Hugh Downman and smallpox inoculation

Professor Dunn quotes Downman’s approval of Lady Mary Wortley’s in his fascinating account of the Exeter physician. Her contemporaries, however, were often less generous. This beautiful and literary lady contracted smallpox in 1715 and probably knew of the Turkish practice of "engrafting" or "variolation" against the disease from her own doctors. As Fellows of the Royal Society they may well have heard an account of it passed on from Timonius of Constantinople. The following year she had the opportunity of travelling to Turkey with her husband who had been appointed ambassador to the Ottoman Empire. Receptive towards Islamic culture she was struck by the relative absence of smallpox and learned that this was attributed to the deliberate infecting of subjects with material from smallpox victims. In March 1718 she summoned the nurse who was Constantinople’s “general surgeon” for inoculation. The nurse pricked the wrist of Lady Mary’s young son with a needle, laid a tiny droplet of smallpox matter on the skin and mixed it with a drop of blood from the puncture. Some eight days later he became febrile and developed about 100 spots on his body. These quickly resolved without leaving scars.

Subsequently, the chequered success of variolation in the hands of English physicians, careless of the finer details of Turkish practice exemplified by Lady Mary, contributed to lifelong controversy. Most cruelly, her former friend Alexander Pope implied in one of his satires that she left people “pox’d by double entendre as well as an attack on the safety of variolation.

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The use of sodium res囟ion in pseudohypoaldosteronism

We describe the use of sodium resonium in a patient with pseudohypoaldosteronism (PHA). PHA is a rare genetic abnormality characterised by raised plasma aldosterone, but mineralocorticoid resistance causing hyperkalaemia and hypotenaemia. Severe recessive type 1 PHA is due to defective epithelial amiloride sensitive sodium channels (ENaC).

Our patient presented aged 14 days with hyponatraemia (130 mmol/l) and hyperkalaemia (9.4 mmol/l). He made no response to hydrocortisone or fludrocortisone. His plasma aldosterone level during crisis was extremely high (3820 pmol/l), confirming a diagnosis of PHA. People’s sibling died neonatally with a presumptive diagnosis of PHA, suggesting autosomal recessive inheritance compatible with an ENaC defect.

Our patient was managed on intermittent rectal calcium res囟ion when hyperkalaemia, and daily solution G (a preparation containing high levels of sodium (13.3 mmol/ml)). The sodium requirement was 45 mmol/kg/day. Due to its unpalatability, solution G was given via gastrostomy. Despite this he had episodes of sudden collapse, precipitated by minor infections, with hyponatraemia and life threatening hyperkalaemia, including a cardiac arrest. Discharge proved impossible.

After 18 months we changed his treatment to sodium res囟ion 0.25 g/kg twice daily via gastrostomy on advice from Professor Dillon (Great Ormond Street Hospital). Our patient
improved and the electrolytes became tightly controlled (see fig 1). Our patient was so much better that after two years inpatient stay, we managed to discharge him home on treatment. The sodium requirement is 19 mmol/kg/day, and he has had no further electrolyte decompensation.

We have treated six patients with recessive PHA in the past 10 years, previously treating them with calcium resonium rectally at times of hyperkalaemic crisis. This treatment has not controlled electrolytes, and two of our patients died. Our experience with recessive PHA is that the defect does not improve with age; one of our patients died at the age of 21 years, and the survivors remain on vast daily intakes of sodium. Sodium resonium has improved our current patient’s quality of life after two years inpatient care. Our experience with recessive PHA in PHA was first described in 1984, it is not widespread. This case should prompt greater use of sodium resonium in PHA.

We must acknowledge the huge number of nurses and doctors that contributed to a successful outcome in this case.

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References


Once upon a time …

I very much enjoyed reading Storr and Once upon a time …

It’s bigger, but is it better?

For me and my colleagues the answer is certainly, “Yes”.

Our Child and Adolescent Mental Health Service (CAMHS) now has all four editions of Child and Adolescent Psychiatry, “the child psychiatrist’s bible”, from the first edition, 1997 (Oxford, J Porter, M Kershaw, J Kirk, N Trevelyan, NJ Sh); to the second edition, 1995 (phone directory size); the paperback third edition, 1995 (Yellow Pages size, plus); and now this, the fourth edition, 2002, which is almost Data Sheet supplement size. These awesome dimensions and the task of reviewing it (“you’ll have to read it, you fool” said a colleague) was somewhat off putting, but now that we’ve opened it and used it we do not want to be without it. This book is the child and adolescent psychiatry textbook and a must have for any district hospital or postgraduate library. The sort of emotional behavioural and developmental problems encountered by paediatricians in hospital and in the community, in the interface between paediatrics and psychiatry, are covered in sufficient detail to be of real use to the clinician, whether he is going it alone or has the luxury of cross referral. The authors are experts in their fields, mainly eminent child and adolescent psychiatrists or psychologists from both sides of the Atlantic, but there are contributions from paediatricians in the chapter on soiling, for example.

Perhaps there is too much detail for the exam driven paediatrician in training, but then some only seem to grasp the value of developing some psychological frameworks later in their careers. Neither is this a quick fix alternative to the psychological component of the DCH and here again I suspect the size factor is likely to intimidate. Even SHOs in adult psychiatry who are now required by the Royal College to spend 10% of their time in child and adolescent psychiatry may be deterred and reach for a more MCQ orientated text in order to pass their exams.

For a hard pressed CAMHS team this book provides both the academic substrate and the clinical detail to be of real value in many everyday clinical scenarios. The reference lists are excellent and up to date and the index works, so that we have already checked out and accessed clinical information and papers we have been meaning to chase but never got round to. The new and esoteric stuff seems to be there as well as the genuine advances we have heard about on CPD days, such as the guidelines for assessment and treatment of childhood depression, clinically useful rating scales, and developments in the management of ADHD and autistic spectrum disorders, using an evidence based approach wherever feasible. What I have read and found fascinating and so it seems that, in this electronic age, there is still a place for a comprehensive reference book on the department shelf.

Much of all this is new and just not in the 1995 edition, which is why the book is so big and why your library should buy it.

M A Griffiths