

**Prevalence of Gluten Sensitivity in a Population of Peripheral Neuropathy Patients**  
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**Objective:** To determine the prevalence of gluten sensitivity as defined by anti-gliadin antibody positivity in patients seen in our general neurology clinic with peripheral neuropathy symptoms.

**Patients and Methods:** Between September and November 2002, patients examined by a neurologist at Wilford Hall Medical Center Neurology Clinic and felt to have neuropathy signs or symptoms were sent for a standard laboratory screen. In addition to the standard evaluation, the patient's blood was also sent for an anti-gliadin antibody panel and any other special tests that the neurologist deemed necessary.

**Results:** To date, 22 patients have been screened. There were 4 patients who had either IgG positivity or had both IgA and IgG positivity. The prevalence rate in this small sample is 18% compared to an estimated prevalence rate of .4% of gluten enteropathy in the United States.

**Conclusion:** Anti-gliadin antibodies are elevated in a number of patients seen in a general neurology clinic with paresthesias.

**Discussion:** Peripheral neuropathy is a common problem in this country, leading to significant morbidity. Peripheral neuropathy, typically a distal, symmetric axonal sensory neuropathy, is associated with celiac disease, and was previously felt to be secondary to fat-soluble vitamin deficiency. However, recent studies indicate that the neuropathy can occur in the absence of vitamin deficiency and responds to a gluten-free diet. Recent reports from England note a 40% incidence of gluten sensitivity in patients sent to a peripheral neuropathy referral clinic. The results of this small pilot study are provocative and consistent with those reports, showing a greatly enhanced prevalence of gluten sensitivity in neuropathy patients as compared to the general population. It is unknown at this time whether the patients have established celiac disease. It has been postulated that gliadin itself is the inciting cause of disease and that neurological manifestations can occur secondary to direct toxicity of gluten, without concomitant celiac disease. These observations might offer alternative treatment modalities, including dietary modification, to the usual therapy for idiopathic peripheral neuropathy.

**Hereditary Neuropathy with Predilection for Pressure Palsies Presenting in a  
Marine Recruit Declared Physically Unfit for Duty**  
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**Background:** Countless number of recruits will fail basic training yearly. This case report highlights an atypical cause for failure of a military recruit to perform to standards secondary to a rare neurologic disorder.

**Case Report:** 17-year-old WM USMC recruit who presents with bilateral arm weakness. Patient reports weakness starting after holding push-up position for one hour several days prior. He now reports that he is unable to perform simple activities of daily living and now has right wrist drop. Patient denies any recent illness, trauma, pain, numbness, tingling, lower extremity weakness, or previous history of weakness. Physical exam was significant for: 1) RUE strength – deltoid/biceps/triceps 4-/5, wrist flexor/extensor 3/5, finger flexor/extensor 3/5, hand intrinsics 3/5; 2) LUE strength – deltoid/biceps/triceps 4/5, wrist flexor/extensor 4-/5, finger flexor/extensor 4-/5, hand intrinsics 4-/5; 3) normal tone, bulk; 4) sensory – patchy decreased pinprick and light touch posterior aspect of forearm and dorsum of hand bilaterally; 5) DTR – triceps/biceps/Achilles reflex 0/4 bilaterally, patellar reflex 1/ 4 bilaterally. CBC and CMP were normal. Non-contrast CT head was normal. Nerve conduction studies showed absent conduction along bilateral radial nerves and prolonged latencies along bilateral ulnar/peroneal nerves and right median nerve.

**Conclusion:** Hereditary neuropathy with predilection for pressure palsies (HNPP) belongs to a family of hereditary neuropathies, which has a prevalence of approximately 1 in 2,500. This particular autosomal dominant disease is characterized by isolated nerve palsies by trivial compression on trauma. Electrophysiologic studies show a hallmark pattern of prolonged distal latencies out of proportion to the slowing of the conduction velocity. The most prominent pathologic feature is focal myelin thickening, or tomacula, on light microscopy.

**Giant Arteriovenous Malformation of the Brainstem and Cervical Spine as an  
Unusual Cause of Foramen Magnum Syndrome**  
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Giant arteriovenous malformations involving both the brainstem and spine are extremely rare. Symptoms referable to the central nervous system, when present, are usually due to hemorrhage, compression, or vascular steal phenomenon. Presented here is a case of giant arteriovenous malformation involving the brainstem and spine causing subacute symptoms and exam findings localized to the foramen magnum in a young man.

He was treated with a combination of neurovascular embolization therapy and external proton-beam radiation that resulted in dramatic improvement in his symptoms and neurologic exam. Radiographs depicting the lesion as a cause of spinal cord displacement and myelopathy in this patient are shown.

A description of the anatomy of the foramen magnum is given. Symptoms, exam findings and potential causative lesions of foramen magnum syndrome are reviewed.

## **Encephalopathy Secondary to West Nile Virus**

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**Case Report:** A 40-year-old homeless Caucasian male presented by squad from a homeless shelter with agitation and hallucinations. The patient was known to the Emergency Department for history of homelessness, alcoholism with cirrhosis and previous admission for alcohol withdrawal and delirium tremens (DTs). The patient was started on Lorazepam drip for agitation as well as Ceftriaxone and Vancomycin for empiric coverage of meningitis. Upon further assessment by the Medical Intensive Care physician, a non-contrast head CT scan and a lumbar puncture (LP) were needed. Head CT was unremarkable and a LP was performed. The cerebral spinal fluid (CSF) was consistent with an aseptic meningo-encephalitis. In addition to bacterial gram stain and cultures, viral studies were sent. Due to the patient's uncertain whereabouts prior to the homeless shelter, history of prolonged outdoor exposure and chronic alcohol abuse, the Ohio Department of Health was contacted for stat West Nile Virus serologies. The patient was found to have WNV positive IgG and IgM. All other cultures and titers remained negative. Supportive care was continued. The patient remained in a permanent vegetative state and was transferred to hospice care. He died 1 month and 3 days after initial presentation.

**Discussion:** The West Nile Virus is one of the most widely distributed arboviruses worldwide but is relatively new to North America. Very few of those infected ever become symptomatic and fewer still require hospitalization. Recognition of symptoms with high clinical suspicion can lead to early diagnosis. While treatment remains supportive, early diagnosis can optimize supportive care and minimize unnecessary interventions and treatments.

## **Wound Botulism in an Intravenous Drug User Presenting with Asymmetrical Neurological Deficits**

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**Objective:** To describe a case of documented wound botulism presenting with asymmetrical neurological findings.

**Background:** Botulism is a paralytic illness caused by a neurotoxin produced by the bacterium *Clostridium botulinum*. The four recognized clinical forms of this disease are food borne botulism, infant botulism, adult infectious botulism and wound botulism. Wound botulism was relatively uncommon in the United States until several cases occurred among intravenous drug users in New York and California in the 1980's. A specific association has been noted between this disease and the use of black tar heroin.

**Case:** We present a case of wound botulism in a 33-year-old female admitted to the medical ICU with difficulty swallowing, blurry vision, and head bobbing. The patient reported a history of intramuscular use of black heroin 4 days prior to presentation. Physical exam was notable for inability to adduct the right eye and right-sided ptosis. Strength was decreased in the bilateral sternocleidomastoid muscles. Skin examination was notable for a right anterior thigh abscess that appeared to have spontaneously drained. An endrophonium challenge was negative and CSF parameters were within normal. A diagnosis of botulism was entertained and serum samples were sent for mouse bioassay. Empiric treatment was initiated with Pen G and botulinum anti-toxin. The anti-toxin was administered within 12 hours of presentation to the medical center and the patient experienced gradual improvement over the following weeks. Mouse bioassay with use of neutralizing antibodies confirmed the diagnosis of botulism with type A toxin.

**Conclusion:** Botulinum toxin is systemically absorbed and mediates its paralytic effect through irreversible binding to the pre-synaptic nerve terminals resulting in inhibition of acetylcholine release. The clinical syndrome associated with botulinum intoxication is described as a descending, symmetrical paralysis. However, a minority of recent case reports have documented asymmetric deficits associated with botulism acquired from black tar heroin use. The asymmetrical findings noted with our patient demonstrate the need to consider atypical presentations of this disease and reinforce the need to be cognizant of the association between wound botulism and IVDA.

## **Rapid Neurological Deterioration in a Patient with Varicella Zoster Viral Encephalitis with Negative Neuroimaging**

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**Case Report:** A 65-year-old, previously healthy man was admitted with a four day history of worsening facial cellulitis and uveitis while on outpatient antibiotics. The patient's forehead and right orbit were erythematous and edematous and a black eschar extended from the right forehead to the nasal tip. Vesicular lesions were noted on his trunk and oral acyclovir was started. On hospital day four he became disoriented, restless, and increasingly confused. Neither computed tomography nor magnetic resonance imaging revealed intracranial pathology. Lumbar puncture was significant for an elevated protein (110 mg/dL) with normal glucose and a lymphocytic predominance of white blood cells. Polymerase chain reaction for varicella zoster virus was positive. The patient subsequently received three weeks of intravenous acyclovir with complete resolution of his symptoms.

**Discussion:** Neurologic manifestations of varicella zoster virus are not uncommon, but true encephalitis in the adult population is not well reported in the United States. The neuropathological mechanism of this disease process has not been fully elucidated; however, it is believed that direct viral invasion or an indirect immune-mediated process plays a major role. Early diagnosis of varicella zoster encephalitis clearly depends on having a high degree of clinical suspicion. Although neuroimaging can be a valuable diagnostic tool, false negative results early in the course limit its utility. Diagnosis is confirmed by cerebrospinal fluid analysis with culture or polymerase chain reaction. Early administration of intravenous acyclovir is imperative. Even with appropriate therapy, however, only half of affected patients fully recover. The diagnosis of varicella zoster encephalitis must be entertained in any patient who presents with suggestive signs, symptoms, and positive laboratory and imaging studies and treatment should be rapidly initiated to minimize permanent sequelae.

## **Patients With Alzheimer's Disease Have Increased Antibody Responses to Amyloid-Beta (Ab) Peptide that Promotes AB Aggregation**

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Animal models for Alzheimer's disease (AD) show that immune responses to Ab can clear amyloid plaques and prevent neurotoxicity. However, immune responses to Ab are poorly characterized in patients with AD.

Antibody levels were determined by ELISA to either preaggregated or unaggregated Ab in serum samples from patients with AD (n=16) and compared to age-matched controls (n=31), patients with multiple sclerosis (n=11) or patients with HIV infection (n=11). Antibody levels were also determined in another 17 patients with ad and 31 matched controls. T cell proliferation was monitored in response to Ab in AD (n=11) and controls (n=4). To determine if antisera to Ab can modulate its aggregation, IgG was purified and incubated with freshly dissolved Ab for 24 hours and the amount of aggregation measured using thioflavin T dye.

**Results:** Antibody titers to Ab in patients with AD were markedly elevated ( $P < 0.003$ ) while no significant difference was found in the two groups for antibodies to unaggregated Ab. 3/17 postmortem CSF samples from AD patients and 0/31 controls had antibodies to Ab. No significant T cell proliferation to Ab was seen in AD or control subjects. IgG was localized in several amyloid plaques either in the center, periphery, or throughout the plaque. Purified IgG from patients with Ad caused increased aggregation of Ab in the aggregation assay compared to controls ( $p < 0.005$ ).

**Conclusions:** Patients with AD have increased humoral immune responses to Ab in the serum and CSF. Antibodies to Ab can be localized to the amyloid plaque where they likely enhance aggregation of the peptide.

**Subcortical Dementia Caused by Bilateral Internal Carotid Artery Occlusion  
(Moyamoya) in a 47-Year-Old Hispanic Woman**

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Moyamoya is an idiopathic progressive vasculopathy of the intracranial vessels, and ultimately leads to their occlusion. Moyamoya is classically described as a disease affecting young Asians and presenting with focal neurological deficits due to ischemic strokes or intracranial hemorrhage. Cases of Moyamoya in other ethnic groups and those associated with atypical neuropsychiatric presentations have not been reported often.

Calcium channel blockers, steroids, and anti-platelet agents have been used with mixed success. A randomized clinical trial failed to show benefit of surgical revascularization over medical therapy. There is some evidence to suggest that a subgroup of patients with more advanced hemodynamic failure may benefit from surgical vascularization. It may be possible to identify these patients with certain studies employing single-photon emission computed tomography (SPECT) or positron emission tomography (PET).

We present a case of a 47-year-old female patient with no history of hypertension, cardiac, or peripheral vascular disease that presented with subcortical vascular dementia (VaD) secondary to bilateral internal carotid artery occlusion (Moyamoya disease).

Moyamoya may be an under recognized case of VaD in the young. This may suggest that clinicians consider including intracranial MRA in the evaluation of younger patients of any ethnicity with acquired neuropsychiatric impairment.

The case reported is accompanied by a literature review, and a discussion of possible guidelines for additional diagnostic testing and available treatment alternatives.