Sweet food preference in amyotrophic lateral sclerosis

Martin R Turner, Kevin Talbot

An elderly female developed anarthria with prominent emotionality over an 18-month period before specialist neurological assessment. Although tongue electromyography was normal, her corticobulbar signs were consistent with amyotrophic lateral sclerosis (ALS), a pattern that, in the absence of functional impairment outside of speech and swallowing, is appropriately termed progressive bulbar palsy. Such patients, often elderly females, may remain ambulant and independent for many months, sometimes years, despite typically rapid anarthria. Electromyography may be insensitive to denervation, even when genioglossus is sampled, and this can contribute to diagnostic delay in patients with corticobulbar presentations of ALS who are frequently referred to ‘TIA’ or ENT clinics. When asked about her nutritional state, she revealed a collection of pictures of her favourite foods carried in her handbag to facilitate communication (figure 1).

ALS has pathological overlap with frontotemporal dementia through the common feature of cytoplasmic inclusions containing TDP-43. A hexanucleotide expansion in C9orf72 is associated with both ‘pure’ and mixed cases of ALS and frontotemporal dementia which may occur within the same pedigree. Overt dementia is not common in ALS (up to 15% in population-based studies), and is typically an early feature coincident with motor signs when it occurs. However, up to 50% of patients with ALS show a spectrum of more subtle cognitive and behavioural change, though most of these will not go on to develop dementia during the course of their disease. There have been criteria developed to reflect this broader phenotypic range of extramotor involvement in ALS.

An acquired preference for sweet foods, often with a narrowed repertoire, is included in the criteria for frontotemporal dementia. In ALS cases, it is a clue to frontotemporal involvement, and part

Figure 1 Packaging cut-outs carried by the patient to indicate her preferred food in the absence of speech. It was noted that all of them were confectionery items, consistent with observations that patients with amyotrophic lateral sclerosis may develop an exaggerated preference for sweet foods as a manifestation of frontal lobe pathology.
of an emerging array of metabolic disturbances common to both disorders.\textsuperscript{5} It should prompt more detailed neuropsychological assessment if there are wider concerns about behaviour or capacity.

**Contributors** MRT saw the patient, conceived and drafted the manuscript. KT saw the patient and edited the manuscript.

**Competing interests** None declared.

**Provenance and peer review** Not commissioned; externally peer reviewed. This paper was reviewed by Jonathan Rohrer, London, UK.

**Open Access** This is an Open Access article distributed in accordance with the terms of the Creative Commons Attribution (CC BY 4.0) license, which permits others to distribute, remix, adapt and build upon this work, for commercial use, provided the original work is properly cited. See: http://creativecommons.org/licenses/by/4.0/

© Article author(s) (or their employer(s) unless otherwise stated in the text of the article) 2017. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

**REFERENCES**


Sweet food preference in amyotrophic lateral sclerosis

Martin R Turner and Kevin Talbot

Pract Neurol 2017 17: 128-129 originally published online January 10, 2017
doi: 10.1136/practneurol-2016-001554

Updated information and services can be found at:
http://pn.bmj.com/content/17/2/128

These include:

References
This article cites 4 articles, 0 of which you can access for free at:
http://pn.bmj.com/content/17/2/128#BIBL

Open Access
This is an Open Access article distributed in accordance with the terms of the Creative Commons Attribution (CC BY 4.0) license, which permits others to distribute, remix, adapt and build upon this work, for commercial use, provided the original work is properly cited. See:
http://creativecommons.org/licenses/by/4.0/

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/