Teaching NeuroImage: Bilateral Nucleus Tractus Solitarius Lesions in Neurogenic Respiratory Failure

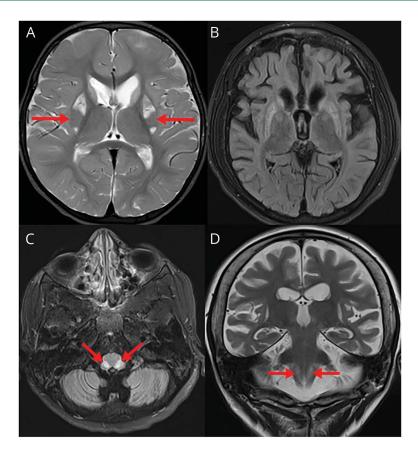
Bindu Parayil Sankaran, MD, DM, FRACP, PhD, Saskia B. Wortman, MD, PhD, Michel A. Willemsen, MD, PhD, and Shanti Balasubramaniam, FRACP

Neurology[®] 2022;98:e103-e104. doi:10.1212/WNL.00000000012614

Correspondence

Dr. Parayil Sankaran bindu.parayilsankaran@ health.nsw.gov.au

Figure Brain MRI



Brain MRI at age 2 years shows the classic "putaminal eye" sign (A, arrows). MRI at age 7 years shows progressive atrophy and gliosis of basal ganglia and cortical atrophy (B) and bilateral symmetrical signal changes of the nucleus tractus solitarius on fluid attenuated inversion recovery sequence (C, arrows) and T2-weighted images (D, arrows).

A 7-year-old girl with MEGD(H)EL [3-methylglutaconic aciduria, dystonia-deafness, (hepatopathy), encephalopathy, Leigh-like syndrome, SERAC1]¹ presented with worsening respiratory compromise. The evaluation showed type II respiratory failure (e.g., hypercapnic) necessitating mechanical ventilation. Cerebral MRI demonstrated progression of known changes in MEGD(H)EL (Figure, A and B) and symmetric nucleus tractus solitarius (NTS)

MORE ONLINE

Teaching slides http://links.lww.com/ WNL/B485

Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

Copyright © 2021 American Academy of Neurology. Unauthorized reproduction of this article is prohibited.

From the Genetic Metabolic Disorders Service (B.P.S., S.B.), Western Sydney Genetics Program, The Children's Hospital at Westmead, Sydney; The Children's Hospital at Westmead Clinical School (B.P.S.), Sydney Medical School, The Faculty of Medicine and Health, The University of Sydney, NSW, Australia; Department of Pediatric Neurology (S.B.W., M.A.W.), Amalia Children's Hospital, Radboud University Nijmegen Medical Center, Nijmegen, the Netherlands; Department of Pediatrics (S.B.W.), University Children's Hospital, Salzburg, Austria; and Discipline of Genetic Medicine (S.B.), Sydney Medical School, University of Sydney, NSW, Australia.

Appendix Authors

Name	Location	Contribution
Bindu Parayil Sankaran, MD, DM, FRACP, PhD	Children's Hospital at Westmead, NSW, Australia	Designed and conceptualized study, analyzed and interpreted the data, and drafted the article for intellectual content
Saskia B. Wortmann, MD, PhD	Amalia Children's Hospital, Nijmegen, the Netherlands and Department of Pediatrics, University Children's Hospital, Salzburg, Austria	Designed and conceptualized study, analyzed and interpreted the data, and drafted the article for intellectual content
Michel A. Willemsen, MD, PhD	Amalia Children's Hospital, Nijmegen, the Netherlands	Designed and conceptualized study, analyzed and interpreted the data, and drafted the article for intellectual content
Shanti Balasubramaniam, FRACP	Children's Hospital at Westmead, NSW, Australia	Designed and conceptualized study, analyzed and interpreted the data, and drafted the article for intellectual content

involvement (Figure, C and D). She was ventilator dependent and subsequently died from the effect of the disease.

Bilateral NTS involvement is a rare occurrence in a neurologic setting.² NTS plays a crucial role in the continuous modulation of chemoreceptor-mediated respiration and other respiratory reflexes.² This case illustrates the neuroimaging correlation of central neurogenic respiratory failure.

Study Funding

The authors report no targeted funding.

Disclosure

B. Parayil Sankaran reports receipt of the clinical research fellowship from Mito Foundation Australia. S.B. Wortmann, M.A. Willemsen, and S. Balalsubramaniam report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

References

- Wortmann SB, van Hasselt PM, Baric I, et al. Eyes on MEGDEL: distinctive basal ganglia involvement in dystonia deafness syndrome. *Neuropediatrics*. 2015;46(2): 98-103.
- Cutsforth-Gregory JK, Benarroch EE. Nucleus of the solitary tract, medullary reflexes, and clinical implications. *Neurology*. 2017;88(12):1187-1196.

Neurology®

Teaching NeuroImage: Bilateral Nucleus Tractus Solitarius Lesions in Neurogenic Respiratory Failure

Bindu Parayil Sankaran, Saskia B. Wortman, Michel A. Willemsen, et al. Neurology 2022;98;e103-e104 Published Online before print August 10, 2021 DOI 10.1212/WNL.00000000012614

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/98/1/e103.full	
References	This article cites 2 articles, 1 of which you can access for free at: http://n.neurology.org/content/98/1/e103.full#ref-list-1	
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Pediatric http://n.neurology.org/cgi/collection/all_pediatric Metabolic disease (inherited) http://n.neurology.org/cgi/collection/metabolic_disease_inherited Mitochondrial disorders http://n.neurology.org/cgi/collection/mitochondrial_disorders 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	

This information is current as of August 10, 2021

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 © 2021 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

