy  
E. Ulegyria  
F. Hydranencephaly  
G. Holoprosencephaly  
H. Syringohydranencephaly  
I. Encephalocele

40. Which of the following is/are pathologic features of Alzheimer's disease:

A. Hirano bodies  
B. amyloid plaques  
C. neurofibrillary tangles  
D. Lewy bodies  
E. Alzheimer type II astrocyte  
F. granulovacuolar degeneration  
G. neuronal loss  
H. microglial nodules  
I. Pick Body
Part III. Matching. Choices may be used once, more than once, or not at all. (1/2 point each match)

41. Match the following diseases (a-e) with clinical, pathologic characteristics (1-5)

a. Adrenoleukodystrophy
   1. Storage disease, autosomal recessive inheritance, cherry red spot on retina, higher prevalence in Ashkenazi Jews
b. Tay Sachs Disease
   2. Storage disease, cells with lacy striated cytoplasm in white matter, spleen, and elsewhere, dementia, spasticity, splenomegaly
c. Metachromatic leukodystrophy
   3. Dysmyelination, X linked inheritance, axonal degeneration and segmental demyelination, elevated very long chain fatty acid levels
d. Alexander's disease
   4. Dysmyelination, autosomal recessive inheritance, membrane-bound vacuoles containing crystalloid structures which shift absorbance spectrum of certain dyes
e. Gaucher's disease
   5. Dysmyelination, rosenthal fibers, dementia, macrocephaly

42. Match the following tumors (a-e) with the neuropathological description (1-5):

a. Ependymoma
   1. Childhood tumor arising in the cerebellum; micropathology shows very primitive, undifferentiated cells
b. Oligodendroglioma
   2. A tumor typically arising in the fourth ventricle; micropathology includes perivascular pseudorosettes
c. Pilocytic astrocytoma
   3. Often calcified, arises in the cerebral hemispheres; micropathology "fried egg" appearing cells
d. Medulloblastoma
   4. Poorly defined tumor, infiltrative; micropathology includes satellitosis or a tendency to surround neurons
e. Astrocytoma
   5. Slow growing tumors, with often favorable prognosis; micropathology includes bipolar cells with hair like processes
43. Match the disease (a-e) with the pattern(s) of neuropathy (1-5) it produces

a. diabetes  
   1. mononeuropathy

b. compression  
   2. mononeuritis multiplex

c. vasculitis  
   3. polyneuropathy

d. alcohol  
   4. 1,2

e. immune-mediated  
   5. 1,2,3

44. Match the disease (a-e) with its site of prominent pathology (1-5):

a. Huntington's disease  
   1. cerebral cortex

b. Parkinson's disease  
   2. midbrain tegmentum

c. Diffuse Lewy body disease  
   3. caudate nucleus

d. Wernicke's encephalopathy  
   4. lateral and dorsal columns

e. B12 deficiency  
   5. mammillary bodies

45. Match the ability (a-e) with the expected age of achievement(1-5):

a. Pincer grasp  
   1. 2 months

b. Stands alone  
   2. 6 months

c. Walks independently  
   3. 10 months

d. Rolls over from prone to supine  
   4. 12 months

e. Sits securely  
   5. 15 months
46. Match the following clinical features (a-f) with the most likely underlying cause of dementia (1-6):

a. dementia with involuntary movements of limbs and trunk
   1. Vascular dementia

b. rapidly progressive dementia with early appearance of myoclonus
   2. Jacob Creutzfeld disease

c. stepwise progression of dementia with focal neurologic symptoms
   3. Frontotemporal Dementia

d. triad of dementia, urinary incontinence and gait disturbance
   4. Normal pressure hydrocephalus

e. parkinsonism, fluctuating cognition and visual hallucinations
   5. Huntington's disease

f. prominent and early changes in personality judgment and behavior at onset of illness
   6. Diffuse Lewy Body disease

Part IV. Short Answer

47. Name 3 other causes of dementia not listed in question 46: (2 points)
   [Alzheimer's disease, Progressive supranuclear palsy, 21-hydroxylase deficiency, Alcohol, Trauma, many others]

48. What pathologic characteristic distinguishes glioblastoma multiforme from anaplastic astrocytoma? (1 point)

49. A 25 year old male college student returned to the ER for the third time in 2 days complaining of inability to walk. He was perfectly healthy until 3 weeks ago when he suffered gastroenteritis. Three days ago he noted vague tingling of his feet. Two days ago the tingling became worse and his feet seemed clumsy. He came to the ER, was told he had a virus and was discharged home. Yesterday he was so weak he could barely get out of bed and walk. The numbness of his legs had reached his thighs. Once again he came to the ER but was discharged home. He returned on the day of admission, brought by his family on a stretcher, unable to move his legs, short of breath and complaining of tingling of the arms.

On exam his vital signs were notable for a regular HR of 120, a blood pressure of 120/80, a respiratory rate of 30 and no fever. His mental status was normal. Cranial nerve exam was notable for weakness of the face bilaterally. Motor exam revealed plegic lower extremities with 3/5 strength in the arms. Sensory exam revealed only moderately impaired vibration and position sense up to the knees bilaterally. No deep tendon reflexes or plantar responses were elicitable.
49a. Are this patient’s symptoms and signs localized to the central nervous system or the peripheral nervous system? (1 point)

49b. What would you expect his muscle tone to be? (1 point) decreased

49c. Why was this man tachycardic? (1 point) autonomic nerve involvement

49d. What is the most likely diagnosis? (1 point) Guillain Barre Syndrome aka Acute Immune-mediated Demyelinating Polyneuropathy

49e. What testing might be helpful in this patient? (1 point) BMG-NCS most helpful; LP with CSF analysis to find elevated protein in absence of cells (albuminocytologic dissociation) also helpful

49f. What is the prognosis? (1 point) Most patients recover completely

49g. Is any treatment helpful in this disorder? (1 point) Plasmapheresis and IVIG double shortens course and limits respiratory dependence

49h. How might the history of gastroenteritis be important? (1 point) Many cases of GBS are preceded by viral infection implicating an autoimmune pathogenesis