

Some More Dickensian Diagnoses

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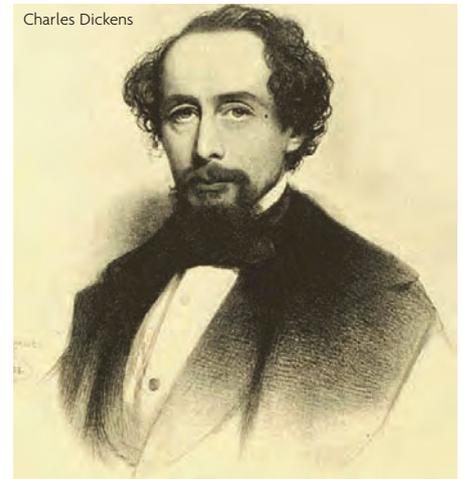
There is a tradition of distinguished clinical neurologists attempting to explain some of the vivid character descriptions in the works of Charles Dickens (1812-1870) in terms of neurological diagnoses: Lord Brain,¹ Macdonald Critchley,² David Perkin.³ Less distinguished figures have also dabbled in the field.⁴ Here we tentatively suggest some further possible Dickensian diagnoses, our justification for doing so being Critchley's statement that "Both as a stylist, and as a recorder of the *comédie humaine*, Charles Dickens is still insufficiently acclaimed".² To be sure, some of his insights have been acknowledged, for example this account from *David Copperfield* (1850) which may describe the phenomenon of *déjà vu*:^{5,6}

We have all some experience of a feeling which comes over us occasionally, of what we are saying and doing having been said or done before, in a remote time – of our having been surrounded dim ages ago, by the same faces, objects, and circumstances – of our knowing perfectly what will be said next, as if we suddenly remembered it.

Both Grandfather Smallweed (*Bleak House*, 1853), who needs to be carried everywhere, and Mrs Clennam (*Little Dorrit*, 1857), who is confined to a chair in her room, have been cited as examples of paraplegia by Lord Brain.¹ As regards the latter, there is an interesting description following her dramatic recovery of the ability to walk at the end of the novel (chapter 31):

There Mrs Clennam dropped upon the stones; and she never from that hour moved so much as a finger again, or had the power to speak one word. For upwards of three years she reclined in her wheeled chair, looking attentively at those about her, and appearing to understand what they said; but, the rigid silence she had so long held was evermore enforced upon her, and, except that she could move her eyes and faintly express a negative and affirmative with her head, she lived and died a stone.

This description may call to the neurologist's mind the locked-in syndrome (de-efferentation) following a ventral brainstem stroke, which typically leaves patients alert and able to perceive sensory stimuli, but unable to move other than some preservation of eyelid and sometimes ocular movements; the head movements apparent in Mrs Clennam would be unusual. Bauby, a famous sufferer of locked-in syndrome (and now the subject of a motion picture), cited a possible literary case of this condition in Alexandre Dumas's



Charles Dickens

novel *The Count of Monte Cristo* (1894),⁷ but not this possible, and prior, case report by Dickens.

The Smallweeds (*Bleak House*) are a peculiar clan. Mrs Smallweed, wife of Grandfather, is "weak in her intellect" and accordingly has been identified as suffering from dementia.¹ Macdonald Critchley cites two fragments of her speech as examples of senile verbigeration (inappropriate recurrent utterances, speech iteration).⁸ And what can one make of young Mr Bartholemew Smallweed, aka Small, or Chick Weed?

Whether Young Smallweed ... was ever a boy, is much doubted .. of small stature and weazen features ... he is a weird changeling, to whom years are nothing ... a kind of fossil Imp (Chapter 20)

There has been only one child in the Smallweed family for several generations. Little old men and women there have been, but no child ... until grandmother ... became weak in her intellect, and fell (for the first time) into a childish state (Chapter 21).

This early appearance of features typically associated with ageing in the apparent absence of cognitive impairment may suggest a diagnosis of progeria (Hutchinson-Gilford progeria syndrome, HGPS). Short stature, skin changes, facial features that resemble aged persons, but with normal cognitive development are typical of this syndrome. Classical HGPS follows an autosomal dominant pattern of inheritance, although almost all cases represent spontaneous mutations.⁹

Krook (*Bleak House*), brother of Mrs Smallweed, is famed for his untimely demise by means of spontaneous combustion, a storyline which involved Dickens in some controversy, not all his readers wishing to suspend disbelief. But a

possible pre-mortem diagnosis may be suggested by this description of Krook:

He was short, cadaverous, and withered, with his head sunk sideways between his shoulders, and the breath issuing in visible smoke from his mouth, as he were on fire within. His throat, chin and eyebrows were so frosted with white hairs, and so gnarled with veins and puckered skin, that he looked from his breast upwards, like some old root in a fall of snow.

Dilatation or engorgement of neck and face veins is one of the characteristic physical findings in superior vena cava obstruction.

Richard Carstone (*Bleak House*) rejects several career opportunities, including medicine, before becoming embroiled in the legal case of Jarndyce and Jarndyce, all the proceeds of which are consumed by lawyers' fees before judgement is finally given, during which time Carstone's health fails leading to his untimely death. Perhaps unwittingly, Dickens, familiar as he was with the workings of the law from his time at Doctors Commons as a young man, may be reporting here a case of "Chancery cachexia", described as such by the nineteenth century Irish physician Jonathan Osborne (Chancery was formerly one of the courts of justice, presided over by the Lord

This description may call to the neurologist's mind the locked-in syndrome

High Chancellor).¹⁰ In his paper on the subject,¹¹ Osborne described a clergyman whose fatal wasting was "occasioned by the delays and vexations in legal proceedings" in much the same way that Richard Carstone fades away having pursued his suit against the advice of John Jarndyce, and preyed upon by the venal lawyer Mr Vholes who makes much money at Carstone's expense.

Miss Havisham (*Great Expectations*, 1861) merits an eponymous syndrome in the writings of Macdonald Critchley,² as the prototype of comparable cases of young women, usually of aristocratic or well-to-do parentage, who suffer a major shock or rebuff and who then become reclusive, opt out of life, and try to make time stand still. Queen Victoria's behaviour after the death of Prince Albert is cited as a possible example. ♦

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