

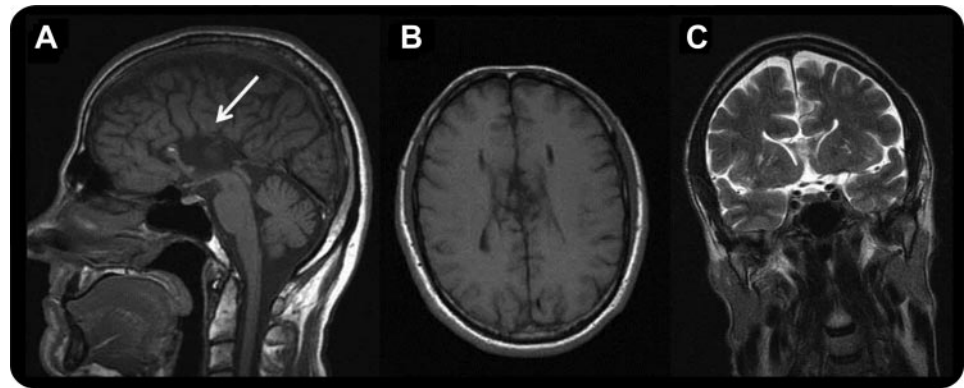
Teaching NeuroImages: Hypothermia and corpus callosum agenesis in Shapiro syndrome

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.

REFERENCES

1. Shapiro WR, Williams GH, Plum F. Spontaneous recurrent hypothermia accompanying agenesis of the corpus callosum. *Brain* 1969;92:423–436.
2. Klein CJ, Silber MH, Halliwill JR, Schreiner SA, Suarez GA, Low PA. Basal forebrain malformation with hyperhidrosis and hypothermia: variant of Shapiro's syndrome. *Neurology* 2001;56:254–256.

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.

Neurology[®]

Teaching *NeuroImages*: Hypothermia and corpus callosum agenesis in Shapiro syndrome: Too cold, even for a Viking

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

Neurology 2012;79:e78

DOI 10.1212/WNL.0b013e318266fc66

This information is current as of August 27, 2012

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/79/9/e78.full
References	This article cites 2 articles, 1 of which you can access for free at: http://n.neurology.org/content/79/9/e78.full#ref-list-1
Citations	This article has been cited by 1 HighWire-hosted articles: http://n.neurology.org/content/79/9/e78.full##otherarticles
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Autonomic diseases http://n.neurology.org/cgi/collection/autonomic_diseases MRI http://n.neurology.org/cgi/collection/mri
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2012 by AAN Enterprises, Inc.. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

