Algrove syndrome: Mutations in ALADIN causing motor neuron disease

Goizet et al. describe a patient with progressive bulbospinal amyotrophy as part of the disease called Triple A (adrenal insufficiency, achalasia, and alacrima) syndrome, also know as Allgrove's syndrome. The disorder is caused by mutations in the gene for ALADIN, which is proposed to play a role in regulating the function of peroxisomes.

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The accompanying editorial by Richard Orrell and Adrian Clark (who was a member of one of the teams that discovered the gene for Triple A syndrome) reviews this autosomal recessive disorder, together with the recently reported genetic mutations in another recessive condition, which includes ALS and primary lateral sclerosis presentations.

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Treatment of ciguatera fish-poisoning

Ciguatera poisoning is the most frequent nonbacterial food poisoning worldwide. The Schnorf et al. randomized, controlled study found that mannitol, the standard treatment of ciguatera poisoning, is no more effective than normal saline in improving neurologic symptoms and signs at 24 hours.

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Is the prevalence of migraine changing?

A large epidemiologic study by Lipton et al. showed that migraine prevalence has not changed in the past decade. However, patients have sought medical consultation at more than twice the previous rate. Migraine patients were also taking more prescription medications.

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Familial aggregation of psychosis in Alzheimer disease (AD)

Sweet et al. found that the risk for an individual developing psychotic symptoms during the course of AD was increased when a sibling with AD had also manifested psychotic symptoms. The demonstration of a familial risk for psychosis in AD suggests an underlying genetic basis.

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Sleep apnea following acute stroke

Iranzo et al. performed polysomnography the first night after acute cerebral infarction in 50 patients. Sleep apnea was found in 62% of the subjects and was associated with stroke onset during sleep and with early neurologic worsening, but had no effect on prognosis in terms of infarct size or functional outcome at 6 months.

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Sleep deprivation for laboratory diagnosis of sleepwalking

Joncas et al. examined the value of sleep deprivation as a diagnostic tool for adult sleepwalking. Sleep deprivation resulted in a significant increase in the frequency and complexity of somnambulistic events recorded in the sleep laboratory.

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The mystery of the Doctor's son

The syndrome of infantile spasms (West syndrome) was first reported by Dr. West in his own son. This Historical Neurology by Eling et al. considers West's 1841 *Lancet* report and the history of West and his son.

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Flying foxes, cycad neurotoxins and ALS-PDC in Guam

In a Medical Hypothesis, Cox and Sacks propose that villagers who ate flying foxes unwittingly ingested large amounts of cycad neurotoxins, leading to the observed 100-fold increase in incidence of the ALS-Parkinson dementia complex in Guam. There, flying foxes actively forage on cycad seeds. If the flying foxes sequester cycad neurotoxins or their metabolites, the prominence of flying fox flesh in the Chamarro diet may account for ALS-PDC being the leading cause of adult death in some villages.

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Screening for the Huntington's disease (HD)–like 2 mutation

The majority of patients with a HD phenotype have CAG expansion of the huntingtin gene on chromosome 4. Stevanin et al. report that the CAG/CTG repeat expansion in the gene encoding junctophilin-3 accounts for ~0.2% of the patients with HD and appears restricted to patients of African ancestry. Further genetic heterogeneity of HD is expected.

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