book reviews

Parkinson's disease in the older patient, 2nd edition

Edited by Jeremy Playfer and John Hindle. Radcliffe Publishing, Abingdon 2008. 428 pp. £65.00

Thank goodness for fields such as gerontology which so enhance the single organ-based specialties and gene-centric approaches. Such cross-sectional disciplines improve all round management of individuals, as exemplified in this book, and ensure that we keep up to speed on life history, ecological and evolutionary concepts of disease.

This is important as the last 50-year track record of finding causes and cures of diseases of ageing, particularly in the face of rapid unexplained increases in longevity in rich nations, is not (lets be honest) that impressive considering the enormous effort largely from a laboratory-based research industry. Laboratory studies typically control the environment, unlike the real world, and that is an uncertain approach particularly given that genes have been known to be largely regulated by diet and stress signals since the birth of molecular biology and the discovery of genetic switches and the lac operon.[1] Degenerative diseases that were solved earlier, such as B12 or B3 deficiency, affect more than one organ and involve both the supply of nutrients and personal genetic make-up. Neuroscientists working on sub-acute combined degeneration of the cord or pellagrous dementia armed with a modern brain and DNA bank but with minimal ecological data might well have been tempted down blind mechanistic alleys and struggled to figure out the root cause, yet this is now the central strategy for solving diseases of ageing.

In this edition the chapter authors ably inform us that we know a great deal about the interdisciplinary management of the ageing patient with Parkinson's disease and how it should be implemented often by breaking down artificial professional ties. Good chapters span conventional drug and surgical approaches and non-motor complications. The effect on quality of life gets ample room as do rightful concerns over nutrition, the carers, rehabilitation and palliation along with organisation of services and the vital role of nurse practitioners. Energetic management encouraging a 'use it or lose it' approach is implicit.

Tantalising hints as to the aetiology of Parkinson's disease are capably documented. I liked the book's cover picturing a forest showing light as the source of fluctuant energy and information forecasts from circadian rhythms illuminating both wood and trees. Is the human super-organism so different or just more complex with different sources of energy, more compartments and a greater panoply of homeostatic responses?

Medicine and education of the population in 50 years will surely be highly developed. Our long-lived, better developed and disease-free descendants may have to thank the theoreticians of ageing and development for seeing the common bioenergentic threads and available trade-offs between health and disease.

Reference

1 Ptashne M, Gann A. *Genes and signals.* New York: Cold Spring Harbor Laboratory Press, 2002.

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Royal maladies: inherited diseases in the ruling house of Europe

By Alan R Rushton. Trafford Publishing, Victoria, Canada 2008. 261 pp. £10.30

This book discusses two familial diseases affecting the royal houses of Europe: haemophilia and variegate porphyria. On haemophilia (16 pages) the author is on reasonably firm ground. He examines the effects of haemophilia on the royal houses in Russia and Spain with devastating political consequences for both. Nevertheless it would be useful, for the medical and non-medical readership, to have more detail about the process of the blood clotting and, in particular, the nature of the clotting-defect in the families (haemophilia A or B). A new mutation in Queen Victoria's elderly father Edward, Duke of Kent (aged 50 when she was conceived) is postulated quite reasonably as the source of the problem. More discussion on paternal age effects, mechanism of X chromosome inactivation and the molecular genetics of haemophilia would increase the value of this aspect of the book.

The major part of Rushton's study (84 pages) is, however, devoted to variegate porphyria. Here the author is on shaky ground. There are serious errors both of omission and comission. Thus he attempts to outline the pathway of haem biosynthesis, vital to the understanding of the nature, severity and cause of the acute porphyrias. In 1980 Battersby and colleagues clarified the steps between porphobilinogen and the uroporphyrins. These steps are not shown and thus the reader is confused on the nature of the defect in acute intermittent porphyria and its key relationship to the symptomatology of variegate porphyria.

Rushton bases his diagnosis of porphyria on the work of the psychiatrists and amateur historians, mother and son duo, Ida Macalpine and Richard Hunter, who categorically stated that King George III was not 'mad' at all but suffered from the undiagnosed medical condition acute intermittent porphyria. In a second paper they altered the diagnosis, again categorically, to the milder condition variegate porphyria on the history of skin lesions in the King (adolescent acne, an instance of sunburn and a possible episode of urticaria).

In spite of authoritative and well argued doubts expressed at the time by experts in the field, this dubious claim has entered the canon of historical fact and is a widely-accepted interpretation that features in academic and popular history books, television documentaries and notably Alan Bennett's award-winning

play and film. Only recently has a detailed re-evaluation of the King's medical records shown that the claims of Macalpine and Hunter and their adherents are seriously flawed and they were guilty of being 'economical with the truth'.[1]

Rushton has not taken on board these serious doubts and has based his book on a diagnosis of variegate porphyria in the King. He has wandered over the European relatives of George III and, using any symptoms of sudden or early death, an episode of diarrhoea or constipation, 'spasms', mental health issues and any skin problems as diagnostic of variegate porphyria in the individual. On this basis he has 'traced' George's defect backwards to William the Conqueror and his antecedents, and forward to Victoria's great, great grandson William, Duke of Kent. Rushton has undertaken a large amount of work: a morning in the British Library would have exposed the fallacy of the porphyria hypothesis.

Rushton also claims to have identified the molecular defect in George III in an American family some 21 ill-defined generations distant from the King! This defect in living patients is quite distinct from that claimed in a possible DNA sample from Queen Victoria's granddaughter, Princess Fedora, recently analysed by John Rohl and his colleagues and thus further confounds the issue.[2]

This book provides some interesting 'what if' speculations about these diseases and their political consequences but as a serious authoritative work leaves much to be desired.

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References

1 Peters TJ, Wilkinson D. King George III and porphyria: a clinical re-examination of the historical evidence. *History of Psychiatry* 2009.
2 Rohl JCG, Warren M, Hunt D. *Purple secret. Genes, 'madness' and the royal houses of Europe.* Bantam Press: London, 1998:192–207.