

Preparing for the American Board  
of Pathology (ABPath)  
Examination of Fundamental  
Knowledge and Skills

## Neuropathology

---

*Content Specifications*

---



### 3      Overview:

#### 4                   Neuropathology Content Specifications

5      This guide outlines the content that may appear on the American Board of Pathology's Neuropathology  
6      Subspecialty exam. It provides a framework based on the knowledge and skills typically covered in  
7      Fellow-level training, along with applicable Core and Advanced Resident topics from residency training  
8      that the trainee is expected to know or be able to perform.

9                   **Key to Designations:**

10                  C = Core/Foundational Knowledge

11                  AR = Advanced Resident Knowledge

12                  F = Fellow/Advanced Practitioner Knowledge

13      The exam assesses the knowledge, judgment, skills, and abilities necessary to identify specific entities,  
14      properly process specimens, and diagnose and monitor diseases using methods common in the practice  
15      of Neuropathology. The specific diseases, tests, and concepts listed in this document are important for  
16      candidates to know, but it is not possible to create a fully comprehensive list of all the material needed  
17      for certification and effective practice. Candidates should use this guide as a reference when preparing  
18      for certification and professional practice.

19                   Table of Contents

### 20      Contents

21      1. General: Neuroanatomy, Histology, Pathologic Responses, and Diagnostic Considerations .....	2
22      2. Developmental Neuropathology .....	3
23      3. Epilepsy .....	5
24      4. Vascular Disorders .....	6
25      5. Trauma .....	7
26      6. Infections and Inflammatory Diseases.....	8
27      7. Demyelinating Diseases .....	9
28      8. Complications of Systemic Disorders.....	9
29      9. Aging and Neurodegenerative Diseases .....	11
30      10. Prion Disease.....	12
31      11. Neoplasms.....	12
32      12. Skeletal Muscle .....	17
33      13. Peripheral Nerve .....	18
34      14. Ophthalmic Pathology.....	19

35

36

## 1. General: Neuroanatomy, Histology, Pathologic Responses, and Diagnostic Considerations

### a. Neuroanatomy

- i. Neocortex, White Matter, and Entorhinal Cortex/Hippocampus C
- ii. Deep (Basal) Nuclei, Brain Stem, and Cerebellum C
- iii. Spinal Cord and Vascular Supply C
- iv. Pituitary, Pineal, and Tracts AR

### b. Cell Types

- i. Neurons, Astrocytes, Oligodendroglia, and Blood Vessels C
- ii. Ependyma, Microglia and Mononuclear Cells AR
- iii. Choroid Plexus and Meninges AR

### c. Cerebrospinal Fluid

### d. Pathologic Responses in Neurons and Axons

- i. Acute Ischemic (Hypoxic) Cell Change AR
- ii. Apoptosis AR
- iii. Axonal Degeneration/Spheroid Reaction F
- iv. Central Chromatolysis F
- v. Tract Degeneration F
- vi. Swollen / Ballooned Neurons F
- vii. Trans-synaptic Neuronal Degeneration F
- viii. Olivary Hypertrophy F
- ix. Protein Aggregation F
- x. Protein Degradation / Ubiquitin Pathway F

### e. Neuronal Nuclear Inclusions

- i. Cowdry Type A (e.g., CMV) AR
- ii. Marinesco Bodies F
- iii. Other Viral Inclusions F
- iv. Inclusions due to Neurodegenerative Disorders F

### f. Neuronal Cytoplasmic Inclusions

- i. Cytoskeleton and Filamentous Inclusions
  - 1. Neurofibrillary Tangles AR
  - 2. Hirano Bodies, Lewy Bodies, and Pick Bodies F
  - 3. Eosinophilic Thalamic Inclusions F
  - 4. Rod-Like Cytoplasmic Inclusions F
  - 5. Filamentous Inclusions F
  - 6. Motor Neuron Disease Inclusions F
- ii. Cytosolic Inclusions
  - 1. Lafora Bodies F
  - 2. Eosinophilic Inclusions in Inferior Olives F
  - 3. Storage Products in Neurometabolic Disease F
- iii. Membrane Bound Inclusions
  - 1. Lipofuscin AR

78	2. Colloid Inclusions and Bunina Bodies	F
79	3. Inclusions from the Acid Vesicle System	F
80	4. Granulovacuolar Degeneration	F
81	5. Storage Products in Neurometabolic Disease	F
82	g. Pathologic Reactions of Astrocytes	
83	i. Reactive Gliosis	AR
84	ii. Rosenthal Fibers	AR
85	iii. Corpora amylacea	AR
86	iv. Astrocyte swelling	F
87	v. Eosinophilic Granular Bodies	F
88	vi. Alzheimer's Type II Gliosis	F
89	h. Pathologic Reactions of Oligodendrocytes	
90	i. Demyelination / Remyelination	AR
91	ii. Dysmyelination	F
92	iii. Intramyelinic Vacuolization	F
93	i. Pathologic Reactions of Ependymal Cells	
94	i. Subventricular gliosis (Granular ependymitis/ependymal granulations)	F
95	ii. Subependymal rosettes / tubules	F
96	j. Pathologic Reactions of Microglia	
97	i. Microglial Activation	F
98	ii. Microglial (Babes) Nodule	F
99	k. Mineralization in the Brain	
100	i. Dystrophic Calcification (e.g., "egg shell" leptomeningeal)	F
101	ii. Secondary mineralization associated with other disorders (e.g., calcium metabolism, infection, ischemia, etc.)	F
102	l. Edema and Herniation	AR
103	m. Artifacts of Tissue Handling and Foreign Material	AR
104	n. Staining Methods and Special Microscopy	AR
105	o. Molecular Techniques	AR

108

## 109     **2. Developmental Neuropathology**

110	a. Fetal and Neonatal Hypoxic-Ischemic Lesions (General Considerations)	
111	i. Hydranencephaly, Basket Brain, and Porencephaly	F
112	ii. Schizencephaly and Multicystic Encephalopathy	F
113	iii. Acute / Subacute White Matter Lesions	
114	1. Diffuse White Matter Gliosis	F
115	2. Periventricular Leukomalacia	F
116	iv. Acute Gray Matter Lesions (General Considerations)	
117	1. Cerebral Cortical Necrosis and Pontosubicular Necrosis	F
118	2. Basal Ganglia and Thalamic Lesions	F
119	3. Cerebellar, Brain Stem, and Spinal Cord Lesions	F
120	b. Perinatal Hemorrhages	
121	i. Subependymal Germinal Plate / Matrix Hemorrhage	AR

122	ii.	Subdural, Subarachnoid, and Supial Hemorrhages	F
123	iii.	Cerebral parenchymal hemorrhage	F
124	iv.	Periventricular hemorrhagic infarct	F
125	v.	Cerebellar hemorrhage	F
126	vi.	Choroid and Intraventricular hemorrhage	F
127	c.	Chronic Lesions	
128	i.	Laminar necrosis	AR
129	ii.	Post-hemorrhagic lesions	F
130	iii.	Periventricular cysts and/or gliosis	F
131	iv.	Ulegyria	F
132	v.	Status marmoratus	F
133	vi.	Unilateral hypertrophy of the pyramidal tract	F
134	vii.	Post-hypoxic / ischemic brainstem injury	F
135	d.	Malformations	
136	i.	Defects of Neural Tube Closure	
137	1.	Anencephaly	AR
138	2.	Myelomeningocele	F
139	3.	Rachischisis	F
140	ii.	Herniation of Neural Tube through Mesodermal Defects	
141	1.	Encephalocele	AR
142	2.	Meningocele	AR
143	3.	Occult Spina Bifida	AR
144	iii.	Chiari Malformations	
145	1.	Chiari Type I Malformation	AR
146	2.	Chiari Type II (Arnold-Chiari) Malformation	F
147	3.	Chiari Type III Malformation	F
148	iv.	Disorders of Forebrain Induction	
149	1.	Holoprosencephaly (Alobar, Semilobar, Lobar)	F
150	2.	Olfactory Aplasia	F
151	3.	Atelencephaly and Aprosencephaly	F
152	4.	Agenesis of the Corpus Callosum	F
153	5.	Anomalies of the Septum Pellucidum	F
154	6.	Septo-optic Dysplasia	F
155	7.	Cavum septi pelludici and cavum vergae	F
156	v.	Neural Migration Defects	
157	1.	Agyria	F
158	2.	Lissencephaly Type I	F
159	3.	Lissencephaly Type II (Cobblestone Lissencephaly)	F
160	4.	Pachygryria	F
161	5.	Polymicrogyria	F
162	6.	Neuronal Heterotopias	F
163	7.	Cortical Microdysgenesis	F
164	8.	Focal Cortical Dysplasias, ILAE Classification	F
165	vi.	Microcephaly	F
166	vii.	Chromosomal and Single Gene Defects	

167	1.	Down Syndrome	AR
168	2.	Fragile X Syndrome	F
169	3.	Adult Polyglucosan Body Disease (GBE1 mutation)	F
170	viii.	Megalencephaly	F
171	ix.	Malformations of the Cerebellum	
172	1.	Cerebellar Agenesis	F
173	2.	Dandy-Walker Syndrome	F
174	3.	Joubert Syndrome	F
175	4.	Pontoneocerebellar Hypoplasia	F
176	5.	Cerebellar Hypoplasia in Other Contexts	F
177	6.	Granule Cell Aplasia	F
178	7.	Cerebellar Heterotopias	F
179	8.	Cerebellar Cortical Dysplasia	F
180	9.	Chiari Type I Malformation	F
181	10.	Lhermitte-Duclos Disease	F
182	x.	Malformations of the Spinal Cord	
183	1.	Tethered Cord	AR
184	2.	Diastematomyelia	F
185	3.	Syringomyelia/Hydromyelia	F
186	e.	Arthrogryposis Multiplex Congenita	F
187	f.	Dysgenetic Syndromes	
188	i.	Sturge-Weber Syndromes	AR
189	ii.	Tuberous Sclerosis (Bourneville Disease)	AR
190	iii.	Neurofibromatosis Type 1 (NF1)	AR
191	iv.	Neurofibromatosis Type 2 (NF2)	AR
192	v.	Schwannomatosis	F
193	g.	Hydrocephalus	AR
194	h.	Metabolic/Environmental/Iatrogenic Factors	
195	i.	Maternal Infection (e.g., CMV, HSV)	AR
196	ii.	Kernicterus	F
197	i.	Disorders that Primarily Affect White Matter	
198	i.	Pelizaeus-Merzbacher Disease	F
199	ii.	Canavan Disease	F
200	iii.	Alexander Disease	F
201	j.	Cerebellum	
202	i.	Menkes Disease	F
203	ii.	Ataxia-Telangiectasia	F
204	k.	Spinal Muscular Atrophy	F
205	l.	Pediatric Trauma	F
206	m.	Sudden Infant Death Syndrome	F
207			
208	<b>3. Epilepsy</b>		
209	a.	Hippocampal (Mesial Temporal) Sclerosis	AR
210	b.	Rasmussen encephalitis	F

211

212     **4. Vascular Disorders**

213	a. Adult Hypoxic and Ischemic Lesions	
214	i. Cerebral Blood Flow	C
215	ii. Hypoxic Insult	AR
216	iii. Ischemic Insult	AR
217	iv. Hypoxic-Ischemic Encephalopathy (Acute/Subacute/Chronic)	AR
218	v. Borderzone Hypoxic-Ischemic Damage	AR
219	vi. Laminar Necrosis	AR
220	vii. Hippocampal Ischemic Injury	AR
221	b. Vascular Disease and Infarcts	
222	i. Atherosclerosis	C
223	ii. Arteriolosclerosis	C
224	iii. Arterial Dissection	AR
225	iv. Hypertensive Vascular Changes	AR
226	v. Moyamoya Disease	F
227	vi. Ischemic Leukoencephalopathy	F
228	vii. Siderocalcinosis/Ferrugination of Microvessels (Fahr Disease)	F
229	viii. Binswanger Disease	F
230	ix. CADASIL	F
231	x. CARASIL	F
232	xi. COL4A1	F
233	xii. Retinocochleocerebral Vasculopathy (Susac Syndrome)	F
234	xiii. TTP	F
235	c. Angiitis and Vasculitis	
236	i. Giant Cell/Temporal Arteritis	AR
237	ii. Primary Angiitis of the CNS	F
238	iii. Secondary Angiitis due to Systemic Vasculitides	F
239	iv. Vasculitis of the Peripheral Nervous System	F
240	v. A-beta-related Angiitis (ABRA)	F
241	d. Embolic Disorders	
242	i. Non-Infectious (Atheroemboli, Air, Fat, and Iatrogenic)	C
243	ii. Infectious	AR
244	e. Cerebral Venous Thrombosis	F
245	f. CNS Infarct (Acute/Subacute/Chronic)	
246	i. Infarcts caused by Thromboembolic Occlusion of Large Arteries	AR
247	ii. Watershed or Borderzone Infarcts	AR
248	iii. Lacunar Infarcts	AR
249	iv. Hemorrhagic Infarcts	AR
250	v. Iatrogenic Infarcts	AR
251	vi. Spinal Cord Infarcts	F
252	g. Spontaneous Hemorrhage	
253	i. Spontaneous Subdural Hemorrhage	AR
254	ii. Spontaneous Subarachnoid Hemorrhage	AR

255	iii.	Hypertensive Brain Hemorrhage	AR
256	iv.	Brain Hemorrhage secondary to Systemic Disease or Therapy	AR
257	h.	Cerebral Amyloid Angiopathy	
258	i.	Age-Related	F
259	ii.	Brain Hemorrhage	F
260	iii.	A-beta-related Angiitis (ABRA)	F
261	iv.	Inherited Amyloidoses (Non-A-beta)	F
262	v.	Systemic Amyloidoses	F
263	i.	Aneurysms	
264	i.	Berry (Saccular) Aneurysms	AR
265	ii.	Infective Aneurysms	F
266	j.	Vascular Malformations	
267	i.	Arteriovenous Malformations	AR
268	ii.	Cavernous Hemangioma	AR
269	iii.	Venous Angioma	F
270	iv.	Capillary Telangiectasia	F
271	v.	Arteriovenous Fistula	F
272	vi.	Vein of Galen Malformation	F
273	k.	Miscellaneous Vascular Disorders	
274	i.	Vascular Dementia	F

275

## 276 5. Trauma

277	a.	Fractures, Skull, and Spine	F
278	b.	Craniocerebral Trauma	
279	i.	Coup Lesion	AR
280	ii.	Contra-Coup Lesion	AR
281	iii.	Traumatic Epidural Hematoma	AR
282	iv.	Traumatic Subdural Hematoma	AR
283	v.	Contusion	F
284	vi.	Laceration	F
285	vii.	Traumatic Subarachnoid Hemorrhage	F
286	viii.	Intraparenchymal (Ball/Streak) Hemorrhages	F
287	c.	Traumatic Axonal Injury	
288	i.	Brain Swelling and Raised Intracranial Pressure	AR
289	ii.	Diffuse Axonal Injury	F
290	iii.	Diffuse Vascular Injury	F
291	iv.	Missile Head Injury	F
292	v.	Blast Head Injury	F
293	d.	Sequelae of Head Injury	
294	i.	Chronic Traumatic Encephalopathy (CTE-MND)	F
295	ii.	Ischemic Damage	F
296	e.	Traumatic Spinal Cord Injury	F

297

298	<b>6. Infections and Inflammatory Diseases</b>	
299	a. Acute Viral Infections	
300	i. Herpes Simplex Virus Infections	AR
301	ii. Rabies	AR
302	iii. Aseptic Meningitis	F
303	iv. Poliomyelitis	F
304	v. Enteroviral Encephalitis	F
305	vi. Other Herpes Viral Infections (VZV, EBV, CMV)	F
306	vii. Adenovirus Infections	F
307	viii. Paramyxovirus Infections	F
308	ix. Rubella Encephalitis	F
309	x. Arbovirus Infections	F
310	xi. West Nile Encephalitis	F
311	b. Chronic and Subacute Viral Infections of the CNS	
312	i. Measles Inclusion Body Encephalitis	F
313	ii. Subacute Sclerosing Panencephalitis	F
314	iii. Progressive Multifocal Leukoencephalopathy -Includes PML and PML-IRIS associated with MS therapy	F
315	c. Human Immunodeficiency Virus Infection (HIV)	
316	i. HIV encephalitis/leukoencephalitis	F
317	ii. HIV-associated Neurologic Disease (HAND)	F
318	iii. Neurological and other Disorders Increased in HIV	F
319	iv. Therapy-Associated Disorders in Patients with HIV	F
320	d. Bacterial Infections	
321	i. Acute Bacterial Meningitis	AR
322	ii. Brain Abscess	AR
323	iii. Tuberculosis	AR
324	iv. Subdural Empyema	F
325	v. Epidural Abscess	F
326	vi. Syphilis	F
327	vii. Lyme Neuroborreliosis	F
328	viii. Whipple Disease	F
329	ix. Nocardia	F
330	e. Fungal Infections	
331	i. Aspergillosis	AR
332	ii. Mucorales Infections	AR
333	iii. Fusarium and Other Hyaline Molds	AR
334	iv. Cryptococcosis	AR
335	v. Candidiasis	AR
336	vi. Blastomycosis	AR
337	vii. Coccidioidomycosis	AR
338	viii. Histoplasmosis	AR
339	ix. Phaeohyphomycosis	F
340	f. Parasitic Infections	
341	i. Amebic Infections	
342		

343	1. Primary Amebic Meningoencephalitis	AR
344	2. Granulomatous Amebic Encephalitis	AR
345	ii. Cysticercosis and Other Cestodes	AR
346	iii. Cerebral Malaria	F
347	iv. Cerebral Toxoplasmosis	F
348	g. Other Inflammatory Diseases	
349	i. Neurosarcoidosis	AR
350	ii. Rasmussen Encephalitis	F
351	iii. Autoimmune Encephalitis	F
352	iv. Paraneoplastic Disorders	
353	1. Paraneoplastic Encephalomyelitis	F
354	2. Paraneoplastic Cerebellar Degeneration	F
355	3. Paraneoplastic Opsoclonus-Myoclonus	F
356	4. Paraneoplastic Myositis	F
357	5. Paraneoplastic Neuropathy	F
358	v. Idiopathic Hypertrophic Pachymeningitis	F
359	vi. IgG4-Related Disease	F
360	vii. Autoimmune (i.e., Lymphocytic) Hypophysitis	F
361	viii. Non-Neoplastic Pituitary Disorders	
362	1. Infectious Hypophysitis	AR
363	2. Pituitary Apoplexy	AR
364	3. Pituitary Hyperplasia	F
365	4. Autoimmune (Lymphocytic) Hypophysitis	F

366

## 7. Demyelinating Diseases

368	a. Multiple Sclerosis	
369	i. Classic (Charcot-Type) MS	AR
370	ii. Acute (Marburg-Type) MS	F
371	iii. Concentric Sclerosis (Balo)	F
372	iv. Neuromyelitis Optica (Devic Disease)	F
373	1. Aquaporin-4 in NMO	F
374	b. Other Demyelinating Diseases	
375	i. Acute Disseminated Encephalomyelitis (ADEM)	F
376	ii. Acute Hemorrhagic Leukoencephalopathy	F
377	iii. Guillain-Barré Syndrome (AIDP)	F
378	iv. Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)	F
379	v. Central Pontine Myelinolysis	F

380

## 8. Complications of Systemic Disorders

382	a. Vitamin Deficiencies	
383	i. Thiamine Deficiency and Wernicke Encephalopathy	F
384	ii. Vitamin B12 Deficiency and Subacute Combined Degeneration	F

385	iii.	Folic Acid Deficiency	F
386	b.	Systemic Metabolic Disease	
387	i.	Hypoglycemia	F
388	ii.	Hyperglycemia	F
389	c.	Disorders of Serum Electrolytes	
390	i.	Central Pontine and Extrapontine Myelinolysis	F
391	ii.	Calcium Disturbances – Siderocalcinosis/Ferrugination Microvessel (Fahr Disease)	F
393	d.	Liver Disease	
394	i.	Acquired Hepatic Encephalopathy	F
395	ii.	Hepatolenticular Degeneration (Wilson Disease)	F
396	e.	Lysosomal and Peroxisomal Disorders	
397	i.	Lysosomal Disorders	
398	1.	GM2 Gangliosidosis	F
399	2.	GM1 Gangliosidosis	F
400	3.	Ceroid Lipofuscinosis (Batten Disease)	F
401	4.	Niemann-Pick Disease, including Type C	F
402	5.	Gaucher Disease	F
403	6.	Acid Beta-Glucuronidase-Associated Parkinson Disease	F
404	7.	Mannosidosis	F
405	8.	Mucopolysaccharidosis	F
406	9.	Fabry Disease	F
407	10.	Type II Glycogenesis (Pompe Disease)	F
408	11.	Farber Disease	F
409	12.	Krabbe Disease	F
410	13.	Metachromatic Leukodystrophy	F
411	ii.	Peroxisomal Disorders	
412	1.	Zellweger Cerebroheptorectal Syndrome	F
413	2.	Adrenoleukodystrophy	F
414	f.	Mitochondrial Disorders	
415	i.	Mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like symptoms (MELAS)	F
417	ii.	Mycoclonic Epilepsy with Ragged-Red Fibers (MERRF)	F
418	iii.	Leber Hereditary Optic Neuropathy, Bilateral Striatal Necrosis, and Multiple Sclerosis-Like Mitochondrial Disease (LHON)	F
419	iv.	Neuropathy, Ataxia, and Retinitis Pigmentosa (NARP)	F
421	v.	Kearns-Sayre Syndrome (KSS) and Chronic Progressive External Ophthalmoplegia	F
423	vi.	Myoneurogastrointestinal Encephalopathy (MNGIE)	F
424	vii.	Leigh Disease	F
425	g.	Toxicity	
426	i.	Ethylene Glycol	F
427	ii.	Methanol	F
428	iii.	Toluene	F
429	iv.	Carbon Monoxide	F

430	v.	Calcineurin Inhibitors (e.g., Cyclosporin A and Tacrolimus)	F
431	vi.	Nucleoside Analogs	F
432	vii.	Phenytoin	F
433	viii.	Chloroquine/Hydroxychloroquine	F
434	ix.	Statin	F
435	x.	Cocaine	F
436	xi.	Heroin	F
437	xii.	1-Methyl-4-Phenyl Tetrahydropyridine (MPTP)	F
438	xiii.	Inhaled Solvents	F
439	h. Toxicity/Secondary Effect due to Radiation Therapy		
440	i.	Radionecrosis	F
441	ii.	Radiation-Induced Neoplasia	F

442

## 443 9. Aging and Neurodegenerative Diseases

444	i.	Aging	
445	i.	Normal Aging	F
446	ii.	Pathologic Aging	F
447	j.	Alzheimer Disease (AD)	
448	i.	AD Posterior Variant	F
449	ii.	AD with Hippocampal Sparing	F
450	iii.	AD with Cotton Wool Plaques	F
451	iv.	AD Familial Forms	
452	k.	Tauopathies, including Frontotemporal Lobar Degeneration-Tau	
453	i.	Frontotemporal Dementia-Parkinsonism Linked to Chromosome 17	F
454	ii.	Progressive Supranuclear Palsy (Steele-Richardson-Olszewski)	F
455	iii.	Corticobasal Degeneration	F
456	iv.	Argyrophilic Grain Disease	F
457	v.	Pick Disease	F
458	vi.	Parkinsonism-Dementia Complex of Guam	F
459	vii.	Postencephalitic Parkinsonism	F
460	viii.	Tangle-Only Dementia	F
461	ix.	Diffuse Neurofibrillary Tangles with Calcification	F
462	x.	Hippocampal Sclerosis Demential Tauopathy	F
463	xi.	Chronic Traumatic Encephalopathy (CTE-MND)	F
464	l.	Synucleinopathies	
465	i.	Lewy-Body Disease (LBD) Spectrum Disorders	
466	1.	LBD, Brainstem Type (Parkinson Disease)	F
467	2.	LBD, Limbic Type	F
468	3.	LBD, Neocortical Type (Dementia with Lewy Bodies)	F
469	4.	Parkinson Disease-Dementia (PD-D)	F
470	ii.	Multiple System Atrophy	F
471	m.	Hippocampal Sclerosis	F
472	n.	Trinucleotide Repeat Disorders	
473	i.	Huntington Disease	F

474	ii.	Spinocerebellar Ataxia	F
475	iii.	Friedreich Ataxia	F
476	iv.	Dentatorubral Pallidoluysian Atrophy	F
477	v.	Spinal-Bulbar Muscular Atrophy/ X-linked Bulbospinal Neuronopathy (Kennedy Disease)	F
478	vi.	Myotonic Dystrophy	F
479	vii.	Oculopharyngeal Muscular Dystrophy	F
480	viii.	Machado-Joseph Disease	F
481	o.	Frontotemporal Lobar Degeneration (FLD) with/without ALS	
482	i.	FLD with TDP-43-Immunoreactive Lesions	F
483	ii.	FLD with Neuronal Intermediate Filament Inclusions	F
484	iii.	FLD with FUS-Immunoreactive Lesions	F
485	iv.	FLD with Motor Neuron Disease	F
486	v.	Valosin-containing Protein (IBMPFD)	F
487	p.	Motor Neuron Disease	
488	i.	SMA Type I (Werdnig Hoffmann)	F
489	ii.	SMA Type II ( Intermediate)	F
490	iii.	SMA Type III (Kugelberg-Welander)	F
491	iv.	Hereditary Spastic Paraparesis	F
492	v.	Primary Lateral Sclerosis	F
493	vi.	Amyotrophic Lateral Sclerosis (ALS)	
494	1.	ALS with TDP-43	F
495	2.	ALS with TDP-Tau	F
496	3.	ALS with SOD	F
497	q.	Neuroaxonal Dystrophy	
498	i.	Neurodegeneration with Brain Iron Accumulation	F
499	ii.	Sporadic Adult-Onset Leukoencephalopathy with Neuroaxonal Spheroids (Neuroaxonal Leukodystrophy)	F
500			
501			

## 10. Prion Disease

502	a.	Sporadic Creutzfeld-Jakob Disease	AR
503	b.	Inherited Creutzfeld-Jakob Disease	F
504	i.	Gerstmann-Sträussler-Scheinker Disease	F
505	ii.	Fatal Familial/Sporadic Insomnia	F
506	iii.	Other Inherited Creutzfeld-Jakob Disease	F
507	c.	Iatrogenic Creutzfeld-Jakob Disease	F
508	d.	Protease-Sensitive Prionopathy	F
509	e.	Variant Creutzfeldt-Jakob Disease	F
510			

## 11. Neoplasms

511	a.	Adult-Type Diffuse Astrocytomas	
512	i.	Astrocytoma IDH-Mutant, CNS WHO Grade 2	AR
513	ii.	Diffuse Astrocytoma, NOS	AR
514	iii.	Astrocytoma, IDH-Mutant, CNS WHO Grade 3	AR
515	iv.	Astrocytoma, IDH-Mutant, CNS WHO Grade 4	AR
516			

517	v.	Glioblastoma, IDH-Wildtype	AR
518	vi.	Giant Cell Glioblastoma	F
519	vii.	Small Cell Glioblastoma	F
520	viii.	Gliosarcoma	F
521	ix.	Glioblastoma, Epithelioid Type	F
522	b.	Circumscribed Astrocytic Gliomas	
523	i.	Pilocytic Astrocytoma	AR
524	ii.	Pleomorphic Xanthoastrocytoma, CNS WHO Grade 2 or 3	AR
525	iii.	High-Grade Astrocytoma with Piloid Features	F
526	iv.	Pilomyxoid Astrocytoma with	F
527	v.	Chordoid Glioma	F
528	vi.	Subependymal Giant Cell Astrocytoma	F
529	vii.	Astroblastoma, MN1-Altered	F
530	c.	Oligodendroglial Tumors	
531	i.	Oligodendrogioma, IDH-Mutant and 1p/19q Co-Deleted, WHO Grade 2	AR
532	d.	Ependymal Tumors (General Considerations)	
533	i.	Subependymoma	F
534	ii.	Myxopapillary Ependymoma	F
535	iii.	Supratentorial Ependymoma, ZFTA-Fusion Positive	F
536	iv.	Supratentorial Ependymoma, YAP1-Fusion Positive	F
537	v.	Supratentorial Ependymoma, NOS	F
538	vi.	Posterior Fossa, Group A (PFA) Ependymoma	F
539	vii.	Posterior Fossa, Group B (PFB) Ependymoma	F
540	viii.	Posterior Fossa Ependymoma, NOS	F
541	ix.	Spinal Ependymoma, NOS	F
542	x.	Spinal Ependymoma, MYCN Amplified	F
543	e.	Choroid Plexus Tumors	
544	i.	Choroid Plexus Papilloma	F
545	ii.	Atypical Choroid Plexus Papilloma	F
546	iii.	Choroid Plexus Carcinoma	F
547	f.	Glioneuronal and Neuronal Tumors	
548	i.	Ganglioglioma	AR
549	ii.	Dysplastic Gangliocytoma of the Cerebellum (Lhermitte-Duclos)	F
550	iii.	Desmoplastic Infantile Astrocytoma/Ganglioglioma	F
551	iv.	Dysembryoplastic Neuroepithelial Tumor	F
552	v.	Gangliocytoma/Multinodular Vacuolating Neuronal Tumor	F
553	vi.	Central Neurocytoma	F
554	vii.	Extraventricular Neurocytoma	F
555	viii.	Cerebellar Liponeurocytoma	F
556	ix.	Papillary Glioneuronal Tumor (PGNT)	F
557	x.	Rosette-Forming Glioneuronal Tumor (RGNT)	F
558	xi.	Cauda equina Neuroendocrine Tumor	F
559	xii.	Myxoid Glioneuronal Tumor	F
560	xiii.	Diffuse Glioneuronal Tumor with Oligodendroglial Features	F

562	xiv.	Diffuse Leptomeningeal Glioneuronal Tumor	F
563	g.	Tumors of the Pinal Region	
564	i.	Pineal Parenchymal Tumors	F
565	ii.	Pineocytoma	F
566	iii.	Pineal Parenchymal Tumor of Intermediate Differentiation	F
567	iv.	Pineoblastoma	F
568	v.	Papillary Tumor of the Pineal Region	F
569	vi.	Desmoplastic Myxoid Tumor of the Pineal Region, SMARCB1-Deficient	F
570			
571	h.	Embryonal Tumors	
572	i.	Medulloblastoma, Classic Type	AR
573	ii.	Atypical Teratoid/Rhabdoid Tumor	AR
574	iii.	Medulloblastoma, Desmoplastic/Nodular Type	F
575	iv.	Medulloblastoma with Extensive Nodularity	F
576	v.	Medulloblastoma, Large Cell/Anaplastic Type	F
577	vi.	CNS Neuroblastoma, FOXR2-Activated	F
578	vii.	Embryonal Tumors with Multilayered Rosettes	F
579	viii.	CNS Tumor BCOR Internal Tandem Duplication	F
580	ix.	Embryonal Tumors with Multilayered Rosettes, C19MC-Altered	F
581	x.	Medulloblastoma, WNT-Activated	F
582	xi.	Medulloblastoma, SHH-Activated & TP53 Wild Type	F
583	xii.	Medulloblastoma, SHH-Activated & TP53 Mutant	F
584	xiii.	Medulloblastoma, Non-WHT/Non-SHH	F
585	i.	Tumors of the Cranial and Paraspinal Nerves	
586	i.	Schwannoma (Cellular and Plexiform Types)	AR
587	ii.	Neurofibroma	
588	1.	Plexiform Neurofibroma	AR
589	2.	Atypical Neurofibromatous Neoplasm of Uncertain Biologic Potential (ANNUPB)	AR
590	iii.	Ganglioneuroma	AR
591	iv.	Malignant Peripheral Nerve Sheath Tumor (MPNST)	AR
592	v.	Perineurioma	F
593	vi.	Hybrid Nerve Sheath Tumors	F
594	vii.	Epithelioid Malignant Peripheral Nerve Sheath Tumor	F
595	viii.	MPNST with Divergent Mesenchymal and/or Epithelial Differentiation or Perineural Differentiation	F
596			
597	j.	Meningothelial Tumors	
598	i.	Meningioma, NOS	AR
599	ii.	Atypical Meningioma	AR
600	iii.	Anaplastic Meningioma	AR
601	iv.	Meningioma, Meningothelial Type	F
602	v.	Meningioma, Fibrous (Fibroblastic)	F
603	vi.	Meningioma, Transitional (Mixed)	F
604	vii.	Meningioma, Psammomatous	F
605	viii.	Meningioma, Angiomatous	F
606			

607	ix.	Meningioma, Microcystic	F
608	x.	Meningioma, Secretory	F
609	xi.	Meningioma, Lymphoplasmacyte-Rich	F
610	xii.	Meningioma, Metaplastic	F
611	xiii.	Meningioma, Clear Cell Type	F
612	xiv.	Meningioma, Chordoid Type	F
613	xv.	Meningioma, Papillary Type	F
614	xvi.	Meningioma, Rhabdoid Type	F
615	xvii.	Meningioma, Other Types	F
616	xviii.	Meningoangiomatosis	F
617	k.	Mesenchymal (Non-Meningothelial) Tumors	
618	i.	Chordoma	AR
619	ii.	Hemangioblastoma	AR
620	iii.	Lipoma	F
621	iv.	Solitary Fibrous Tumor	F
622	v.	Chondrosarcoma	F
623	vi.	Mesenchymal Chondrosarcoma	F
624	vii.	Hemangioma	F
625	viii.	Intracranial Mesenchymal Tumor FET::CREB Fusion-Positive	F
626	ix.	CIC Rearranged Sarcoma	F
627	x.	Primary Intracranial Sarcoma DICER1-Mutant	F
628	xi.	Ewing Sarcoma	F
629	l.	Primary Melanocytic Tumors	
630	i.	Melanocytosis	F
631	ii.	Melanocytoma	F
632	iii.	Melanoma	F
633	iv.	Melanomatosis	F
634	m.	Lymphoma and Hematopoietic Tumors of the Central Nervous System	
635	i.	Diffuse Large B-cell Lymphoma of the CNS	F
636	ii.	Langerhans Histiocytosis	F
637	iii.	Non-Langerhans Histiocytosis (Rosai-Dorfman)	F
638	iv.	Immunodeficiency-Associated CNS Lymphomas	F
639	v.	Intravascular Large B-cell Lymphoma	F
640	vi.	MALT Lymphomas of the Dura	F
641	n.	Tumors of the Sellar Region	
642	i.	Rathke Cleft Cyst	AR
643	ii.	Craniopharyngioma, Adamantinomatous Type	AR
644	iii.	Craniopharyngioma, Papillary Type	AR
645	iv.	Pituitary Adenoma/Pituitary Neuroendocrine Tumor (PitNET)	AR
646	1.	Densely Granulated Corticotroph Adenoma	F
647	2.	Sparingly Granulated Corticotroph Adenoma	F
648	3.	Crooke Cell Adenoma	F
649	4.	Densely Granulated Somatotroph Adenoma	F
650	5.	Sparingly Granulated Somatotroph Adenoma	F
651	6.	Mammosomatotroph Adenoma	F

652	7. Mixed Somatotroph-Lactotroph Adenoma	F
653	8. Sparsely Granulated Lactotroph Adenoma	F
654	9. Densely Granulated Lactotroph Adenoma	F
655	10. Acidophil Stem Cell Adenoma	F
656	11. Thyrotroph Adenoma	F
657	12. Gonadotroph Adenoma	F
658	13. Null Cell Adenoma	F
659	14. Plurihormonal PIT1-Positive Adenoma	F
660	v. Pituitary Carcinoma/ Metastatic PitNET	F
661	vi. Pituitary Hyperplasia	F
662	vii. Hypothalamic Hamartoma	F
663	viii. Granular Cell Tumor	F
664	ix. Pituicytoma	F
665	x. Spindle Cell Oncocytoma	F
666	o. Germ Cell Tumors	
667	i. Germinoma	AR
668	ii. Embryonal Carcinoma	AR
669	iii. Yolk Sac Tumor	AR
670	iv. Choriocarcinoma	AR
671	v. Teratoma, Mature	AR
672	vi. Teratoma, Immature	AR
673	vii. Teratoma with Malignant Transformation	AR
674	viii. Malignant Mixed Germ Cell Tumor	AR
675	(Specify Components: Germinoma, Embryonal, etc.)	
676	p. Cysts	
677	i. Epidermoid and Dermoid Cysts	AR
678	ii. Colloid Cyst of the Third Ventricle	AR
679	iii. Endodermal/Enterogenous Cyst	AR
680	iv. Arachnoid Cyst	AR
681	v. Ependymal Cyst	F
682	vi. Pineal Cyst	F
683	q. Metastatic Tumors	
684	i. Neoplasms in Tissues Surrounding the CNS	AR
685	ii. Secondary Neoplasms in the Meninges	AR
686	iii. Secondary Neoplasms in the Brain and Spinal Cord	AR
687	r. Paraneoplastic Disorders	
688	i. Paraneoplastic Encephalomyelitis	F
689	ii. Paraneoplastic Cerebellar Degeneration	F
690	iii. Paraneoplastic Opsoclonus-Myoclonus-Ataxia	F
691	s. Familial Tumor Predisposition Syndromes	
692	i. Tuberous Sclerosis	AR
693	ii. Neurofibromatosis, Type 1 (NF1)	AR
694	iii. Neurofibromatosis, Type 2 (NF2)	AR
695	iv. von Hippel-Lindau Disease	AR
696	v. Li-Fraumeni Syndrome	AR

697	vi.	Cowden Disease	AR
698	vii.	Lynch Syndrome	AR
699	viii.	Nevvoid Basal Carcinoma Syndrome	AR
700	ix.	Rhabdoid Tumor Predisposition Syndrome	AR
701	x.	Carney Complex	AR
702	xi.	Familial Adenomatous Polyposis	AR
703	xii.	DICER1 Syndrome	AR
704	xiii.	Familial Retinoblastoma	AR
705	xiv.	BAP1 Tumor Predisposition Syndrome	AR
706	xv.	Constitutional Mismatch Repair Deficiency (CMMRD) Syndrome	F
707	xvi.	Melanoma Astrocytoma Syndrome	F
708	xvii.	Fanconi Anemia	F
709	xviii.	ELP1 Medulloblastoma Syndrome	F
710	t.	Pediatric-Type Diffuse Low-Grade Gliomas	
711	i.	Diffuse Astrocytoma, MYB or MYB-1 altered	F
712	ii.	Angiocentric Glioma	F
713	iii.	Polymorphous Low-Grade Neuroepithelial Tumor of the Young	F
714	iv.	Diffuse Low-Grade Glioma, MAPK Pathway Altered	F
715	u.	Pediatric-Type Diffuse High-Grade Gliomas	
716	i.	Diffuse Midline Glioma, H3 K27-Altered	F
717	ii.	Diffuse Hemispheric Glioma, H3 G34-Mutant	F
718	iii.	Diffuse Pediatric-Type High-Grade Glioma, H3-Wild Type and 1. IDH-Wildtype	F
719	iv.	Infant-Type Hemispheric Glioma	F

721

## 12. Skeletal Muscle

722	v.	Muscle Biopsy	
723	i.	Handling	AR
724	ii.	Muscle Biopsy Techniques, IHC/Special Stains	F
725	iii.	Skeletal Muscle Ultrastructure	F
726	w.	Myopathic Processes	
727	i.	Myopathic Histopathologic Features	F
728	x.	Inflammatory Myopathies	
729	i.	Dermatomyositis	F
730	ii.	Inclusion Body Myositis	F
731	iii.	Sarcoid Myopathy	F
732	iv.	Bacterial Myositis	F
733	v.	Viral Myositis	F
734	vi.	Parasitic Myositis	F
735	vii.	Fungal Myositis	F
736	viii.	Vasculitis	F
737	ix.	Paraneoplastic Myositis	F
738	y.	Metabolic Diseases	
739	i.	Glycogenoses	F

741	1. Type II (Acid Maltase Deficiency/Pompe Disease)	F
742	2. Type V (Myophosphorylase Deficiency/McArdle Disease)	
743	ii. Mitochondrial Myopathy	F
744	iii. Lipid Storage Myopathies	F
745	z. Channelopathies	F
746	aa. Toxic Myopathies	
747	i. Steroid Myopathy/ Acute Care Myopathy	F
748	ii. Chloroquine / Hydroxychloroquine Vacuolar Myopathy	F
749	bb. Muscular Dystrophies	
750	i. Dystrophinopathies	F
751	ii. Emery-Dreifuss Muscular Dystrophy	F
752	iii. Facioscapulohumeral Dystrophy (FSHD)	F
753	iv. Myotonic Dystrophy	F
754	v. Oculopharyngeal Muscular Dystrophy	F
755	vi. Limb-Girdle Muscular Dystrophy	F
756	vii. Congenital Muscular Dystrophy	F
757	cc. Autophagic Vacuolar Myopathies	F
758	dd. Myofibrillar Myopathies	F
759	ee. Distal Myopathies, including Valsoin-Containing Protein (IBMPFD)	F
760	ff. Congenital Myopathies	
761	i. Nemaline Myopathies	F
762	ii. Core Myopathies	F
763	iii. Centronuclear Myopathies	F
764	iv. Congenital Fiber Type Disproportion Myopathies	F
765	gg. Neuropathic (Neurogenic) Diseases	
766	i. Neuropathic Histopathologic Features	F
767	ii. Motor Neuron Diseases, Spinal Muscular Atrophy	F
768		

### 13. Peripheral Nerve

769	a. Peripheral Nerve Biopsy	
770	i. Handling	AR
771	b. Peripheral Nerve Ultrastructure	F
772	c. Major Pathologic Processes	
773	ii. Axonal (Wallerian) Degeneration	F
774	iii. Distal ("Dying Back") Axonopathy	F
775	iv. Demyelination (Segmental)	F
776	d. Traumatic Neuroma	AR
777	e. Inflammatory Neuropathies	
778	i. Vasculitic Neuropathy	AR
779	ii. Sarcoid Neuropathy	AR
780	iii. Guillain-Barré Syndrome (AIDP)	F
781	iv. Chronic Inflammatory Demyelinating Polyradiculopathy (CIDP)	F
782	v. Paraneoplastic Neuropathy	F
783	f. Infectious Neuropathies	
784		

785	i. Leprosy	F
786	g. Hereditary Motor and Sensory Neuropathies (HMSN)	F
787	h. Nutritional Deficiency/Toxic Neuropathies	F
788	i. Neuropathies Associated with Systemic Diseases	
789	i. Diabetic Neuropathy	AR
790	ii. Amyloid Neuropathy	AR

791

#### **14. Ophthalmic Pathology**

792	a. Diseases of the Orbit	F
793	b. Diseases of the Conjunctiva	F
794	c. Diseases of the Cornea	F
795	d. Diseases of the Anterior Segment	F
796	e. Diseases of the Uvea	F
797	f. Retina and Vitreous	
798	i. Retinal Neoplasms	
799	1. Retinoblastoma	AR
800	2. Melanoma	AR
801	ii. Retinal Detachment	F
802	iii. Retinal Vascular Disease	F
803	iv. Age-Related Macular Degeneration	F
804	v. Other Retinal Degeneration	F
805	vi. Retinitis	F
806	g. Diseases of the Optic Nerve	F
807	h. Phthisis Bulbi	F
808	i. Therapeutic or Iatrogenic Complications	F