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A summary of recently published articles in the *Neurology*® Resident & Fellow Section

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July 3, 2018 issue

This issue starts off with a Clinical Reasoning case that reviews the differential diagnosis of acute headache in an adult. The Pearls & Oysters is a pediatric case discussing a genetic syndrome with paroxysms that may be difficult to distinguish from seizures. There are also 2 Teaching NeuroImages that describe a rare cause of ophthalmoparesis and visual anosognosia.

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Clinical Reasoning: A 41-year-old man with thunderclap headache

A 41-year-old man with history of low testosterone on androgen therapy presented to the emergency department complaining of acute onset of the worst headache of his life. Thunderclap headache presents multiple diagnostic possibilities, such as subarachnoid hemorrhage, pituitary apoplexy, reversible cerebral vasoconstriction syndrome, meningitis, venous sinus thrombosis, and hemorrhagic or ischemic stroke.

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Pearls & Oysters: Alternating hemiplegia of childhood mimics focal epilepsy and paroxysmal dyskinesia in infancy

This is a detailed account of a case of alternating hemiplegia of childhood (AHC) and the challenges associated with the diagnosis and treatment. Our case highlights the fact that in AHC, the classic hemiplegic attacks are preceded by ocular abnormalities and dyskinesia by several months. These spells often mimic seizures, which can further delay the diagnosis.

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Teaching NeuroImages: Presentation of diffuse large B-cell lymphoma with bilateral sequential oculomotor neuropathy

A 56-year-old man with HIV on combination antiretroviral therapy had sequential, bilateral oculomotor nerve palsy after recent travel to Africa. Nonexpansile enhancement of the oculomotor nerves was noted on MRI.

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Teaching NeuroImages: Distinct visual anosognosia after serial lesions of Meyer loop and the lateral geniculate body

A clinical experience of a patient with a sequence of anosognosia for quadrantanopia is presented. Previously unaware of an upper right quadrantanopia after left temporal glioma resection, this man complained of a novel visual field defect restricted to that quadrant. Surprisingly, MRI did not show recurrence of glioma, but a new ischemic lesion of the left-sided lateral geniculate body.

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July 10, 2018, issue

Pediatric autoimmune neurologic diseases can have a variety of presentations including encephalopathy, seizures, and movement disorders. The Child Neurology article in this month's issue describes a case of a rare autoimmune movement disorder. The Clinical Reasoning reviews the differential diagnosis of diplopia in a pregnant woman. Finally, the Teaching NeuroImages describe MRI in prion disease and a rare lysosomal storage disease.

Child Neurology: Treatable bilateral striatal lesions related with anti-dopamine 2 receptor autoimmunity

A 16-month-old healthy boy was admitted to the emergency department due to sudden onset of altered mental status and abnormal posturing of his limbs. He showed marked irritability, poor eye contact, sustained flexion of his left limbs, and extension of his right limbs alternating with mixed choreic-dystonic movements of the 4 limbs and mandible.

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Clinical Reasoning: Onset of diplopia in pregnancy

A 32-year-old woman presented at 27 weeks gestation with new-onset diplopia. She awoke with persistent binocular horizontal diplopia. She denied headache, ocular pain, or other associated neurologic symptoms. There were no previous episodes.

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Teaching NeuroImages: Cerebral cortex swelling in Creutzfeldt-Jakob disease with V180I mutation

A 74-year-old woman developed amnesia that rapidly progressed over 6 months. She exhibited no apparent neurologic abnormalities, except for cognitive decline. Mini-Mental State Examination score was 20/30, mainly involving orientation and recent memory. EEG revealed no periodic synchronous discharge.

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Teaching NeuroImages: Brain MRI and DaT-SPECT imaging in adult GM1 gangliosidosis

We describe a 58-year-old woman with genetically confirmed diagnosis of adult GM1 gangliosidosis, which clinically started at age 20 years with a generalized dystonic syndrome later followed by an akinetic rigid parkinsonism.

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July 17, 2018, issue

The Child Neurology article highlights the role of genetic testing in children with developmental delay and seizures. There are 3 Teaching NeuroImages cases that include an atypical encephalitis, a rare cause of cervicothoracic syrinx, and sequelae of Guillain-Barré syndrome.

Child Neurology: Siblings with infantile epilepsy and developmental delay: A circuitous path to genomic diagnosis

We report 2 full siblings, a brother and sister, with a unique familial 2.4 Mb microdeletion at 14q13.1–14q13.3 by chromosomal microarray. Both children presented with infantile spasms that evolved to intractable epilepsy and profound developmental delay. They share distinctive dysmorphic features.

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Teaching NeuroImages: Isolated pontine involvement in subacute sclerosing panencephalitis

A 16-year-old boy, with history of measles at age 4, developed generalized myoclonic jerks and progressive decline in cognition and sensorium over 6 months. Generalized, periodic, high-amplitude discharges on EEG and elevated immunoglobulin G anti-measles antibody in CSF suggested subacute sclerosing panencephalitis.

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Teaching NeuroImages: Spinal cord syrinx secondary to a spinal dural arteriovenous fistula

A 60-year-old man presented with a 2-year history of progressive lower limb weakness, numbness, urinary retention, and chronic constipation with superimposed episodes of severe paraparesis. MRI showed a cervicothoracic syrinx with flow voids.

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Teaching Video NeuroImages: Propriospinal myoclonus as a sequela of Guillain-Barré syndrome

A previously healthy 27-year-old woman developed Guillain-Barré syndrome (GBS) with severe tetraplegia, requiring immunoglobulins. During remission, nonrhythmic, stimulus-sensitive abdominal jerks, propagating to the hips, appeared, worsening in supine position. Surface EMG confirmed propriospinal myoclonus 1 originating from Th8.

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July 24, 2018, issue

This issue starts off with 2 difficult cases. The first is a Pearls & Oy-sters describing a rapidly progressive neuromuscular disease. The second is a Clinical Reasoning case describing a patient with double vision. There is a Teaching NeuroImages of Alexander disease and a Teaching Video NeuroImages localizing a focal epilepsy onset.

Pearls & Oy-sters: A curable myopathy manifesting as exercise intolerance and respiratory failure

A young patient presented with exercise intolerance and progressive dyspnea with normal initial evaluation that quickly

evolved into a dramatic respiratory insufficiency and severe muscle weakness. Muscle MRI and muscle biopsy depicted a fatty infiltration and lipid droplets accumulation, respectively.
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Clinical Reasoning: A patient with multiple cranial nerve palsies and areflexic paraparesis

A 62-year-old hypertensive man presented with a 1.5-month history of bilateral facial nerve palsy evolving over 5 days, followed by double vision and restriction of eye movements for 1 month. He had progressive deterioration of vision of 20 days duration.

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Teaching NeuroImages: Alexander disease with features of both frontal and bulbo-spinal involvement

A 28-year-old woman, who was considered to have Alexander disease at 14 months of age, presented with bulbo-spinocerebellar symptoms from the age of 22 years.

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Teaching Video NeuroImages: The frontal eye field

Finding the ictal onset zone in frontal lobe epilepsy is a challenge because many different signs are usually associated during the seizures. Among them, contralateral eye deviation indicates an involvement of the small region of the frontal eye field. This video shows a case of pure frontal eye field seizure.

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July 31, 2018, issue

Innovation in resident and fellow education is one of the main goals of the Resident & Fellow Section. This Education Research article describes a novel model of curriculum development. There are 3 Teaching NeuroImages that cover topics such as a rare adult case of necrotizing encephalitis, isolated brain overgrowth syndrome in a child, and a video of gaze palsy in spinocerebellar ataxia.

Education Research: Resident education through adult learning in neurology: Implementation and impact

This study evaluates the efficacy and feasibility of an adult learning theory-based curricular program in neurology education. The results show robust and sustainable benefit for residents in training without imposing a financial or logistical burden on programs. Resident Education through Adult Learning in Neurology could serve as a model for curricular reform in other programs across subspecialties.

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Teaching NeuroImages: Radiographic evolution in an adult case of acute necrotizing encephalopathy

A healthy 37-year-old woman developed delirium and was comatose on admission after a 3-day history of chills, fever, and myalgias. Neuroimaging showed lesions affecting bilateral thalami and pons, and normal venous sinuses.

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Teaching NeuroImages: A case of isolated hemithalamic overgrowth

We report on a 17-month-old boy who came to our attention for a left hemiparesis, without any other neurologic disorder. Brain MRI revealed a remarkable hypertrophy of the left thalamus. There were no differences vs the right thalamus before or after using contrast, hypersignal in fluid-attenuated inversion recovery, or restriction at diffusion-weighted imaging.

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Teaching Video NeuroImages: Upward gaze palsy is a sign of spinocerebellar ataxia type 3

A 32-year-old woman presented with a 5-year history of ataxia, dysphagia, and dysarthria; family history was positive for spinocerebellar ataxia type 3. Examination revealed bulging eyes, upward gaze palsy with vertical nystagmus, bilateral horizontal nystagmus, slow saccades, dysmetria, and gait ataxia.

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Resident & Fellow Rounds

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