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# AJNR

## ***PANK2* Mutation Screening Recommended to Confirm Diagnosis of Pantothenate Kinase –Associated Neurodegeneration**

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## Neurointerventional Guide Sheath

We read with interest Luzardo et al's<sup>1</sup> technical note in the December issue of the *AJNR*. Although we are pleased that the new Envoy 6F guide catheter (Cordis, Miami Lakes, Fla) facilitated a dual-microcatheter procedure with a single arterial access, it should be noted that this is by no means a new technique. The 6F Envoy guide catheter with an inner diameter of 0.070 inches and outer diameter of 2.0 mm is comparatively new, but we have been using the same approach to perform balloon-assisted coiling with a 5F Shuttle guide sheath (Shuttle-SL; Cook, Bloomington, Ind) for more than 4 years. The 5F Shuttle guide sheath is currently our preferred guide sheath for endovascular management of intracranial aneurysms. The 5F Shuttle guide sheath has an inner diameter of 0.074 inches and an outer diameter of 0.090 inches (2.3 mm). For the purpose of balloon-assisted coil embolization, one could easily pass a compliant balloon catheter such as the HyperForm or HyperGlide (Micro Therapeutics, Irvine, Calif) and a standard 0.010- or 0.014-inch microcatheter through the Shuttle. We have used this method for other dual-catheter techniques (eg, temporary proximal balloon occlusion with nondetachable balloon catheters and coaxial coil and foreign body retrieval with the Goose Neck Snare [Microvena, Vadnais Heights, Minn] or the Retriever device [Target Therapeutics/Boston Scientific, Fremont, Calif]).

The Shuttle guide sheath is a hydrophilic-coated braided sheath that provides excellent stability during the procedures. It has a soft, radiopaque tip and is kink resistant. Cook's Shuttle sheaths have 2 inner dilators with the 5F system (0.018 or 0.038 inch), which allows for a better transition between the wire, inner dilator, and sheath. It also comes with a side port, which facilitates contrast injection. In addition, a Tuohy rotating hemostatic valve can be connected to the side port, through which one could pass the Micro Therapeutics balloon catheter facilitating individual control of each microcatheter. Because our patients typically receive anticoagulants, on completion of the procedure we exchange the 5F Shuttle guide sheath for a standard 5F femoral arterial access sheath, which remains sutured in place overnight. We would also like to point out that, at the end of the procedure, the hole in the femoral artery is 5F when a Shuttle is used as opposed to 6F when the Envoy guiding catheter is used. As mentioned in the article, we agree that the use of this technique has resulted in reduced periprocedural risk when compared with separate femoral arterial catheterizations. We have experienced no technical complications with the use of the 5F Shuttle in conjunction with 2 microcatheters.

## Reference

1. Luzardo GD, Ross IB, Gal G. **Balloon-assisted coiling through a single 6F guiding catheter.** *AJNR Am J Neuroradiol* 2006;27:190–91

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## PANK2 Mutation Screening Recommended to Confirm Diagnosis of Pantothenate Kinase–Associated Neurodegeneration

In a recent report in the *AJNR*,<sup>1</sup> Koyama and Yagashita describe brain perfusion studies on single-photon emission CT in a 6-year-old boy diagnosed with pantothenate kinase–associated neurodegeneration (PKAN). Although the authors state that this patient was diagnosed on the basis of clinical findings, including the eye-of-the-tiger

sign on T2-weighted MR imaging, there is no mention of diagnostic molecular testing of the causative gene, *PANK2*. The T2 image demonstrating the eye-of-the-tiger sign was not provided. A verbal description of the T2 findings without an accompanying image makes it difficult to know whether this patient has PKAN or another form of neurodegeneration with brain iron accumulation. Because the authors are reporting a new finding, it is critical for the reader to be certain of which disorder the patient has. On the basis of our extensive experience with PKAN, we recommend using molecular testing to confirm the diagnosis even in the presence of an eye-of-the-tiger sign. The current standard of care for a suspected diagnosis of PKAN is to obtain *PANK2* mutation analysis. Although MR imaging can be strongly suggestive of PKAN, the diagnosis is not established without mutation studies.

## Reference

1. Koyama M, Yagashita A. **Pantothenate kinase-associated neurodegeneration with increased lentiform nuclei cerebral blood flow.** *AJNR Am J Neuroradiol* 2006;27:212–13

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## Maintenance of Certification

*“But he has nothing on at all,” said a little child at last. “Good heavens! listen to the voice of an innocent child,” said the father, and one whispered to the other what the child had said. “But he has nothing on at all,” cried at last the whole people. That made a deep impression upon the emperor, for it seemed to him that they were right; but he thought to himself, “Now I must bear up to the end.” And the chamberlains walked with still greater dignity, as if they carried the train which did not exist.<sup>1</sup>*

Congratulations to Dr. David Hackney for his article regarding Maintenance of Certification (MOC) in the January 2006 issue of the *AJNR*,<sup>2</sup> who exclaims, “Where are the ABR's clothes?”

When I took the examination 10 years ago, I rationalized that the \$3200 assessment was necessary to offset expenses and compensate the dedicated examiners who spent much of the week in Louisville. The reasons for the current fee and subsequent yearly dues, as well as requirements for distant travel to a testing site, are a mystery because the examination requires only a proctor (who gets \$100 per day plus expenses)<sup>3</sup> and is electronically graded. The rationale given on the ABR Website is that the cost is based on analysis of past expenses on a cost-per-candidate basis; this hardly seems relevant in light of the new format of an electronic examination on an ordinary PC.

Has the ABR reached out to the organizers of the ASNR, RSNA, ACR, and other major organizations to offer the examination at major medical meetings (San Diego, anyone)? Noooo. Neuroradiologists are eager to participate in the process, as evidenced by the immense crowds at the Neuroradiology SAM (self-assessment module) course on Monday of the 2005 RSNA. There were so many people at this interactive course that attendance at the scientific sessions suffered, much to the chagrin of the presenters and organizers, who spent months preparing those sessions in Chicago.

Maintenance of competency, at least in our practice, requires a familiarity with advanced imaging modalities, including diffusion/perfusion, rapid triage for appropriate angiography/interventional cases, spectroscopy and positron-emission tomographic imaging, and MR and CT angiography. If the board wants to measure compe-