

## Parry–Romberg syndrome with migraine and intracranial aneurysm

Pichiecchio et al. report a patient with progressive facial hemiatrophy associated with migraine and intracranial aneurysm. Aneurysm in this rare disease may be related to a neural crest migration disorder.

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## The neural crest, aneurysms, and dissections

Commentary by Wouter I. Schievink, MD

What causes an intracranial aneurysm? This commonly asked question is difficult to answer succinctly. Risk factors include smoking cigarettes, hypertension, and binge drinking; genetic factors are also important.<sup>1</sup> Some insight into the development of intracranial aneurysms can be gleaned from their association with rare disorders of the extracellular matrix such as Ehlers–Danlos syndrome type IV or autosomal dominant polycystic kidney disease. The article by Pichiecchio et al. suggests a role for the neural crest in the development of intracranial aneurysms. A neural crest disorder had been implicated previously to explain the association of intracranial aneurysms and cervical artery dissections with neural crest-derived disorders such as cutaneous lentiginosis, bicuspid aortic valve, and aortic coarctation.<sup>2,3</sup> The neural crest

occurs transiently at a very early stage of embryogenesis<sup>4</sup> and is unique because its cells migrate to all parts of the embryo and differentiate into many specific cell types, such as Schwann cells and melanocytes. Most of the mesenchymal structures of the head and neck, including the tunica media of the aortic arch and cervicocerebral vessels, are of neural crest origin.

Many genes are involved in neural crest formation and further molecular studies will likely detect a heterogeneous group of disorders predisposing to the development of intracranial aneurysms.

Should patients with Parry–Romberg syndrome be screened for intracranial aneurysms? I believe these patients should be informed of the possibly increased risk for harboring an aneurysm and screening should

be offered. Screening with MR angiography has no risk and the potential benefits are unquestionable.

### References

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3. Schievink WI, Mokri B, Piepgras DG, Gittenberger-de Groot AC. Intracranial aneurysms and cervicocephalic arterial dissections associated with congenital heart disease. *Neurosurgery* 1996;39:685–690.
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## Clinical trial of magnesium, diazepam, or both after out-of-hospital cardiac arrest

Most patients resuscitated from out-of-hospital cardiac arrest do not regain consciousness. Longstreth et al. examined whether magnesium, diazepam, or both, when administered in the field by paramedics would improve neurologic recovery compared with placebo. No significant benefit or harm was detected.

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*The accompanying editorial by Schneider and Diringier considers the challenges inherent in research on resuscitation of sudden cardiac death, noting that this study gives evidence suggesting that the participating paramedics somehow became unblinded and biased treatment assignments. There is still suboptimal treatment for the more than 400,000 people per year who suffer cardiac arrest. Because there are many candidates for treatment under development, the Longstreth study is a prototype for the cooperation of emergency medicine, critical care physicians, internists, and neurologists.*

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## Neurologic benefit resulting from treatment of stroke in rats

Li et al. found significant functional recovery but no reduction in volume of cerebral infarction after stroke when rats were given IV human bone marrow stromal cells (hMSC). The hMSC appear to function as mini-factories, producing neurotrophic factors in the compromised brain. These factors may then induce proliferation of endogenous cells and reduce apoptosis in the lesion boundary.

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*The accompanying editorial by Rempe and Kent notes the important implications of this animal study. Because hMSC injected IV can home to the area of ischemic injury, they may serve as vectors to deliver substances that promote recovery. Whether hMSCs can actually be integrated into damaged tissue and function as neurons is not established.*

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## Systolic blood pressure: Strong predictor of carotid atherosclerosis

Ultrasound-detected carotid artery intima-media thickness (IMT) is regarded as a valid index of atherosclerosis. In Weber's longitudinal study of 483 healthy men, age, cigarette smoking in the past, and systolic blood pressure were predictors of IMT. Interestingly, even high-normal systolic blood pressure levels were associated with thicker IMT.

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## Surgery: A risk factor for sporadic Creutzfeldt–Jakob disease (CJD)

Ward et al. found that a history of surgery was a risk factor for sporadic CJD; the risk was not dependent on number of operations and was greater in women. Neurologic operations did not increase risk, whereas risk was increased with gynecological and other operations.

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*The accompanying editorial by Mastrianni and Roos notes that although these data do raise concerns about the adequacy of surgical instrument sterilization, the fact that neither the number of surgeries nor neurosurgical procedures increased risk is puzzling if indeed contaminated surgical instruments account for this increased risk.*

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## Primary care costs before the diagnosis of Alzheimer's disease

Albert et al. identified incident AD in a population-based sample and then examined Medicare records for respondents who developed AD and those who did not. They found that people who developed AD had higher outpatient and ambulatory care costs 1 to 2 years before diagnosis. Projected to the US population of adults aged  $\geq 75$  years, the prodromal period of AD was associated with an excess primary care costs of \$128.5 to \$194.7 million.

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## Benign hereditary chorea

Breedveld et al. followed up on their earlier work on benign hereditary chorea (BHC) testing additional BHC families for genetic linkage to chromosome 14q12-q13. Three out of seven families showed linkage to chromosome 14q12-q13. Four other BHC families were not linked. Comparison of the clinical features demonstrated clinical heterogeneity between the families linked to 14q12-q13 on the one hand and the unlinked families on the other. Benign hereditary chorea linked to 14q12-q13 is a well-defined clinical and genetic entity.

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## Neurologic complications in alcoholic liver disease

Buis et al. found a postoperative acute confusional state in 48% of liver transplant patients with prior alcoholism. Recipients with preoperative hyperammonia and shorter duration of abstinence were at greatest risk.

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## Headache aggravation during interferon beta therapy in MS

Pöllmann et al. demonstrated worsening of pre-existing headaches in patients with MS on IFN $\beta$ . Headaches were much more frequent with IFN $\beta$  than glatiramer acetate.

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## Increased CSF hypocretin in restless leg syndrome?

Allen et al. found that subjects with the restless leg syndrome (RLS) have increased CSF hypocretin-1 in the evening. The increased levels were present only in early-onset cases. This observation raises the possibility that increased hypothalamic hypocretin transmission may contribute to RLS.

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## Sinus foreign body and typical cluster headache



Scorticati et al. describe maxillary sinusitis secondary to a foreign body, associated with typical cluster headache with seasonal fluctuations. Unresponsive to several drugs, pain remitted after surgery.

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