

# PostScript

## LETTERS

### Informed choice: why measuring behaviour is important

Olusanya *et al* debate the principles of informed choice within the context of infant hearing screening.<sup>1</sup> In doing so they draw on our conceptualisation and measure of informed choice. Unfortunately they draw an erroneous conclusion, namely that it is inappropriate to measure uptake as part of assessing informed choice. This is based on a misinterpretation of both our definition of informed choice and its measurement.

Based on the decision making literature,<sup>2</sup> we have proposed an operational definition of informed choice: "one that is based on relevant knowledge, consistent with the decision maker's values and behaviourally implemented".<sup>3</sup> There are two types of informed choice: an informed choice to decline screening, where someone with good knowledge and negative attitudes towards themselves undergoing screening does not undergo screening; and an informed choice to accept screening where someone with good knowledge and positive attitudes towards themselves undergoing screening, undergoes screening. An assessment of informed choice therefore requires an assessment of knowledge, attitudes, and the consistency between attitudes and screening behaviour, to determine whether screening behaviour, usually referred to as uptake, reflects the attitudes of the person offered screening. This definition and model places no value on whether the choice made is to accept or to decline screening: both such choices can be informed and therefore represent a positive outcome of screening.

Olusanya *et al* have also misinterpreted the policy of informed choice in the context of screening.<sup>4</sup> The goal of an informed choice strategy for screening is not for everyone to have positive attitudes towards undergoing the procedure, but rather that people act consistently with their own values, not those of others, including healthcare professionals. There are some situations where there is one clear choice and healthcare professionals recommend a course of action; for example, the need for an emergency laparotomy or the need to reduce a diuretic dose in someone with a low serum potassium.<sup>5</sup> Undergoing a screening test does not fall into this category.

We hope this serves to clarify the misunderstanding that Olusanya and colleagues appear to be labouring under, with regard to both the concept of informed choice and its operationalisation.

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Competing interests: none declared

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### Authors' reply

#### Is negative parental attitude towards infant hearing screening justifiable?

Marteau *et al* took exception to the following phrase in our paper:

"Our model differs from the three-dimensional typology proposed by Marteau *et al*, which incorporated uptake as a measure of informed choice. In our view, uptake represents a consequence rather than the goal of informed choice and was therefore excluded as a measure."

and went on to raise the following issues which we wish to address in this reply:

- That we drew on their conceptualisation and measure of informed choice in a way that misrepresented their definition and measure of informed choice
- That the definition and the measure of informed choice must include a measure of uptake or "behaviour"
- That the goal of informed choice is not for everyone to have positive attitudes towards undergoing the procedure, but rather that people act consistently with their own values (*whatever they are*).

Firstly, our two-dimensional (knowledge and attitude) model was adapted from our previous work on the social change that underpins public health interventions.<sup>1</sup> It preceded the three dimensional (knowledge, attitude, and uptake/behaviour) model by Marteau and colleagues<sup>2</sup> and was first presented at an international conference in Manchester in 1999.<sup>3</sup> It was conceived from an earlier work on the management of corporate change by Professor Paul Strebler of the International Institute for Management Development (IMD) in Switzerland as was acknowledged in the earlier report.<sup>1</sup> We have simply contextualised that earlier model for infant hearing screening in this paper and highlighted the difference with a generic model.<sup>2</sup> The authors' subtle claim to originality is therefore presumptuous.

The last two points on the definition, goal, and measure of informed choice are inter-

related. The word "choice" in the context of our paper is defined as "the act of choosing" (*Webster Collegiate Dictionary*) rather than the actual choice that is made among available options. The expectation in any screening programme is that there is high uptake and that this is based on informed consent. But this must not be confused with the goal of our paper, which was to examine the route to informed choice/decision making. The General Medical Council (GMC) for instance stipulates that healthcare workers "must take appropriate steps to find out what patients want to know and ought to know about their condition and its treatment".<sup>4</sup> To suggest that parents should be allowed to act consistently with their values (*whatever they are*) shows a lack of understanding of the challenge of offering public health intervention, particularly where parental doubts exist.<sup>5-8</sup> It also overlooks cases where parental perception towards non-life threatening conditions such as infant hearing loss may be nonchalant.<sup>1</sup> The readiness or willingness to accept screening is reflected in parental attitude (positive or negative) towards screening. It is immoral to ignore a negative attitude towards a public health intervention that is in the patient's best interest, especially when it emanates from personal or cultural values, or even unfavourable past experience.<sup>9</sup> Moreover, an "assessment of the consistency between attitudes and screening behaviour" is an academic exercise that is irrelevant for our purpose. A recent article perhaps sums up the principle underlying our model: a doctor believes in facts, but a manager believes in perceptions.<sup>10</sup>

In summary, our model is a simple and practical tool that is intended as a guide for healthcare workers to facilitate a positive attitude towards infant hearing screening in cross-cultural settings without attempting to coerce or frighten parents into giving consent. From this point, it is entirely the parents' responsibility to give or withhold consent and to accept the consequences of that decision. Sadly, the authors failed to comprehend this crucial context, which perhaps explains their inability to relate their commentary to child health interventions or specifically to infant hearing screening. However, we are pleased to observe a common ground on this subject—that knowledge and attitude are key determinants of parental decision making (and perhaps also, readiness) for infant hearing screening.

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Competing interests: none declared

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### Home management versus inpatient care of adolescents with newly diagnosed type 1 diabetes mellitus: survey of current practice

I read with interest the article written by Lowes and Gregory<sup>1</sup> on the above subject. In 2003, I carried out a postal survey in order to document the current practice of paediatricians when managing adolescents newly diagnosed with type 1 diabetes mellitus. A total of 117 consultant paediatricians, mainly members of the British Society of Paediatric Endocrinology and Diabetes, were asked to complete a questionnaire indicating their current practice. Statistical analysis was made by MINITAB;  $\chi^2$  and trend  $\chi^2$  were used to examine the factors that affected the management option chosen by the paediatricians.

Sixty eight per cent (80/117) replied. When faced with a well 12 year old child newly diagnosed with diabetes mellitus, the majority (47.5%) will offer a short admission (<2 days) followed by home based management and outpatient reviews, while 32.5% will offer complete home based management with no hospital admission. A minority (20%) did not offer any home based management at all. There was no significant difference between paediatricians that worked in a tertiary hospital and those that worked in a district general hospital in the choice of management ( $p=0.4$ ). In addition, the ratio of diabetes nurse specialists to number of patients in the clinic made no difference to choice ( $p=0.09$ ). However, using trend  $\chi^2$ , we found that units that had two or more diabetes nurse specialists were significantly more likely to offer home based care ( $p=0.007$ ).

Since the earlier survey in 1988,<sup>2</sup> it appears that more UK paediatricians are offering home based management of newly diagnosed children with diabetes mellitus. In this study, the majority are choosing to offer this after a short hospital admission.

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Competing interests: none declared

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### Cystic fibrosis is no longer an important cause of childhood death in the UK

We have previously reported the survival of the UK cystic fibrosis (CF) population.<sup>1</sup>

Funding for active surveillance ceased in 1997, leaving incomplete ascertainment for the post-1993 cohorts and thus preventing accurate survival calculations for these cohorts. However, as the number of CF births in the UK is reasonably constant, being related to the total birth rate, a knowledge of CF deaths by age can give an insight into the survival of young children.

Death certification data for the UK were obtained for 1994 to end 2003 (ICD-9 codes 2270, 7770, and 7484, and ICD-10 codes E84.0, E84.1, E84.8, and E84.9). Non-nationals and obvious miscodings were removed. Table 1 presents deaths by year of birth and age.

The 1994 data are consistent with a 97% survival to age 10 (304/311) based on live births (750 000) and CF incidence (1 in 2416). Deaths in the first year of life average three (29/10) per cohort, while subsequently there is about one death every three completed years (12/34).

It would be difficult to assert that these figures could be bettered without the most detailed investigation of the circumstances surrounding each death.

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Competing interests: none declared

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## BOOK REVIEWS

### ADHD: the facts

Edited by Mark Selikowitz. Oxford: Oxford University Press, 2004, pp 235, £11.99. ISBN 0 19 852628 8



This book opens badly, with two long histories that, for some reason, are presented in tiny font size. Already irritated I was then dismayed to find the cases described maintaining the tired stereotypes of the dreamy inattentive *girl* and the hyperactive, impulsive *boy*. The boy

is, disappointingly, also violent and aggressive. There are girls who are hyperactive and impulsive, and hyperactive, impulsive children of both sexes who are neither violent nor aggressive. Unfortunately the media stereotype, reinforced here, is not a helpful one for most children with ADHD trying to make sense of themselves.

The relation between real and administrative prevalence, or the political and social factors which can influence both, are not discussed. ADHD is, at the severe end of the scale, a disabling disorder with clear neurobiological deficits. However it is also a dimensional disorder with no boundary between “normal” and “ADHD”; at the cusp it becomes, in part, a socially constructed disorder. This has such profound implications for the appropriateness of how we treat and teach all children that it should at least have had some mention.

The book attempts, with considerable success, to explain simply the neurodevelopmental basis of ADHD. But there is confusion between the neuropsychological deficits found in ADHD and those found in other disorders with which it may be occasionally co-morbid. For example, the book fails to explain the role of attentional difficulties in the aetiology of social clumsiness found in ADHD, which is quite different in character to the primary socialisation difficulties of autism, arising as they do from deficits in communication, empathy, and theory of mind. To add to this, Asperger's syndrome

**Table 1** Deaths in the UK cystic fibrosis population by year of birth and age

Year of birth	Age at death (y)									
	0-<1	1-<2	2-<3	3-<4	4-<5	5-<6	6-<7	7-<8	8-<9	9-<10
1994	4	1	0	1	0	0	0	0	1	0*
1995	3	1	1	0	0	0	0	1	0*	
1996	1	0	1	0	1	0	1	1*		
1997	4	1	0	0	0	0	0*			
1998	3	1	0	0	0	0*				
1999	6	0	1	0	0*					
2000	1	0	0	1*						
2001	3	0	0*							
2002	2	0*								
2003	2*									

\*Denotes partially observed years.

is described within a section on emotional disorders characteristic in ADHD, which is highly misleading. Indeed, throughout the book core and non-core symptomatology are muddled. The introductory section makes no distinction between the core symptoms of inattention, hyperactivity, and impulsiveness, and co-morbidity such as clumsiness and inflexibility—core for developmental coordination disorder and ASD respectively, but not for ADHD. In section 2, core deficits are again described with the same emphasis as, for example, defiance and emotional disorders. I think this can only confuse readers as to the true nature of ADHD.

Subsequent chapters, on diagnosis, treatment, school and home management, and adult ADHD, provide a useful and well balanced source of information, although recent developments result in the comments on SSRIs being now out of date.

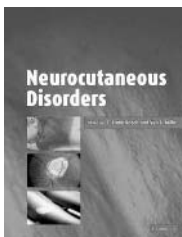
The author appears to explain *all* behaviours in children with ADHD using exclusively neuropsychological, rather than behavioural, theory with no discussion of the impact of a child's developmental context on how both personality and disorder are manifest. For example, chapter 7 ("Low self esteem") discusses this phenomenon as having an entirely neuropsychological basis arising from faulty self appraisal mechanisms (although these theoretically could provide one with a super-ego instead). Only in chapter 9, in an aside, is the impact of negative experiences, of which these children have plenty, acknowledged in the development of poor self esteem. The quality of attachment, maternal depression, exposure to violence are all known to have a profound impact on how children develop, whether or not they have neurodevelopmental problems. It may be that children with ADHD are even more susceptible to the impact of early adverse experiences, but nowhere in this book is this explored. This is, presumably, an understandable but over-reaction to the still pervasive tendency to blame ADHD on poor parenting. But we need a more balanced view than that provided here.

The blurb on the back claims this book to be useful to parents and professionals alike. Unfortunately the tabloid approach and lack of referenced primary sources really preclude its relevance to a professional audience. Although it does provide a wealth of useful information for a lay audience, it would have been much improved if there was at least some indication in the text of when a statement made is well supported by good quality research, for example, the high heritability of ADHD, from another which sets down a "fact" with, as far as I can ascertain, no research base at all. For example, page 47: "adolescent boys with ADHD are particularly averse to taking instruction from a woman teacher". With some reservations this book will be useful for parents and other carers wanting a fairly detailed text they can dip into as the need arises but it is not one I could recommend to colleagues.

E V J Webb

## Neurocutaneous disorders

Edited by E Steve Roach, Van S Miller. Cambridge: Cambridge University Press, 2004, pp 325, £120.00 (hardback). ISBN 0 52 78153 1



Most doctors either love or hate "the neurocutaneous disorders". These words can provoke flashbacks to examination agony, or to diagnostic triumph. They plunge some into uncertainty, and transport others to scholarly heights.

But whatever your experience, this book is for you, or at least for your library. The editors, American neurologists, amply achieve their aim "to provide readily accessible information about the clinical features and natural history of these rare conditions as well as an understanding of their genetic basis and molecular mechanisms".

Introductory chapters providing useful lists and background genetics are followed by comprehensive coverage of individual disorders. Some are well known—for example, neurofibromatosis, tuberous sclerosis, and Sturge-Weber syndrome. Rarer classics that roll off the dermatological tongue find their place here: ataxia telangiectasia, incontinentia pigmenti, xeroderma pigmentosum. Inclusion of conditions that uncharacteristically affect the nervous system does not detract: any disorder of blood vessels can affect the brain—for example, Ehlers-Danlos syndrome, blue rubber bleb nevus syndrome, and hereditary haemorrhagic telangiectasia. In some cases the neurological component is limited to the eyes (pseudoxanthoma elasticum) or peripheral nerve (macroductily-nerve fibrolipoma).

The material is well organised and well edited, with each chapter following the same plan: clinical manifestations (dermatological, neurological, radiological, etc), followed by genetic basis and management issues. Gorlin or Gorlin's syndrome is, as ever, a delight. Well chosen references and accurate indexing combine to make the book both authoritative and user friendly. Most genetics books are out of date as soon as they are published, but every relevant new gene has been included in this one. The sparse, mostly black and white illustrations are at first sight a disappointment. However, the pictures are well chosen to illustrate relevant points: restrained compared with dermatological atlases, but entirely adequate.

Errors and omissions are few and relatively unimportant. Epidermal naevus due to keratin 1 or 10 mutation is not (as stated on page 20) a neurocutaneous disorder, because keratins are not expressed in the nervous system. Linear and whorled hypermelanosis, contrary to the statement in the section on incontinentia pigmenti, is sometimes associated with neurological abnormalities. Perhaps those skin disorders associated with deafness should be included as neurocutaneous disorders, such as KID syndrome (keratitis/ichthyosis/deafness) and deafness with palmoplantar keratoderma due to connexin mutations. Two steroid sulphatase deficiency disorders with developmental delay are omitted, namely X-linked ichthyosis with contiguous deletion of a mental retardation gene, and multiple sulphatase deficiency.

This book radiates enthusiasm and excitement. These are the disorders where clinical diagnostic skills really matter. The accessibility of the skin helps us to understand the inaccessible nervous system. The future is bright, the future is genetic. For most of these conditions the molecular basis has been

elucidated in the past 10 years, leading not only to more accurate diagnosis, and more informed genetic counselling, but also to a greater understanding, and the hope of effective treatment for these once incurable disorders. You may still be unable to list four cutaneous markers of tuberous sclerosis, but at least you know where to look them up.

C Moss

## Paediatric and adolescent gynaecology: a multidisciplinary approach

Edited by Adam H Balen, Sarah M Creighton, Melanie C Davies, Jane MacDougall, Richard Stanhope. Cambridge: Cambridge University Press, 2004, pp 540, £150.00. ISBN 0 521 80961 4



The management of young and adolescent girls with disorders of the genital tract and associated illnesses has long fallen between many subspecialties. With few adolescent gynaecologists in this country, paediatric surgeons

and physicians, adult gynaecologists, and endocrinologists work with (or not as the case may be) clinical geneticists and psychologists to diagnose, explain, and manage sometimes simple and sometimes extremely complex problems. Pre-adolescent and adolescent gynaecology has always been on the margins of most professionals' knowledge and practice and there is a dearth of supporting reference texts. The range of specialties involved may have made the area unappealing for reference publication except for a small chapter in a text largely on something else. Thus, textbooks in this area are scarce and modern thought even more so. This book has thus found a good potential niche for publication.

Naturally, however, in trying to draw together expert authors from multiple disciplines there are areas of overlap and gap, perhaps more so here than is common even in this type of text. A more consistent structured chapter template, and for those readers with an image based memory, a greater number of diagrams and images, would have been helpful in almost every chapter. To please a target readership from paediatric surgeons and paediatricians to adult gynaecologists and psychologists will always be difficult and I suspect that all groups will be frustrated at some level. I think this may be inevitable and should not detract from the value of the book as a modern and broad resource.

The first part covers normal development well and is a good source of reference. Finding a pattern for the bulk of the book was clearly difficult and like many reviewers I would have organised it differently. While much of what one needs is present I found the layout confusing, perhaps due to the theme of an integrated approach. I would expect those general paediatricians and surgeons treating simple gynaecological problems to appreciate those chapters based on presentations such as pelvic pain, genital dermatology, and vaginal discharge and amenorrhoea being drawn

together, and an adjacent separate section to those chapters on ovarian disorders and fertility together, etc while devoting a section to all the aspects of intersex. Grouping the chapters concerning intersex, which although always interesting, is a further subspecialty practiced now by progressively fewer doctors, would allow a progressive development of the diagnosis, management, and outcomes as viewed by each discipline.

One of the practical issues that many doctors dealing with children and adolescents, particularly in the area of gynaecology and fertility face relates to the medico-legal aspects of "consent", especially to that pertaining to genetic material, and should there be a second edition I would request more information than that presented within the chapter on "Preservation of fertility before cancer therapy".

In general, however, the book is written and edited by experts and provides a broad resource of information, for both the simple and complex problems which may be encountered in paediatric gynaecology. Its integrated multidisciplinary approach does make it an essential reference text for all doctors and psychologists involved in the care of children and young adults with complex gynaecological conditions and I am pleased to have it on my shelf to partially replace a loose leaf file stuffed with old and new articles gleaned from a variety of publications of variable quality and reliability.

R J Hitchcock

### Immunological disorders in infants & children, 5th edition

Edited by E Richard Stiehm, Hans D Ochs, Jerry A Winkelstein. Philadelphia: Elsevier Saunders, 2004, pp 1458, £174.00 (hardcover). ISBN 0 7216 8964 7



The latest edition of this definitive text is certainly not intended for cover-to-cover reading, but aims to contain "everything you always wanted to know (and probably much more) about paediatric immuno-

logical disorders". It contains a vast amount of information written by many of the leading names in the field, and undoubtedly succeeds in its aim as far as any textbook covering a rapidly evolving field can hope to do.

The first edition was published in 1973, and this fifth edition has been published just one year after the death of Robert Good, author of the first chapter of the first edition, "Crucial experiments of nature that have guided analysis of the immunologic apparatus". These "experiments of nature", as well as a great deal of basic science, have since then continued to provide insights into the immense complexity of the immune system. The advances in understanding of both basic and clinical immunology even since publication of the last edition in 1996 have been extensive, and this is reflected in the addition of much new material. The book is, as previously, intended for a wide readership, including paediatric sub-specialists, general paediatricians, paediatric trainees, and medical students. It will also be of considerable interest to adult immunologists. It is divided into four sections: Development and Function of the Immune System, Primary Immunodeficiencies, Secondary Immunodeficiencies, and Immunologic Aspects of Paediatric Illness.

The section on ontogeny and fundamental immunology may appear relevant only to sub-specialists. However, increasing recognition of the role of various forms of immunological dysregulation in the pathogenesis of a wide variety of disorders in many paediatric disciplines means that these chapters are a valuable reference resource. New chapters in the first section include one on innate immunity, which contains much recent information about mechanisms of natural immunity, such as the mannose binding lectin system, Toll-like receptors, defensins, and the roles of the major cytokines. Also new is a chapter on the immunology of pregnancy.

Detailed discussions of over 100 defined primary immunodeficiencies in Section II are preceded by a useful overview, including discussion of clinical immunology investigations. The huge expansion in understanding of the basic mechanisms underlying these disorders is reflected in the increase from one to four chapters on combined and isolated T cell deficiencies. Many recently identified

molecular defects are discussed. Likewise the chapter on primary antibody deficiency includes descriptions of more molecularly defined disorders, including the four defined forms of hyper-IgM syndrome. The X linked form (CD40 ligand deficiency) arguably belongs with T cell disorders given that many of its manifestations reflect the basic T cell defect rather than antibody deficiency, but this is a minor point. A completely new chapter for this edition focuses on disorders of apoptosis, which manifest as autoimmune lymphoproliferative syndrome.

The third and fourth sections of the book are likely to be of most general interest. An extensive section on the immunodeficiency of immaturity may be of particular interest to neonatologists. Discussions of every aspect of neonatal immunology are each followed by a succinct summary for those without the time or the inclination to struggle with basic immunology. A wide variety of genetic syndromes with immunodeficiency as a component are described in another new chapter, and there are expanded discussions of immunodeficiency associated with malnutrition, splenic deficiency, and paediatric HIV infection, as well as immunological aspects of surgical and anaesthetic stress. The final section covers immunological aspects of many paediatric disorders affecting virtually every system, including descriptions of the periodic fever syndromes, the molecular basis for several of which have recently been elucidated, infection in immunocompromised children, immunisation, and comprehensive coverage of both solid organ and bone marrow transplantation in children.

This is an impressive reference text that provides an appealing balance between specialised descriptions of complex immunology and rare primary immunodeficiencies, and the roles played by immune mechanisms in a wide variety of both rare and relatively frequent paediatric disorders. Although it is inevitably already out of date—for example, in the areas of most recent antifungals and monoclonal antibody therapies—this does not detract from its value. It will undoubtedly retain the position of the major comprehensive reference work in paediatric immunology.

A Jones