

Monks's Medical Condition: A Note

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CHARLES DICKENS took a keen interest in medicine and the medical profession and his contribution to the understanding of medicine has been increasingly recognised. Dickens had excellent powers of observation and description and he employed these skills to graphic effect when describing some unusual medical conditions in his novels. In *The Pickwick Papers* Dickens describes the Pickwickian syndrome¹ which today is known as obesity hypoventilation syndrome. In another example, in *Bleak House*, Dickens depicts the Smallweed family with their short stature, preserved intellect and early aging that might represent progeria.²

One of the most interesting cases in which Dickens seems to describe a previously unrecognised medical condition appears in *Oliver Twist*. Here it seems that Dickens pre-empted doctors in describing the main features of one particular medical condition: Sturge-Weber Syndrome.



Julian Rhind-Tutt as Edward Leeford, alias Monks: BBC TV adaption of *Oliver Twist* (2007).

In the 2007 BBC television production of *Oliver Twist*, Julian Rhind-Tutt, played the part of Edward Leeford alias Monks, with a port-wine stain on the left side of his face. This was plainly an interpretation by the programme's producers and make-up department of what the character might

have looked like. Although Dickens does not specifically refer to a port-wine stain in the text of *Oliver Twist*, the suggestion is made clear when Mr Brownlow tells Monks ‘...you, who from your cradle were gall and bitterness to your own father’s heart, and in whom all evil passions, vice, and profligacy, festered till they found a vent in a hideous disease which had made your face an index even to your mind – you, Edward Leeford, do you brave me still!’³ It is also clear that Monks suffers from epilepsy from Nancy’s description of him to Mr Brownlow: ‘His lips are often discoloured and disfigured with the marks of teeth, for he has desperate fits, and sometimes even bites his hands and covers them with wounds...’⁴

Joanne Eysell has argued that ‘given the prevalence of sexually transmitted diseases, especially various forms of syphilis, in the Victorian Age, it appears likely that Dickens was referring to syphilis when Mr Brownlow confronted Monks’.⁵ Another possible diagnosis for Leeford’s condition, perhaps, could be tuberose sclerosis, a rare multi-system genetic disease that causes non-malignant tumours to grow in the brain and other organs. Its symptoms may include seizures, behavioural problems and skin abnormalities. However, in 1998 a panel of international experts revised the diagnostic criteria of tuberous sclerosis complex to remove epilepsy and mental retardation from the indicators of this condition.⁶

A more likely diagnosis for Leeford’s condition, therefore, could be Sturge-Weber Syndrome, a rare congenital neurological and skin disorder. The condition is often associated with port-wine stains on the face, glaucoma, seizures, mental retardation, and ipsilateral leptomeningeal angioma (a tumour made up of blood vessels on the surface of the brain on the same side as the port-wine stain on the face). It is also characterized by abnormal blood vessels on the brain surface although normally only one side of the brain is affected.

A paper which first linked epilepsy to a lesion on the brain, entitled ‘A Case of Partial Epilepsy, apparently due to a Lesion of one of the Vaso-motor Centres of the Brain’ was presented to the Clinical Society of London in 1879 by William Allen Sturge, a physician at the Royal Free Hospital, London. The patient was a girl with no history of hereditary nervous diseases or ‘fits’ in the

family and born with a very extensive ‘mothers mark’ on the right side of the head and face. After her birth the child enjoyed good health until she was six months old, at which point she developed epilepsy. In his concluding remarks Sturge said: ‘I have no intention of going into the difficult question of the pathology of port-wine mark. The point to which I wish to call particular attention is the probable relationship between the mark and the fits.’⁷

The condition was explored further in 1922, when Frederick Parkes Weber, a dermatologist at Mount Vernon Hospital, London, and a personal physician to Queen Victoria, published a paper entitled ‘Right-Sided Hemi-Hypotrophy Resulting From Right-Sided Congenital Spastic Hemiplegia, With a Morbid Condition of the Left Side of the Brain, Revealed by Radiograms’. The patient in question was a 22-year-old woman with a right-sided congenital spastic hemiplegia, sexual infantilism and a very widespread vascular naevus, chiefly of the superficial ‘port-wine stain’ type. (‘Naevus’ refers to any localised area of pigmentation or collection of blood vessels of the skin which are usually benign and congenital). Weber concluded that: ‘It is highly probable that the congenital cerebral disease is in some way connected to the presence of a vascular naevus of the meninges of the brain on the left side – of the same nature as the extensive naevus of the patient’s body.’⁸ The condition was later named ‘Sturge-Weber disease’ by Brushfield and Wyatt.⁹

In *Oliver Twist*, Dickens may have made a connection between the propensity of the vascular lesion on Monks’s face to redden and blanch in accordance with his changing emotions and epilepsy in a way that Sturge had failed to do. Assuming the diagnosis of Sturge–Weber Syndrome is correct, Dickens therefore described the features of the condition in *Oliver Twist* in 1839 - a full 89 years before it was given its eponym in 1927.

¹ Burwell CS, Robin ED, Whaley RD, Bicklemann AG. Extreme obesity associated with alveolar hypoventilation; a Pickwickian syndrome. *American Journal of Medicine*. 1956; **21** 5: 811–8

² Singh V. Description of a family with progeria by Charles Dickens. *Neurology*. 2010; **75** 6: 571

³ *Oliver Twist*, London; Penguin, 2002, p.414

⁴ Ibid, p.387

⁵ Eysell J. *A Medical Companion to Dickens's Fiction*, Frankfurt, Peter Lang, 2005, p.89

⁶ Roach ES, Sparagana SP. *Journal of Child Neurology*. 2004; **19** 9: 643-649

⁷ Sturge WA. A case of partial epilepsy, apparently due to a lesion of one of the vasomotor centres of the brain. *Transactions of the Clinical Society of London*, 1879 **12**: 162-7

⁸ Weber FP. Right-sided hemi-hypertrophy resulting from right-sided congenital spastic hemiplegia, with a morbid condition of the left side of the brain, revealed by radiograms. *Journal of Neurology and Psychopathology*, London, 1922 **3**: 134-9.

⁹ Brushfield T, Wyatt W. Sturge-Weber disease. *British Journal of Children's Diseases*, 1927, **24**: 98-106; 25: 96-101

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