

Teaching Video NeuroImage: Chvostek sign with Fahr syndrome in a patient with hypoparathyroidism



Sunil K. Narayan, DM
P. Sivaprasad, MD
Ram Niranjan Sahoo,
MD
V. Bhuvaneshwari, FRCR

Address correspondence and reprint requests to Dr. Sunil K. Narayan, Professor and Head of the Department of Neurology, Jawaharlal Institute of Postgraduate Medical Education and Research, Pondicherry, India 605 006
sunil.narayan@jipmer.edu.in
sknarayan@gmail.com

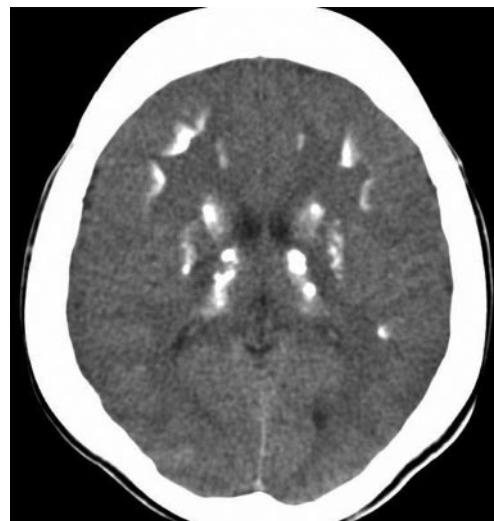
Figure 1 Chvostek sign: Repeated gentle taps in front of tragus over the superficial part of facial nerve elicits infatigable ipsilateral facial muscle contractions with each tap (see video)



A 17-year-old short-statured girl (figure 1) presented with seizures, progressive cataracts, and tetany. Further evaluation revealed positive Chvostek sign (video), cerebrostriothalamic calcifications or Fahr syndrome, persistent hypocalcemia (total serum calcium 6.5 [normal 9–10.5] mg/100 mL), and primary hypoparathyroidism (serum parathyroid hormone <3.0 [normal 12–72] pg/mL).

Chvostek sign, seen in hypocalcemia, hypomagnesemia, or alkalosis, is also reported in children with epilepsy.¹ Calcifications in Fahr syndrome (figure 2) are distinguishable from those of physiologic and

Figure 2 CT scan of the brain with plain axial cuts shows bilateral, symmetric, mirror-image cerebrostriothalamic calcifications (Fahr syndrome)



other causes, including hyperparathyroidism; phacomatoses; gliosis from CNS infections, trauma, or strokes; and neoplasms. Clinically, Fahr syndrome can be associated with rigidity, seizures, psychosis, and dementia. Short stature and primary hypoparathyroidism prompted investigations for a mitochondrial disorder,² but a muscle biopsy was normal.

REFERENCES

1. Ahmed MA, Martinez A, Mariam S, Whitehouse W. Chvostek's sign and hypocalcaemia in children with seizures. *Seizure* 2004;13:217–222.
2. Tengan CH, Kiyomoto BH, Rocha MS, Tavares VL, Gabbai AA, Moraes CT. Mitochondrial encephalomyopathy and hypoparathyroidism associated with a duplication and a deletion of mitochondrial deoxyribonucleic acid. *J Clin Endocrinol Metab* 1998;83:125–129. Review.

Supplemental data at
www.neurology.org

From the Department of Neurology (S.K.N., R.N.S.), Department of Internal Medicine (S.P.), and Department of Radiology (V.B.), Jawaharlal Institute of Postgraduate Medical Education and Research, Pondicherry, India.

Disclosure: The authors report no disclosures.

Neurology[®]

Teaching Video *NeuroImage*: Chvostek sign with Fahr syndrome in a patient with hypoparathyroidism

Sunil K. Narayan, P. Sivaprasad, Ram Niranjana Sahoo, et al.

Neurology 2008;71:e79

DOI 10.1212/01.wnl.0000336974.93499.ad

This information is current as of December 8, 2008

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/71/24/e79.full
Supplementary Material	Supplementary material can be found at: http://n.neurology.org/content/suppl/2008/12/07/71.24.e79.DC1
References	This article cites 2 articles, 0 of which you can access for free at: http://n.neurology.org/content/71/24/e79.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Neuromuscular Disease http://n.neurology.org/cgi/collection/all_neuromuscular_disease Clinical neurology examination http://n.neurology.org/cgi/collection/clinical_neurology_examination CT http://n.neurology.org/cgi/collection/ct Endocrine http://n.neurology.org/cgi/collection/endocrine Metabolic disease (inherited) http://n.neurology.org/cgi/collection/metabolic_disease_inherited
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 . All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

