The "Other Neuro Diagnoses" Coffee Club:

Polyneuropathies, Myopathies, and West Nile Virus Encephalitis

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Objectives and Purpose

Abstract/Purpose Statement: The purpose of this presentation is providing the listener with the necessary information to recognize and understand course and treatment of acute and chronic demyelinating polyneuropathy, immune mediated myopathy and West Nile encephalopathy.

Objectives:
1) Upon completion of this program, the participant should be able to: Recognize signs of acute demyelinating polyneuropathy and understand the difference between acute and chronic disease processes
2) Recognize and understand disease and treatment course of necrotizing autoimmune myopathy
3) Recognize and understand disease and treatment course of West Nile encephalopathy

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  - April 2019 – On With Life

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Imagine this:

- You're feeling spunky and decide to go for that gas station sushi
- Within 24 hours you're strongly regretting that decision and worshiping the porcelain god (or goddess)
- Over the course of the next week, you notice progressive weakness of your legs which eventually effects your ability to ambulate
- You start to notice difficulty breathing and present to the nearest ER

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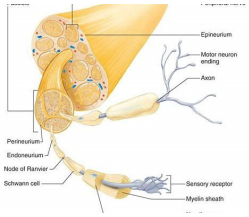
Acute Immune-mediated Polyneuropathies

Also known as Guillain-Barre Syndrome

- Immune response to a preceding infection
- Antibodies cross react with components of the peripheral nerves – myelin sheath or axons



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Antecedent Events

Infection
- Campylobacter jejuni
- Cytomegalovirus
- Epstein-Barr Virus
- HIV
- Zika Virus

Other Triggers
- Surgery
- Immunizations
- Trauma
- Bone-marrow transplantation

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Epidemiology

"If you hear hooves in Iowa, you think horses not zebras"

- It's a zebra
  - 1-2 cases/100,000 per year
  - Incidence increases 20% with every 10-year increase in age beyond the first decade of life
  - Males > females

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Clinical features

- Progressive, symmetric muscle weakness
  - Usually starts in legs
  - Starts in arms or face (10%)
  - Respiratory weakness – 10-30% require ventilator support
  - Facial nerve palsies (50%)
  - Oropharyngeal weakness (50%)
  - Oculomotor weakness (15%)
- Absent/Depressed deep tendon reflexes (90-100%)

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Clinical features continued

- Parasthesias of the hands and feet (80%)
- Pain due to nerve root inflammation (66%)
  - Back and extremities
- Dysautonomia
  - Diarrhea/constipation
  - Hyonatremia (including SIADH)
  - Bradycardia/Tachycardia
  - Urinary retention

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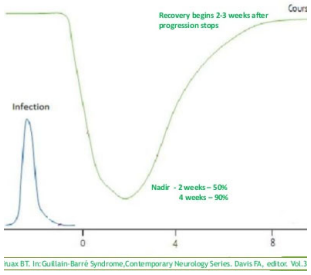
Disease Progression

- Progresses over about a 2-week period
- 90% have reached nadir of disease by 4 weeks
- Progression for more than 4 weeks is consistent with chronic form of demyelination polyneuropathy (more to come)



Kuax BT. In:Guillain-Barré Syndrome,Contemporary Neurology Series. Davis FA, editor. Vol.1

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Diagnosis:
- Clinical features
- CSF evaluation
  - Elevated protein with normal WBC
- EMG/NCV
- Antibody testing
- MRI
  - Thickening of spinal nerve roots

Brighton criteria for case definition of Guillain-Barrè syndrome

Level 1 of diagnostic certainty	Level 2 of diagnostic certainty	Level 3 of diagnostic certainty
▪ Bilateral and flaccid weakness of the limbs; **AND** ▪ Decreased or absent deep tendon reflexes in weak limbs; **AND** ▪ Monophasic illness pattern; and interval between onset and nadir of weakness between 12 hours and 28 days; and subsequent clinical plateau; **AND** ▪ Absence of identified alternative diagnosis for weakness; **AND** ▪ Cytoalbuminologic dissociation (ie, elevation of CSF protein level above laboratory normal value and CSF total white cell count <50 cells/microL; **AND** ▪ Electrophysiologic findings consistent with GBS	▪ Bilateral and flaccid weakness of the limbs; **AND** ▪ Decreased or absent deep tendon reflexes in weak limbs; **AND** ▪ Monophasic illness pattern; and interval between onset and nadir of weakness between 12 hours and 28 days; and subsequent clinical plateau; **AND** ▪ Absence of identified alternative diagnosis for weakness; **AND** ▪ CSF total white cell count <50 cells/microL (with or without CSF protein elevation above laboratory normal value); **OR** electrophysiologic studies consistent with GBS if CSF not collected or results not available	▪ Bilateral and flaccid weakness of the limbs; **AND** ▪ Decreased or absent deep tendon reflexes in weak limbs; **AND** ▪ Monophasic illness pattern; and interval between onset and nadir of weakness between 12 hours and 28 days; **AND** ▪ Absence of identified alternative diagnosis for weakness

CSF: cerebrospinal fluid; GBS: Guillain-Barré syndrome.

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Differential diagnosis of Guillain-Barré syndrome

Differential diagnosis
Cerebral
Acute disseminated encephalomyelitis (ADEM)
Bilateral strokes
Psychogenic symptoms
Cerebellar
Acute cerebellar ataxia syndrome
Posterior fossa structural lesion
Spinal
Anterior spinal artery syndrome
Compressive myelopathy
Neuromyelitis optica
Poliomyelitis
Transverse myelitis
Other infectious causes of acute myelitis (eg, West Nile virus, coxsackieviruses, echoviruses)
Peripheral nervous system
Acute onset chronic inflammatory demyelinating neuropathy (CIDP)
Critical illness polyneuropathy
Cytomegalovirus-related radiculitis
Diphtheria
HIV-related radiculitis
Leptomeningeal malignancy
Lyme disease
Metabolic and electrolyte disorders (eg, hypoglycemia, hypophosphatemia)
Porphyria
Thiamine deficiency (beriberi)
Tick paralysis
Toxic neuropathy
Vasculitis
Neuromuscular junction
Botulism
Myasthenia gravis
Neuromuscular blocking agents
Muscle disease
Acute inflammatory myopathies (eg, dermatomyositis, polymyositis)
Acute viral myositis
Acute rhabdomyolysis
Critical illness myopathy
Metabolic myopathies (eg, hypokalemic, hyperkalemic)
Mitochondrial myopathies
Periodic paralyses

Data from:
1. Evans OB. Guillain-Barré syndrome in children. Pediatr Rev 1986; 8:69.
2. Jones HR. Childhood Guillain-Barré syndrome: clinical presentation, diagnosis, and therapy. J Child Neurol 1996; 11:4.
3. Yuki N, Hartung HP. Guillain-Barré syndrome. N Engl J Med 2012; 366:2294.
4. Willison HJ, Jacobs BC, van Doorn PA. Guillain-Barré syndrome. Lancet 2016; 388:717.

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GBS Variants

Acute inflammatory demyelinating polyneuropathy	Acute motor axonal neuropathy	Acute motor and sensory axonal neuropathy	Miller Fisher Syndrome
• 85-90% US cases • Classical presentation • Myelin is targeted at nerve roots • Widespread leading to paralysis • Remyelination occurs over weeks to months	• Frequent in Asia • More frequent in summer • Deep tendon reflexes preserved • Selective involvement of motor neurons • Axonal involvement • Worse prognosis than GBS	• More severe • Motor and sensory nerves	• Ophthalmoplegia with ataxia and areflexia • ¼ develop extremity weakness • Antibodies to ganglioside nerve component

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Treatment Approach

Initial stabilization

- Respiratory, cardiac and hemodynamic monitoring
  - up to 30% require ventilator support
    - Onset to admission <7 days
    - Inability to cough, stand, lift head or arms, elevated liver enzymes
  - Cardiovascular management of dysautonomia
    - Hypo/hypertension
    - arrhythmias
- DVT prophylaxis, bowel and bladder care, rehabilitation
- Symptomatic treatments for pain
  - Gabapentin/carbamazepine
  - Opiate and non-opiate analgesics

Disease-Modifying Approaches

- Plasmapheresis
  - Removes circulating antibodies, complement and biological response modifiers
  - Earlier improvement in muscle strength, decreased ventilator needs and better recovery
  - Most effective when started within 7 days of symptom onset
  - Complications: hypotension, sepsis, IV access issues
- IVIG
  - Providing anti-idiotypic antibodies
  - As effective as plasmapheresis
  - Adherence may be better
  - Side effects: aseptic meningitis, rash, acute renal failure, hyperviscosity

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Clinical course

Continued progression for about 2 weeks

Plateau Phase for 2-4 weeks

Recovery of function

Time from onset to recovery shortened by 40-50% with plasma exchange or IVIG

19-49 days to show improvement with treatment

53-85 days to walk unaided following treatment

Relapses with increased weakness occur in up to 10%

Confirm diagnosis to rule out error

Usually not as severe as initial symptoms

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Prognosis

- 80-84% walk independently at 6-12 months
- 60% have full recovery of motor strength at 12 months
- 5-10% have prolonged course with several months of ventilator support
  - 3-7% will die
  - 20% of those ventilator dependent will die
- Factors associated with poorer prognosis
  - Older age
  - Rapid onset
  - Severe muscle weakness on admission
  - Need for ventilatory support
  - More severe EMG findings
    - Axonal degeneration
    - Markedly reduced distal motor responses
  - Preceding diarrheal illness
- 2-5% will develop chronic inflammatory demyelination polyneuropathy

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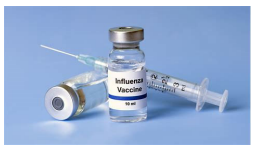
Immunization associated GBS



- 1-2 excess GBS cases per 1,000,000 vaccinated
  - Risk much less than that of disease
- Recommendations for those with immunization associated GBS
  - Do not vaccinate in the 1st year after diagnosis
  - Future avoidance of the particular vaccine that triggered GBS
  - After 1-year, other immunizations need not be withheld

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Imagine this:

- You notice gradual, unexplained weight loss over the course of 2-3 months
- Eventually, you notice weakness and parasthesias of your hands and feet
- After this progresses, you present to the ED and after thorough evaluation, you are diagnosed with presumptive Guillain-Barre syndrome
- You undergo standard treatment and although you progress, your symptoms wax and wane

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Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

- Acquired disorder of peripheral nerves and nerve roots
- Involves cellular and humoral immunity factors
- Provoking antigens have not been previously identified but circulating antibodies to nerve components exist
- 1.5-3.6 new cases/1,000,000 each year
  - 2-5% of those diagnosed initially with Guillain-Barre go on to be diagnosed with CIDP

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Clinical Features

Looks and acts similar to GBS

- Proximal and distal motor weakness
- Diminished or absent deep tendon reflexes
- Sensory involvement
  - Vibratory and position sense > pain and temperature
  - Sensory loss in distal to proximal pattern
- Painful dysesthesias
- Autonomic involvement generally mild
- Slow progressive course

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AIDP vs CIDP

Guillain-Barre (AIDP)

- Monophasic subacute illness that reaches it's nadir in a 4 weeks
- Onset easily identifiable
- At least 70% have antecedent event
- More motor involvement
- Increased ventilator needs

CIDP

- Continues to progress or has relapses for over 8 weeks
  - 3 or more episodes of clinical deterioration
  - Only observation over time clarifies the course
- Precise onset less clear
- Only about 30% have antecedent event
- Prominent sensory signs
  - More likely mild disease course with retained ability to walk independently

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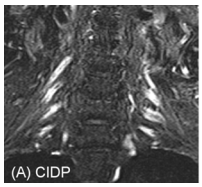
Medical Evaluation



- EMG/NCV
  - Confirm presence of demyelination
- CSF analysis
  - Elevated protein and normal white cell count
- Nerve Biopsy
  - Select a nerve clinically and electrophysiologically affected by the disorder
  - Onion bulb formation from recurrent demyelination/remyelination
- MRI
  - Look for enlarged or enhancing nerves
  - Can guide nerve choice for biopsy
- Laboratory Studies
  - No specific lab test finding support CIDP
  - Rule in or out disorders that can mimic CIDP

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Diagnostic Criteria

- Progression over at least 2 months
- Sensory > weakness
- Symmetric involvement of arms and legs
- Proximal and distal muscles
- Reduced deep tendon reflexes
- Increased CSF protein
- EMG/NCV evidence of demyelination
- Nerve Biopsy demyelination

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Treatment

Initial Treatment

- IVIG
  - Rapid improvement
  - Less likely than steroids to result in remission
  - Expensive
- Pulse Glucocorticoids
  - Inexpensive
  - Chronic use limited by side effects
  - Most likely to produce clinical remission
- Plasma exchange
  - Expensive, invasive and available at specialized centers
- Patients should be evaluated every 2-3 months
- If patients fail to respond, therapy should be adjusted

Response to Treatment

- Success or failure of treatment should be judged using objective measures
  - Grip strength dynamometry
  - 10-meter walk test
  - Measures of impairment
    - Rasch-built Overall Disability Scale
    - Overall Neuropathy Limitations Scale

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Classifying Disease Activity

- Unstable Active Disease (18%)
  - Abnormal exam with progressive or relapsing course
  - Treatment naïve or < 3 months of treatment
- Improvement (7%)
  - Between 3 months and 1 year on treatment
  - Stable or improving exam
- Stable active disease (44%)
  - >1 year on treatment with stable/improving exam
- Remission (20%)
  - <5 years off treatment with stable/improving exam
- Cure (11%)
  - > 5 years off treatment with stable/improving exam
- 30% achieve cure or remission
- Treatment can be discontinued after 1 year of sustained remission
  - Treatment should be resumed if relapses occur
- Patients with refractory disease activity to standard treatments can consider other immunosuppressant options
  - Methotrexate, cyclosporine, cyclophosphamide

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Prognosis

- Data is limited on long-term CIDP prognosis
- 67% initially respond well to single standard treatment
- 10-15% are resistent to therapy

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Imagine this

- You've been on statin therapy for years for hyperlipidemia without issue
- All the sudden, you notice weakness of your shoulders and hips
- This progresses, limiting your mobility and ability to climb stairs.
- You then notice difficulty eating and swallowing
- You present to the ED for evaluation

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Necrotizing Autoimmune Myopathy (NAM)

- Very rare autoimmune myopathy (about 200,000 cases in the US)
- Characterized by subacute proximal limb muscle weakness and high creatinine kinase levels
- Pathologically, there is marked muscle fiber necrosis and regeneration without inflammation
- Age of onset between 30 and 70
- Most commonly idiopathic but can be associated with statin exposure, cancer, connective tissue disease and rarely viral infections (HIV)

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Clinical Features

Physical Exam Findings

- Proximal muscle weakness
  - Generally worse in lower limbs
  - Neck muscles included
  - Distal muscle weakness does occur
- Dysphagia
  - Weight loss
- Respiratory and cardiac abnormalities
  - Dyspnea
  - Neuromuscular respiratory weakness
- Muscle pain

Diagnostic Findings

- Elevated creatinine kinase
- Elevated serum troponin (94%)
- Muscle biopsy revealing necrosis (100%) and regeneration (95%)
- Some will have circulating antibodies
- EMG studies show fibrillation potentials and myotonic discharges
- MRI may show evidence of muscle inflammation
  - Help choose muscle to biopsy

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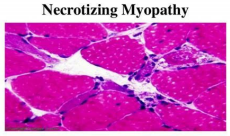
Diagnostic Findings



Scattered necrotic myofibres with myophagocytosis in the paucity of T-lymphocytic infiltration.

- Elevated creatinine kinase
- Elevated serum troponin (94%)
- Aldolase, ALT, AST, lactate dehydrogenase
- Muscle biopsy revealing necrosis (100%) and regeneration (95%) – Gold Standard
- Circulating antibodies
  - ANA
  - anti-SRP or anti-HMGCR antibodies
- EMG studies show fibrillation potentials and myotonic discharges
- MRI may show evidence of muscle inflammation
  - Help choose muscle to biopsy
- Cancer screening to rule out para-neoplastic cause

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Treatment and Prognosis

Immune Modulating Therapy
- Corticosteroids
- IVIG
- Methotrexate
- Mycophenolate mofetil
- A combination of IVIG, corticosteroids and a steroid-sparing immunosuppressant for at least 3 months
- Long term treatment with steroid-sparing immunosuppressant

Prognosis
- > 50% recover markedly or improve to normal
- 10% have little or no improvement
- Relapse rate of about 55% as treatments are tapered or discontinued
- Predictors of favorable outcomes: males, use of 2 more immunosuppressants within 3 months

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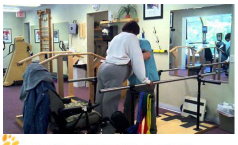
Approaches to Therapy

Patient Specific Considerations
- Fear and Anxiety
- Depression and Guilt
- Pain
- Low motivation due to feelings that recovery is not directly related to effort



Evaluations
- Clinical features can vary widely from person to person
- Patient/Caregiver interviews
- Sensory assessment
  - Sensation, touch, pressure, touch localization
- Skin Inspection
- Joint ROM
- Muscle Testing
- Functional Testing
- Mobility
- Consider
  - Respiration/diaphragm function
  - Autonomic Dysfunction
  - DVT
  - Endurance

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Goals of Therapy

1 – Achieve optimal muscle use and tolerable pain levels

Therapy does not facilitate nerve and muscle repair

It does teach the body to utilize optimal muscle control until nerves/muscles repair

Prevent secondary complications

The body can only do what it can do at this point

2 – Use supportive equipment and functional adaptations for those with residual impairments to allow to return to as much independence as possible

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Specific Considerations at different Disease Stages

Acute GBS/CIDP Flare

- Patient may not be able to tolerated active motion
- Therapy aimed at education and prevention of contracture, skin breakdown and DVT
  - Positioning considerations
  - Gentle PROM as tolerated
  - Breathing exercises
  - Adaptive equipment needs
- Building rapport

Recovery of Function

- PROM to AAROM to AROM
- Begin strengthening – increasing repetitions before adding resistance
- Avoid exercising to point of exhaustion
  - Provide multiple rest breaks
  - Back off if reports of fatigue last more than 12-24 hours
  - Teach energy conservation
- Transition to functional daily and ADL activities
- Home exercise program
- Consider sensory desensitization if warranted

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Patient resources

AIDP/GBS/CIDP

- GBS/CIDP Foundation International
  - https://www.gbs-cidp.org/
    - Provides centers of excellence in diagnosis and treatment
- Guillain-Barre Syndrome Awareness Society
  - http://www.gbsas.org/
- Facebook support groups

NAM

- Myositis Support and Understanding
  - https://understandingmyositis.org/
  - Provides patient and caregiver support
  - Financial support
  - Current research
- Facebook support groups

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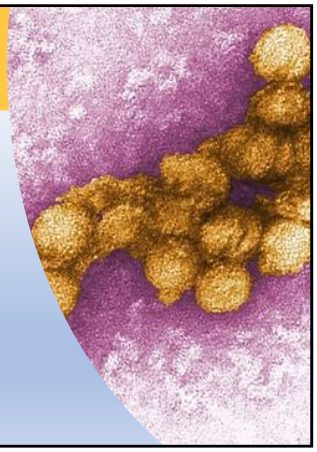
West Nile Virus



- RNA arbovirus
- Leading cause of domestically acquired arbor viral disease
- Increased incidence secondary to higher than average temperatures
  - Cases peak late summer to early fall
- Incidence strongly underreported as most have mild symptomology
  - 1:150-250 develop neuroinvasive disease

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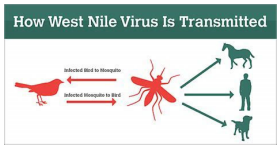
Transmission



*** Can be transmitted in utero, through breastmilk, needle sticks, organ transplant, blood transfusions and through conjunctival exposure***

- Culex species
  - Culex pippin's pipiens in Northern US/Canada
  - Culex pipiens quinquefasciatus in the Southern US
  - Culex tarsalis in Western US/Canada

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Clinical Manifestations

West Nile Fever (25%)

- Most common presentation – asymptomatic or subclinical illness
  - Only 20-40% infected have symptoms
- Incubation period is 2-14 days
- After acute infection, persistent symptoms can exist
  - Fatigue, headache, memory issues, weakness, balance issues
- Once recovered, immunity is thought to be life long

- Self limiting illness
  - Fever, headache, malaise, back ache, myalgias, and anorexia
  - Less commonly: eye pain, pharyngitis, nausea, vomiting, diarrhea and abdominal pain
- Generalized lymphadenopathy
- Rash (20-25%)
  - Chest, back and arms
  - Maculopapular rash
  - Associated with decreased incidence of neuroinvasive disease and death
  - Lasts for about a week
- Acute symptoms last 3-10 days
- Risks for developing, increased viral load and female gender

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Clinical Manifestations cont..

Neuroinvasive Disease (1:150-250)

- Meningitis
  - Fever, headache, meningeal signs and photophobia
- Encephalitis
  - Range from mild self-limiting confused state to coma and death
  - Tremor and myoclonus
  - Parkinsonian features: rigidity, postural instability and bradykinesia
  - Cognitive difficulties can last up to 1 year
- Acute flaccid paralysis syndrome
  - Asymmetric weakness of the limbs that progresses quickly over 48 hours from onset
  - With or without meningitis/encephalitis
  - 1/3 recover to normal, 1/3 modestly improve and 1/3 fail to improve
  - Recovery mostly occurs within 6-8 months

Risk Factors for Development of Neuroinvasive Disease

- Advancing Age
- Malignancy
- Persons infected through organ transplant
- Certain host genetic factors
  - chemokine receptor CCR5 deficiency
- Diabetes, HTN, renal disease, alcohol abuse, male gender

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Less common presentations

- brachial plexopathy, demyelinating neuropathy, motor axonopathy, axonal polyneuropathy, involvement of ventral spinal roots, myasthenia gravis, and a disorder similar in character to Guillain-Barré syndrome
- cranial nerve palsies resulting in facial weakness, vertigo, dysarthria, seizures, cerebellar ataxia, and dysphagia
- Ocular manifestations: chorioretinitis, retinal hemorrhages, and vitreitis
- Rhabdomyolysis, hemorrhagic fever, hepatitis and pancreatitis, myocarditis, myositis, orchitis
- Congenital infections

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Diagnosis

WN virus should be suspected in patients who present with neurologic changes consistent with encephalitis or asymmetrical flaccid paralysis plus development of a maculopapular rash on the trunk and extremities during mosquito season

Laboratory

- Most blood tests do not distinguish West Nile from other viral infections
- With CNS involvement
  - CSF has elevated protein, increased white cell count with predominant lymphocytes

Imaging

- CT scan unlikely to show evidence of acute disease
- MRI scan can show changes but often takes weeks

EEG

- CNS involvement shows generalized slowing worse to temporal and frontal regions

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- **MAC-ELISA**: IgM antibody-capture enzyme-linked immunosorbent assay
  - positive serum or CSF is sufficient to confirm diagnosis in most
- **PRNT**: plaque reduction neutralization test
  - Help discern WN from other infections
  - Positive PRNT and MAC-ELISA is confirmatory

United States Centers for Disease Control and Prevention laboratory criteria for diagnosis of West Nile virus disease

A CONFIRMED case of West Nile virus infection in a patient with clinically compatible disease is determined by meeting one of the following criteria:
• Isolation of virus from, or demonstration of specific viral antigen or nucleic acid in, tissue, blood, CSF, or other body fluid, OR
• Fourfold or greater change in virus-specific quantitative antibody titers (PRNT) in paired sera, OR
• Virus-specific IgM antibodies (MAC-ELISA) in serum with confirmatory virus-specific neutralizing antibodies (PRNT) in the same or a later specimen, OR
• Virus-specific IgM antibodies in the CSF (MAC-ELISA) and negative IgM in the CSF for other arboviruses endemic to the region where the exposure occurred
A PROBABLE case of West Nile virus is considered if:
• Virus-specific IgM antibodies (MAC-ELISA) are present in CSF or serum but no other testing (eg, PRNT, PCR) is performed

A diagnosis of probable disease is sufficient for the vast majority of patients. This is particularly true for those with a clinically compatible illness and epidemiologic risk factors for West Nile virus (eg, no travel history outside the United States and/or presenting during outbreaks). In such patients, there is little need for further PRNT confirmatory testing. Refer to the UpToDate topic review that discusses the diagnosis of West Nile virus for additional information on diagnostic testing.

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Treatment and Prevention

Treatment

- Mainly supportive
- Potential pharmacologic intervention
  - Interferon
    - Potential benefit
  - Ribavirin
    - In animal models – no known efficacy in humans
  - IVIG
    - Potential benefit if no contraindications

Prevention

- Personal protection
- Mosquito control programs
  - Eliminate breeding sites
  - Application of larvicides
  - Ariel spraying
- Blood donor screening
- Vaccine development

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