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Mitchell S.V. Elkind, MD, MS

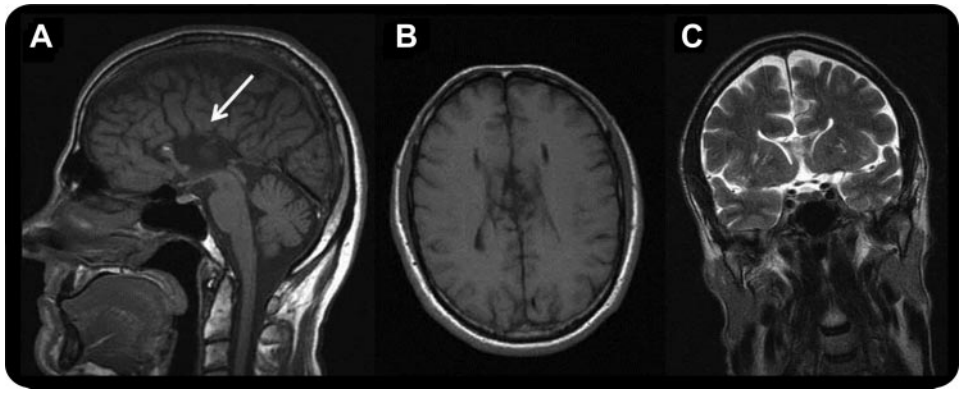
Teaching Neuro*Images*: Hypothermia and corpus callosum agenesis in Shapiro syndrome

Too cold, even for a Viking

Daniel L. Kenney, MD
Michel Toledano, MD
Brian D. Moseley, MD

Correspondence & reprint requests to Dr. Moseley: briandmoseley@gmail.com

Figure MRI of the brain



(A) Sagittal T1-weighted and (B) axial T1-weighted images showing corpus callosum agenesis (arrow) suggestive of Shapiro syndrome. No pituitary abnormalities were noted. (C) Coronal T2-weighted image showing the Viking helmet or moose head appearance assumed by the lateral ventricles in the absence of the corpus callosum.

A 58-year-old man presented with daily hour-long spells of hypothermia, hyperhidrosis, and hypotension. Results of laboratory studies, including thyroid-stimulating hormone, free thyroxine, total/bioavailable testosterone, morning cortisol, and urine osmolality, were normal. MRI of the brain revealed corpus callosum agenesis (figure, A and B) consistent with Shapiro syndrome.

Shapiro syndrome is a rare disorder described in fewer than 60 patients. It consists of periodic hypothermia, hyperhidrosis, and corpus callosum agenesis.[1,2] Any condition with corpus callosum agenesis can cause the lateral ventricles to assume a Viking helmet appearance (figure, C). The hypothermia in Shapiro syndrome is probably due to hypothalamic dysfunction and may be responsive to clonidine.[2]

AUTHOR CONTRIBUTIONS

Drs. Kenney and Moseley made substantive contributions to the design of the study and drafting of the manuscript. Drs. Toledano made substantive contributions to the revision of the manuscript. All authors gave final approval to the version to be published.

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From the Division of Child and Adolescent Neurology, Department of Neurology (D.L.K.) and Department of Neurology (M.T., B.D.M.), Mayo Clinic, Rochester MN.

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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Daniel L. Kenney, Michel Toledano and Brian D. Moseley

Neurology 2012;79;e78
DOI 10.1212/WNL.0b013e318266fc66