

- 2 **De Michele G**, Filla A, Cavalcanti F, *et al*. Late onset Friedreich's disease: clinical features and mapping of mutation to the FRDA locus. *J Neurol Neurosurg Psychiatry* 1994;**57**:977–9.
- 3 **Campuzano V**, Montermini L, Maltò MD, *et al*. Friedreich's ataxia: autosomal recessive disease caused by an intronic GAA triplet repeat expansion. *Science* 1996;**271**:1423–7.
- 4 **Cossee M**, Durr A, Schmitt M, *et al*. Friedreich's ataxia: point mutations and clinical presentation of compound heterozygotes. *Ann Neurol* 1999;**45**:200–6.
- 5 **Filla A**, De Michele G, Cavalcanti F, *et al*. The relationship between trinucleotide (GAA) repeat length and clinical features in Friedreich's ataxia. *Am J Hum Genet* 1996;**59**:554–60.
- 6 **Mateo I**, Llorca J, Volpini V, *et al*. GAA expansion size and age at onset of Friedreich's ataxia. *Neurology* 2003;**61**:274–5.
- 7 **Koutnikova H**, Campuzano V, Foury F, *et al*. Studies of human, mouse and yeast homologues indicate a mitochondrial function for frataxin. *Nat Genet* 1997;**16**:345–51.
- 8 **Bidichandani SI**, Ashizawa T, Patel PI. The GAA triplet repeat expansion in Friedreich ataxia interferes with transcription and may be associated with an unusual DNA structure. *Am J Hum Genet* 1998;**62**:111–21.
- 9 **Ohshima K**, Montermini L, Wells RD, *et al*. Inhibitory affects of expanded GAA TCC triplet repeats from intron 1 of the Friedreich ataxia gene on transcription and replication in vivo. *J Biol Chem* 1998;**273**:14588–95.
- 10 **Cossee M**, Campuzano V, Koutnikova H, *et al*. Frataxin fracos. *Nat Genet* 1997;**15**:337–8.
- 11 **Campuzano V**, Montermini L, Lutz Y, *et al*. Frataxin is reduced in Friedreich ataxia patients and is associated with mitochondrial membranes. *Hum Mol Genet* 1997;**6**:1771–80.
- 12 **Livak KJ**, Schmittgen TD. Analysis of relative gene expression data using real-time quantitative PCR and the 2(-Delta Delta C(T)) method. *Methods* 2001;**25**:402–8.
- 13 **Pfaffl MW**, Horgan GW, Dempfle L. Relative expression software tool (REST) for group-wise comparison and statistical analysis of relative expression results in real-time PCR. *Nucleic Acids Res* 2002;**30**:e36.

HISTORICAL NOTE

Lewis Carroll's Humpty Dumpty: an early report of prosopagnosia?

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Prosopagnosia is a rare form of visual agnosia characterised by impaired recognition of familiar faces (or equivalent stimuli). The term was coined by Bodamer in 1947, although the phenomenon had been described towards the end of the 19th century by Quaglino (1867), Hughlings Jackson (1872, 1876), and Charcot (1883).^{1,2} Brief accounts thought to be suggestive of prosopagnosia have been identified in writings from classical antiquity by Thucydides and Seneca.³ While there are dangers in this type of retrospective case identification, nonetheless I venture to suggest another early description of prosopagnosia.

The account is taken from *Through the looking-glass and what Alice found there* (1872) by Lewis Carroll (pseudonym of the Reverend Charles Lutwidge Dodgson). In chapter 6, Alice notices that the egg that she has just purchased

had eyes and a nose and mouth; and when she had come close to it, she saw clearly that it was HUMPTY DUMPTY himself. "It can't be anybody else!" she said to herself. "I'm as certain of it, as if his name were written all over his face."

Discussion follows, in which Humpty Dumpty, sitting precariously balanced upon a wall, gives his famous definition of the meaning of a word ("just what I choose it to mean") and coins the term "portmanteau word". As Alice takes her leave of Humpty Dumpty, the subject of facial recognition recurs, in the following exchange:

"Good-bye, till we meet again!" she said as cheerfully as she could.

"I shouldn't know you again if we *did* meet," Humpty Dumpty replied in a discontented tone, giving her one of his fingers to shake: "you're so exactly like other people." "The face is what one goes by, generally," Alice remarked in a thoughtful tone. "That's just what I complain of," said Humpty Dumpty. "Your face is the same as everybody else has—the two eyes, so—" (marking their places in the air with his thumb) "nose in the middle, mouth under. It's always the same. Now if you had the two eyes on the same side of the nose, for instance—or the mouth at the top—that would be some help." "It wouldn't look nice," Alice objected.

Humpty Dumpty reports an inability to recognise a familiar face, yet is able to recognise eyes, nose, and mouth and their correct positions, as is also the case with prosopagnosics. In developmental or congenital prosopagnosia, where the neuropsychological deficit is perhaps most pure because acquired cases following pathological insults such as cerebrovascular disease may not respect functional boundaries and may be accompanied by additional neurological signs such as visual field defects, there are impairments in face identity matching tasks but the ability to identify sex, age, emotional facial expression, and eye gaze direction is preserved.^{4,5} As in these cases, Humpty Dumpty's account seems to indicate preserved componential but impaired configural processing. There is also a suggestion that Humpty Dumpty might be able to use extraneous information to assist in facial recognition, his example being two eyes on one side of the nose or the mouth at the top of the face. Prosopagnosics may use extraneous visual cues such as spectacles, facial jewellery, and hair colour or style to aid facial recognition.^{3–5}

Whether Dodgson wrote this passage purely from imagination, or he based it upon observation of a prosopagnosic individual is not known. He did occasionally parody human idiosyncrasies, for example he himself appears as the Dodo because of his stammer ("Do-do-Dodgson") in *Alice's Adventures in Wonderland* (1865, chapters 2 and 3).

A J Larner

Cognitive Function Clinic, Walton Centre for Neurology and Neurosurgery, Lower Lane, Fazakerley, Liverpool, L9 7LJ, UK; a.larner@thewaltoncentre.nhs.uk

References

- 1 **Pryse-Phillips W**. Companion to clinical neurology. 2nd edn. Oxford: Oxford University Press, 2003:783.
- 2 **Della Sala S**, Young AW. Quaglino's 1867 case of prosopagnosia. *Cortex* 2003;**39**:533–40.
- 3 **De Haan EHF**. Covert recognition and anosognosia in prosopagnosic patients. In: Humphreys GW, ed. *Case studies in the neuropsychology of vision*. Hove: Psychology Press, 1999:161–80.
- 4 **Nunn JA**, Postma P, Pearson R. Developmental prosopagnosia: should it be taken at face value? *Neurocase* 2001;**7**:15–27.
- 5 **Larner AJ**, Downes JJ, Hanley JR, *et al*. Developmental prosopagnosia: a clinical and neuropsychological study [Abstract P591]. *J Neurol* 2003;**250**(suppl 2):II156.