



Letterbox

The improbable 'Elephant Man'

Since the death of Joseph Merrick in 1890, many clinicians have sought to explain his pathology, yet the case remains unresolved.

The observations of Merrick's Physician and friend, Frederick Treves, prompted various retrospective diagnoses, the most enduring of which was Neurofibromatosis 1 (Nf1). Medical developments have led to further suggestions including Fibrous Dysplasia, Paget's Disease and most recently Proteus syndrome (PS).

Radiologist Amita Sharma (Royal London Hospital) has used CAT Scan and X-Ray to examine Merrick's skeleton. The results partially supported each theory, thereby precluding a single diagnosis. DNA-analysis of dental tissue by Geneticist Nalin Thacker (St Mary's Hospital, Manchester) also proved inconclusive due to insufficient organic material.

Such considerations have led other medical experts, such as Consultant Dermatologist John Harper (Great Ormond Street Hospital), to concede that the case is so singular as to perhaps warrant a separate diagnosis *i.e.* 'Merrick's Disease'.

PS is a rare congenital disorder (1/9 000 000 births). Symptoms include abnormally long digits, an enlarged skull, darkened skin and asymmetrical growth. Only 120 case histories are known (mainly from industrialised countries), some 50% of these indicate reduced cognition and/or longevity.

Nf1 is both a genetic and congenital disorder (1/2500 births). Initial diagnosis is often based upon the observation of six or more 'cafe-au-lait' skin discoloration, which occur to a lesser extent in some 10% of the US population. Most patients diagnosed and treated at an early stage have a relatively 'normal' prognosis, but some will require regular excision of bone and/or skin lesions.

Given the incidence of Nf1, it is feasible that Merrick had combined PS/Nf1 and that these disorders acted synergistically to produce the uniqueness of his pathology (1/22 500 000 000 births).

This hypothesis is theoretically testable through comparative anatomical surveys involving patients with PS, Nf1, combined PS/Nf1, and Merrick's skeleton. The reality,

however, is that PS/Nf1 probability is too low to permit the collection of a statistically viable data set.

Nevertheless, it is interesting to speculate that if Joseph Merrick did have a combined condition, then in accordance with UN median projections of population growth, the next case should present in approximately ten generations.

Further comments and references are available from the online letterbox.

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T. gondii – a reply

We were surprised that our article on *Toxoplasma* should be a cause for 'increasing concern'. While *Toxoplasma* can cause severe disease in 'at risk groups' (pregnant women and immunocompromised individuals), the vast majority of infections are benign.

Concerning the specific points raised in John Timmis's letter, the oocysts are not particularly 'sticky'. Cats are fastidious creatures who like to bury their faeces and they often defecate in loose soil (vegetable plots) or sand (child play areas). The oocysts sporulate and disperse through the soil or sand. This is why it is recommended that children's sandpits be covered when not in use. The oocysts are not specifically deposited on vegetation but could be splashed on to such by heavy rain along with soil particles. Therefore there is no danger from 'greens' as long as they are washed sufficiently to remove adhering soil. Very basic hygiene such as washing of hands and fresh vegetables before preparing food will remove the oocysts.

The cat can get a 'bad press' because of *Toxoplasma*. In Britain, between 19% and 46% of cats are infected with *Toxoplasma*. However, it is important to realise that these cats are not continually excreting oocysts; these are only produced for approximately one week a few days after initial infection. Thereafter the cat develops immunity and rarely produces oocysts again, even if reinfected. Recent research points to the tissue cysts in meat being a more important source of human infection. Once again, infection can be prevented by good kitchen hygiene and proper cooking of meat to kill

the tissue cysts. It would help if a vaccine for cats was available and certain companies are at present working on such a vaccine. However, this would not reduce the environmental contamination due to feral cats, nor would it reduce the risk associated with the tissue cyst.

Concerning the possible complications of the tissue cysts present in the brain: Again there is no well-characterised adverse consequence of chronic infection in humans with healthy immune systems. Although a few isolated reports do exist of *Toxoplasma* infection being associated with certain behavioural changes in humans, little hard evidence exists. Similarly there is no evidence of any involvement in the development of dementia or senility. It all comes down to relative risk. There is a high risk that you may be infected but a very low risk that any disease symptoms will result. As a chronically infected individual, who understands *Toxoplasma*, I am not losing any sleep over my infection.

DJP Ferguson
JM Wastling

Foot and Mouth Disease – the other factors

In his Foot and Mouth disease article in the April *Biologist*, Paul Barnett talks about 'stamping out'. This presumably refers to the whole grisly and contentious business of slaughtering and then, somehow, disposing of the dead carcasses. It is the waste disposal factor that I feel raises grave biological issues that should not be left out of any discussion on this disease.

Health concerns have led to carcass burning being stopped or not undertaken in certain areas, including my home town. In this country we have banned farmers from burning straw and instituted strict controls on the incineration of waste and yet, when it officially suits us, we seem happy to burn large quantities of dead livestock on crude open funeral pyres. The sight of smoke pouring from these fires sends a strange signal from a country reputedly an enthusiast for a cleaner environment.

The alternative to burning is to bury dead carcasses. The environmental problems presented by this approach have been well documented in the press and do not need to be repeated here. This whole issue of environmentally and biologically acceptable waste disposal needs to assume a much higher profile in the factors for consideration in controlling this disease. I would have thought that the waste disposal industry could put some figures on the amount of dead carcasses that can reasonably be handled within current rules and guidelines. It would then be necessary to develop a more sophisticated and progressive National Foot and Mouth policy, probably including vaccination and revised compensation arrangements, to fit in with the waste disposal possibilities.

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