Neurology®

The most widely read and highly cited peer-reviewed neurology journal
The Official Journal of the American Academy of Neurology

Neurology Publish Ahead of Print
DOI: 10.1212/WNL.00000000000013020

Teaching Video NeuroImage: Aurora and Dusk of the Lentiform Fork Sign in a Patient With Reversible Dystonia

Author(s):

Bruna Kroeff, MD¹; Eduardo Hummelgen, MD¹; Giorgio Fabiani, MD^{1,2}; Ana Rosa Martins Cervellini, MD, MSc¹

Corresponding Author:

Giorgio Fabiani
giorgiofabiani@icloud.com

Neurology® Published Ahead of Print articles have been peer reviewed and accepted for publication. This manuscript will be published in its final form after copyediting, page composition, and review of proofs. Errors that could affect the content may be corrected during these processes.

Affiliation Information for All Authors: 1. Department of Neurology, Hospital Angelina Caron, CG do Sul, PR. ; 2. Department of Neurology, CHC-UFPR - Hospital de Clinicas da UFPR, Curitiba, PR.

Contributions:

Bruna Kroeff: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data

Eduardo Hummelgen: Drafting/revision of the manuscript for content, including medical writing for content; Analysis or interpretation of data

Giorgio Fabiani: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data

Ana Rosa Martins Cervellini: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data

Number of characters in title: 85

Abstract Word count: 100

Word count of main text: 100

References: 2

Figures: 1

Tables: 1

Supplemental: Patient consent form

Search Terms: [120] MRI, [147] Electrolyte, [162] Dystonia, [165] Parkinson's disease/Parkinsonism

Acknowledgements: Walter Oleschko Arruda, MD, PhD, for patient reading and last suggestions.

Study Funding: The authors report no targeted funding

Disclosures: The authors report no disclosures relevant to the manuscript.

The lentiform fork sign (LFS) is an uncommon imaging finding. Accepted hypotheses include vasogenic edema and metabolic acidosis affecting the lentiform nuclei^{1,2}. MRI imaging in a 46-year-old female undergoing regular hemodialysis showed three stages of the LFS over nine months. Her initial neurological examination revealed severe dystonia [Video 1]. Laboratory testing revealed mild compensated metabolic acidosis with nearly normal bicarbonate and anion gap [Table 1]. MRI sequence scans demonstrated [symmetric hyperintensities of the BG

and hyperintense rim delineating the lentiform] [Figure 1]. This case demonstrates that LFS can exist without metabolic acidosis. Vasogenic edema is considered a likely underlying mechanism.

Video 1-<http://links.lww.com/WNL/B646>

Teaching Slides-<http://links.lww.com/WNL/B647>

References:

- 1) Hamed S, Mohamed K, Abd Elhameed S, et al. Movement Disorders Due to Selective Basal Ganglia Lesions with Uremia. *Can J Neurol Sci.* 2020 May;47(3):350-365.
- 2) Fabiani G, Teive HA, Munhoz RP. Lentiform fork sign and fluctuating, reversible parkinsonism in a patient with uremic encephalopathy. *Mov Disord.* 2013 Jul;28(8):1053.

Video 1 legend: In segment 1 of the video, the patient showed severe dystonic movements with *geste antagoniste*, dysarthria, and was unable to remain seated. In Segment 2, the patient showed an improvement in the dystonic movements. The patient can now sit in an upright and comfortable position, and the *geste antagoniste* disappeared. There are mild to moderate signs of dystonia and bradykinesia, discretely worst on the left side.

Figure 1, Brain MRI and the three stages of LFS: 2020/02 - Brain MRI axial T2-FLAIR and STIR-FSE A.a, B.a, C.a no evidence of the LFS; 2020/08 - A.b, B.b, C.b bilateral, symmetric hyperintensities of the caudate, putamen, and thalamus, with the LFS, the hyperintense rim delineating the lentiform nucleus. (white arrowheads) and, 2020/10 - A.c, B.c, C.c show an impressive reduction of the bilateral hyperintensities, and the LFS just vanished (white arrowheads).

Laboratory	Timeline		
	2020/02	2020/08	2020/10
Glucose (mg/dL)	269	245	246
Sodium level (Na ⁺) (meq/L)	137,0	129,9	133,3
K ⁺ (meq/L)	3,7	6,67	4,91
ion Ca ⁺⁺ (mmol/L)	1,11	1,22	1,17
Bicarb level (HCO ₃) (meq/L)	20,4	19,9	24,7
Chloride level (Cl ⁻) (meq/L)	96	97	98
pH	7,35	7,398	7,45
pO ₂ (mmHg)	53,1	66,4	159,5
pCO ₂ (mmHg)	39.1	33	39,8
BE (meq/L)	-5,0	-4,30	-0,6
Anion gap (meq/L)	20,6	13	10,6
Lactate (meq/L)	1,2	5,95	2,65
Hgb (g/d)	9,7	9,6	11,6
Hematocrit (%)	31,5	28,0	34,0
Creatinine (mg/dL)	4,93	7,91	3,79
Urea (mg/dL)	66	159	30

1 **Table 1:** The central laboratory result

ACCEPTED

Neurology®

Teaching Video NeuroImage: Aurora and Dusk of the Lentiform Fork Sign in a Patient With Reversible Dystonia

Bruna Kroeff, Eduardo Hummelgen, Giorgio Fabiani, et al.

Neurology published online October 29, 2021

DOI 10.1212/WNL.00000000000013020

This information is current as of October 29, 2021

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/early/2021/10/28/WNL.00000000000013020.citation.full
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Dystonia http://n.neurology.org/cgi/collection/dystonia Electrolyte http://n.neurology.org/cgi/collection/electrolyte MRI http://n.neurology.org/cgi/collection/mri Parkinson's disease/Parkinsonism http://n.neurology.org/cgi/collection/parkinsons_disease_parkinsonism
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 © 2021 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.