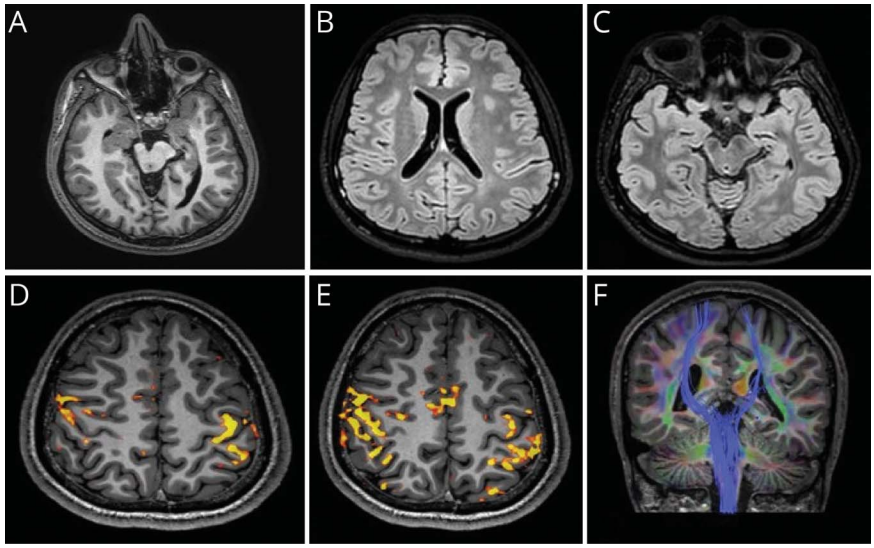
Teaching Video NeuroImage: Congenital Hemidystonia–Hemi-midbrain Atrophy Syndrome

Kanchana Soman Pillai, MD, DM, Mahendra Thakre, MD, DNB, Dnyaneshwar Asole, MD, DNB, Anil Venkitachalam, MD, DM, Uma Sundar, MD, Ashwini P. Sankhe, MD, Anagha R. Joshi, MD, and Vikrant P. Firke, MD

Correspondence
Dr. Pillai
kanchanapillai@gmail.com

Neurology® 2022;98:860-861. doi:10.1212/WNL.0000000000200513

Figure MRI of the Brain (T1 and T2 FLAIR), fMRI, and MR Tractography Findings



(A–C) Right-sided hemiatrophy of the midbrain with slight right hemispheric white matter volume loss. (D and E) Bilateral cortical activation on fMRI with right and left finger taps, respectively. (F) Magnetic resonance tractography shows normal decussation of corticospinal tracts.

An 18-year-old man with normal birth and development and a negative family history complained of nonprogressive clumsiness and posturing of his left hand since early childhood. His left limbs were shorter than his right but showed no evidence of atrophy (eFigure 1, A-C, links.lww.com/WNL/B902). Examination revealed dystonic posturing with mirror dystonia of the left hand and subtle mirror movements in his feet (Video 1). MRI of the brain showed right hemiatrophy of the midbrain, bilateral cortical activation with unilateral finger taps on fMRI, and normal corticospinal tract decussation (Figure, A-F). Levodopa response was present, suggesting dopaminergic denervation due to a congenital hemi-midbrain insult.[1] This case represents hemidystonia-hemiatrophy syndrome.[2] Subtle limb shortening and mirror dystonia can aid in diagnosis.

MORE ONLINE

Video

Teaching slides
links.lww.com/WNL/B902.

Study Funding
No targeted funding reported.

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

From the Department of Neurology (K.S.P., M.T., D.A., A.V., U.S.) and Department of Radiology (A.P.S., A.R.J., V.P.F.), Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai.

Go to Neurology.org/N for full disclosures.

Publication History

Received by *Neurology* October 5, 2021. Accepted in final form February 28, 2022. Submitted and externally peer reviewed. The handling editor was Whitley Aamodt, MD, MPH.

Appendix Authors

Name	Location	Contribution
Kanchana Soman Pillai, MD, DM	Department of Neurology, Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or intepretation of data
Mahendra Thakre, MD, DNB	Department of Neurology, Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; and analysis or intepretation of data
Dnyaneshwar Asole, MD, DNB	Department of Neurology, Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai	Drafting/revision of the manuscript for content, including medical writing for content, and analysis or intepretation of data
Anil Venkitachalam, MD, DM	Department of Neurology, Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai	Drafting/revision of the manuscript for content, including medical writing for content, and study concept or design
Uma Sundar, MD	Department of Neurology, Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai	Drafting/revision of the manuscript for content, including medical writing for content, and study concept or design
Ashwini P Sankhe, MD	Department of Radiology, Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai	Major role in the acquisition of data and analysis or intepretation of data
Anagha R Joshi, MD	Department of Radiology, Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai	Major role in the acquisition of data and analysis or intepretation of data
Vikrant P Firke, MD	Department of Radiology, Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai	Major role in the acquisition of data and analysis or intepretation of data

References

1. Vidailhet M, Dupel C, Lehéricy S, et al. Dopaminergic dysfunction in midbrain dystonia: anatomoclinical study using 3-dimensional magnetic resonance imaging and fluorodopa F 18 positron emission tomography. *Arch Neurol*. 1999; 56(8):982-989.
2. He C, Zhang P, Li Y, et al Hemiparkinsonism or hemidystonia with hemiatrophy Syndrome: a case series with follow-up. *Front Neurosci*. 2020;14:64.

Teaching Video NeuroImage: Congenital Hemidystonia–Hemi-midbrain Atrophy Syndrome

Kanchana Soman Pillai, Mahendra Thakre, Dnyaneshwar Asole, et al.

Neurology 2022;98;860-861 Published Online before print March 29, 2022
DOI 10.1212/WNL.0000000000200513