In Focus

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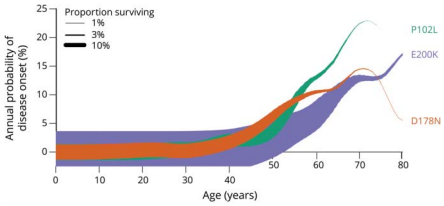
Notable in *Neurology* this week

This issue features an article that assesses the impairment in and developmental vs progressive character of spinocortical proprioceptive pathways in Friedreich ataxia; another investigates the effect of earlier or later resective surgery on seizure outcome. A featured Views & Reviews article discusses the areas of agreement and opportunity in the Food and Drug Administration's industry draft guidance for amyotrophic lateral sclerosis (ALS) drug development, the ALS community recommendations, and ALS Clinical Trial Guidelines.

Articles

Age at onset in genetic prion disease and the design of preventive clinical trials



Genetic prion disease is rare and patients' age at onset varies widely. This study suggests that these factors may make it numerically infeasible to design a randomized prevention trial following healthy mutation carriers to disease onset. Preventive approaches in prion disease will likely need to rely on biomarker endpoints.

Page 54

Use of β2-adrenoreceptor agonist and antagonist drugs and risk of Parkinson disease

β2-adrenoceptor agonists/antagonists modify the risk for Parkinson disease, but the public health implications are unclear. The authors replicated the association of β2-adrenoreceptor agonists and antagonists with reduced and increased Parkinson risk, respectively. However, the data suggest that smoking causes the β2-adrenoreceptor agonist association, and reverse causation underlies the β2-antagonist association.

Page 55

Risks of 23 specific malformations associated with prenatal exposure to 10 antiepileptic drugs

This French nationwide cohort study investigated the risks of 23 specific major congenital malformations associated with prenatal exposure to antiepileptic drugs. The results confirm valproate teratogenicity and the association between topiramate exposure and the risk of cleft lip. No association was found for lamotrigine, levetiracetam, carbamazepine, oxcarbazepine, or gabapentin.

Page 59

A randomized controlled trial with everolimus for IQ and autism in tuberous sclerosis complex

Previous contributions to better outcome in children with tuberous sclerosis complex (TSC) offered only supportive care and early seizure control. The authors investigated everolimus to determine the effect on intellectual disability, autism, and other neuropsychological deficits. With no-effect results, use of everolimus in children with TSC should not be encouraged.

Page 62

Continued

MORE ONLINE

Editor's Summary
Audio summary of highlighted articles.
NPub.org/edsum

From editorialists Ess & Franz: "The findings of Overwater et al. should pave the way for another rigorous clinical trial where very young children with TSC are treated with mTOR inhibitors if possible prior to, or congruent with, developmental delay onset."
Page 51

NB: "Pearls & Oy-sters: Adolescent-onset adrenomyeloneuropathy and arrested cerebral adrenoleukodystrophy," p. 81. To check out other Resident & Fellow Section Pearls & Oy-sters articles, point your browser to Neurology.org/N and click on the link to the Resident & Fellows Section. At the end of the issue, check out the Resident & Fellow Section Teaching Video NeuroImage illustrating tongue myokymia in hypoglossal neuropathy. This week also includes a Reflections: Neurology and the Humanities piece titled "Hearing about your diagnosis while walking on the beach."

NEW EPISODE

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July 9, 2019

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Use of ß2-adrenoreceptor agonist and antagonist drugs and risk of Parkinson disease (see p. 55)

1. Use of ß2-adrenoreceptor agonist and antagonist drugs and risk of Parkinson disease
2. What's Trending: The US Food and Drug Administration's Authorization of the First Cannabis-Derived Pharmaceutical

In the first segment, Dr. Jeffrey Ratliff talks with Dr. Franziska Hopfner about her paper on the use of ß2-adrenoreceptor agonist and antagonist drugs and risk of Parkinson disease. In the second part of the podcast, Dr. Jason Crowell focuses his discussion with Dr. Jerzy Szaflarski on his *JAMA Neurology* paper discussing the US Food and Drug Administration's authorization of the first cannabis-derived pharmaceutical. The paper can be accessed here: https://jamanetwork.com/journals/jamaneurology/article-abstract/2714720.

Disclosures can be found at Neurology.org.

Spotlight on the July 9 issue

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