Content Specifications are intended to guide diplomates in selecting their MOC Part II and Part IV activities. They are not intended to be study guides for the MOC Part III exam, although the exams may include some questions on topics and references included in the Content Specifications. Please refer to the MOC Study Guides for exam preparation.

New Validated Practical Knowledge

I. Normal Development, Structure, and Function
   Role of specific genes in brain development

II. Congenital Diseases of the Nervous System
   Identification of teratogens causing CNS malformations
   Periventricular heterotopia

III. Degenerative and Demyelinating Diseases
   Frontotemporal degeneration (FTLD)
   - Categorization of FTLD by protein deposition
   - FTLD-chromosome 17 with tau-positive inclusions
   - FTLD with TDP-43-positive inclusions
   - FTLD-U with ubiquitin-positive/tau negative inclusions
   - FTLD negative for both tau and ubiquitin inclusions
   - Consensus criteria for FTLD
   Clinical and pathologic overlap between FTLD and ALS
   Third consensus conference for the diagnosis of Lewy body disorders
   Utility of new marks (tau, alpha-synuclein, TDP-43) in neurodegenerative diseases
   Inflammation and vascular aspects of Alzheimer disease
   New concepts in multiple sclerosis including demyelination in grey matter
   NMO (Devic disease) new concepts
   - Role of aquaporin 4
   - NMO serum testing
   Abeta-related-angiitis (ABRA)
   CADASIL

IV. Infectious and Inflammatory Diseases
   New concepts about prion disease
   - Use of western blot and immunohistochemistry
   Utility of in-situ hybridization for identification/confirmation of infectious agents
   - PML
   - HSV
Etc.

V. Trauma
Amyloid precursor protein in head injury
Chronic traumatic encephalopathy

VI. Cerebrovascular Disease
Distribution of arterial supply

VII. Neoplasm
Use of molecular testing for prognosis and/or treatment in specific brain tumors
- IDH1/IDH2/hTERT mutations
- oligodendrogliomas - 1p/19q
- INI-1/snf5 in AT/RT
- EGFR amplification in malignant gliomas
- PTEN loss in high grade astrocytomas
- Decreased MGMT expression in high grade astrocytomas
- Aberrant beta catenin and impaired p63 in craniopharyngiomas

Newly described neoplasms
- Pituicytoma
- Chordoid glioma
- Liponeurocytoma
- Extraventricular neurocytoma
- Desmoplastic infantile ganglioglioma
- Chordoid meningioma
- Papillary ganglioneurocytoma
- Dysembryoplastic neuroepithelial tumor
- PXA with anaplastic features
- Atypical teratoid/rhabdoid tumor
- Medulloblastoma variants
- Rosetteglioneuronal tumor
- Embryonal tumor with abundant neuropil and true rosettes (ETANTR)

Meningioma variants
- EBV in situ hybridization for lymphomas and disorders in immunosuppressed patients
- DNA repair enzymes involved in therapeutic resistance
  - decreased MGMT expression in high grade astrocytomas

VIII. Neuromuscular
Classification of Molecular Muscle Pathology
- Genetic classification of myopathies
  - Congenital myopathies
  - Muscular dystrophies
  - Distal myopathies
Myofibrillar myopathies
Mitochondrial myopathies
Autophagic vacuolar myopathies

Muscle biopsy evaluation to assist in molecular diagnosis of genetic myopathies
Use of combined histochemistry, immunostaining, and electron microscopy to identify structural abnormalities in congenital myopathies
Use of combined histochemistry, immunostaining, and electron microscopy to identify myofibrillary myopathies and define the abnormal protein
Use of immunostaining to identify muscular dystrophy subtypes

IX. Other
Genetic markers associated with epilepsy

REFERENCES


ONLINE REFERENCES

1. GeneTable (http://www.musclegenetable.org) – a nearly comprehensive list of genetic neuromuscular diseases maintained by the World Muscle Society (NOTE: on 6/20/12 this site was still under construction)
3. Leiden Muscular Dystrophy pages (http://www.dmd.nl/) – a registry of muscular dystrophy and some congenital myopathy gene mutations