

POLYNEUROPATHY + NEURONOPATHY: Points in Differential Diagnosis

1. Selective Functional Involvement

<p>Motor Motor Neuropathy: Multifocal (MMN); Distal Motor Neuron Disorders Lower Motor Neuron (LMN): BVVL; Fazio-Londe; Madras Spinal Muscular Atrophies (SMA): 5q; BSMA; Infantile; HMN ALS: Sporadic; Hereditary Toxic: Pb; Dapsone; Botulism; Tick Acute: Porphyria; Axonal GBS (AMAN); Polio; West Nile Diabetic amyotrophy</p>	<p>Autonomic Diabetes Mellitus; Amyloid; Paraneoplastic Immune: Acute; GBS; Vasculitis Multisystem: Shy-Drager; IOH; Mitochondrial Hereditary: Riley-Day; Sensory PN; HSN II Infections: Chagas; Leprosy; HIV Toxic: Vacor; Perhexiline; Vincristine Hyperhidrosis: Arsenic; Hg; Acrylamide Localized: RSD; Horner's</p>	<p>Treatable neuropathies Diabetes; Vasculitis; CIDP; Lymphoma; Infection; Toxic; Compression Deficiency: B₁₂; E; Cu Antibody: GM1; Sulfatide; MAG; GALOP; Cryoglobulin; M-protein</p>
<p>Sensory: Large fiber & Ataxic Tabes dorsalis; Vitamin deficiency: B12; E Toxic: Vitamin B6; cis-platinum; Taxol; MeHg Hereditary: an-α-lipoproteinemia; Friedreich's; Ataxia telangiectasia; Biemond; OPCA; HSN Immune: MAG; GALOP; GD1b; Miller-Fisher Neuronopathies: Hu; Sjögren's Rule out: Myelopathy; HTLV1; HIV</p>	<p>Sensory: Small fiber Leprosy; Amyloid; Idiopathic Toxic: Kepone; Ciguatera Metabolic: Diabetes; Increased Triglycerides Hereditary HSN I & IV; Tangier's, Fabry's; Indifference; Thermoanalgesia; Navajo HSN + Paraparesis (Cavanagh)</p>	<p>Neuropathy + Pain Diabetic amyotrophy; Vasculitis; Idiopathic; Polio; HIV; Ca meningitis; Toxic: EtOH; Arsenic; Ifosfamide cis-Platinum; Thallium; Thalidomide Hereditary: Fabry's; HSN I; Erythromalgia Focal: Median & Post tibial n. Other: Roots; Plexus</p>

2. Selective Anatomical Distribution: Most are symmetric and maximal distally in the lower extremities

<p>Extraocular muscle Botulism Diabetes Miller-Fisher Diphtheria Rule out: MG; Myopathy</p>	<p>Proximal Motor Immune Demyelinating: GBS; CIDP SMA; Porphyria Plexopathy: Brachial; Lumbar Rule out: Joint pain; Myopathy</p>	<p>Proximal Sensory Hereditary: Porphyria; Tangier Neuronopathy: Hu; Sjögren's Thoracic neuropathy Rule out: Myelopathy</p> <p>Skin temperature-related Leprosy</p>	<p>Upper extremity Immune: MMN; Vasculitis CIDP variant Amyloid: Carpal tunnel Entrapment: HNPP; Other Lead ALS; LMN Rule out: Spinal; CNS</p>	<p>Asymmetric Mononeuritis multiplex Neuronopathy: ALS; Sensory Entrapments Plexopathies Toxic</p>
<p>CNS Spinal: Organophosphate; Hexacarbon; AMN; MLD; Lymphoma; Cuban; Vernant's Optic: Disulfiram; CS₂; Hg; Drugs; NARP; CMT6; Post col & RP; Cuban; Vernant's Hearing loss: HMSN X, 1A, 1B, 4D, 6; Mitochondrial; Sarcoid Cerebellum: FA; AT; MLD; Refsum; A-β-lipoproteinemia; SCA 2, 3, 4; IOSCA; Hu & CV-2 Supratentorial: Mitochondrial; Thyroid; Hu; B12; Vasculitis; Neoplastic; Sarcoid Infection: Lyme; HIV; Rabies; Syphilis; West Nile Hereditary: Polyglucosan; Fabry; HexA; Porphyria; Prion; ALS; Cowchock; NAD; Krabbe; MLD</p>		<p>Face Bell's Palsy Melkersson; Tangier Polyradiculopathies: Sarcoid; Lyme; GBS Motor neuron disorders: ALS; Kennedy's; Möbius Rule out: MG; Myopathy</p>		

3. Time Course

<p>Acute Immune: GBS (Demyel & Axon); Vasculitis Toxic: Botulism; Thallium; Vacor; Drugs; Org-phos Infections: Tick; Lyme; HIV; Leptospirosis; Rabies; CMV; West Nile Porphyria; Paraneoplastic (Anti-Hu) Rule out: Myopathy; MG; Spinal</p>	<p>Hereditary CMT Demyel Dom: IA PMP-22, 17p11; IB & IE P0, 1q22; IC LITAF, 16p13; ID EGR2, 10q21; IF NFL 8p21 HNPP PMP-22, 17p11; Thermosensitive; SOX10 22q13; DNM2 19p12; YARS 1p35; 10q24 III (PMP-22; P0; EGR2); CMT + Intermid NCV: 19p12; 10q24; CMT2-P0; CMT-X (Semi-dominant) CMT Demyel Rec: III; 4A GDA P1 8q21; 4B MTMR2 11q22; 4B2 SBF2 11p15; 4C SH3TC2 5q23; 4D NDRG1 8q24 4E EGR2 10q21; 4F Prx 19q13; 4G HK1 10q22; 4H FGD4 12q12; 4J FIG4 6q21; CCFDN CTD1P1 18qter CMT X-linked: Connexin-32 Xq13; Xp22.2; Xq26; Cowchock Xq24; PRPS1 Xq22; AUNX1 Xq23 CMT Axonal Dominant: 2A KIF1B or MFN2, 1p36; 2B RAB7, 3q; 2C 12q23; 2D GARS, 7p14; 2E NFL, 8p21; 2F HSPB1 7q11; 2G 12q12; 2L: HSPB8 12q24; DNM2 19p12; HMSN-P 3p14; 5; 6; CNS; HNA1 SEPT9 17q25 CMT Axonal Rec (AR-CMT): 2A Lamin A/C 1q21; 2B 19q13; 2K GDAP1 8q21; Cowchock; P0 CMT + CNS: Andermann KCC3 15q13; Deafness; Optic Δ; NAD; GAN Gigaxonin, 16q24; AR-CMT2 + Pyramidal 8q21; Lethal Neonatal; TDP1 14q31 Myelin, Recessive: Cockayne's; Refsum (PAO & Peroxin-1); Krabbe; MLD; CMT 4E; P0; EGR-2 ALS: AD SOD 21q22; SETX 9q34; VAPB 20q13; ANG 14q11; NFH 22q12; TDP-43 1p36; FIG4 6q21; FUS; 18q21, 16q, 20p AR NF-H; Alsin 2q33; PRPH 12q12; FUS 16p11; 15q15; ALS-FTD CHMP2B 2p11 SMA: Androgen receptor Xq12; SMN 5q12; Infant Xp11; Congenital 12q23; P 3q13 HMN: HSPB8 12q; GARS 7p; BSCL2 11q; 2q; 7q; 9p; Xq13; DCTN1 2p; SETX 9q; PLEKHG5 1p; HSPB1 7q; IGHMBP2 11q Sensory neuropathy, Dominant: HSN1 SPTLC1 9q22; Sens + Deaf Connexin-31 1p35; ARHGEF10 8p23 Sensory neuropathy, Recessive: HSN2 HSN2 12p13; HSN3 (Riley-Day) IKBKAP 9q31; Tangier; SCN9A 2q24 HSN4 NTRK1 1q21; HSN5; NGF-β 1p13; ATM 11q22; FA FRDA 9q13; Fabry Xq22; DHH 12q12; NARP; 5p15; 3p22</p>
<p>Relapsing CIDP \pm GBS Hereditary: Porphyria; HNPP; HNA; Thermosensitive; Refsum</p>	
<p>Childhood Onset Immune: CIDP; GBS Hereditary: NAD; HMSN I & III; MLD Giant Axonal; Lethal neonatal; Tangier CCFDN; Congen Hypomyelin; Krabbe</p>	

4. Other Features

<p>Paraneoplastic Myopathy: Necrotizing; Dermatomyositis; Type II atrophy; Decorin Antibody; Rippling muscle; Metabolic; Scleromyxedema NMJ: MG (Thymoma); LEMS (Small cell Ca) Nerve: Hu; Motor neuronopathy; Isaac's; M-protein Neuromyopathy (wt loss > 15%); Autonomic (Hu) Spinal: Necrotizing; Stiffman (Breast +) Cerebellar: Yo (Gyn); Tr (Lymphoma); Hu; LEMS; Ma Opsoclonus/Myoclonus - Child (Neuroblastoma), Adult, Ri Encephalopathy: Hu; Thymoma; Ma2 (Ta)</p>	<p>GI disorders Neuropathy Hereditary: Hirschsprung; MEN; Pseudoobstruction Infection: Polio; Chagas MNGIE; Diabetes; Amyloid; Hu; POEMS; Porphyria Vitamin \downarrow: B₁₂; E Myopathy: Inflammatory; \uparrow Thyroid; Visceral Dystrophy: Myotonic; Duchenne; Oculopharyngeal; Oculopharyngodistal Spinal: Syringomyelia</p>	<p>Large nerves Leprosy; CIDP; Amyloid; Acromegaly; Entrapment Hereditary: Refsum; HMSN I & III; Noonan Neoplasm: Focal; Diffuse: Lymphoma Local: Entrapment; Tumors; Mononeuropathy</p>	
<p>Demyelination Immune: GBS; CIDP; MMN; MAG; Sulfatide; GALOP; POEMS Toxic: Amiodarone; Perhexiline; Diphtheria; Na cyanate; Buckthorn Hereditary: HMSN I, III, IV; HNPP; Krabbe; MLD; Refsum Mitochondrial: MNGIE; NARP</p>	<p>Spontaneous Activity Complex repetitive Dystonia Fasciculations ALS; MMN; Normal Hyperekplexia (Startle) Myokymia Isaac's; Radiation Neuromyotonia Myotonia Stiffman Syndrome Tetanus</p>	<p>Antibodies: For M-protein use immunofixation IgM vs GM1 or GalNAc-GD1a: Motor neuropathy IgG vs GM1, GM1b, GalNAc-GD1a: AMAN IgM vs GM2: Sensory > Motor; Demyelinating IgM vs GD1b: Sensory; Axonal; CANOMAD IgM vs TS-HDS: Sensory; Pain; Axonal IgG vs GQ1b: Miller-Fisher; Ophthalmoplegia IgM vs MAG: Sensory > Motor; Ataxia; Tremor IgM vs Sulfatide or GALOP: Sensory; Gait Δ IgG vs Hu: Sensory neuron IgM vs Tubulin: CIDP; Asymmetric IgG vs Yo, Tr, Hu: Cerebellar Cryoglobulins: Mononeuritis multiplex</p>	<p>Nerve biopsy: \uparrow Utility Asymmetric PN Age > 65 + Disabled Sensory loss Abnormal NCV Diagnoses Vasculitis; \pm CIDP Systemic: Leprosy; Amyloid; Sarcoid Hereditary: MLD Giant axonal; Fabry; Krabbe</p>